

Nervous System

CHAPTER
6

A. CASE SHEET FORMAT

HISTORY TAKING

Name:

Age:

Sex:

Residence:

Occupation:

Chief complaints:

1. _____ × days
2. _____ × days
3. _____ × days

History of presenting illness:

HIGHER MENTAL FUNCTION

Altered state of consciousness:

- Onset
- Any seizures and blackouts
- Any fall/injuries
- Any ear or nose bleed
- Fever
- Any ear pain or discharge
- Drug history
- Any addictions.

Mental state and cognition:

- Changes in the memory
- State of alertness and drowsiness
- Changes in the mood and affect (loss of spontaneity)
- Language changes
- Loss of spatial orientation
- Diminished ability to carry out routine activities of daily living.

Other higher mental functions:

- Speech difficulty
- Difficulty to recognize people or objects
- Inappropriate crying or laughter
- Lack of interest
- Social disinhibition
- Delusions/hallucinations.

CRANIAL NERVE DYSFUNCTION

Ask about:

- Loss of vision, smell, and taste
- Alteration in facial feeling
- Double vision/visual symptoms
- Problems with swallowing and chewing
- Speech alterations
- Vertigo/hearing abnormalities
- Hoarseness of voice, dysphagia, nasal regurgitation, and nasal intonation of speech
- Pain/difficulty in neck movements.

Example

Left lower motor neuron (LMN) 7th nerve palsy: History of retroauricular pain followed by abrupt onset deviation of angle of mouth to right with slurring of speech and difficulty in left eye closure with history of hyperacusis.

MOTOR DYSFUNCTION

Weakness

Distribution of weakness:

- Is it symmetrical/asymmetric:
- Paresis or plegia:
- Limbs involved:
- Ipsilateral or contralateral:
- Patterned weakness.

Example

Right middle cerebral artery (MCA) territory embolic infarct: History of sudden onset, complete loss of power in left upper limb and lower limb. Weakness maximum at onset and nonprogressive.

Onset and progression:

- Acute, subacute, or chronic

Progression of the weakness:

- Ascending weakness or descending weakness
- Ellsberg phenomenon
- Variation throughout the day
- Muscles/limb(s) involved.

Proximal upper limb—shoulder/arm:	Difficulties in combing hair, reaching for high objects, winging of scapula
Distal upper limb—forearm/hand:	Finger/wrist drop, poor hand grip, cannot open jar, difficulty in buttoning/unbuttoning
Proximal lower limb—pelvic/thigh:	Cannot rise from chair or squatting position, waddling gait
Distal upper limbs—leg/foot:	Difficulty in gripping chappals, cannot walk on heels/toes foot drop
Neck muscles	Dropped head/broken neck

Trunk

Inability to roll on the bed

Example

Guillain–Barré syndrome (GBS): History of preceding gastrointestinal (GI) infection followed by acute onset difficulty in getting up from squatting position, difficulty walking, progressing to involve upper limbs (difficulty combing hair), and neck muscle weakness. No sensory symptoms.

Wasting/Loss of Muscle Bulk

- Wasting—present/absent
- Fasciculations—present/absent

Stiffness of Limbs

- Stiffness—present/absent
- Heaviness—present/absent

Gait Abnormalities

- Limp or dragging foot
- Scissoring/circumduction.

Involuntary Movements

- Type
- Symmetrical/asymmetrical
- Part of the body involved
- Present at rest
- Functional disability.

SENSORY DYSFUNCTION

- Numbness/loss of feeling
- Altered feeling:
 - Paresthesia
 - Dysesthesias (tingling and pin-needles)
 - Spontaneous pain
- Pattern of sensory loss.

CEREBELLAR HISTORY

- Swaying to one side
- Tremors while reaching objects
- Lack of coordination of activities
- Overshooting acts
- Abnormal involuntary eye movements (oscillopsia/nystagmus).

HISTORY SUGGESTING MENINGITIS/RAISED INTRACRANIAL PRESSURE

- Headache
- Neck pain
- Projectile vomiting
- Blurring of vision
- Seizures
- Photophobia.

HISTORY SUGGESTING AUTONOMIC DYSFUNCTION

- Dryness of skin
- Palpitations
- Perspiration
- Syncopal attacks/postural giddiness
- Bladder dysfunction:
 - Urinary retention
 - Loss of awareness of bladder control
 - Frequency, urgency
 - Urge/overflow incontinence.

REVIEW OF COMMON NEUROLOGICAL SYMPTOMS

Headaches

- Onset and duration of headache
- Location of headache, unilateral versus bilateral
- Severity
- Frequency
- Radiation
- Quality of headache (dull and diffuse)
- Types:
 - a. Continuous
 - b. Pulsating
 - c. Stabbing
 - d. Sharp
 - e. Throbbing
 - f. Dull
 - g. Thunderclap
- Alleviating factors
- Triggers for the headache/aggravating factors
- Temporal association (headache not worse in mornings)
- Association with nausea/vomiting/tearing of eyes/redness of eyes
- Vision changes before or during headache
- Precipitating factors:
 - Stress
 - Menses
 - Allergens

- Sleep deprivation
- Coughing
- Straining
- Bending forwards
- Associated motor/sensory symptoms: Weakness, numbness, and tingling in upper or lower extremities
- Photophobia/phonophobia
- Systemic symptoms—weight loss, low energy, and anorexia
- Fever and neck stiffness
- History of head trauma
- History of migraine
- Family history of migraines
- Effect on daily activities
- Use of oral contraceptive pills
- Caffeine intake
- Smoking and alcohol history.

Example

Classical migraine: Visual aura followed by insidious onset, unilateral, severe pulsating type of headache lasting for >4 hours associated with nausea and photophobia. Repeated such attacks every month with history of some identifiable precipitating factors and a positive family history of migraine.

Seizures

- Onset and duration
- Frequency
- Factors which precipitate these episodes
- Injury sustained as a result of the seizure
- Postictal symptoms: Confusion
- Associated sensory deficits
- Associated motor deficits
- Associated cognitive deficits
- Muscle spasms
- Anatomical progression of motor involvement (e.g. Jacksonian March)
- Symptoms suggesting aura
- Associated incontinence
- Tongue biting and salivation
- Automatisms associated with these episodes
- History of head trauma
- Perinatal infection
- Drug history
- History of seizure disorder
- Family history of seizure disorders
- Effect on daily activities.

Example

Generalized tonic clonic seizure (GTCS): Abrupt onset tonic clonic contraction of muscle associated with tongue bite and urinary incontinence. Patients generally regain consciousness within few minutes with postictal confusion and headache.

Past history:

- Asthma
- Chronic obstructive airway disease
- Tuberculosis
- History of contact with tuberculosis
- Diabetes mellitus (DM)
- Hypertension (HTN)
- Ischemic heart disease (IHD)
- Seizure disorder and drugs used (in detail).

Family history:

(draw pedigree chart representing three generations)

Personal history:

- Bowel habits
- Bladder habits
- Appetite
- Loss of weight
- Occupational exposure
- Sleep
- Dietary habits and taboo
- Food allergies
- Smoking (in smoking Index or Pack years)
- Alcohol history (___ grams of alcohol/day or ___ units of alcohol/week).

Menstrual and obstetric history:

- G___P___L___A___
- Age of menarche ___
- Menopause at ___
- Flow—amenorrhea/oligorrhea/menorrhagia.

Summarize:

Differential diagnosis:

- 1.
- 2.
- 3.

GENERAL EXAMINATION

Patient

- Conscious
- Cooperative
- Obeying commands

Body Mass Index (BMI)

- Wt (kg)/Ht² (meters)
- Grading according to WHO for Southeast Asian countries

Vitals

- **Pulse**
 - Rate
 - Rhythm
 - Volume
 - Character
 - Vessel wall thickening
 - Radio-radial delay and radio-femoral delay
 - Peripheral pulses
- **Carotid and vertebral bruit**
- **Blood pressure**
 - Right arm
 - Left arm
 - Leg—right/left
- **Respiratory rate**
 - Regular
 - Abdominothoracic (male) or thoracoabdominal (female)
 - Usage of accessory muscles
- **Jugular venous pulse**
 - Waveform
- **Jugular venous pressure**
 - ____ cm of blood above sternal angle (+ 5 cm water)

On Physical Examination

- Pallor
- Icterus
- Cyanosis
- Clubbing
- Lymphadenopathy
- Edema

Others Head to Toe

- Nerve thickening
- Neurocutaneous markers
- External markers of atherosclerosis
- Signs of nutritional deficiency, alcoholism, etc.
- Any other general examination finding

NERVOUS SYSTEM EXAMINATION

- Right/left handed person
- Education

HIGHER MENTAL FUNCTIONS

- Consciousness—if impaired document using Glasgow coma scale
- Orientation to time/place/person
- Memory:
 - Immediate (repetition—30 seconds)
 - Recent (up to 5 minutes—recall)
 - Remote (> 5 minutes)
- Intelligence
- Mood/emotion
- Concentration and calculation (subtract seven from 100)
- Speech:
 - Spontaneous speech—comprehension
 - Fluency
 - Repetition
 - Reading
 - Writing
 - Naming objects
 - Phonation
 - Aphasia
 - Dysarthria
- Apraxias—present/absent
- Hemineglect—present/absent
- Hallucinations and delusions—present/absent

Cranial nerves	R	L
Olfactory—I nerve: Sense of smell (peppermint, soap, coffee, lemon peel or vanilla) *Both eyes shut, one nostril checked at a time Appreciate smell ± identify it		
Optic—II nerve: Visual acuity (perception of light/hand movements and finger counting/Snellen's chart at 6 meters/Jaeger's chart at 14 inches) Visual field (confrontation method/menace reflex)—mention defects, if any Color vision (Ishihara's test) Fundus		
Oculomotor, trochlear, abducens—III, IV, VI nerves: Eyelids (any ptosis) Position of eyeballs at rest (any deviation, exophthalmos, enophthalmos) Extraocular movements: <ul style="list-style-type: none">I. Binocular movements<ul style="list-style-type: none">— Saccadic:— Pursuit:— Reflex (doll's eye, caloric stimulation)II. Unicocular movements (#Comment on ophthalmoplegia, if present—supranuclear, internuclear, individual nerves, or muscles) Pupil <ul style="list-style-type: none">• Size (in mm)• Shape		

<ul style="list-style-type: none"> • Reaction • Direct light reflex • Consensual light reflex • Accommodation reflex <p>Nystagmus (Describe whether spontaneous or provoked/type—horizontal, vertical, rotatory, pendular)</p>	
<p>Trigeminal nerve—V nerve:</p> <ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – Touch – Pain – Temperature <p>(To be checked on all three divisions around the jawline, on the cheek, and on the forehead)</p> • Motor: <ul style="list-style-type: none"> – Jaw deviation – Hollowing above and below zygoma – Clenching teeth (feel temporalis and masseter) – Open mouth against resistance – Side to side movement of jaw (pterygoid) • Reflexes: <ul style="list-style-type: none"> – Corneal—present/absent (superficial reflex, 5th nerve afferent, 7th nerve efferent) – Jaw jerk—present/absent/exaggerated (deep reflex, afferent and efferent, both 5th nerve, center mid-pons) 	
<p>Facial nerve—VII nerve: Facial asymmetry (look for absence of wrinkling, drooping of corner of mouth, obliteration of nasolabial fold, widened palpebral fissures)</p> <ul style="list-style-type: none"> • Motor: <ul style="list-style-type: none"> – Frontalis (raise the eyebrows) – Orbicularis oculi (shut the eyes tight) – Buccinator (show teeth, smile, blow check, whistle) – Orbicularis oris (close lips, pronounce labials “p”, “b”, “m”) – Platysma (pull down the corners of mouth) <p>(## Look for Bell's phenomenon)</p> • Sensory: <ul style="list-style-type: none"> – Anterior 2/3rd tongue taste (sugar, lime, salt, quinine) <p>Lacration Hyperacusis—present/absent Emotional fibers checking—emotions preserved or not</p>	
<p>Vestibulocochlear nerve—VIII nerve: The ability to hear the sound produced by rubbing the thumb and forefinger together is then tested for each ear at distances up to a few centimeters</p> <ul style="list-style-type: none"> • Rinne's test—air conduction/bone conduction (AC/BC) • Weber's test—lateralized/centralized • Caloric test [Irrigates one external auditory canal with cool (about 30°C) or warm (40°C) water. Normally, cool water in one ear produces nystagmus on the opposite side. Warm water produces it on the same side] 	
<p>Glossopharyngeal, vagus IX, X nerve: Note the patient's ability to drink water and eat solid food and also see the character, volume and sound of the patient's voice.</p> <ul style="list-style-type: none"> • Position of uvula • Movement of uvula on saying “ah”—any deviation • Gag reflex—present/absent/exaggerated (taste over the posterior third of the tongue and can be tested) 	
<p>Spinal accessory—XI nerve:</p> <ul style="list-style-type: none"> • Sternocleidomastoid (instruct the patient to rotate head against resistance applied to the side of the chin to tests the function of the opposite sternocleidomastoid muscle. To test both sternocleidomastoid muscles together, the patient flexes the head forward against resistance placed under the chin) 	

<ul style="list-style-type: none"> Trapezius (shrugging a shoulder against resistance) <p>Hypoglossal nerve—XII:</p> <p>Inspection (inside the mouth):</p> <ul style="list-style-type: none"> Size of tongue Symmetry/any wasting Fasciculation (on protrusion) Deviation—side Tremors <p>Palpation:</p> <ul style="list-style-type: none"> Tone Power Speech 		
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MOTOR SYSTEM

Attitude

- Upper limb
- Lower limb

Bulk

Inspection: Symmetry, generalized wasting comment on small muscle wasting, deformities, claw hand, foot drop, if any.

Measurement in cm	<i>R</i>	<i>L</i>
Arm (10 cm above olecranon)		
Forearm (10 cm below olecranon)		
Thigh (18 cm above the superior border of patella)		
Leg (10 cm below the tibial tuberosity)		

Note: Bilateral similar distance from fixed bony points till the maximum bulk of muscle.

Tone

	<i>R</i>	<i>L</i>
Upper limb		
Lower limb		

Note: Comment whether normal, hypotonia or hypertonia (spasticity/rigidity).

Power

Checked both isometric (resistance against movement) and isotonic (resistance at end of movement).

0	Complete paralysis
1	A flicker of contraction only
2	Power detectable only when gravity is excluded by postural adjustment
3	Limb can be held against gravity but not resistance
4	Limb can be held against gravity and some resistance

5 Normal power

Muscle	R	L
Neck <ul style="list-style-type: none"> Flexors (SCM, platysma, scalene, suprathyoid, infrahyoid, longus colli and capitis, rectus capitis) Extensors (trapezius and paravertebral muscles—splenius, erector spinae, transversospinalis, interspinal intertransverse) <p><i>Note:</i> Avoid active movement checking if cervical cord injury suspected</p>		
Shoulder <ul style="list-style-type: none"> Abduction (0–15°—supraspinatus, 15–90°—middle fibers of deltoid, above 90°—trapezius and serratus anterior) Adduction (pectoralis major, latissimus dorsi and teres major) Flexion (biceps brachii (both heads), pectoralis major, anterior deltoid, and coracobrachialis) Extension (posterior deltoid, latissimus dorsi, and teres major) 		
Elbow <ul style="list-style-type: none"> Flexion (biceps brachii) Extension (triceps brachii) 		
Wrist <ul style="list-style-type: none"> Flexion (FCR, FCU) Extension (ECRL, ECRB, ECU) 		
Hand grip (long flexors)		
Small muscles of hand		
Trunk (rectus abdominis, transversus abdominis, oblique, pyramidalis) <ul style="list-style-type: none"> Elevation of head or leg in supine position Beevor's sign if present Abdominal binding to check for intercostal muscle weakness Intercostal binding to check for diaphragmatic weakness 		
Hip <ul style="list-style-type: none"> Flexion (iliopsoas) Extension (gluteus maximus) Abduction (gluteus medius and minimus, tensor fascia lata) Adduction (adductor longus, brevis, and magnus) 		
Knee <ul style="list-style-type: none"> Flexion (hamstrings) Extension (quadriceps) 		
Ankle <ul style="list-style-type: none"> Plantar flexion (gastrocnemius, soleus) Dorsiflexion (tibialis anterior) 		
Small muscles of foot , EHL if needed		

REFLEXES

Superficial reflexes	R	L
Corneal (cranial nerve V and VII)		
Abdominal: <ul style="list-style-type: none"> Epigastric (T6–T9) Mid-abdominal (T9–T11) 		

• Hypogastric (T11–L1)		
Cremasteric (L1, L2)		
Anal reflex (S2, S3)		
Plantar:		
<ul style="list-style-type: none"> Reflexogenic zone—S1 Afferent nerve—tibial nerve SC segments—L4, L5, S1, S2 		
Chaddock's (lateral aspect of foot from below up), Gordon's (calf), Oppenheim's (anterior tibia), Schaffer's (Achilles tendon), Gonda's (press down 4th toe), Stransky's (adduct little toe), Bing's (pinprick on dorsolateral foot)		

Deep tendon reflexes	R	L
Jaw jerk (afferent and efferent both 5th nerve and center mid pons)		
Biceps (C5, C6)		
Brachioradial/supinator/radial periosteal (C5, C6)		
Triceps (C6, C7, C8)		
Knee jerk/quadriceps/patellar reflex (L2, L3, L4)		
Ankle jerk (L5, S1, S2)		
Clonus—present/absent		
<ul style="list-style-type: none"> Patellar Ankle 		
Latent reflexes (suggest pyramidal lesion if present unilaterally)		
Tromner's/finger flexor reflex/Hoffmann's sign		
Wartenberg's sign		
By convention the deep tendon reflexes are graded as follows:		
<ul style="list-style-type: none"> 0 = no response; always abnormal 1+ = a slight but definitely present response; may or may not be normal 2+ = a brisk response; normal 3+ = a very brisk response; may or may not be normal 4+ = a tap elicits a repeating reflex (clonus); always abnormal 		
Please do reinforcement maneuvers before saying DTR's are absent		
Primitive reflexes		
<ul style="list-style-type: none"> Glabellar tap Palmomenatal (both sides) Sucking Rooting Pout and snout Grasp 		

Involuntary movements (describe in detail)

Coordination (described later under cerebellum)

SENSORY SYSTEM

Primary sensation	R	L
Touch		

Pain		
Temperature		
Vibration		
Joint position sense		
Any sensory level		
Pattern of sensory loss (graded/dissociative/crossed/hemi)		

Cortical sensation (to be tested only in the presence of primary sensation intact)	R	L
Tactile localization (topognosis)		
Two point discrimination		
Stereognosis		
Graphesthesia (figure identification)		
Sensory extinction		

Romberg's test:

CEREBELLAR SIGNS

<i>Upper extremity</i>	R	L
Limb ataxia: • Outstretched arm test • Finger nose test • Nose-finger-nose test • Finger-finger test		
Rapid alternating movements: • Rapid hand tapping • Pronation-supination • Thigh slapping		
Pointing and past pointing		
Writing (macrographia)		
Rebound phenomenon (arm)		
Tremors (intention)		

<i>Lower limbs</i>	R	L
Heel knee test		
Pendular knee jerk		
Finger toe test		
Rapid alternating movements—foot tapping		

<i>General</i>
Titubation
Nystagmus

Tremors
Hypotonia
Truncal ataxia
Tandem walking
Gait

GAIT

- Base—wide or narrow
- Slow/rapid
- Falling to sides
- Look which part of foot touches ground first (toe/heel)
- How high foot lifted above ground?
- Hand swing
- Turning around
- Position of hip, sound produced while foot touches ground.

Signs of Involvement of Autonomic Nervous System

- Dryness of skin/excessive sweating/spoon test
- Postural hypotension
- Heart rate—baseline, on respiration, on standing
- Palpable bladder
- Pupillary reactions
- Valsalva maneuver.

Signs of Meningeal Irritation

- Neck stiffness
- Kernig's sign
- Brudzinski's sign—neck, leg, and pubis.

Skull and Spine

- Deformities
- Tenderness
- Short neck.

SOFT NEUROLOGICAL SIGNS

- **Pyramidal drift** describes a tendency for the hand to move upward and supinate if the hands are held outstretched in a pronated position (palms downward), or to pronate downward if the hands are held in supination.
- **Cerebellar drift** is generally upward with excessive rebound movements if the hand is suddenly displaced downward by the examiner.
- **Parietal drift** is an outward movement on displacing the ulnar border of the supinated hand.

OTHER SYSTEMS

Respiratory system:

- Inspection:
- Palpation:
- Percussion:
- Auscultation:

Cardiovascular system:

- Inspection:
- Palpation:
- Percussion:
- Auscultation:

Gastrointestinal system:

- Inspection:
- Palpation:
- Percussion:
- Auscultation:

B. DIAGNOSIS FORMAT

GENERAL FORMAT

Nature of Disease

- Onset: Sudden/acute/subacute/chronic (sudden—vascular, acute—demyelinating, subacute— infections/space occupying lesions, chronic—degenerative)
- Deficit: Monoplegia/hemiplegia/quadruplegia/paraplegia/nerve palsies/ataxia/sensory disturbance/movement disorders.

Site of Involvement of Nervous System

- Upper motor neuron disease—intracranial (brain or cerebellum) or extracranial (spinal cord)
- Lower motor neuron disease—anterior horn cell disease, radiculopathies, neuropathies, neuromuscular junction diseases, and myopathies.

FOR CEREBROVASCULAR ACCIDENT

Sudden onset, right-sided dense hemiplegia with right upper motor neuron (UMN) facial palsy due to cerebrovascular accident possible thrombotic in etiology with site of lesion being left internal capsule, possible involving the lenticulostriate branch of middle cerebral artery (MCA). Patient is in state of neuronal shock. Patient has following risk factors _____.

FOR NEUROPATHY

Acute onset of symmetrical flaccid quadriplegia (ascending) with no evidence of sensory, bowel, bladder involvement with bilateral lower motor neuron (LMN) facial palsy, possible site of lesion in the peripheral nerve, pathology being demyelination—acute inflammatory demyelinating polyneuropathy (AIDP).

FOR SPINAL CORD DISEASE

Subacute onset of symmetrical spastic paraparesis with involvement of sensory, bladder, and bowel; with no involvement of cranial nerves with vertebral tenderness at T4-5, possible site of lesion is spinal cord, the disease being compressive myelopathy.

- Horizontal level
 - Extradural extramedullary
- Vertical level
 - Motor level: Above T10
 - Sensory level: At T8
 - Autonomic level: Above T12
 - Reflex level: Above T10
 - Spinal level: T8
 - Vertebral level: T5.

Possible etiology: Tuberculosis—Pott's spine.

FOR EXTRAPYRAMIDAL (PARKINSON'S DISEASE)

Insidious onset, slowly progressive, degenerative disease involving the motor system (in the form of rigidity and tremors) with no evidence of sensory, cranial nerves or bowel, bladder, we would consider involvement of extrapyramidal system probably parkinsonism with no evidence of secondary causes, no signs or symptoms of Parkinson's plus syndromes, functional status—Stage III (Hoehn and Yahr staging system).

FOR ATAXIA

Insidious onset, slowly progressive, symmetrical ataxia and cerebellar signs of trunk and limbs with no evidence of sensory, cranial nerve or autonomic involvement. I would like to consider the possibility of degenerative cerebellar ataxia possibly inherited (family history +ve).

C. CENTRAL NERVOUS SYSTEM: DISCUSSION ON CARDINAL SYMPTOMS

DISCUSSION ON CARDINAL SYMPTOMS

Taking a Neurological History

The neurological history should be a focused, goal-directed exercise that seeks to answer the following questions:

1. Which part of the nervous system is affected by “a pathological process” and is causing the symptoms (where is the lesion)? Is it a single lesion or are there multiple diffuse lesions? Alternatively, is there a diffuse problem affecting many neurological systems?
2. What is the underlying pathological process (e.g. vascular, inflammatory, degenerative)?
3. Is this a purely neurological problem or a neurological manifestation of a systemic disease?

Note:

- Ask the patient to tell their story in their own words
- Explore each symptom in detail, evaluating the evolution and the way the symptoms affect the ability to function
- Ask for an eyewitness account when cognition or consciousness is involved
- If you cannot make a neurological diagnosis, take the history again before arranging investigations.

Pathology of neurological diseases		
Acute	Subacute	Chronic
Vascular—stroke	Infection	Degeneration
Demyelination	Space occupying lesions	
Metabolic	Metabolic	

HIGHER MENTAL FUNCTION

Altered State of Consciousness

- Onset
- Any seizures, blackouts
- Any fall/injuries
- Any ear or nose bleed
- Fever
- Any ear pain or discharge
- Drug history
- Any addictions.

Other Higher Mental Functions

- Speech difficulty
- Difficulty to recognize people or objects
- Memory defects
- Inappropriate crying or laughter

- Lack of interest
- Social disinhibition
- Delusions/hallucinations.

Mental State and Cognition

- Changes in the memory
- State of alertness and drowsiness
- Changes in the mood and affect (loss of spontaneity)
- Language changes
- Loss of spatial orientation
- Diminished ability to carry out routine activities of daily living.

CRANIAL NERVE DYSFUNCTION

Ask about:

CN	Symptoms
1	Smell disturbance
2, 3, 4, 6	Diplopia, blurred vision, blindness, difficulty in opening eyelid (CN3)
5	Difficulty in chewing, loss of sensations over face
7	Deviation of angle of mouth, accumulation of food at one side of the mouth, dribbling of saliva, loss of taste sensation, hyperacusis
8	Tinnitus, hearing loss, dizziness, loss of balance
9, 10	Nasal intonation, nasal regurgitation of food, dysphagia, difficulty in speech, hoarseness of voice
11	Difficulty in neck/shoulder movements
12	Difficulty in mixing food in the mouth, difficulty in speech

For example: Left LMN 7th nerve palsy—history of retroauricular pain followed by abrupt onset deviation of angle of mouth to right with slurring of speech and difficulty in left eye closure with history of hyperacusis.

MOTOR DYSFUNCTION

Weakness

Distribution of Weakness

- Is it symmetrical/asymmetric?
- Plegia—complete loss of power—0/5 vs paresis—incomplete loss of power
- One limb: Monoparesis.
- Two limbs, same side: Hemiparesis.
- Both lower limbs: Paraparesis.
- All four limbs: Quadriplegia (or tetraparesis).
- **Pentaplegia** is a spinal cord injury at or above C4 level, resulting in complete loss of motor functions below the injury level and paralysis of respiratory muscles.

- Two (contralateral to each other) or three limbs (upper and lower limbs), e.g. right upper limb and left lower limb or left arm and both legs, both arms and one leg.
- Patterened weakness:
 - The pattern of pyramidal weakness is weakness of upper limbs extensors and lower limbs flexors.

For example: Right MCA territory embolic infarct—history of sudden onset, complete loss of power in left upper limb, lower limb associated with left UMN facial palsy. Weakness—maximum at onset, nonprogressive.

Causes of monoplegia affecting the lower limb	Causes of monoplegia affecting the upper limb
<ol style="list-style-type: none"> 1. Stroke, affecting anterior cerebral artery territory. 2. Cerebral venous sinus thrombosis affecting superior sagittal sinus. 3. Trauma, head injury, with contusion in the frontal lobe. 4. Infection, such as granuloma affecting frontal lobe. 5. Trauma to the lumbosacral plexus, diabetic lumbosacral plexopathy. 6. Functional or psychogenic. 	<ol style="list-style-type: none"> 1. Stroke, affecting superior division of contralateral middle cerebral artery territory, affecting parietal lobe, or unpaired anterior cerebral artery. 2. Head injury, with contusion in the parietal lobe. 3. Trauma to the brachial plexus. 4. Injury to multiple cervical nerve roots. 5. Functional or psychogenic.
Causes of hemiplegia	
<ol style="list-style-type: none"> 1. Ischemic or hemorrhagic stroke, affecting contralateral cerebral hemisphere, internal capsule, brainstem or ipsilateral upper cervical cord. 2. Cerebral venous sinus thrombosis with venous infarction of contralateral cerebral hemisphere. 3. Acute central nervous system infection, such as meningitis or encephalitis, brain abscess, granulomatous infections. 4. Head injury causing contusion/bleeding in the contralateral cerebral hemisphere, internal capsule, basal ganglia, or brainstem. 5. Tumor affecting cerebral hemisphere, internal capsule, basal ganglia, brainstem or cervical cord. 6. Bleeding into a brain tumor on the contralateral side. 7. Demyelinating illness, such as acute disseminated encephalomyelitis (ADEM) or multiple sclerosis (MS). 8. Todd's paresis. 9. Mill's hemiplegic variant of motor neuron disease (MND). 	

Causes of Quadriplegia (Table 6C.1)

Table 6C.1: Causes of quadriplegia.	
UMN causes	LMN causes
<ul style="list-style-type: none"> • Cerebral palsy • Bilateral brainstem lesion (glioma) • Craniovertebral junction anomaly • High cervical cord compression • Multiple sclerosis • Motor neuron disease 	<ul style="list-style-type: none"> • Acute anterior poliomyelitis • GB syndrome • Peripheral neuropathy • Myopathy or polymyositis • Myasthenia gravis • Periodic paralysis • Snake bite, organophosphorous poisoning, etc.

Causes of Paraplegia

Causes of Flaccid Paraplegia (LMN type)

- **UMN lesion in shock stage**, i.e. sudden onset or history of long duration as in extradural transverse myelitis and spinal injury
- **Lesion involving anterior horn cells:**
 - Acute anterior poliomyelitis

- Progressive muscular atrophy (a variety of motor neuron disease)
- **Diseases affecting nerve root:** tabes dorsalis, radiculitis, GB syndrome
- **Diseases affecting peripheral nerves:**
 - Acute infective polyneuropathy (GB syndrome)
 - High cauda equina syndrome
 - Disease of peripheral nerves involving both the lower limbs
 - Lumbar plexus injury (psoas abscess or hematoma)
- **Diseases affecting myoneural junction:**
 - Myasthenia gravis, Lambert-Eaton syndrome
 - Periodic paralysis due to hypo- or hyperkalemia
- **Diseases affecting muscles:** Myopathy.

Onset and Progression

- Acute, subacute, or chronic.
- Reversible, stable nonreversible, fluctuating, stuttering or step-ladder, or progressive.
- **Ascending weakness**—first lower limbs→upper limbs→GB syndrome, extramedullary compressive myelopathy
- **Descending weakness**—first upper limbs→lower limbs→Miller Fisher variant of GB syndrome, intramedullary compressive myelopathy.
- **Ellsberg phenomenon**—compressive lesions near the high cervical cord produce weakness of the ipsilateral shoulder and arm followed by weakness of the ipsilateral leg, then the contralateral leg, and finally the contralateral arm, an “anticlock-wise” pattern that may begin in any of the four limbs.

Table 6C.2: Causes of spastic paraplegia [upper motor neuron (UMN) type lesion].

A. Gradual onset	B. Sudden onset
Cerebral causes	
<ul style="list-style-type: none"> • Parasagittal meningioma • Hydrocephalus 	Thrombosis of unpaired anterior cerebral artery or superior sagittal sinus
Spinal causes	
<p>Compressive or transverse lesion in the spinal cord: Cord compression</p> <p>Noncompressive or longitudinal lesion or systemic disease of the spinal cord</p> <ul style="list-style-type: none"> • Motor neuron disease (MND), e.g. amyotrophic lateral sclerosis • Multiple sclerosis, Friedreich's ataxia • Subacute combined degeneration (i.e. from vitamin B₁₂ deficiency) • Lathyrism, Syringomyelia, Erb's spastic paraplegia, Tropical spastic paraparesis • Radiation myelopathy 	<p>Compressive causes</p> <ul style="list-style-type: none"> • Injury to the spinal cord (fracture-dislocation or collapse of the vertebra) • Intervertebral disc prolapse • Spinal epidural abscess or hematoma <p>Noncompressive causes</p> <ul style="list-style-type: none"> • Acute transverse myelitis • Thrombosis of anterior spinal artery • Hematomyelia (from arteriovenous malformation, angiomas, or endarteritis)

Muscles/Limb(s) Involved

Proximal upper limb—shoulder/arm:	Difficulties combing hair, reaching for high objects, winging of scapula
Distal upper limb—forearm/hand:	Finger/wrist drop, poor hand grip, cannot open jar, difficulty in buttoning/unbuttoning
Proximal lower limb—pelvic/thigh:	Cannot rise from chair or squatting position, waddling gait
Distal lower limbs—leg/foot:	Difficulty in gripping chappals, cannot walk on heels/toes, foot drop

Neck muscles	Dropped head/broken neck
Trunk	Inability to roll on the bed

- **Variation throughout day—fatigability:** In postsynaptic neuromuscular junction disorders like myasthenia gravis the weakness worsens on exertion.
- **Wasting/loss of muscle bulk**—wasting is a feature of LMN disease. Florid wasting is seen in motor neuron disease. Usually associated with fasciculations. In late stages of UMN disease disuse atrophy may be seen.
Wasting of muscles also results in undue prominence of underlying bones.
- **Stiffness of limbs**—increased tone of the limbs resulting in stiffness and heaviness of limbs is a characteristic feature of UMN disease. Patients may complain that the limbs are heavy as log of wood in spasticity, while they may say that the limbs are floppy in LMN diseases.
- **Gait abnormalities:** It may aid in the diagnosis.
 - Limp or dragging foot—might suggest LMN disease/foot drop
 - Scissoring/circumduction may suggest UMN disease.
- **Involuntary movements:**
 - Type
 - Symmetrical/asymmetrical
 - Part of the body involved
 - Present at rest
 - Functional disability.

SENSORY DYSFUNCTION

- Numbness/loss of feeling
- Altered feeling:
 - Paresthesia
 - Dysesthesias (tingling, pin-needles)
 - Spontaneous pain
- Pattern of sensory loss:

Pattern of sensory loss	Site of the lesion
Hemisensory loss—same side face and body	Internal capsule/thalamus
Crossed sensory—one side face, opposite side body	Lateral medulla
Ascending sensory loss—lower limbs → upper limb	Extramedullary compressive myelopathy
Descending sensory loss—upper limbs → lower limb	Intramedullary compressive myelopathy
Dissociative sensory loss (only pain and temperature lost, posterior column sensations preserved)	Intramedullary compressive myelopathy Lateral medullary syndrome Anterior cord syndrome
Definite sensory level (below which all sensations lost)	Suggestive of spinal cord disease
Graded sensory loss—glove and stocking	Suggestive of peripheral neuropathy

Positive and Negative Symptoms

Abnormal sensory symptoms can be divided into two categories: positive and negative.

Positive Symptoms

- Altered sensation that are described as pricking, bandlike, lightning-like shooting feelings (lancinations), aching, knifelike, burning, scarring, electrical. Such symptoms are often painful.
- Positive phenomena usually result from trains of impulses generated at sites of lowered threshold or heightened excitability along a peripheral or central sensory pathway.
- Because positive phenomena represent excessive activity in sensory pathways, they may or may not be associated with a sensory deficit (loss) on examination.

Negative Symptoms

- Represent loss of sensory function and are characterized by diminished or absent feeling that often is experienced as numbness and by abnormal findings on sensory examination.
- It is estimated that at least one-half of the afferent axons innervating a particular site are lost or functionless before a sensory deficit can be demonstrated by clinical examinations.
- Subclinical degrees of sensory dysfunction may be revealed by sensory nerve conduction studies.
- Whereas sensory symptoms may be either positive or negative, sensory signs on examination are always a measure of negative phenomena.

Sense	Test device	Endings activated	Fiber size mediating
Pain	Pin prick	Cutaneous nociceptors	Small
Temperature (heat)	Warm metal object	Cutaneous thermoreceptors for hot	Small
Temperature (cold)	Cold metal object	Cutaneous thermoreceptors for cold	Small
Touch	Cotton wisp, fine brush	Cutaneous mechanoreceptors, also naked endings	Large and small
Vibration	Tuning fork, 128 Hz	Mechanoreceptors, especially Pacinian corpuscles	Large
Joint position	Passive movements of specific joints	Joint capsule tendon endings, muscle spindles	Large

CEREBELLAR EXAMINATION

Coordination and Balance

1. Difficulty in walking
2. Unsteadiness
3. Falls
4. Staggering
5. Loss of balance in dark.

AUTONOMIC DYSFUNCTION

Bladder Dysfunction (Table 6C.3)

- History of:

- Urinary retention
- Loss of awareness of bladder control
- Frequency, urgency, urge and overflow maintenance.

MENINGEAL SIGNS

- Headache
- Projectile vomiting
- Photophobia
- Neck pain

OTHERS

Dizziness, vertigo, blackouts, and fatigue

Dizziness: It covers many complaints, from a vague feeling of unsteadiness to severe, acute vertigo. It is frequently used to describe lightheadedness felt in panic and anxiety, during palpitations, and in syncope or chronic ill-health. The real nature of this symptom must be determined.

Vertigo: An illusion of movement—is more definite. It is a sensation of rotation, or tipping. The patient feels that the surroundings are spinning or moving. It is distinctly unpleasant and often accompanied by nausea or vomiting.

Blackout like dizziness, is a descriptive term implying either altered consciousness, visual disturbance or falling. Epilepsy, syncope, hypoglycemia, anemia must be considered. However, commonly no sinister cause is found. A careful history from an eyewitness is essential.

Fatigue is another common symptom of neurological disorders.

Table 6C.3: Various causes of neurogenic bladder.

Type	<i>Uninhibited bladder/detrusor hyperreflexia</i>	<i>Automatic bladder/detrusor sphincteric dyssynergia</i>	<i>Autonomous bladder/detrusor areflexia</i>	<i>Sensory atonic bladder</i>	<i>Motor atonic bladder</i>
Site of lesion	Suprapontine neurologic disorder, mostly frontal lobe	UMN disorder of the suprasacral spinal cord	LMN lesion at the sacral cord	LMN lesion—peripheral nerve	
Causes	Frontal tumors, parasagittal meningioma, ACA aneurysm, NPH	Spinal cord trauma, compressive myelopathy, myelitis	Cauda equina syndrome, conus medullaris lesion, spinal shock	Diabetes mellitus, amyloidosis, tabes dorsalis	Lumbosacral meningocele, tethered cord syndrome, lumbar canal stenosis
Bladder sensation	Preserved	Interrupted	Absent	Absent	Intact
Size of bladder	Normal	Small	Large	Large	Large
Ability to initiate voiding	Present	Absent	Absent	Present	Lost
Type of incontinence	Urge/social disinhibition	Urge	Overflow	Overflow	Overflow
Residual urine	Nil	Small	Large amount	Large	Large

Anal sphincter tone	Normal	Normal	Lost	Normal	Lost
Perianal sensation	Normal	Normal	Absent	Absent	Preserved
Bulbocavernous/anal reflex	Normal	Normal	Absent	Absent	Preserved
Treatment	Anticholinergic medication	Self-intermittent catheterization	Continuous catheterization		

NECK PAIN

Deformities: Infantile torticollis	Infections of bone: TB of cervical spine. Pyogenic infection of cervical spine	Tumors: Benign and malignant tumors in relation to cervical spine and nerve roots
Arthritis of spinal joints: Rheumatoid arthritis-ankylosing spondylitis (RA-AS) Cervical spondylosis	Mechanical derangement: <ul style="list-style-type: none"> • Prolapsed cervical disc • Cervical spondylolisthesis • Whiplash injury • Cervical spine fracture • Neck muscle strain • Neck sprain 	Referred pain: <ul style="list-style-type: none"> • Ear • Throat • Brachial plexus • Angina (pain extends to neck) • Aortic aneurysm • Meningismus

BACKACHE

Musculoskeletal	Infectious
<ul style="list-style-type: none"> • Nonspecific musculoskeletal back pain • Spondylolysis/spondylolisthesis • Scoliosis • Scheuermann disease • Disc degeneration and/or prolapsed 	<ul style="list-style-type: none"> • Discitis • Vertebral osteomyelitis including tuberculosis (Pott disease) • Epidural abscess • Sacroiliac joint infection
Others	Nonspinal infection
<ul style="list-style-type: none"> • Intervertebral disc calcification • Congenital absence of pedicle • Vertebral apophyseal fracture • Aneurysmal bone cyst • Sacroiliac joint stress reaction • Idiopathic juvenile osteoporosis 	<ul style="list-style-type: none"> • Paraspinous muscle abscess • Pyelonephritis • Pneumonia • Pelvic inflammatory disease • Endocarditis • Viral myalgias
Inflammatory	Neoplastic
<ul style="list-style-type: none"> • Ankylosing spondylitis • Psoriatic arthritis • Inflammatory bowel disease-associated arthritis • Reactive arthritis 	<ul style="list-style-type: none"> • Osteoid osteoma • Leukemia or lymphoma • Solid malignancy, primary or metastatic • Other benign tumor: Neurofibroma, vascular malformation
Others	
<ul style="list-style-type: none"> • Appendicitis • Sickle cell pain crisis • Syringomyelia • Cholecystitis • Pancreatitis 	<ul style="list-style-type: none"> • Chronic recurrent multifocal osteomyelitis • Psychosomatic illness • Nephrolithiasis • Ureteropelvic junction obstruction

RED FLAGS FOR ACUTE LOW BACK PAIN

<i>History</i>
<ul style="list-style-type: none">• Cancer• Unexplained weight loss• Immunosuppression• Prolonged use of steroids• Intravenous drug use• Urinary tract infection• Pain worse at night or when supine• Fever• Significant trauma related to age• Bladder or bowel incontinence• Urinary retention (with overflow incontinence)
<i>Physical examination</i>
<ul style="list-style-type: none">• Saddle anesthesia• Loss of anal sphincter tone• Major motor weakness in lower extremities• Fever• Vertebral tenderness• Limited spinal range of motion• Neurologic findings persisting beyond 1 month

NOTES

D(i). GENERAL EXAMINATION IN NEUROLOGY

GENERAL PHYSICAL EXAMINATION IN NERVOUS SYSTEM

Pulse

- Decreased pulse rate—increased intracranial pressure (ICP)—Cushing reflex
- Resting tachycardia autonomic dysfunction
- Irregularly irregular—atrial fibrillation (AF)
- Feeble pulse, carotid bruit—atherosclerosis.

Blood pressure

- Increased BP—intracranial (IC) bleed—reactionary hypertension
- Cushing's reflex.
- Orthostatic hypotension

Jugular Venous Pressure

Increased in high output states.

Fever

- Meningitis
- Encephalitis
- CVA
- Brain abscess
- Epidural abscess
- Vasculitis
- ADEM
- Complex partial seizures
- Normal pressure hydrocephalus
- Myotonic dystrophy
- Hypothalamic dysfunction.

Pallor

- Vitamin B₁₂ deficiency
- Pica, restless leg syndrome—iron deficiency
- Chronic liver disease (CLD), chronic kidney disease (CKD)—encephalopathy.

Icterus

- Hepatic encephalopathy
- Kernicterus.

Clubbing

- Syringomyelia
- Chronic hemiplegia
- Median nerve injury.

Lymphadenopathy

- Lymphoma—neuropathy, cerebellar ataxia, intracranial metastasis
- Paraneoplastic syndrome:
 - Lung carcinoma—Lambert–Eaton Myasthenic syndrome
 - Lymphoma.
- Drug induced—phenytoin.

Pedal Edema

- Chronic liver disease
- Chronic kidney disease
- Autonomic dysfunction.

Signs of Nutritional Deficiency

Discussed earlier.

NEUROCUTANEOUS SYNDROMES/PHAKOMATOSES

The neurocutaneous syndromes include a heterogeneous group of disorders characterized by abnormalities of both the integument and central nervous system (CNS).

Most disorders are familial and believed to arise from a defect in differentiation of the primitive ectoderm.

Common neurocutaneous syndromes	
<ul style="list-style-type: none">• Neurofibromatosis I and II• Tuberous sclerosis• Von Hippel–Lindau disease• Sturge–Weber syndrome• Klippel–Trenaunay–Weber syndrome• Osler–Weber–Rendu syndrome• PHACE syndrome• Wyburn–Mason syndrome• Linear nevus sebaceous syndrome• Neurocutaneous melanosis• Waardenburg syndrome type 1 and 2• Fabry's disease	<ul style="list-style-type: none">• Lentiginosis, deafness, cardiopathy syndrome• Hypomelanosis of Ito• Ataxia-telangiectasia (Louis–Bar syndrome)• Xeroderma pigmentosum• Cockayne's syndrome• Rothmund-Thomson syndrome• Sjögren-Larsson syndrome• Neuroichthyosis• Werner syndrome and progeria• Incontinentia pigmenti• Neurocutaneous melanosis• Retinal—neurocutaneous cavernous hemangioma syndrome (Weskamp-Cotlier syndrome)

NEUROFIBROMATOSIS [FIG. 6D(I).1]

Two types of neurofibromatosis (type 1 and type 2).



Fig. 6D(i).1: Neurofibromas.

Neurofibromatosis 1

Synonyms: von Recklinghausen disease and Watson disease.

Most prevalent neurocutaneous syndrome.

- Autosomal dominant
- The *NF1* gene on chromosome region 17q11.2 encodes a protein also known as neurofibromin. Neurofibromin acts as an inhibitor of the oncogene Ras.

Diagnostic Criteria

<i>Two out of the following seven signs</i>
1. Six or more café-au-lait macules over 5 mm in greatest diameter in prepubertal individuals and over 15 mm in greatest diameter in postpubertal individuals. 2. Axillary or inguinal freckling. 3. Two or more Iris Lisch nodules [Fig. 6D(i).2]. 4. Two or more neurofibromas or one plexiform neurofibroma. 5. A distinctive osseous lesion, such as sphenoid dysplasia (which may cause pulsating exophthalmos) Or cortical thinning of long bones with or without pseudarthrosis. 6. Optic gliomas. 7. A first-degree relative with NF1 whose diagnosis was based on aforementioned criteria.

Conditions with Café-au-lait Macules [Fig. 6D(i).3]

- Neurofibromatosis type 1 and 2
- McCune–Albright syndrome
- Ataxia telangiectasia
- Bloom's syndrome
- Familial Café-au-lait macules.

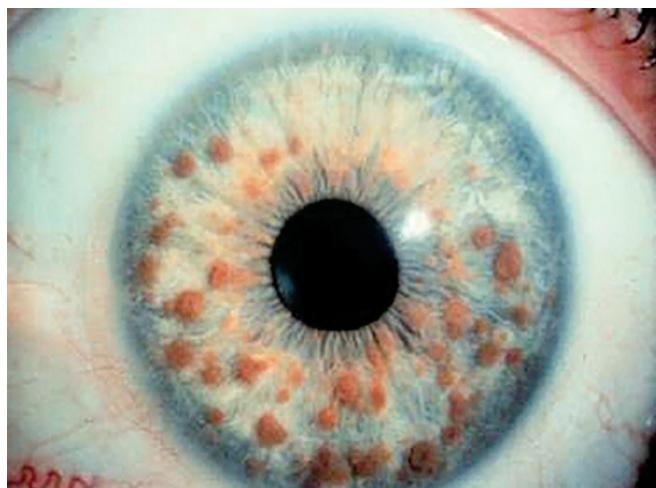


Fig. 6D(i).2: Iris nodules (Lisch nodules).



Fig. 6D(i).3: Café-au-lait macules (CALM).

Neurofibromatosis 2

The *NF2* gene (also known as merlin or schwannomin) is located on chromosome 22q1.11.

Diagnostic Criteria for Neurofibromatosis 2

<i>One of the following three features is present</i>
1. Bilateral vestibular schwannomas 2. A parent, sibling, or child with NF2 and either unilateral vestibular schwannoma or any two of the following: meningioma, schwannoma, glioma, neurofibroma, or posterior subcapsular lenticular opacities 3. Multiple meningiomas (two or more) and unilateral vestibular schwannoma or any two of the following: schwannoma, glioma, neurofibroma, or cataract.

TUBEROUS SCLEROSIS [TABLE 6D.(I).1)]

- Also called Bourneville disease
- Autosomal dominant

- Widespread hamartomas—brain, eyes, skin, kidneys, liver, heart, and lungs.
- Clinical triad described by Vogt:

EPI-LOI-A

- » Epilepsy
- » Low intelligence
- » Adenoma sebaceum [Figs. 6D(i).4A to C].

STURGE–WEBER SYNDROME [FIG. 6D(I).5]

- Results from anomalous development of the primordial vascular bed in the early stages of cerebral vascularization.
- As a result, brain becomes atrophic and calcified, particularly in the molecular layer of the cortex.

Clinical Manifestations

- Facial capillary malformation—Port-wine stain
- Unilateral facial nevus
- Buphthalmos and glaucoma of the ipsilateral eye
- Seizures in the 1st year of life in most patients.

Skull Radiograph

Serpentine or railroad track intracranial calcification in the occipitoparietal region.

Table 6D(i).1: Diagnostic criteria for tuberous sclerosis complex (TSC).

Major features	Minor features
<ul style="list-style-type: none"> • Facial angiofibromas or forehead plaque • Nontraumatic ungual or periungual fibroma (Koenen's tumour) • Shagreen patch (connective tissue nevus) (Fig. 6D(i).4A) • Hypomelanotic macules (more than three) (Fig. 6D(i).4B) • Multiple retinal nodular hamartomas • Cortical tuber • Subependymal nodule • Subependymal giant cell astrocytoma • Cardiac rhabdomyoma, single or multiple • Lymphangiomyomatosis • Renal angiomyolipoma 	<ul style="list-style-type: none"> • Multiple randomly distributed pits in dental enamel • Hamartomatous rectal polyps • Bone cysts • Cerebral white matter migration lines • Gingival fibromas • Non-renal hamartoma • Retinal achroic patch • “Confetti” skin lesions • Multiple renal cysts

Definite TSC: Either two major features or one major feature with two minor features

Probable TSC: One major feature and one minor feature

Possible TSC: Either one major feature or two or more minor features



Figs. 6D(i).4A to C: (A) Shagreen patch; (B) Ash leaf-shaped macule is a hypopigmented macule oval at one end and pointed at the opposite end; (C) Adenoma sebaceum.



Fig. 6D(i).5: Sturge–weber syndrome.

VON HIPPEL–LINDAU DISEASE

- Autosomal dominant trait
- von Hippel-Lindau (VHL) tumor suppressor gene located on 3p25-26.

Clinical Features

- Cerebellar hemangioblastoma
- Retinal angioma
- Cystic lesions of the kidneys, pancreas, liver, and epididymis
- Pheochromocytoma.

PHACE SYNDROME

- Posterior fossa malformation
- Hemangiomas ipsilateral to the aortic arch
- Arterial anomalies
- Coarctation of the aorta, aplasia or hypoplasia of carotid arteries, aneurysmal carotid dilatation, aberrant left subclavian artery
- Eye abnormalities—glaucoma, cataracts, microphthalmia, and optic nerve hypoplasia.

ATAXIA TELANGIECTASIA

- Autosomal recessive
- Chromosome 11
- Cerebellar atrophy
- Telangiectasia appears on bulbar conjunctiva and skin
- Sinopulmonary infections
- Lymphoreticular malignancies
- Immune deficiency.

NERVE THICKENING

Detecting enlargement of accessible nerves is very helpful in assessing patients with peripheral nerve disorders, as only a few types of neuropathy lead to nerve thickening. Clinical landmarks and sites of palpable nerves are given in **Table 6D(i).2** and **Figure 6D(i).6**.

Table 6D(i).2: Clinical landmarks of palpable nerves.

Nerve	Anatomical site	Palpated against
Supraorbital [Fig. 6D(i).7]	Forehead	Orbital ridge of frontal bone
Infraorbital	Cheek	Zygomatic bone
Greater auricular [Figs. 6D(i).8 and 6D(i).9]	Neck, anterior branch across the sternocleidomastoid, posterior branch over the sternocleidomastoid	Sternocleidomastoid
Ulnar [Fig. 6D(i).10]	Elbow joint	Behind medial epicondyle in olecranon groove
Superficial radial	Above wrist joint	Against lateral border of radius
Median	Near wrist joint, proximal to the flexor retinaculum	Against carpal bones
Common peroneal [Fig. 6D(i).11]	Knee joint	Against fibular head
Posterior tibial	Ankle joint, below and behind medial malleolus	Against calcaneus
Sural	Lateral side of lower third of leg	Fibula

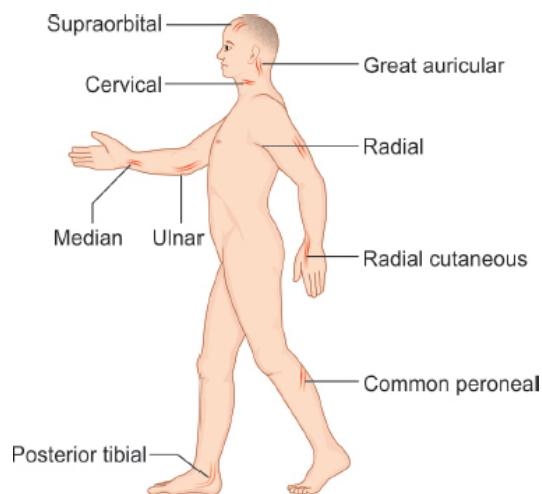


Fig. 6D(i).6: Sites of palpable nerves.

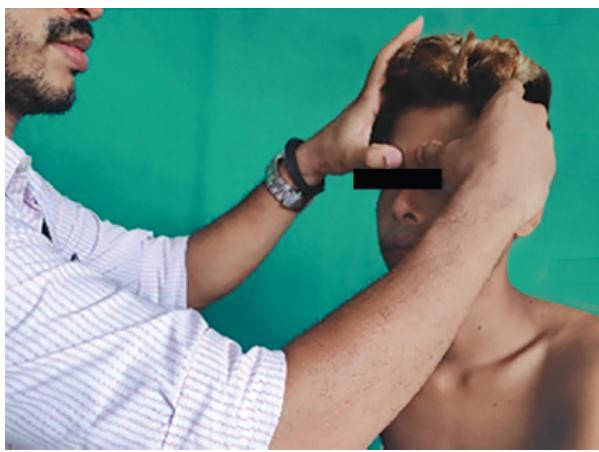


Fig. 6D(i).7: Supraorbital nerve.



Fig. 6D(i).8: Greater auricular nerve.



Fig. 6D(i).9: Greater auricular nerve of neck.



Fig. 6D(i).10: Ulnar nerve.



Fig. 6D(i).11: Common peroneal nerve.

Causes of Nerve Thickening

Infective

Leprosy

Hereditary

- Hereditary motor and sensory neuropathy types 1 and 3 (Charcot–Marie–Tooth neuropathy, Dejerine–Sottas syndrome)
- Refsum's disease.

Acquired immune mediated

- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
- Chronic inflammatory sensory polyradiculopathy (CISP)
- Multifocal acquired demyelinating sensory and motor polyneuropathy (MADSAM)
- Relapsing Guillain-Barre syndrome (GBS).

Tumors of nerves or nerve sheath

- Localized hypertrophic neuropathy
- Schwannoma
- Neurofibromatosis 1 and 2.

Nerve infiltrations

- Neurolymphomatosis
- Acromegaly
- Amyloidosis
- Sarcoidosis.

NOTES

D(ii). HIGHER MENTAL FUNCTIONS

NERVOUS SYSTEM EXAMINATION

Handedness

Handedness	
<i>Right handed (90–95%)</i>	<i>Left handed (5–10%)</i>
99% have—left dominant hemisphere 1% have—right dominant hemisphere	60–70% have—left dominant hemisphere 15–20% have—right dominant hemisphere 15–20% have—mixed dominance

Examination

Any of the following methods can be adopted:

- Ask the patient to kick a football, normally the dominant side leg is used.
- Ask the patient to peep through a keyhole, normally the dominant side eye is used.
- Ask the patient to fold the arms in front one over the other, the dominant hand is the one which lies anteriorly.
- Ask the patient to “stand at ease” position, the dominant hand is the one which lies posteriorly.

Clinical Implications

- Handedness is important for rehabilitation of the patient (right-handed individuals—dominant left hemisphere needs to be aggressively rehabilitated so as to have minimal residual deficit).
- Degenerative diseases like Huntington’s disease have been postulated to be more common in individuals with right dominant cortex.
- Failure to develop clear hemispheric dominance has been implicated in dyslexia, stuttering, mirror writing, learning disability, and general clumsiness.

Education

- Formal education up to standard _____.
- It is important for testing components of higher mental functions like calculation, reading, and writing.

CONSCIOUSNESS

The ascending reticular activating system (RAS) arising from the reticular formation of the brainstem, primarily the paramedian tegmentum of the upper pons and midbrain, and projects to the paramedian, parafascicular, centromedian, and intralaminar nuclei of the thalamus. This is the primary control of consciousness.

The hypothalamus is also important for consciousness; arousal can be produced by stimulation of the posterior hypothalamic region.

Coma

- It is a state of complete loss of consciousness from which the patient cannot be aroused by ordinary stimuli.
- There is complete unresponsiveness to self and the environment.
- The patient in coma has no awareness of themselves, makes no voluntary movements, and has no sleep-wake cycles.

Stupor	<ul style="list-style-type: none"> It is a state of partial or relative loss of response to the environment in which the patient's consciousness may be impaired to varying degrees. The patient can be aroused only with vigorous or unpleasant stimuli (e.g. sharp pressure or pinch, or rolling a pencil across the nail bed). No significant voluntary verbal or motor responses. Mass movement responses may be observed in response to painful stimuli or loud noises. <p>For example:</p> <ul style="list-style-type: none"> Bilateral cerebral hemisphere disease Upper brainstem diseases
Lethargy/drowsiness	Patient can usually be aroused or awakened and may then appear to be in complete possession of their senses, but promptly falls asleep when left alone. It resembles normal sleepiness. For example: High brainstem disturbances
Obtundation	Refers to moderate reduction in the patient's level of awareness such that stimuli of mild-to-moderate intensity fail to arouse; when arousal does occur, the patient is slow to respond.
Minimally conscious (vegetative) state	<ul style="list-style-type: none"> Return of irregular sleep-wake cycles and normalization of the so-called vegetative functions—respiration, digestion, and blood pressure control. The patient may be aroused, but remains unaware of his or her environment. There is no purposeful attention or cognitive responsiveness.
Persistent vegetative state	Individuals who remain in a vegetative state 1 year or longer after traumatic brain injury (TBI) and 3 months or more after anoxic brain injury.
Confusional state	<p>Patients may appear alert, but are confused and disoriented. It is usually tested in three dimensions:</p> <ol style="list-style-type: none"> Time Place Person.
Delirium	<p>It is an acute organic mental disorder characterized by confusion, restlessness, incoherence, inattention, anxiety, or hallucinations which may be reversible with treatment.</p> <p>For example:</p> <ul style="list-style-type: none"> Toxicity (alcohol) Infections
Catatonia	<ul style="list-style-type: none"> Symptom of psychotic state in which the patient is otherwise normal. He does not follow movements, does not appear to pay attention to surroundings and will often have aplastic rigidity of limbs which may remain in any position in which they are placed (however bizarre the position may be).

It is preferable to describe the patient's state of responsiveness or use an objective and well-defined scheme, such as the Glasgow Coma Scale (GCS).

Glasgow Coma Scale (GCS)					
Eye opening		Best verbal response		Best motor response	
				Obeys commands	6
		Oriented and converses	5	Localizes pain	5
Open spontaneously	4	Converses, but disoriented, confused	4	Exhibits flexion withdrawal	4
Open only to verbal stimuli	3	Uses inappropriate words	3	Decorticate rigidity	3
Open only to pain	2	Makes incomprehensible sounds	2	Decerebrate rigidity	2
Never open	1	No verbal response	1	No motor response	1
Maximum score = 15 Minimum score = 3 Coma is equal to GCS of 8 or less.					

Mnemonic (GCS → EVM = 4, 5, and 6)

Note: In intubated patients, verbal response is denoted as V_T.

Glasgow coma scale—pupils score

- The Glasgow coma scale-pupils score (GCS-P) was described in 2018 as a strategy to combine the two key indicators of the severity of traumatic brain injury into a single simple index
- Calculation of the GCS-P is by subtracting the pupil reactivity score (PRS) from the Glasgow coma scale (GCS) total score:

$$\text{GCS-P} = \text{GCS} - \text{PRS}$$

- The pupil reactivity score is calculated as follows:

Pupils unreactive to light	Pupil reactivity score
Both pupils	2
One pupil	1
Neither pupil	0

- The GCS-P score can range from 1 and 15 and extends the range over which early severity can be shown to relate to outcomes of either mortality or independent recovery.

ORIENTATION

Time	Ask for year, season, month, date, and time
Place	Ask for country, state, city, hospital name, and floor/ward
Person	<ul style="list-style-type: none">• What is your name?• How old are you?• Where were you born?• What is the name of your wife/husband?

Findings are documented in the medical record as follows: Patient is alert and oriented × 3 (time, person, and place) or × 2 (person, place) depending on the domains correctly identified.

An additional domain that can be examined is **circumstance**.

(What happened to you? What kind of a place is this? Why do people come here?)

APPEARANCE/BEHAVIOR

- Mood and affect
- Thought and perception

These have been discussed under Chapter 9—Approach to Psychiatric Illness.

MEMORY

Classification of Memory

Explicit memory (declarative memory)	Implicit memory
Involves conscious recall and requires integrity of various cortical regions	Does not require conscious recall. Involves basal ganglia and cerebellum
Can be tested bedside	Cannot be tested bedside
It includes: • Immediate (prefrontal cortex) • Recent (medial temporal structures) • Remote (widespread neocortical areas).	It includes: • Procedural memory (basal ganglia)—like riding a car • Classical conditioning (cerebellum) • Probabilistic classification learning (basal ganglia).

Examination of Explicit Memory

Types of memory	Description and testing	Areas in brain
Immediate (working memory)	<ul style="list-style-type: none"> Digit span is a test of immediate memory, a very short-term function in which the material is not actually committed to memory Ask patient to repeat series of random digits forward and backward Normal digit span is 7 ± 2 	Dorsolateral frontal lobe, prefrontal cortex, and perisylvian cortex
Recent (short-term)	<ul style="list-style-type: none"> Recent, or short-term memory is tested by giving the patient items (pen, phone, and bottle) to recall After ensuring the patient has registered the items, proceed with other testing. After approximately 5 minutes, ask the patient to recall the items 	<ul style="list-style-type: none"> Mammillothalamic tract Hippocampus Parahippocampal cortex (spatial memory) Amygdala (emotional aspects) Perirhinal cortex (for visual) Medial temporal structures and connections
Remote (long-term)	<ul style="list-style-type: none"> A patient's fund of information reflects their remote memory. The fund of information includes schooling details, famous personalities, major events in history, etc. 	<ul style="list-style-type: none"> Widespread Neocortical areas
Episodic memory refers to the system involved in remembering particular episodes or experiences, such as the movie you saw last weekend or the meeting you attended yesterday. Semantic memory refers to the type of long-term memory concerned with factual details outside of personal details Budson and Price concept of memory systems: The frontal lobe can be considered as filing clerk, deciding which information has to be filed or retrieved. The medial temporal lobes are the actual filing cabinets for recent memories and the neocortical regions are filing cabinets for remote memories Wernicke's encephalopathy—g lobal confusion, ophthalmoplegia and ataxia (mnemonic—goa). Korsakoff's psychosis: Recent memory loss + confabulation (anteromedial thalamus)		

Amnesia

Anterograde amnesia	Impaired registration and recall of new information
Retrograde amnesia	Impaired recall of information registered within a certain interval before the disease onset

ATTENTION

- Attention is the directing of consciousness to a person, thing, perception, or thought.
- It depends on the capacity of the brain to process information from the environment or from long-term memory.
- An individual with intact selective attention is able to screen and process relevant sensory information about both the task and the environment while screening out irrelevant information.

- Selective attention can be examined by asking the patient to attend to a particular task.
- For example, the doctor asks the patient to repeat a short list of numbers forward or backward (digit span test).
- Normally, individuals can recall seven forward and five backward numbers.
- **Sustained attention (or vigilance)** is examined by determining how long the patient is able to maintain attention on a particular task (time on task).
- **Alternating attention (attention flexibility)** is examined by requesting the patient to alternate back and forth between two different tasks (e.g. add the first two pairs of numbers, then subtract the next two pairs of numbers).
- Requesting the patient to perform two tasks simultaneously determines divided attention.
- For example, the patient talks while walking (Walkie-Talkie test).

INTELLIGENCE/CALCULATION

Serial sevens, or spelling of any word backward.

COGNITION ASSESSMENT TOOLS

- Mini Mental Status Examination (MMSE)—Folstein's

O	Orientation	Place Time	10
R	Registration	Name 3 objects	3
A	Attention and calculation	Serial 7/word backward	5
R	Registration recall	Recall previously named 3 objects	3
L	Language	3 stage command Name two objects Read and follow Draw a pentagon Repetition Write a sentence	9

- MMSE total score:
 - 21–24: Mild cognitive dysfunction
 - 10–20: Moderate
 - Less than 10: Severe.
- Montreal cognitive assessment (MoCA)
- Cognitive state test (COST)
- Addenbrooke's cognitive examination (ACE)
- Cambridge cognitive examination (CAMCOG)
- Brief cognitive assessment tool (BCAT), and
- Short test of mental status (STMS).

SPEECH

Definitions

Phonation	It is defined as the production of vocal sounds without word formation; it is entirely a function of the larynx
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Vocalization	It is the sound made by the vibration of the vocal folds, modified by working of the vocal tract
Speech	It consists of words which are articulate vocal sounds that symbolize and communicate ideas
Articulation	It is the enunciation of words and phrases; it is a function of organs and muscles innervated by the brainstem
Language (Fig. 6D(ii).1)	<ul style="list-style-type: none"> • It is a mechanism for expressing thoughts and ideas as follows: • By speech (auditory symbols) • By writing (graphic symbols), or • By gestures and pantomime (motor symbols) • Language may be regarded as any means of expressing or communicating feeling or thought using a system of symbols. • It is a function of the cerebral cortex
Aphasia	Aphasia is an acquired disorder with loss or defective language content of speech resulting from damage to the speech centers within the dominant (usually left in 97%) hemisphere
Paraphasia	Substitution in the components of speech, e.g. foon for spoon
Neologism	Use of words which are nonexistent. Classically seen with Wernicke's aphasia
Jargon	Completely meaningless speech containing neologisms and paraphasias. Described in Wernicke's aphasia
Echolalia	Continuous repetition of heard words or sentences. Seen with transcortical sensory and transcortical mixed aphasias.
Alexia	It is the impairment of visual word recognition, in the context of intact auditory word recognition and writing ability
Agraphia	It is the inability to write, as a language disorder resulting from brain damage
Anomia	In this, word approximates the correct answer but it phonetically inaccurate (plentil for pencil)—phonemic paraphasia. When the patient cannot say the appropriate name when an object is shown but can point the object when the name is provided, it is known as one way or retrieval-based naming deficit
Mutism	Unable to speak or make sound
Aphonia	Unable to produce sound
Aphemia	Loss of speech

Slurred speech can be because of aphasia or dysarthria.

Aphasia	Dysarthria
Aphasia is a disorder of language	Dysarthria is a disorder of the motor production or articulation of speech
Usually due to cerebral dysfunction/lesions	Dysarthria is defective articulation of sounds or words of neurologic origin (usually brainstem)
Aphasia usually affects other language functions, such as reading and writing	In dysarthria, there are often other accompanying bulbar abnormalities, such as dysphagia

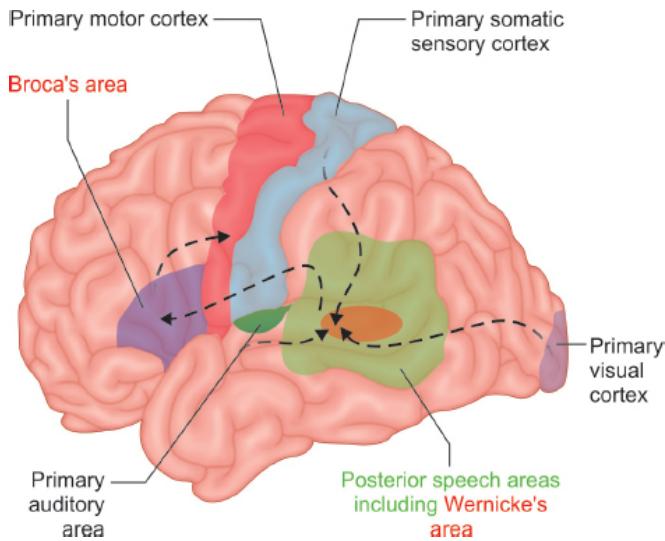


Fig. 6D(ii).1: Language and the brain.

Wernicke's area (area 22)	Arcuate fasciculus	Broca's area (area 44)
Decoding of sounds into language information (comprehension)	Communication between the Broca's and Wernicke's area. Needed for speech repetition	Responsible for spontaneous speech output (i.e.) fluency. Approximate number words produced per minute is 100/min for males and 150/min for females

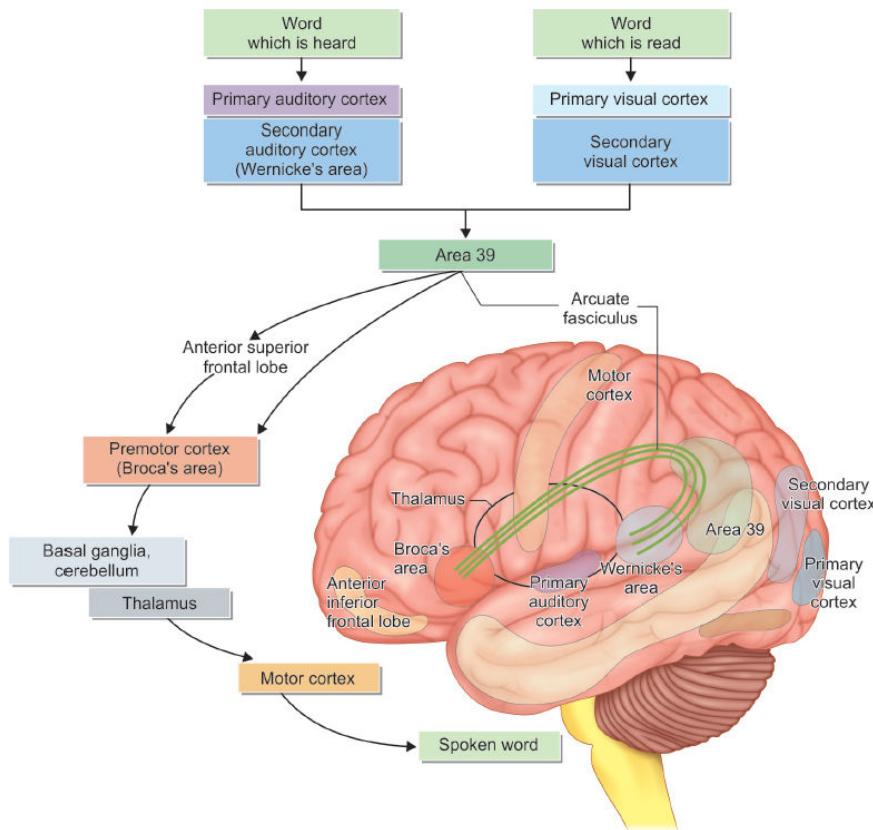


Fig. 6D(ii).2: Genesis of speech.

APHASIAS

- Aphasia is an acquired disorder with loss or defective language content of speech resulting from damage to the speech centers within the dominant (usually left in 97%) hemisphere.
- A language disturbance occurring after a right hemisphere lesion in a right hander is known as crossed aphasia.
- It includes defect in or loss of the power of expression by speech, writing, or gestures or a defect in or loss of the ability to comprehend spoken or written language or to interpret gestures.
- Aphasia may be categorized according to whether the speech output is fluent or nonfluent.
 - **Fluent aphasias** (receptive aphasias) are impairments mostly due to the input or reception of language with difficulties either in auditory verbal comprehension or in the repetition of words, phrases, or sentences spoken by others. For example, Wernicke's aphasia.
 - **Nonfluent aphasias** (expressive aphasias) are difficulties in articulating with relatively good auditory, verbal comprehension. For example, Broca's aphasia [Fig. 6D(ii).3].
- **Normal fluency** 100–150 words/min, sentence length >7 words.
- Reduced fluency in Broca's aphasia, transcortical motor, global aphasia, and primary progressive aphasia.

Domains of Language

1. Spontaneous speech/fluency
2. Comprehension
3. Repetition

4. Reading
5. Writing
6. Naming.

C—Comprehension (requires intact Wernicke's and transcortical sensory area)

R—Repetition (requires intact Wernicke's, arcuate fibers, and Broca's area)

F—Fluency (requires intact Broca's and transcortical motor area) [Flowchart 6D(ii).1].

	Aphasia	Site of lesion	C	R	F
1	Wernicke's—sensory/receptive/posterior	Infarction of inferior division of middle cerebral artery	—	—	+
2	Broca's—motor/expressive/anterior	Infarction of superior frontal branch of middle cerebral artery	+	—	—
3	Conduction/arcuate	Arcuate fasciculus	+	—	+
4	Transcortical sensory	Posterior watershed zone	—	+	+
5	Transcortical motor	Anterior watershed zone	+	+	—
6	Isolation aphasia (mixed transcortical aphasia)	Both anterior and posterior watershed areas	-	+	—
7	Global aphasia	Dominant frontal, parietal and superior temporal lobe	-	—	—

Note:

C—Comprehension

R—Repetition

F—Fluency

Once the comprehension, repetition, and fluency are intact, we look for reading, writing, and naming disorders associated with reading, writing, and naming.

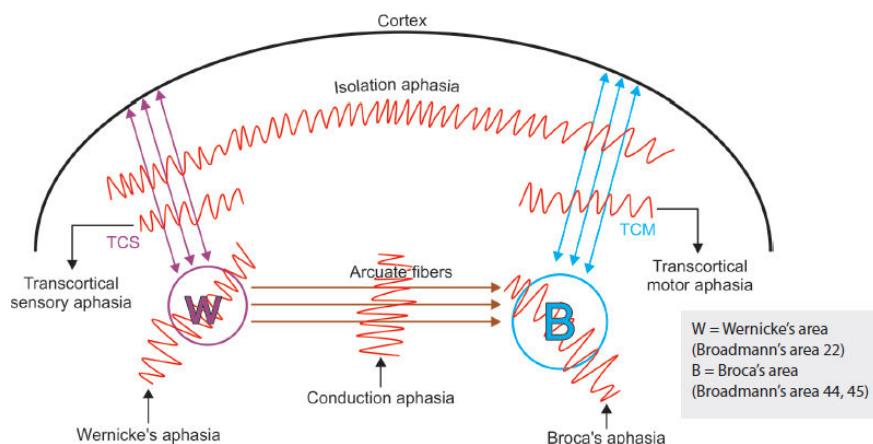
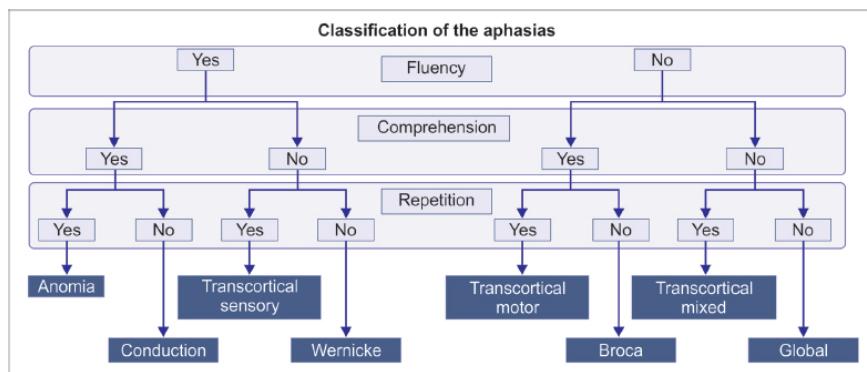


Fig. 6D(ii).3: Schematic representation of aphasias and associated lesions.

Flowchart 6D(ii).1: Approach for aphasias.



			R	W	N
8	Alexia without agraphia	Occipitotemporal region	-	+	+
9	Alexia with agraphia	Left angular gyrus	-	-	+
10	Nominal/anomic/amnesic	Temporoparietal	+	+	-

- Lesions in the anterior limb of internal capsule/basal ganglia can produce Broca's like aphasia.
- Lesions in the thalamus can produce Wernicke's like aphasia.
- Most common type of aphasia seen in stroke: Broca's aphasia.
- Overall most common type of aphasia is anomic aphasia.

DYSARTHRIAS

Production of sounds requires:

1. Normal respiration
2. Muscles of articulation (labial, lingual, and palatal muscles)
3. Phonation (by larynx)
4. Resonance (by nasopharynx).

Articulated Sounds

Articulated labials (b, p, m, and w) are formed principally by the lips.

Modified labials (o and u, and to a lesser extent i, e, and a) are altered by lip contraction.

Labiodentals (f and v) are formed by placing the teeth against the lower lip.

Linguals are sounds formed with tongue action.

T, d, l, r, and n are tongue point, or alveolar sounds formed by touching the tip of the tongue to the upper alveolar ridge.

S, z, sh, zh, ch, and j are dentals, or tongue blade sounds.

To hear distorted linguals, place the tip of your tongue against the back of your bottom teeth, hold it there and say "top dog," "go jump", and "train".

To hear distorted labials, hold your upper lip between the thumb and forefinger of one hand and your bottom lip similarly with the other and say "my baby".

Gutturals (velars, or tongue back sounds, such as k, g, and ng) are articulated between the back of the tongue and the soft palate.

Palatals (German ch and g, and the French gn) are formed when the dorsum of the tongue approximates the hard palate.

Types of dysarthrias		
Types	Description	Cause
Flaccid (lingual, buccal, and guttural)	LMN weakness of facial, lingual, or pharyngeal muscles. <ul style="list-style-type: none">• Facial paralysis causes difficulty with labials, such as b, p, m, and w.• Tongue paralysis affects a large number of sounds, particularly l, d, n, s, t, and x.• Palatal paralysis produces a nasal twang in speech.	Cerebrovascular accidents (especially brainstem lesions)
Spastic (hot potato voice)	Strained, slurred hot potato-like voice	UMN weakness (bilateral), e.g. pseudobulbar palsy
Ataxic speech	Scanning speech: Undue separation of syllables (monosyllable speech)	Cerebellar diseases
	Staccato speech: Explosive type of speech with emphasis on syllables	
Hypokinetic	Slow monotonous, low voice with inappropriate silence	Extrapyramidal (parkinsonism)
Hyperkinetic dysarthria	Distorted speech with continuous change in articulation	Chorea, athetosis, and dyskinesias
Myasthenic dysarthria	Voice is normal in the beginning but becomes weak as sentences progress	Myasthenia gravis

APRAXIA

Definition

Apraxia is impaired ability (inability) to carry out (perform) skilled, complex, and organized motor activities in the presence of normal basic motor, sensory, and cerebellar functions.

Examples of complex motor activities: Dressing, using cutlery, and geographical orientation.

Types	
Ideomotor apraxia	Most common. It is the inability to perform a specific motor command/act (e.g. cough, lighting a cigarette with a matchstick) in the absence of motor weakness, incoordination, and sensory loss or aphasia. Site of lesion is bilateral parietal lobe. Buccofacial apraxia involves apraxic deficits in movements of the face and mouth. Limb apraxia encompasses apraxic deficits in movements of the arms and legs
Dressing apraxia	Site of lesion is nondominant parietal lobe. It is inability to wear his/her dress
Constructional apraxia	It is inability to copy simple diagrams or build simple blocks. Site of lesion is nondominant parietal lobe
Ideational apraxia	It is a deficit in the execution of a goal-directed sequence of movements even with real object (e.g. asked to pick up a pen and write, the sequence of uncapping the pen, and placing the cap at the opposite end). This is commonly associated with confusion and dementia rather than focal lesions associated with aphasic conditions
Gait apraxia (Bruns ataxia)	Seen in normal pressure hydrocephalus (NPH)
Gaze apraxia	Part of Balint syndrome
Other apraxias	Speech apraxia, conceptual apraxia, and conduction apraxia

AGNOSIA

Definition

Agnosia is failure to recognize objects (e.g. places, clothing, persons, sounds, shapes, or smells), despite the presence of intact sensory system.

Site of lesion: Contralateral parietal lobe.

Types of agnosias	
Visual agnosia	Failure to recognize what is seen with eyes despite the presence of intact visual pathways. The individual can describe the shape, color, and size without naming it. Site of lesion is in the posterior occipital or temporal lobes
Prosopagnosia	A type of visual agnosia in which patient cannot identify familiar faces, sometimes the reflection of his or her own face in the mirror even including their own. Site of lesion is parieto-occipital lobe
Simultanagnosia	It is inability to perceive more than one object at a time
Autotopagnosia	It is a form of agnosia, characterized by an inability to localize and orient different parts of the body
Pseudopolymelia	The feeling of false—the feeling of false extremities. More frequent, the patients feel the extremities. More frequent, the patients feel the third hand
Anosognosia	It is an inability or refusal to recognize a defect or disorder that is clinically evident
Auditory agnosia	It consists of the loss of ability to know objects or sounds characteristic for them (clock—on ticking)

DELUSIONS

Definition

Delusion is a belief held with strong conviction despite superior evidence to the contrary (strongly held false beliefs).

It is a disorder of content of thought.

Types of delusion (based on their content)	
Persecutory delusions	Conviction that others are out to get me
Grandiose delusions	Belief that one has special powers or status
Nihilistic delusions	Conviction that "my head is missing/rotting", "I have no body", and "I am dead"
Erotomanic delusions	Believing a movie star loves them
Somatic delusions	Believing head is filled with air/worms
Delusions of reference	Believing story in a book is referring to them
Delusions of control/passivity	Believing one's thoughts and movements are controlled by aliens
Other delusions are	Delusions of misinterpretation, hypochondrial delusions, fantastic/bizarre delusions, delusions of passivity, delusions of jealousy

HALLUCINATIONS

Definition

Hallucinations are perceptions without external stimuli (**wakeful sensory experiences of content that is not actually present**). They can occur in any sensory modality, most common being **visual or auditory**.

For example, hearing voices when no one else is present, or seeing “visions”. Other types include tactile (cocaine bug), olfactory, gustatory, command kinesthetic/psychomotor, and lilliputian and complex hallucinations.

Pseudohallucinations

These are hallucinations that are perceived as originating in the external world, not in the patient's own mind.

Hypnagogic and Hypnopompic Hallucinations

In narcolepsy 2, specific hallucinations are seen. **Hypnagogic:** They occur when falling asleep. **Hypnopompic:** They occur on waking up from sleep.

(mnemonic—hypno**G**Otic hallucinations are perceived while **GO**ing to sleep).

Hallucinations	Illusions
Perceptions without external stimuli	Misperceptions of real external stimuli
For example, hallucinating that someone is talking to them when there is no actual stimulus	For example, mistaking a rope for snake

Functions and effects of damage to various lobes of cerebral hemispheres are listed in **Table 6D(ii).1** and **Figure 6D(ii).4**.

Table 6D(ii).1: Functions and effects of damage to various lobes of cerebral hemispheres.		
Lobe	Function	Cognitive/behavioral effects of damage
Frontal Please SMILE (MNEMONIC)	Personality	
	Social behavior	Antisocial behavior
	Micturition	Incontinence
	Intelligence	
	Language	Expressive dysphasia
	Emotional response	Disinhibition
Parietal: Dominant side	Language	Dysphasia, dyslexia
	Calculation	Acalculia
	Others	Apraxia, agnosia
Parietal: Nondominant side	Spatial orientation	Spatial disorientation, neglect of contralateral side
	Constructional skills	Constructional apraxia, dressing apraxia
Temporal: Dominant side	Auditory perception	Receptive aphasia
	Language	Dyslexia
	Verbal memory	Impaired verbal memory
	Smell	

	Balance	
Temporal: Nondominant side	Auditory perception	Impaired nonverbal memory
	Melody/pitch perception	Impaired musical skills (tonal perception)
	Nonverbal memory	
	Smell	
	Balance	
Occipital	Visual processing	Visual inattention, visual loss, visual agnosia (Anton–Babinski syndrome)

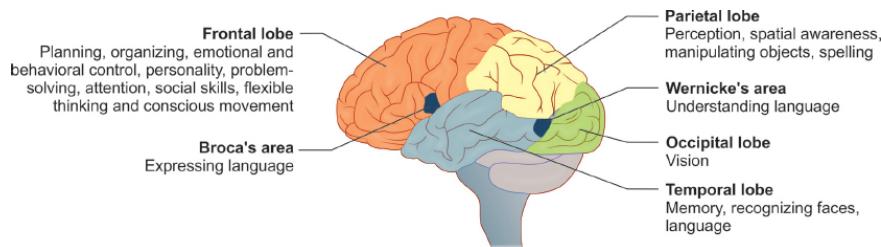


Fig. 6D(ii).4: Various lobes of cerebral hemispheres.

LESIONS OF NONDOMINANT (RIGHT) HEMISPHERE

Neglect

Definition → directed inattention, or a relative lack of attention, paid to one hemisphere; patients are less aware (or completely unaware) of objections or actions in one side of the world (usually the left).

Diagnosis

- *Severe forms* → patients completely ignore left side, denying that, such as side even exists; they may leave their left side ungroomed, unshaven, and undressed; may leave food on left side of plate uneaten; may deny they have a left hand, and when confronted with it, may claim that it is actually the examiner's.
- *Milder forms* → may perform actions with their left side only with encouragement or after repeated prodding.
- *Most sensitive sign* → extinction to double simultaneous stimulation; sensory stimuli applied singly to either side are properly felt, but when both sides are stimulated simultaneously, only the non-neglected side is felt; extinction may exist with tactile, visual, or auditory stimulation.
- *Etiology* → lesions in right hemisphere (frontal or parietal lobe), most commonly an acute finding after stroke.
 - *Frontal lobe* lesion → more of a motor neglect in which patient has tendency to not use left side for motor actions
 - *Parietal lobe* lesion → more of a sensory neglect in which stimuli from the left side tend to be ignored.

Others

- **Prosody** → while semantic elements of language (pure meaning) reside in dominant hemisphere, some other elements of successful oral communication (e.g. proper voice inflection) reside in nondominant hemisphere
- **Anosognosia** → tendency to be unaware of one's deficits in some patient's w/right hemispheric lesions
 - For example, patient with complete left hemiplegia may insist on immediate discharge from hospital because he feels nothing is wrong
 - For example, patient with dense left hemianopia may wonder why she keeps bumping into others since she notices nothing wrong with her vision.

NOTES

D(iii). CRANIAL NERVES

CRANIAL NERVE I—OLFACTOORY NERVE

Prerequisites for Examination

- Rule out nose blocks
- Close eyes while examining
- Test each nostril separately.

Substances Which Can be Used for Testing

- Peppermint
- Soap
- Coffee beans
- Lemon peel
- Vanilla.

Note: Avoid irritants like ammonia as they directly stimulate the trigeminal nerve endings.

Method of Examination

- Examine each nostril separately while occluding the other [Fig. 6D(iii).1].
- With the patient's eyes closed and one nostril occluded, bring the test substance near the open one.
- Instruct the patient to sniff repetitively and to tell you when an odor is detected, identifying the odor, if recognized.
- Bring the test odor up to within 30 cm or less of the nose.
- Repeat for the other nostril and compare the two sides.

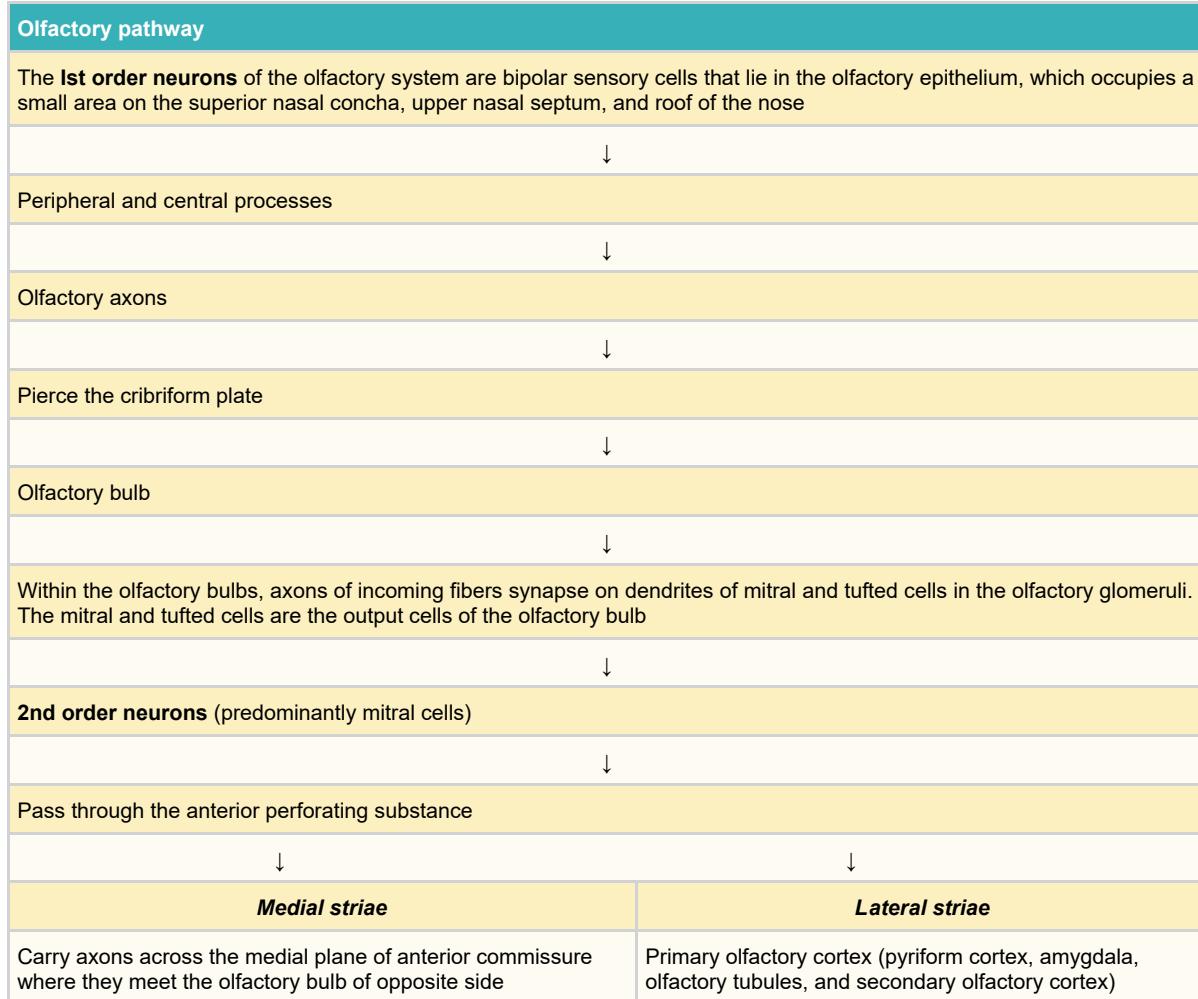
Note: The side that might be abnormal should be examined first.



Fig. 6D(iii).1: Method of examination of olfactory nerve.

Interpretation

- Patient able to detect smell, recognize, and name
- Patient able to detect smell, recognize but not name
- Patient able to detect, but not recognize or name.



Note:

- The olfactory nerves are the **unmyelinated filaments** that pass through the cribriform plate.
- The bulbs and tracts are part of the rhinencephalon.

Disturbances in olfaction
Anosmia
Local causes:
<ul style="list-style-type: none"> • Acute rhinitis (most common cause) • Heavy smoking • Atrophy of bulb
Systemic causes:
<ul style="list-style-type: none"> • Parkinsonism • Meningitis • Head trauma • Intracranial tumors • Endocrine diseases:

- Diabetes mellitus
- Hypothyroidism
- Kallmann syndrome
- Turner syndrome
- Vitamin B₁₂ deficiency
- Chronic kidney diseases.
- Refsum's disease

Syndromes associated:

- **Foster-Kennedy syndrome** (anosmia, optic atrophy of one eye, and contralateral eye papilledema due to tumor in brain)
- **Pseudo-Foster-Kennedy syndrome** (above features in absence of tumor)

Impaired smell

K: Korsakoff

B: Basilar meningitis

C: Chorea huntington's

A: Anterior cerebral artery diseases

S: Spinocerebellar ataxia

H: Hydrocephalus

Other miscellaneous points

- Anosmia is commonly associated with hypogeusia/ageusia
- Olfactory hallucinations: Usually of unpleasant odors like burned rubber, can occur in temporal lobe epilepsy, migraine and schizophrenia
- Hyperosmia: May be seen with Addison's disease, cystic fibrosis or pituitary tumors
- Merciful anosmia—atrophic rhinitis.

Note:

- Olfactory is the only nerve which does not process through thalamus.
- Olfactory and optic are the two nerves which do not pass through brainstem.
- Loss of smell is usually associated with loss of taste sensation (Aguesia/hypogeusia).

CRANIAL NERVE II—OPTIC NERVE

1. Visual acuity
2. Visual field
3. Color vision
4. Fundus examination.

Visual Acuity

Assessment of visual acuity is usually done by asking the patient to read the specific charts as described below. The least possible distance with best vision is considered as the viewing distance.

Visual acuity	
For far vision	For near vision
Snellen chart [Fig. 6D(iii).2]	Jaeger chart [Fig. 6D(iii).3]
Examined at 6 m	Examined at 30 cm
Described as x/y → x (numerator—suggests the viewing distance of patient) and y (denominator—viewing distance of normal person)	<ul style="list-style-type: none"> • Describes as J₁, J₂, etc. • Normal range of near vision is J₁ to J₄

Note: In absence of Snellen's chart finger counting can be done.

Defects in visual acuity may be due to:

- Refractive errors
- Cataract
- Vitreous opacity, etc.



Fig. 6D(iii).2: Snellen's chart for far vision.

0.37 M	I walked up the cheapness I bade her give me three sort, he gave me three puffy rolls. any worth of any sort, he gave me three puffy rolls. I was sort, he gave me three puffy rolls. I took it, and walked off with a rod under each arm of with a rod under each arm up Market Street as far as Fourth Street, passing by the house.	J2
0.50 M	the difference of money and the greater cheapness I bade him give me three penny worth of any sort, he gave me three puffy rolls. I was surprised at the quantity but I took it, and walked off with a rod under each arm. Thus I walked up Market Street as far as Fourth Street, passing by the house.	J3
0.62 M	of Mr. Read, my future wife's father. She, standing at the door, saw me and thought I made a most awkward appearance, as I certainly did. Then I turned and went down Chestnut street and a part of Walnut Street. Being filled with one of my rolls, I gave the other two to a woman	J4
0.75 M	and her child. But this time the street hand many clean and well dressed people in it, all walking the same way. I joined them and was led into the great meeting house of the Quakers'. I sat down among them and after looking around a while and hearing nothing said.	J5
1.00 M	I fell fast asleep, this was the first house I was in, or slept in, in Philadelphia. Looking in the faces of people, I met a young man whose countenance I liked, and asked	J7
1.25 M	if he would tell me where a stranger could get lodging. "Here", and he, "is one place that entertains strangers."	J8

Fig. 6D(iii).3: Jaeger's chart for near vision.

Visual Field Testing

Confrontation Method

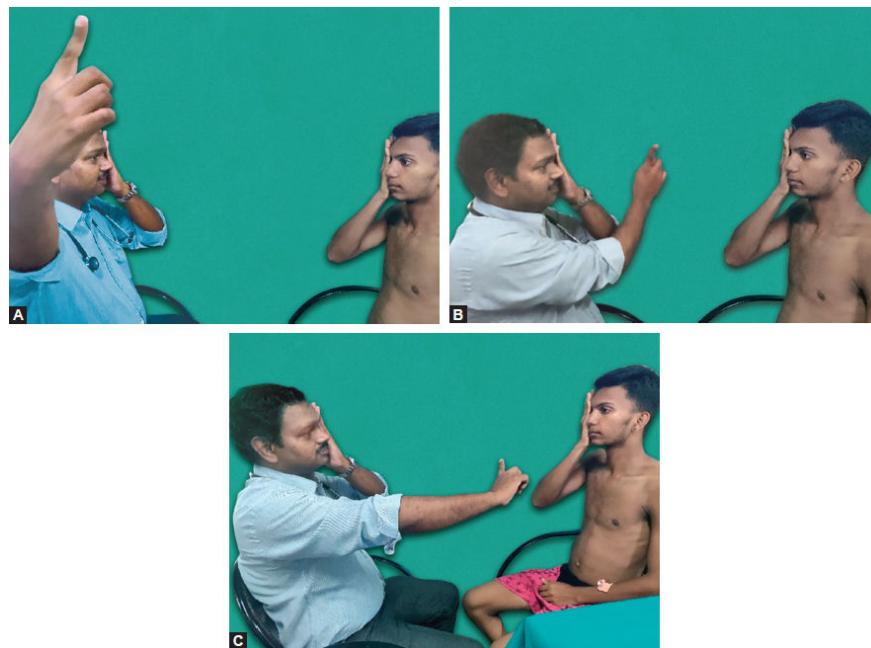
Testing distance: 1 m or one full hands distance [Figs. 6D(iii).4A to C]

Instructions:

- Subject and examiner should be sitting at the same height with each one looking into each other's eye separated by distance of 1 m.
- For checking the visual field of right eye of the subject, he is instructed to close his left eye with his left hand while the examiner closes his right eye with right hand. Now, the examiner brings in the flickering index finger of left hand from extremes of all four directions/quadrants diagonally toward the center of the visual field.
- The subject is instructed to give the signal at the first instance of perceiving the flickering finger movement.
- Normal extent of visual field of individual eye:
 - Vertically up 60°
 - Vertically down 75°
 - Medially 60°
 - Laterally 100°
- Normal extent of visual field in binocular vision:
 - Horizontally = 200°
 - Vertically = 140°

Shortcomings of Confrontation Method

1. Field and defects [Figs. 6D(iii).5 and 6D(iii).6]:



Figs. 6D(iii).4A to C: Method of examination (confrontation method).

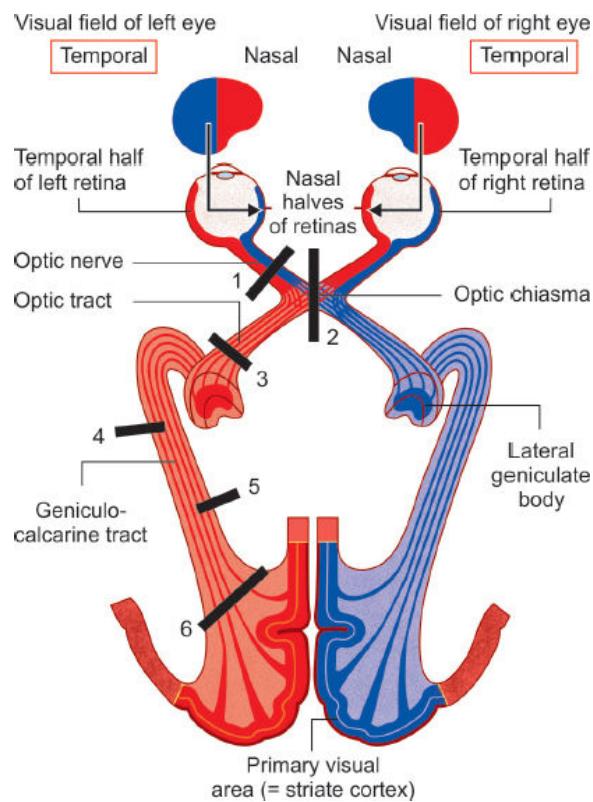


Fig. 6D(iii).5: Sites of lesions causing visual field defects.

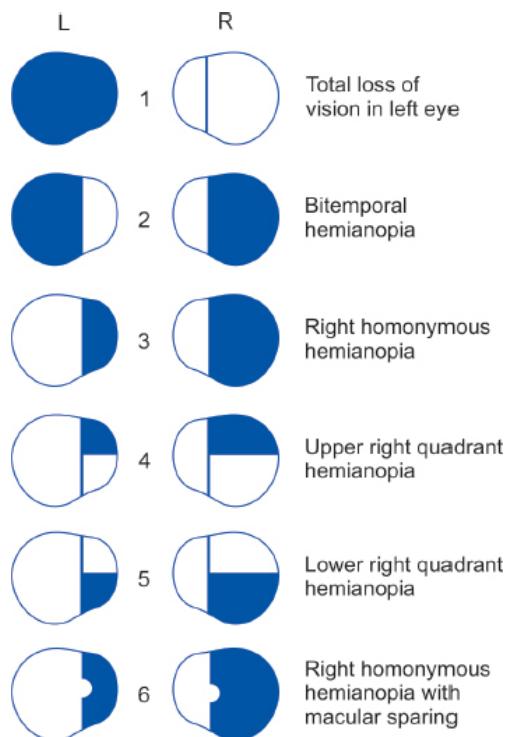


Fig. 6D(iii).6: Visual field defects.

Visual field defect		
	Site of lesion	Type of defect
1	Optic nerve	Total loss of vision in left eye
2	Optic chiasma	Bitemporal hemianopia
3	Optic tract	Right homonymous hemianopia
4	Geniculocalcarine tract	Upper right quadrantanopia
5	Geniculocalcarine tract	Lower right quadrantanopia
6	Macula	Right homonymous hemianopia with macular sparing

Note : Visual field defect produced by papilledema—enlarged blind spot

Color Vision (Red/Green/Blue)

Chart used: Ishihara chart [Figs. 6D(iii).7 and 6D(iii).8]

Congenital anomalies:

- **Red and green** = chromosome X (mnemonic: remember Red, Green and Symbol X all are traffic symbols)
- **Blue** = chromosome 7 (mnemonic: remember sky is **blue** which has rainbow containing **7** colors).

Acquired defects: Color vision occur in macular and optic nerve diseases, and due to certain drugs (e.g. ethambutol, chloroquine, digitalis, and sildenafil).



Fig. 6D(iii).7: Method of examining color vision.

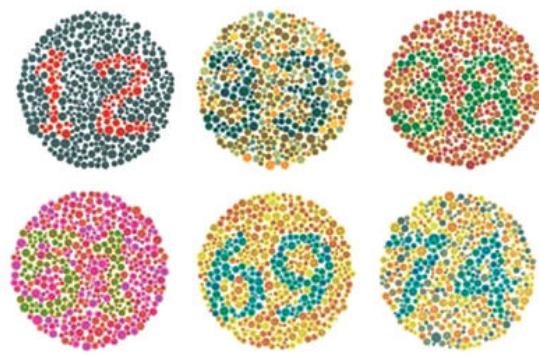


Fig. 6D(iii).8: Ishihara chart for color vision.

Fundus Examination

Instrument used: Direct ophthalmoscope.

How to use:

- The subject should be examined in sitting or lying down position.
- Examination room should be semidark.
- Keep the eye as still as possible.
- Hold ophthalmoscope in same hand as eye you are looking at, and looking through (e.g. hold ophthalmoscope in the left hand for examining patients left eye, through your left eye) [Figs. 6D(iii).9 and 6D(iii).10].
- Hold head steady with thumb above eyebrow, or hold shoulder.
- At about 30 cm distance with light on eye, locate red reflex (seen as an orange glow in the pupil).
- Follow red reflex into the eye as this will get you directly into the optic disc.
- If you cannot find the disc, trace any blood vessels back to it.
- Examine vessels in all four quadrants of eye (upper and lower, nasal and temporal quadrants).
- Identify macula—slightly darker pigmented area, two optic disc widths lateral away from the optic disc.



Fig. 6D(iii).9: Fundus examination of right eye.



Fig. 6D(iii).10: Fundus examination of left eye.

Look for optic atrophy and papilledema.

Also watch for feature of retinopathy like hemorrhages, exudates, cotton wool spots, and arteriolar changes.

Fundoscopic Finding

Papilledema is a disease entity which refers to the swelling of the optic disc due to elevated intracranial pressure (ICP) [Fig. 6D(iii).11].

Grade	Description
1	Disruption of the normal radial arrangement of nerve fiber bundles with a blurring of the nasal border of the optic disc and normal temporal margin
2	Nasal and temporal (circumferential) blurring of the optic disc with more pronounced changes from grade 1
3	The elevated and blurred disc margin borders obscure one or more major retinal vessel segments
4	More pronounced changes than from grade 3 and with total obscuration of a segment of the central retinal artery or vein
5	More pronounced changes than from grade 4 and with total obscuration of all disc vessels



Fig. 6D(iii).11: Papilledema.

Causes of Papilledema

Space-occupying lesions:

- Intracranial mass
- Abscess
- Hemorrhage
- Arteriovenous malformation

Focal or diffuse cerebral edema:

- Trauma
- Toxic
- Anoxia

Blockage of CSF flow: Noncommunicating hydrocephalus

Reduction in CSF reabsorption:

- Meningitis
- Elevated cerebral venous sinus pressure
- Elevated CSF protein—Guillain–Barré syndrome

Pseudotumor cerebri

Systemic causes:

- Hypercarbia
- Hypertension
- Hypercalcemia
- Hypoparathyroidism.

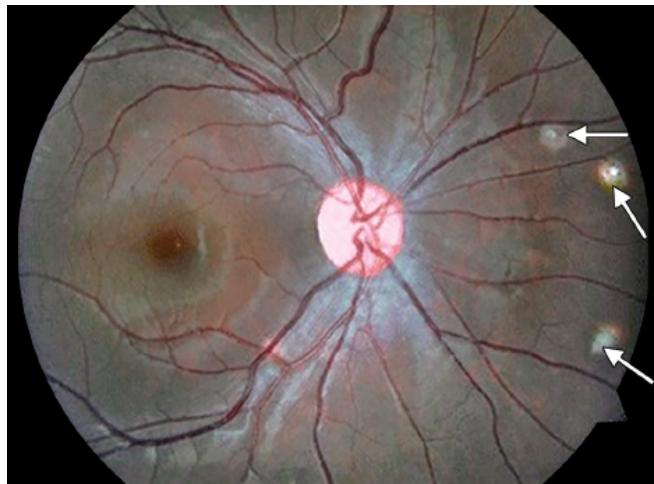


Fig. 6D(iii).12: Choroid tubercles in tuberculosis.

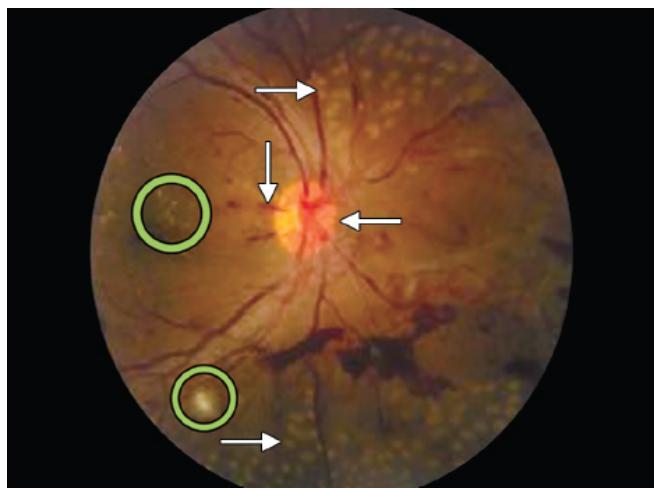


Fig. 6D(iii).13: Proliferative diabetic retinopathy with panretinal photocoagulation.

STAGES OF HYPERTENSIVE RETINOPATHY [FIGS. 6D(III).14 TO 6D(III).17]

Keith-Wagener-Barker Classification

- Group 1: Slight constriction of retinal arterioles
- Group 2: Group 1 + focal narrowing of retinal arterioles + AV nicking
- Group 3: Group 2 + flame-shaped hemorrhages + cotton-wool spots + hard exudates and copper wiring
- Group 4: Group 3 + optic disc swelling and silver wiring.

STAGES OF DIABETIC RETINOPATHY

Nonproliferative Diabetic Retinopathy

Very mild: Microaneurysms only.

Mild

Any or all of: Microaneurysms, retinal hemorrhages, cotton wool spots.

Moderate

- Severe retinal hemorrhages in 1–3 quadrants or mild IRMA
- Significant venous beading in no more than 1 quadrant
- Cotton wool spots.

Severe

The 4-2-1 rule:

- Severe retinal hemorrhages in all 4 quadrants
- Significant venous beading in ≥ 2 quadrants
- Moderate IRMA in ≥ 1 quadrants.

Very severe: ≥ 2 of the criteria for severe.

Proliferative Diabetic Retinopathy [Fig. 6D(iii).13]

Mild-moderate

- New vessels on the disc (NVD) < 1/3 disc area
- New vessels elsewhere (NVE) <1/2 disc area.

High-risk

- NVD >1/3 disc area
- Any NVD with vitreous or preretinal hemorrhage
- NVE >1/2 disc area with vitreous or preretinal hemorrhage.

Advanced diabetic eye disease

- Preretinal (retrohyaloid) and/or intragel hemorrhage
- Tractional retinal detachment
- Tractional retinoschisis
- Rubeosis iridis (iris neovascularization).



Fig. 6D(iii).14: Focal arteriolar narrowing.

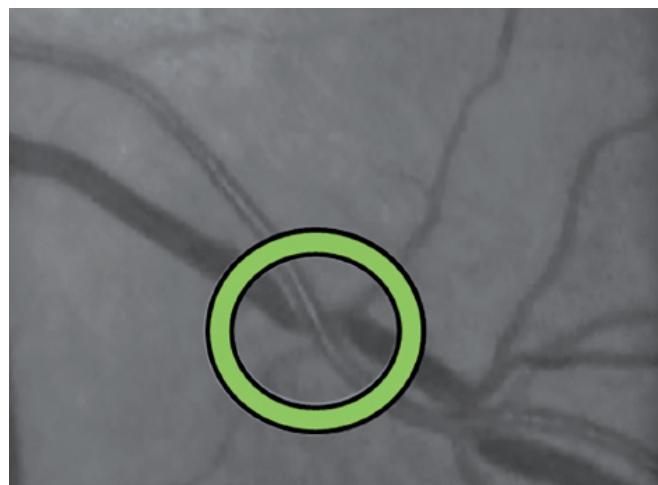


Fig. 6D(iii).15: AV nipping.

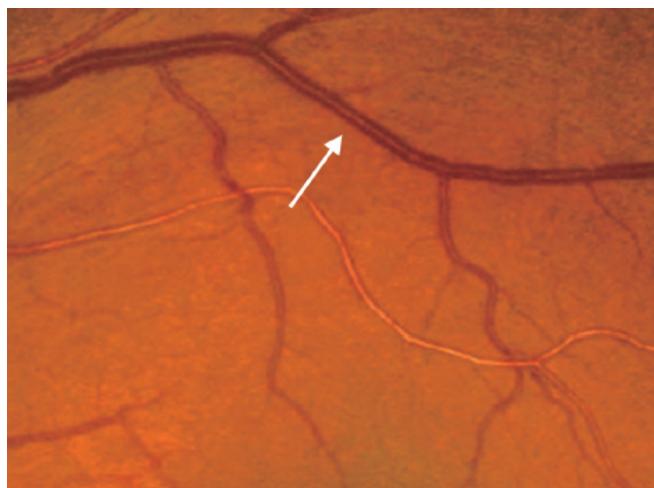


Fig. 6D(iii).16: Copper wiring.

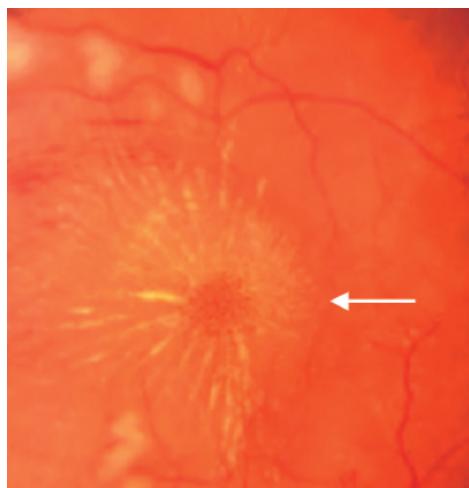


Fig. 6D(iii).17: Cotton wool spots and exudates forming macular star.

Background diabetic retinopathy (bdr)

- It is the earliest phase of diabetic retinopathy (DR).
- Characterized by microaneurysms, dot and blot hemorrhages and exudates.

Diabetic maculopathy: Refers to presence of any retinopathy at the macula.

Preproliferative diabetic retinopathy (PPDR): Cotton wool spots, venous changes, intraretinal microvascular abnormality (IRMA) and deep retinal hemorrhages.

Diabetic papillopathy: It is a form of optic neuropathy seen in young type I diabetics. It is unrelated to glycemic control or any other known feature of diabetes.

CAUSES OF OPTIC ATROPHY

1. Inflammation
2. Ischemia

3. Compression, including raised ICP
4. Nutritional deficiencies/effect of toxins
5. Trauma
6. Hereditary conditions and childhood optic atrophy.

CRANIAL NERVES III, IV AND VI—OCULOMOTOR, TROCHLEAR AND ABDUCENS

Anatomy:

Nuclei	Location	Additional points
III	Upper midbrain	<ul style="list-style-type: none"> • Four paired nuclei (SR, IR, MR, and IO muscles) • One unpaired nuclei (LPS muscles of both sides)
IV	Midbrain	At level of inferior colliculus (SO muscle)
VI	Mid to lower pons	LR muscle

(SR: superior rectus; IR: inferior rectus; SO: superior oblique; IO: inferior oblique; MR: medial rectus; LR: lateral rectus; LPS: levator palpebrae superioris)

Examined under following headings:

1. Eyelids
2. Eyeballs at rest
3. Extraocular muscles
4. Pupils
5. Nystagmus.

Eyelids

Ptosis: The narrowing of the palpebral fissures due to inability to open an upper eyelid is called ptosis.

Ptosis can be due to	
↓	↓
Paralysis of levator palpebrae superioris (LPS)	Paralysis of tarsal muscle
LPS supplied by III cranial nerve	Tarsal muscle supplied by sympathetic system
LPS is paralyzed and the patient cannot voluntarily rise the eyelid, he compensates by contracting frontalis muscle and thus there is wrinkling of forehead seen in long-standing cases	Here since the III nerve is intact and LPS is not paralyzed, ptosis disappears on voluntary contraction of LPS

Cause of ptosis

1. Congenital ptosis	
2. Acquired ptosis	
Neurogenic	<ul style="list-style-type: none"> • Horner's syndrome • III nerve palsy
Neuromuscular disorder	<ul style="list-style-type: none"> • Myasthenia gravis (fatigable ptosis)

	<ul style="list-style-type: none"> • Poisoning (snake bite/botulism)
Myogenic	<ul style="list-style-type: none"> • Mitochondrial myopathy • Oculopharyngeal muscle dystrophy • Myotonic dystrophy
Mechanical ptosis	Due to eyelid edema or tumors

Unilateral and bilateral ptosis

<i>Unilateral ptosis</i>	<i>Bilateral ptosis</i>
<ul style="list-style-type: none"> • Third cranial nerve lesion • Lesion of cervical sympathetic pathway (Horner's syndrome) • Lesions of the upper eyelid 	<ul style="list-style-type: none"> • Myopathies • Myasthenia gravis • Bilateral Horner's syndrome • Snake bite • Botulism



Fig. 6D(iii).18: Neurogenic ptosis.



Fig. 6D(iii).19: Mechanical ptosis secondary to edema.

Ptosis and pupil size

Ptosis with	
Small pupil	Horner's syndrome
Large pupil	IIIrd nerve palsy (compressive lesions)
Normal pupillary size	Infarction of IIIrd nerve, myasthenia gravis, myopathies or Guillain–Barré syndrome

Lid retraction:

- Lid is buried under the brow
- Sclera clearly visible above iris
- Example—hyperthyroidism, large doses of anticholinesterases
- Collier's sign: Seen in Parinaud's syndrome. Produces retraction nystagmus.

Reversible ptosis: Myasthenia gravis—**ice pack test [Figs. 6D(iii).20A to C]**

- The ice pack test is cheap, safe, and very quick to perform as it can be carried out at the bedside in approximately 3–5 minutes
- Positive test is the improvement of ptosis by >2 mm or more. This transient improvement in ptosis is due to the **cold** decreasing the acetylcholinesterase breakdown of acetylcholine at the neuromuscular junction.

Position of Eyeballs at Rest

Exophthalmos:

- Proptosis of eye
- Most commonly seen in hyperthyroidism

Unilateral exophthalmos:

- Carotid-cavernous fistula (pulsatile exophthalmos)
- Thyroid disorder—hyperthyroidism
- Orbital mass lesion
- Cavernous sinus thrombosis
- Sphenoid wing meningioma



Figs. 6D(iii).20A to C: Reversible ptosis (ice pack test).

- Meningocele
- Mucormycosis.

Enophthalmos: Enophthalmos can be defined as a relative, posterior displacement of a normal-sized globe in relation to the bony orbital margin. Causes are trauma, microphthalmia, post radiation, Horner's syndrome (apparent enophthalmos), Marfan syndrome, Duane's syndrome, or phthisis bulbi.

Extraocular Muscles

Functions of extraocular muscles [Fig. 6D(iii).21]:

	Primary function	Secondary function	Tertiary function
--	------------------	--------------------	-------------------

SR	Elevation	Intorsion	Adduction
IR	Depression	Extorsion	Adduction
SO	Intorsion	Depression	Abduction
IO	Extorsion	Elevation	Abduction
MR	Adduction		
LR	Abduction		

(SR: superior rectus; IR: inferior rectus; SO: superior oblique; IO: inferior oblique; MR: medial rectus; LR: lateral rectus)

Mnemonic: **S** in **R** ad

- All **S**uperiors are **I**ntortors
- All **R**ecti are **A**dductors except lateral rectus
- Function of **R**ecti is **r**egular (superior rectus is for elevation)
- Function of **O**blique is **o**pposite (superior oblique is for depression)
- In adducted eye—elevation is by inferior oblique and depression is by superior oblique
- In abducted eye—elevation is by superior rectus and depression is by inferior rectus.

Note: position of testing the muscle and actual action of the muscle usually is opposite with respect to horizontal gaze.

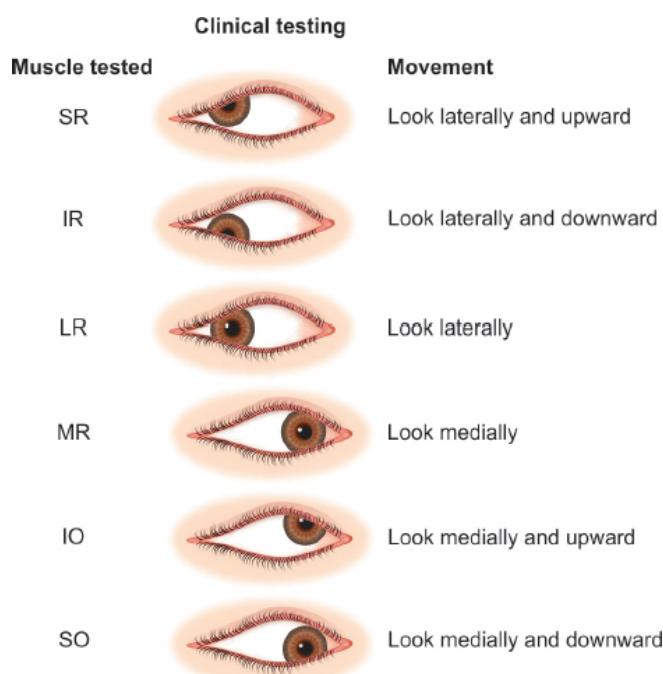


Fig. 6D(iii).21: Extraocular movements.

(SR: superior rectus; IR: inferior rectus; SO: superior oblique; IO: inferior oblique; MR: medial rectus; LR: lateral rectus)

Binocular Movements

Center for conjugate eye movements: Frontal eye field area number 8.

Saccades:

- Conjugate rapid eye movements
- Frontal lobe (premotor area number 6) controls saccadic movements.

Pursuits:

- Slow and smooth movement of eye following a moving target
- Occipital lobe is connected to the PPRF which is responsible for the horizontal pursuit movements.

Reflexes:

- Dolls eye reflex (oculocephalic reflex)
- Caloric stimulation test (vestibuloocular reflex).

Uniocular Movements**Nerve involved and features**

Nerve involved	Clinical features
III cranial nerve	<ul style="list-style-type: none"> • Down and out eye • Divergent squint • Ptosis • Dilatation of pupil
IV cranial nerve	<ul style="list-style-type: none"> • Defective downward eye movement • Outward rotation of eyeball by unopposed action of inferior rectus • Compensated by head tilt to opposite side
VI cranial nerve	<ul style="list-style-type: none"> • Defective lateral gaze • Medial squint • Patient may have diplopia on lateral gaze • Compensated by head turn to same side

In the oculomotor nerve [Fig. 6D(iii).22], the parasympathetic fibers lying on the peripheral part have dual blood supply via vasa nervosum and vessels on the sheet. In compressive lesions from outside (tumor and hematoma), pupils are involved early.

In ischemic lesions, pupils are spared since the center of the nerve is affected early.

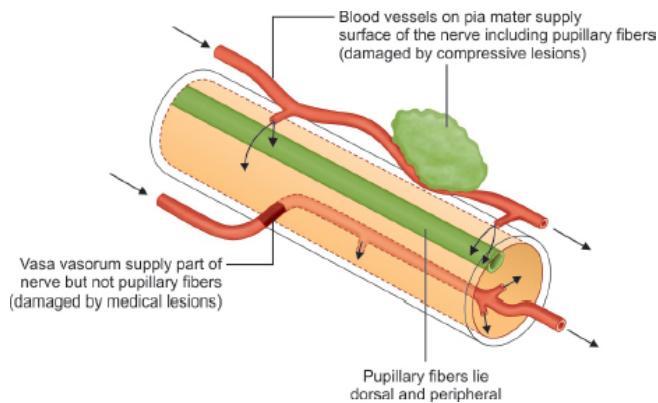


Fig. 6D(iii).22: Oculomotor nerve.

OCULAR MOVEMENT TESTING

Ask the patient to follow the examiner's finger or a red topped hat pin which is kept 60 cm away from the patient's face in all directions [Figs. 6D(iii).23 and 6D(iii).24].

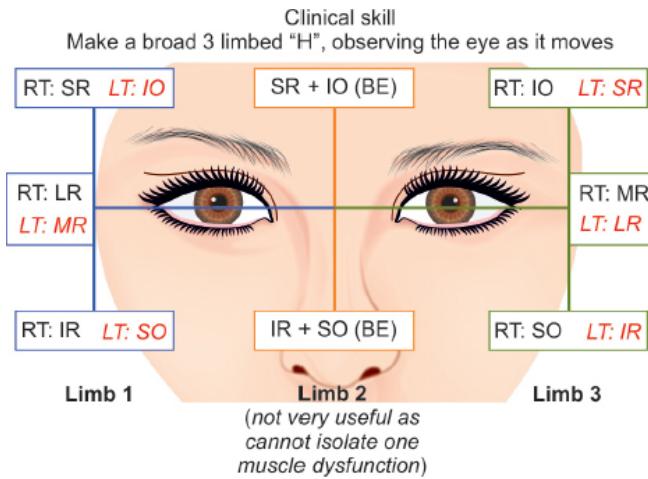


Fig. 6D(iii).23: Ocular movements testing method.

(RT: right; SR: superior rectus; IO: inferior oblique; LT: left; LR: lateral rectus; MR: medial rectus; IR: inferior rectus; SO: superior oblique; IO: inferior oblique)

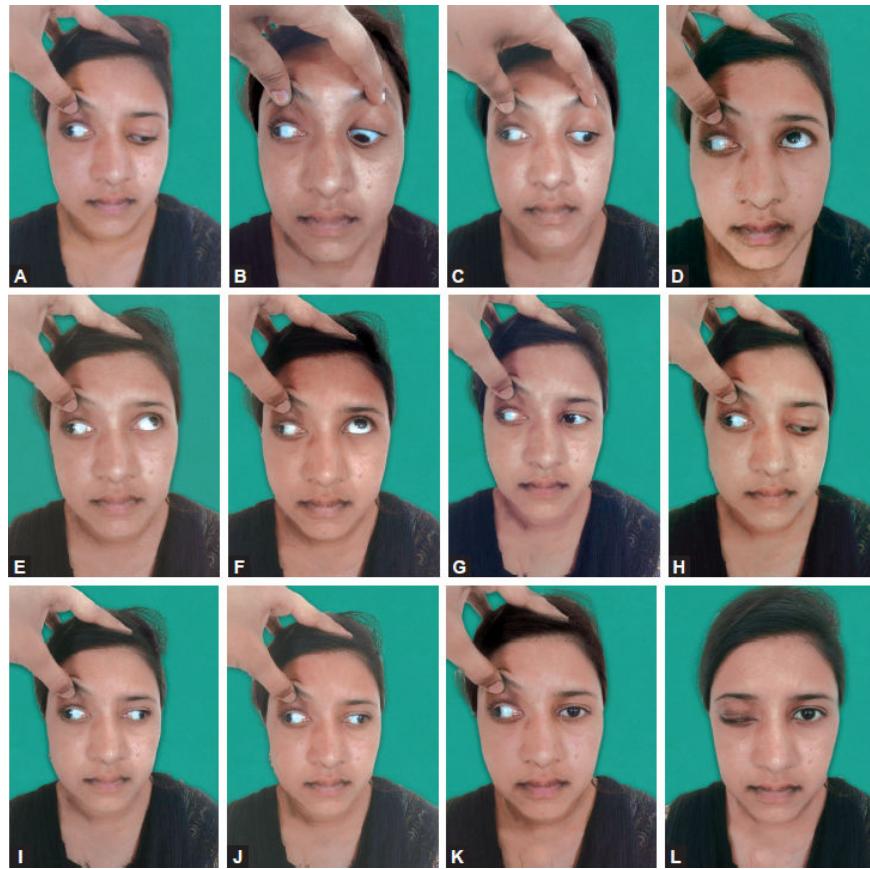
Etiology of III, IV, and VI nerve palsies		
	<i>Medical palsy</i>	<i>Surgical palsy</i>
III nerve ophthalmoplegia	Pupil sparing	Pupil involving
	Due to vascular causes where in the central part of nerve is involved (as visualized from the cut section)	Due to compression from the outside on the peripheral part of nerve (as visualized from the cut section)
	<ul style="list-style-type: none"> • Diabetes • Vasculitis • Myasthenia gravis • Myopathy 	<ul style="list-style-type: none"> • Posterior communicating aneurysm • Tumors of base of skull
	Nuclear lesion	
IV nerve palsy		
VI nerve palsy	<ul style="list-style-type: none"> • Pontine lesions • False localizing sign wherein raised ICT is the cause for palsy 	

DIPLOPIA

Diplopia means double vision. Most common subjective complaint elicited by lesions in the oculomotor system. Occurs more frequently with lesions of the extraocular muscles or oculomotor nerves than with supranuclear lesions which result in gaze palsies.

Monocular Diplopia

- The first point to clarify is whether diplopia persists in either eye after covering the fellow eye. If it does, the diagnosis is monocular diplopia.



Figs. 6D(iii).24A to L: Ocular movements testing in a patient with right complete ophthalmoplegia.

- The cause is usually intrinsic to the eye. For example, corneal aberrations, uncorrected refractive error, cataract, foveal traction or foreign body in the aqueous or vitreous may give rise to monocular diplopia.

Binocular Diplopia

Diplopia improved by covering one eye is binocular diplopia and is caused by disruption of ocular alignment. Occurs only if both eyes are open.

Binocular diplopia occurs from a wide range of processes: For example, infectious, neoplastic, metabolic, degenerative, inflammatory, and vascular.

Assessment of Diplopia

- Cover one of the patient's eyes with a transparent red shield. Move a point of light in the direction of action of each muscle.
- Ask the patient if he sees one object or two.
- If double, do the images lie side by side or one above the other?
 - Side by side—medial rectus (MR)/lateral rectus (LR)
 - One above the other—superior rectus (SR)/inferior rectus (IR) and superior oblique (SO)/inferior oblique (IO)
- Which is the red image?
- In which position the images are the farthest.

Points to note:

- In diplopia two images, one real and one false are formed. The real image is closer to the eye and distinct; the false image is farther away from eye and indistinct.
- Separation of images is maximum in the direction of action of weak muscle.

Muscle	Movement affected	Squint	Diplopia	
LR	Abduction	Convergent	Uncrossed	Maximum on looking laterally
MR	Adduction	Divergent	Crossed	Maximum on looking medially
SO	Downward movement in adduction	Convergent—in elevation and extorsion	Uncrossed	Maximum on looking down and medially
IO	Upward movement in adduction	Convergent—in depression and intorsion	Uncrossed	Maximum on looking up and medially
SR	Upward movement in abduction	Divergent—in depression and extorsion	Crossed	Maximum on looking up and laterally
IR	Downward movement in abduction	Divergent—in elevation and intorsion	Crossed	Maximum on looking down and laterally

(LR: lateral rectus; MR: medial rectus; SO: superior oblique; IO: inferior oblique; SR: superior rectus; IR: inferior rectus)

STRABISMUS/SQUINT

- Loss of parallelism of eyeball resulting in abnormal position of eyes.
- Primary deviation—deviation in the paralyzed eye
- Secondary deviation—deviation in the normal eye.

Types of Squint

Paralytic	Nonparalytic/concomitant
Secondary deviation > primary deviation	Secondary deviation = primary deviation
Acquired	<ul style="list-style-type: none"> • Usually congenital • Starts in childhood
Diplopia present	No diplopia
Ocular movements affected	Ocular movements are full in all directions

Pupils

Miosis and mydriasis

Large pupils	Small pupils
Unilateral: <ul style="list-style-type: none"> • Physiological • Pharmacological • Oculomotor nerve palsy • Adie's pupil • Uncal herniation • Traumatic sphincter paralysis • Iris ischemia • Ocular siderosis 	Unilateral: <ul style="list-style-type: none"> • Physiological • Horner's syndrome • Anterior uveitis • Long standing Adie's pupil • Pharmacological
Bilateral:	Bilateral:

<ul style="list-style-type: none"> • Pharmacological • Parinaud's dorsal midbrain syndrome • Benign periodic mydriasis • Brainstem death 	<ul style="list-style-type: none"> • Physiological senile miosis • Pharmacological • Argyll Robertson pupil • Lepromatous miosis • Congenital microcoria • Myotonic dystrophy
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Cranial nerve palsy	Examination findings—evidence of incomitance (i.e. angle of squint varies with position of gaze)		
	Direction of gaze	Primary position	Direction of gaze
Right 3rd nerve palsy	 Smaller angle of horizontal squint	 Right eye turns downwards and outwards	 Unable to adduct right eye Larger angle of squint Double vision further apart
Right 4th nerve palsy	 No obvious squint	 Right eye turns upwards	 Right eye elevates more as it moves medially Double vision further apart
Right 6th nerve palsy	 Unable to abduct right eye Larger angle of squint Double vision further apart	 Right eye turns medially	 Able to adduct right eye No obvious squint

Fig. 6D(iii).25: Cranial nerve 3, 4, and 6 palsy.

Light reflex

- Mediated by retinal photoreceptors.
 - Subserved by four neurons [**Fig. 6D(iii).26**]
1. First (sensory)—connects each retina with both pretectal nuclei, nasal fibers decussate, and temporal fibers uncrossed
 2. Second (internuncial)—connects each pretectal nucleus to both Edinger-Westphal nuclei —indirect reflex
 3. Third (preganglionic motor)—connects Edinger-Westphal nucleus to ciliary ganglion.
 - Parasympathetic fibers pass through III nerve inferior division and reach the ciliary ganglion via the nerve to the inferior oblique muscle.
 4. Fourth (postganglionic motor) leaves the ciliary ganglion and passes in the short ciliary nerves to innervate the sphincter pupillae.

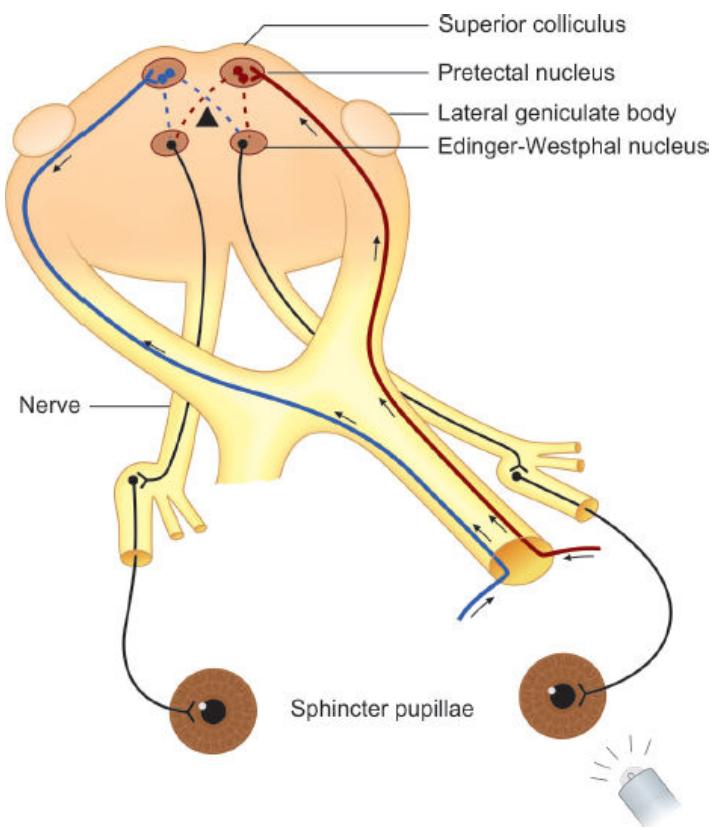


Fig. 6D(iii).26: Light reflex pathway.

- Tested in each eye individually
- Patient fixing at a distance
- Light shown to the eye obliquely.
- Cover uncover technique—uses ambient light
- Normal response: Brisk constriction—slight dilatation back to an intermediate state.
- Can be recorded: prompt, sluggish, and absent—graded 0–4+

The accommodation reflex

- Relax accommodation by gazing at a distant object.
- Shifting gaze to some near object.
- The primary stimulus for accommodation is blurring.
- Response: Accommodation, convergence, and miosis.

Pathway similar to light reflex till [Fig. 6D(iii).27]

- Fibers of Edinger-Westphal nucleus when entering the eye will cause constriction of the pupil and stimulation of ciliary muscle, so the parasympathetic causes the two changes (constriction of the pupil and contraction of ciliary muscle that increases the thickness of the lens thus increasing its power).
- The third change is convergence (adduction of both eyes by stimulating medial rectus on both sides); this is achieved by the Vergence center that affects the oculomotor nucleus in

the midbrain on this side and the other. Fibers coming from the oculomotor nucleus will enter and stimulate the medial rectus on both sides, when both eyes are adducted, the image will be on the same area (focus) of the retina.

PUPILLARY ABNORMALITIES

Argyll Robertson Pupil

- Small irregular pupil having light near dissociation [Fig. 6D(iii).28]
Characteristic feature:
 - In dim light, both pupils are small and may be irregular.
 - In bright light, neither pupil constricts.
 - On accommodation both pupils constrict (light near dissociation).
 - After instillation of pilocarpine 0.1% into both eyes, neither pupil constricts.
- Described for neurosyphilis.
- Lesion in periaqueductal region, pretectal, and rostral midbrain.

Other causes: Diabetes mellitus, chronic alcoholism, multiple sclerosis, and sarcoidosis.

Reverse Argyll Robertson Pupil

In this accommodation, reflex on the pupil is absent.

Cause: Diphtheria and tumors at corpora quadrigemina.

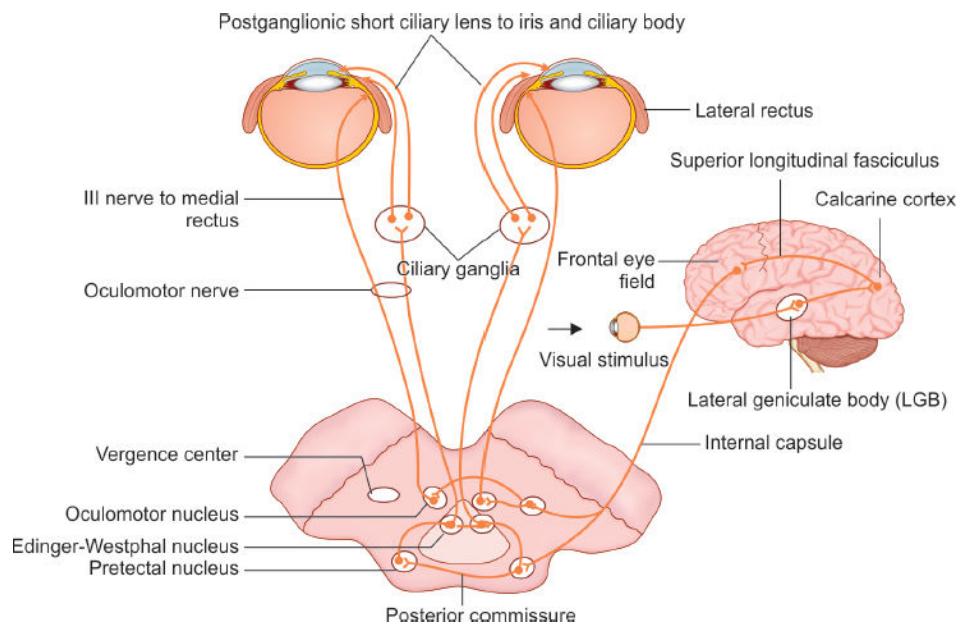


Fig. 6D(iii).27: Accommodation reflex pathway.

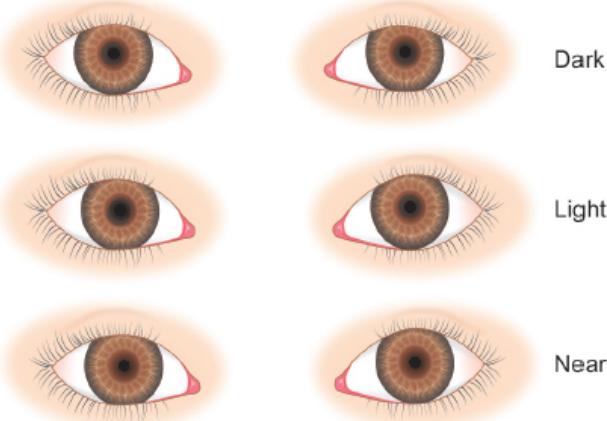


Fig. 6D(iii).28: Argyll Robertson pupil.

Wernicke's Hemianopic Pupil

- It indicates lesion of the optic tract.
- In this condition, light reflex (ipsilateral direct and contralateral consensual) is absent when light is thrown on the temporal half of the retina of the affected side and nasal half of the opposite side; while it is present when the light is thrown on the nasal half of the affected side and temporal half of the opposite side.

The Adie's Tonic Pupil

In this condition, reaction to light is absent and to near reflex is very slow and tonic.

- The affected pupil is larger (anisocoria).
- Its exact cause is not known.
- It is usually unilateral, associated with absent knee jerk and occurs more often in young women.
- Adie's pupil constricts with weak pilocarpine (0.125%) drops, while normal pupil does not.
- In long-standing cases, the pupil may become small ("little old Adie").
- In some cases, are diminished deep tendon reflexes (Holmes-Adie syndrome).

Afferent Pupillary Defect or Marcus Gunn Pupil

- The status of the light reflex must be judged by comparing the two eyes [Fig. 6D(iii).29]
- Indicator of optic nerve function
- Swinging flashlight test: Light is held about 1 inch from the eye and just below the visual axis; the light is rapidly alternated.
 - The examiner attends only to the stimulated eye.
 - Comparing the amplitude and velocity of the initial constriction in the two eyes.
- The reaction is relatively weaker when the bad eye is illuminated.

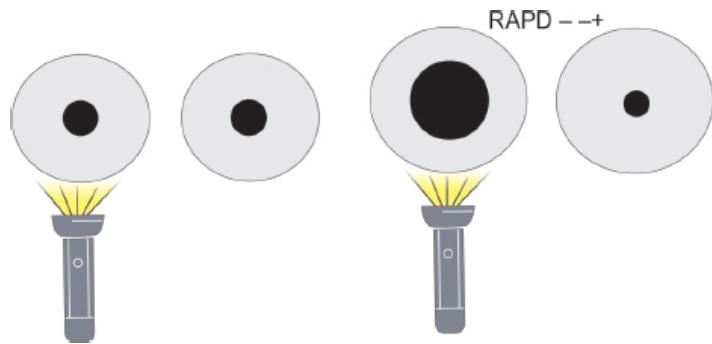


Fig. 6D(iii).29: Relative afferent pupillary defect (RAPD)/Marcus Gunn pupil.

- The brain detects a relative diminution in light intensity and the pupil may dilate a bit in response.
- Bring out the dynamic anisocoria.
- The weaker direct response or the paradoxical dilation of the light-stimulated pupil is termed as an afferent pupillary defect (APD).

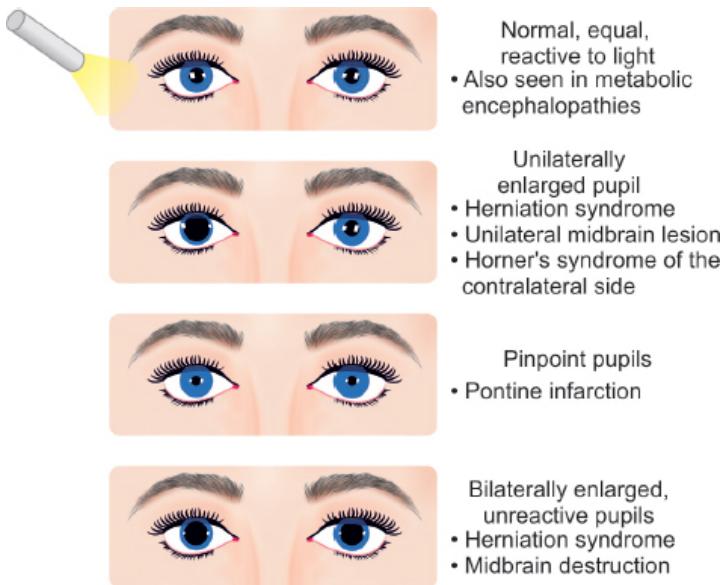


Fig. 6D(iii).30: Pupillary abnormalities in coma.

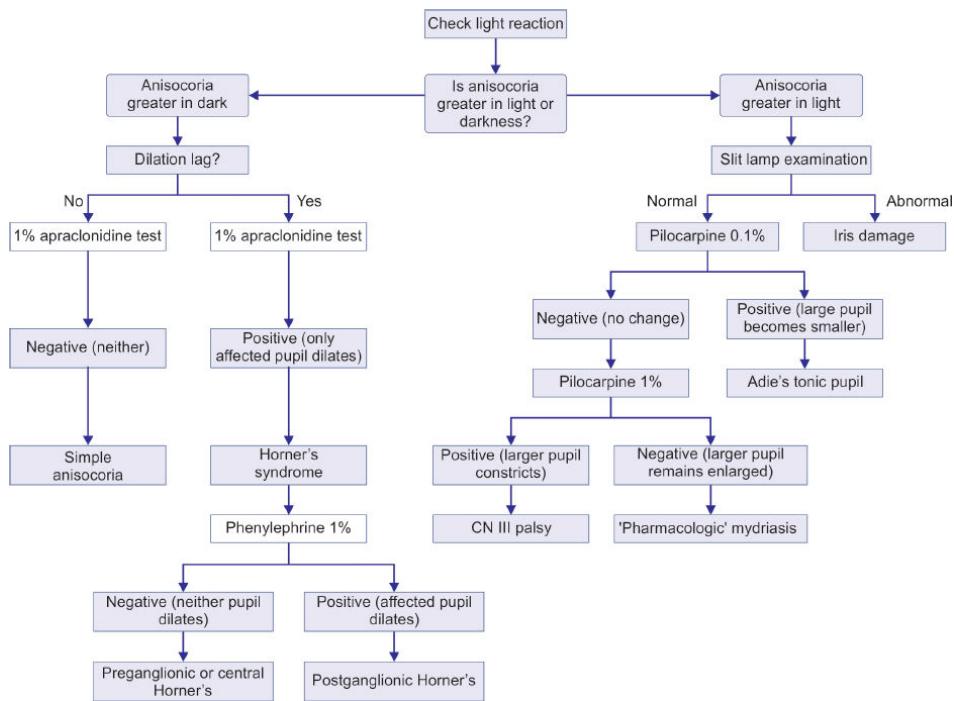


Fig. 6D(iii).31: Approach to pupillary abnormalities.

- Trace APD: Pupil that has an initial constriction, but then it escapes to a larger intermediate position than in the other eye.
- 1 to 2+ APD: No change in pupil size initially, then dilation.
- 3 to 4+ APD: Immediate dilation of the affected pupil.

Hutchinson's Pupil

- Seen in comatose patients
- Dilated poorly reactive pupil
- Due to expanding intracranial supratentorial mass causing uncal herniation and III nerve compression.

Hippus

- Irregular rhythmic visible pupillary oscillations 2 mm/more in amplitude irregular dilating and constricting movements are observed
- Also called as pupillary athetosis
- **Cause:** Myasthenia gravis.

Tectal Pupils

Large pupils with light near dissociation: Seen in lesions affecting the upper midbrain.

Horner's Syndrome: Oculosympathetic Palsy

- **Ptosis:** Denervation of Müller's muscles
- **Miosis:** Denervation of dilators
- **Enophthalmos:** Narrowing of palpebral fissure

- Anhydrosis: Sympathetic denervation
- Loss of ciliospinal reflex.

Mnemonic—Protein **MEAL** [Fig. 6D(iii).33].

Usually unilateral: The smooth muscle fibers of the lower eyelid retractors also lose their sympathetic supply in patients with Horner's syndrome and, thus, the lower eyelid appears slightly elevated. This appearance has been termed “**upside-down ptosis**” or “**reverse ptosis**”.

- Hypochromic heterochromia (iris of different color—Horner is lighter) may be seen if congenital or long-standing. Sympathetic innervation is thought to be required for the formation of melanin by stromal melanocytes.
- Reduced ipsilateral sweating if the lesion is below the superior cervical ganglion, because the sudomotor fibers supplying the skin of the face run along the external carotid artery.
- Horner's syndrome is usually characterized by “**partial ptosis**” and “**apparent enophthalmos**”

Unilateral (dilated)		Reaction to light (direct)	Associated signs
Third nerve palsy		None	Ptosis (partial or complete), external ophthalmoplegia
Holmes-Adie syndrome		Slow	Better response to accommodation, lower limb areflexia
Marcus Gunn pupil		Slow and incomplete	Normal consensual response, optic atrophy, central scotoma, impaired color vision
Local lesion of the iris		Variable depending on extent of local damage	Irregular pupil
Unilateral (constricted)			
Horner's syndrome		Reduced dilatation to shade	Ptosis (partial), ipsilateral facial anhidrosis, "enophthalmos"
Bilateral (dilated)			
Midbrain lesion		None	Mid-position pupils; impaired vertical gaze
Iatrogenic/atropine, tricyclic antidepressants		None or reduced	
Bilateral (constricted)			
Senile		None or reduced	
Iatrogenic/pilocarpine drops		None or reduced	
Pontine lesion		None	Pin-point pupils, coma, Cheyne-Stokes respiration
Argyll-Robertson		None	Irregular pupils, normal accommodation

Fig. 6D(iii).32: Summary of pupillary abnormalities.

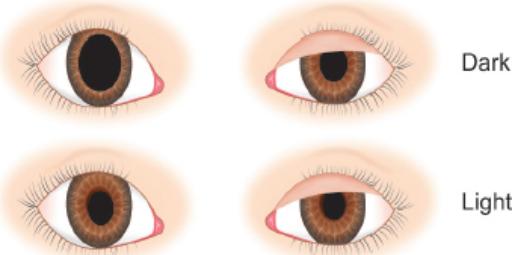


Fig. 6D(iii).33: Horner's syndrome.

Causes of Horner's Syndrome [Fig. 6D(iii).34]

Unilateral	Bilateral
<p>Central (1st order neurons): Brainstem disease (tumor, vascular, and demyelination), syringomyelia, lateral medullary (Wallenberg) syndrome, spinal cord tumor, and base of skull tumors/injury</p> <p>Preganglionic (2nd order neuron):</p> <ul style="list-style-type: none"> • Pancoast tumor, carotid and aortic aneurysm and dissection, neck lesions (glands, trauma, and postsurgical) • Birth trauma with lower brachial plexus injury and cervical rib <p>Postganglionic (3rd order neuron): Cluster headaches (migrainous neuralgia), internal carotid artery dissection, nasopharyngeal tumor, otitis media, cavernous sinus mass, Raeder syndrome (paratrigeminal syndrome), and carotid cavernous fistula</p>	<p>Diabetic autonomic neuropathy, amyloidosis, pure autonomic failure, Anderson–Fabry disease, familial dysautonomia, and paraneoplastic syndrome</p>

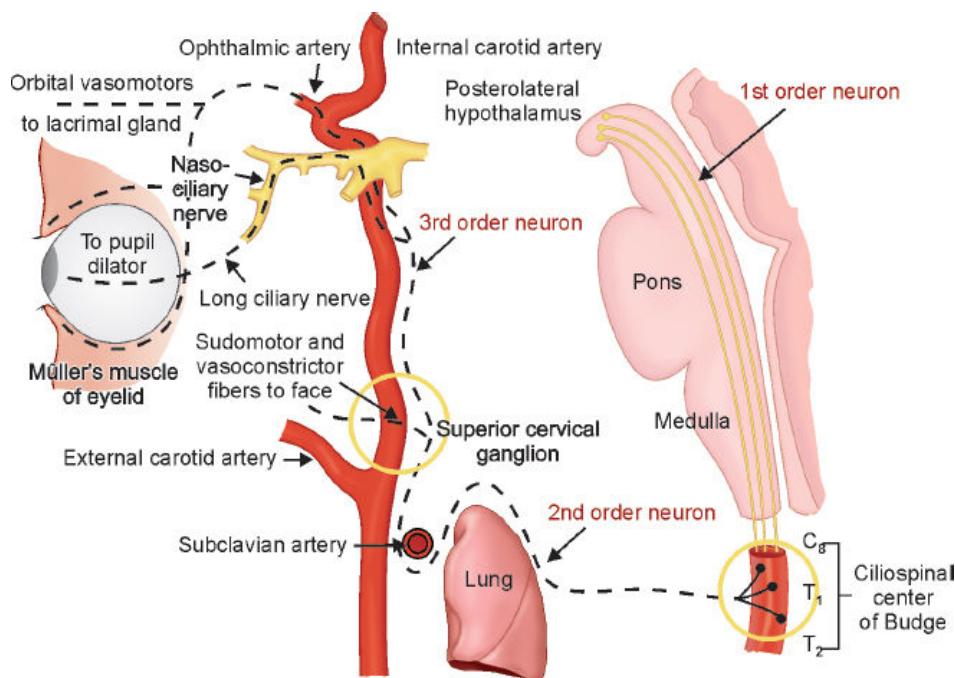


Fig. 6D(iii).34: Diagrammatic representation of sites of involvement of Horner's syndrome.

OPHTHALMOPLEGIA

Definitions:

- **Supranuclear ophthalmoplegia:** Also called as gaze palsies. It is due to involvement of corticonuclear fibers of the III, IV, and VI cranial nerves.
- **Internuclear ophthalmoplegia:** It is due to involvement of MLF and PPRF which connect the III nerve to the contralateral VI nerve.
- **Nuclear/intranuclear ophthalmoplegia:** Involvement of individual cranial nerves (CN III, IV, and VI).

1st neuron: Associated symptoms of brainstem involvement, such as dizziness, vertigo, transient ischemic attacks suggestive of hemianopia with/without long tract signs

- Hydroxyamphetamine—dilates both pupils
- Phenylephrine—dilates both pupils
- Cocaine—Horner's pupil dilates more poorly than normal pupil

2nd neuron: Chest mass with arm pain, phrenic nerve paralysis, supraclavicular nodes, neck mass, thyroid enlargement, neck surgery, neck injury, cervical osteoarthritis with bone spurs

- Hydroxyamphetamine—dilates both pupils
- Phenylephrine—dilates both pupils
- Cocaine—Horner's pupil dilates more poorly than normal pupil

3rd neuron: History of vascular headache (migraine, Raeder's, cluster), carotid artery disease with ipsilateral visual loss and contralateral motor and sensory signs. Sweating present if above bifurcation of carotid artery and absent if below bifurcation

- Hydroxyamphetamine—Horner's pupil dilates less or not at all
- Phenylephrine—Horner's pupil dilates more
- Cocaine—Horner's pupil dilates more poorly or not at all

Fig. 6D(iii).35: Differentiating features of 1st order, 2nd order, and 3rd order Horner's syndrome.



Fig. 6D(iii).36: Horner's syndrome.

- **Internal ophthalmoplegia:** Paralysis of constrictor pupillae and ciliary muscle
- **External ophthalmoplegia:** Paralysis of extraocular muscles
- **Total ophthalmoplegia:** Combination of external and internal ophthalmoplegia.

Gaze Palsies/Supranuclear Ophthalmoplegia

Vertical Gaze Palsies

Upward gaze palsy:

- Lesions at the superior colliculus—Parinaud's syndrome
- Progressive supranuclear palsy
- Parkinson's disease
- Wernicke's encephalopathy
- Thalamic hemorrhage (Sunset sign).

Downward gaze palsy:

- Huntington's chorea
- Niemann–Pick disease
- Olivopontocerebellar ataxia
- Progressive supranuclear palsy
- Parkinson's disease.

Combined upward and downward gaze palsy:

- Bilateral frontal lobe lesions
- Progressive supranuclear palsy
- Parkinson's disease.



Fig. 6D(iii).37: Reptilian stare in progressive supranuclear palsy.

Horizontal Gaze Palsies

- | | |
|--|--|
| <ul style="list-style-type: none"> • Frontal eye field (Area number 8) • Destructive lesion—both eyes will turn toward the side of lesion (Vulpian sign) • Irritative lesion—both eyes will turn to opposite side | <ul style="list-style-type: none"> • Pontine lateral gaze center • Destructive lesion—loss of lateral gaze to the same side • Irritative lesion—eyes deviate to the same side as lesion |
|--|--|

Internuclear Ophthalmoplegia

- Caused by a lesion of the medial longitudinal fascicle (MLF), which carries signals from the abducens nucleus to the contralateral medial rectus oculomotor subnucleus [Fig. 6D(iii).38].

- The abducens nerve and MLF coordinate conjugate horizontal eye movements with co-contraction of ipsilateral lateral rectus and contralateral medial rectus muscles.
- Classic signs of unilateral internuclear ophthalmoplegia include impaired adduction of the ipsilesional eye and abducting nystagmus of the contralateral eye.
- Despite ipsilateral adduction weakness with direct motility testing, adduction is often intact with convergence because convergence signals to the medial rectus nucleus are distinct from the MLF.
- Multiple sclerosis and microvascular brainstem ischemia are the most common causes.

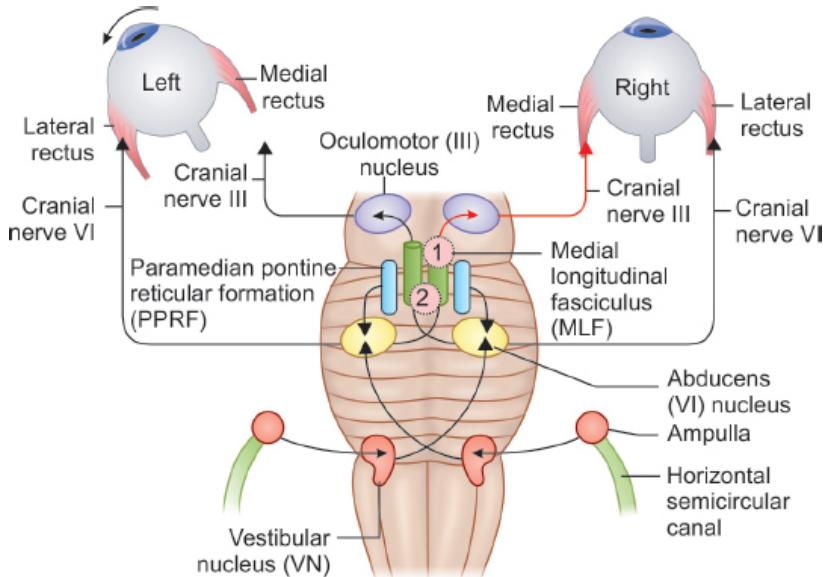


Fig. 6D(iii).38: Internuclear ophthalmoplegia.

Superior INO (Lhermitte's syndrome)	Lesions in the brainstem
Inferior INO (Lutz syndrome)	Lesions in the pontine lateral gaze center → to the abducens nucleus
Pseudo-INO	<ul style="list-style-type: none"> Myasthenia Gravis Miller Fisher syndrome
Webino syndrome (wall eyed bilateral INO)	Bilateral MLF and bilateral medial rectus nucleus
Wemino syndrome (wall eyed mono-ocular INO)	Unilateral MLF and unilateral medial rectus nucleus
One and a half syndrome	Involvement of pontine PPRF and adjacent MLF
Eight and a half syndrome	One and a half syndrome + 7th nerve palsy

(INO: internuclear ophthalmoplegia; MLF: medial longitudinal fasciculus; PPRF: paramedian pontine reticular formation; WEBINO: wall-eyed bilateral internuclear ophthalmoplegia; WEMINO: wall-eyed mono-ocular internuclear ophthalmoplegia)

Etiology of Nuclear or Infranuclear Palsy

Site	Occulomotor nerve	Trochlear nerve	Abducens nerve palsy
------	-------------------	-----------------	----------------------

	palsy	palsy	
Brainstem	<ul style="list-style-type: none"> Weber's syndrome Nothnagel syndrome Benedict's syndrome Claude's syndrome 	Midbrain syndromes	<ul style="list-style-type: none"> Millard-Gubler syndrome Raymond-Cestan syndrome Foville's syndrome Möbius syndrome
Subarachnoid space	+	+	-
Petros apex—Dorello's canal	-	-	+ (Gradenigo's syndrome)
Cavernous sinus	+	+	+
Superior orbital fissure	+	+	+
Orbit	+/-	+	-

Painful Ophthalmoplegia

- Cavernous sinus thrombosis
- Superior orbital fissure syndrome—Tolosa–Hunt syndrome
- Ophthalmoplegic migraine
- Pituitary apoplexy
- Orbital cellulitis
- Orbital tumors.

	Supranuclear ophthalmoplegia	Nuclear/intranuclear ophthalmoplegia
Movements affected	Gaze	Individual muscle movements
Diplopia and squint	Absent	Present
Pupils	Normal	May or may not be involved
Vestibulo-ocular reflex (cold caloric)	+	-

NYSTAGMUS

Definition: Nystagmus is involuntary, conjugate, repetitive, and rhythmic movements of eyeball.

Method of examination: Eyes should be deviated in all four directions for at least 5 seconds and deviation should not be of extremes.

Grading/degrees of nystagmus	
I	Nystagmus only on deviation of eyes
II	Nystagmus on looking forward
III	Direction of nystagmus opposite to the fast beating component

Types of Nystagmus

Pendular nystagmus	Jerk nystagmus			Nystagmus of dissociated rhythm
	Horizontal	Vertical	Rotatory	
In this type of amplitude of nystagmus is equal in either directions	In this type of nystagmus, there is slow component followed by fast (jerk) component due to cortical correction			Usually gaze evoked nystagmus
These are predominantly seen in congenital conditions especially due to visual defects from earlier years	<ul style="list-style-type: none"> Labyrinthine disorders Cerebellar disorders Uppermost cervical lesion 	<ul style="list-style-type: none"> Never labyrinthine Cerebellar disorders Brainstem lesions Drugs like benzodiazepines and barbiturate 	<ul style="list-style-type: none"> Labyrinthine disorders Brainstem lesions 	<ul style="list-style-type: none"> MLF lesions Multiple sclerosis

Other Common Types of Nystagmus

	Description	Condition seen
Seesaw nystagmus	Upward deflection of one eyeball with downward deflection on the contralateral eyeball	Suprasellar region anterior to III ventricle
Up beat nystagmus	Fast movement upward	Lesions in the vermis of the cerebellum
Down beat nystagmus	Fast component is down	Foramen magnum lesions
Optokinetic nystagmus	Railway track nystagmus	Deep parietal lobe lesions
Convergence retraction nystagmus	Attempted upgaze provokes jerk nystagmus with fast component in inward convergent manner	Lesion at superior colliculus—Parinaud's syndrome

Non-nystagmus Oscillations of Eyeball

Ocular flutter	Periodic horizontal saccades	Cerebellar and PPRF lesions
Opsoclonus	Irregular oscillations with different amplitude and directions	<ul style="list-style-type: none"> Toxins Encephalitis
Ocular bobbing	Rapid downstroke followed by slow uprise of eyeball	Pontine destruction
Ocular dipping	Slow downstroke followed by rapid uprise of the eyeball	Toxic encephalopathy

(PPRF: paramedian pontine reticular formation)

	Central nystagmus	Peripheral nystagmus
Fast component	Fast component is toward same side of pathology	Fast component is to the opposite of the pathology

Duration of episode	Long lasting	Acute and transient
Vertigo	Less prominent	Usually associated
Suppression on fixation using Frensel lens	Not suppressed	Suppressed
Pursuits and saccades	Usually present	Absent
Other clinical finding	CNS involvement is seen	Hardness of hearing and tinnitus is seen

(CNS: central nervous system)

CRANIAL NERVE V—TRIGEMINAL NERVE

- Largest among cranial nerves
- Most complex of the cranial nerves

We shall discuss trigeminal nerve under:

1. Sensory component and motor components
2. Reflexes
3. Disorders of trigeminal nerve dysfunction

Sensory and Motor Component

Component	Sensory part	Motor part
Size	Larger	Smaller
Nuclei	Three nuclei	One nuclei
Distribution	<ul style="list-style-type: none"> • Face (except angle of mandible) • Teeth • Oral cavity • Nasal cavity • Scalp to vertex • Intracranial dura • Cerebral vasculature • Proprioception to muscles of mastication 	Muscles of mastication

Distribution [Fig. 6D(iii).39]: The distribution of CN V3 does not extend to the jaw line; there is a large “notch” at the angle of the jaw innervated by the greater auricular nerve (C2-3).

Nuclei and functions:

Nuclei	Location	Function
Motor nuclei	Pons	<ul style="list-style-type: none"> • Muscles of mastication • Mylohyoid • Anterior belly of digastric • Tensor veli palatini • Tensor tympani

Principle sensory nucleus	Pons	<ul style="list-style-type: none"> • Pressure • Touch • Vibration
Mesencephalic nuclei	Extends to midbrain	Proprioception of muscles of mastication, EOM, facial expression
Spinal nucleus	Extends to spinal nucleus (C3, 4) via medulla — quintothalamic tract	<ul style="list-style-type: none"> • Pain • Temperature

Note:

- All the sensory supply relay via trigeminal ganglion which is also called as Gasserian ganglion or semilunar ganglion.
- It is largest ganglion located at Meckel's cave, lateral to ICA and posterior to cavernous sinus.
- It is analogous to dorsal root ganglion.

Testing of sensory component:

- Test the sensation of the face for touch, pain, and temperature in each of the divisions.
- Sensation should be compared in each trigeminal division, and the perioral region compared to the posterior face to exclude an onion skin pattern (**Figs. 6D(iii).40 to 6D(iii).43**)
- Pain or temperature should be compared with touch to exclude dissociated sensory loss (a common finding in lateral medullary syndrome).

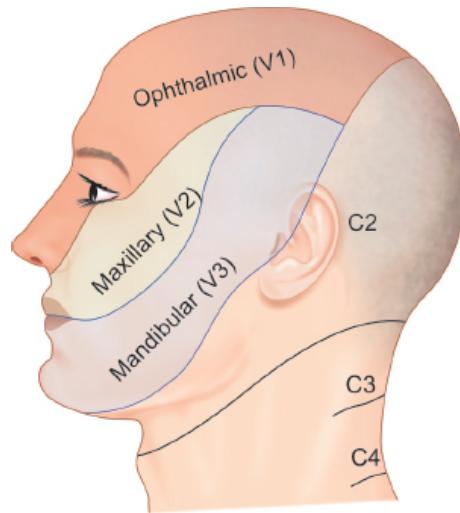


Fig. 6D(iii).39: Image showing sensory distribution of three divisions of trigeminal nerve.

- On the trunk, organic sensory loss typically stops short of midline because of the overlap from the opposite side, and crossing of the midline suggests nonorganic nature of the symptoms. However, this finding is not reliable on the face because there is less midline overlap, so organic facial sensory loss may extend to the midline.



Fig. 6D(iii).40: Examination of sensory component of trigeminal nerve.



Fig. 6D(iii).41: Examination of ophthalmic division of trigeminal nerve.



Fig. 6D(iii).42: Examination of maxillary division of trigeminal nerve.



Fig. 6D(iii).43: Examination of mandibular division of trigeminal nerve.

Testing of motor component [Figs. 6D(iii).44 and 6D(iii).45]:

- Motor component can be gauged by palpating these muscles as the patient clinches the jaw. An effective technique is to place the examining fingers along the anterior or lateral border of the masseters bilaterally.
- When the jaw is clenched, the fingers will move forward (when fingers placed anteriorly) or sideward (when fingers placed laterally); this movement should be symmetric on the two sides.
- Unilateral trigeminal motor weakness causes deviation of the jaw toward the weak side on opening, due to the unopposed action of the contralateral lateral pterygoid. Careful observation of jaw opening is often the earliest clue to the presence of an abnormality.
- It is occasionally difficult to be certain whether the jaw is deviating or not. Note the relationship of the midline notch between the upper and lower incisor teeth; it is a reliable indicator.

Unilateral weakness of CN V innervated muscles	Bilateral weakness of the muscles of mastication with inability to close the mouth (dangling jaw)
Suggests: <ul style="list-style-type: none"> • The brainstem • Gasserian ganglion • The motor root of CN V at the base of the skull 	Suggests: <ul style="list-style-type: none"> • Motor neuron disease • Neuromuscular transmission disorder • Myopathy



Fig. 6D(iii).44: Examination of motor component of trigeminal nerve (masseter muscle).



Fig. 6d(iii).45: Examination of motor component of trigeminal nerve (pterygoid muscle).

Rule of 17 (10 + 7 and 12 + 5)

- **10 + 7** → In facial nerve weakness and vagus nerve involvement, the deviation will be toward the normal side
- The levator anguli oris (in CN 7) and palatopharyngeus (in CN 10) are ‘pulling’ muscles. Hence, the normal side ‘pulls’ the angle of mouth/uvula toward the normal side
- **12 + 5** → In trigeminal nerve and hypoglossal nerve weakness, the deviation will be toward the affected side
- The lateral pterygoid (CN 5) and the genioglossus (CN 12) are ‘pushing’ muscles. Hence, the normal side ‘pushes’ the angle of jaw/tongue toward the affected side

Reflexes

Reflexes associated with V nerve:

1. Jaw jerk [**Fig. 6D(iii).46**]
2. Sternutatory reflex
3. Corneal reflex
4. Conjunctival reflex

Jaw jerk or Masseter or Mandibular reflex

Theory: Sensory fibers → mesencephalic nucleus → reflex center in pons → motor nucleus → motor fibers

Normal	Minimal or absent response
Limb hyperreflexia due to cervical spinal lesion	Normal jaw reflex
Generalized hyperreflexia	Exaggerated jaw reflex

Note: Exaggerated reflex is due to lesion in the bilateral corticobulbar tracts above motor nucleus, e.g. pseudobulbar palsy or amyotrophic lateral sclerosis.



Fig. 6D(iii).46: Illustration showing examination of jaw jerk.

Testing [Fig. 6D(iii).47]:

- Examiner places the index finger or thumb over the middle of patient's chin, holding the mouth open about midway with jaw relaxed and then taps the finger with reflex hammer.
- The response is upward jerk of mandible.

Other methods:

- For bilateral response:
 - Tapping chin directly
 - Placing the tongue blade over the tongue or lower incisor and tapping the protruding end.
- For unilateral response:
 - Tapping the angle of the jaw
 - Placing the tongue blade over the lower molar teeth of one side and tapping the protruding end.



Fig. 6D(iii).47: Examination of jaw jerk.

Sternutatory/Nasal/Sneeze Reflex

Primary clinical use is to cross check the corneal reflex.

Method: Stimulation of nasal mucous membrane with cotton, a spear of tissue or similar object → wrinkling of nose, eye closure, and often a forceful exhalation resembling a feeble sneeze.

Theory: The ophthalmic division of trigeminal innervates the nasal septum and anterior nasal passages.

Afferent limb	Center	Efferent limb
V1	Brainstem and upper spinal cord	V VII IX X

Corneal Reflex

- Elicited by lightly touching the cornea with wisp of cotton or tissue [Fig. 6D(iii).48].
- Stimulus is ideally delivered to upper cornea because the lower cornea may be innervated by CN V2 in some individuals.
- Stimulus should be ideally brought in from the side so that patient cannot see it.
- Stimulus must be delivered to cornea but not sclera

Afferent limb	Efferent limb
V1	VII

Conjunctival Reflex

- Same as corneal reflex [Fig. 6D(iii).48]
- However, the sensitivity of corneal reflex is more.

Trigeminal lesion (complete)		
	Direct reflex	Consensual (indirect) reflex

Stimulus to involved eye	Absent	Absent
Stimulus to opposite eye	Present	Present
Facial nerve lesion (complete)		
	<i>Direct reflex</i>	<i>Consensual (indirect) reflex</i>
Stimulus to involved eye	Absent	Present
Stimulus to opposite side	Present	Absent



Fig. 6D(iii).48: Demonstration of corneal/conjunctival reflex.

Disorders of V Nerve Dysfunction

1. Motor Dysfunction

- Unilateral UMN lesion—generally no weakness observed.
- Bilateral UMN lesion—pseudobulbar palsy—marked weakness seen with exaggerated jaw jerk.
- Myasthenia gravis—masticatory fatigue (not to be confused with claudication pain of giant cell arteritis)
- ALS: Jaw drop with diminished jaw jerk—dysphagia and difficulty in swallowing their own saliva.
- Involuntary movements include—dystonia (extrapyramidal symptoms of antipsychotic drugs), Meige syndrome (oromandibular dystonia with blepharospasm), and trismus.

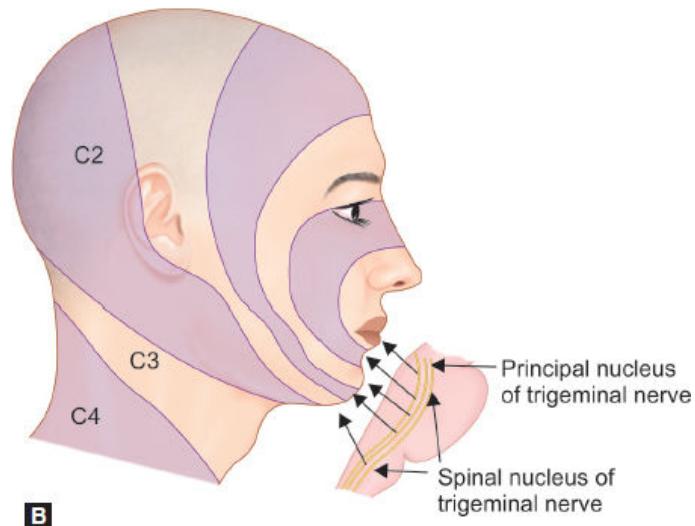
Causes of trigeminal nerve involvement

- Supranuclear—bilateral (pseudobulbar) palsy
- Nuclear—syringobulbia
- Nerve root—cerebellopontine angle tumor
- Gasserian ganglion—Gradenigo syndrome, otitis media, meningitis, and aneurysms of internal carotid artery
- Cavernous sinus—thrombosis/tumor
- Superior orbital fissure—Tolosa–Hunt

- Individual branches involvement

2. Sensory Dysfunction

Site of lesion	Disease	Manifestation
Parietal lobe or sensory radiation (supranuclear lesion)	Stroke/tumors	May raise the sensory threshold of contralateral face
Thalamic lesion	Stroke/tumors	Facial hypoesthesia with hyperpathia or allodynia
Principal sensory nucleus: • Pressure • Touch • Vibration	Stroke/tumors	Diminished tactile sensation of skin and mucous membrane of that side
Spinal nucleus	Lateral medullary or pontine lesion/tumors	Pain and temperature loss
Intramedullary lesion	Syringomyelia/syringobulbia/tumors	Dissociative loss of sensation



Figs. 6D(iii).49A and B: (A) Balaclava helmet and (B) Dejerine onion skin distribution seen in syringobulbia.

Trigeminal Neuralgia (Also known as Fothergill's disease Tic douloureux)

- Most common disorder to involve trigeminal sensory function.

- Paroxysms of fleeting but excruciating unilateral facial pain—usually involves II and III division and rarely I division.
- Pain lasts for few seconds but may occur many times per day.
- Trigger for pain may be talking, chewing, brushing, exposure to cold or by wind on face.
- Most common cause for compression of sensory root by ecstatic arterial loop of the basilar artery (AICA or superior cerebellar artery)
- Other causes include MS, tumors of CP angle—bilateral is suggestive of MS.

3. Postherpetic Neuralgia

Acute herpes zoster is extremely painful.

- Usually in CN V1—pain in vesicles in forehead, eyelid, and cornea but may affect other division also.
- Persistent neuralgic pain syndrome after 1 month of acute eruption is appropriately labeled as postherpetic neuralgia. It is a dysesthetic with burning component, constant but with superimposed paroxysm of lancinating pain that may be provoked by touching certain spots with affected area.
- There may be hypo- or hyperesthesia.

4. Facial Numbness

- *Numb chin syndrome*: In distribution of mental nerve—due to metastatic process in mental foramen.
- *Numb cheek syndrome*: Involvement of infraorbital nerve.

5. Other Trigeminal Nerve Disorders

Marcus-gunn phenomenon or jaw winking phenomenon	Seen in congenital ptosis: Opening the mouth, chewing or lateral jaw movements cause an exaggerated reflex elevation of the ptotic lid due to proprioceptive impulses from the pterygoid muscles being misdirected to the oculomotor nucleus
Reversed Gunn phenomenon or inverse jaw winking or Marin-Amat sign	Synkinesis due to aberrant regeneration of facial nerve where there is involuntary closure of one eye on mouth opening
Frey syndrome	Flushing, warmth, and excessive perspiration over the cheek and pinna on one side following ingestion of spicy food—due to misdirection of secretory fibers to parotid gland to the sweat glands and vasodilator ending in the auriculotemporal nerve distribution—usually follows trauma or infection of parotid gland or local nerve injury
Sturge–Weber or Weber–Dimitri disease	Congenital nevi or angiomas over the side of face in the trigeminal distribution with associated ipsilateral leptomeningeal angiomas and intracortical calcification with attendant neurologic complications
Raeder's paratrigeminal syndrome	<ul style="list-style-type: none"> • Unilateral oculosympathetic paresis (differential diagnosis with Horner) • Ipsilateral trigeminal involvement
Gradenigo's syndrome	<ul style="list-style-type: none"> • Damage to V1 division of trigeminal nerve • Ipsilateral 6th nerve palsy
Cavernous sinus syndrome	3, 4, 6 nerves with V1 and V2 (less often)

Superior orbital fissure syndrome	Never involving V2, other than that similar to cavernous sinus syndrome. Exophthalmos and blindness can be present
V1: Bilateral corneal anesthesia	Diabetic neuropathy
V2: Numb cheek syndrome	<ul style="list-style-type: none"> Infraorbital nerve Distribution: Squamous cell carcinoma, skin and LASIK
V2: Trumpet player's neuropathy	Anterior superior alveolar nerve
V3: Tongue numbness	<ul style="list-style-type: none"> Lingual nerve in temporal Arteritis
V3: Numb chin syndrome/roger's sign	Mental neuropathy: Cancer of breast and lung, giant cell arteritis, Burkitt lymphoma, and sickle cell disease

FACIAL NERVE

Motor (70%)	Sensory	Parasympathetic
<ul style="list-style-type: none"> Muscles of facial expression Scalp Ear Buccinators Platysma Stapedius Stylohyoid Posterior belly of digastrics 	<p>Taste: Anterior 2/3</p> <p>Exteroceptive:</p> <ul style="list-style-type: none"> Eardrum EAC <p>Proprioception: From the muscles supplied by it</p> <p>GVS:</p> <ul style="list-style-type: none"> Salivary glands Mucosa of nose and pharynx 	<ul style="list-style-type: none"> Submandibular Sublingual Lacrimal Mucous membrane of oral and nasal mucosa

(EAC: external auditory canal)

Note:

- There is anatomical segregation of motor component from sensory and autonomic fibers.
- Sensory root (nervus intermedius of Wrisberg)—contains both sensory and autonomic fibers.

Examination of Motor Function

Inspection:
<ul style="list-style-type: none"> Facial asymmetry, nasolabial fold with forehead wrinkles, and movements during spontaneous facial expression Tone of the muscles of facial expression Atrophy and fasciculations Abnormal muscle contractions and involuntary movements Spontaneous blinking for frequency and symmetry
Testing the temporal branches of the facial nerve: Patient is asked to frown and wrinkle his or her forehead
Testing the zygomatic branches of the facial nerve: Patient is asked to close their eyes tightly

Testing the buccal branches of the facial nerve:

- Puff up cheeks (buccinator)
- Smile and show teeth (orbicularis oris)
- Tap with finger over each cheek to detect ease of air expulsion on the affected side

Muscle tested	Instruction	Response in palsy
Frontal belly of occipitofrontalis (Fig. 6D(iii).50)	Ask the patient to wrinkle his/her forehead	Asymmetry as he/she cannot wrinkle his forehead on the side of palsy in lower motor neuron (LMN) palsy
Orbicularis oculi [Fig. 6D(iii).51]	Ask the patient to close his/her eyes forcibly while you try to open the eyelids with your fingers	In LMN palsy, eyelids do not close completely. Instead the eyeball rolls up. This is known as Bell's phenomenon. In healthy individuals, eyelids cannot be opened with mild force against patient's resistance
Levator anguli oris, zygomatic major and minor, depressor anguli oris, buccinator, and risorius [Fig. 6D(iii).52]	Ask the patient to show his/her teeth or smile	Angle of mouth deviates toward normal side
Orbicularis oris and buccinators [Fig. 6D(iii).53]	Ask the patient to blowout cheeks with mouth closed, i.e. puff the cheeks and assess power by your attempt to deflate the cheek. Ask the patient to whistle	Patient cannot blowout his cheek as air escapes from affected side
Platysma [Fig. 6D(iii).54]	Ask the patient to clench his/her teeth and simultaneously depress the angles of mouth	Folds of platysma is seen in the neck as flat



Fig. 6D(iii).50: Examination of frontal belly of occipitofrontalis.



Fig. 6D(iii).51: Examination of orbicularis oculi.



Fig. 6D(iii).52: Examination of levator anguli oris.



Fig. 6D(iii).53: Examination of buccinator.



Fig. 6D(iii).55: Examination of taste sensation.



Fig. 6D(iii).54: Examination of platysma.

Examination of Sensory System

Anterior two-thirds of tongue [Fig. 6D(iii).55]

- Tongue protruded
- Hold with soft gauze
- With applicator's tip apply over the dorsum of the tongue
- Rinse after each test with water
- Sensations from the tip to deep—follow sweet → salt → sour → bitter (last)
- Fifth modality—umami appreciated with compounds of some amino acids
- Normally taste is appreciated within 10 seconds
- Artificial sweeteners make better test substances than ordinary sugar.

Aguesia

Complete inability to perceive taste

Hypogeusia	Blunted or delayed taste
Parageusia	Perversions of taste
Impaired taste	Lesion is proximal to junction with chorda tympani
Not affected	Lesion is at or distal to stylomastoid foramen

Secretory Function

1. Lacrimation: Schirmer's test → 10 mm is normal
2. Nasolacrimal test: By diluted solution of ammonium and formaldehyde—trigeminal nerve → greater superficial petrosal nerve.

Reflexes
<i>Orbicularis oculi reflex</i>
Percussion causes reflex contraction of the eye muscle. The reflex is known as the supraorbital, glabellar, or nasopalpebral reflex, depending upon the site of the stimulus. Both eyes usually close, with the contralateral response being weaker. The trigeminal nerve is the afferent side and the facial nerve the efferent side of the reflex. Light and sound can also produce the reflex, with the optic and acoustic nerves providing the afferent side
The response is weak or abolished in nuclear and peripheral lesions, and present or exaggerated in supranuclear lesions. It is exaggerated in Parkinsonism and cannot be voluntarily inhibited
<i>Palpebral oculogyric reflex</i>
The eyeballs deviate upward when the eyes are closed, both when awake and asleep. The afferent arc is proprioceptive impulses carried through the facial nerve to the medial longitudinal fasciculus. The oculomotor nerve to the superior rectus muscles forms the efferent side
In peripheral and nuclear lesions, an exaggeration of this reflex is known as Bell's phenomenon
<i>Orbicularis oris reflex</i>
Percussion on the side of the nose or the upper lip causes ipsilateral elevation of the angle of the mouth and upper lip. The reflex arc is composed of the fifth and seventh nerves. <i>Synonyms:</i> nasomental, buccal, oral, or perioral reflex
This reflex disappears after about the first year of life, recurring with supranuclear facial nerve lesions and with extrapyramidal diseases, such as Parkinsonism
<i>Snout reflex</i>
Tapping the upper lip lightly with a reflex hammer, tongue blade, or finger causes bilateral contraction of the muscles around the mouth and base of the nose. The mouth resembles a snout
This is an exaggeration of the orbicularis oris reflex. It is present with bilateral supranuclear lesions and in diffuse cerebral diseases, such as various causes of dementia
<i>Sucking reflex</i>
Sucking movements of lips, tongue, and mouth are brought about by lightly touching or tapping on the lips. At times, merely bringing an object near the lips produces the reflex
Occurs in patients with diffuse cerebral lesions. The snout reflex occurs in similar circumstances
<i>Palmental reflex</i>
A stimulus of the thenar area of the hand causes a reflex contraction ipsilaterally of the orbicularis oris and mentalis muscles

A number of normal individuals have this reflex, and also patients with diffuse cerebral disease. It is significant when other similar reflexes are also present

Corneal reflex

Stimulation of the cornea with a wisp of cotton produces reflex closure of both ipsilateral (strongest) and contralateral eyelids. The fifth nerve carries the afferent impulses, and the facial nerve the efferent impulses

Site of cranial nerve 7 lesion and associated manifestations

Lesion location	Manifestations
Above the facial nucleus (supranuclear lesion)	Contralateral paralysis of lower facial muscles with relative preservation of upper muscles. Lesion located cortex, internal capsule or midbrain
Pons (nuclear or fascicular lesion)	Ventral pontine lesion (of Millard–Gubler): Ipsilateral facial monoplegia, lateral rectus palsy (VI), and contralateral hemiplegia (corticospinal fibers). Pontine tegmentum lesion (of Foville): Ipsilateral facial monoplegia; contralateral hemiplegia (corticospinal fibers); paralysis of conjugate gaze to side of lesion (pontine paramedian reticular formation)
Cerebellopontine angle (peripheral nerve lesion)	Ipsilateral facial monoplegia, loss of taste to anterior two-thirds of tongue, impairment of salivary and tear secretion, hyperacusis (if VIII is not affected). Additional cranial nerves may be involved: deafness, tinnitus, and vertigo (VIII); sensory loss over face and absence of corneal reflex (V); ipsilateral ataxia (cerebellar peduncle)
Facial canal between internal auditory meatus and geniculate ganglion (peripheral nerve type lesion here and subsequently)	Same as above except cranial nerves other than VII are not involved
Facial canal between geniculate ganglion and nerve to stapedius muscle	Facial monoplegia; impaired salivary secretion; loss of taste; and hyperacusis
Facial canal between nerve to stapedius and leaving of chorda tympani	Facial monoplegia; impaired salivary secretion; and loss of taste
After branching of chorda tympani	Facial paralysis, distribution related to site of lesion

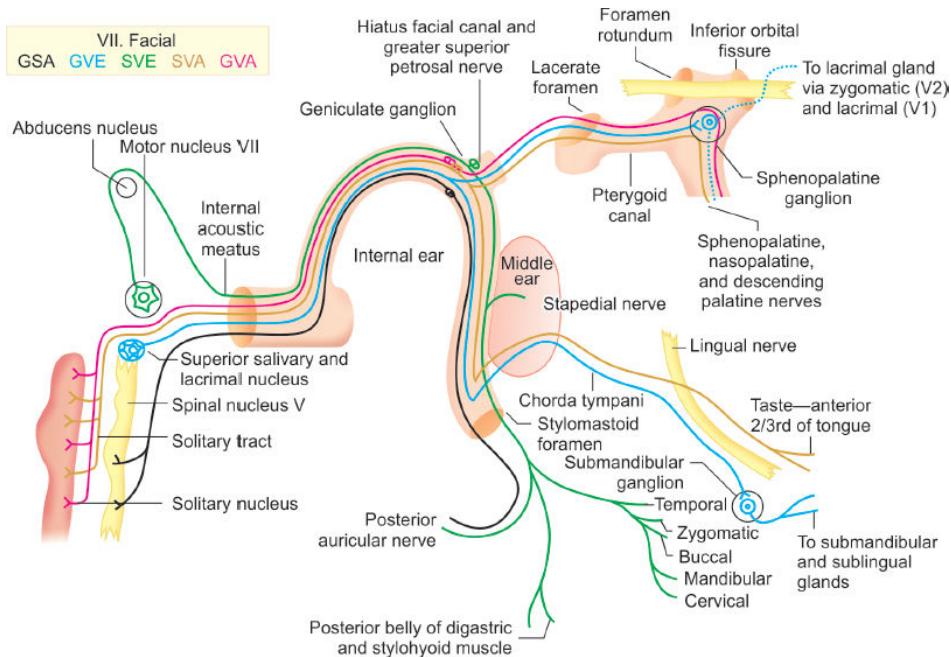


Fig. 6D(iii).56: Facial nerve pathway.

FACIAL NERVE PALSY

Peripheral Facial Palsy

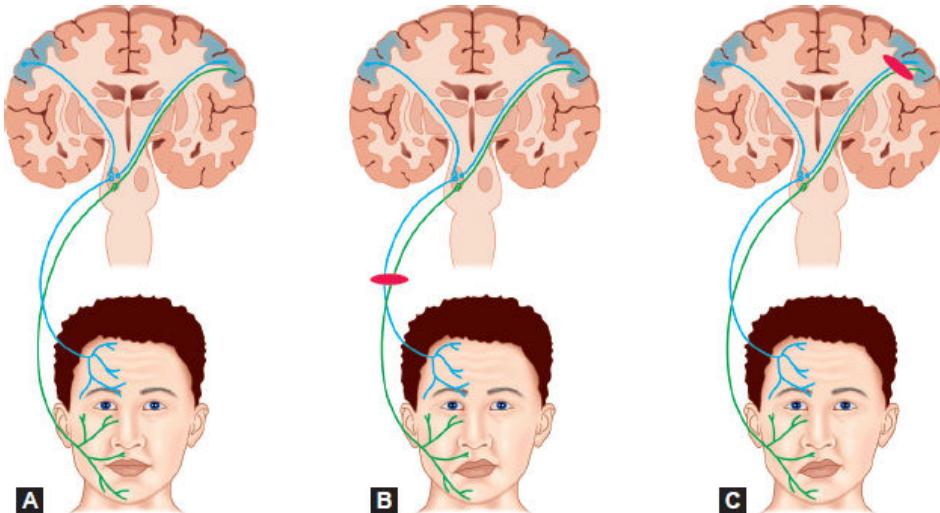
There is flaccid weakness of all the muscles of facial expression on the involved side, both upper and lower face, and the paralysis is usually complete.

Signs in LMN Facial Palsy

Bell's phenomenon	Attempting to close involved eye causes a reflex upturning of the eyeball
Levator sign of Dutemps and Céstan	Patient look down, then close the eyes slowly; because the function of levator palpebrae superioris is no longer counteracted by orbicularis oculi, upper lid on the paralyzed side moves upward slightly
Negro's sign	Eyeball on the paralyzed side deviates outward and elevates more than the normal one when the patient raises her eyes
Bergara-Wartenberg sign	Loss of the fine vibrations palpable with the thumbs or fingertips resting lightly on the lids as the patient tries to close the eyes as tightly as possible
Platysma sign of Babinski	Asymmetric contraction of the platysma, less on the involved side, when the mouth is opened

House-Brackmann grading system of LMN facial palsy	
Grade I	Normal
Grade II	Mild dysfunction, slight weakness on close inspection, and normal symmetry at rest

Grade III	Moderate dysfunction, obvious but not disfiguring difference between sides, eye can be completely closed with effort
Grade IV	Moderately severe, normal tone at rest, obvious weakness or asymmetry with movement, incomplete closure of eye
Grade V	Severe dysfunction, only barely perceptible motion, and asymmetry at rest
Grade VI	No movement



Figs. 6D(iii).57A to C: Innervation by facial nerve.

Causes of LMN Facial Palsy

Congenital:

- Möbius syndrome
- Goldenhar syndrome
- Melkersson–Rosenthal syndrome

Birth related: Forceps delivery

Idiopathic: Bell's palsy

Infection:

- Viral infection, i.e. varicella zoster (Ramsay Hunt), herpes zoster, herpes simplex, and HIV
- Otitis media
- Cholesteatoma
- Necrotizing otitis externa
- Skull base osteomyelitis
- Lyme disease
- Leprosy

Trauma:

- Temporal bone fracture
- Gunshot or penetrating injury
- Laceration

Neoplastic:

- Schwannoma
- Meningioma
- Hemangioma
- Parotid malignancy

Iatrogenic: Brain, middle ear, mastoid, parotid or facial surgery

Neurological:

- Lacunar or brainstem infarct
- Guillain–Barré syndrome
- Myasthenia gravis
- Multiple sclerosis

Metabolic:

- Diabetes mellitus
- Hypertension
- Pregnancy
- Vitamin A deficiency

Central Facial Nerve Palsy (UMN Facial Nerve Palsy)

Facial weakness of central origin/UMN facial palsy	
Volitional or voluntary	Emotional or mimetic
Weakness of the lower face, with relative sparing of upper face	Upper face is not necessarily completely spared, but it is always involved to a lesser degree than the lower face
Lesion of the cortical center in the lower third of the precentral gyrus that controls facial movements, or the corticobulbar tract	Thalamic or striatocapsular lesions, usually infarction
Weakness more marked on voluntary contraction, when patient is asked to smile or bare her teeth	Facial asymmetry more apparent with spontaneous expression, as when laughing

Differences between UMN and LMN type of facial nerve palsy

	UMN type	LNM type
Facial motor function	Wrinkling of forehead preserved (frontalis unaffected)	Total face is involved
Bell's phenomenon [Fig. 6D(iii).60A to C]	Absent	Present
Facial muscles	Not atrophied	Fasciculations, Atrophied
Taste sensation	Preserved	May be lost

Corneal reflex	Preserved	Lost
Hemiplegia	Contralateral	Ipsilateral
Babinski reflex	Present	Absent

(UMN: upper motor neuron; LMN: lower motor neuron)



Fig. 6D(iii).58: Image showing deviation of angle of mouth.



Fig. 6D(iii).59: Weakness of orbicularis oculi.

Bilateral VII Nerve Palsy

Bilateral UMN palsy	Bilateral LMN palsy
<ul style="list-style-type: none"> Emotional fibers—spared Emotional incontinence—present Associated with bilateral long tract signs 	<ul style="list-style-type: none"> Bell's phenomenon present Emotional fibers—affected Long tract signs—absent

- tract signs
- Jaw jerk—exaggerated
- Corneal reflex—present
- Taste sensation—spared
- Gag reflex—exaggerated

- Jaw jerk—normal
- Corneal reflex—absent
- Taste sensation—absent

(UMN: upper motor neuron; LMN: lower motor neuron)

- **Causes of bilateral facial nerve palsy:**
- Diabetes
- Bilateral Bell's palsy
- Borreliosis
- *Mycoplasma pneumoniae* infection
- Guillain-Barré syndrome* and Miller-Fisher syndrome
- Sarcoidosis
- Möbius syndrome
- Leukemia
- Viral infections (Herpes simplex)
- Syphilis
- Basal skull fractures
- Pontine gliomas
- Leprosy
- Mononucleosis
- Brainstem encephalitis
- Hansen's disease
- Cryptococcal meningitis
- Pontine tegmental hemorrhage

*Most common cause

Syndromes of Facial Palsy

Syndromes with facial nerve palsy

- Foville's syndrome
- Millard-gubler syndrome
- Möbius syndrome
- Ramsay hunt syndrome
- Melkersson-rosenthal syndrome [triad of recurrent infranuclear facial paralysis, orofacial edema (predominately of the lips), and lingua plicata]
- Guillain–barré syndrome
- Progressive hemifacial atrophy (parry–romberg syndrome)
- Meige syndrome (blepharospasm oromandibular dystonia, orofacial cervical dystonia, and brueghel's syndrome)
- Uveoparotid fever (heerfordt's disease)
- Goldenhar syndrome
- Crocodile tear syndrome
- Frey's syndrome



Figs. 6D(iii).60A to C: Bell's phenomenon.

CRANIAL NERVE VIII—VESTIBULOCOCHLEAR NERVE

Contains two components	
Vestibular component	Cochlear component
↓	↓
Responsible for equilibrium	Responsible for hearing
Pathway	
For linear accelerations	Organ of corti
Macula	
Utricle	↓
Saccule	
For angular acceleration	Cochlear nuclei
Ampulla	
	↓
	Inferior colliculus
	↓
	Lateral lemnisci
↓	↓
Vestibular ganglia	Medial geniculate body
↓	↓

Vestibular nerve

Brodmann areas 41 and 42 (transverse temporal gyrus of Heschl)

Examination	
Vestibular component	Cochlear component
Rotational test	Rubbing fingers
Calorie test (Fig. 6D(iii).61)	Rinne's test and Weber's test
Electronystagmography	Audiometric tests: • Pure tone audiometry • Tone decay • Bekesy audiometry

Testing for vertigo and nystagmus

In sitting position, turn the head to one side by 45°

↓

Make the patient to lie down abruptly with the head hanging down from the edge of cot

↓

This position is maintained for at least a minute

↓

Watch for nystagmus

↓

Fast component is toward the lower ear suggests following possibilities

↓

↓

Benign paroxysmal positional vertigo**Central cause**

Starts after short latency (3–10 sec), patient will have nystagmus associated with vertigo	Immediate nystagmus
Rapid adaptation	No adaptation

Testing the vestibular component of VIII nerve**Rotational test**

Patient is seated in a chair that can be rotated with his head well supported and fixed in head rest

↓

To test

Horizontal canal—head in flexed at 30°

Vertical canal—head is flexed at 120°

↓

Chair is rotated 10 times in 20 seconds

↓

Normally when the rotation to the right has stopped, there is nystagmus with its slow phase to the right and vice

versa
Calorie test
The patient is placed supine with the head tilted up by 30°. In this way, the horizontal semicircular canal is oriented in a vertical plane
↓
250 mL of water (or air at controlled temperature) is irrigated through the external auditory meatus over period of 40 seconds, first using 30°C and later using 44°C
↓
Patient fixes his eyes on the given point immediately above his head
↓
After ceasing the irrigation, the time in seconds is measured during which nystagmus on the forward gaze persist
↓
Now the test is repeated on the other ear
↓
Normal response is cold water produces fast component toward the opposite side and warm water produces a fast component toward the same side (mnemonic— COWS)

Interpretation	
No response (canal paresis)	<ul style="list-style-type: none"> Meniere's disease Acoustic nerve tumor Vestibular neuronitis Lesions of vestibular nuclei
Directional preponderance	<ul style="list-style-type: none"> Lesions of peripheral or central vestibular apparatus Cerebellum Corticofugal fibers deep in the temporal lobe
Combination of above two	Vestibular nerve or labyrinth lesions

Testing the Cochlear Component of VIII Nerve

Rinne's and Weber's test [Figs. 6D(iii).62 to 6D(iii).65]

- Done with 256/512 Hz tuning fork
- The prongs should be put equidistant on either ears while examining
- Examination should be done in quite room

Rinne's test	Weber test
<p>By two methods:</p> <ol style="list-style-type: none"> An activated fork may be place first on the mastoid process, then immediately beside the ear and patient asked which is louder Traditional method where—place the tuning fork on the mastoid and when no longer heard there move it beside the ear, where it should still be audible 	A vibrating tuning fork is place in the midline on the vertex of the skull. Normally the sound is heard equally in both ears

Interpretation	
<i>In conductive hearing loss</i>	
BC > AC (Rinne negative)	Lateralized to abnormal side
<i>In sensorineural hearing loss</i>	
AC > BC (Rinne positive)	Lateralized to normal side

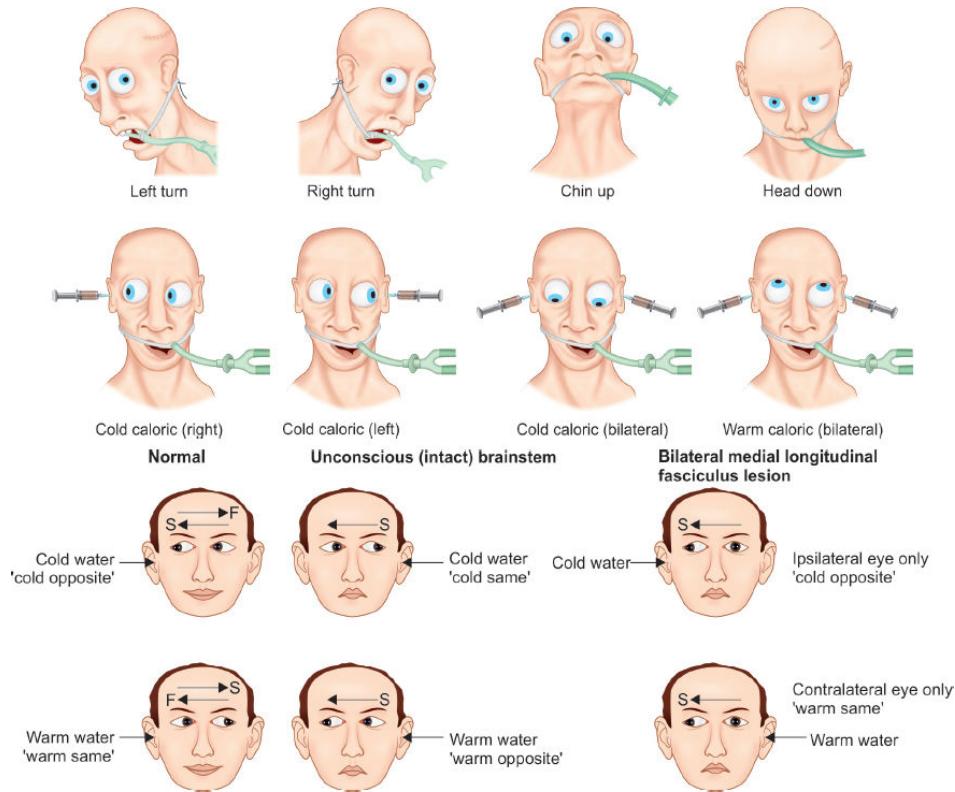


Fig. 6D(iii).61: Illustration demonstrating caloric test.

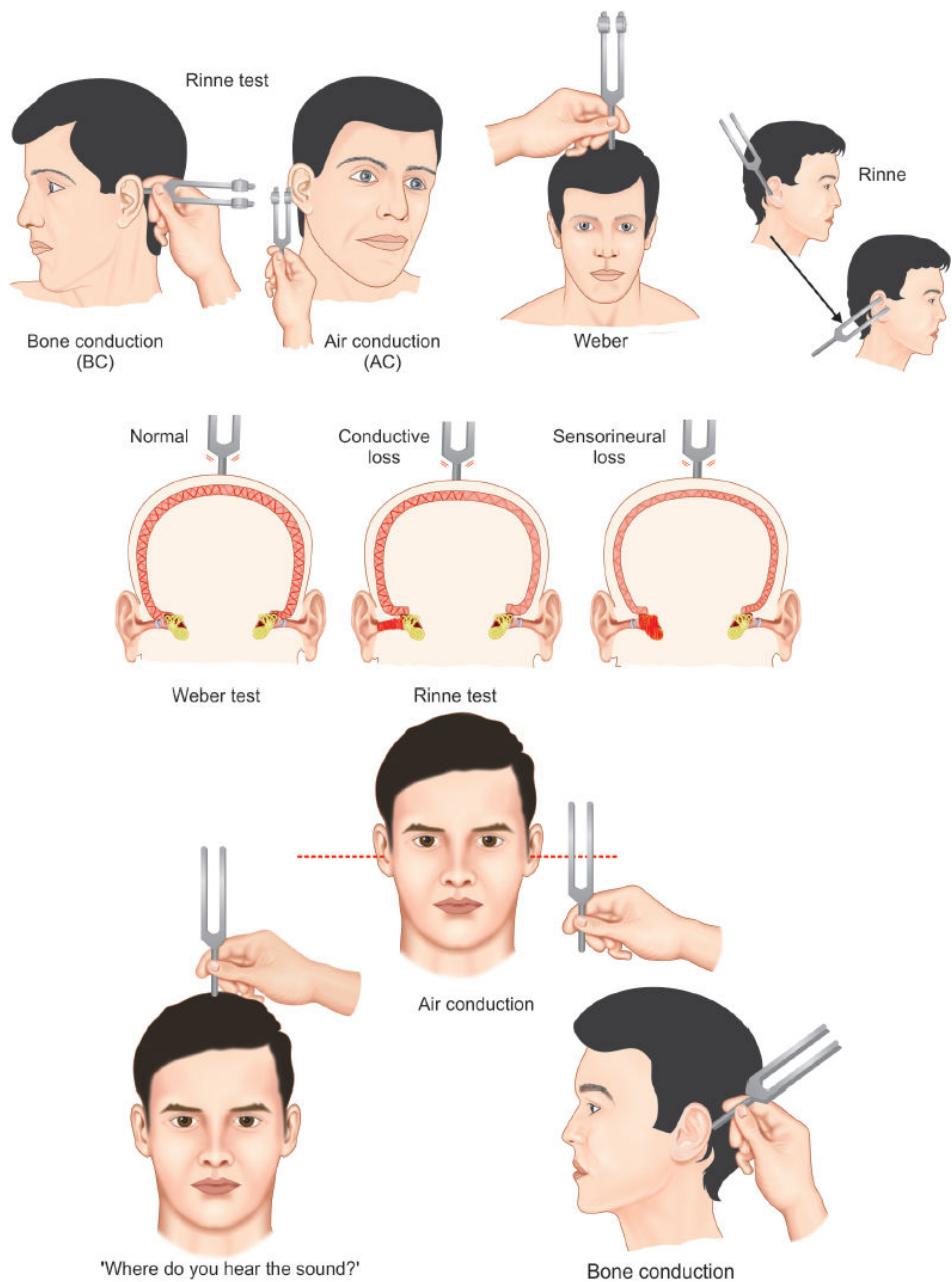


Fig. 6D(iii).62: Illustration showing demonstration of Rinne's test and Weber's test.



Fig. 6D(iii).63: Rinne's test: Placement of tuning fork on the mastoid process.



Fig. 6D(iii).64: Rinne's test: Placement of tuning fork beside the ear parallel to tympanic membrane.

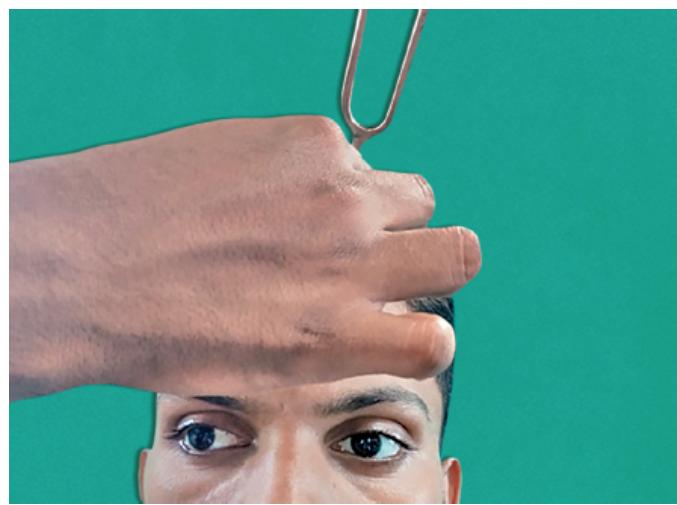


Fig. 6D(iii).65: Weber's test: Placement of tuning fork in midline on the vertex.

Causes of VIII Nerve Dysfunction Based on Site of Involvement

Vestibular component	Cochlear component
<p>At level of labyrinth:</p> <ul style="list-style-type: none"> • Meniere's disease • Motion sickness • Drug toxicity • Migraine <p>Vestibular nerve: Vestibular neuronitis</p> <p>Brainstem:</p> <ul style="list-style-type: none"> • Vascular insufficiency • Cerebellar tumors • IV ventricle tumors • Acute demyelinating diseases <p>Temporal lobe: As epileptic manifestation</p>	<p>Conduction defects:</p> <ul style="list-style-type: none"> • External meatus obstruction • Middle ear pathology • Eustachian tube block • Intracranial infection • Middle ear infection <p>Cochlear pathology:</p> <ul style="list-style-type: none"> • Meniere's disease • Osteosclerosis • Internal auditory meatus occlusion <p>Nerve trunk:</p> <ul style="list-style-type: none"> • Old age • Meningitis • Cerebellopontine angle tumors <p>Brainstem:</p> <ul style="list-style-type: none"> • Vascular pathology • Demyelination disease <p>Cerebrum: Temporal disease</p>

Unilateral and Bilateral Causes of VIII Nerve Dysfunction

Vestibular component		Cochlear component	
Unilateral	Bilateral	Unilateral	Bilateral
<ul style="list-style-type: none"> • Tumor (cerebellopontine angle and acoustic neuroma) • Fracture of the petrous temporal bone • Vascular disease of the internal auditory artery 	<ul style="list-style-type: none"> • Industrial deafness • Presbycusis • Drug toxicity (gentamicin, salicylate, etc.) • Meniere's disease 	<ul style="list-style-type: none"> • Tumor (cerebellopontine angle and acoustic neuroma) • Fracture of the petrous temporal bone • Vascular disease of the internal auditory artery 	<ul style="list-style-type: none"> • Demyelinating illness, e.g. multiple sclerosis • Migraine

- Brainstem lesion (e.g. stroke)
- Vestibular neuritis

The “doll’s eye” oculocephalic reflex

- Tests the vestibulocochlear nerve, the brainstem nuclei of the vestibulocochlear nerve, the fibers to the cerebellum, the fibers from the cerebellum, the medial longitudinal fasciculus (MLF), and the 3rd and 6th cranial nerves.
- The cause of the unconsciousness in a patient with a negative oculocephalic reflex is some sort of destructive brainstem pathology or brain death. Conversely, an intact oculocephalic reflex suggests that the coma is of a nonstructural cause, because much of the brainstem must be intact.

CRANIAL NERVE IX AND X—GLOSSOPHARYNGEAL AND VAGUS

The two nerves:

- Have motor and autonomic branches with nuclei of origin in the medulla.
- Both conduct general somatic afferent (GSA) as well as general visceral afferent (GVA) fibers to related or identical fiber tracts and nuclei in the brainstem.
- Both have a parasympathetic, or general visceral efferent, and a branchiomotor, or special visceral efferent (SVE), component
- Both leave the skull together
- Remain close in their course through the neck
- Both supply some of the same structures.
- They are often involved in the same disease processes
- Involvement of one may be difficult to differentiate from involvement of the other.

For these reasons, the two nerves are discussed together.

Muscles innervated by cranial nerve IX and X

IX nerve	
Muscular branch	Stylopharyngeus
X nerve	
Pharyngeal branch [Fig. 6D(iii).66]	<ul style="list-style-type: none"> • Musculus uvulae (azygos uvulae) • Levator veli palatini • Palatopharyngeus • Salpingopharyngeus • Palatoglossus • Superior, middle, and inferior constrictors of the pharynx
Superior laryngeal nerve	Cricothyroid
Recurrent laryngeal nerve	<ul style="list-style-type: none"> • Posterior cricoarytenoids • Lateral cricoarytenoids • Thyroarytenoids (vocalis) • Arytenoid

GLOSSOPHARYNGEAL NERVE IX

Functions:

Glossopharyngeal nerve: Sensory supply to posterior one-third of tongue, taste sensation, and pharyngeal mucosa.

Testing of IX Nerve

Cranial nerve IX is difficult to examine because most or all of its functions are shared by other nerves and because many of the structures it supplies are inaccessible.

Gag reflex [Fig. 6D(iii).67]

- The gag reflex is protective; it is designed to prevent noxious substances or foreign objects from going beyond the oral cavity.
- Components of gag reflex:** There are three motor components: elevation of the soft palate to seal off the nasopharynx, closure of the glottis to protect the airway, and constriction of the pharynx to prevent entry of the substance.
- Pathway:** The afferent limb of the reflex is mediated by CN IX and the efferent limb through CNs IX and X. The reflex center is in the medulla.
- Testing of gag reflex:** The reflex is elicited by touching the lateral oropharynx in the region of the anterior faucial pillar with a tongue blade, applicator stick, or similar object (pharyngeal reflex), or by touching one side of the soft palate or uvula (palatal reflex). The reflex also occurs with touching the base of the tongue or posterior pharyngeal wall.
- Clinical implication:** May be bilaterally absent in some normal individuals. Unilateral absence signifies a lower motor neuron lesion. Like most bulbar muscles, the pharynx receives bilateral supranuclear innervation, and a unilateral cerebral lesion does not cause detectable weakness. A hyperactive gag reflex may occur with bilateral cerebral lesions, as in pseudobulbar palsy and amyotrophic lateral sclerosis (ALS).



Fig. 6D(iii).66: Examination of deviation of uvula.



Fig. 6D(iii).67: Examination of gag reflex.

Disorders of IX Cranial Nerve

- **Unilateral supranuclear lesions** cause no deficit because of the bilateral corticobulbar innervation.
- **Bilateral supranuclear lesions** may cause pseudobulbar palsy.
- **Nuclear and infranuclear processes** that may affect CN IX include intramedullary and extramedullary neoplasms and other mass lesions (e.g. glomus jugulare tumor), trauma (e.g. basilar skull fracture or surgical dissection), motor neuron disease, syringobulbia, retropharyngeal abscess, demyelinating disease, birth injury, and brainstem ischemia.

The most important lesion of the ninth nerve is glossopharyngeal (or vagoglossopharyngeal) neuralgia or “tic douloureux of the ninth nerve”. In this condition, the patient experiences attacks of severe lancinating pain originating in one side of the throat or tonsillar region and radiating along the course of the eustachian tube to the tympanic membrane, external auditory canal, behind the angle of the jaw, and adjacent portion of the ear. The pain may be brought on by talking, eating, swallowing, or coughing. It can lead to syncope, convulsions, and rarely to cardiac arrest because of stimulation of the carotid sinus reflex.

CRANIAL NERVE X—VAGUS

The vagus (in Latin means “wandering,” because of its wide distribution) is the longest and most widely distributed.

The vagus emerges from the medulla as a series of rootlets just below those of the glossopharyngeal.

CN X leaves the skull through the jugular foramen in the same neural sheath as the cranial root of CN XI and behind CN IX. In the jugular foramen, the nerve lies close to the jugular bulb, a dilatation of the internal jugular vein that houses the glomus jugulare (tympanic body). The glomus jugulare has functions similar to the carotid body.

Branches of cranial nerves: There are 10 major terminal branches that arise at different levels: (a) meningeal, (b) auricular, (c) pharyngeal, (d) carotid, (e) superior laryngeal, (f) recurrent laryngeal, (g) cardiac, (h) esophageal, (i) pulmonary, and (j) gastrointestinal.

Motor: The vagus, with a contribution from the bulbar portion of CN XI, supplies all the striated muscles of the soft palate, pharynx, and larynx except for the stylopharyngeus (CN IX) and tensor veli palatini (CN V).

Parasympathetic: The vagus is the longest parasympathetic nerve in the body and a vagal discharge causes bradycardia, hypotension, bronchoconstriction, bronchorrhea, increased peristalsis, increased gastric secretion, and inhibition of adrenal function. The vagal centers in the medulla that control these functions are themselves under the control of higher centers in the cortex and hypothalamus. Inhibition of vagal function produces the opposite effects.

Sensory: Both vagal ganglia are sensory. The superior ganglion primarily conveys somatic sensation, and most of its communication is with the auricular nerve. The inferior ganglion relays general visceral sensation and taste.

Normal functions mediated by CNs IX and X include swallowing, phonation, and airway protection and modulation.

Examination

Motor function: The character of the voice and the ability to swallow provide information about the branchiomotor functions of the vagus.

Clinical implications:

A unilateral vagal lesion causes weakness of the soft palate, pharynx, and larynx. Acute lesions may produce difficulty swallowing both liquids and solids and hoarseness or a nasal quality to the voice. Sensory change is anesthesia of the larynx due to involvement of the superior laryngeal nerve. The gag reflex is absent on the involved side. Autonomic reflexes (vomiting, coughing, and sneezing) are not usually affected.

Bilateral complete vagal paralysis is incompatible with life. It causes complete paralysis of the palate, pharynx, and larynx, with marked dysphagia and dysarthria; tachycardia; slow, irregular, and respiration; vomiting; and gastrointestinal atonia.

Disorders of Cranial Nerve X

Unilateral supranuclear lesions generally cause no dysfunction because of bilateral innervation.

Bilateral supranuclear lesions, as from pseudobulbar palsy, cause dysphagia and dysarthria.

Extrapyramidal disorders may produce difficulty with swallowing and talking. Patients with Parkinson's disease typically have a hypokinetic dysarthria. Laryngeal spasm with stridor may occur in Parkinson's disease.

Nuclear lesions bulbar ALS, syringomyelia, and some neoplasms, may cause fasciculations in the palatal, pharyngeal, and laryngeal muscles.

Infranuclear Extramedullary and intracranial involvement can occur in processes involving the meninges, extramedullary tumors, aneurysms, trauma, sarcoidosis, and skull fractures.

Lesions at the jugular foramen or in the retroparotid space usually involve some combination of IX, X, XI, XII, and the cervical sympathetics.

Palatal myoclonus: Seen in lesions at Mollaret triangle.

Jacobson's neuralgia: Involvement of tympanic branch of CN 9.

Recurrent laryngeal nerve palsy:

Causes:

- Unilateral:
 - Mitral stenosis
 - Bronchogenic carcinoma
 - Aortic aneurysm
 - Hodgkin's disease
- Bilateral:

- Guillain–Barré syndrome
- Thyroidectomy
- Lymphomas

CRANIAL NERVE XI—SPINAL ACCESSORY

The spinal accessory (SA) nerve, cranial nerve XI (CN XI), is actually two nerves that run together in a common bundle for a short distance [Fig. 6D(iii).68].

Cranial part (ramus internus): The smaller cranial portion is a special visceral efferent (SVE) accessory to the vagus. It emerges from the medulla laterally as four or five rootlets caudal to the vagal filaments. The cranial root runs to the jugular foramen and unites with the spinal portion, traveling with it for only a few millimeters to form the main trunk of CN XI. The cranial root communicates with the jugular ganglion of the vagus, and then exits through the jugular foramen separately from the spinal portion. It is distributed principally with the recurrent laryngeal nerve to sixth branchial arch muscles in the larynx.

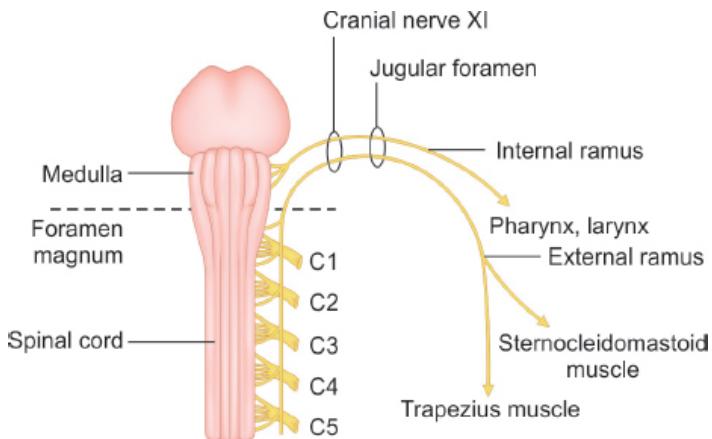


Fig. 6D(iii).68: Anatomy of spinal accessory nerve.

Spinal part (ramus externus): The major part of CN XI is the spinal portion. Its function is to innervate the sternocleidomastoid (SCM) and trapezius muscles. The fibers of the spinal root arise from SVE motor cells in the SA nuclei in the ventral horn from C2 to C5, or even C6. These unite into a single trunk, which ascends between the denticulate ligaments and the posterior roots. The nerve enters the skull through the foramen magnum, ascends the clivus for a short distance, and then curves laterally. The spinal root joins the cranial root for a short distance, probably receiving one or two filaments from it. It exits through the jugular foramen in company with CNs IX and X.

C1-2 supplies sternocleidomastoid.

C3-4 supplies trapezius.

Testing the Spinal Accessory Nerve

Cranial Part

The functions of the cranial portion of CN XI cannot be distinguished from those of CN X, and examination is limited to evaluation of the functions of the spinal portion.

Spinal Part

Testing SCM [Figs. 6D(iii).69 and 6D(iii).70]:

Testing one muscle at a time: To assess SCM power, have the patient turn the head fully to one side and hold it there, then try to turn the head back to midline, avoiding any tilting or leaning motion. The muscle usually stands out well, and its contraction can be seen and felt. Significant weakness of rotation can be detected if the patient tries to counteract firm resistance.

Testing two muscle at a time: The two SCM muscles can be examined simultaneously by having the patient flex his neck while the examiner exerts pressure on the forehead or by having the patient turn the head from side to side. Flexion of the head against resistance may cause deviation of the head toward the paralyzed side.



Fig. 6D(iii).69: Examination of sternocleidomastoid muscle (testing one muscle at a time).



Fig. 6D(iii).70: Examination of sternocleidomastoid (testing both muscles at a time).

Interpretation: With unilateral paralysis, the involved muscle is flat and does not contract or become tense when attempting to turn the head contralaterally or to flex the neck against resistance. Weakness of both SCMs causes difficulty in anteroflexion of the neck, and the head may assume an extended position.

Testing trapezius muscle (Fig. 6D(iii).71):

Inspection: With trapezius atrophy, inspection findings include:

- Depression or drooping of the shoulder contour
- Flattening of the trapezius ridge
- Sagging of the shoulder

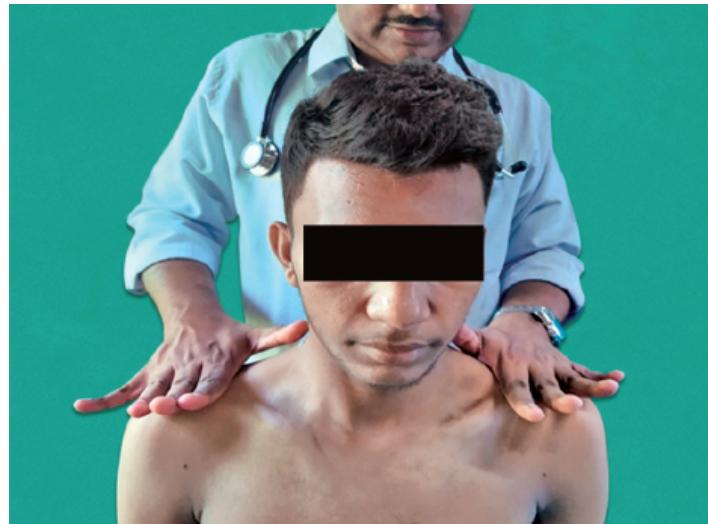


Fig. 6D(iii).71: Traditional method of assessing trapezius muscle (shrugging shoulders against resistance).

- The resting position of the scapula shifts downward
- The upper portion of the scapula tends to fall laterally while inferior angle moves inward (this scapular rotation and displacement are more obvious with arm abduction).

Palpation:

Traditional method: The strength of the trapezius is traditionally tested by having the patient shrug the shoulders against resistance. However, much of shoulder shrugging is due to the action of the levator scapulae.

Newer methods:

- **For upper trapezius:** Resisting the patient's attempt to approximate the occiput to the acromion. Impairment of upper trapezius function causes weakness of abduction beyond 90°.
- **For middle and lower trapezius:** Place the patient's abducted arm horizontally, palm up, and attempt to push the elbow forward. Muscle power should be compared on the two sides. Weakness of the middle trapezius muscle causes winging of the scapula.

Clinical implication: Weakness of the muscles supplied by CN XI may be caused by supranuclear, nuclear, or infranuclear lesions.

- **Supranuclear involvement:** Irritative supranuclear lesions may cause head turning away from the discharging hemisphere. This turning of the head (or head and eyes) may occur as part of a controversial, ipsiversive, or Jacksonian seizure and is often the first manifestation of the seizure. Extrapyramidal lesions may also involve the SCM and trapezius muscles, causing rigidity, akinesia, or hyperkinesis.
- **Nuclear involvement** of the SA nerve may occur in motor neuron disease, syringobulbia, and syringomyelia. In nuclear lesions, the weakness is frequently accompanied by atrophy and fasciculations.
- **Infranuclear or peripheral lesions**—either extramedullary but within the skull, in the jugular foramen, or in the neck—are the most common causes of impairment of function of the SA nerve. Tumors in the foramen magnum, lesions of the cerebellopontine angle, basal skull fractures, and meningitis.

“Dropped Head Syndrome”/Floppy Head Syndrome/Broken Neck Sign

This syndrome, characterized by weakness of the extensor muscles of neck with or without involvement of neck flexors, can be caused by:

- Myasthenia gravis
- Inflammatory myopathy—polymyositis
- Guillain–Barré syndrome
- Amyotrophic lateral sclerosis (ALS)/Bulbar polio
- Facio-scapulo-humeral dystrophy
- Neurotoxic snake bite/Organophosphorous compound poisoning.

CRANIAL NERVE XII—HYPOGLOSSAL NERVE

Function: CN XII supplies the intrinsic muscles, and all of the extrinsic muscles of the tongue except the palatoglossus.

Anatomy [Fig. 6D(iii).72]: Nucleus located in medial medulla. Distribution of fibers from rostral to caudal, the innervation is intrinsic tongue muscles, then genioglossus, hyoglossus, and styloglossus.

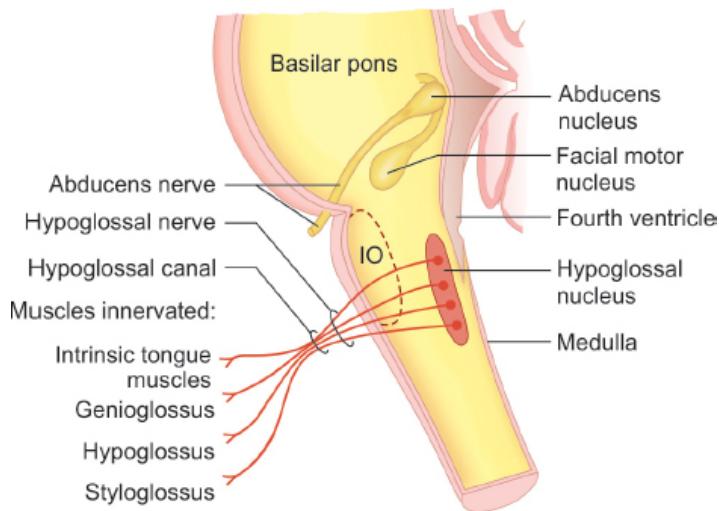


Fig. 6D(iii).72: Location of hypoglossal nerve.

Examination

The clinical examination of hypoglossal nerve function consists of evaluating the strength, bulk, and dexterity of the tongue—looking especially for weakness, atrophy, abnormal movements (particularly fasciculations), and impairment of rapid movements.

Inspection:

- **Tongue deviation:** To look for tongue deviation by asking the patient to protrude the tongue and also to move the tongue to either sides.
- **Fasciculations:** Ask the patient to open the mouth and with the tongue inside the mouth look for the fasciculations.

Palpation:

- Hold the tongue with gauze and palpate the tongue with gloved finger to examine the consistency of the tongue [Fig. 6D(iii).73].
- To examine the power of the tongue patient is instructed to push the tongue against the cheek while giving the counter resistance from outside [Fig. 6D(iii).74].

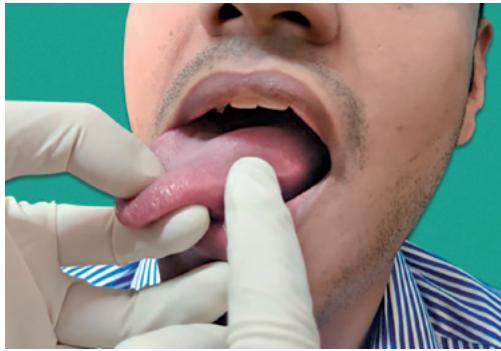


Fig. 6D(iii).73: Palpation of tongue.



Fig. 6D(iii).74: Examining the motor power of tongue.

Interpretation

On inspection:

- **Tongue deviation [Fig. 6D(iii).75]:** When unilateral weakness is present, the tongue deviates toward the weak side on protrusion because of the action of the normal genioglossus. And also there is impairment of the ability to deviate the protruded tongue toward the opposite side.
- **Fasciculations:** Presence of fasciculations suggests LMN paralysis of the 12th cranial nerve.

On palpation:

- **Small and stiff tongue:** Suggestive of UMN type of 12th nerve palsy.
- **Flabby tongue with fasciculations:** Suggestive of LMN type of 12th nerve palsy.

Other clinical aspects: The neck-tongue syndrome, consisting of pain in the neck and numbness or tingling in the ipsilateral half of the tongue on sharp rotation of the head, has been attributed to damage to lingual afferent fibers traveling in the hypoglossal nerve to the C2 spinal roots through the atlantoaxial space.



Fig. 6D(iii).75: Tongue deviation to the left suggestive of weakness of left hypoglossal muscle.

Bulbar palsy	Pseudobulbar palsy
<p>Etiology:</p> <ul style="list-style-type: none"> • Motor neuron disease • Syringobulbia • Guillain-Barré syndrome • Poliomyelitis • Subacute meningitis (carcinoma and lymphoma) • Neurosyphilis • Brainstem CVA 	<p>Etiology: The most common cause is bilateral CVAs affecting the internal capsule Other causes include:</p> <ul style="list-style-type: none"> • Multiple sclerosis • Motor neuron disease • High brainstem tumors • Head injury
<ul style="list-style-type: none"> • Bilateral damage or injury of the nerve nuclei of cranial nerves IX, X, XI, and XII • Lower motor neuron palsy of the respective muscles • Gag reflex—absent • Tongue—wasted, fasciculations • “Wasted, wrinkled, thrown into folds, and increasingly motionless” • Palatal movement—absent • Jaw jerk—absent or normal • Speech—nasal “Indistinct (flaccid dysarthria), lacks modulation, and has a nasal twang” • Emotions – normal • Other—signs of the underlying cause, e.g. limb fasciculations 	<ul style="list-style-type: none"> • Bilateral damage or injury of corticobulbar tracts to nerve nuclei of cranial nerves V, VII, X, XI, and XII • Upper motor neuron palsy of the respective muscles • Gag reflex—increased or normal • Tongue—spastic <p>“It cannot be protruded, lies on the floor of the mouth and is small and tight”</p> <ul style="list-style-type: none"> • Palatal movement—absent • Jaw jerk—increased • Speech—spastic: “A monotonous, slurred, high-pitched, ‘Donald Duck’, dysarthria” that “sounds as if the patient is trying to squeeze out words from tight lips”. “Hot potato voice” • Emotions—labile • Other—bilateral upper motor neuron (long tract) limb signs. Bilateral extensor plantar and bilateral exaggerated reflexes

MULTIPLE CRANIAL NERVE PALSIES

Cranial nerve	Cavernous sinus thrombosis	Superior orbital fissure syndrome	Orbital apex syndrome	Jaccoud's (retro-sphenoid space)syndrome	Petrous apex gradinigo syndrome	Tolosa-Hunt, lateral cavernous sinus syndrome	CP angle tumor	Vernet jugular foramen syndrome	Villaret, post-retroparotid syndrome	sy
II			✓	✓						
III	✓	✓	✓	✓		✓				

IV	✓	✓	✓	✓		✓					
V1	✓			✓	✓	✓	✓	✓			
V2	✓		✓	✓	✓						
V3				✓	✓						
VI	✓	✓	✓	✓	✓	✓	✓	✓			
VII									✓		
VIII								✓			
IX									✓	✓	
X									✓	✓	
XI									✓	✓	
XII											✓
Horner	✓										✓

NOTES

D(iv). MOTOR SYSTEM EXAMINATION

Motor system examination includes examination of:

1. Attitude of the limbs
2. Bulk/nutrition
3. Assessment of tone
4. Examination of power
5. Reflexes
6. Coordination
7. Gait

Reflexes, coordination, and gait have been discussed separately in the successive sections.

ATTITUDE

Attitude is the position of the limbs which it adopts when the patient is in resting position.

In a patient with hemiplegia	
Upper limb	Lower limb
<ul style="list-style-type: none">• Adduction at shoulder• Flexion at elbow• Semipronated• Thumb tucked into the palm	<ul style="list-style-type: none">• Extended at hip and knee• Externally rotated at hip• Foot inverted• Plantar flexed

Few common attitudes

Paraplegia	Bilateral lower limbs are: <ul style="list-style-type: none">• Extended at hip and knee• Externally rotated at hip• Foot inverted• Plantar flexed
Erb's palsy	On the affected side: <ul style="list-style-type: none">• Arm: Adducted and internally rotated• Forearm: Extended and pronated• Wrist: flexed• "Waiter's tip deformity"

MUSCLE BULK/NUTRITION

- Muscle bulk is assessed by inspection as well as measurements at corresponding sites in the extremities.
- Symmetry is important with consideration given to handedness and overall body habitus.
- Wasting is considered if there is >1 cm reduction on the dominant extremity and >2 cm in the nondominant extremity. In some areas, just inspection is adequate (thenar eminence, hypothenar eminence, shoulder) whereas in other areas (thighs, legs, arms and forearms) measurement is required.
- Measurements of the circumferences of the limb are done at corresponding areas at fixed distances from bony landmarks, which are part of that limb. Example: 10 cm below the olecranon [Fig.]

6D(iv).1], 10 cm above the medial humeral epicondyle [**Fig. 6D(iv).2**], 18 cm above the patella, and 10 cm below the tibial tuberosity.



Fig. 6D(iv).1: Measurement of bulk in the forearm.

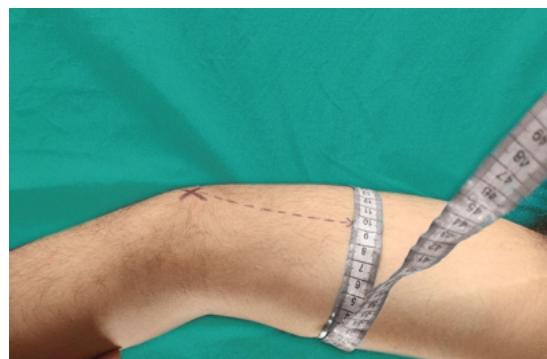


Fig. 6D(iv).2: Measurement of bulk in the arm.

Causes for Muscle Hypertrophy (Usually in the Calf) [Fig. 6D(iv).3]

True hypertrophy	Pseudohypertrophy (due to increased fat in muscle)
Exercise	<ul style="list-style-type: none">Duchene's muscular dystrophyBecker's muscular dystrophyMyotonia congenita—Thomson's diseaseKugelberg Welander spinal muscular atrophyHypothyroidism (infantile Hercules/Kocher–Debré–Semelaigne syndrome)Storage disorders
Localized muscle swelling —muscle hemorrhage, myositis ossificans, abscess, tumor, muscle rupture or cysts (cysticercosis)	



Fig. 6D(iv).3: Pseudohypertrophy of calf muscle.

Causes of Muscle Wasting

Generalized wasting	Proximal wasting	Distal wasting
<ul style="list-style-type: none"> Malignancy Cachexia Tuberculosis Thyrotoxicosis Addison's disease HIV/AIDS 	<ul style="list-style-type: none"> Motor neuron disease: Juvenile SMA (Kugelberg Welander) Muscular dystrophy: FSHD [Fig. 6D(iv).4], limb girdle dystrophy Inflammatory myopathies Brachial plexopathy Axillary neuropathy 	<ul style="list-style-type: none"> Anterior horn cell disease—polio, motor neuron disease Syringomyelia, intramedullary tumors Peripheral neuropathies—leprosy, Carpal tunnel syndrome Myotonic dystrophy Plexopathies—lower brachial plexus Arthritis—rheumatoid Disuse atrophy



Fig. 6D(iv).4: Proximal muscle wasting seen in facioscapulohumeral dystrophy (FSHD).

Causes of hand muscle wasting [Fig. 6D(iv).5]

Anterior horn cell disease	<ul style="list-style-type: none"> Motor neuron disease Syringomyelia Polio Spinal muscular atrophy
Nerve root	<ul style="list-style-type: none"> T1 compression by disc lesion. Pachymeningitis

	<ul style="list-style-type: none"> Cervical spondylosis Syphilitic amyotrophy C8-T1 tumors
Brachial plexus	<ul style="list-style-type: none"> Pancoast tumor Thoracic outlet obstruction, cervical rib Trauma, Klumpke's paralysis Other—Infiltration, irradiation
Lesions of peripheral nerve (ulnar or median)	<ul style="list-style-type: none"> Trauma Acute compression (coma, anesthesia, deep sleep) Chronic compression (entrapment) Acute ischemia (collagen vascular disease, diabetes)
Muscle disease	<ul style="list-style-type: none"> Myotonic dystrophy Distal myopathy—Welander, Udd, Miyoshi, Nonaka, Markesberry
Others	<ul style="list-style-type: none"> Rheumatoid arthritis Disuse atrophy Rarely—parietal lobe lesions



Fig. 6D(iv).5: Small muscle wasting of the hand.

The Split Hand Sign

- It is highly specific for amyotrophic lateral sclerosis (ALS).
- It is due to a lesion in the ulnar nerve or the lower trunk, which will cause predominant wasting of first dorsal interossei and hypothenar muscles with preserved thenar muscles (which are innervated by the median nerve).
- It is called split hand sign as it preferentially affects lateral part of the hand (abductor pollicis brevis and first dorsal interossei) and spares the medial part of the hand.
- This pattern of dissociated wasting does not correspond to a nerve or plexus or root distribution.
- This is in contrast to a C8-T1 root lesion, which will cause wasting of both thenar and hypothenar muscle as both median and ulnar nerves receive C8-T1 innervation.

MUSCLE TONE

Definition

Tone is defined as partial state of contraction of the muscle at rest which is demonstrated by resistance offered by the muscle to passive movement across the joint.

Tone is examined in the upper limb (wrist and elbow joint) and the lower limb (knee and ankle joint).

Testing for Tone in the Legs [Figs. 6D(iv).6 and 6D(iv).7]

- With the patient relaxed, place your hands on the thigh and roll the whole leg. Observe the movement of the foot
- With the patient in a supine position, place your hands behind the patient's knee, and lift the leg in a sudden motion. Observe if the heel drags along the bed. With normal muscle tone, the heel will drag along the surface of the bed. However, if there is an increased tone or spasticity, the foot may not make contact with the bed.
- Alternatively flex and extend the knee. Feel for the extensors during flexion and flexors during extension.

Testing for Tone in the Arms [Figs. 6D(iv).8 to 6D(iv).10]

- Lift the arm and let it drop. See the speed and smoothness.
- At the elbow, check for tone in biceps and triceps. Feel the biceps while extending the arm, and feel the triceps while flexing the arm.



Fig. 6D(iv).6: Assessment of tone in the lower limbs.



Fig. 6D(iv).7: Assessment of tone in the lower limbs.

- At the wrist, take the hand as if to shake it. First pronate and supinate the forearm. Then roll the hand around at the wrist. This demonstrates cog wheel rigidity [Fig. 6D(iv).11].



Fig. 6D(iv).8: Examining tone of triceps.



Fig. 6D(iv).9: Examining the tone of biceps.



Fig. 6D(iv).10: Examining the tone in the upper limb.



Fig. 6D(iv).11: Examining for cog wheeling/rigidity.

Abnormalities of Tone

Hypotonia—decreased tone.

Causes:

- Lower motor neuron (LMN) disease
- Cerebellar disease
- Hypothyroidism
- Upper motor neuron (UMN) disease in a state of neuronal shock
- Chorea
- Hypermagnesemia
- Down syndrome
- Anesthesia and muscle relaxants.

Hypertonia—increased tone. Two principal types:

1. Spasticity
2. Rigidity

	Spasticity	Rigidity
Synonym	Clasp-knife	Lead-pipe/Cog-wheel
Diseases	Pyramidal	Extrapyramidal
Pathophysiology	Increased gamma activity	Increased gamma and alpha activity
Description	<ul style="list-style-type: none"> • Tone increased in the initial part of movement followed by sudden release—clasp-knife effect* • Supination-pronation of the forearm will reveal the so-called supinator catch 	<ul style="list-style-type: none"> • Increased tone present continuously throughout the complete range of movement—lead-pipe • With associated tremors—cog-wheel**
Muscles involved	Anti-gravity muscles (flexors in the UL and extensors in the LL)	Both groups of muscles
Velocity	Velocity dependent (more with fast movements)	Velocity independent
Associated features	Hyperreflexia, extensor plantar	Tremors, bradykinesia

***Clasp-knife phenomenon:** The muscles at rest do not have excessive tone but a brisk stretch will produce a catch at about mid-length of the muscle followed by a sudden release of the catch and

relaxation of the muscle. The giving away or the release portion of the clasp-knife phenomenon is due to the increased firing of the inhibitory Golgi tendon organs. To elicit this phenomenon, the clinician extends the patient's knee using a constant velocity, but as the patient's knee nears full extension, the muscle tone of the quadriceps muscles increases dramatically and completes the movement, just as the blade of a pocket knife opens under the influence of its spring.

**Cog-wheel rigidity: Lead pipe rigidity superimposed with tremors (Negro sign).

Causes of hypertonia:

- UMN disease—pyramidal and extrapyramidal
- Tetanus
- Tetany
- Strychnine poisoning
- Tonic phase of seizure
- Catatonia (seen in schizophrenia where there is increased tone for all movements)

Paratonia—altered tone seen in psychiatric diseases and frontal lobe dysfunction which is characterized by inability to relax the muscle during muscle tone assessment. Can be of two types:

1. Oppositional paratonia (**Gegenhalten**)—where the subjects involuntarily resist passive movements
2. Facilitatory paratonia (**Mitgehen**)—where the subject involuntarily assists passive movement.

Paratonia is present in bilateral frontal lobe dysfunction and diffuse cerebellar disorders.

Myotonia—Slow relaxation of muscle after voluntary contraction or contraction provoked by muscle percussion. Examples: myotonic dystrophy, congenital myotonia, hypothyroidism, neuromyotonia congenita, Issac syndrome [Fig. 6D(iv).12].

Myoedema

Stationary muscle mounding after muscle percussion without electrical muscle activity is called myoedema. Myoedema is due to prolonged muscle contraction caused by delayed calcium reuptake by sarcoplasmic reticulum, following local calcium ion release brought out by percussion or pressure.

Can be seen in hypothyroidism, chronic debilitating diseases, severe cachexia as in TB.

MOTOR POWER

Prerequisites

- Explain the test and the movements you are planning to do clearly to the patient before performing the test.



Fig. 6D(iv).12: Demonstration of myotonia.

- Position the patient according to the muscle which is being tested.

State of Muscle during Examination

- Fully contracted muscle
 - Muscle is at maximum advantage (small muscle)
- Fully relaxed muscle
 - Muscle at maximum disadvantage (may detect mild degrees of weakness)
- Mid-contracted muscle
 - Most feasible method
 - Used for most large muscles

Qualitative Assessment of Weakness (MRC Grading)

- Grade 0—no contraction
Grade 1—Flicker or trace of contraction
Grade 2—active movement, with gravity eliminated
Grade 3—active movement against gravity
Grade 4—active movement against gravity and resistance
Grade 5—normal power
- Grades 4-, 4, and 4+ may be used to indicate movement against slight, moderate, and strong resistance, respectively.

Muscle of neck	
Flexion of neck (sternocleidomastoid/platysma)	The patient attempts to flex his neck against resistance while supporting the chest [Fig. 6D(iv).13]
Extensor of neck	The patient attempts to extend their neck against resistance; contraction of the trapezius and other extensor muscles can be seen and felt, and strength of movement can be judged [Fig. 6D(iv).14]
Upper limb	
Supraspinatus—C5	Patient initiates abduction of arm from side against resistance [Fig. 6D(iv).15]
Deltoid—C5	Patient holds his hand at 60° against resistance [Fig. 6D(iv).16]
Infraspinatus—C5	The patient flexes his elbow, examiner holds the elbow to his side, and then attempts external rotation of the forearm against resistance [Fig. 6D(iv).17]

Rhombooids—C5	With hands on hip ask the patient to force the elbow backward [Fig. 6D(iv).18]
Serratus anterior—C5, 6, 7	The patient pushes his arms forward against firm resistance [Fig. 6D(iv).19]
Pectoralis major—C6, 7, 8	<ul style="list-style-type: none"> Placing hand on hip and pressing inward, sternocostal part of muscle can be seen and felt to contract [Fig. 6D(iv).20] Raising the arm forward above 90° and attempting to adduct clavicular portion can be felt
Latissimus dorsi—C7	<ul style="list-style-type: none"> While palpating muscles ask the patient to cough Resist the patients attempt to adduct the arm when abducted to above 90° [Fig. 6D(iv).21]
Biceps—C5	Ask the patient to flex at the forearm with hand in supine position, against resistance [Fig. 6D(iv).22]
Brachioradialis—C5,6	The patient is asked to flex the elbow with the forearm midway between pronation and supination [Fig. 6D(iv).23]
Triceps—C7	The patient attempts to extend elbow against resistance [Fig. 6D(iv).24]
Extensor carpi radialis longus—C6, 7	The patient makes a fist and extends the wrist towards the radial side [Fig. 6D(iv).25]
Extensor carpi ulnaris—C7	The patient makes a fist and extends the wrist towards the ulnar side [Fig. 6D(iv).26]
Extensor digitorium—C7	The examiner attempts to flex the patient's extended fingers at the metacarpophalangeal joints [Figs. 6D(iv).27A and B]
Flexor carpi radialis—C6, 7	The examiner attempts to flex the wrist toward the radial side [Fig. 6D(iv).28]
Flexor carpi ulnaris—C8	Best seen while testing the abductor digiti minimi when it fixes its point of origin [Figs. 6D(iv).29A and B]
Abductor pollicis longus—C8	Patient maintains their thumb in the abduction against the examiner's resistance [Fig. 6D(iv).30]
Extensor pollicis brevis—C8	The patient attempts to extend the thumb while the examiner attempts to flex it at the metacarpophalangeal joint [Fig. 6D(iv).31]
Extensor pollicis longus—C8	The patient attempts to extend the thumb while the examiner attempts to flex it at the interphalangeal joint
Opponens pollicis—T1	The patient attempts to touch the little finger with the thumb [Fig. 6D(iv).32]
Abductor pollicis brevis—T1	Place an object between the thumb and base of forefinger to prevent full adduction Patient attempts to raise the edge of the thumb vertically against the resistance [Fig. 6D(iv).33]
Flexor pollicis longus—C8	Tested by attempting to extend the distal phalanx of the thumb against resistance, while holding the proximal phalanx [Fig. 6D(iv).34]
Adductor pollicis—T1	The patient attempts to hold a piece of paper between the thumb and the palmar aspect of forefinger and examiner tries to pull the paper [Fig. 6D(iv).35]
Lumbricals—C8, T1	The patient tries to flex the extended fingers at the metacarpophalangeal joints [Fig. 6D(iv).36]
Dorsal interossei	The patient attempts to keep the fingers abducted against resistance [Fig. 6D(iv).37]
First dorsal interossei and palmar interossei	Place the hand flat on table and the patient tries to abduct and adduct the forefinger against the resistance [Figs. 6D(iv).38 and 6D(iv).39]
Flexor digitorum sublimis—C8	The patient flexes the fingers at the proximal interphalangeal joint against resistance from the examiner's fingers placed on the middle phalanx [Fig. 6D(iv).40]
Flexor digitorum profundus—C8	The patient keeps his hand on a flat surface. The examiner holds the middle phalanx

	down; the patient flexes the distal phalanx against resistance [Fig. 6D(iv).41]
Flexor digiti minimi—T1	The back of hand is placed on the table and the little finger abducted against resistance. (often the only sign of an ulnar lesion)
Trunk muscles	
Abdominal muscles	The recumbent patient attempts to raise his head against resistance [Fig. 6D(iv).43]
Extensors of spine	The patient, lying prone, attempts to raise the head and upper part of the chest [Fig. 6D(iv).44]
Lower limb	
Iliopsoas—L1, 2, 3	The patient lies supine and attempts to flex the thigh against resistance [Fig. 6D(iv).45]
Adductor femoris—L5, S1 (Adductor magnus, longus and brevis)	The patient attempts to adduct the leg against resistance [Fig. 6D(iv).46]
Gluteus medius and minimus—L2, 3	Patient in prone, flexes the knee, and then forces the foot outward against resistance [Fig. 6D(iv).47]
Gluteus maximus—L5, S1	Patient in prone raises the thigh against resistance with the knee flexed to minimize the contribution from the hamstrings [Fig. 6D(iv).48]
Hamstrings—L4, 5, S1, 2 (biceps, semimembranosus, and semitendinosus)	Patient in prone and attempts to flex the knee against resistance [Fig. 6D(iv).49]
Quadriceps femoris—L3, 4	Patient is supine and extends the knee against resistance [Fig. 6D(iv).50]
Tibialis anterior—L4, 5	The patient dorsiflexes the foot against the resistance of examiner [Fig. 6D(iv).51]
Tibialis posterior—L4	The patient plantar flexes the foot slightly and then tries to invert it against resistance [Fig. 6D(iv).52]
Peronei—L5, S1	The patient everts the foot against resistance [Fig. 6D(iv).53]
Extensor digitorum longus—L5	Patient asked to dorsiflex the foot against resistance [Fig. 6D(iv).54]
Flexor digitorum longus—S1, 2	Patient asked to flex the terminal phalanges against resistance [Fig. 6D(iv).55]
Extensor hallucis longus—L5, S1	Patient asked to dorsiflex the great toe against resistance [Fig. 6D(iv).56]
Extensor digitorum brevis—S1	The patient dorsiflexes the toes against resistance [Fig. 6D(iv).57]



Fig. 6D(iv).13: Flexion of neck (sternocleidomastoid/platysma).



Fig. 6D(iv).14: Extensor of neck.



Fig. 6D(iv).15: Supraspinatus—C5. Patient initiates abduction of arm from side against resistance.



Fig. 6D(iv).16: Deltoid C5.



Fig. 6D(iv).17: Infraspinatus—C5.



Fig. 6D(iv).20: Pectoralis major—C6, 7, 8.



Fig. 6D(iv).18: Rhomboids—C5.

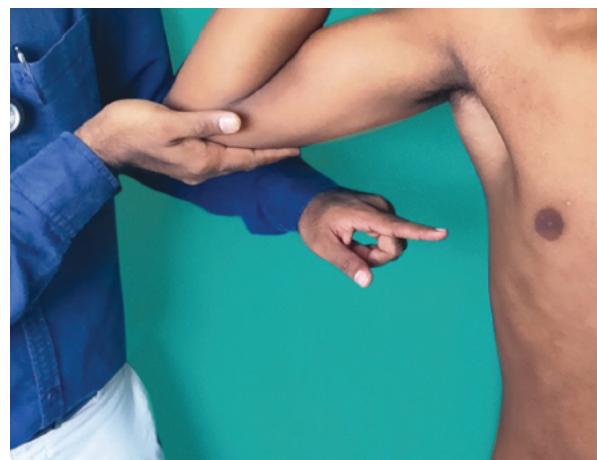


Fig. 6D(iv).21: Latissimus dorsi—C7.

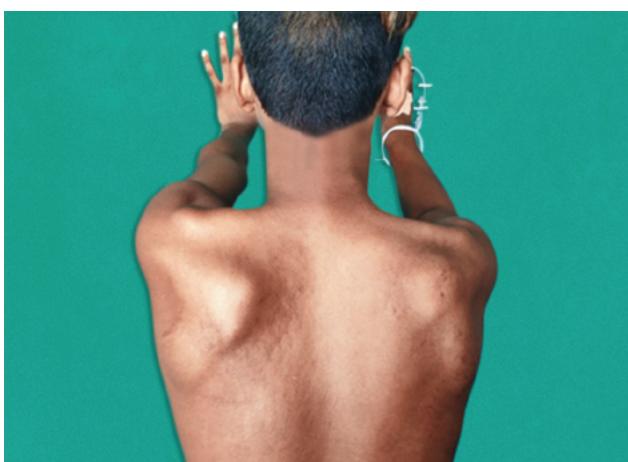


Fig. 6D(iv).19: Serratus anterior—C5, 6, 7.



Fig. 6D(iv).22: Biceps—C5.



Fig. 6D(iv).23: Brachioradialis—C5, 6.



A



Fig. 6D(iv).24: Triceps—C7.



Figs. 6D(iv).27A and B: Extensor digitorum—C7.



Fig. 6D(iv).25: Extensor carpi radialis longus—C6, 7.



Fig. 6D(iv).26: Extensor carpi ulnaris—C7.



Fig. 6D(iv).28: Flexor carpi radialis—C6, 7.



A



B

Figs. 6D(iv).29A and B: Flexor carpi ulnaris—C8.



Fig. 6D(iv).31: Thumb extension.



Fig. 6D(iv).32: Opponens pollicis—T1.



Fig. 6D(iv).30: Thumb abduction.



Fig. 6D(iv).33: Abductor pollicis brevis—T1.

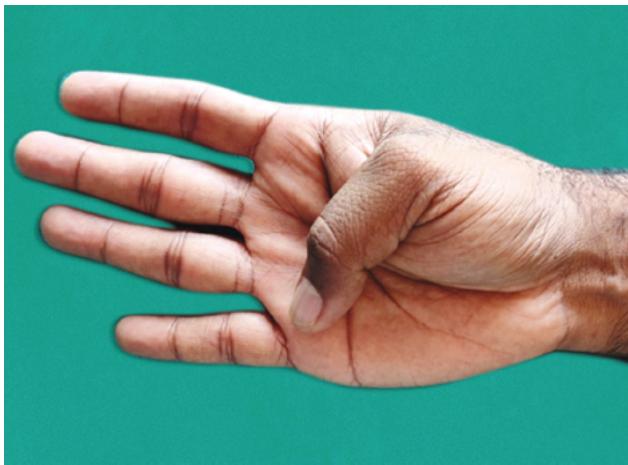


Fig. 6D(iv).34: Thumb flexion.



Fig. 6D(iv).37: Dorsal interossei.



Fig. 6D(iv).35: Thumb adduction.



Fig. 6D(iv).38: Palmar interossei.

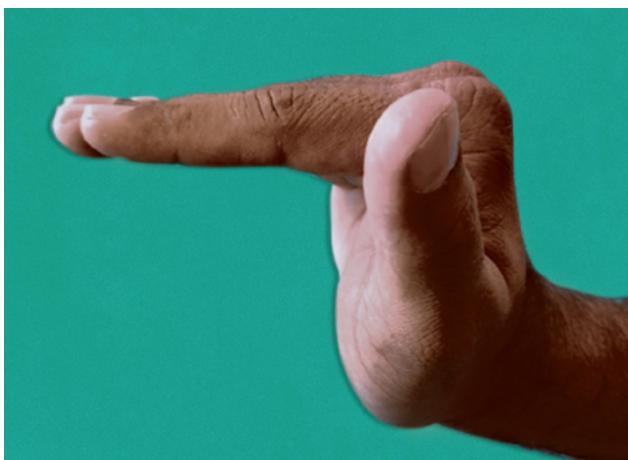


Fig. 6D(iv).36: Lumbricals—C8, T1.



Fig. 6D(iv).39: Card test for palmar interossei.

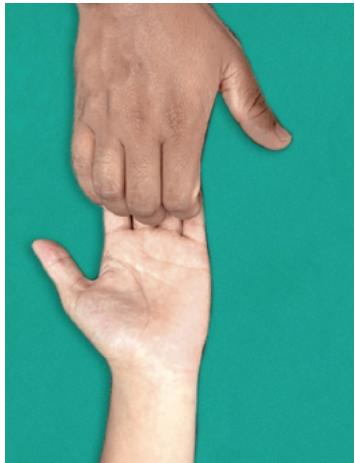


Fig. 6D(iv).40: Flexor digitorum sublimis.



Fig. 6D(iv).43: Abdominal muscles T5–L1.

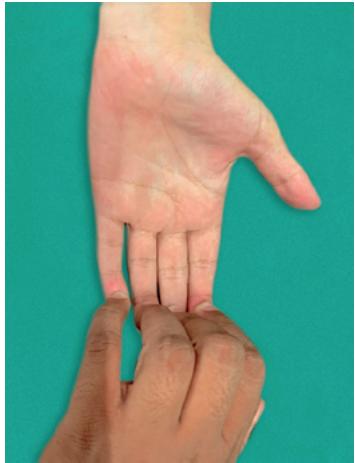


Fig. 6D(iv).41: Flexor digitorum profundus.



Fig. 6D(iv).44: Extensors of spine.



Fig. 6D(iv).42: Abductor digiti minimi.



Fig. 6D(iv).45: Iliopsoas—L1, 2, and 3.



Fig. 6D(iv).46: Adductor femoris—L5, S1.

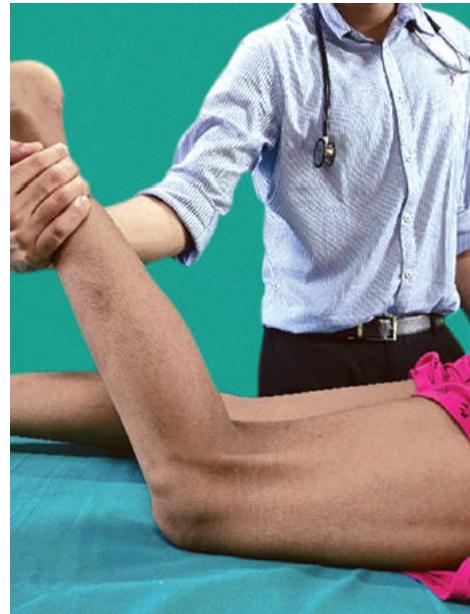


Fig. 6D(iv).49: Hamstrings—L4, 5, S1, 2 (biceps, semimembranosus, and semitendinosus).

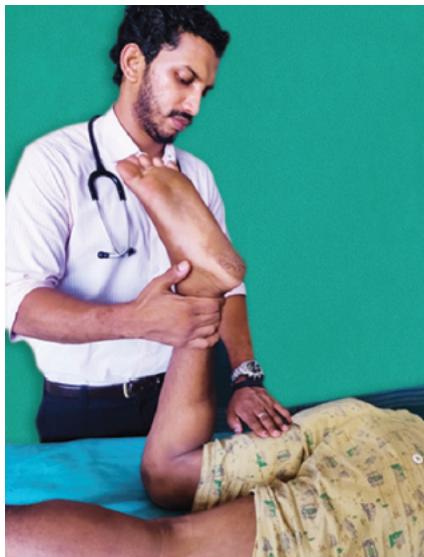


Fig. 6D(iv).47: Gluteus medius and minimus—L2, 3.

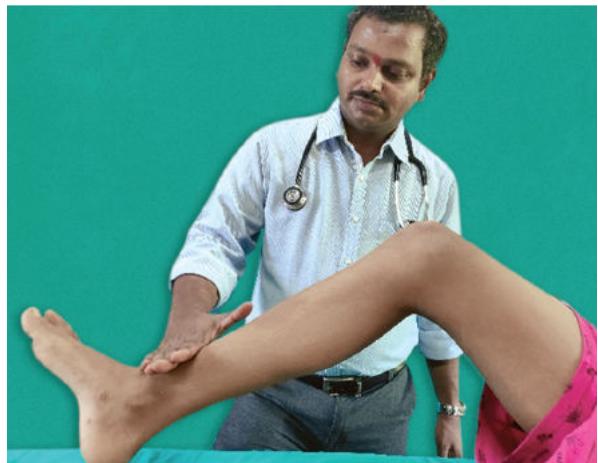


Fig. 6D(iv).50: Quadriceps femoris—L3, 4.



Fig. 6D(iv).48: Gluteus maximus—L5, S1.



Fig. 6D(iv).51: Tibialis anticus—L4, 5.



Fig. 6D(iv).52: Tibialis posticus—L4.



Fig. 6D(iv).55: Flexor digitorum longus—S1, 2.



Fig. 6D(iv).53: Peronei—L5, S1.

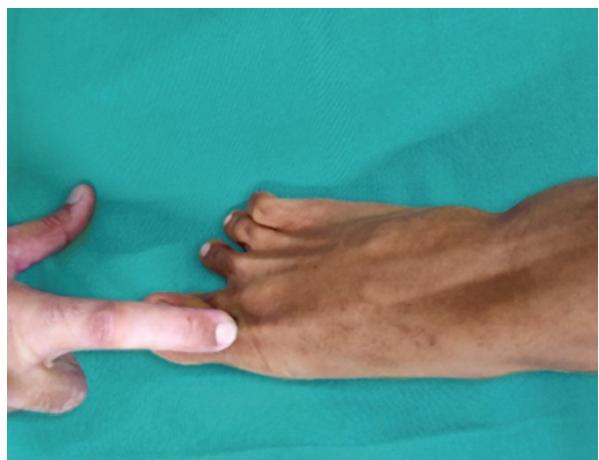


Fig. 6D(iv).56: Extensor hallucis longus—L5, S1.



Fig. 6D(iv).54: Extensor digitorum longus—L5.



Fig. 6D(iv).57: Extensor digitorum brevis—S1.

EXAMINATION FOR SUBTLE HEMIPARESIS [FIG. 6D(IV).58]

1. Pronator drift (Barre's sign)

- The patient stretches out both arms directly in front of him or her with palms upright (i.e. forearms supinated) and closes his or her eyes.
- This position is held for 20–30 seconds.

Normal response:

- Palm will remain flat, elbows straight and the limbs horizontal OR
- Symmetrical deviation from this position (i.e. on both the sides—dominant hand may pronate slightly more than the non-dominant hand)

Positive pronator drift: Components of pronator drift as mentioned above are seen in the weaker side (asymmetric response) which indicates a lesion in contralateral cortex

- Positive with eyes open: Motor deficit
- Positive with eyes closed: Sensory deficit (posterior column)
- Outward and upward drift: Cerebellar drift
- “Updrift” (involved arm rising overhead without patient awareness): Parietal lobe lesions (loss of position sense)
- Drift without pronation: Functional upper limb paresis (conversion disorder)

2. Forearm rolling test [Fig. 6D(iv).59]

- The patient bends each elbow and places both forearms parallel to each other.
- He or she then rotates the forearms about each other, first in one direction and then the other.
- In the abnormal response, the forearm contralateral to the lesion appears fixed while the other arm rotates around it.

3. Rapid finger tapping test

- The patient rapidly taps the thumb and index finger repeatedly at a speed of about two taps per second.
- Hemispheric lesions cause the contralateral finger and thumb to tap more slowly and with diminished amplitude.

4. Foot tapping test

- The seated patient taps one forefoot at a time for 10 seconds on the floor, as fast as possible, while the heel maintains contact with the floor.
- A discrepancy of more than five taps between the left and right foot indicates cerebral disease contralateral to the slower foot.

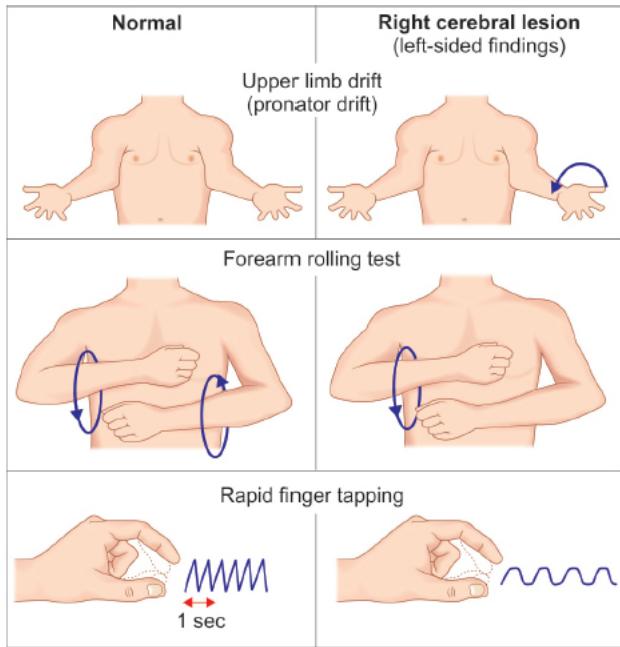


Fig. 6D(iv).58: Examination for subtle hemiparesis.

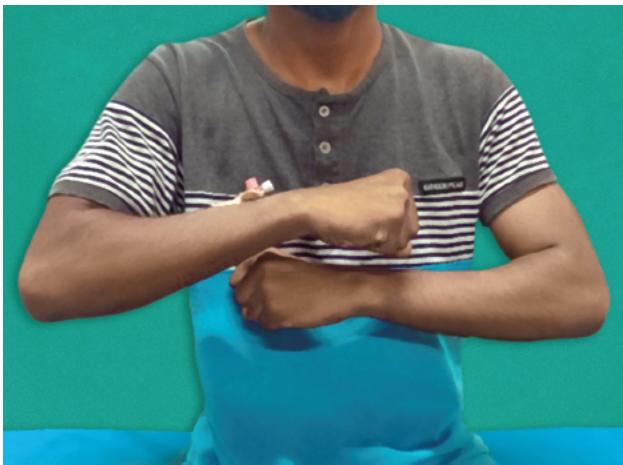


Fig. 6D(iv).59: Forearm rolling test.

D(v). REFLEXES

DEFINITION

A reflex is an involuntary response to a sensory stimulus.

MECHANISM OF REFLEX GENERATION [FIG. 6D(V).1]

Afferent impulses arising in a sensory organ produce a response in the effector organ. The response can be sensory, motor or autonomic.

It has two components:

Segmental component	Suprasegmental component
It consists of a local reflex center in the spinal cord or brainstem and its afferent and efferent connections	It is made up of descending central pathways that control, modulate, and regulate the segmental activity
	Diseases may increase the activity of some reflexes, decrease activity of others, and causes reflexes to appear that are not normally seen

TYPES OF REFLEXES

1. Deep tendon reflexes (monosynaptic reflex)
2. Superficial reflex (polysynaptic reflex)
3. Plantar reflex
4. Latent reflex
5. Primitive reflexes
6. Inverted and perverted reflexes.

GRADING OF REFLEXES (FOR DTR'S) NINDS SCALE

Absent reflex (even after reinforcement)	Grade 0
Present but diminished	Grade 1+
Normal	Grade 2+
Increased but not necessarily to pathologic degree	Grade 3+
Markedly hyperactive, pathologic, often with extrabeats or accompanying sustained clonus	Grade 4+

REINFORCEMENT MECHANISM AND METHODS

Mechanism

Normally, when a muscle spindle is stimulated two kinds of responses are seen via the following nerves:

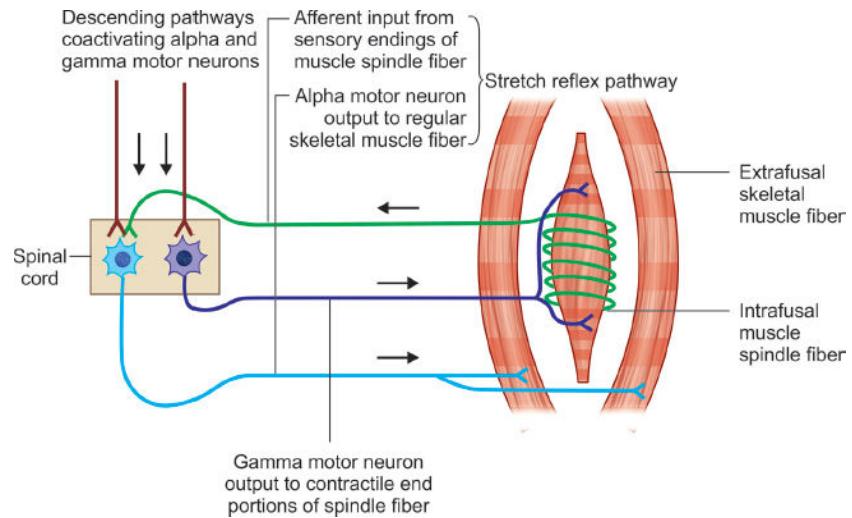


Fig. 6D(v).1: Schematic representation of innervation of muscle fiber and pathways.

Alpha motor neurons	Gamma motor neuron*	Inhibitory neuron
Causes: contraction of Extrafusal fibers of muscle	Causes: contraction of intrafusal fibers of muscle	Causes: inhibition of reciprocal muscle contraction

*Normally gamma motor neurons are under the inhibitory control of upper motor neurons and reinforcement maneuvers remove the inhibitory effect on gamma motor neurons [Fig. 6D(v).1].

Note: Mnemonic—AntiEpileptics cause gastro intestinal disturbance. (**A**: Alpha neuron, **E**: Extrafusal fibers), (**G**: Gamma neuron, **I**: Intrafusal fibers).

Reinforcement Maneuvers for Deep Tendon Reflexes (DTRs)

Distraction	Talk to the patient and cause diversion of thought process
Clenching the teeth or clenching the fist of the other arm [Fig. 6D(v).2]	Traditionally done for upper limb
Jendrassik maneuver (interlocking the flexed fingers of the two hands and pull one against each other) [Fig. 6D(v).3]	Preferably done for lower limb



Fig. 6D(v).2: Clenching the teeth for reinforcement of upper limb reflexes.

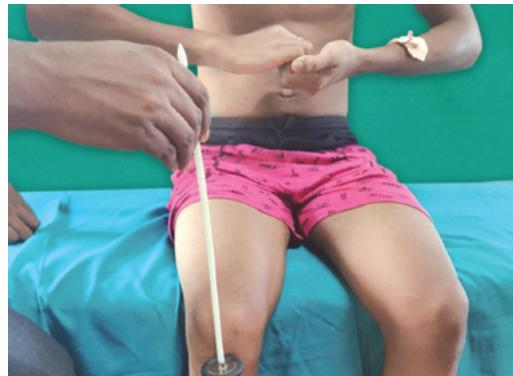


Fig. 6D(v).3: Jendrassik maneuver for reinforcement of lower limb reflexes.

DEEP TENDON REFLEXES

These are monosynaptic reflexes.

Prerequisite for examination:

- Good knee hammer (preferably Queen Square reflex hammer)
- Expose adequately the muscle to be tested
- Make sure patient is not anxious
- The muscle should be placed in optimum position, slightly on stretch, but with plenty of room for contraction.

The most commonly used specialized reflex hammers are grouped into three types by the shape of the head: triangular/tomahawk shaped (Taylor), T-shaped (Tromner, Buck), or circular (Queen square, Babinski)

Tromner neurological reflex hammer



Taylor hammer



Babinski neurological reflex hammer



Queen square neurological reflex hammer	
Buck neurological reflex hammer	

Reflex	Root value
Biceps	C5C6 (musculocutaneous nerve)
Supinator (brachioradialis)	C5C6 (radial nerve)
Triceps	C7C8 (radial nerve)
Knee	L3L4 (femoral nerve)
Ankle	S1S2 (medial popliteal nerve)
Mnemonic—S1,2: L3,4: C5,6:C7,8 (in sequence from below)	
Few others	
Pectoral	C5-T1 (medial and lateral pectoral nerves)
Finger flexion	C6-T1 (median nerve)

Reflex	Method of elicitation	Normal response
Biceps [Figs. 6D(v).4A to C]	Press the forefinger gently on the biceps tendon in the antecubital fossa and then strike the finger with the hammer	Flexion of the elbow with visible contraction of the biceps muscle
Supinator [Figs. 6D(v).5A to C]	Strike the lower end of the radius about 5 cm above the wrist and watch for the movement of forearm and fingers	Contraction of brachioradialis and flexion of elbow
Triceps [Figs. 6D(v).6A to D]	By holding the patient's hand draw the arm across the trunk and allow it to lie loosely in the new position. Then strike the triceps tendon 5 cm above the elbow	Extension of elbow with visible contraction of triceps muscle
Knee [Figs. 6D(v).7A to C]	For right-handed examiner, the left arm is under both the knees in order to flex them together and tap the patellar tendon lightly on each side and compare the movements of lower leg and of quadriceps muscle	Extension of the knee and visible contraction of the quadriceps (in case of lower leg amputation keep finger just above the patella with legs extended and strike it in peripheral direction and look for upward pull of patella)
Ankle [Figs. 6D(v).8A to E]	<ul style="list-style-type: none"> Patient's leg should be externally rotated and slightly flexed at the knee. Examiner uses the left hand to dorsiflex the foot. For the left leg move to the other side of the bed 	Plantar flexion of foot and contraction of gastrocnemius

	<ul style="list-style-type: none"> The Achilles tendon is then struck 	
Few others		
Pectoral [Fig. 6D(v).9]	With patients arm in the mid position between adduction and abduction hook your index finger on the tendon of the pectoralis major muscle in the anterior fold of axilla and strike with hammer	Adduction of the arm and visible contraction of the pectoralis major
Finger flexion test [Fig. 6D(v).10]	Allow the patient's hand to rest palm upwards, the fingers slightly flexed. The examiner interlocks his fingers with patient's fingers and strikes them with the hammer	Slight flexion of all the fingers and of the interphalangeal joint of the thumb



Fig. 6D(v).4A: Demonstration of biceps reflex (right hand).



Fig. 6D(v).4B: Demonstration of biceps reflex supine position (right hand).



Fig. 6D(v).4C: Demonstration of biceps reflex (left hand).



Fig. 6D(v).5C: Demonstration of supinator reflex in supine position.



Fig. 6D(v).5A: Demonstration of supinator reflex (right).



Fig. 6D(v).6A: Demonstration of triceps reflex (right hand).



Fig. 6D(v).5B: Demonstration of supinator reflex (left).



Fig. 6D(v).6B: Demonstration of triceps reflex (right hand) in sup position.

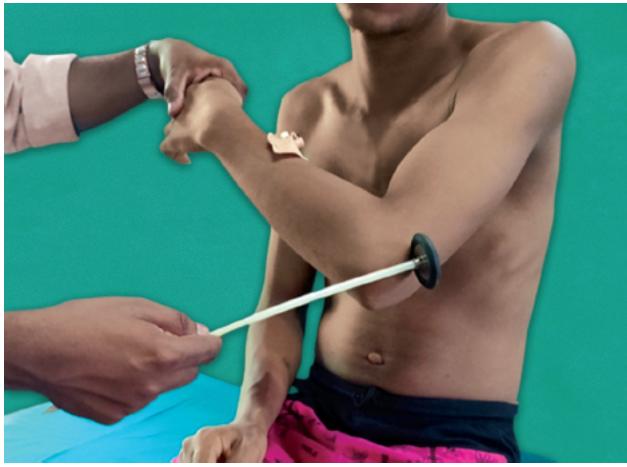


Fig. 6D(v).6C: Demonstration of triceps reflex (left hand).



Fig. 6D(v).7B: Demonstration of right knee jerk in supine position.



Fig. 6D(v).6D: Demonstration of triceps reflex (left hand) in supine position.



Fig. 6D(v).7C: Demonstration of knee jerk (for comparing both sides).



Fig. 6D(v).7A: Demonstration of knee jerk sitting position (for pendular movement).



Fig. 6D(v).8A: Demonstration of ankle reflex of right leg.



Fig. 6D(v).8B: Demonstration of ankle reflex of left leg.



Fig. 6D(v).8E: Demonstration of ankle reflex with foot dangling c
the edge of table.



Fig. 6D(v).8C: Demonstration of ankle reflex of left leg.

Fig. 6D(v).9: Demonstration of pectoral reflex.



Fig. 6D(v).8D: Demonstration of ankle reflex in prone position.



Fig. 6D(v).10: Demonstration of finger flexion reflex.

Clonus

Clonus is a series of rhythmic involuntary muscular contractions induced by the sudden passive stretching of a muscle or tendon.

Clonus	Demonstration
Ankle clonus [Figs. 6D(v).12A and B]	Examiner supports the leg, preferably with one hand under the knee, grasps the foot from below with the other hand, and quickly dorsiflexes the foot while maintaining slight pressure on the sole at the end of the dorsiflexion <ul style="list-style-type: none"> The leg and foot should be well relaxed, the knee and ankle in moderate flexion, and the foot slightly everted Right ankle clonus is examined by standing on the right side of the patient and left ankle clonus by standing on the left side Unsustained clonus fades away after a few beats; sustained clonus persists as long as the examiner continues to hold slight dorsiflexion pressure on the foot
Patellar clonus [(Figs. 6D(v).11A and B)]	Examiner grasps the patella between index finger and thumb and executes a sudden, sharp, downward thrust, holding downward pressure at the end of the movement
Wrist clonus	Sudden passive extension of the wrist produces wrist clonus



Fig. 6D(v).11A: Demonstration of right patellar clonus.



Fig. 6D(v).11B: Demonstration of left patellar clonus.



Fig. 6D(v).12A: Demonstration of right ankle clonus.



Fig. 6D(v).12B: Demonstration of left ankle clonus.

SUPERFICIAL REFLEXES

These are the responses to stimulation of either the skin or mucous membrane.

Clinical Significance

Superficial reflexes are abolished by pyramidal tract lesions.

Superficial reflex	Deep tendon reflex
Polysynaptic reflexes	Monosynaptic reflexes
Respond slowly	Faster response
Latency is longer	Latency is slower
Fatigue easily	Fatigue slowly
Not as consistently present as deep tendon reflexes	Consistently present
Abolished by pyramidal tract lesions	Exaggerated by pyramidal tract lesions

Superficial reflex	Elicitation
Corneal (cranial nerve V and VII)	Lightly touching the upper cornea with wisp of cotton or tissue, brought in from the side so the patient cannot see
Abdominal [Fig. 6D(v).13] • Epigastric (T6-T9) • Mid abdominal (T9-T11) • Hypogastric (T11-L1)	Stimulus is delivered by stroking the abdominal wall (preferably towards the umbilicus) and watch for contractions
Cremasteric [Fig. 6D(v).14] (L1, L2)	Stroking the skin in upper inner aspect of thigh and watch for the upward movement of testes in scrotum
Anal reflex (S2, S3)	Contraction of external sphincter in response to stroking the skin or mucous membrane in the perianal region
Bulbocavernosus reflex (S2, S3) [Fig. 6D(v).15]	Contraction of anal sphincter which is best appreciated by a gloved finger in the rectum on stimulation of glans penis or clitoris



Fig. 6D(v).13: Demonstration of abdominal reflex.

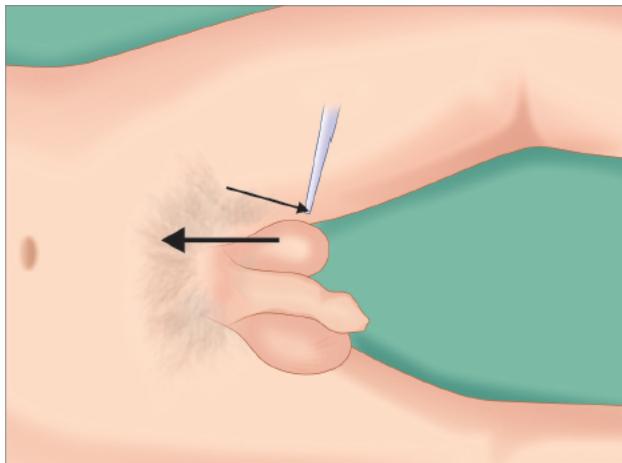


Fig. 6D(v).14: Direction of stimulus and movement of testes in cremasteric reflex.

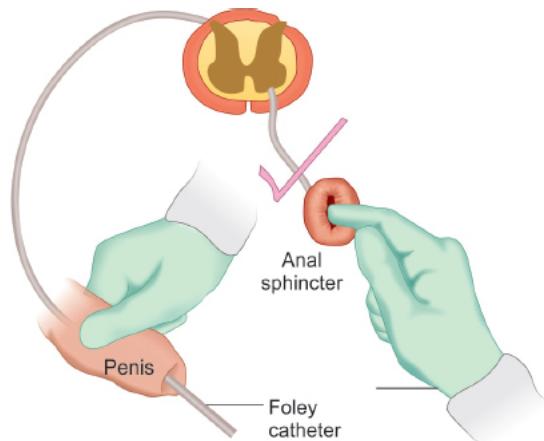


Fig. 6D(v).15: Pictorial representation of bulbocavernosus reflex.

PLANTAR REFLEX AND VARIATIONS

Plantar Reflex

Stroking the plantar surface of foot from the heel forward is normally followed by plantar flexion of foot and toes.

Babinski Sign

It is the pathologic variation of plantar reflex (i.e. extensor plantar response). It is part of primitive flexion reflex. In higher vertebrates, the flexion response includes flexion at hip, flexion at knee, and dorsiflexion of ankle (all of which help in removing the threatened part from danger). Normally the descending motor pathway suppresses the primitive flexion response.

Positioning of patient [Fig. 6D(v).16]	<ul style="list-style-type: none"> Best position is supine Knee must be extended Heels should rest on the bed
Prerequisites	Rule out ankylosis of great toe
Stimulating agent	<ul style="list-style-type: none"> Applicator stick Blunt key

	<ul style="list-style-type: none"> • Hand of reflex hammer • Broken tongue blade • Thumb nail
Strength of stimulus	Variable strength with strong stimulus for thick soles and minimal stimulation when response is strongly extensor
Site of stimulus	<ul style="list-style-type: none"> • Reflexogenic area of S1 • Stimulus should begin near the heel on the lateral aspect of sole and carried up to metatarsophalangeal joint of little toe and then carried medially falling short of 1st metatarsophalangeal joint [Fig. 6D(v).17]
Normal response	Flexion of the great toe and other toes
Abnormal response (Babinski sign)	<ul style="list-style-type: none"> • Dorsiflexion of great toe and small toes • Fanning of toes • Dorsiflexion of ankle • Flexion of knee joint • Flexion at hip joint • Contraction of tensor fascia lata
Reinforcement of plantar reflex	By asking patient to rotate the head to opposite side



Fig. 6D(v).16: Position of leg for demonstration of plantar reflex.



Fig. 6D(v).17: Direction of stimuli for demonstrating the plantar reflex.

Variants of Plantar Response

Equivocal response	<ul style="list-style-type: none"> Rapid extension followed by flexion Only great toe extension Extension of great toe with flexion of fingers No response to the plantar stimulus Flexion at hip and knee, but no movement of toes
Minimal plantar response	<ul style="list-style-type: none"> No toe movement Contraction of tensor fascia lata with mild internal rotation and abduction of hip
Pseudo Babinski	<ul style="list-style-type: none"> Voluntary extension of great toe due to hyperesthesia or strong painful stimulus Dystonic posturing of great toe

Other method of obtaining plantar reflex

Method	Elicitation
Chaddock [Fig. 6D(v).18]	<ul style="list-style-type: none"> Elicited by stimulating the lateral aspect of the foot, not the sole, beginning about under the lateral malleolus near the junction of the dorsal and plantar skin, drawing the stimulus from the heel forward to the small toe The Chaddock is the only alternative toe sign that is truly useful It may be more sensitive than the Babinski but is less specific It produces less withdrawal than plantar stimulation
Reverse Chaddock	The stimulus moves from the small toe toward the heel
Oppenheim [Fig. 6D(v).19]	<ul style="list-style-type: none"> Dragging the knuckles heavily down the anteromedial surface of the tibia from the infrapatellar region to the ankle. The response is slow and often occurs toward the end of stimulation
Shaeffer's sign [Fig. 6D(v).20]	Deep pressure on Achilles tendon
Gordon's sign [Fig. 6D(v).21]	Squeezing of calf muscles
Bing's sign [Fig. 6D(v).22]	Pricking dorsum of foot with a pin
Moniz' sign [Fig. 6D(v).23]	Forceful passive plantar flexion at ankle

Throckmorton's sign	Percussing over dorsal aspect of metatarsophalangeal joint of great toe just medial to EHL tendon
Stransky	Small toe forcibly abducted, then released
Szapiro	Pressure against dorsum of second through fifth toes, causing firm passive plantar flexion while stimulating plantar surface of foot
Strümpell's phenomenon	Forceful pressure over anterior tibial region
Cornell response	Scratching dorsum of foot along inner side of EHL tendon

Combining two methods may elicit minimal reflexes

[Fig. 6D(v).24]



Fig. 6D(v).18: Chaddock's sign.



Fig. 6D(v).19: Openheim's technique.



Fig. 6D(v).20: Shaeffer's technique.



Fig. 6D(v).21: Gordon's technique.



Fig. 6D(v).22: Bing's sign.



Fig. 6D(v).23: Moniz's sign.



Fig. 6D(v).24: Eliciting plantar by simultaneous stimulus from Openheim's and plantar strike.

LATENT REFLEXES OF UPPER LIMB

Reflex	Elicitation
Wartenberg's reflex [Fig. 6D(v).25]	Patient's fingers are interlocked with examiner's fingers and pulled apart. Normally thumb extends. However in pyramidal lesions thumb is adducted and flexed. This sign is equivalent of Babinski of lower limb
Hoffmann's reflex [Fig. 6D(v).26]	Flexion of the interphalangeal joint of middle finger of patient produces flexion response in other fingers along with adduction of thumb
Tromner's reflex [Fig. 6D(v).27]	Examiner holds the patient's partially extended middle finger, letting the hand dangle, then, with the other hand, thumps or flicks the finger pad. The response is the same as that in the Hoffmann test



Fig. 6D(v).25: Wartenberg's sign.



Fig. 6D(v).26: Hoffmann's reflex.



Fig. 6D(v).27: Tromner's reflex.

PRIMITIVE REFLEXES

Reflex	Elicitation
Glabellar tap (Myerson's sign) [Fig. 6D(v).28]	Repetitive tapping of the forehead between the eyebrows causing blinking, which usually stops within few taps. However if blinking persists, it suggests positive frontal release sign. <i>Note:</i> To avoid visual stimulus bring the hand from above and behind
Palmomental reflex of Marinesco– Radovici [Fig. 6D(v).29]	<ul style="list-style-type: none"> Stroke the thenar eminence in a proximal to distal direction using a sharp object such as the pointed end of a reflex hammer, key, paper clip, or fingernail and watch for twitch of chin muscle This reflex does not have any localizing value, and is commonly seen in elderly patients with degenerative disease of the cortex
Sucking reflex [Fig. 6D(v).30]	Sucking reflexes may be seen in response to tactile stimulation in the oral region, or in response to the insertion of an object (for example, a spatula) into the mouth
Rooting reflex [Fig. 6D(v).31]	Rooting responses are seen when the mouth turns towards an object gently stroking the cheek (tactile rooting), or towards an object (for example, tendon hammer) brought into the patient's field of view (visual rooting)
Pout and snout reflex	The snout reflex is present when the lips pucker in response to gentle pressure over the nasal philtrum

[Fig. 6D(v).32]

Grasp reflex
[Fig. 6D(v).33]

- If the examiner's fingers are placed in the patient's hand, especially between the thumb and forefinger, or if the palmar skin is stimulated gently, there is slow flexion of the digits
- The patient's fingers may close around the examiner's fingers



Fig. 6D(v).28: Glabellar tap.



Fig. 6D(v).29: Palmomental reflex.



Fig. 6D(v).30: Sucking reflex.



Fig. 6D(v).31: Rooting reflex.



Fig. 6D(v).32: Pout reflex.



Fig. 6D(v).33: Grasp reflex.

INVERTED AND PERVERTED REFLEXES

Reflex	Description and example
Inverted reflex	<p>Contractions opposite to that of expected For example:</p> <p>An inverted brachioradialis reflex:</p> <ul style="list-style-type: none"> • When the supinator reflex elicits finger flexion and not elbow flexion • Is associated with an absent biceps jerk and an exaggerated triceps jerk • Is indicative of a spinal cord lesion at C5 or C6, e.g. due to trauma, syringomyelia, or disc prolapse <p>Inversion of biceps reflex</p> <ul style="list-style-type: none"> • On eliciting bicep reflex the following are noticed: <ul style="list-style-type: none"> – There is no flexion at the elbow – But instead there is extension at the elbow due contraction of the triceps muscle • Presence of this reflex indicates that the lesion is at the level of C5 segment <p>Inversion of triceps reflex</p> <p>With disc protrusions at C6/7 there is a “paradoxical triceps reflex” with forearm muscles acting to flex the elbow against no triceps resistance</p>

	Inversion of knee reflex <ul style="list-style-type: none"> • On eliciting the knee jerk • There is no extension of the knee joint • But instead there is flexion of the knee due to contraction of the hamstring muscles • Presence of this indicates that the lesion is at the level of L3, 4
Perverted reflex	<p>It is false inverted reflex where there is an alteration in the response rather than true inversion</p> <p>For example: When supinator jerk is elicited there is a perverted response of finger flexion. (<i>Note:</i> In the presence of brachioradialis reflex this phenomenon is called as spread of reflex, while in the absent of brachioradialis reflex this is considered as pseudo inverted reflex or perverted reflex)</p>

Other Causes of Altered Reflexes	
Thyroid disease	
Woltman's sign	of myxedema, is the delayed relaxation phase of the muscle stretch reflex. In hypothermia or β-blockade, the relaxation phase of the ankle jerk may be prolonged.
Chorea: "Hung-up" knee jerk	is a specific but rarely appreciated clinical sign of Huntington disease (HD) and Sydenham chorea. During an elicited knee jerk, the extended lower leg may not relax immediately but may remain elevated for several seconds due to sustained contraction of the quadriceps femoris.
Very brisk reflexes	—even with a few beats of clonus can be seen in anxious individuals, as well as in hyperthyroidism and in tetany.
Electrolyte disturbances	<ul style="list-style-type: none"> • Absent reflexes is seen with hypermagnesemia. • In the Holmes Adie syndrome, absent deep tendon reflexes are seen.

D(vi). SENSORY SYSTEM EXAMINATION

SENSORY SYSTEM EXAMINATION

Sensations can be grossly divided into primary and secondary modalities

Primary modalities	Secondary modalities (cortical sensation)
Touch	Tactile localization
Pressure	2 point discrimination
Pain	Sensory inattention
Temperature	Stereognosis
Joint position sense	Graphesthesia
Vibration	These require secondary association area in parietal lobe

Note: When primary sensations are normal but secondary modalities are lost it implies a parietal lobe lesion.

Sherrington classification of sensory system	
Exteroceptive system	Information about the external environment, including somatosensory functions and special senses
Proprioceptive system	Senses the orientation of the limbs and body in space
Interoceptive system	Information about internal functions, blood pressure, or the concentration of chemical constituents in bodily fluids

PRIMARY MODALITIES

Examination of Exteroceptive System (Spinothalamic Tract)

Pain

- Ask the patient to close his eyes.
- Sharp end of pin is applied mildly sufficient to produce pain but not to penetrate the skin [Fig. 6D(vi).1].
- Compare adjacent normal area and corresponding area on the opposite side.
- Indicate whether sensation is normal, decreased (or absent) or increased.
- In peripheral nerve disease, there is anesthesia more than analgesia.
- In spinal cord disease, there is analgesia more than anesthesia.
- Commonly used objects are the safety pin or broken wooden applicator stick.
- Avoid too sharp objects and hypodermic needles.
- A useful trick is to hold the pin or shaft of the applicator stick lightly between thumb and fingertip and allow the shaft to slide between fingertip and thumb. This ensures consistent stimulus intensity.



Fig. 6D(vi).1: Examination of pin prick sensation.

Temperature [Fig. 6D(vi).2]

- With the patient's eyes closed, apply the warm and cold test tubes randomly over the skin in dermatomal pattern.
- Instruct the patient to say what he feels—hot/cold/no response.
- Cold = 5°C to 10°C (41°F to 50°F) (crushed ice can be used).
- Warmth = 40°C to 45°C (104°F to 113°F) (warm water can be used).
- Temperature much lower or higher than these elicit pain rather than temperature sensations.
- In lesions of leprosy, temperature may be lost prior to pain.



Fig. 6D(vi).2: Examination of temperature.

Tactile Sensation

- Light touch can be tested with a:
 - Wisp of cotton [Fig. 6D(vi).3]
 - Feather
 - Soft brush [Fig. 6D(vi).4]
 - Light touch of the fingertip.

- For diabetic neuropathies
 - Von Grey's hairs
 - Semmelweis monofilament
- With patient's eyes closed, gently touch the skin (preferably non-hairy region) without exerting pressure.
- Ask the patient whether he can feel the touch.
- Tactile response can be graded as per international spinal injury standards as
 - 0 = absent
 - 1 = altered response (impaired/increased)
 - 2 = normal/intact response.



Fig. 6D(vi).3: Examination of tactile sensation with wisp of cotton.



Fig 6D.vi.4: Examination of tactile sensation with soft brush.

Examination of Proprioceptive System

Proprioception (Proprioception refers to either the sense of position of a body part or motion of a body part)	
Conscious component	Unconscious component
Travels with the fibers subserving fine, discriminative touch.	Via spinocerebellar tract

These include:
 Motion
 Position
 Vibration
 Pressure

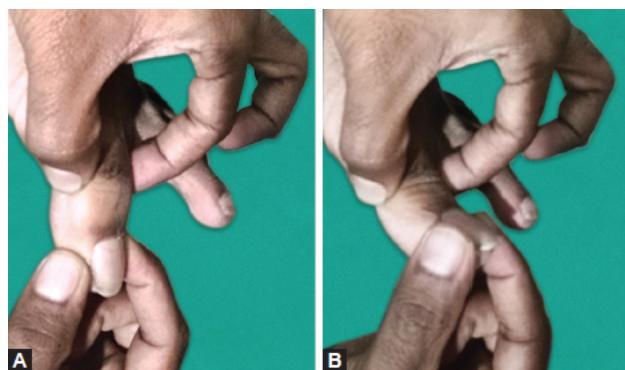
Examination of different components of proprioception: Joint motion and position:

- Usually tested together
- In the lower extremity [Figs. 6D(vi).5A and B]:
- Tested at the metatarsophalangeal joint of the great toe,
- In the upper extremity [Figs. 6D(vi).6A to C]:

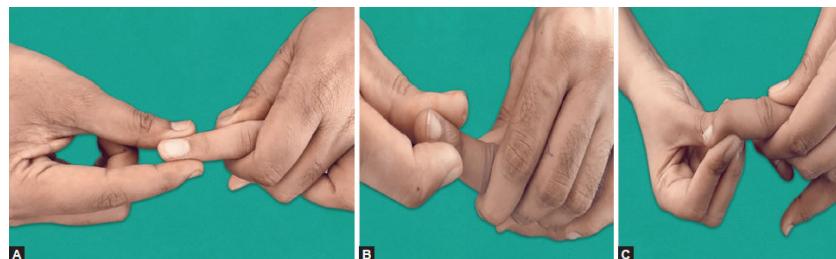
At one of the distal interphalangeal joints. If these distal joints are normal, there is no need to test more proximally.

Joint motion:

- Testing is done with the patient's eyes closed.
- It is extremely helpful to instruct the patient, eyes open, about the responses expected before beginning the test.
- Show the patient up or down movements and instruct him to reply "up" or "down".
- The examiner should hold the patient's completely relaxed digit on the sides, away from the neighboring digits, parallel to the plane of movement, exerting as little pressure as possible to eliminate clues from variations in pressure.
- The part is then passively moved up or down, and the patient is instructed to indicate the direction of movement from the last position.
- Healthy young individuals can detect great toe movements of about 1 mm, or 2° to 3°; and in the fingers virtually invisible movements, 1° or less, at the distal interphalangeal joint are accurately detected.



Figs. 6D(vi).5A and B: Examination of joint sense in the lower limb.



Figs. 6D(vi).6A to C: Examination of joint sense in upper limb.

Position sense:

- Tested by placing the fingers of one of the patient's hands in a certain position (like "OK" sign) [Fig. 6D(vi).7] while his eyes are closed, and then asking him to imitate it with the other hand OR do passive movement in one hand and ask the patient to do in similar way in other hand [Fig. 6D(vi).8].
- This is sometimes referred to as **parietal copy**. Light touch can be tested with a wisp of cotton, tissue paper, a feather, a soft brush, light stroking of the hairs, or even using a very light touch of the fingertip. Both parietal lobes (and their connections) must be intact: one side to register the position and the other side to copy it.

Vibration (pallesthesia) [Figs. 6D(vi).9A to C]: Preferentially using a tuning fork of 128Hz due to slow decay (256 Hz is used to detect early changes in cases like subacute combined cord degeneration).



Fig. 6D(vi).7: Examination of position sense (OK sign).

- Explain procedure to patient clearly.
- Strike the tuning fork and place on the forehead and explain the difference between vibration and plain touch of tuning fork, by dampening the vibration by holding the prongs.



Fig. 6D(vi).8: Examination of position sense by asking to copy passive movement.



Fig. 6D(vi).9A: Demonstration of vibration over proximal great toe.



Fig. 6D(vi).9B: Demonstration of vibration over medial malleolus.

- Keep the vibrating tuning fork, starting from the distal most bony prominence and proceed proximally.
- Ask the patient to say when he ceases to feel the vibration.

Timed vibration test:

- It is the most sensitive and simple method to quantify defects in vibration.
- Note the time duration of perception of vibration after the tuning fork is set into vibration.
- Normally
 - ≥ 10 sec in lower limb.
 - ≥ 20 sec in upper limb.

Rhomberg's sign [Figs. 6D(vi).10A and B]:

- It is a sign of posterior column dysfunction.
- Ask the patient to stand upright with feet/heels close together, arms by the side and eyes open.
- Any significant swaying is noted.
- Now, ask the patient to close the eyes while taking adequate measures to make sure patient does not fall and hurt himself.
- Watch for swaying
 - Minimal swaying is normal.

- Immediate gross swaying is considered as positive test.

Pseudoathetosis [Fig. 6D(vi).11]:

- It is an upper limb equivalent of examination of posterior column dysfunction.
- Ask the patient to hold the upper limb in extended position and close the eyes.
- Watch for slow writhing movements of fingers (piano-playing movement) which disappear on opening the eyes.

Pressure pain:

- Tested by squeezing the Achilles tendon or calf muscle.
- Abadie's sign is loss of deep pain (seen with diseases affecting the posterior column like neurosyphilis—tabes dorsalis).

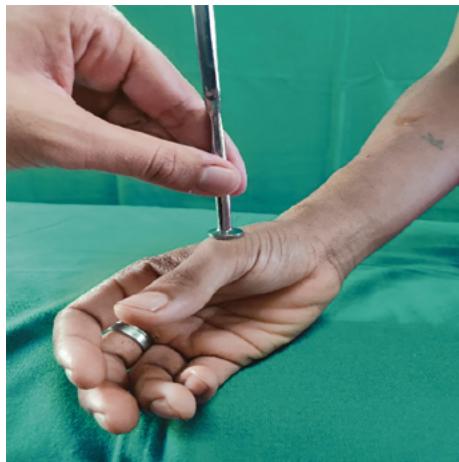


Fig. 6D(vi).9C: Demonstration of vibration over the proximal 1st metacarpopharyngeal joint.



Figs. 6D(vi).10A and B: Demonstration of Rhomberg's sign.



Fig. 6D(vi).11: Demonstration of pseudoathetosis in upper limb.

SECONDARY MODALITIES

Cortical Sensations

Cortical sensations cannot reliably be tested unless primary sensation is intact bilaterally.

Two-point discrimination [Fig. 6D(vi).12]: Ability to recognize simultaneous stimulation by two blunt points. Measured by the distance between the points required for recognition. The normal distances at which two points can be discriminated on various body parts:

- Tongue tip: 1 mm
- Fingertip: 2 to 4 mm
- Dorsum of fingers: 4 to 6 mm
- Palm: 8 to 12 mm
- Dorsum of hand: 20 to 30 mm
- Skin over the back : 30–40 mm.

Tactile localization (topognosia):

Ability to localize stimuli to parts of the body. Topagnosia is the absence of this ability.

Graphesthesia [Fig. 6D(vi).13]:

Ask the patient to close their eyes and identify letters or numbers that are being traced onto their palm or the tip of their finger.

Stereognosis [Figs. 6D(vi).14A and B]:

Ask the patient to close their eyes and identify various objects by touch using one hand at a time.

Tactile extinction (double simultaneous stimulation) [Figs. 6D(vi).15A and B]

- Ability to perceive a sensory stimulus when corresponding areas on the opposite side of the body are stimulated simultaneously. Loss of this ability is termed sensory extinction (perceptual rivalry/sensory suppression).
- The site of lesion is contralateral parietal lobe.

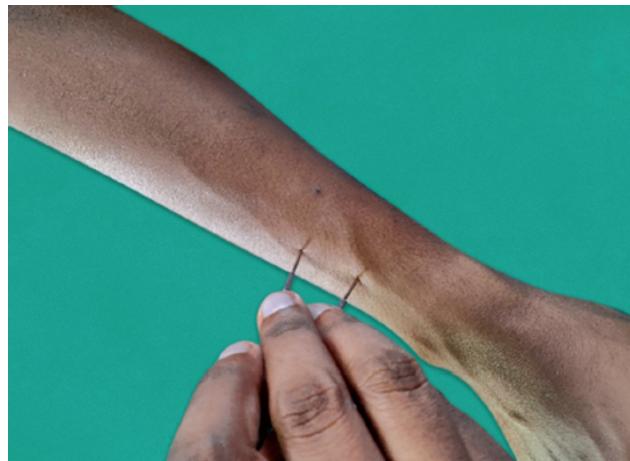


Fig. 6D(vi).12: Demonstration of 2 point discrimination.



Fig. 6D(vi).13: Demonstration of graphesthesia.



Fig. 6D(vi).14A: Demonstration of stereognosis with key.



Fig. 6D(vi).14B: Demonstration of stereognosis with coin.



Fig. 6D(vi).15A: Demonstration of tactile extinction in upper limb.



Fig. 6D(vi).15B: Demonstration of tactile extinction in lower limb

Disorders of touch	
Anesthesia	Absence of touch appreciation.
Hypoesthesia	Decrease in touch appreciation.
Hyperesthesia	Exaggeration of touch sensation, which is often unpleasant.
Paresthesia	Abnormal sensations perceived without specific stimulation. They can include wide variety of abnormal sensation except pain; episodic or constant.
Hyperpathia	Exaggerated reaction to any stimuli (touch/pressure/pain).
Disorders of pain	
Analgesia	Absence of pain appreciation.
Hypoalgesia	Decrease in pain appreciation.
Hyperalgesia	Exaggeration of pain appreciation, which is often unpleasant.
Allodynia	Perception of non-painful stimulus as painful.

Causalgia	Persistent pain, allodynia or hyperalgesia along with abnormal pseudomotor activity (edema and blood flow changes). It is also called as reflex sympathetic dystrophy.
Phantom limb pain	Individuals who have had a limb amputated may experience pain or tingling sensations that feels as if they were coming from the amputated limb, just as if that limb were still present. These individuals experience pain or tingling sensations that feel as if they were coming from the amputated limb, just as if that limb were still present.
Central or thalamic pain	Spontaneous, inexplicable, agonizing pain and other unusual sensations in the anesthetic parts.
Disorders of temperature	
Thermanesthesia	Absence of temperature appreciation
Thermypoesthesia	Decrease of temperature appreciation
Thermhyperesthesia	Exaggeration of temperature sensation, which is often unpleasant
Disorders of posterior column sensations	
Arthranesthesia	Absence of joint position sense (Arthesthesia —perception of joint position sense)
Apallesthesia/Pallanesthesia	Absence of vibration sense

Barognosis (recognition of weight)

- The ability to recognize different weights.
- A set of discrimination weights consisting of small objects of the same size and shape but of graduated weights are used.

HOMUNCULUS, SENSORY PATHWAY, DERMATOMES AND CLINICAL PATTERNS OF SENSORY LOSS

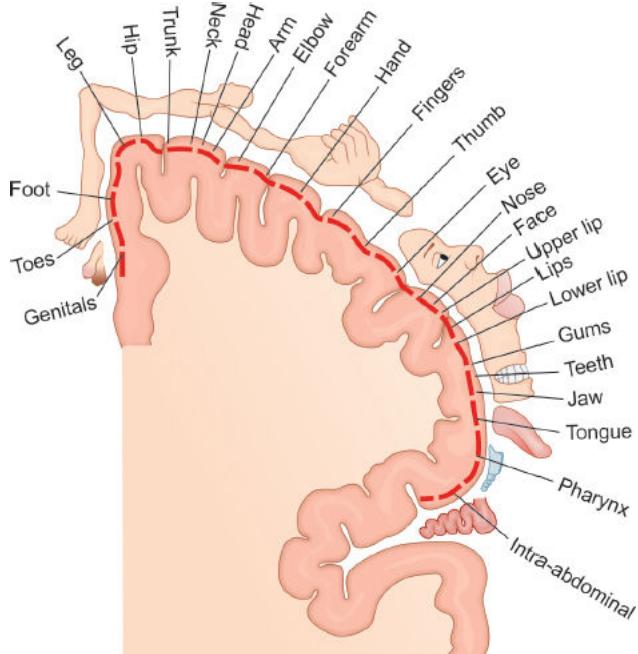


Fig. 6D(vi).16: Sensory homunculus.

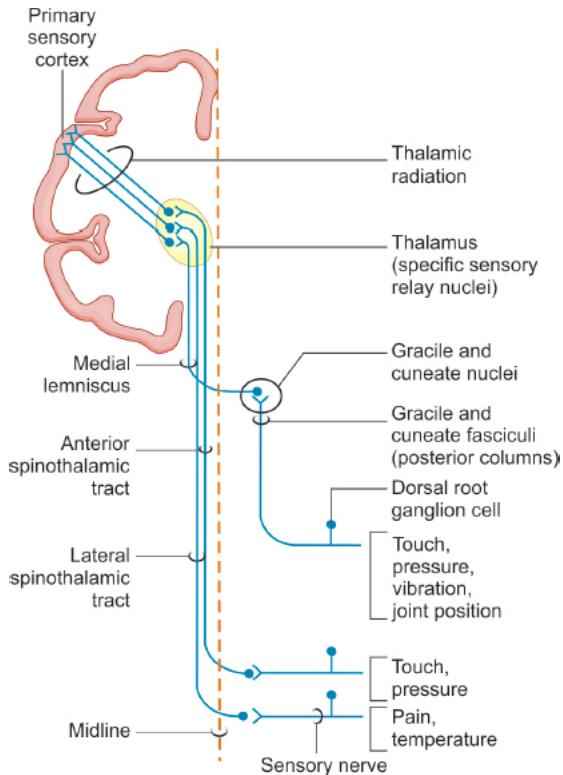


Fig. 6D(vi).17: Sensory pathway.

Sensation	Receptor	Pathway	Decussion
Pain and thermal sense from the body	A δ and C fiber endings	Spinothalamic tract of anterolateral system (ALS)	Anterior white commissure
Nondiscriminative (crude) touch and superficial pressure from the body	Free nerve endings, Merkel's disks, peritrichial nerve endings	Spinothalamic tract of ALS	Anterior white commissure
Two-point discriminative (fine) touch, vibratory sense, proprioceptive sense from muscles and joints of body	Meissner's corpuscles, Pacinian corpuscles, muscle stretch receptors, Golgi tendon organs	First order fibers: Fasciculi gracilis and cuneatus Second order fibers: Medial lemniscus	Medial lemniscal decapsulation

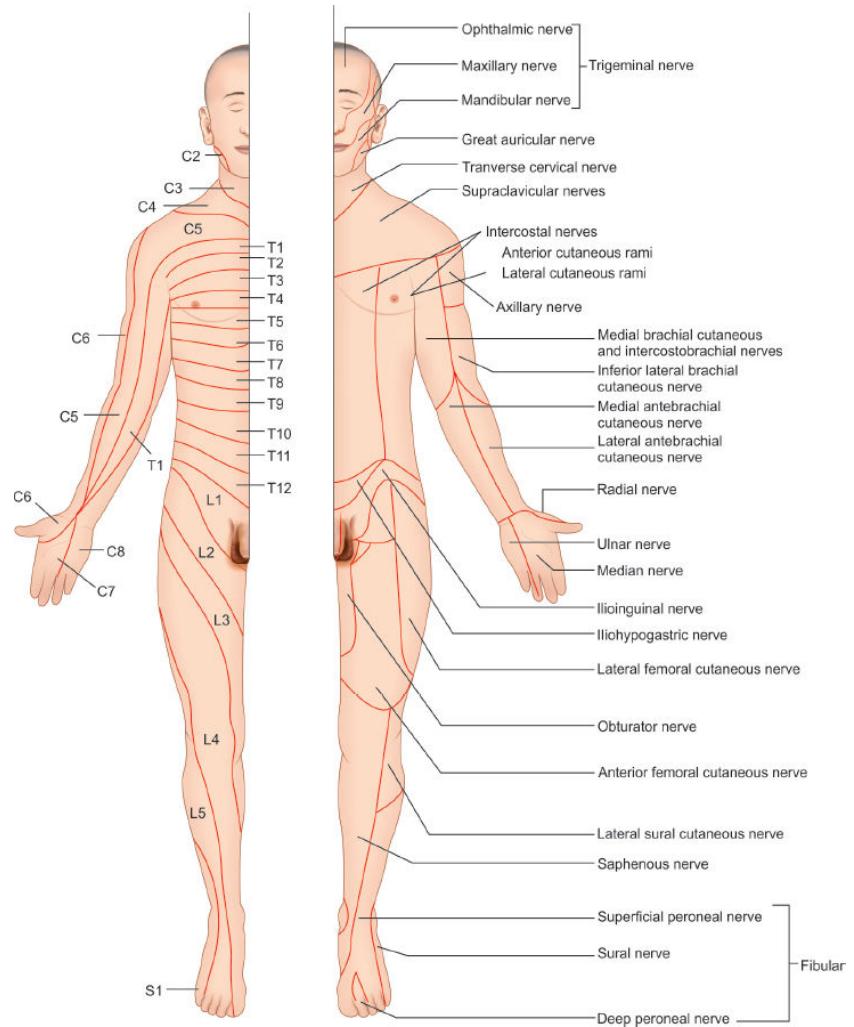


Fig. 6D(vi).18: Anterior view of skin segment innervation.

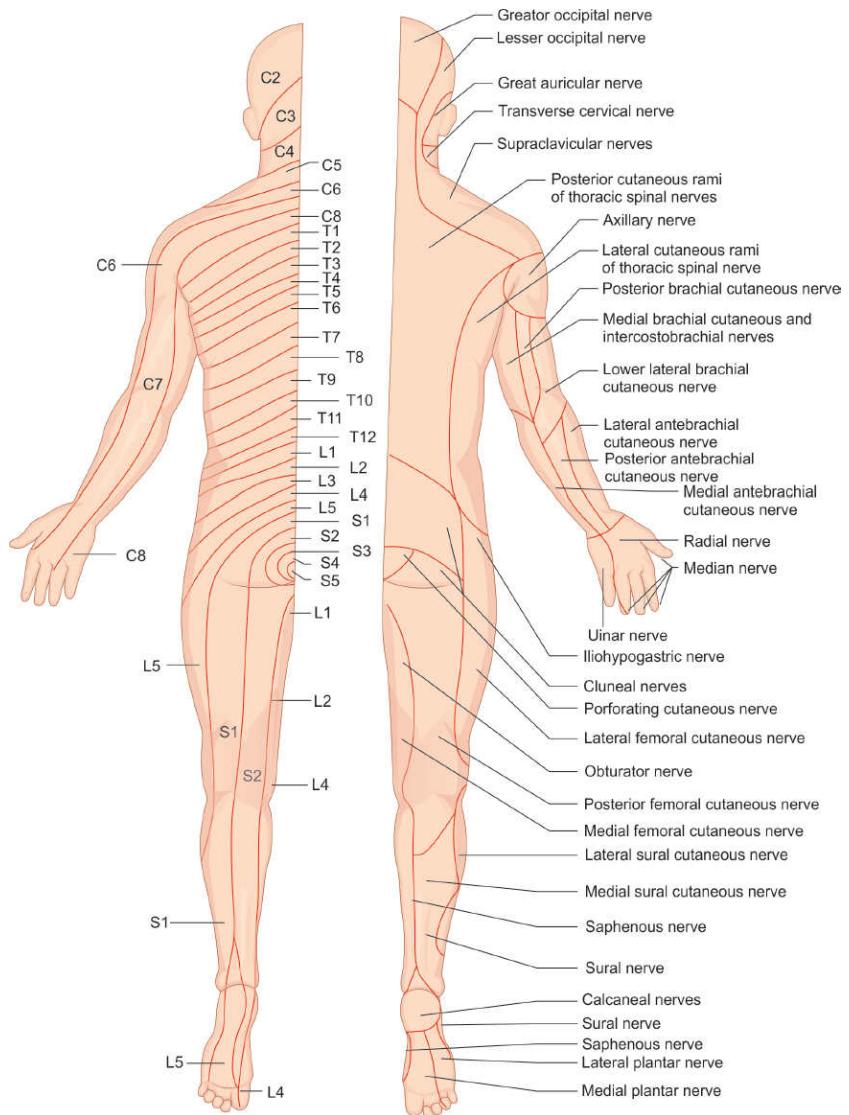
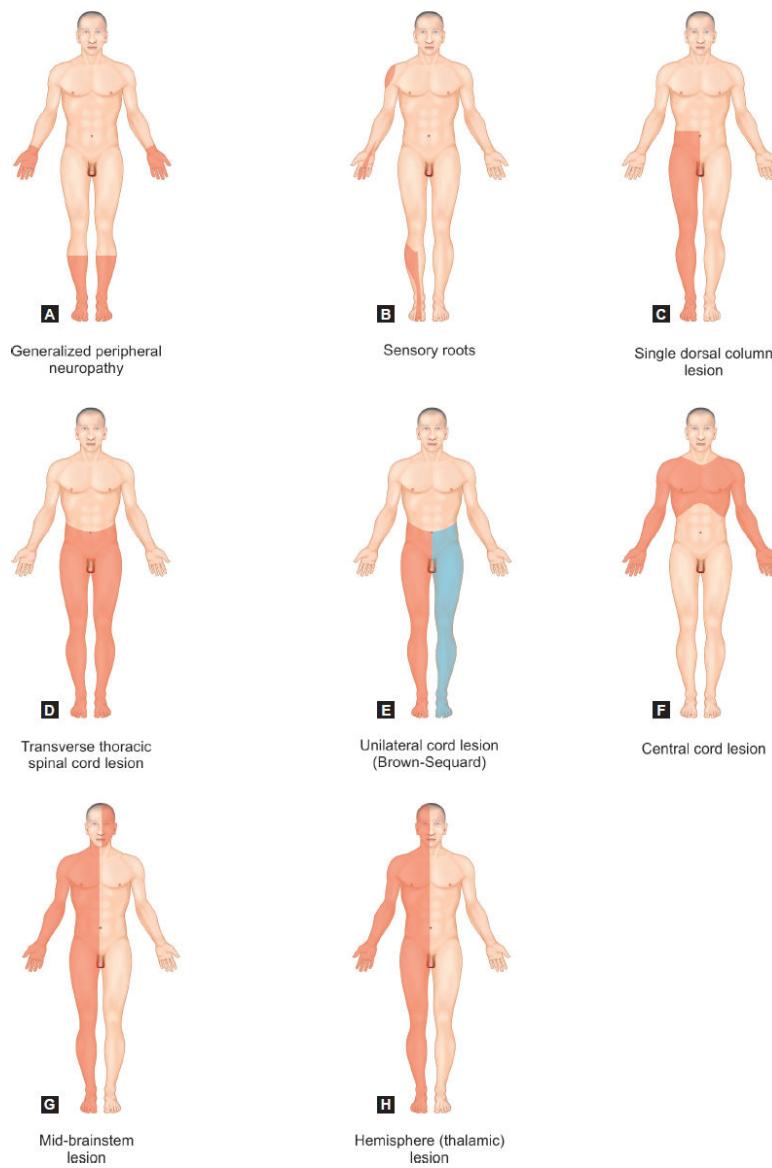


Fig. 6D(vi).19: Posterior view of skin segment innervation.

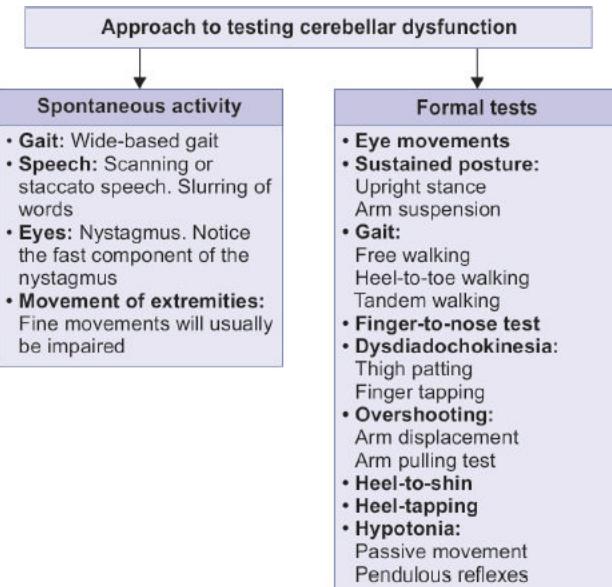


Figs. 6D(vi).20A to H: Clinical patterns of sensory dysfunction.

D(vii). CEREBELLUM AND COORDINATION

SIGNS OF CEREBELLAR DISORDERS

Deficit	Manifestation
Ataxia	Reeling, wide-based gait.
Decomposition of movement	Inability to sequence fine, coordinate acts correctly. <i>This is usually tested while performing the finger-nose test which requires a fine coordination between shoulder, elbow, and wrist joint. Patients with a cerebellar lesion will find it difficult to perform such movements.</i>
Dysarthria	Inability to articulate words correctly, usually manifesting as slurring and/or inappropriate phrasing.
Dysdiadochokinesia	Inability to perform rapid, alternating movements.
Dysmetria	Inability to control or limit the range of movement.
Hypotonia	Decrease in muscle tone.
Nystagmus	Involuntary rapid oscillation of eyeballs in a horizontal, vertical or rotational fashion with the fast component of nystagmus maximal towards the side of the cerebellar lesion.
Scanning/Staccato speech	Slow explosive enunciation with a tendency to hesitate at the beginning of each word or each syllable. <i>Asking the patient to pronounce a word with multiple syllables, such as Mississippi or Venkataramana will elicit distinct pauses before each syllable.</i>
Tremor	Rhythmic, alternating, oscillatory movements which affects a limb as it approaches a target (Intention tremor) or of proximal musculature when attempting to bear weight (postural tremor).



Hypotonia

- Usually accompanies acute hemispheric lesions.
- Interestingly, it is seen less often in chronic lesions.
- Ipsilateral to the side of a cerebellar lesion.
- More noticeable in upper limbs and proximal muscles.
- Pendular knee jerk: Leg keeps swinging after knee jerk more than 4 times (4 or less is considered normal).

Ataxia

- Defective timing of sequential contraction of agonist/antagonist muscles.
- Results in a disturbance in smooth performance of voluntary acts (errors in rate, range, force, duration).
- May affect limbs, trunk, gait (depends on the part of cerebellum involved).

Asynergia

Lack of synergy of various muscles while performing complex movements (movements are broken up into isolated, successive parts. This is known as decomposition of movement).

Dysmetria OR abnormal excursions in movement

Finger-to-nose test

- With eyes open, the patient is asked to partially extend elbow and rapidly bring tip of index finger in a wide arc to tip of his nose.
- In cerebellar disease, the action may manifest an intention tremor.
- With eyes closed, sense of position in the shoulder and elbow is tested.

Heel-to-shin test

- Patient is asked to place one heel on opposite knee and slide the heel down the tibia with foot dorsiflexed.
- Movement should be performed accurately.
- In cerebellar disease, the arc of the movement is jerky/wavering.
- The slide down the shin may manifest an action tremor.

Dysdiadochokinesia OR impaired performance of rapidly alternating movement

Normal coordination includes ability to arrest one motor impulse and substitute the opposite.

There are several simple clinical methods to test this:

- Alternating movements (pronate and supinate forearm and hand quickly): In cerebellar disease, the movements tend to overshoot or are inadequate resulting in irregular or inaccurate movements.
- Rapidly tap fingers on the table.
- Open and close fists.
- Stewart-Holmes rebound sign.

Have the patient pull on your hand and when they do, slip your hand out of their grasp. Normally the antagonists muscles will contract and stop their arm from moving in the desired direction. A positive sign is seen in a spastic limb where the exaggerated "rebound" occurs with movement in the opposite direction. However, in cerebellar disease, this response is completely absent causing the limb to continue moving in the desired direction. (Be careful that you protect the patient from the unrestricted movement causing them to strike themselves).

Past pointing

Overshoot is also commonly seen as part of ataxic movements and is sometimes referred to as past pointing, when the patient overshoots while reaching target (finger-to-nose test)

Cerebellar dysarthria

- Abnormalities in articulation and prosody (together or independent).
- Scanning, slurring, staccato, explosive, hesitant, garbled speech.
- Hemisphere lesions are associated with speech disorders more often than vermal lesions.
- Causes enunciation of individual syllables: "*the British Parliament*" becomes "*the Brit-ish Par-la-ment*."

Intention tremor—occurs during goal-directed movements. Intention tremor results when the antagonist activation that normally stops a goal-directed movement as the goal is approached is inappropriately sized or timed.

Oculomotor dysfunction

- Nystagmus frequently seen in cerebellar disorders.
- Gaze-evoked nystagmus, upbeat nystagmus, rebound nystagmus, optokinetic nystagmus may all be seen in midline cerebellar lesions.

Gait

- In cerebellar disease, the gait is staggering/lurching/wavering.
- Lesion in mid-cerebellum: Movements are in all directions.
- Lesion in lateral cerebellum: Staggering/falling is toward the side of the lesion.
- Somewhat steadied by standing or walking on a wide base.

Position of feet

Ataxia from cerebellar disease is less when the patient stands on a broad base (feet widely apart).

Eyes open or closed

Cerebellar ataxia is not improved by visual orientation; ataxia from posterior column disease (disordered proprioception) is worsened with the eyes closed.

Direction of Falling

Disease of lateral lobe of cerebellum causes falling to ipsilateral side.

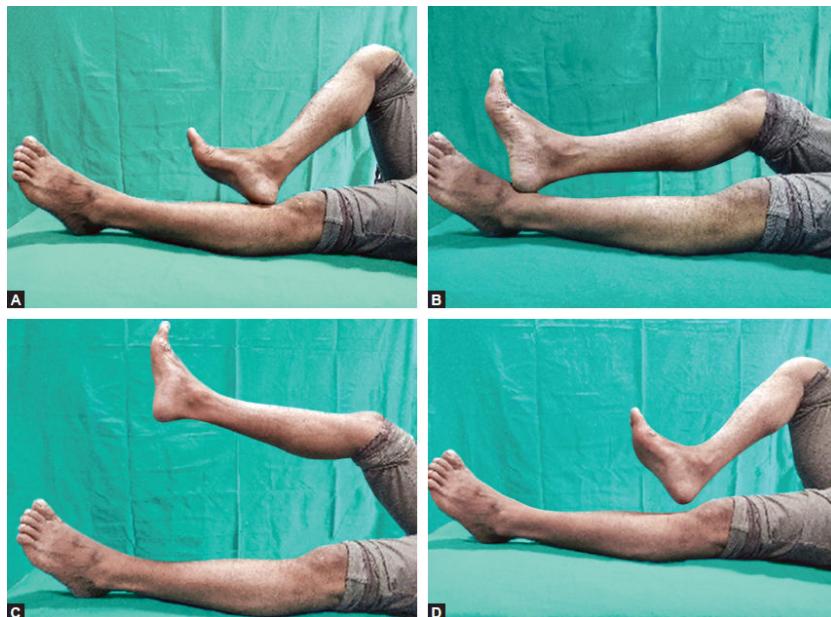
Lesions of midline/vermis cause indiscriminate falling depending on initial stance of the patient.

Titubation

Consists of a rhythmic body or head tremor. There is a rotatory, rocking or bobbing movement. *Clinically, this does not have significant value in localizing the lesion with respect to the part of the cerebellum involved.*

HEEL KNEE TEST [FIGS. 6D(VII).1A TO D]

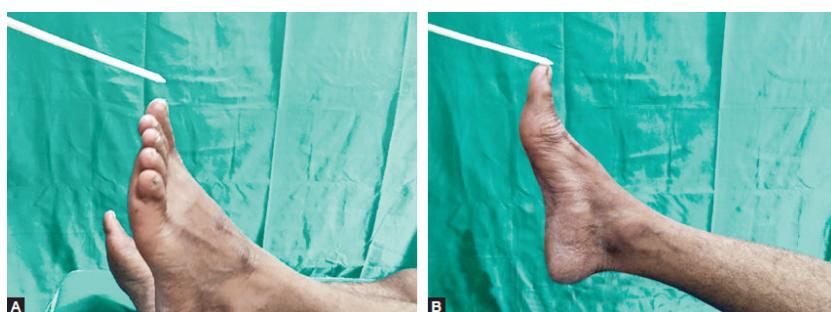
The patient is asked to touch the heel of one foot to the opposite knee and then to drag their heel in a straight line all the way down the front of their shin and back up again. In order to eliminate the effect of gravity in moving the heel down the shin, this test should always be done in the supine position.



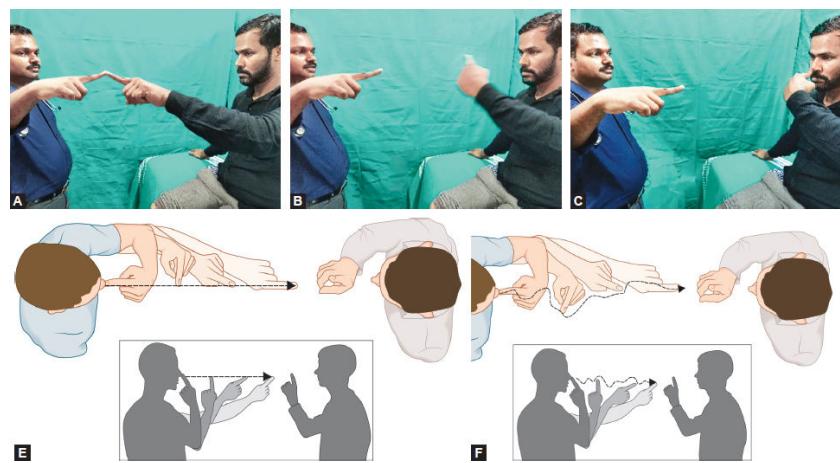
Figs. 6D(vii).1A to D: Demonstration of heel knee test.

TOE FINGER TEST [FIGS. 6D(VII).2A AND B]

Patient lies in bed and is asked to touch his great toe to the examiners fingers or any object held above the bed within his reach.



Figs. 6D(vii).2A and B: Demonstration of toe finger test.



Figs. 6D(vii).3A to E: Showing demonstration of nose finger nose test.

Nose-finger-nose test [Figs. 6D(vii).3A to E] in which the patient is asked to alternately touch their nose and the examiner's finger as quickly as possible. Abnormality of this is called as dysmetria.

FINGER NOSE TEST [FIGS. 6D(VII).4A AND B]



Figs. 6D(vii).4A and B: Demonstration of finger nose test.

Rebound Phenomenon [Fig. 6D(vii).5]



Fig. 6D(vii).5: Demonstration of rebound phenomenon.

DYSIDIADOKOKINESIA [FIGS. 6D(VII).6A TO D]



Figs. 6D(vii).6A to D: Demonstration of dysdiadochokinesia.

FOOT TAPPING/FOOT PAT TEST [FIGS. 6D(VII).7A TO C)]

Patient is made to sit on chair with feet touching the floor flat. He is asked to pat the floor with his forefoot. The rate, rhythm and speed of patting is compared on both sides. Even minimum cerebellar disease can be picked up by this test.



Figs. 6D(vii).7A to C: Demonstration of foot tapping.

STRAIGHT LINE WALKING [FIGS. 6D(VII).8A AND B]



Figs. 6D(vii).8A and B: Straight line walking.

TANDEM WALKING [FIGS.6D(VII).9A AND B]



Figs. 6D(vii).9A and B: Demonstration of tandem walking.

ROMBERG TEST [FIGS. 6D(VII).10A AND B]

Patient stands still with their heels together. Ask the patient to remain still and close their eyes. If the patient loses their balance immediately, the test is positive.

To achieve balance, a person requires 2 out of the following 3 inputs to the cortex: 1. Visual confirmation of position, 2. Nonvisual confirmation of position (including proprioceptive and vestibular input), and 3. A normally functioning cerebellum.

Therefore, if a patient loses their balance after standing still with their eyes closed, and is able to maintain balance with their eyes open, then there is likely to be lesion in sensory input.



Figs. 6D(vii).10A and B: Demonstration of Romberg's sign.

APPROACH TO ATAXIA

- Ataxia, defined as impaired coordination of voluntary muscle movement affecting the rate, range, direction and force of movements.
- It is a physical finding, not a disease.
- Types of ataxia:
 1. Cerebellar
 2. Sensory
 3. Vestibular
 4. Optic
 5. Frontal

Type of ataxia	Cerebellar	Sensory	Frontal
Stance and support	Wide based	Narrow based; looking down	Wide based
Velocity	Variable	Slow	Very slow
Stride	Irregular, lurching	Regular with path deviation	Short, shuffling
Romberg	+/-	Unsteady; patient falls	+/-
Heel-shin	Abnormal	+/-	Normal
Initiation	Normal	Normal	Hesitant
Postural instability	+	+++	++++
Falls	Late event	Frequent	Frequent
Turns	Unsteady	+/-	Multisteped; hesitant

Sensory ataxia is due to a severe sensory neuropathy, ganglionopathy or lesions of the posterior column of the spinal cord, e.g. Sjogren's syndrome, cisplatin, chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), paraneoplastic disorders, subacute combined degeneration (SACD), tabes dorsalis, Miller Fischer syndrome, celiac disease.

- Ataxia more at night or while walking through narrow passages (coffee plantations).

- A history of falling into the sink or imbalance when splashing water on the face (wash-basin sign), passing a towel over the face or pulling a shirt over the head should also be sought.
- Pseudoathetosis—"piano-playing" movements—when the patient has his arms outstretched and eyes closed, the affected arm will wander from its original position.
- Vibration and position sense are usually lost together.
- Positive Romberg's test is a hallmark of sensory ataxia.

Vestibular ataxia is due to lesion of vestibular pathways resulting in impairment and imbalance of vestibular inputs, e.g. vestibular, neuronitis, and streptomycin toxicity.

- Vertigo and associated tinnitus and hearing loss.
- Direction of the nystagmus is away from the lesion.

Optic ataxia was first described in a man with lesions of the posterior parietal lobe on both sides of the brain, later known as **Balint syndrome**.

- Among the symptoms that characterize the syndrome are a restriction of visual attention to single objects and a paucity of spontaneous eye movements.
- Patients have difficulty in completing visually guided reaching tasks in the absence of other sensory cues.

Frontal lobe ataxia (Brun's ataxia) is due to involvement of subcortical small vessels, Binswanger's disease, multi infarct state or normal pressure hydrocephalus (NPH).

- The gait may appear to be a combination of awkward, magnetic (stuck to the floor), cautious, slow, and shuffling. This is also known as a frontal gait disorder, referring to the frontal lobe conditions which often cause **gait apraxia**.

CEREBELLAR ATAXIA

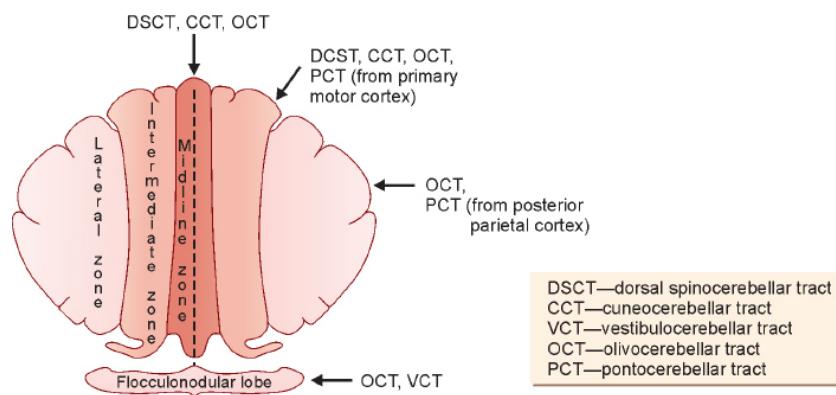


Fig. 6D(vii).11: Anatomical and functional areas of cerebellum.

Zone [Fig. 6D(vii).11]	Corresponding anatomical site	Function	Loss of function
Midline zone	Anterior and posterior parts of the vermis, fastigial nucleus	Posture, locomotion, position of head relative to trunk, control of extraocular movements	Disorders of stance/gait, truncal postural disturbances, rotated postures of the head, disturbances of eye movements
Intermediate zone	Paravermal region of cerebellum and interposed nuclei (<i>emboliform, globose</i>)	Control of velocity, force and pattern of muscle activity	—
Lateral zone	Cerebellar hemisphere and dentate nucleus	Planning of fined and skilled movement (<i>in connection with neurons in the Rolandic region of the cerebral cortex</i>).	Hypotonia, dysarthria, dysmetria, dysdiadochokinesia, excessive rebound, impaired check, kinetic and static tremors, past pointing

CAUSES OF CEREBELLAR ATAXIA

Symmetrical Cerebellar Ataxias

Acute	Subacute	Chronic
<ul style="list-style-type: none"> Drugs: Phenytoin, phenobarbitone, lithium, Chemotherapeutic agents Alcohol Infectious: Acute viral cerebellitis, post-infectious Toxins: Toluene, glue, gasoline, methyl mercury 	<ul style="list-style-type: none"> Alcohol, or Nutritional (B₁, B₁₂) Paraneoplastic Antigliadin or anti-GAD antibody Prion diseases 	<ul style="list-style-type: none"> MSA-C Hypothyroidism Phenytoin toxicity

(GAD: glutamic acid decarboxylase; MSA-C: multiple system atrophy with cerebellar ataxia)

Asymmetrical Cerebellar Ataxias

Acute	Subacute	Chronic
<ul style="list-style-type: none"> Vascular: Cerebellar infarction or hemorrhage, subdural hematoma Infectious: Abscess 	<ul style="list-style-type: none"> Neoplastic: Glioma, metastases, lymphoma Demyelination: MS HIV related: Progressive multifocal leukoencephalopathy 	<ul style="list-style-type: none"> Congenital lesions: Arnold Chiari malformation, Dandy Walker syndrome

Treatable Causes of Ataxia

- Hypothyroidism
- Ataxia with vitamin E deficiency (AVED)
- Vitamin B₁₂ deficiency
- Wilson's disease
- Ataxia with antigliadin antibodies and gluten sensitive enteropathy
- Ataxia due to malabsorption syndromes
- Lyme's disease
- Mitochondrial encephalomyopathies, aminoacidopathies, leukodystrophies and urea cycle abnormalities
- Wernicke's encephalopathy

Cerebellar Syndromes	
Rostral vermis syndrome (anterior lobe) For example, alcoholics	Wide-based stance and gait. Ataxia of gait; proportionally less ataxia is seen on performing Heel-shin test while the patient is lying down. Normal or slightly impaired arm coordination. Infrequent hypotonia, nystagmus and/or dysarthria.
Caudal vermis syndrome (flocculonodular, posterior lobe) For example, tumors (medulloblastoma)	Axial disequilibrium; staggering gait. Little or no limb ataxia. Spontaneous nystagmus might be seen. Rotated postures of head.
Hemispheric syndrome (Posterior lobe, anterior variants also possible) For example, infarcts, neoplasms, abscesses.	Incoordination of ipsilateral limb movements. More noticeable with fine motor skills. Incoordination affects most noticeably muscles involved in speech and finger movements.
Pancerebellar syndrome For example, infectious/parainfectious processes, hypoglycemia, paraneoplastic disorders, toxic-metabolic disorders	Combination of all the other syndromes. Bilateral signs of cerebellar dysfunction involving trunk, limbs, cranial musculature.

LOCALIZATION OF CEREBELLAR LESIONS

Signs and symptoms	Most probable region of involvement
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Higher cognitive changes	Lateral hemispheres
Action tremor	Dentate and interposed nuclei OR cerebellar outflow to ventral thalamus
Palatal tremor	Dentate nucleus, Guillain Mollaret triangle
Titubation	Any zone; especially anterior vermis and associated deep nuclei
Dysarthria	Posterior left hemisphere and vermis
Gait ataxia	Anterior vermis
Limb ataxia	Lateral hemispheres
Saccadic dysmetria	Dorsal vermis
Square wave jerks	Cerebellar outflow
Gaze evoked nystagmus	Flocculus and paraflocculus

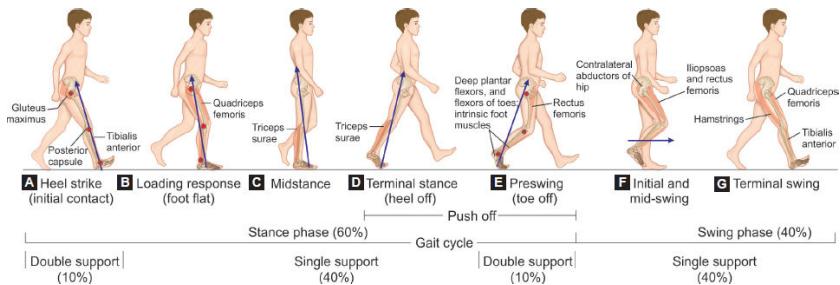
Mnemonics for cerebellar signs

Danish pen	Vanishd
Dysdiadochokinesia Ataxic gait Nystagmus Intention tremor Scaning/Staccato speech Hypotonia/Heel-shin test Pendular knee jerk	Vertigo Ataxia Nystagmus Intentional tremor Scanning speech Hypotonia Dysdiadochokinesia

D(viii). GAIT

NORMAL GAIT CYCLE [FIGS. 6D(VIII).1A TO G]

The gait cycle is the time interval or sequence of motions occurring between two consecutive initial contacts of the same foot, i.e. cycle of stance and swing by one foot.



Figs. 6D(viii).1A to G: Normal gait cycle.

Observation to be noted while the patient walks:

1. Posture of the body while walking
2. The regularity of the movement
3. The position and movement of the arms
4. The relative ease and smoothness of the movement of the legs
5. The distance between the feet both in forward and lateral directions
6. The ability to maintain a straight course
7. The ease of turning
8. Stopping
9. Position of feet and posture just before initiation of gait.

ABNORMALITIES OF GAIT

Neurogenic gait disorders should be differentiated from those due to skeletal abnormalities (characterized by pain producing an antalgic gait, or limp).

Gait abnormalities incompatible with any anatomical or physiological deficit may be due to functional disorders.

Pyramidal (Circumduction/Hemiplegic) Gait [Fig. 6D(viii).2]

- Lesions of the upper motor neuron lesions produce characteristic extension of the affected leg. There is tendency for the toes to strike the ground on walking and outward throwing/swing of lower limbs. This movement occurring at the hip joint is called circumduction. There is leaning towards the opposite normal side. The arm of the affected side is adducted at the shoulder and flexed at the elbow, wrist, and fingers.
- In hemiplegia/hemiparesis, there is a clear asymmetry between affected and normal sides on walking, but in paraparesis both lower legs swing slowly from the hips in extension and are stiffly dragged over the ground (walking in mud).



Fig. 6D(viii).2: Circumduction gait.

Foot Drop (High Stepping/Slapping Gait) [Fig. 6D(viii).3]

In normal walking, the heel is the first part of the foot to hit the ground. A lower motor neuron lesion affecting the leg will cause weakness of ankle dorsiflexion, resulting in a less controlled descent of the foot, which makes slapping noise as it hits the ground. In severe cases, the foot will have to be lifted higher at the knee to allow room for the inadequately dorsiflexed foot to swing through, resulting in a high-stepping gait. Cause, e.g. common peroneal nerve palsy.

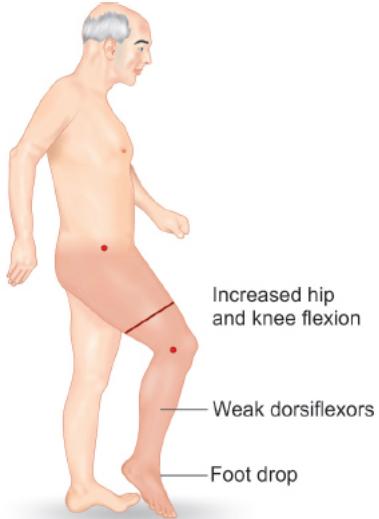


Fig. 6D(viii).3: High stepping gait.

Myopathic Gait/Waddling Gait [Fig. 6D(viii).4]

- During walking, alternating transfer of the body's weight through each leg, needs adequate hip abduction.
- **Causes:** Weakness of proximal lower limb muscles (e.g. polymyositis and muscular dystrophy) causes difficulty rising from sitting. The hips are not properly fixed by these muscles and trunk movements are exaggerated, and walking becomes a waddle or rolling. The pelvis is poorly supported

by each leg. This may be seen with bilateral congenital dislocation of hip (**Trendelenburg gait**). The patient walks on a broad base with exaggerated lumbar lordosis.

Gluteus Medius Gait or Abductor Lurch

Lurch of body towards affected side in every stance phase (abductor lurch). Seen with congenital coxa vara, gluteus medius paralysis, polio, and Perthes disease.

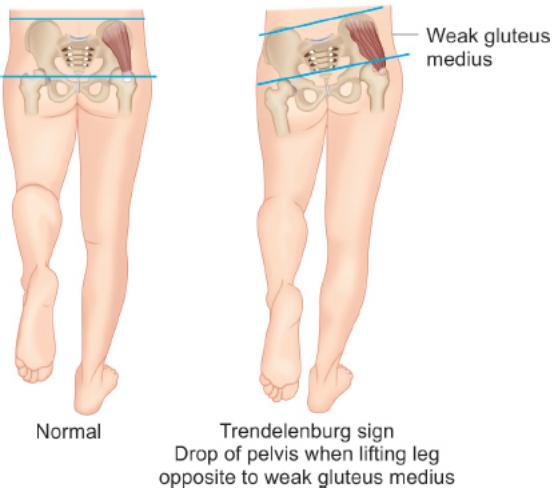


Fig. 6D(viii).4: Waddling gait.

Ataxic Gait (Cerebellar Ataxia: Broad-based Gait) [Fig. 6D(vii).5]

- In this type of gait, the patient, unstable, tremulous and reels in any direction (including backwards) and walks on a broad base. Ataxia describes this incoordination. The patient finds difficulty in executing tandem walking.
- Causes:** Lesions of the cerebellum, vestibular apparatus or peripheral nerves. When walking, the patient tends to veer to the side of the affected cerebellar lobe. When the disease involves cerebellar vermis, the trunk becomes unsteady without limb ataxia, with a tendency to fall backwards or sideways and is termed truncal ataxia.

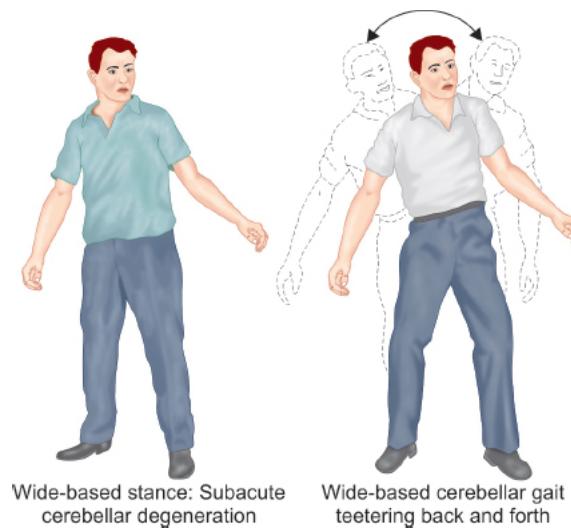


Fig. 6D(vii).5: Cerebellar/ataxic gait.

Apraxic Gait

- In an apraxic gait, the acquired walking skills become disorganized. On examination of the legs, the power, cerebellar function, and proprioception are normal. Leg movement is normal when sitting or lying and the patient can carry out complex motor tasks (e.g. bicycling motion). But patient cannot initiate and organize the motor act of walking. The feet appear stuck to the floor and the patient cannot walk.
- **Causes:** Diffuse bilateral hemisphere disease or diffuse frontal lobe disease (e.g. tumor, hydrocephalus, and infarction).

Marche à petits pas

- It is characterized by small, slow steps, and marked instability. In contrast to the festination found in Parkinson's disease, it lacks increasing pace and freezing.
- **Cause:** Small-vessel cerebrovascular disease and accompanying bilateral upper motor neuron signs.

Extrapyramidal/Shuffling/Festinant Gait [Fig. 6D(viii).6]

- It is characterized by stooped posture and gait difficulties with problems initiating walking and controlling the pace of the gait. Patients make a series of small, flat footed shuffles, and become stuck while trying to start walking or when walking through doorways (freezing). The center of gravity will be moved forwards to aid propulsion and difficulty in stopping. It is characterized by muscular rigidity throughout extensors and flexors. Power is preserved, pace is shortened and slows to a shuffle, and its base remains narrow. There is a stoop and diminished arm swinging and gait becomes festinant (hurried) with short rapid steps. Patient will be having difficulty in turning quickly and initiating movement. Retropulsion, i.e. small backward steps are taken involuntarily when a patient halts.
- **Cause:** Parkinsonism.

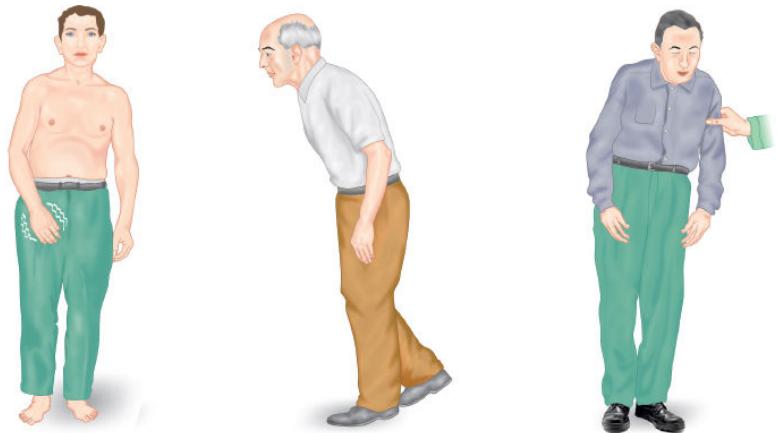
[**Kinesia paradoxa**—presented in Parkinson's disease patients, who generally cannot move but under certain circumstances of need exhibit a sudden, brief period of mobility (walking or even running)]

Scissoring Gait [Figs. 6D(viii).7A and B]

Seen classically with cerebral palsy due to bilateral spasticity.

Sensory Ataxia: Stamping Gait [Fig. 6D(viii).8]

- It is characterized by broad based, high stepping, stamping gait, and ataxia due to loss of proprioception (position sense). This type of ataxia becomes more prominent by removal of sensory input (e.g. walks with eyes closed) and becomes worse in the dark. Romberg's test is positive.
- **Cause:** Peripheral sensory (large fiber) lesions (e.g. polyneuropathy), posterior column lesion (vitamin B₁₂ deficiency or tabes dorsalis).

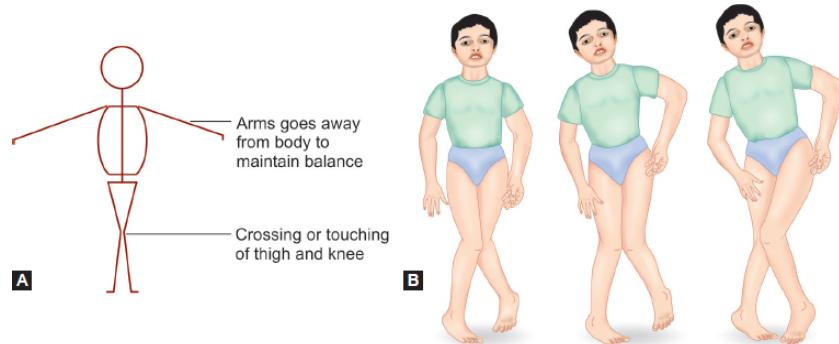


Stage 1: Unilateral involvement; early masking of facial expression; affected arm is semiflexed position with tremor

Stage 2: Bilateral involvement with early postural changes; slow, shuffling gait with decreased excursion of legs

Stage 3: Pronounced gait disturbances and moderate generalized disability; postural instability with tendency to fall

Fig. 6D(viii).6: Stages of Parkinson's gait.



Figs. 6D(viii).7A and B: Scissoring gait.



Fig. 6D(viii).8: Sensory ataxia.

Choreiform Gait (Hyperkinetic Gait)

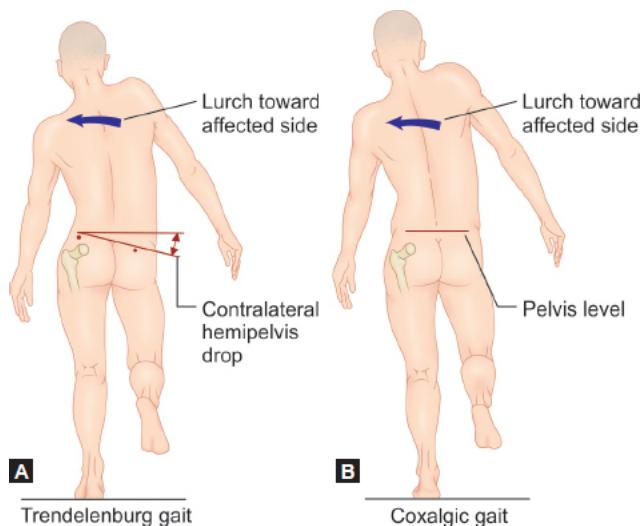
- The patient will display irregular, jerky, and involuntary movements in all extremities. Walking may
- accentuate their baseline movement disorder.
 - **Cause:** Sydenham's chorea, Huntington's disease, and other forms of chorea, athetosis or dystonia.

Antalgic or Painful Gait

Decreased duration of stance phase as the painful limb is unable to bear full weight. It is seen in any painful lesion of the lower extremity, i.e. foot, knee, and hip.

Coxalgic Gait [Figs. 6D(viii).9A and B]

In patients with hip pain, the upper trunk is typically shifted towards the affected side during the stance phase on the affected leg. This is an unconscious adaptive maneuver which reduces the force exerted on the affected hip during the stance phase.



Figs. 6D(viii).9A and B: Trendelenburg gait versus coxalgic gait.

Toe-walking or Equinus Gait

Heel strike is avoided. It is seen in patients with heel pain, clubfoot, congenital short Achilles tendon, and cerebral palsy.

Quadriceps Weakness Gait

Inability to maintain knee extension at heel-strike and patient may push on thigh to extend the knee and lock. It is seen in quadriceps paralysis.

Astasia-Abasia

It is a psychogenic pattern of walking in which the patient seems to alternate between a broad base for stability and a narrow, tightrope-like stance, with contortions of the trunk, and limbs that give the appearance of an imminent fall.

Alderman's Gait

Patient walks with chest and head thrown backwards with protuberant abdomen and legs thrown wide apart. It is seen in tuberculosis of lower thoracic and upper lumbar vertebra.

GAIT ABNORMALITIES ANALYSIS



Gait initiation, maintenance, and termination	Difficulty starting	PD, atypical parkinsonism
	Freezing of gait	PD, atypical parkinsonism
	Inability to stop (festination)	PD, atypical parkinsonism
Stance width	Narrowed base of support	PD, spastic paraparesis
	Widened base of support	Cerebellar ataxia, sensory ataxia, vestibular ataxia
	Scissoring of the legs	Spastic paraparesis
	Unable to walk in a straight line, sideways deviation (veering) of gait	Unilateral vestibular ataxia, unilateral cerebellar ataxia
Step length, height, and cadence	Reduced step height	PD, parkinsonism; foot drop
	Small steps	PD, atypical parkinsonism, normal pressure hydrocephalus
	Irregular step size	Cerebellar ataxia, vestibular ataxia, chorea
	Reduced stance phase on the affected side (limping)	Pain (antalgic gait)
Arm swing	Unilaterally reduced	Hemiparesis, dystonia, PD
	Bilaterally reduced	PD, parkinsonism, dystonia
	Excessive	Chorea, levodopa-induced dyskinesias, NPH
	Tremor appearing in hand during walking	PD, parkinsonism
Movement fluidity	Dropped foot, lifting the leg higher than normal (steppage gait)	Neuropathy of common fibular nerve or sciatic nerve, L5 radiculopathy, Charcot-Marie–Tooth disease
	Knees giving way (buckling of the knees)	Quadriceps weakness (for example, limb-girdle myopathy, IBM)
	Locking of the knees	Cerebellar ataxia
	Pelvis drop at side of the swing leg, resulting in alternating lateral trunk movements (waddling gait and bilateral Trendelenburg gait)	Bilateral proximal muscle weakness in the leg and hip girdle
	Bizarre gait pattern	Chorea
Gait speed	Slow	PD
	Fast	Vestibular disease, Alzheimer's disease

(PD: Parkinson's disease; NPH: normal pressure hydrocephalus; IBM: inclusion body myositis)

BEDSIDE TESTS TO DIAGNOSE PES CAVUS AND PES PLANUS

Wet Test [Fig. 6D(viii).10]

There are three basic foot types, each based on the height of the arches. The quickest and easiest way to determine your foot type is by taking the "wet test," below. (1) Pour a thin layer of water into a shallow pan. (2) Wet the sole of your foot. (3) Step onto a shopping bag or a blank piece of heavy paper. (4) Step off and look down. Observe the shape of your foot



Fig. 6D(viii).10: Wet test and appearance.

D(ix). APPROACH TO INVOLUNTARY MOVEMENTS

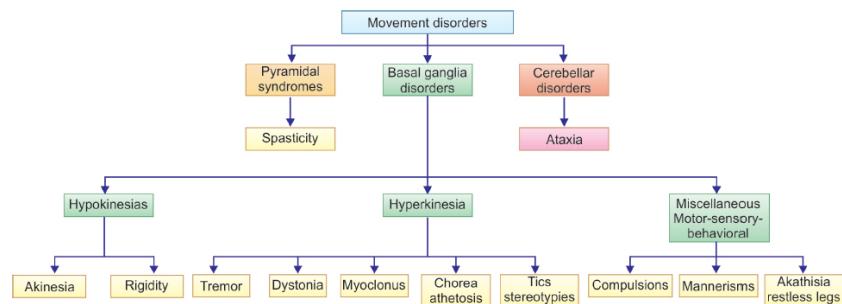
MOVEMENT DISORDERS

Dyskinesia is abnormal uncontrolled movement and is a common symptom of many movement disorders [Flowcharts 6D(ix).1 and 6D(ix).2].

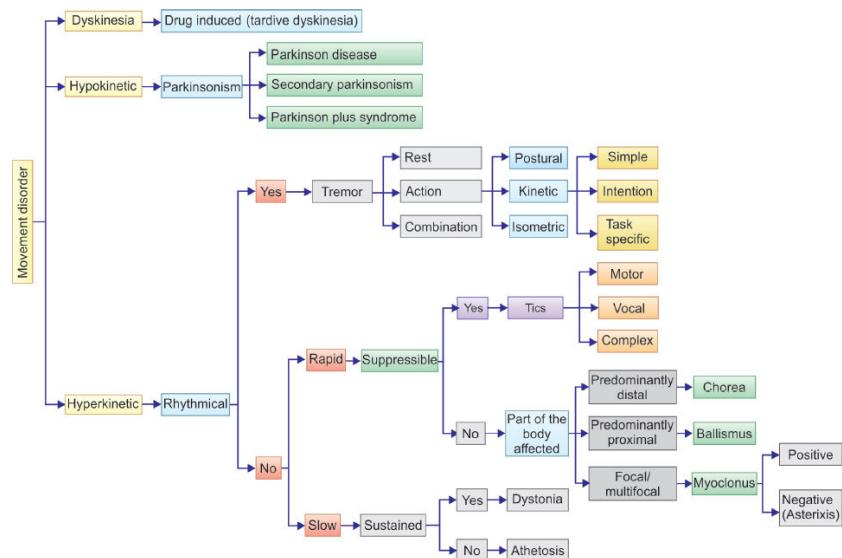
Movement disorders disrupt motor function by:

1. Abnormal, involuntary, unwanted movements (hyperkinetic movement disorders).
2. Curtailing (restricting) the amount of normal free flowing, fluid movement (hypokinetic movement disorders).

Flowchart 6D(ix).1: Categorization of movement disorders.



Flowchart 6D(ix).2: Systematic approach to movement disorders.



Site of Lesion

1. Parkinsonism → Contralateral substantia nigra
2. Unilateral hemiballismus → contralateral subthalamic nucleus
3. Chronic chorea → Caudate nucleus/putamen
4. Athetosis, dystonia → Contralateral putamen or thalamus
5. Myoclonus → Cerebellar cortex/thalamus

6. Rhythmic palatal/facial myoclonus → Central tegmental tract, inferior olivary nucleus, olivodentate fibers.

TREMOR

Tremor: Series of involuntary, relatively rhythmic, purposeless, oscillatory movements due to intermittent muscle contractions:

- Simple tremor involves only a single muscle group
- Compound tremor involves several muscle groups
 - Several elements in combination
 - Resulting in a series of complex movements
- May be unilateral or bilateral
- Most commonly involves distal parts of the extremities—fingers or hands
- May also affect the arms, feet, legs, tongue, eyelids, jaw, and head
- May occasionally involve the entire body
 - Rate may be slow, medium, or fast
 - » Slow: Oscillations of 3 to 5 Hz
 - » Rapid: Oscillations of 10 to 20 Hz
 - Amplitude may be fine, coarse, or medium
 - The relationship to rest or activity is the basis for classification into two primary tremor types:
 - » Resting
 - » Action

Action Tremors		
Resting (Static)		
<p>Postural tremors become evident when the limbs are: Maintained in an antigravity position (e.g. arms outstretched) Types of postural tremor:</p> <ul style="list-style-type: none"> • Enhanced physiological tremor (EPT) • Essential tremor (ET) 	<p>Kinetic tremor: Appears when making a voluntary movement May occur at the beginning, during or at the end of the movement. For example, intention (terminal) tremor seen primarily in cerebellar disease</p>	<p>Task specific tremor: Occurs when performing highly skilled, goal-oriented tasks. For example, while writing or speaking</p>

CHOREA

- Characterized by involuntary, irregular, purposeless, random, nonrhythmic hyperkinesias.
- Movements are spontaneous, abrupt, brief, rapid, jerky, and unsustained.
- Movements are actually random and aimless:
 - Rather than disrupting a voluntary task, it appears as if fragments of movements intrude; in some cases, there is loss of motor tone, known as **“motor impersistence”**, which appears due to lapses in the ability to perform desired action.

- When asked to hold the hands outstretched, there may be constant random movements of individual fingers (**piano-playing** movements).
- If the patient holds the examiner's finger in her fist, there are constant twitches of individual fingers (**milkmaid grip**):
 - **"Jack in the box" tongue**/ harlequin's tongue: Patient is unable to maintain tongue in protruded state and the tongue moves in and out.
- Blink rate is increased.

Causes

- Hereditary: Huntington's disease, benign chorea
- Drugs: Antiparkinsonian drugs, oral contraceptives
- Toxin: Alcohol, carbon monoxide poisoning
- Infections: Sydenham's chorea, encephalitis
- Metabolic: Hyperthyroidism, hypocalcemia
- Immunological: SLE, polyarteritis nodosa
- Vascular
- Pregnancy (Chorea gravidarum)

ATHETOSIS

- Involuntary, irregular, coarse, somewhat rhythmic, and writhing or squirming in character (twisting).
- Hyperkinesias are slower, more sustained, and larger in amplitude than those in chorea.
- May involve the extremities, face, neck, and trunk.
- In the extremities, they affect mainly the distal portions, the fingers, hands, and toes:
 - Affected limbs are in constant motion (athetosis means "without fixed position")
 - Choroathetosis refers to movements that lie between chorea and athetosis in rate and rhythmicity, and may represent a transitional form.

Causes

- Cerebral palsy
- Congenital due to perinatal injury to the basal ganglia.

HEMIBALLISMUS

Dramatic neurologic syndrome of wild, flinging (forceful), incessant (uninterrupted or continuous) movements that occur on one side of the body.

Due to infarction or hemorrhage in the region of the contralateral subthalamic nucleus.

- More rapid and forceful
- Involve the proximal portions of the extremities.
- When fully developed, there are continuous, violent, swinging, flinging, rolling, throwing, flailing (thrashing) movements of the involved extremities.
- They are usually unilateral, and involve one entire half of the body.
- Rarely, they are bilateral (biballismus or paraballismus) or involve a single extremity (monoballismus).

MYOCLONUS

Single or repetitive, abrupt, brief, rapid, lightning-like, jerky, arrhythmic, asynergic, involuntary contractions, involving portions of muscles, entire muscles, or groups of muscles.

- Seen principally in the muscles of the extremities and trunk, but the involvement is often multifocal, diffuse, or widespread.

- May involve the facial muscles, jaws, tongue, pharynx, and larynx.
- Myoclonus may appear symmetrically on both sides. Such synchrony may be an attribute unique to myoclonus.

Myoclonus has been classified in numerous ways including the following:

- i. Positive versus negative;
- ii. Epileptic versus nonepileptic;
- iii. Stimulus sensitive (reflex) versus spontaneous;
- iv. Rhythmic versus arrhythmic;
- v. Anatomically (peripheral, spinal, segmental, brainstem, or cortical)
- vi. By etiology (physiologic, essential, epileptic, and symptomatic)

- Encephalitis
- Juvenile myoclonic epilepsy (JME, Janz syndrome)
- Drug overdose
- Hypnic jerks (appear during the process of falling asleep)
- Hiccup
- Creutzfeldt–Jakob disease
- Subacute sclerosing panencephalitis (SSPE)
- Anoxic encephalopathy (Lance-Adams syndrome)

TIC

A “tic” is an involuntary movement or vocalization that is usually sudden onset, brief, repetitive, stereotyped but nonrhythmic in character, can be suppressed.

Types

Motor tics are associated with movements. Categorized as simple or complex.

Simple motor tics involve only a few muscles usually restricted to a specific body part.

- Examples of simple motor tics include: Eye blinking, shoulder shrugging, facial grimacing, neck stretching, mouth movements, jaw clenching, and spitting.

Vocal/phonic tics are associated with sound

Simple vocal tics consist of sounds that do not form words, such as, throat clearing, grunting, coughing, and sniffing.

Common complex vocal tics include: Repeating words or phrases out of context.

- Coprolalia: Use of socially unacceptable words, frequently obscene.
- Palilalia: Repeating one's own sounds or words.
- Echolalia: Repeating the last-heard sound, word, or phrase.

Gilles de la Tourette syndrome—associated with chronic motor and phonic tics.

DYSTONIA

- Refers to a syndrome of involuntary sustained or spastic muscle contractions involving cocontraction of the agonist and the antagonist.
- The movements are usually slow and sustained, and they often occur in a repetitive and patterned manner.
- They can be unpredictable and fluctuate.

Partial or focal	Generalized
<ul style="list-style-type: none"> • Spasmodic torticollis • Blepharospasm • Oromandibular dystonia • Writers cramp • Hemiplegic dystonia after stroke 	<ul style="list-style-type: none"> • Dystonia musculorum deformans (idiopathic torsion dystonia) • Dopamine responsive dystonia: In childhood and generally involves the legs only. • Drug-induced dystonia (metoclopramide, phenothiazine, haloperidol, chlorpromazine) • Symptomatic dystonia (after encephalitis, Wilsons disease)

Blepharospasm and Oromandibular Dystonia

Involuntary prolonged tight eye closure (blepharospasm) is associated with dystonia of mouth, tongue or jaw muscles (jaw clenching and tongue protrusion).

Writer's Cramp = Mogigraphia = Scrivener's Palsy

Symptoms usually appear when a person is trying to do a task that requires fine motor movements such as writing or playing a musical instrument.

MYOKYMIA

Myokymia, a form of involuntary muscular movement, usually can be visualized on the skin as vermicular or continuous rippling movements.

AKATHISIA

Akathisia is a movement disorder characterized by a feeling of inner restlessness and a compelling need to be in constant motion, as well as by actions such as:

- Rocking while standing or sitting.
- Lifting the feet as if marching on the spot.
- Crossing and uncrossing the legs while sitting.

RESTLESS LEGS SYNDROME/“EKBOM’S SYNDROME”

- Spontaneous, continuous leg movements associated with paresthesia.
- These sensations occur only at the rest and relieved by movement.
- Causes: Familial, lumbar root disease, polyneuropathy, renal failure, and iron deficiency.

SYNKINESIS/MIRROR MOVEMENTS

Mirror movements are characterized by involuntary movements on one side of the body mirroring voluntary movements of the other side.

FASCICULATIONS

Fasciculations are visible, fine and fast, sometimes vermicular contractions of fine muscle fibers that occur spontaneously and intermittently but usually do not generate sufficient force to move a limb. Described as verminosis, because they look like worms moving below the dermis.

Involuntary contraction of the muscle fibers innervated by a motor unit.

Causes of Fasciculations

Fasciculations in healthy subjects	Coffee; exhaustive physical activity/fatigue; stress; benign fasciculations
Fasciculations associated with movement disorders	Spinocerebellar degeneration-type 3; spinocerebellar degeneration-type 36; Parkinsonism (multiple system atrophy, ALS-plus syndromes)
Motor neuron diseases	Amyotrophic lateral sclerosis; progressive spinal muscular atrophies; benign monomelic amyotrophy; postpolio syndrome; Kennedy disease
Systemic diseases	Hyperthyroidism; hypophosphatemia, calcium disorders secondary to hyperparathyroidism, paraneoplastic myopathy

Drugs and/or intoxications by heavy metals pollutants

Organophosphorus poisoning; neostigmine; corticosteroids; succinylcholine; elemental mercury intoxication; atropine, lithium, nortriptyline; flunarizine; isoniazid

SIGNS OF MENINGEAL IRRITATION

Nuchal Rigidity/Meningeal Stiffness

Meningeal tightness is a contracture of the paravertebral muscles, a defense against the secondary pain stemming from inflammation of the meninges.

Painful and permanent, it sometimes presents with the subject lying down, curled up with his or her back to the light, head back, and extremities half-bent. All attempts to flex the head provoke insurmountable and painful resistance. There is extreme neck stiffness; rotational and side-to-side movements are possible but aggravate the headache [Fig. 6D(x).1].



Fig. 6D(x).1: Examination of neck stiffness.

In **Kernig's sign**, patient is kept in supine position, hip and knee are flexed to a right angle, and then knee is slowly extended by the examiner. The appearance of resistance or pain during extension of the patient's knees beyond 135° constitutes a positive Kernig's sign [Figs. 6D(x).2 and 6D(x).3].

Brudzinski's Sign

Josef Brudzinski described 4 maneuvers for the clinical diagnosis of meningitis: The cheek sign, symphyseal sign, Brudzinski's leg sign/reflex, and Brudzinski's neck sign.

1	The cheek sign	A positive cheek sign is elicited by applying pressure on both cheeks inferior to the zygomatic arch that leads to spontaneous flexion of the forearm and arm
2	Symphyseal sign [Fig. 6D(x).4]	A positive symphyseal sign occurs when pressure applied to the pubic symphysis elicits a reflex hip and knee flexion and abduction of the leg
3	Brudzinski's leg sign/reflex [Fig. 6D(x).5]	Brudzinski's contralateral reflex sign consists of reflex flexion of a lower extremity after passive flexion of the opposite extremity
4	Brudzinski's neck sign [Figs. 6D(x).6 and 6D(x).7]	Brudzinski's neck sign is performed with the patient in the supine position. The examiner keeps one hand behind the patient's head and the other on chest in order to prevent the patient from rising. Reflex flexion of the patient's hips and knees after passive flexion of the neck constitutes a positive Brudzinski's sign



Fig. 6D(x).2: Demonstration of Kernig's sign.

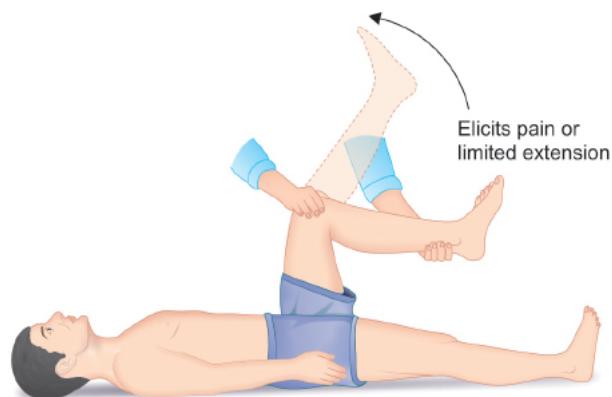


Fig. 6D(x).3: Illustration of Kernig's sign.



Fig. 6D(x).4: Symphyseal sign.



Fig. 6D(x).7: Brudzinski's neck sign.



Fig. 6D(x).5: Brudzinski's leg sign/reflex.



Fig. 6D(x).8: Tripod sign (Amoss's sign).

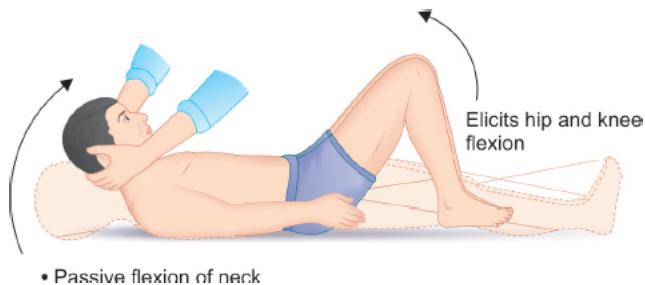


Fig. 6D(x).6: Illustration of Brudzinski's sign.

Tripod sign, also known as the “Amoss’s sign”, is a useful sign of meningeal irritation.

The patient is asked to sit up in bed. This action requires active movement involving flexion of the neck. Although a normal patient sits up without supporting himself, a patient with meningeal irritation tries to sit up by supporting himself with his hands placed far behind him in the bed (like a tripod), in order to take the weight off the spine and prevent its flexion [Fig. 6D(x).8]. Severe meningeal irritation may result in the patient assuming the tripod position with the knees and hips flexed, the back arched lordotically, the neck extended, and the arms brought back in a plane posterior to the pelvis to support the thorax.

MENINGISM

Meningism, also called meningismus or pseudomeningitis, is a set of symptoms similar to those of meningitis but not caused by meningitis. Whereas meningitis is inflammation of the meninges (membranes that cover the central nervous system), meningism is caused by nonmeningitic irritation of the meninges usually associated with acute febrile illness, especially in children and adolescents.

Causes

Meningism:

- Meningitis
- Subarachnoid hemorrhage.

Other conditions that mimic meningism (also resist cervical rotation):

- Cervical spondylosis
- After cervical fusion
- Parkinson's disease
- Raised intracranial pressure especially if there is impending tonsillar herniation
- Acute dystonic reaction

- Tetanus
- Strychnine poisoning.

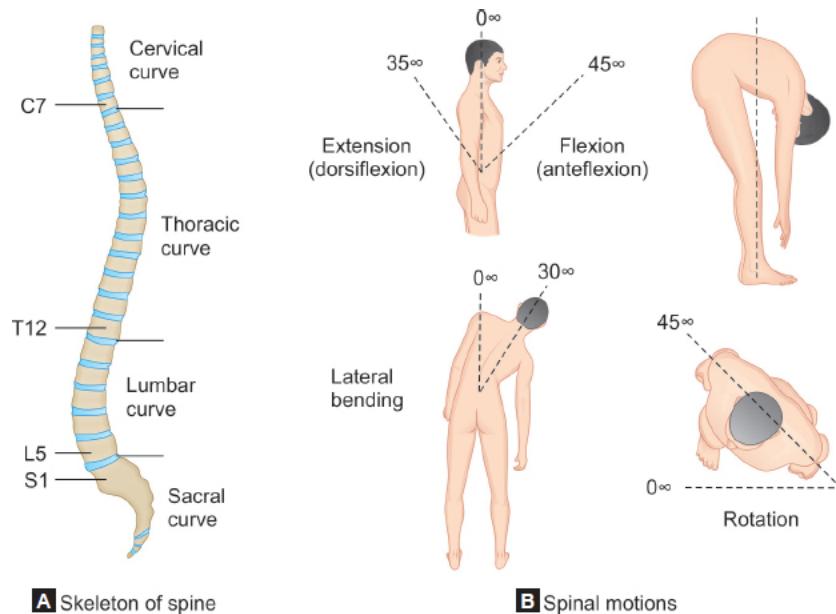
Intermittent neck stiffness is characteristic of Arnold-Chiari malformation.

EXAMINATION OF SKULL

- Size of skull—microcephaly, macrocephaly
- Shape/deformities
- Tenderness—fracture/metastasis
- Crackpot sound on percussion—hydrocephalus
- Bruits on auscultation—arteriovenous malformation (AVM), hemangioma.

EXAMINATION OF SPINE

- Inspection—deformities, curvature—kyphosis, scoliosis, lordosis, dimple, tuft of hair, Pott's spine, and meningioma
- Palpation—tenderness, paraspinal spasm, and deformities
- Movements [Figs. 6D(x).9A and B].



Figs. 6D(x).9A and B: Movements of spine. (Details discussed under rheumatology section)

AUTONOMIC NERVOUS SYSTEM TESTING

Common autonomic symptoms	Signs
<ul style="list-style-type: none"> • Orthostatic intolerance • Dizziness • Lightheadedness • Fatigue 	<ul style="list-style-type: none"> • Pupils—mid-dilated sluggish reacting pupil • Pedal edema • Resting tachycardia • Postural hypotension • Palpable urinary bladder • Sweating abnormalities

<ul style="list-style-type: none"> • “Coat hanger” headache • Nausea • Palpitations • Near syncope and syncope <p>Genitourinary</p> <ul style="list-style-type: none"> • Bladder urgency or frequency • Incontinence • Nocturia • Erectile dysfunction • Ejaculatory disturbances 	
Common autonomic symptoms	Signs
<p>Gastrointestinal</p> <ul style="list-style-type: none"> • Diarrhea • Constipation • Fecal incontinence • Postprandial fullness, cramping, or bloating <p>Sudomotor</p> <ul style="list-style-type: none"> • Hyperhidrosis • Hypohidrosis and anhidrosis 	
	Tests
<p>Cardiovagal innervation (parasympathetic innervation)</p> <ul style="list-style-type: none"> • Heart rate (HR) response to deep breathing • Valsalva ratio, and • HR response to standing (30:15 ratio) 	<p>“Spoon test”: A kitchen soup spoon, with its curved surface resting on the skin, was held between the thumb and forefinger, and was drawn slowly on the skin, using sufficient energy to overcome its weight without lifting it from the skin. When “sympathectomized” skin was crossed, the pull was smooth and unopposed; but where sweat gland innervation and sympathetic function was intact, the skin was moist, and the flow of the spoon was interrupted, and became sticky requiring readjustment of the strength of pull</p>
<p>Adrenergic</p> <ul style="list-style-type: none"> • Beat-to-beat blood pressure (BP) responses to the Valsalva maneuver, sustained handgrip/diastolic hand grip test ** and • BP and HR responses to tilt-up or active standing 	<p>“Sustained handgrip test (SHT): This parameter indicates cardiac sympathetic response and DBP response to the sustained handgrip test—taken as the difference between the DBP just before release of handgrip and the mean of three resting DBP readings. The change in mean DBP in response to sustained handgrip test was interpreted as:</p> <ul style="list-style-type: none"> • ≥ 16 mm Hg was taken as normal • 11–15 mm Hg as borderline • ≤ 10 mm Hg as abnormal
<p>Sudomotor:</p> <ul style="list-style-type: none"> • Quantitative sudomotor axon reflex test (QSART) • Thermoregulatory sweat test (TST) • Sympathetic skin response (SSR), and • Silastic sweat imprint 	

Head-Up Tilt-Table Testing

The patient lies supine on the tilt table. Beat-to-beat and oscillometric BP instruments are attached to each arm. ECG monitoring should take place throughout the test. Once the patient is comfortable, with feet resting on the footboard, a baseline BP is recorded for at least 3 minutes. The patient is then slowly tilted upright to an angle of 60–80°.

During testing, the patient is asked to report any symptoms. Both BP and HR are recorded throughout tilt-table testing, after which the patient is returned to a horizontal supine position.

Three well-described patterns of neurally-mediated syncope can occur during head-up tilt-table testing:

1. Vasodepression resulting in hypotension without bradycardia.
2. Cardioinhibition with a marked bradycardia (fewer than 40 beats/min) with or without significant hypotension.
3. Mixed, with both bradycardia and hypotension.

Valsalva Ratio

The Valsalva maneuver consists of respiratory strain which increases intrathoracic and intra-abdominal pressures and alters hemodynamic and cardiac functions.

- The patient is supine or with head slightly elevated to about 30°.
- Have the patient strain against 40 mm Hg applied for 15 seconds by blowing into a mouthpiece attached to a sphygmomanometer.
- Following cessation of the Valsalva strain, the patient relaxes and breathes at a normal comfortable rate.
- The ECG is monitored during the strain and 30–45 seconds following its release.
- The maximal heart rate of phase II actually occurs about 1 seconds following cessation of the strain.
- The minimal heart rate occurs about 15–20 seconds after releasing the strain.

DISEASES ASSOCIATED WITH AUTONOMIC DYSFUNCTION [TABLE 6D(X).1]

Table 6D(x).1: Diseases commonly associated with autonomic dysfunction.

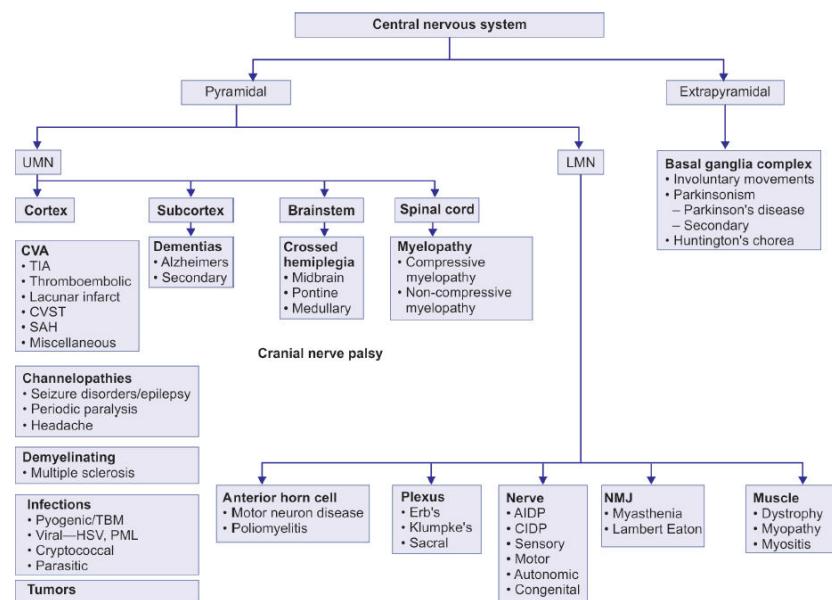
- **Preganglionic autonomic failure:**
 - Multiple system atrophy
 - Parkinson's disease with autonomic failure
- **Ganglionic and postganglionic disorders**
 - Pure autonomic failure
- **Peripheral neuropathies and neuronopathies with autonomic dysfunction**
 - *Acute and subacute (preganglionic and postganglionic):*
 - Acute pandysautonomia
 - Guillain-Barré syndrome
 - Paraneoplastic pandysautonomia
 - Others (porphyria, toxins, durgs)
 - *Chronic small-fiber (postganglionic) neuropathies:*
 - Diabetes
 - Amyloidosis
 - Hereditary (familial dysautonomia, Fabry's disease)
 - *Subacute or chronic sensory and autonomic ganglionopathies:*
 - Paraneoplastic
 - Sjogren's syndrome
 - *Other peripheral neuropathies:*
 - Infections (human immunodeficiency virus)
 - Connective tissue disease (systemic lupus erythematosus)
 - Metabolic-nutritional (alcohol, uremia, vitamin B₁₂ deficiency)

E. APPROACH TO COMMON NEUROLOGICAL CASES

Approach to following cases have been discussed in this section:

1. Approach to cerebrovascular accident
2. Approach to spinal cord diseases
3. Approach to neuropathy
4. Approach to movement disorders

Flowchart 6E.1: Diseases stratification of nervous system.



(UMN: upper motor neuron; LMN: lower motor neuron; CVA: cerebrovascular accident; TIA: transient ischemic attack; CVST: cerebral venous sinus thrombosis; SAH: subarachnoid hemorrhage; TBM: tuberculous meningitis; HSV: herpes simplex virus; PML: promyelocytic leukemia; SACD: subacute combined degeneration; AIDP: acute inflammatory demyelinating polyneuropathy; CIDP: chronic inflammatory demyelinating polyneuropathy; NMJ: neuromuscular junction)

1. APPROACH TO CEREBROVASCULAR ACCIDENT

Table 6E.1: Signs of upper and lower motor neuron disease.

Sign	Upper motor neuron	Lower motor neuron
Atrophy	None (rarely disuse atrophy)	Severe wasting
Fasciculations	None	Common
Tone	Hypertonia—rigidity/spasticity	Decreased (hypotonia)
Distribution of weakness	Distal predominant/regional	Predominantly proximal (except neuropathy)/segmental
Tendon reflexes	Exaggerated/hyperactive	Hypoactive/lost
Babinski sign	Present	Absent

Flexor spasms, clonus	Present	Absent
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- A stroke (cerebrovascular accident is a vague term which should be avoided) is defined as a syndrome of rapid (abrupt) onset of a neurologic deficit that is attributable to a focal vascular cause (**Flowchart 6E.2**).
- World Health Organization (WHO) definition: Stroke is a “rapidly developing clinical signs of focal (or global) disturbance of cerebral function, with symptoms lasting for 24 hours or longer or leading to death, with no apparent cause other than of vascular origin”.
- **Progressing stroke (or stroke in evolution)**: It is a stroke in which the focal neurological deficit worsens after the patient first presents. It may be due to increasing volume of infarction, secondary hemorrhage in the infarcted area, or increasing cerebral edema.
- **Complete stroke**: Rapid onset with persistent focal neurological deficit which does not progress beyond 96 hours.
- **Evolving stroke**: Gradual stepwise development of neurological deficits. Focal cerebral deficits that develop slowly (over weeks to months) are unlikely to be due to stroke and are more suggestive of tumor or inflammatory or degenerative disease.

Terminologies

Several terms are used to classify strokes mainly based on the duration and evolution of symptoms.

- **Transient ischemic attack (TIA)**: Described later
- **Reversible ischemic neurological deficit (RIND)**: In some cases, deficits last for longer than 24 hours but resolve completely or almost completely within a few days.
- **Stuttering hemiplegia**: Internal carotid lesions are characterized by repeated episodes of TIA followed by fully evolved stroke.

Flowchart 6E.2: Types of stroke.

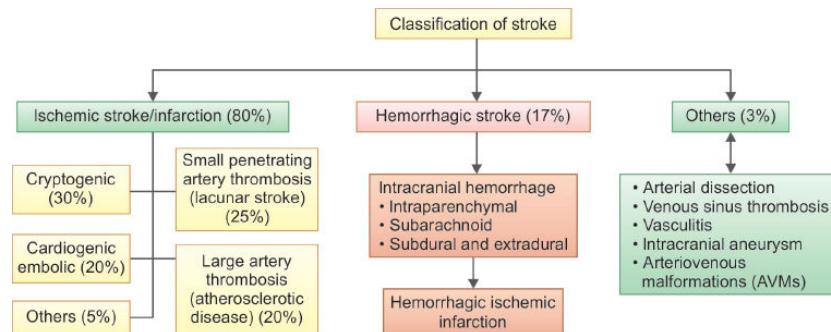


Table 6E.2: Risk factor for stroke.

Risk factors in patients of all age groups	
High-risk	
<ul style="list-style-type: none"> • Hypertension (including isolated systolic) • Smoking • Diabetes mellitus • Atrial fibrillation • Drugs: Cocaine, amphetamine • Dilated cardiomyopathy • Endocarditis 	<ul style="list-style-type: none"> • High cholesterol • Obesity • Vasculitis: Systemic vasculitides [e.g. polyarteritis nodosa—PAN], granulomatosis with polyangiitis (Wegener's) etc., primary CNS vasculitis • Meningitis (syphilis, tuberculosis, fungal, bacterial, zoster)
Low-risk	
Migraine	<ul style="list-style-type: none"> • Recent myocardial infarction

• Oral contraceptives or alcohol	• Prosthetic valve
• Patent foramen ovale	• Sleep apnea
Additional risk factors that are more common in young patients	
Hypercoagulable disorders	
• Protein C and S deficiencies	• Sickle-cell anemia
• Antithrombin III deficiency	• Hyperhomocysteinemia
• Antiphospholipid antibody syndrome	• Thrombotic thrombocytopenic purpura
• Factor V Leiden mutation	• Arterial dissection
• Prothrombin G20210A heterozygous mutation	• Infections (e.g. syphilis, HIV)
	• Systemic malignancy

(CNS: central nervous system; HIV: human immunodeficiency virus)

Table 6E.3: Causes for young stroke.

<ul style="list-style-type: none"> Cardiac <ul style="list-style-type: none"> Congenital heart disease, patent foramen ovale Atrial myxoma Atrial fibrillation and other arrhythmia Cardiomyopathy, myocarditis, myocardial infarction Cardiac surgery, cardiac catheterization Endocarditis, rheumatic heart disease Prosthetic valve Hematologic <ul style="list-style-type: none"> Sickle cell disease, iron deficiency anemias, polycythemia vera Hypercoagulable states <ul style="list-style-type: none"> Inherited prothrombotic states, protein C and S deficiency, antithrombin III deficiency, factor V Leiden gene mutation, prothrombin gene mutation Antiphospholipid antibody syndrome Hyperhomocysteinemia Myeloproliferative disorders (e.g. leukemia, lymphoma) Pregnancy exposure to hormonal treatments, such as anabolic steroids and erythropoietin, nephrotic syndrome 	<ul style="list-style-type: none"> Vascular <ul style="list-style-type: none"> Noninflammatory <ul style="list-style-type: none"> Arterial dissection Secondary to connective tissue disease (Ehlers-Danlos, Marfan) Moyamoya disease Hypertension Radiation vasculopathy Vasculitis and postinfectious vasculopathy Migraine Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) Fibromuscular dysplasia, Susac's syndrome, Sneddon's syndrome, Fabry's disease Inflammatory <ul style="list-style-type: none"> Takayasu arteritis Giant cell arteritis Kawasaki disease Polyarteritis nodosa Human immunodeficiency virus (HIV) Bacterial meningitis <p>Illicit drug use: Cocaine, amphetamine</p>
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Table 6E.4: Differences between hemorrhagic, thrombotic, and embolic strokes.

Feature	Hemorrhagic stroke (Intracerebral or subarachnoid hemorrhage)	Ischemic stroke	
		Thrombotic	Embolic
Time of onset of stroke	During activity	Suddenly and often during sleep or in the early morning (4 AM)	Any time (usually during activity)
Rapidity of onset and progression	Over minutes and hours	On waking up or over hours	Rapid within seconds deficit maximum at onset
Transient ischemic attacks (TIAs)	Absent	Precedes stroke	Precedes stroke
Vomiting	Recurrent	Absent or occasional	Absent or occasional
Headache	Severe and prominent	Mild or absent	Mild or absent
Early resolution (within minutes or days)	Unusual	Variable	Possible
Meningeal irritation	May be present	Absent	Absent

Carotid bruit and absence of pulse	Not observed	Highly supports the diagnosis	Possible
Valvular heart disease and atrial fibrillation	Not found	Unusual	Highly supports the diagnosis
CT scan findings	Hemorrhage	<ul style="list-style-type: none"> • Early stage: Normal • Later: Pale infarct 	<ul style="list-style-type: none"> • Early stage: Normal • Later: Pale infarct

Localization of Stroke

Site of lesion	Predominant clinical features
Cortex	<ul style="list-style-type: none"> • Monoplegia common (brachial-MCA territory; crural-ACA territory) • Hemiplegia (may be present but never dense) • Contralateral 7th cranial nerve palsy (UMN variant) • Seizures • Aphasias (in dominant hemisphere) • Apraxias (in nondominant hemisphere)
Subcortical (usually secondary to hypoperfusion)	<ul style="list-style-type: none"> • Monoplegias common • Transcortical aphasias common
Internal capsule lesion	<ul style="list-style-type: none"> • Contralateral hemiplegia (dense) • Contralateral hemisensory loss • 7th cranial nerve palsy (UMN variant) • Homonymous hemianopia • Broca's like aphasia (only site to have subcortical aphasia). <p>Note: Most common etiology being ischemic and hence is territory specific. Since different parts of internal capsule has blood supply from different blood vessels, all the above-mentioned features may not be present at same time. However, if present, it suggests hemorrhage or tumor compressing internal capsule</p>
Brainstem lesion	<ul style="list-style-type: none"> • Discussed in separate table
High cervical cord lesion (Brown-Séquard syndrome)	<ul style="list-style-type: none"> • Ipsilateral hemiplegia • Ipsilateral loss of posterior column sensation • Contralateral loss of pain and temperature sensation • Usually no cranial nerve involvement

(ACA: anterior cerebral artery; MCA: middle cerebral artery; UMN: upper motor neuron)

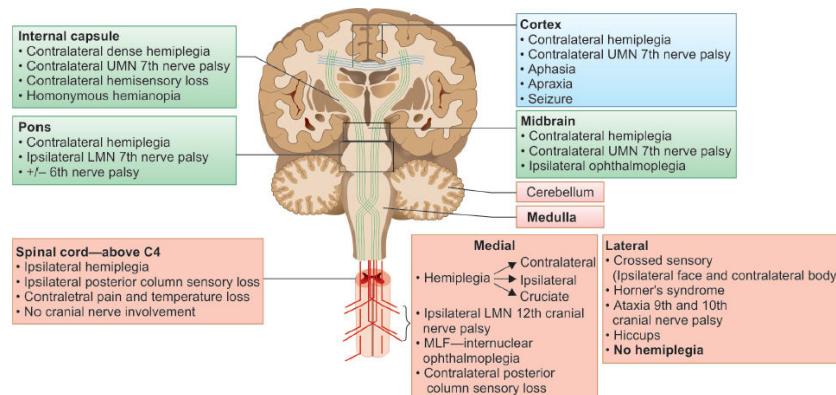


Fig. 6E.1: Localization of hemiplegia.

(UMN: upper motor neuron; LMN: lower motor neuron; MLF: medial longitudinal fasciculus)

Middle cerebral artery lesions and clinical features				
<i>Internal carotid artery</i>		<i>M1 branch of MCA</i>	<i>Stem of MCA</i>	<i>M2 branches of MCA</i>
Both anterior cerebral artery (ACA) and middle cerebral artery (MCA) territory involved along with ophthalmic artery causing amaurosis fugax		<ul style="list-style-type: none"> • Global aphasia • Dense hemiplegia (as internal capsule is also involved due to involvement of lenticulostriate branches of MCA) 	<ul style="list-style-type: none"> • Global aphasia • Internal capsule spared 	<ul style="list-style-type: none"> • Superior division • Inferior division (differences described below)
M2 stroke				
<i>Division of M2</i>		<i>Superior division</i>	<i>Inferior division</i>	
Motor involvement		Face, arm > leg	Nil	
Sensory		Face, arm	Nil	
Vision		Nil	Quadrantanopia	
Language		Broca's aphasia	Wernicke's aphasia	
Nondominant		Hemineglect	Constructional apraxia	
Brainstem syndromes				
<i>Site of lesion/syndrome</i>	<i>Blood supply and tracts involved</i>		<i>Ipsilateral features</i>	<i>Contralateral features</i>
Midbrain				
Benedict's syndrome (Claude's + Weber)	Interpeduncular branches of basilar artery, PCA—posterior cerebral artery (midbrain tegmentum—CN III fibers; red nucleus; CST; SCP)		Ipsilateral CN III palsy	Ataxia + Hyperkinesia and tremor ("rubral tremor") + Hemiparesis
Claude's syndrome	PCA (midbrain tegmentum—CN III fibers; red nucleus; SCP)		Ipsilateral CN III palsy	Ataxia + Tremor ("rubral tremor")
Weber's syndrome	Paramedian branches of the basilar artery, PCA		Ipsilateral CN III palsy	Hemiparesis
Nothnagel syndrome	Basilar penetrating artery, mesencephalic artery (midbrain tectum Ipsilateral or bilateral CN III)		Oculomotor palsies; ataxia	
Parinaud syndrome	Midbrain dorsum (quadrigeminal plate region; pretectum; periaqueductal gray matter)		Impaired upgaze; convergence retraction nystagmus; dilated pupils with light near dissociation	
Top of basilar artery syndrome	<ul style="list-style-type: none"> • Midbrain • Thalamus • Portion of temporal and occipital lobe involved 		<ul style="list-style-type: none"> • Behavioral abnormalities • Ocular finding • Visual defects • Pupillary abnormalities • Motor deficits 	
Artery of Percheron stroke	Single thalamic perforating artery from the proximal PCA		<ul style="list-style-type: none"> • Altered sensorium • Vertical gaze palsy • Memory impairment 	
Pons				
Raymond Ceston	Long circumferential branch of basilar artery (CN VI; CST)		6th nerve palsy	Hemiparesis

syndrome			
Millard-Gubler syndrome	Basilar artery (CN VII; CST)	7th nerve palsy (\pm Lateral rectus palsy)	Hemiparesis
Foville's syndrome	Basilar artery (CN VII; lateral gaze center, CST)	7th nerve palsy + Horizontal gaze palsy	Hemiparesis
Pierre-Marie-Foix syndrome	AICA	<ul style="list-style-type: none"> • 6th + 7th nerve palsy • Horner's syndrome 	Hemiparesis
Medulla			
Wallenberg syndrome (lateral medullary syndrome)	Vertebral artery > PICA (Lateral medullary) Tegmentum—spinal tract of CN V and its nucleus; nucleus ambiguus; emerging fibers of CNs IX and X; LST; descending sympathetic fibers; vestibular nuclei; inferior cerebellar peduncle; afferent spinocerebellar tracts; lateral cuneate nucleus)	<ul style="list-style-type: none"> • Loss of pain and temperature of face • Ipsilateral decreased corneal reflex • Ipsilateral weakness of soft palate • Ipsilateral loss of gag reflex • Ipsilateral paralysis of vocal cord • Ipsilateral central Horner's syndrome • Nystagmus • Cerebellar ataxia of ipsilateral limbs • Lateropulsion • Hiccups 	Loss of pain and temperature of body
Dejerine syndrome (medial medullary syndrome)	Vertebral > anterior spinal artery	Ipsilateral tongue weakness	Hemiparesis
Avellis' syndrome	Medullary tegmentum	Ipsilateral palatal and vocal cord weakness;	Loss of pain and temperature
Jackson's syndrome	Medullary tegmentum	Ipsilateral flaccid paralysis of soft palate, pharynx, and larynx; flaccid weakness and atrophy of SCM and trapezius (partial), and of the tongue	
Schmidt's	Lower medullary tegmentum	Ipsilateral paralysis of soft palate, pharynx, and larynx; flaccid weakness and atrophy of SCM and trapezius (partial)	
Céstan-Chenais	Due to vertebral artery occlusion below origin of the PICA; (nucleus ambiguus; ICP; sympathetics; CST; ML)	Ipsilateral weakness of soft palate, pharynx, and larynx; cerebellar ataxia; Horner's syndrome	Contralateral hemiparesis with loss of posterior column function
Internuclear ophthalmoplegia (INO)	MLF lesion in the midbrain	Ipsilateral adduction palsy	Contralateral gaze evoked nystagmus
Wall eyed bilateral internuclear ophthalmoplegia (WEBINO)	Bilateral MLF lesion in the brain	Bilateral adduction deficit and primary gaze position exotropia	
PCA syndromes			
Gerstmann syndrome	Parietal lobe	Inability to write (dysgraphia or agraphia), the loss of the ability to do mathematics (acalculia), the inability to identify one's own or another's fingers (finger	

		agnosia), and inability to make the distinction between the right and left side of the body.	
Anton syndrome	Bilateral occipital cortex involvement due to bilateral PCA infarct	Anton's syndrome describes the condition in which patients deny their blindness despite objective evidence of visual loss, and moreover confabulate to support their stance	Anosognosia (or lack of awareness of defect) and confabulation
Balint syndrome	Parieto-occipital lobes on both sides of the brain	Inability to perceive the visual field as a whole (simultanagnosia), difficulty in fixating the eyes (oculomotor apraxia), and inability to move the hand to a specific object by using vision (optic ataxia)	

(CN: cranial nerve; CST: corticospinal tract; SCP: superior cerebellar peduncle; AICA: anterior inferior cerebellar artery; PICA: posterior inferior cerebellar artery; LST: lateral spinothalamic tract ; SCM: sternocleidomastoid muscle; ICP: intracranial pressure; CST: corticospinal tract ; ML: medial lemniscus ; MLF: medial longitudinal fasciculus; PCA: posterior cerebral artery)

Transient Ischemic Attacks

Transient ischemic attack (TIA) is characterized by a brief episode of neurological dysfunction (sudden loss of function) in which symptoms and signs resolve completely after a brief period within 24 hours (usually within 30 minutes).

- Transient ischemic attack is defined as a transient episode of neurologic dysfunction caused by focal brain, spinal cord, or retinal ischemia, without acute infarction. However, TIAs may herald a stroke.
- Newly proposed definition classifies those with new brain infarction as ischemic strokes regardless of whether symptoms persist.

Clinical features: Hemiparesis and aphasia are most common. Other features include amaurosis fugax (sudden transient loss of vision in one eye), hemisensory loss, hemianopic visual loss, diplopia, vertigo, vomiting, choking and dysarthria, ataxia, etc.

Types of Transient Ischemic Attack

- Large artery low-flow TIA—recurrent, short lasting episodes of stereotyped symptoms (shotgun TIA/thrombotic TIA)
- Embolic TIA—longer lasting less frequent episodes with varied symptoms, changing territories
- Lacunar TIA.

Small Vessel (Lacunar) Stroke

- Small penetrating arterial branches of 200–800 µm in diameter, supply the deep brain parenchyma. Each of these small branches can be occluded either by atherosclerotic disease at its origin or by the development of occlusive vasculopathy—lipohyalinotic thickening (consequence of hypertension) (**Table 6E.5**).
- Thrombosis of these vessels causes small infarcts that are referred to as lacunae. These infarcts range in size from 0.2 mm to 15 mm in diameter.

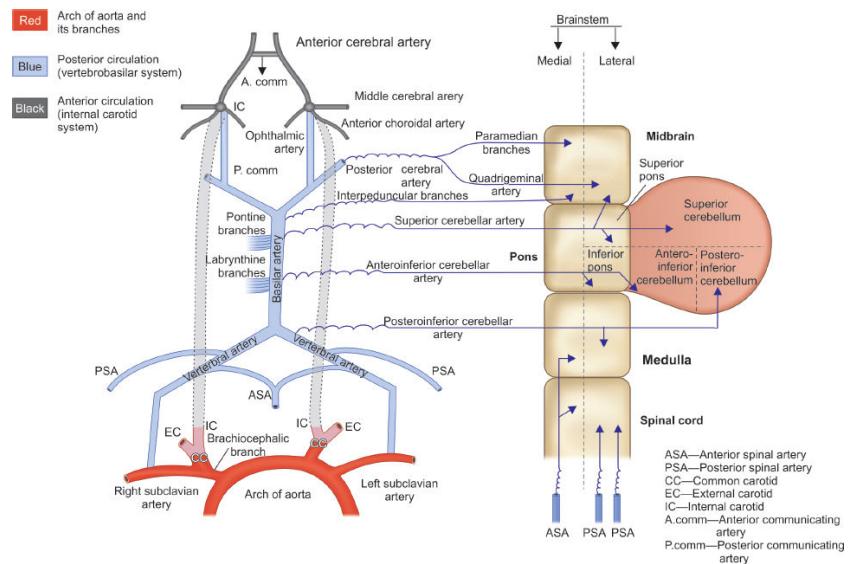


Fig. 6E.2: Cerebrovascular system (a comprehensive diagram of arterial system).

Internal capsule	Anterior limb	Genu	Posterior limb	Sub-lentiform	Retro-lentiform
Upper part	Lenticulostriate branches of MCA				
Lower part	ACA (Recurrent artery of Heubner)	ACA IC P. Comm	AChA	AChA PCA	PCA

Fig. 6E.3: Blood supply of internal capsule.

(ACA: anterior cerebral artery; MCA: middle cerebral artery; PCA: posterior cerebral artery; AChA: anterior choroidal artery; IC: internal carotid artery (direct branches); P. Comm: posterior communicating artery)

Table 6E.5: Signs and symptoms of lacunar stroke depending on location of lesion.			
Syndrome	Signs/symptoms	Localization	Vascular supply
Pure motor	Contralateral hemiparesis or hemiplegia. Affects face, arm and leg equally	<ul style="list-style-type: none"> Posterior limb of internal capsule Corona radiata-Basis pontis 	Lenticulostriate branches of the middle cerebral artery (MCA) or perforating arteries from basilar artery
Pure sensory	Contralateral hemisensory loss. Persistent or transient numbness and/or tingling on one side of the body	<ul style="list-style-type: none"> Ventral posterolateral (VPL) nucleus of thalamus 	Lenticulostriate branches of MCA. Small thalamoperforators of posterior cerebral artery (PCA)
Mixed sensorimotor	Contralateral weakness and numbness.	Thalamus and	Lenticulostriate branches of MCA

	Hemiparesis or hemiplegia with ipsilateral sensory impairment	adjacent posterior limb of internal capsule	
Dysarthria-clumsy hand	Slurred speech and weakness of contralateral hand (fine motor)	Basis pontis	Basilar artery perforators
Ataxic-hemiparesis	Combination of cerebellar and motor symptoms. Contralateral hemiparesis and ataxia out of proportion to weakness	<ul style="list-style-type: none"> Internal capsule-posterior limb Basis pontis Corona radiata 	<ul style="list-style-type: none"> Lenticulostriate branches of MCA Perforating arteries of basilar artery
Hemiballismus/hemic chorea	Contralesional limb flailing/dyskinesia	Subthalamic nucleus	Perforating arteries of anterior choroidal or posterior communicating artery (PCom)

2. APPROACH TO SPINAL CORD DISEASES

Spinal Cord Anatomy

The spinal cord originates at the medulla and continues caudally to terminate at the filum terminale, a fibrous extension of the conus medullaris that terminates at the coccyx.

The adult spinal cord is approximately 45 cm long, oval or round in shape, and enlarged in the cervical and lumbar regions, where neurons that innervate the upper and lower extremities, respectively are located. The meninges that cover the spinal cord are continuous with those of the brainstem and cerebral hemispheres.

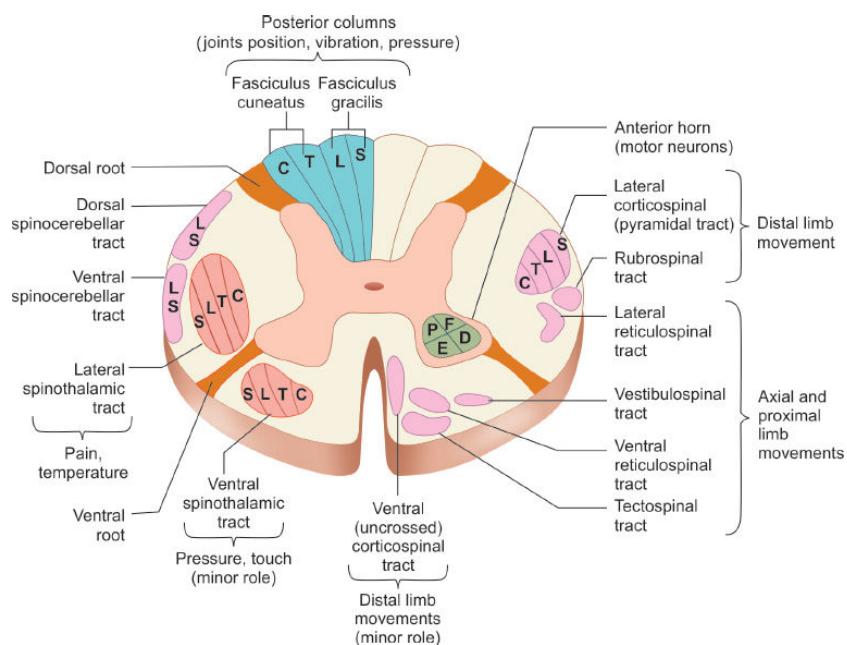


Fig. 6E.4: Tracts of spinal cord.

- The adult cord consists of 31 segments, each containing an exiting ventral motor root and entering dorsal sensory root.
- During embryologic development, growth of the cord lags behind that of the vertebral column, and in the adult spinal cord ends at approximately the first lumbar vertebral body. The lower spinal nerves take an increasingly downward course to exit via the appropriate intervertebral foramina.

- The first seven pairs of cervical spinal nerves exit above the same-numbered vertebral bodies, whereas all the subsequent nerves exit below the same-numbered vertebral bodies; this situation is due to the presence of eight cervical spinal cord segments but only seven cervical vertebrae.
- The approximate relationship between spinal cord segments and the corresponding vertebral bodies is shown in the following table:

Spinal cord level	Corresponding vertebral body
• Upper cervical	• Same as cord level
• Lower cervical	• 1 level higher
• Upper thoracic	• 2 levels higher
• Lower thoracic	• 2 to 3 levels higher
• Lumbar	• T 10 to T11
• Sacral	• T12 to L1
• Coccygeal	• L1

Features Suggestive of Involvement of Spinal Cord

- Presence of sensory deficit and/or motor weakness in both lower limbs and/or upper limbs.
- Bladder and bowel involvement
- Brown-Séquard type of clinical picture
- Presence of definite sensory level
- Vertebral pain.

VASCULAR SUPPLY OF SPINAL CORD (FIG. 6E.5)

- The anterior spinal artery:** Union of the anterior spinal branches of the vertebral artery and descends within the anterior median fissure.
- The two posterior spinal arteries:** Originate from the vertebral arteries and descend in the posterolateral sulcus.
- By themselves not sufficient and depend on feeder arteries that join them along their course (6–10 join the ASA and 10–20 join the PSA).
- Thirty-one pairs of small radicular arteries:** Supply corresponding nerve roots.
- Some of them give a branch to spinal arteries:** The radiculospinal branches.
- C1-4:** Vertebral artery.
- C5-T2:** Ascending and deep cervical artery.
- T3 to T8:** Intercostal artery.
- T9 and below:** Artery of Adamkiewicz—supplies most of the lower one-third of spinal cord; arises from a left-sided intercostal or lumbar artery (T8-L3).

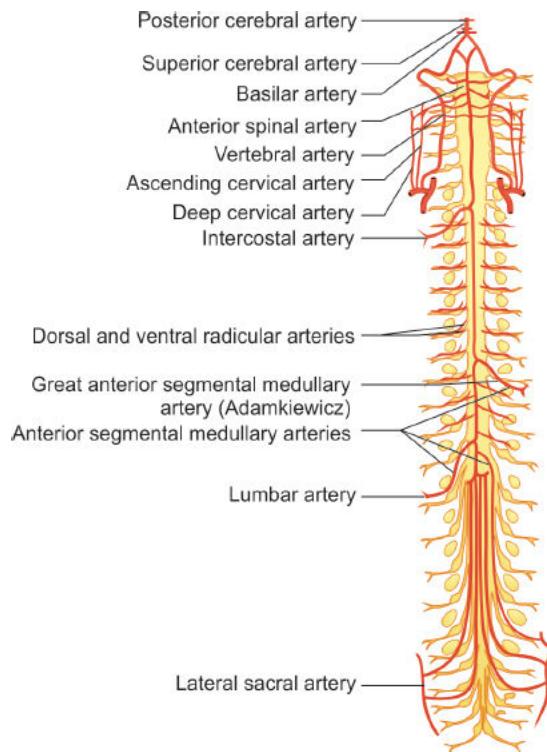


Fig. 6E.5: Vascular supply of spinal cord.

DIFFERENTIATION BETWEEN COMPRESSIVE AND NONCOMPRESSIVE MYELOPATHY

Features	Compressive	Noncompressive
Bony deformity	+	-
Bony tenderness	+	-
Girdle like sensation	+	-
Upper level of sensory loss	+	-
Zone of hyperesthesia	+	-
Root pain	+	-
Onset and progress	Gradual	May be acute
Symmetry	Asymmetrical	Majority are symmetrical
Flexor spasm	Common	Usually absent
Pattern of neurodeficit	U-shaped (Ellsberg phenomenon)	Bilaterally symmetrical
Bladder and bowel movement	Late	Early (acute transverse myelitis)
Selective tract involvement	Rare	Usually seen

Flowchart 6E.3 depicts the types of spinal cord diseases.

Compressive myelopathies examples

<ul style="list-style-type: none"> • Trauma • Tumor • Tuberculosis • Myeloma • Metastasis 		
Extramedullary extradural <ul style="list-style-type: none"> • Caries spine • Metastasis • Intervertebral disc prolapse • Spondylosis • Fluorosis • Trauma to vertebra • Epidural abscess • Epidural hematoma • Hematomyelia 	Extramedullary intradural <ul style="list-style-type: none"> • Meningioma • Neurofibroma • Schwannoma • Patchy arachnoiditis • Arteriovenous malformations • Lipoma • Sarcoma • Dermoid 	Intramedullary <ul style="list-style-type: none"> • Ependymoma • Chordoma • Glioma

Flowchart 6E.3: Types of spinal cord diseases.



Noncompressive myelopathies examples	
Inflammatory	
• Infectious—viral, bacterial, fungal, and parasitic	
• Autoimmune—SLE, Sjogren's, sarcoidosis, Bechet syndrome, MCTD, polyarteritis nodosa, pANCA positive vasculitis	
• Demyelinating—MS, NMO, ADEM, and postviral postvaccinal	
• Paraneoplastic—lung carcinoma, breast, and ovary	
• Encephalomyelitis	
Noninflammatory	
• Inherited—HSP, inherited metabolic disorders	
• Metabolic—vitamin B ₁₂ , copper, folate and vitamin E deficiency—AIDS associated	
• Toxic—cassava, lathyrism, fluorosis, SMON, nitrous oxide, TOCP, and Konzo	
• Vascular—anterior spinal artery thrombosis, AVM, and dural arteriovenous fistula	
• Degenerative—familial spastic paraparesis	
• Physical agents—electrical injury, Caisson's disease, and radiation myelopathy	

(SLE: systemic lupus erythematosus; MCTD: mixed connective tissue disease; pANCA: perinuclear antineutrophil cytoplasmic antibodies; MS: multiple sclerosis; NMO: neuromyelitis optica; ADEM: acute disseminated encephalomyelitis; HSP: hereditary spastic paraparesis; AIDS: acquired immunodeficiency syndrome; SMON: subacute myelo-optic neuropathy; TOCP: triorthocresyl phosphate; AVM: arteriovenous malformation)

Discriminate Between Extramedullary and Intramedullary Lesions

Features	Extramedullary	Intramedullary
Radicular pain	Common Intradural: Unilateral Extradural: Bilateral	Unusual
Vertebral pain	Common (extradural)	Unusual
Funicular pain	Rare	Common

Motor deficit	Ascending motor weakness, i.e. sacral → lumbar → thoracic → cervical	Descending pattern of loss, i.e. cervical → thoracic → lumbar → sacral
Upper motor neuron involvement	Early and prominent	Less pronounced; late feature
Lower motor neuron involvement	Segmental	Marked with widespread atrophy, fasciculations seen
Reflexes	Brisk early feature	Less brisk, later feature
Sensory deficit	Ascending sensory loss, i.e. sacral → lumbar → thoracic → cervical Saddle anesthesia Hemisection—contralateral loss of pain and temperature, ipsilateral loss of joint position	<ul style="list-style-type: none"> Descending pattern of loss, i.e. cervical → thoracic → lumbar → sacral Dissociative sensory loss Suspended sensory loss (Jacket pattern)
Sacral sensation	Lost (early)	Sacral sparing
Autonomic involvement (bladder and bowel)	Late	Early
Trophic changes	Usually not marked	Common
Vertebral tenderness	May be present (extradural)	No bony tenderness in vertebrae
Changes in CSF	Frequent (increased protein, cells)	Rare

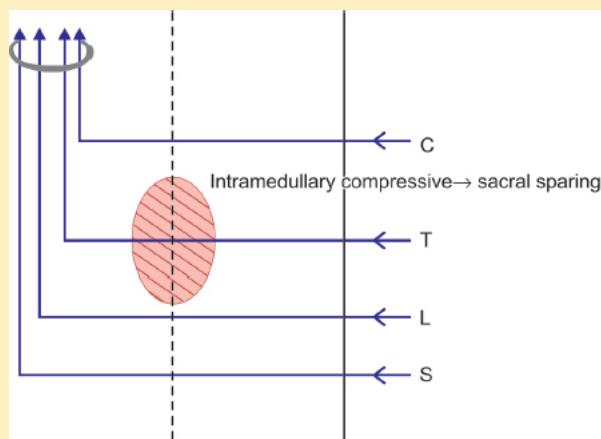


Fig. 6E.6: Arrangement of motor fibers.

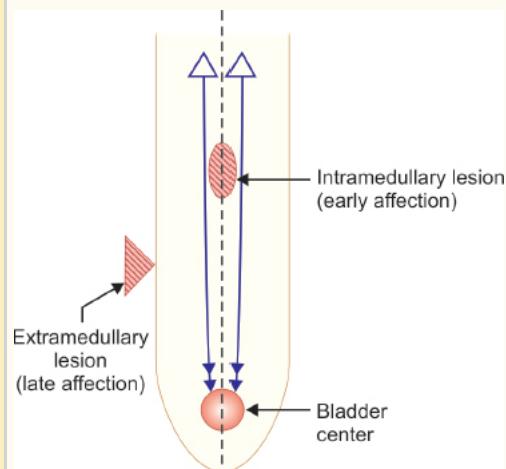


Fig. 6E.7: Bladder involvement in spinal cord disease.

Differences Between Presentation of Intradural and Extradural Lesion

Features	Extradural	Intradural
Mode of onset	Usually symmetrical	Asymmetrical
Root pain	Less common	More common
Spinal tenderness	Common	Uncommon
Spinal deformity	Present	Absent

Patterns of Spinal Cord Disease

1. Complete cord transection syndrome

2. Brown-Sequard syndrome/hemisection of the cord
3. Central cord syndrome (syringomyelia)
4. Posterior column syndrome (tabes dorsalis)
5. Posterolateral cord syndrome (SACDC)
6. Combined AHC—pyramidal tract syndrome (ALS)
7. AHC syndrome
8. Anterior spinal artery occlusion.

Complete Cord Transection

Causes	Features
<ul style="list-style-type: none"> • Trauma • Metastatic carcinoma • Multiple sclerosis • Spinal epidural hematoma • Autoimmune disorders • Postvaccinal syndromes 	<ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – All sensations are affected – Sensory level is usually 2 segments below the level of lesion – Segmental paresthesia occurs at the level of lesion • Motor: <ul style="list-style-type: none"> – Paraplegia due to corticospinal tract involvement – First spinal shock followed by hypertonic hyperreflexia paraplegia – Loss of abdominal and cremasteric reflexes – At the level of lesion LMN signs occur • Autonomic: <ul style="list-style-type: none"> – Urinary retention and constipation – Anhidrosis, trophic skin changes, vasomotor instability below the level of lesion – Sexual dysfunction can occur

Brown-Sequard Syndrome

Due to damage to one lateral half of spinal cord.

Causes	Features
<ul style="list-style-type: none"> • Caused by extramedullary lesions • Usually caused by penetrating injuries (gunshot) or tumor 	<ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – Ipsilateral loss of proprioception due to posterior column involvement – Contralateral loss of pain and temperature due to involvement of lateral spinothalamic tract 1 or 2 segments below • Motor: <ul style="list-style-type: none"> – Ipsilateral spastic weakness due to descending corticospinal tract involvement – Lower motor neuron signs at the level of lesion

Central Cord Syndrome

Causes	Features
<ul style="list-style-type: none"> • Most common cause is syringomyelia • Other causes are hyperextension, injuries of neck, intramedullary tumors and trauma • Associated with Arnold Chiari type 1 and 2 and Dandy Walker malformation 	<ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – Pain and temperature are affected – Touch and proprioception are preserved – Dissociative anesthesia – Shawl like distribution of sensory loss • Motor: <ul style="list-style-type: none"> – Upper limb weakness > Lower limb weakness • Other features include: <ul style="list-style-type: none"> – Horner's syndrome – Kyphoscoliosis – Sacral sparing – Neuropathic arthropathy of shoulder and elbow joint <p>Early bladder involvement (exception—syringomyelia)</p>

Posterior Column Syndrome

Cause	Features
Occurs due to neurosyphilis, diabetes mellitus	<ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – Impaired position and vibration sense in lower limb – Sensory ataxia – Positive Romberg's sign, sink sign and Lhermitte's sign • Abadie's sign positive • Urinary incontinence • Absent knee and ankle jerk (areflexia and hypotonia) • Charcot's joint • Miotic and irregular pupil not reacting to light—Argyll Robertson pupil

Posterolateral Column Disease

Causes	Features
<ul style="list-style-type: none"> • Vitamin B₁₂ deficiency • AIDS • HTLV associated myelopathy • Cervical spondylosis 	<ul style="list-style-type: none"> • Sensory: <ul style="list-style-type: none"> – Paresthesia in feet – Loss of proprioception and vibration in legs – Sensory ataxia – Positive Romberg's sign • Bladder atonia • Motor: <ul style="list-style-type: none"> – Corticospinal tract involvement—spasticity, hyperreflexia, bilateral Babinski sign • AIDS-associated dementia and spastic bladder is present • HTLV associated myelopathy—slowly progressive paraparesis and an increase in CSF IgG antibodies to HTLV1

(AIDS: acquired immunodeficiency syndrome; HTLV: human T-cell lymphotropic virus; CSF: cerebrospinal fluid; IgG: immunoglobulin G)

Anterior Horn Cell Syndromes

Cause	Features
Spinal muscular atrophy (SMA)	<ul style="list-style-type: none"> • Motor: <ul style="list-style-type: none"> – Weakness, atrophy, and fasciculations – Hypotonia with depressed reflexes – Muscles of trunk and extremities are affected • Sensory system is not affected

Anterior Spinal Artery Syndrome

Cause	Features
Occurs due to syphilitic arteritis, aortic dissection, atherosclerosis of aorta, SLE, AIDS, and AV malformation	<ul style="list-style-type: none"> • Motor: <ul style="list-style-type: none"> – Flaccid and areflexic paraplegia • Sensory: <ul style="list-style-type: none"> – Loss of pain and temperature – Preservation of position and vibration • Autonomic: <ul style="list-style-type: none"> – Urinary incontinence – Spinal cord infarction usually occurs in T1 to T4 and L1 segment • Abrupt onset, radicular, or girdle pain

Postspinal Artery Syndrome

Cause	Features

Rare	<ul style="list-style-type: none"> Loss of proprioception and vibratory sense Pain and temperature is preserved Absence of motor deficit
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Anterior Horn Cell and Pyramidal Tract

Cause	Features
ALS—amyotrophic lateral sclerosis	<ul style="list-style-type: none"> LMN signs UMN signs Sensations preserved Onuf's nucleus spared—hence no bladder and bowel involvement

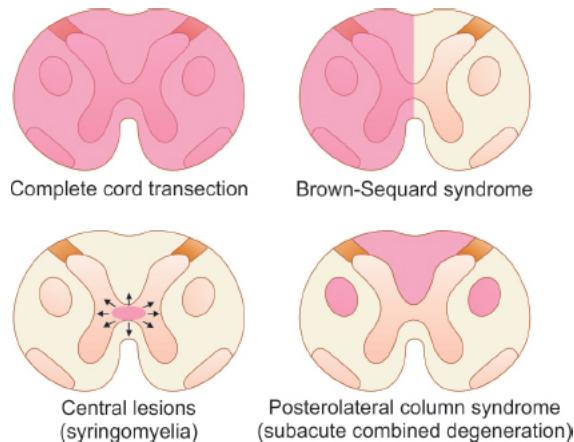


Fig. 6E.8: Spinal cord syndromes 1.

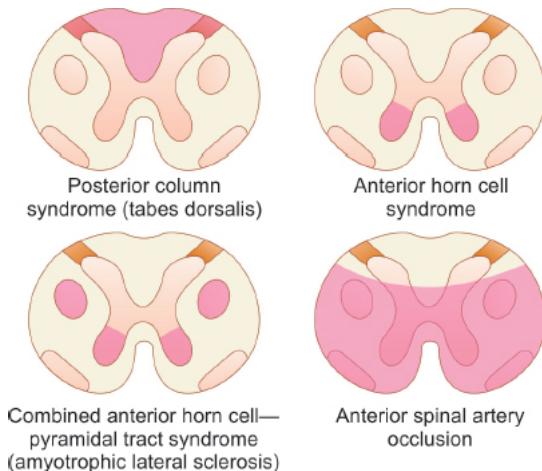


Fig. 6E.9: Spinal cord syndromes 2.

Difference Between Paraplegia in Flexion and Paraplegia in Extension

Features	Paraplegia in extension	Paraplegia in flexion
Definition	Lower limb takes an extension attitude and extensor muscles are spastic	Lower limb muscles take an attitude of flexion
Pathology	Only pyramidal tract involved	Both pyramidal and extrapyramidal tract involved (reticulospinal tracts). Occurs in late stage of paraplegia

Evolution	Early	Late
Tone	Clasp knife spasticity in extensor group	Tone is increased in flexor groups
Deep tendon reflex (DTR)	<ul style="list-style-type: none"> • Deep tendon reflexes are exaggerated • Clonus may be present 	<ul style="list-style-type: none"> • DTR's are present but diminished • No clonus
Plantar reflex	Extensor plantar response	Extensor plantar associated with flexor spasm
Mass reflex**	Absent	Present

Note: ****Mass reflex:** Any stimulation (scratching of skin) below the level of lesion produces an interoceptive response resulting in flexor spasms, spontaneous emptying of bowel and bladder, profuse sweating and piloerection and seminal emission.

Cord Involvement at Multiple Sites

- Arachnoiditis (in tubercular, there is patchy involvement)
- Neurofibromatosis
- Multiple sclerosis
- Secondary deposits
- Cervical spondylitis.

Causes of Spastic Paraplegia (UMN Type Lesion)

A. Gradual onset

- Cerebral causes—parasagittal meningioma, hydrocephalus, etc.
- Spinal causes:
 - Compressive or transverse lesion in the spinal cord
 - Noncompressive or longitudinal lesion or systemic disease of the spinal cord.
- Motor neuron disease (MND), e.g. amyotrophic lateral sclerosis
- Multiple sclerosis, Devic's disease
- Friedreich's ataxia
- Subacute combined degeneration (i.e. from vitamin B₁₂ deficiency)
- Lathyrism
- Syringomyelia
- Hereditary spastic paraplegia
- Erb's spastic paraplegia
- Tropical spastic paraplegia
- Radiation myelopathy.

B. Sudden onset

- Cerebral causes—thrombosis of unpaired anterior cerebral artery, superior sagittal sinus thrombosis
- Spinal causes:

Compressive causes:

- Injury to the spinal cord (fracture-dislocation or collapse of the vertebra)
- Prolapsed intervertebral disc
- Spinal epidural abscess or hematoma.

Noncompressive causes:

- Acute transverse myelitis
- Thrombosis of anterior spinal artery
- Hematomyelia (from arteriovenous malformation, angiomas, or endarteritis)
- Radiation myelopathy electrical injury.

Causes of Flaccid Paraplegia (LMN Type)

- UMN lesion in shock stage, transverse myelitis, spinal injury
- Lesion involving anterior horn cells:
 - Acute anterior poliomyelitis
 - Progressive muscular atrophy (variety of MND).
- Diseases affecting nerve root—tabes dorsalis, radiculitis, Guillain-Barré (GB) syndrome
- Diseases affecting peripheral nerves:
 - Acute infective polyneuropathy (GB syndrome)
 - High cauda equina syndrome
 - Disease of peripheral nerves involving both the lower limbs
 - Lumbar plexus injury (psoas abscess or hematoma).
- Diseases affecting myoneural junction:
 - Myasthenia gravis, Lambert-Eaton syndrome
 - Periodic paralysis due to hypo- or hyperkalemia.
- Diseases affecting muscles—myopathy.

Causes of Quadriplegia

Weakness of all the 4 limbs can occur in the lesions from cortex to C5 level of spinal cord and various LMN lesion affecting anterior horn cells, roots, peripheral nerve, NM junction, and muscles.

Upper motor neuron causes	Lower motor neuron causes
<ul style="list-style-type: none"> • Cerebral palsy • Bilateral brainstem lesion (glioma) • Craniovertebral anomaly • High cervical cord compression • Multiple sclerosis • Motor neuron disease 	<ul style="list-style-type: none"> • Acute anterior poliomyelitis • Guillain-Barré syndrome • Peripheral neuropathy • Myopathy or polymyositis • Myasthenia gravis and crisis • Periodic paralysis • Snake bite, organophosphate poisoning, etc.

SPECIFIC LOCALIZING SIGNS AT VARIOUS LEVELS

Features of Cervical Signs at Cord Lesion

In general, cervical cord disorders are best localized by the pattern of weakness that ensues, whereas sensory deficits have less localizing value.

- High cervical cord lesions (lesions above C5) are frequently life threatening, produce quadriplegia and weakness of diaphragm, the main respiratory muscle innervated by the phrenic nerve (C3-C5).
- Extensive lesions near the junction of the cervical cord and medulla are usually fatal owing to involvement of adjacent medullary centers, which results in vasomotor and respiratory collapse.
- Compressive lesions near the foramen magnum may produce weakness of the ipsilateral shoulder and arm followed by weakness of the ipsilateral leg, then the contralateral leg, and finally the contralateral arm (cartwheel pattern or Ellsberg phenomenon).
- Lesions at C4-C5 produce quadriplegia with preserved respiratory function.
- At the midcervical (C5-C6) level, there is relative sparing of shoulder muscles and loss of biceps and brachioradialis reflexes.
- Lesions at C7 spare the biceps but produce weakness of finger and wrist extensors and loss of the triceps reflex.
- Lesions at C8 paralyze finger and wrist flexion, and the finger flexor reflex is lost.
- Horner's syndrome (miosis, ptosis, and facial hypohidrosis) may also occur ipsilateral to cervical lesions at any level.

Features of Thoracic Cord Lesion

Lesions of the thoracic cord are best localized by identification of a sensory level on the trunk.

- Useful markers in terms of sensory dermatomes are at the nipples (T4), xiphisternum (T6), subcostal margins (T8), umbilicus (T10), and pubic symphysis (T12)
- The abdominal wall musculature, supplied by the lower thoracic nerves is observed during movements of respiration or coughing or by asking the patient to interlock the fingers behind the head in the supine position and attempt to sit up.
- Lesions at T9-T10 paralyze the lower, but spare the upper, abdominal muscles, resulting in upward movement of the umbilicus when the abdominal wall contracts (Beevor's sign) and in loss of lower, but not upper, superficial abdominal reflexes.
- With unilateral lesions, attempts to contract the abdominal wall produce movement of the umbilicus to the normal side; superficial abdominal reflexes are absent on the involved side.
- Midline back pain is a useful localizing sign in the thoracic region.

Feature of Lumbar Cord

Effect of various root lesions in lumbar region:

Roots	Motor deficit (most rapidly demonstrated)
L2	Hip flexion and thigh adduction
L3	Knee extension and thigh adduction
L4	Inversion of foot
L5	Dorsiflexion to toes and foot
S1	Plantar flexion and eversion of foot

- Lesions at L2-L4 paralyze flexion and abduction of the thigh, weaken leg extension at the knee, and abolish the patellar reflex.
- Lesions at L5-S1 paralyze movements of the foot and ankle, flexion at the knee, and extension of the thigh, and abolish the ankle jerk (S1).
- A cutaneous reflex useful in localization of lumbar cord disease is the cremasteric reflex, which is segmentally innervated at L1-L2.

Features of Sacral Cord/Conus Medullaris

The conus medullaris is the tapered caudal termination of the spinal cord, comprising the lower sacral and single coccygeal segments. Isolated lesions of the conus medullaris spare motor and reflex functions in the legs.

The Conus Syndrome (Fig. 6E.10)

- Bilateral saddle anesthesia (S3-S5), prominent bladder and bowel dysfunction (urinary retention and incontinence with lax anal tone), and impotence
- The bulbocavernous (S2-S4) and anal (S4-S5) reflexes are absent
- Muscle strength is largely preserved.

Cauda Equina Syndrome—Asymmetric, Atrophic, and Areflexic Paralysis of Lower Limbs (Fig. 6E.10)

- The cluster of nerves derived from the lower cord as they descend to their exits in the intervertebral foramina (L2-3 to coccygeal nerve roots).
- Cauda equina lesions are characterized by severe low back or radicular pain, asymmetric leg weakness or sensory loss, variable areflexia in the lower extremities, and relative sparing of bowel and bladder function.
- Mass lesions in the lower spinal canal may produce mixed clinical picture in which elements of both cauda equina and conus medullaris syndromes coexist.

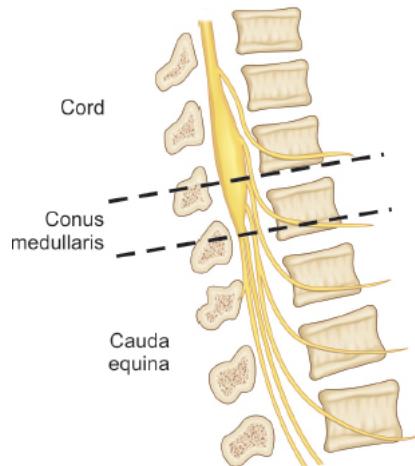


Fig. 6E.10: Conus-cauda equina syndrome.

	Conus medullaris syndrome (S2-4)	Cauda equina syndrome (L3 root and below)
Presentation	Sudden and bilateral	Gradual and unilateral
Reflexes	Knee jerk is preserved but ankle jerk is affected	Both knee and ankle jerks are affected
Radicular pain	Less severe	More severe
Low back pain	More	Less
Sensory symptoms and signs	Numbness is symmetrical and bilateral, sensory dissociation occurs, saddle anesthesia present	Numbness is asymmetrical, may be unilateral, no necessary dissociation
Motor strength	Typically symmetric hyperreflexia, distal paresis of lower limbs	Asymmetric areflexic paraplegia
Impotence	Frequent	Less frequent
Sphincter dysfunction	Overflow urinary incontinence and fecal incontinence, tend to present early in course of disease	Urinary retention tends to present late in course of disease
Trophic changes	Common	Less marked

Epiconus: Lesion of lumbar cord at the level of L4-S2 characterized by a flaccid paralysis of legs (only the roots are affected causing peripheral paralysis, i.e. distal paraparesis). Reflex but not conscious evacuation of the bladder is present, and rectum is preserved. Sexual potency is lost.

What are the Different Types of Spinal Pain?

- Radicular pain is characterized as a unilateral, lancinating, dermatomal pain often exacerbated by cough, sneeze, or Valsalva's maneuver. Radicular pain is common with extradural growths and rare with intramedullary lesions. An example of an extramedullary tumor causing radicular pain is the neurilemmoma (usually an intradural extramedullary lesion).
- Vertebral pain is characterized by an aching pain localized to the point of the spine involved in the compressive process and often accompanied by point tenderness. Spinal pain is common with neoplastic or inflammatory extradural lesions and infrequent with intramedullary or intradural extramedullary lesions.
- Funicular (central) pain is common with intramedullary lesions and very unusual with extradural lesions. It is described as deep, ill-defined painful dysesthesias, usually distant from the affected spinal cord level (and therefore of poor localizing value), probably related to dysfunction of the spinothalamic tract or posterior columns.

- With dysfunction of the posterior columns in the cervical region, neck flexion may elicit a sudden “electric-like” sensation down the back or into the arms (Lhermitte’s sign or “barber’s chair syndrome”).

APPROACH TO PERIPHERAL NEUROPATHY

Various nerve fibers and their functions are depicted in **Figure 6E.11**.

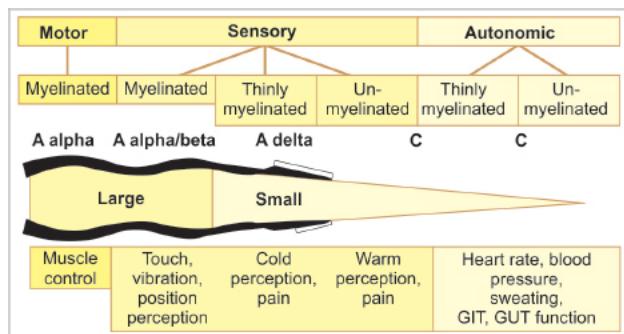


Fig. 6E.11: Various nerve fibers and their function.

Clinical Types of Neuropathy

- 1. Polyneuropathy:** It is the most common variety of neuropathy. The nerve fibers are affected in a length-dependent pattern; toes and soles are affected first and hands later. A majority of these cases occur due to metabolic, toxic, or systemic disorders.

Causes of polyneuropathy

- Diabetes mellitus
- Alcohol
- Nutritional (B_{12} deficiency)
- Guillain-Barré syndrome
- Toxins (Pb, As, Zn, and Hg)
- Hematologic (paraproteins)
- Endocrine (hypothyroid)
- Rheumatologic (systemic lupus erythematosus, rheumatoid arthritis, and vasculitis)
- Amyloid
- Porphyria
- Infectious (syphilis, human immunodeficiency syndrome)
- Sarcoid
- Tumor (paraneoplastic)

“DANG THERAPIST”

- 2. Mononeuropathy:** Mononeuropathy refers to single peripheral nerve involvement and usually occurs due to trauma, compression, or entrapment.

Causes of mononeuropathy

- **Acute:** Sustained pressure, e.g. tourniquet
- **Chronic:** Entrapment.

Causes (according to site of compression)

• Carpal tunnel	Median nerve
• Cubital tunnel	Ulnar nerve
• Spiral groove of humerus	Radial nerve
• Inguinal ligament	Lateral cutaneous of thigh (meralgia paresthetica)
• Neck of fibula	Common peroneal nerve
• Flexor retinaculum (Tarsal tunnel)	Posterior tibial nerve

- Entrapment neuropathies are commonly seen in**
- Endocrin (diabetes mellitus, myxedema, acromegaly)
 - Amyloidosis
 - Hereditary neuropathy susceptible to pressure palsy
 - Pregnancy
 - Arthritis (rheumatoid)

3. Multiple mononeuropathies/mononeuritis multiplex refers to the involvement of multiple, separate noncontiguous peripheral nerves either simultaneously or sequentially.

Causes of mononeuritis multiplex
<ul style="list-style-type: none"> • Leprosy (most common) • Diabetes mellitus • Vasculitis • Sarcoidosis • Amyloidosis • Malignancy • Neurofibromatosis • HIV infection • Idiopathic multifocal motor neuropathy

PATHOLOGIC CLASSIFICATION OF NEUROPATHIC DISORDERS (FIGS. 6E.12A AND B)

1. Neuronopathies (pure sensory or pure motor):

- Sensory neuronopathies (ganglionopathies)
- Motor neuronopathies (motor neuron disease)

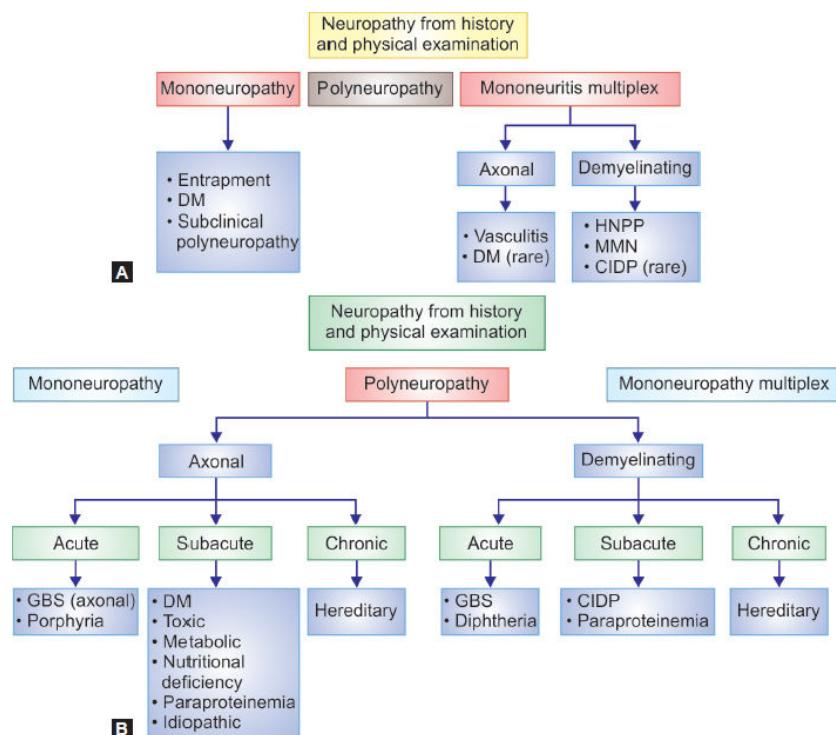
Sensory neuronopathy	Motor neuronopathy
<ul style="list-style-type: none"> • Ganglion cells predominantly affected • Both proximal and distal involvement • Sensory ataxia is common • No weakness • But awkward movement due to sensory disturbances <p>Example:</p> <ul style="list-style-type: none"> • Cancer (paraneoplastic) • Sjogren's syndrome • Cisplatin and other analogs • Vitamin B₆ toxicity • HIV-related sensory neuronopathy 	Disorder of anterior horn cells. Weakness, fasciculation, atrophy not truly a process of peripheral nerves

2. Peripheral neuropathies (usually sensorimotor):

- Myelinopathies
- Axonopathies

Axonal neuropathy	Demyelinating neuropathy
Usually gradual and insidious onset	Usually acute or subacute
Large and long axons are affected early, hence initially lower extremities are affected	Diffuse process, starts in lower limbs. But not always distal
Stocking-glove sensory motor loss results in symmetrical distal clinical signs in legs and arms	Generalized weakness and mild sensory loss
Distal involvement	Proximal and distal involvement

Ankle jerk lost early and proximal tendon reflexes preserved	All reflexes are lost early
Muscle wasting common	Relatively absent
Cerebrospinal fluid (CSF) proteins normal	CSF proteins elevated (since nerve roots are involved)
Slow recovery	Rapid recovery
Residual deformity common	Residual deformity less common
Nerve conduction normal or slightly lowered	Nerve conduction is slowed



Figs. 6E.12A and B: Classification of neuropathy based on history and examination.

(DM: diabetes mellitus; HNPP: hereditary neuropathy with liability to pressure palsies; CIDP: chronic inflammatory demyelinating polyneuropathy; MMN: multifocal motor neuropathy; GBS: Guillain–Barré syndrome)

APPROACH TO POLYNEUROPATHY

What is the onset and temporal evolution?	
	<ul style="list-style-type: none"> • Acute (days to 4 weeks) • Subacute (4–8 weeks) • Chronic (>8 weeks)
Acute onset	<ul style="list-style-type: none"> • Guillain–Barré syndrome • Acute intermittent porphyria • Critical illness polyneuropathy • Thallium toxicity
Subacute onset	<ul style="list-style-type: none"> • Toxins or medications • Nutritional deficiency • Metabolic abnormality

	<ul style="list-style-type: none"> • Paraneoplastic syndrome
Chronic	<ul style="list-style-type: none"> • Hereditary motor and sensory neuropathy (HMSN) • CIDP • CKD
Relapsing/remitting course	<ul style="list-style-type: none"> • Guillain-Barré syndrome • CIDP • HIV/AIDS • Porphyria

(CIDP: chronic inflammatory demyelinating polyneuropathy; CKD: chronic kidney disease; HIV: human immunodeficiency virus; AIDS: acquired immunodeficiency syndrome)

What systems are involved? Motor (or) sensory (or) autonomic (or) mixed	
Motor symptoms	
Negative symptoms	Positive symptoms
<ul style="list-style-type: none"> • Weakness • Wasting • Loss of dexterity <p>In the early stage, weakness in peripheral neuropathy is distal; however, early proximal weakness is a feature of demyelinating neuropathy and porphyric neuropathy</p>	<ul style="list-style-type: none"> • Cramps • Tremors • Fasciculations • Spasms
Neuropathic disorders that may have only motor symptoms at presentation	
<ul style="list-style-type: none"> • Motor neuron disease • Lead intoxication • Acute porphyria • Guillain-Barre Syndrome • Hereditary motor neuropathy • CIDP • Diphtheria • Brachial neuritis • Diabetic lumbosacral plexus neuropathy 	
Sensory symptoms	
Negative symptoms	Positive symptoms
Numbness, loss of sensation in hands and feet	Burning, pain, walking on cotton wool, band-like sensation on feet or trunk, stumbling, tingling, pins, and needles
Large fiber neuropathy—neuropathy of signs/ataxic neuropathy There are few symptoms (numbness, ataxia) but lots of signs (loss of vibration, joint position sense, diminished reflexes, Romberg's sign positive)	Small fiber neuropathy—neuropathy of symptoms Lots of symptoms (PAIN—burning, shock like, stabbing, prickling, shooting, lancinating, allodynia, tight band like pressure. Insensitive to heat and cold) but very few signs (loss of pain, temperature)
Examples: <ul style="list-style-type: none"> • Sjogren's syndrome • Vitamin B₁₂ neuropathy • Cisplatin • Pyridoxine neurotoxicity • Friedreich's ataxia 	Examples: <ul style="list-style-type: none"> • Diabetes • Amyloidosis • Fabry's disease • HIV • Tangier's disease • Hereditary sensory and autonomic neuropathy • Sjogren's syndrome • Chronic idiopathic small fiber sensory neuropathy
Small and large fiber neuropathy—pan sensory: Global sensory loss	
Example:	
<ul style="list-style-type: none"> • Carcinomatous sensory neuropathy 	

- Hereditary sensory neuropathy
- Diabetic sensory neuropathy
- Vacor intoxication
- Xanthomatous neuropathy of primary biliary cirrhosis

Peripheral neuropathies that are often associated with pain

- Cryptogenic sensory or sensorimotor neuropathy
- Diabetes mellitus
- Vasculitis
- Guillain–Barré syndrome
- Amyloidosis
- Toxic (arsenic and thallium)
- HIV related distal symmetrical polyneuropathy
- Fabry's disease

Autonomic symptoms

Enquire if the patient has fainting spells or orthostatic lightheadedness, sweating abnormalities or any bowel, bladder, or sexual dysfunction.

Examples:

Acute:

- Pandysautonomia
- Botulism
- Porphyria
- Guillain-Barré syndrome
- Amiodarone
- Vincristine

Chronic:

- Amyloid
- Diabetes
- Sjogren's
- HSN 1 and 3
- Chagas disease
- Paraneoplastic

PATTERNS OF NEUROPATHY

Pattern 1

Symmetric Proximal and Distal Weakness with Sensory Loss

Inflammatory demyelinating polyneuropathy (GBS and CIDP).

Pattern 2

Symmetric Distal Weakness with Sensory Loss

Metabolic disorders, hereditary toxins drugs.

Pattern 3

Asymmetric Distal Weakness with Sensory Loss

- Multiple nerves—vasculitis
- Single nerves/regions—compressive mononeuropathy and radiculopathy.

Pattern 4

Asymmetric Distal Weakness without Sensory Loss

- Motor neuron disease—with upper motor neuron findings
- Multifocal motor neuropathy—without upper motor neuron findings.

Pattern 5

Asymmetric Proximal and Distal Weakness with Sensory Loss

- Polyradiculopathy or plexopathy due to diabetes mellitus
- Meningeal carcinomatosis.

Pattern 6

Symmetric Sensory Loss without Weakness

Cryptogenic sensory polyneuropathy (CSPN), metabolic (diabetes and others) drugs, and toxins.

Pattern 7

Symmetric Sensory Loss and Distal Areflexia with Upper Motor Neuron Findings

B₁₂ deficiency, HIV, and hepatic disease.

Pattern 8

A Symmetric Proprioceptive Sensory Loss without Weakness

Sensory neuronopathy (ganglionopathy).

Pattern 9

Autonomic Symptoms and Signs

Neuropathies associated with autonomic dysfunction.

Pattern 10

Syndrome of Acute Ascending Motor Paralysis

- Guillain-Barré syndrome/acute idiopathic polyneuritis
- Diphtheria
- Porphyria
- Triorthocresyl phosphate (TOCP) poisoning
- Paraneoplastic
- Postvaccinal.

Pattern 11

Syndrome of Subacute Sensory Motor Neuropathy

- Deficiency—alcoholic beriberi, pellagra, and vitamin B₁₂
- Toxins = arsenic, lead, Hg, and Pb
- Drugs = nitrofurantoin, INH, dapsone, disulfiram, and clioquinol
- Uremic
- DM, PAN and sarcoidosis.

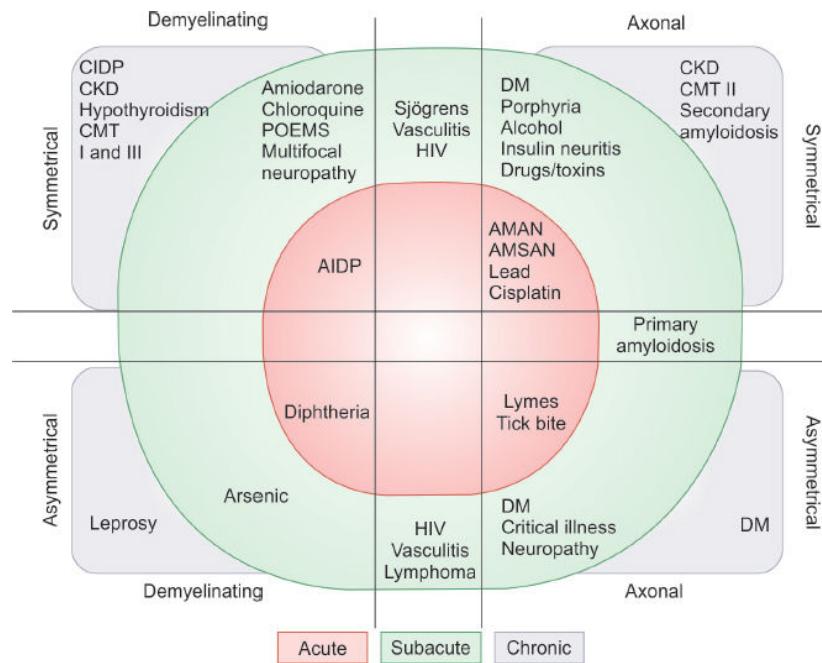


Fig. 6E.13: Simplified diagram showing types of polyneuropathy.

(CIDP: chronic inflammatory demyelinating polyneuropathy; CKD: chronic kidney disease; CMT: Charcot-Marie-Tooth; POEMS: polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes; DM: diabetes mellitus; HIV: human immunodeficiency virus; AIDP: acute inflammatory demyelinating polyneuropathy; AMAN: acute motor axonal neuropathy; AMSAN: acute motor and sensory axonal neuropathy)

General examination in neuropathy	
Purpura, livedo reticularis	Vasculitis
Skin hypopigmentation	Leprosy
Hyperpigmentation	Osteosclerotic myeloma—POEMS
Bullous lesions	Variegate porphyria
Purpura	Vasculitis, cryoglobulinemia
Ichthyosis	Refsum's disease
Mee's lines	Arsenic/thallium intoxication
Alopecia	Thallium poisoning
Curled hair	Giant axonal neuropathy
Nerve thickening	<ul style="list-style-type: none"> • Leprosy • CMT • CIDP • Amyloidosis • Neurofibromatosis • Refsum's disease • Dejerine-Sottas disease • Roussy Levy syndrome • Acromegaly • Idiopathic

(POEMS: polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes; CMT: Charcot-Marie-Tooth; CIDP: chronic inflammatory demyelinating polyneuropathy)

CRANIAL NERVE EXAMINATION IN NEUROPATHY

- Anosmia—Refsum's disease and B₁₂ deficiency
- Optic atrophy—demyelinating disease may suggest an inherited syndrome, B₁₂ deficiency
- Anisocoria and impaired pupillary light reflexes—parasympathetic damage and may be isolated, as in Adie's syndrome, diabetic neuropathy or acute dysautonomia as in GBS
- Impaired ocular mobility suggests botulism or Miller Fisher syndrome
- Facial weakness—GBS, CIDP, Lymes disease, and leprosy
- Trigeminal sensory loss—Sjogren neuropathy
- Lower cranial nerve palsies—Kennedy's disease.

Medications causing neuropathies	
<ul style="list-style-type: none"> • Axonal <ul style="list-style-type: none"> – Vincristine – Paclitaxel – Nitrous oxide – Colchicine – Isoniazid – Hydralazine – Metronidazole – Pyridoxine – Didanosine – Lithium – Dapsone – Phenytoin – Cimetidine – Disulfiram – Chloroquine – Ethambutol – Amitriptyline 	<ul style="list-style-type: none"> • Demyelinating <ul style="list-style-type: none"> – Amiodarone – Chloroquine – Suramin – Gold
	<ul style="list-style-type: none"> • Neuronopathy <ul style="list-style-type: none"> – Thalidomide – Cisplatin – Pyridoxine

COMMON NEUROPATHIES

Guillain-Barré Syndrome (Tables 6E.6 and 6E.7)

Table 6E.6: Diagnostic criteria of GBS.
Required features <ul style="list-style-type: none"> • Progressive weakness in both arms and legs • Areflexia (or hyporeflexia)
Features supportive of diagnosis <ul style="list-style-type: none"> • Progression of symptoms over days to 4 weeks • Relative symmetry • Mild sensory signs or symptoms • Cranial nerve involvement, especially bilateral facial weakness • Recovery beginning 2–4 weeks after progression ceases • Autonomic dysfunction • Absence of fever at onset • Typical CSF (albuminocytologic dissociation) • EMG/nerve conduction studies (characteristic signs of a demyelinating process in the peripheral nerves)
Features casting doubt on the diagnosis

- Asymmetrical weakness
- Persistent bladder and bowel dysfunction
- Bladder or bowel dysfunction at onset
- >50 mononuclear leukocytes/mm³ or presence of polymorphonuclear leukocytes in CSF
- Distinct sensory level

Features that rule out the diagnosis

- Hexacarbon abuse
- Abnormal porphyrin metabolism
- Recent diphtheria infection
- Lead intoxication
- Other similar conditions: Poliomyelitis, botulism, hysterical paralysis, toxic neuropathy

(CSF: cerebrospinal fluid; EMG: electromyogram)

Table 6E.7: Variants of GB syndrome.

Common variants	Less common variants
<ul style="list-style-type: none"> • Acute motor and sensory axonal neuropathy (AMSAN) • Acute motor axonal neuropathy (AMAN) • Miller-Fisher variant • Pure motor variants • Pure sensory variants • Pure dysautonomia variant • Pharyngeal-cervical-brachial variant • Paraparetic variant (Ropper variant) 	<ul style="list-style-type: none"> • Acral paresthesias with diminished reflexes in either arms or legs • Facial diplegia or abducens palsies with distal paresthesias • Isolated postinfectious ophthalmoplegia • Bilateral foot drop with upper limb paresthesias • Acute ataxia without ophthalmoplegia • Bickerstaff's brainstem encephalitis (BBE)

Diabetes Mellitus (Box 6E.1)

Box 6E.1: Classification of diabetic neuropathy.

Polyneuropathy

- Symmetrical, mainly sensory and distal
- Asymmetrical, mainly motor and proximal (including amyotrophy)

Mononeuropathy and mononeuritis multiplex

- Cranial nerve lesions
- Isolated peripheral nerve lesions

Autonomic (visceral) neuropathy

- Cardiovascular
- Gastrointestinal
- Genitourinary
- Sudomotor
- Vasomotor
- Pupillary

Polyradiculopathies

- Diabetic amyotrophy (lumbar polyradiculopathy)
- Thoracic polyradiculopathy
- Diabetic neuropathic cachexia

Treatment-induced neuropathy of diabetes

Neuropathies with HIV Infection

- **Seroconversion**
 - Guillain-Barre syndrome
 - Chronic inflammatory demyelinating polyneuropathy (CIDP).
- **Symptomatic stage:** Mononeuritis multiplex axonal type subacute or chronic
- **Late symptomatic stage:** Distal symmetrical sensory polyneuropathy, most common neuropathy frequently coexists with symptomatic encephalopathy and myopathy
 - Toxic polyneuropathy (drugs)
 - Subacute asymmetrical polyneuropathy of cauda equina, caused by cytomegalovirus.

HEREDITARY NEUROPATHIES

Neuropathy is the sole or primary part of the disease	Neuropathy is part of a more generalized neurological or multisystem disorder
<ul style="list-style-type: none">• <i>Charcot-Marie-tooth disease</i>—CMT1 (demyelinating) and CMT2 (axonal)• HMSN-III (or <i>Dejerine–Sottas neuropathy</i>)• Hereditary sensory and autonomic neuropathy (HSAN)	<ul style="list-style-type: none">• Spinocerebellar atrophy (SCA)—Friedreich ataxia (FA)• Hereditary spastic paraparesis neuropathy (i.e. complicated HSP, HMSN 5)• Familial amyloid (transthyretin, gelsolin, ApoA1)
<ul style="list-style-type: none">• Distal hereditary motor neuropathy (dHMN)• Hereditary brachial plexus neuropathy (HBPN)• Hereditary neuropathy with liability to pressure palsies (HNPP)	<ul style="list-style-type: none">• Leukodystrophy• Lipoprotein deficiency• Porphyrias

APPROACH TO A PATIENT WITH PARKINSON'S DISEASE

Idiopathic Parkinson's Disease (Paralysis Agitans)

It is a chronic, progressive disorder in which idiopathic parkinsonism occurs without evidence of more widespread neurologic involvement.

Clinical Manifestations

Motor symptoms: Always asymmetrical in onset and become bilateral within a year (Table 6E.8).

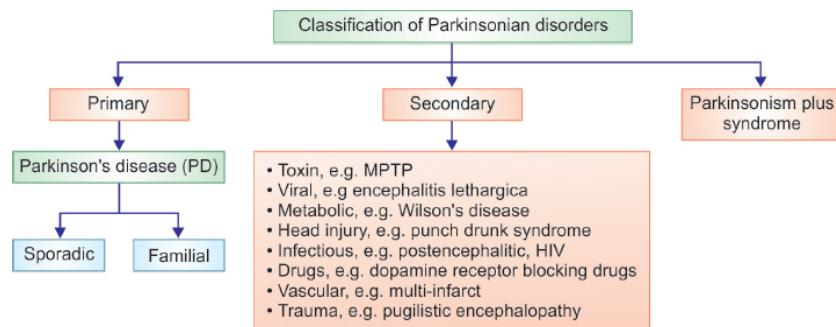
- **Tremor** is an early and presenting symptom in 70% of patients.
 - Frequency is 4–6 Hz tremor and is typically most prominent at rest and worsens with emotional stress.
 - Typically tremor starts with the fingers and hands at rest.
 - Often described as pill rolling of finger and wrist, because the patient appears to be rolling something between thumb and forefinger.
 - Disappears on voluntary movement and sleep.
- **Rigidity:**
 - Stiffness on passive limb movement is described as “lead pipe” rigidity because the increase in muscle tone is present throughout the range of movement. Unlike spasticity, it is not dependent on speed of movement.
 - When tremor is superimposed on the rigidity, a ratchet like jerkiness is felt, described as “cogwheel” rigidity.
- **Akinesia or bradykinesia**
 - Poverty/slowing of movement is the hallmark of parkinson's disease (PD). Slowness/difficulty of initiating voluntary movement and an associated reduction in automatic movements, such as swinging of the arms when walking.

- There is fixity of facial expression (facial immobility—mask like face) with widened palpebral fissures and infrequent blinking.
- Repetitive tapping (at about 2 Hz) over the glabella (glabellar tap) produces a sustained blink response (Myerson's sign), in contrast to the response of normal subject.
- **Postural changes:** A stooped posture is a characteristic feature.
- **Gait changes:** Slow shuffling, freezing and reduced arm swing, small stride length, slow turns, festinating gait (tendency to advance rapid short steps) and catching center of gravity. Feet may be glued to floor. Postural instability and freezing may result in fall forward.
- **Reduced eye blink.**

Table 6E.8: Nonmotor symptoms of Parkinson's disease.

Autonomic dysfunction	Neuropsychiatric	Sensory problems
<ul style="list-style-type: none"> • Orthostatic hypotension • Urinary incontinence • Constipation • Sexual problems 	<ul style="list-style-type: none"> • Anxiety • Depression • Apathy • Psychosis • Dementia 	<ul style="list-style-type: none"> • Reduced sense of smell (hyposmia) • Pain
Sleep disorders	Rheumatological	Other
<ul style="list-style-type: none"> • Restless legs • Insomnia • Daytime somnolence 	<ul style="list-style-type: none"> • Frozen shoulder • Periarthritis • Swan neck deformity 	<ul style="list-style-type: none"> • Seborrhea

Flowchart 6E.4: Classification of Parkinsonian disorder.



(MPTP: manganese, 1-methyl 4-phenyl tetrahydropyridine; HIV: human immunodeficiency virus)

Table 6E.9: Hoehn and Yahr stage of Parkinson's disease.

Stage	Disease state
I	Unilateral involvement only, minimal or no functional impairment
II	Bilateral or midline involvement, without impairment of balance
III	First sign of impaired righting reflex, mild to moderate disability
IV	Fully developed, severely disabling disease; patient still able to walk and stand unassisted
V	Confinement to bed or wheelchair unless aided

Table 6E.10: Causes of secondary Parkinsonism.

Toxin: Manganese, 1-methyl 4-phenyl -1,2,3,6-tetrahydropyridine (MPTP), carbon monoxide, manganese, mercury, carbon disulfide, cyanide, methanol	Drugs: Dopamine receptor blocking drugs, reserpine, tetrabenazine, alpha methyl dopa, lithium, flunarizine, cinnarizine
Viral: Encephalitis lethargica, Creutzfeldt-Jakob disease	Vascular: Multi-infarct, Binswangers disease
Metabolic: Wilson's disease	Trauma: Pugilistic encephalopathy

Head injury: Punch drunk syndrome

Infectious: Postencephalitic, human immunodeficiency virus (HIV), subacute sclerosing panencephalitis (SSPE), Prion diseases

Others: Parathyroid abnormalities, hypothyroidism, brain tumors, paraneoplastic, normal pressure hydrocephalus (NPH), psychogenic

Table 6E.11: Parkinson plus syndromes and its features.

Syndrome	Features
Progressive supranuclear palsy (PSP, Steele-Richardson-Olszewski syndrome)	Slow ocular saccades, eyelid apraxia, and restricted eye movements with particular impairment of downward gaze and reptilian stare [Fig. 6D(iii).37]. Frequently experience hyperextension of the neck with early gait disturbance and falls. MRI may reveal a characteristic atrophy of the midbrain with relative preservation of the pons (the 'hummingbird sign' on midsagittal images)
Multiple-system atrophy (MSA) <ul style="list-style-type: none">• Parkinsonian (MSA-P) or striatonigral degeneration• Cerebellar (MSA-C) or olivopontocerebellar atrophy• Autonomic (MSA-A)• form or Shy-Drager syndrome	Parkinsonism in conjunction with cerebellar signs and/or early and prominent autonomic dysfunction, usually orthostatic hypotension. Cerebellar and brainstem atrophy (the pontine 'hot cross buns' sign in MSA-c)
Corticobasal ganglionic degeneration (Rebeitz-Kolodny-Richardson syndrome)	Asymmetric dystonic contractions and clumsiness of one hand coupled with cortical sensory disturbances manifest as apraxia, agnosia, focal myoclonus, or alien limb phenomenon
Dementia with Lewy bodies	Early onset dementia, visual hallucinations
Parkinsonism-dementia complex of Guam	Motor neuron disease plus Parkinson's
Guadeloupean parkinsonism	Levodopa-unresponsive parkinsonism, postural instability with early falls, and pseudobulbar palsy