Nervous system

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Introduction to nervous system

Upper and lower motor neuron (UMN and LMN)

There are 2 main groups of nerve fibres coming down from the cerebral cortex:

1. Corticospinal fibres
2. Corticonuclear fibres.

These 2 group of fibres relay on the following stations respectively:

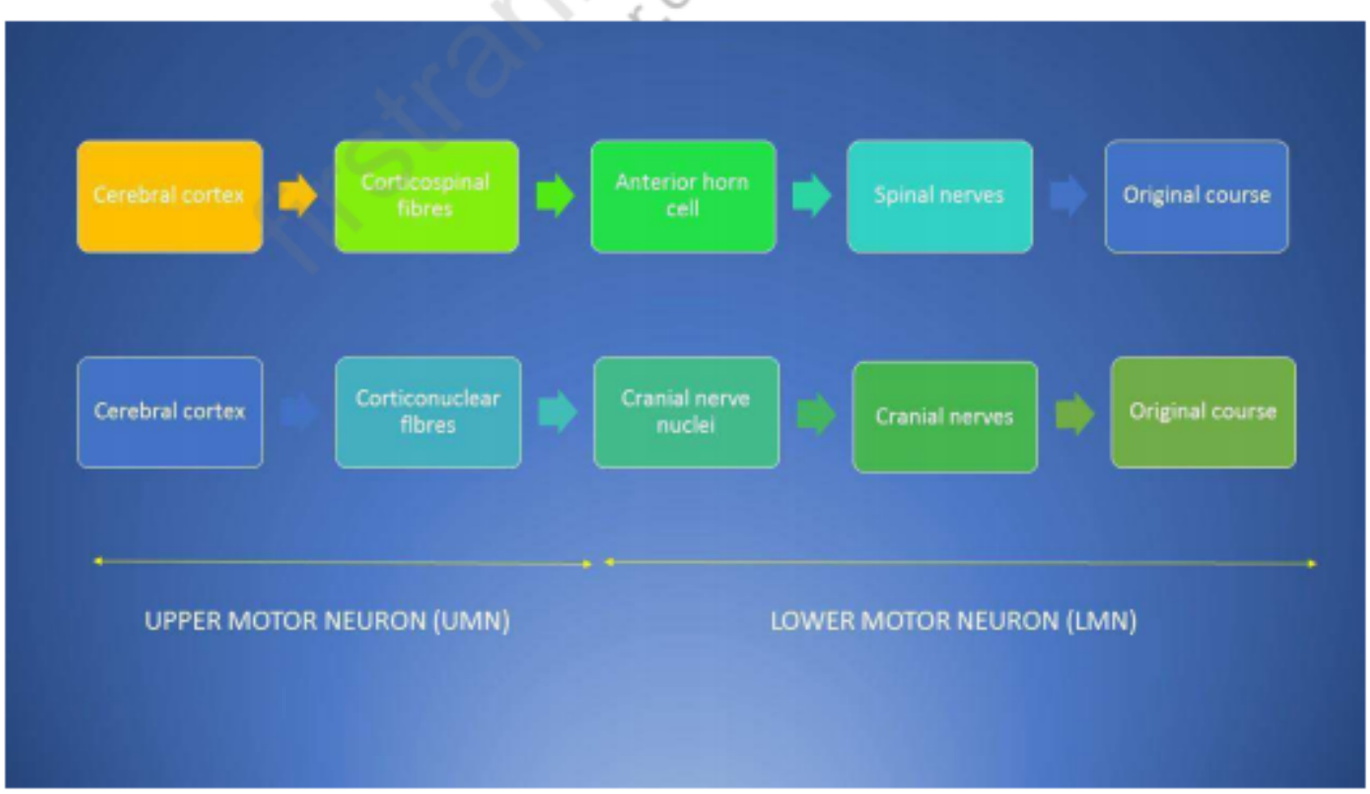
1. Anterior horn cell
2. Cranial nerve nuclei.

Till these 2 stations, the pathway are called 'upper motor neurons'.

After this stations, the following 2 sets of nerves starts respectively and they follow their original course:

1. Spinal nerves
2. Cranial nerves.

They comprise the 'lower motor neurons'.



Actions of UMN and LMN:

- Through upper motor neurons, cortex exerts a negative control over the lower motor neurons. So, the main function of UMN is control of voluntary movements.
- The main function of LMN is control of muscle movements.

Basic feature of UMN and LMN lesions:

- UMN lesion: Loss of voluntary movement
- LMN lesion: Paralysis of muscle.

Details of features of UMN and LMN lesions:

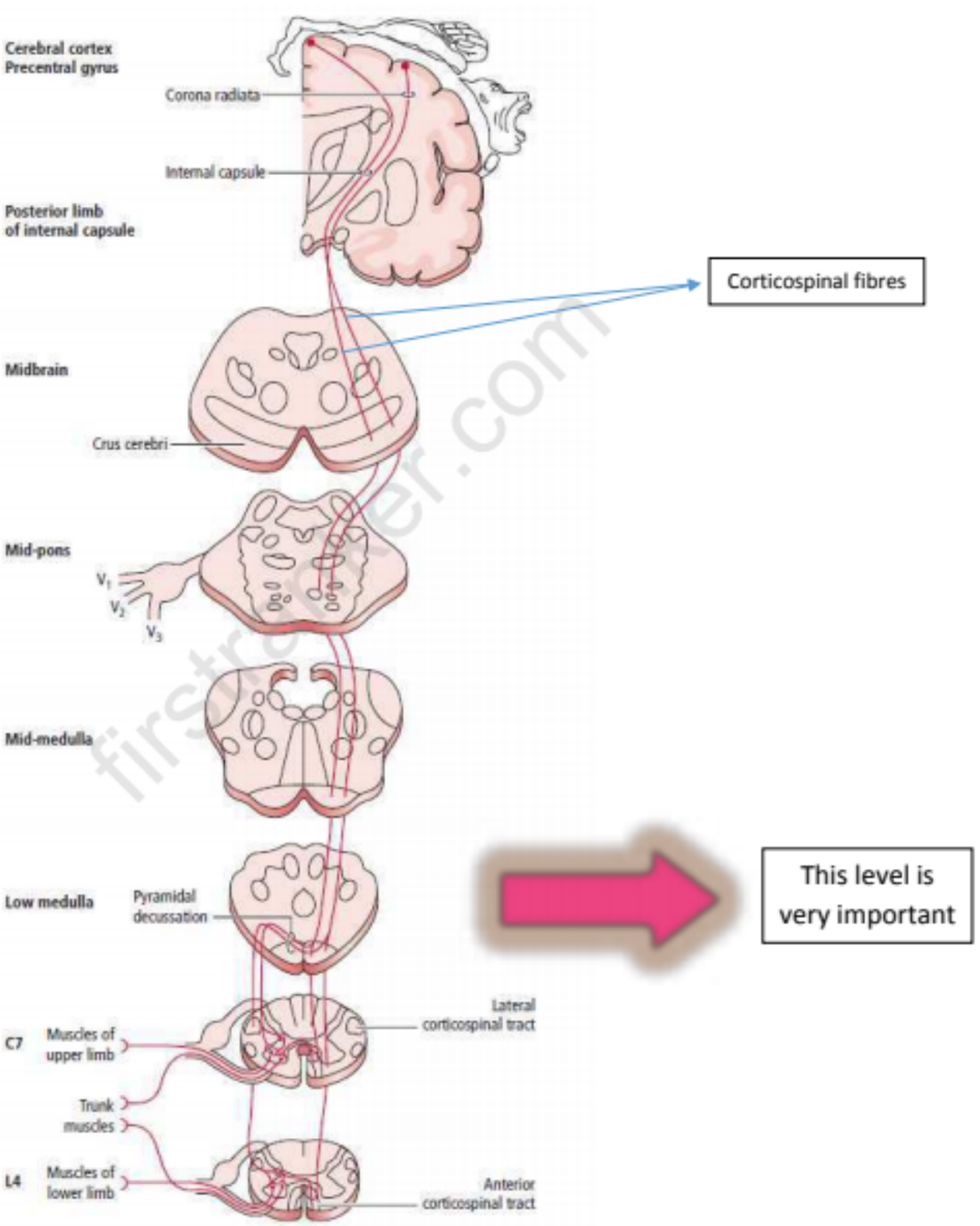
Points	UMN lesion*	LMN lesion*
Muscle power	Decreased	
Muscle tone	Hypertonia/ spasticity	Hypotonia/ flaccidity
Wasting/ atrophy of muscles	Absent	Present (due to denervation)
Deep tendon reflex (DTR)/ Jerk/ Stretch reflex	Exaggerated (clonus may be present)	Absent
Superficial reflex	Planter: Extensor Other reflexes: Lost	Plantar: Unresponsive Other reflexes: Lost
Above signs will be present in that part of the body which is innervated by the affected/ damaged UMN/LMN.		

*UMN may be described as the overhead wire over the railways and LMN may be described as the rail track. If the overhead wire gets cut, there is still a chance of running the train by other methods (like motor/ diesel etc.) but if the rail track is damaged, there is no way to run the train.

Just like that, if UMN is damaged, the muscles can still work (and in reality they work with hyperactivity due to loss of cortical control) but if LMN is damaged, the muscles can't work at all and they become flaccid, atrophied with loss of all reflexes.

Corticospinal fibres

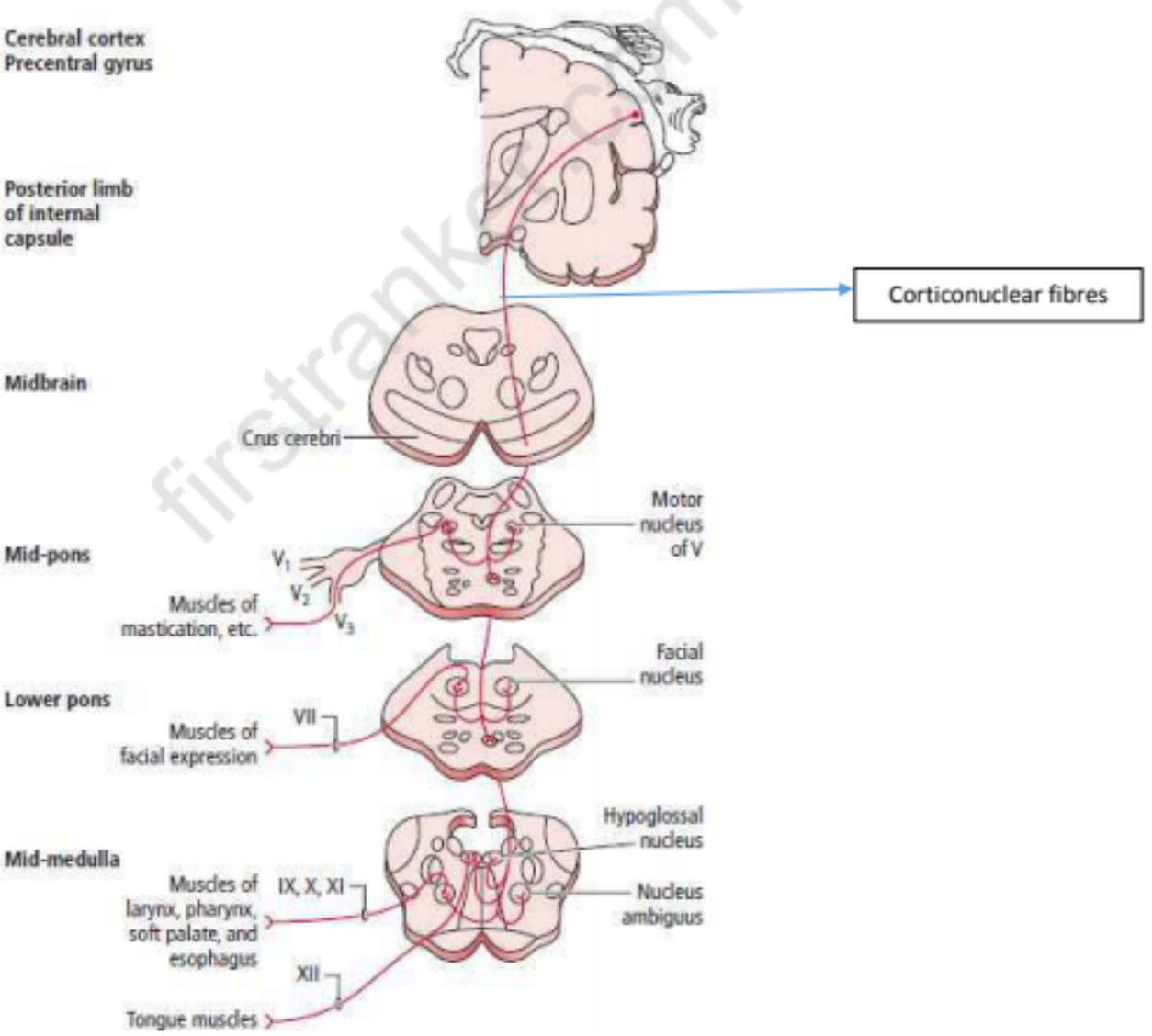
Course



- Corticospinal fibres control movement of contralateral (opposite) half of the body.
- So, if corticospinal tract is damaged in the brain, effect will be contralateral to the side of lesion.
- If corticospinal tract is damaged within the spinal cord, effect will be ipsilateral (same) to the side of lesion.
- So, in a corticospinal tract lesion, UMN signs will be present in that part of the body which is controlled by the fibres below the level of the lesion.

Corticonuclear fibres

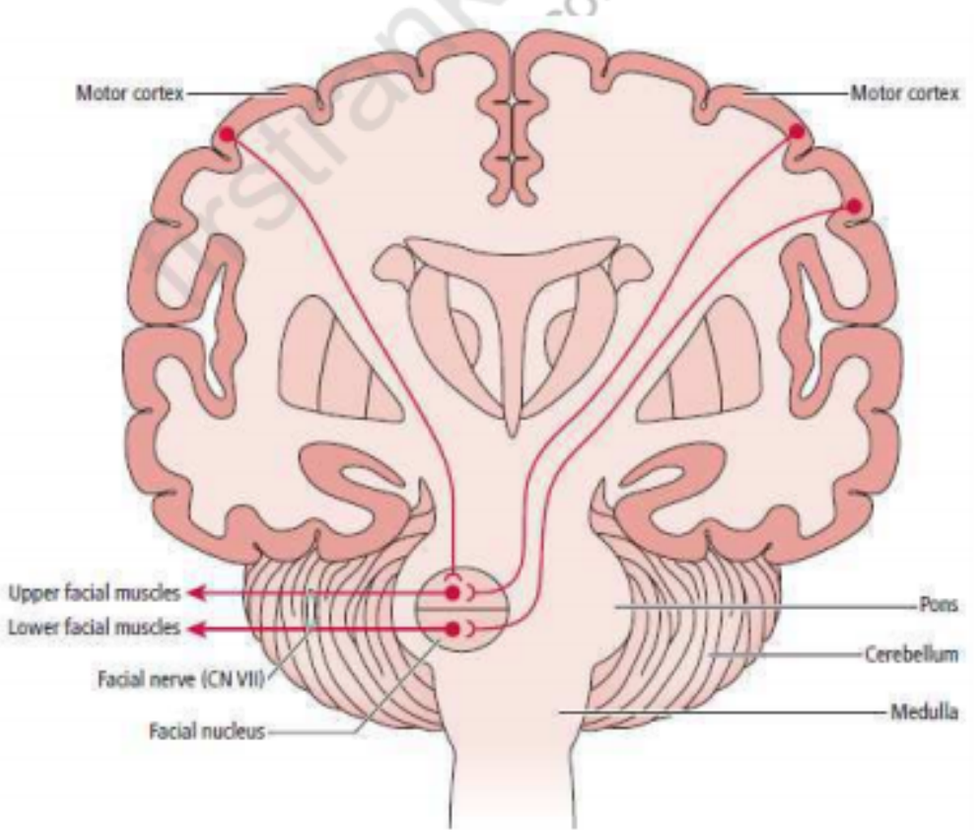
Course



- They arise from cortex and ultimately innervate different motor cranial nerve nuclei in the brainstem, forming UMN pathway of that particular cranial nerve.
- Therefore, all of the motor cranial nerve nuclei are bilaterally innervated by corticonuclear fibres (*except lower half of 7th cranial nerve nucleus*).

The case of 7th cranial nerve corticonuclear fibres

- CN7 nucleus has an upper ½ and a lower ½ from which, upper and lower ½ of facial muscles are innervated.
- Corticonuclear fibres of the upper ½ of the CN7 nucleus act exactly like the other motor cranial nerve nuclei. Therefore, *upper ½ of CN7 nucleus is innervated by corticonuclear fibres from both side of motor cortex*.
- But, corticonuclear fibres destined for lower ½ of CN7 nucleus leave rest of the pyramidal fibres at the level of midbrain and decussate to innervate the nuclei.
- Therefore, *lower ½ of CN7 nucleus has got unilateral corticonuclear innervation which comes from contralateral side*.



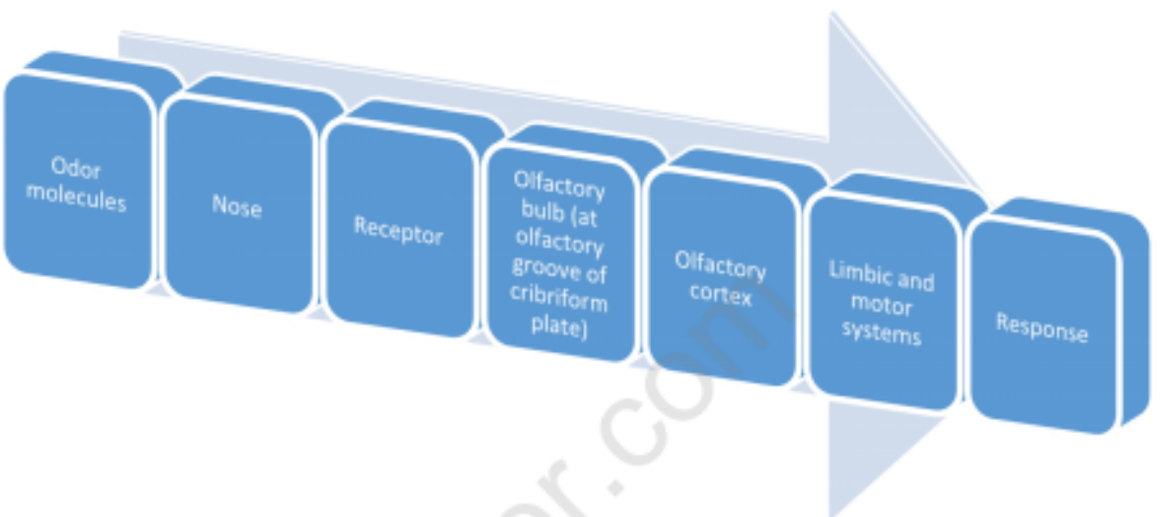
Localization of corticospinal tract lesion (compare with the pictures above)

Site	Structure damaged	Effect
Cortex	Corticospinal tract	Contralateral hemiplegia
	Corticonuclear fibre innervating lower ½ of CN7 nucleus	Paralysis of contralateral lower ½ of face (UMN type CN7 palsy)
	Higher cortical areas	Higher cortical dysfunction (Ex: speech area damage: aphasia)
	Fibres of visual field	Visual field defect
	Because the pyramidal fibres are widely separated in cortex, a cortical lesion often causes isolated monoparesis/ asymmetrical weakness of the limbs/ weakness of contralateral lower ½ of face.	
Internal capsule	Corticospinal tract	Contralateral hemiplegia
	Corticonuclear fibre innervating lower ½ of CN7 nucleus	Paralysis of contralateral lower ½ of face (UMN type CN7 palsy)
Midbrain	Corticospinal tract	Contralateral hemiplegia
	Corticonuclear fibre innervating lower ½ of CN7 nucleus	Paralysis of contralateral lower ½ of face (UMN type CN7 palsy)
	CN3 nucleus	***Ipsilateral LMN type CN3 palsy***
	CN4 nucleus	***Ipsilateral LMN type CN4 palsy***
Pons	Corticospinal tract	Contralateral hemiplegia
	Corticonuclear fibre innervating lower ½ of CN7 nucleus	Paralysis of contralateral lower ½ of face (UMN type CN7 palsy)
	CN6 nucleus	***Ipsilateral LMN type CN6 palsy***
	Principal sensory nucleus of CN5	***Loss of touch sensation from ipsilateral ½ of face***
	Sympathetic trunk	Ipsilateral Horner's syndrome
	Lateral spinothalamic tract	Loss of pain and temperature sensation from contralateral ½ of body
Medulla	Corticospinal tract	Contralateral hemiplegia
	CN 9,10,11,12 nucleus	***LMN type bulbar palsy***
	Sympathetic trunk	Ipsilateral Horner's syndrome
	Lateral spinothalamic tract	Loss of pain and temperature sensation from contralateral ½ of body
	Spinal nucleus of CN5	***Loss of pain and temperature sensation from ipsilateral ½ of face***

Cranial nerves

CN1

Simplified olfactory pathway



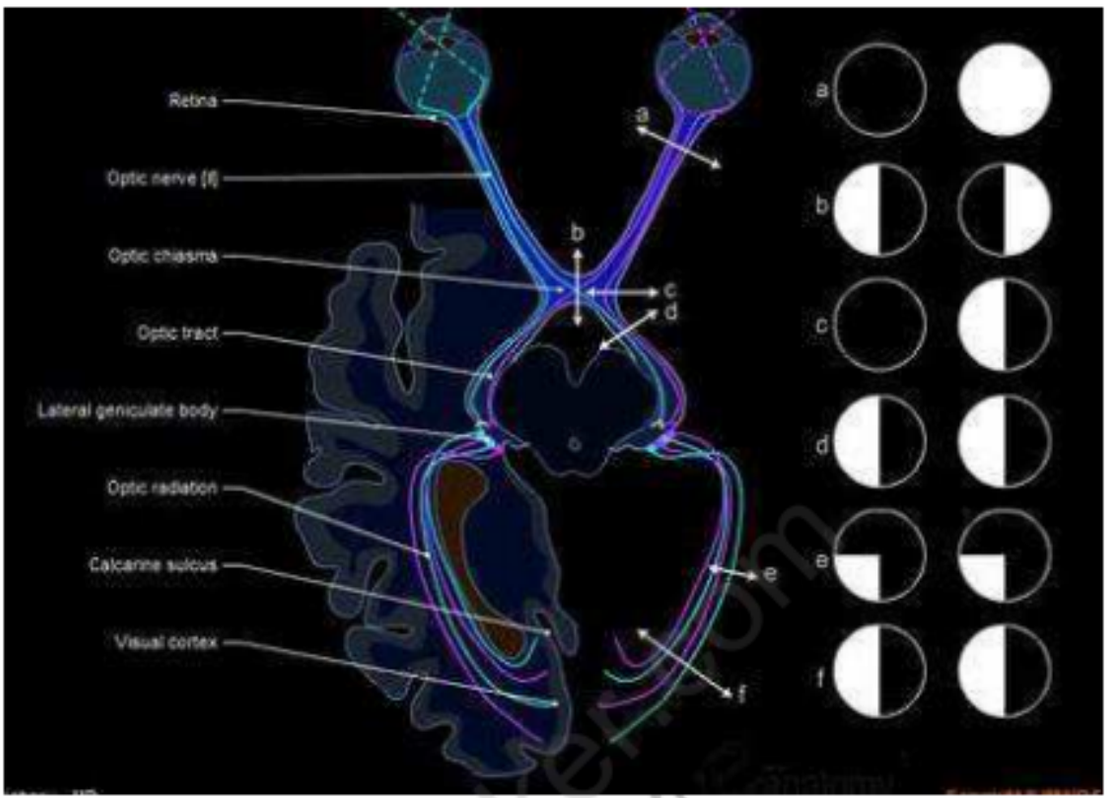
Hyposmia/ Anosmia:

1. Transport defect: DNS/ Polyp/ Rhinitis
2. Receptor defect: Viral infection
3. Neural defect:
   a. Trauma/ fracture of cribriform plate
   b. Tumor: Meningioma of olfactory groove
   c. Alzheimer's disease
   d. Chronic smokers
   e. Kalmann's syndrome (anosmia + hypogonadism)
   f. Foster-Kennedy syndrome (anosmia + ipsilateral optic atrophy + contralateral papilledema).

CN2

Functions of the optic nerve

1. Visual acuity
2. Color vision
3. Visual field.



(White: Lost field of vision)

Lesions at different levels and their effects

Level of lesion	Effect
Optic nerve	Monocular loss of vision from both the fields of the affected side In the affected eye: • Direct light reflex (DLR): Absent • Consensual light reflex (CLR): Preserved - This defect is called RAPD (Relative afferent pupillary defect)/ Marcus Gunn Pupil.
Optic chiasma	Bitemporal hemianopia (both temporal fields are lost) Some of the important causes are: • Pituitary tumor • Supra-sellar tumor • Craniopharyngioma.
Optic tract, LGB and optic radiation lesion: Homonymous hemianopia (identical half of the visual field of each eye is lost), crossed/ contralateral in types (as the contralateral half of the visual field is lost).	

Optic tract	Congruous in type (as the fibres within optic tract are densely arranged; when the visual field loss is mapped out, they look symmetrical/ identical).
Optic radiation	Congruous in type (The fibres within optic radiation are widely separated, *but the fibres responsible for superior and inferior quadrantic vision group together* and pass through parietal and temporal lobes, respectively).

Optic neuritis (SN)

Introduction

Inflammation of optic nerve.

Etiology

1. Multiple sclerosis (MS)
2. Viral infection
3. Devic's disease.

Clinical features

1. Monocular loss of vision
2. Orbital/ retro-orbital pain, particularly on upward gaze due to inflammation of the superior rectus tendon
3. Loss of vision from nasal as well as temporal field
4. Affected eye: DLR lost but CLR present.

Investigation

MRI orbit

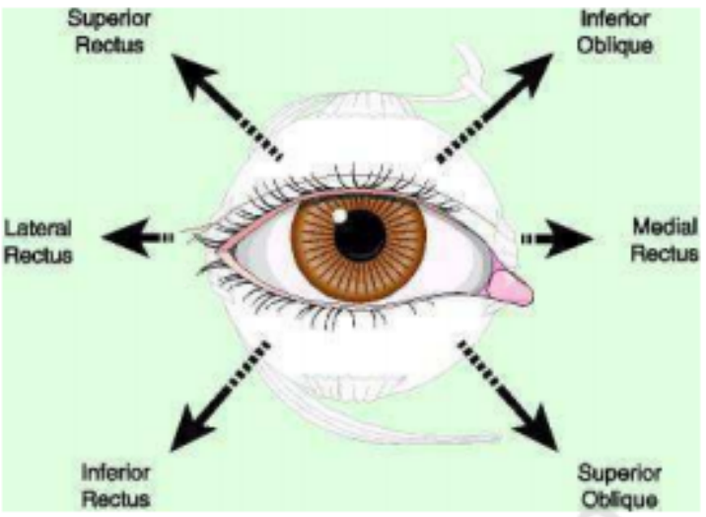
Treatment

High dose corticosteroid

CN3, CN4 and CN6

Function

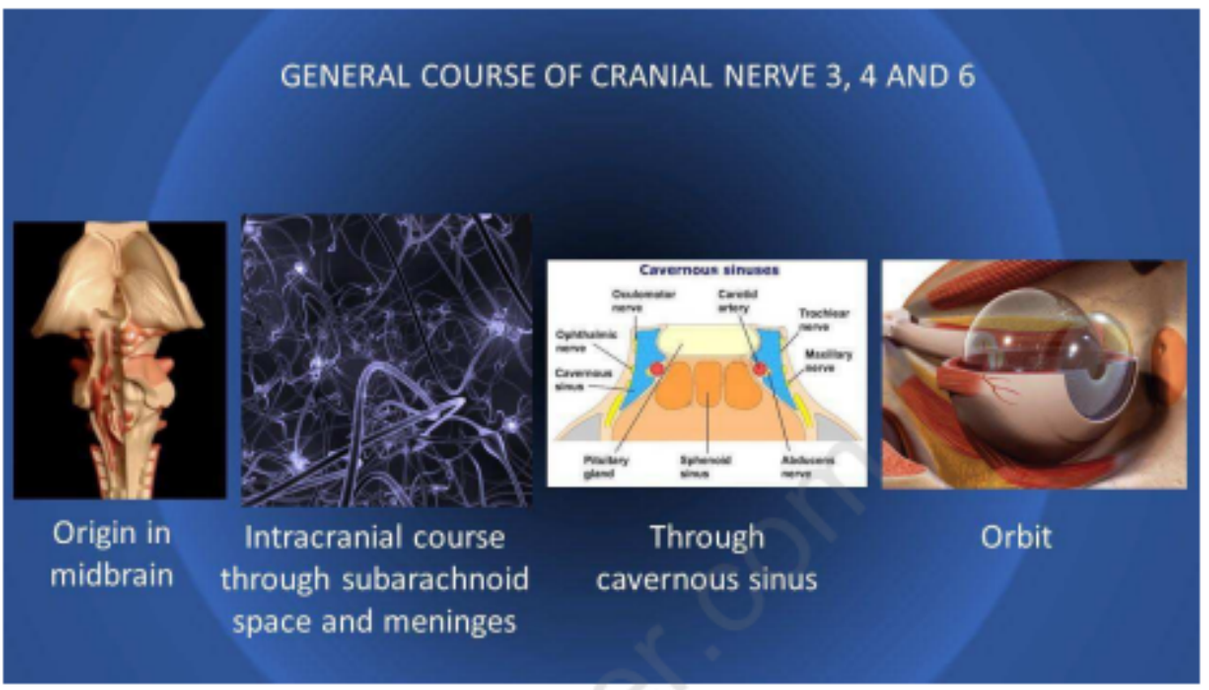
1. Innervate extraocular muscles and thereby mediate movement of eyeball.
   Innervation of EOM: [ALL]3 [SO]4 [LR]6



2. CN3 innervates Levator palpebrae superioris (LPS), which is an eyelid elevator.
3. CN3 causes constrictor pupillae.
4. CN3 mediates accommodation reaction.

Affected nerve	Symptoms and sign of palsy
CN3	Symptoms • Diplopia: When the patient tries to look towards the field of vision of the affected muscle, diplopia occurs. Often diplopia gets masked due to complete ptosis. • Ptosis: Drooping of the upper eyelid. Signs • Attitude of the eyeball: Divergent squint • Weakness of the affected muscle is elicited • Light reflex: Lost (as the efferent pathway is through CN3) • Accommodation reflex: Lost.
CN4	• Diplopia: Particularly when the patient climbs downstairs (Superior oblique action acts more) • Weakness of superior oblique is elicited.
CN6	• Diplopia: On lateral gaze of the affected eye (Lateral rectus acts more) • Attitude of the eyeball: Convergent squint.

Some important causes of CN3, CN4 and CN6 palsy



Causes of CN3 palsy

Group	Cause
Diseases of midbrain	• CVA • Tumor • Abscess
Meningeal diseases	In bacterial meningitis, the inflammatory exudate often strangulate the nerve at the basal meninges.
Diseases of Subarachnoid space	Hemorrhage: Particularly in aneurysm of posterior communicating artery (PCA).
Diseases of Cavernous sinus	• Cavernous sinus thrombosis • Carotid-cavernous fistula • Lateral extension of pituitary (uncommon).
Orbital diseases	• Tumor • Cellulitis.
Others	Mononeuritis multiplex (secondary to diabetes and vasculitis)

Causes of CN4 palsy (rare)

1. Head injury
2. Midbrain diseases
3. Meningeal diseases
4. Cavernous sinus diseases
5. Orbital diseases.

[Point 2-5 are same as CN3 palsy]

Causes of CN6 palsy

1. **Raised ICT:** As CN6 has the longest intracranial course, fibres often get stretched when ICT raises; giving rise to some *false localizing signs* in addition to specific signs of CN6 palsy.
2. Pontine lesion (as the nucleus of CN6 lies in the pons)
3. Meningeal diseases
4. Cavernous sinus diseases
5. Orbital lesions
6. Mononeuritis multiplex.

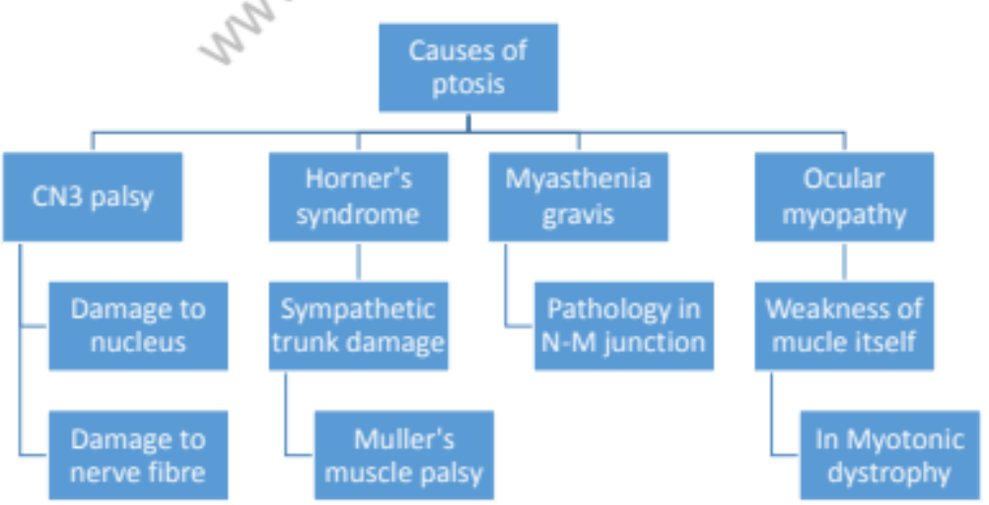
[Points 3-6 are same as CN3 palsy]

Some short notes related to CN3, 4 and 6

Ptosis (SN)

Introduction

Drooping of upper eyelid.



Clinical features according to the causes of ptosis

CN3 palsy	Horner's syndrome	Myasthenia gravis
Complete ptosis	Partial ptosis/ pseudo-ptosis	Fatigable ptosis
Non-fatigable	Non-fatigable	
Usually unilateral	Usually unilateral	Usually bilateral
Pupil: Large	Pupil: Small	Pupil: Normal
Other signs of CN3 palsy are present	Other features of Horner's syndrome are present	Fatigable weakness of other muscles in the body may be present

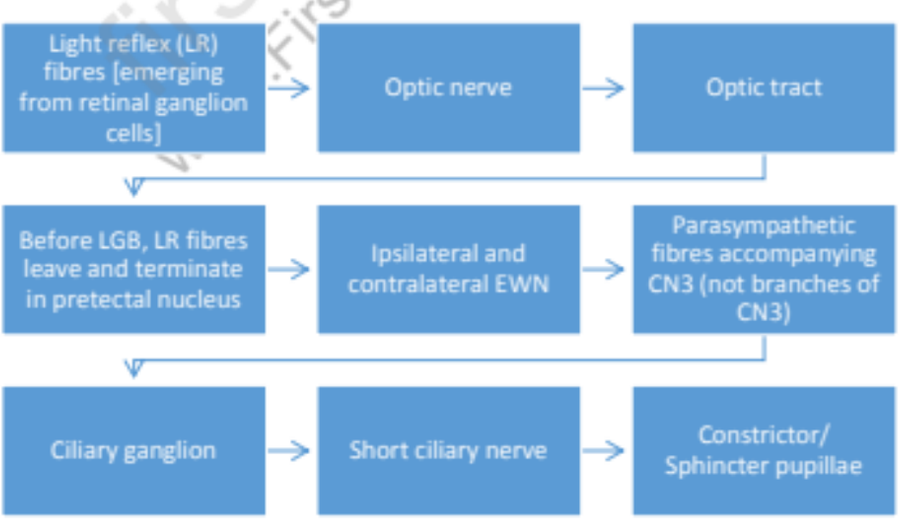
Squint: Brief discussion

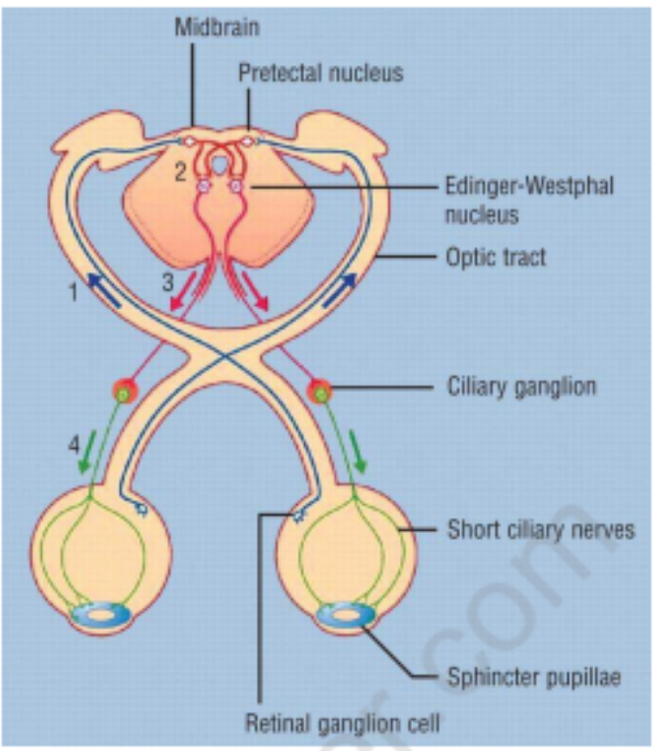
Squint is of 2 types, the features of which are being discussed in brief:

Non-paralytic	Paralytic
Present since birth	Usually develops later
No diplopia	Diplopia present
Movement of eyeball not restricted (as there is weakness of EOM)	Movement of eyeball restricted
No signs of cranial nerve palsy	Signs of cranial nerve palsy present

Light reflex

Pathway of light reflex





Note: Parasympathetic fibres (which constricts pupil) pass along CN3 fibres and therefore, often damaged in CN3 nerve lesions.

Loss of light reflex

Causes:

1. Optic nerve lesion
2. Optic tract lesion (pre-geniculate lesion):
   The affected eye in this case is called "*Wernicke's hemianopic pupil*": DLR is lost in this eye when light is thrown from nasal half, but DLR is present when light is thrown from temporal half. (Remember- W:DT)
   In contralateral eye: DLR is lost when light is thrown from temporal half and DLR is present when light is thrown from nasal half.
3. CN3 palsy
4. Argyll Robertson Pupil (SN):
   It is an abnormality of pupil usually seen in patients of ***neurosyphilis***.
   Component:
   a. Usually bilateral
   b. Pupil is small in size and dilates poorly to mydriatics

c. Light reflex is lost
d. Accommodation reaction: Present (Remember: ARP in ARP).

Accommodation reaction

Component:

1. Medial convergence of eyeball
2. Pupillary constriction
3. Anterior-posterior bulging of lens.

Lost accommodation reaction:

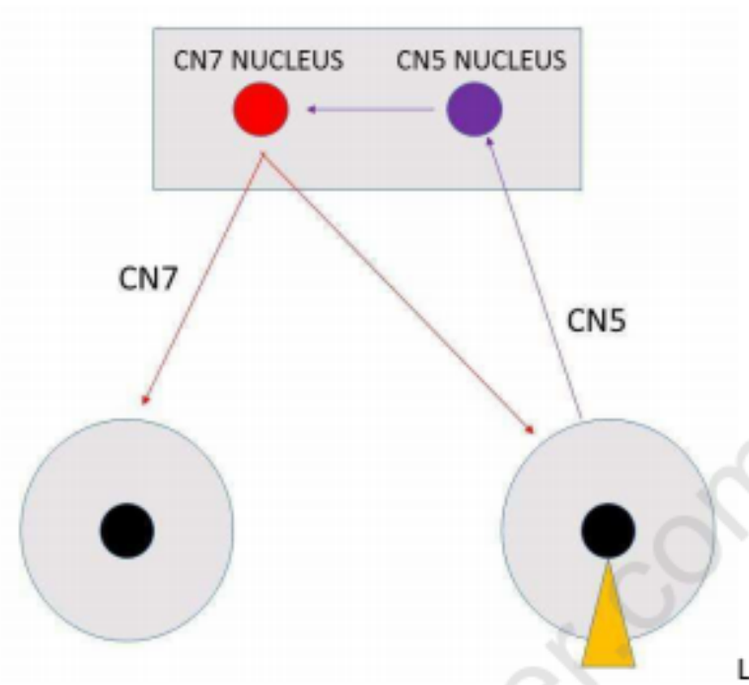
Seen in CN3 palsy (as CN3 supplies both medial rectus and constrictor pupillae).

CN5

Functions:

1. Motor function:
   Innervates masticatory muscles:
   a. Temporalis
   b. Masseter
   c. Pterygoid.
2. Sensory function:
   Carries superficial sensation from ipsilateral half of face through 3 branches:
   a. Ophthalmic
   b. Maxillary
   c. Mandibular.
3. Reflex:
   CN5 mediates the following reflexes:
   a. Corneal reflex
   b. Jaw jerk.

Corneal reflex pathway



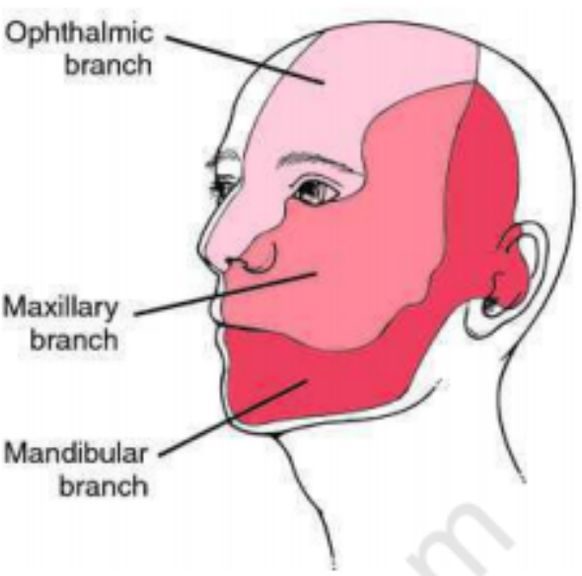
Loss of corneal reflex

Causes:

1. CN5 palsy (damage to ophthalmic division):
   In the affected side, direct corneal reflex will be lost but consensual corneal reflex will be present.
2. CN7 palsy:
   In the affected side, both type of corneal reflex (direct and consensual) will be lost.
3. Deep coma.

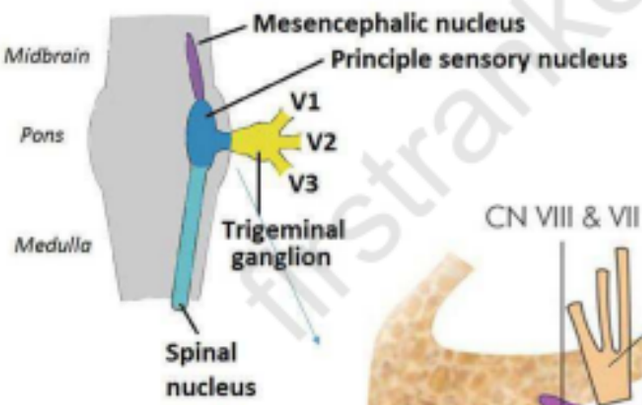
Features of CN5 palsy

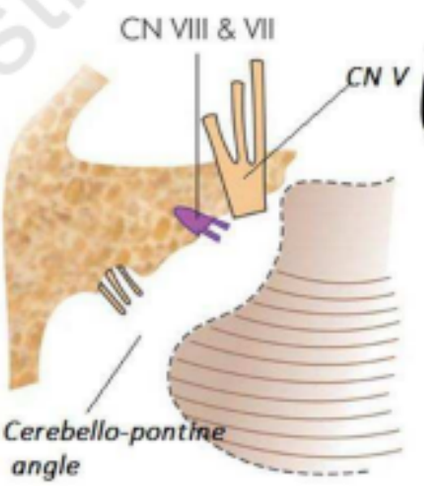
1. Motor dysfunction:
   a. Weakness of masticatory muscles leading to *difficulty in mastication*
   b. Wasting of temporalis and masseter, leading to *hollowing of the face*.
2. Sensory dysfunction:
   Loss/ impairment of sensation from ipsilateral half of face.
   In many cases, only one sensory branch of trigeminal nerve gets damaged, leading to sensory impairment from the distribution of the affected branch only.

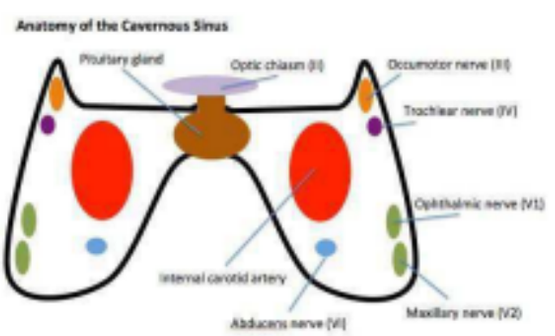


3. Loss of corneal reflex.

Causes of CN5 palsy







General course of CN5

Brainstem nuclei-→ Post. Cranial fossa (CP angle)-→ Trigeminal ganglion-→ Cavernous sinus (V1 & V2 only)-→ Supply the face.

1. Brainstem lesion: CVA
   a. Pontine lesion:
      - Motor paralysis
      - Touch sensation lost

b. Medullary lesion:
   - Pain sensation lost
   - Temperature sensation lost

2. Posterior fossa lesion:
   CP angle tumor (Acoustic neuroma)
3. Trigeminal ganglion lesion:
   a. Tumor
   b. Trauma
   c. Gradenigo's syndrome
4. Cavernous sinus pathology:
   Thrombosis.

Trigeminal neuralgia (SN)

Introduction

It is a condition characterized by excruciating pain along the distribution of trigeminal nerve.

Etiology

1. Idiopathic
2. Multiple sclerosis
3. Intracranial space occupying lesion (IC-SOL): Tumor stretches the nerve fibres
4. In some cases, the nerve gets irritated by an aberrant blood vessel.

Clinical features

Pain:

a. Site: Usually hemifacial pain, particularly in the chick and chin
b. Nature: Severe, often electric shock like pain and typically paroxysmal
c. Triggering factor: Often triggered by face washing, shaving, chewing
d. The pain is so severe that it throws the face; causing *facial spasm* and the patient involuntarily starts *wincing like a tic*; therefore the condition is also called *Tic douloureux**.

[* It has been described as among the most painful conditions known to humankind.]

Investigation

Often not required as it is a clinical diagnosis. Relevant investigations to look for any underlying diseases are performed in selected cases.

Treatment

1. Medical:
   Carbamazepine/ Oxcarbamazepine
2. Interventional:
   a. Radiofrequency ablation of trigeminal ganglion
   b. Microvascular compression of aberrant vessels.

CN7

Functions:

Innervates all muscles of face except masticatory muscles (muscles of facial expression) which are supplied by CN5.

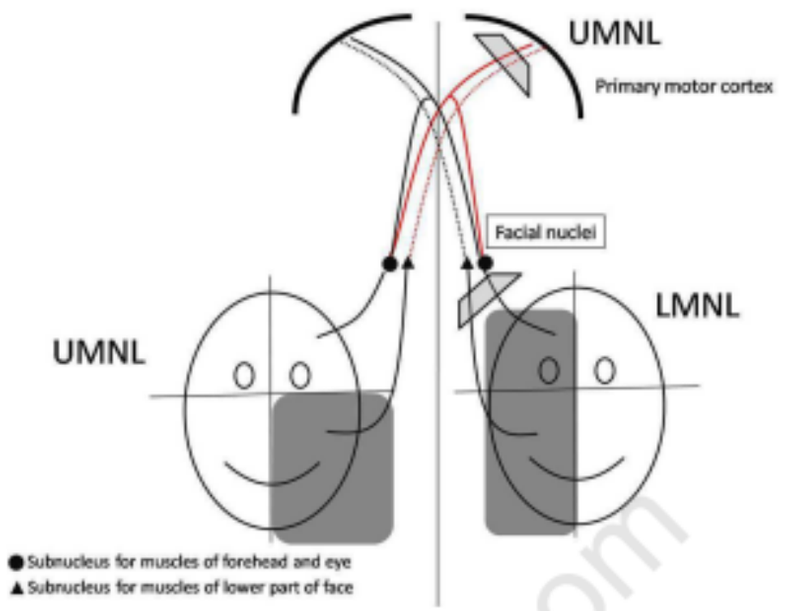
There are 2 other types of fibres which accompany facial nerve almost along its entire course and therefore considered to be a part of CN7:

a. Secretomotor fibres to (lacrimal + sublingual + submandibular) gland
b. Taste fibre from ant. 2/3rd of the tongue (post 1/3rd: supplied by CN9).

CN7 palsy

It is of 2 types:

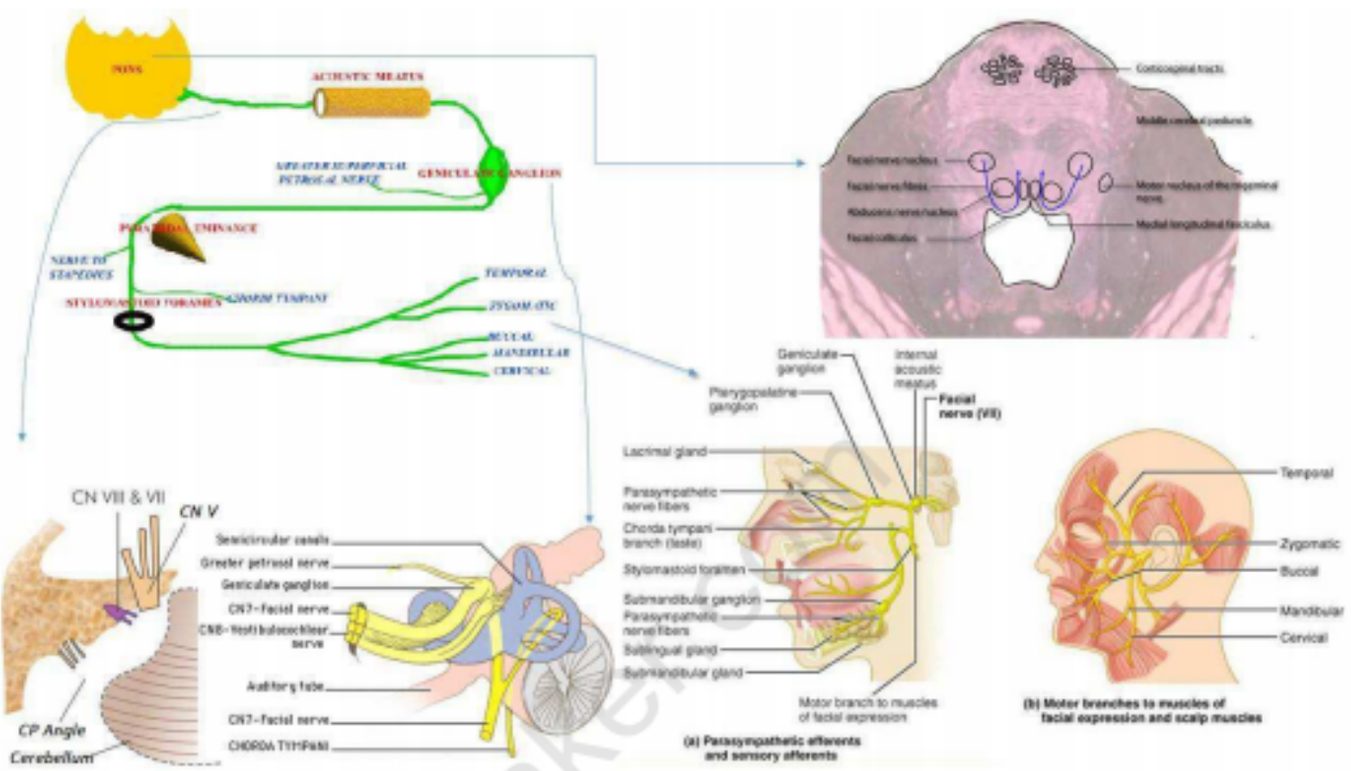
UMN type of CN7 palsy	LMN type of CN7 palsy
Due to lesions of the corticonuclear fibres innervating lower ½ of contralateral CN7 nucleus	Due to nucleus damage/ infra-nuclear lesion
Lesion is anywhere above pons (where CN7 nucleus lies)	Lesion is in the pons/ anywhere along the course of CN7
Feature: Weakness/ paralysis of lower ½ of contralateral side of face	Feature: Weakness/ paralysis of whole of the ipsilateral side of face



Features of CN7 palsy

- ✓ Inability to elevate the eyebrows
- ✓ Loss of forehead furrowing/ wrinkling
- ✓ Inability to frown
- ✓ Inability to close the eyelid:
  Eyeball is seen to be rolled upwards on attempted closure of eye. This is called '*Bell's phenomenon*'. Such an eye is at risk of developing keratitis.
- ✓ Weakness of buccinator leading to following manifestations:
  - On puffing out of chick, air leaks through the angle of the mouth of the affected half
  - Loss of nasolabial fold
  - Lower eyelid sacs down so that punctum no longer remains in contact.
- ✓ Drooping of the angle of the mouth to the affected side: saliva dribbles through angle of mouth
- ✓ Angle of mouth is deviated towards healthy site when patient attempts to smile.

Localization of LMN type of Facial palsy



Site of lesion	Disease	Features (Compare with the picture above)
Pons	CVA	a. Ipsilateral LMN type CN7 palsy b. Ipsilateral CN6 palsy c. Contralateral hemiplegia (CST damage) d. Loss of secretomotor (taste) function.
CP angle	Acoustic neuroma	a. Ipsilateral LMN type CN7 palsy b. Ipsilateral CN5 palsy c. Ipsilateral sensorineural deafness (CN8 damage) d. Ipsilateral cerebellar signs e. Loss of secretomotor function.
Temporal bone	Trauma Tumor Petrositis (Gradenigo's syndrome)	a. Ipsilateral LMN type CN7 palsy b. Loss of secretomotor function c. Hyperacusis (nerve to stapedius)
Geniculate ganglion	Herpes zoster (Ramsey-Hunt syndrome)	Same as temporal bone lesion + Painful herpetic vesicles over external ear and pinna
Stylomastoid foramen	Bell's palsy	Ipsilateral LMN type CN7 palsy
Parotid gland	Tumor	Ipsilateral LMN type CN7 palsy

	Surgery	
Peripheral branches	GB syndrome Neurosarcoidosis Lyme's disease Complication of forceps delivery	Ipsilateral LMN type CN7 palsy

Bell's palsy (SN)

Introduction

It is a rapidly developing LMN type of ipsilateral CN7 palsy due to inflammation of the nerve at/near the stylomastoid foramen.

Etiopathogenesis

It has been postulated that the inflammation of nerve is probably triggered out by HSV infection. Rapidly developing inflammatory exudate strangulates the nerve, leading to palsy. Because there is no permanent structural damage of the nerve in many cases, there is usually complete functional recovery.

Clinical features

1. Onset: Usually acute/ subacute: develops over few days
2. Often there is H/O a preceding exposure to cold air/ an attack of common cold
3. Clinical features of CN7 palsy.

Investigation

None required as it is a clinical diagnosis.

Treatment

1. Drugs:
   A. Systemic corticosteroid: Most effective if given within 24-48 hours of onset of symptoms. Usually a course of 10-14 days is given.
   B. Acyclovir: Although its role is doubtful, many clinicians prefer to prescribe it.
2. Prevention of exposure keratitis:
   A. Protective eyepad
   B. Lubricant eye drop/ ointment.
3. Physiotherapy.

Complications

1. Incomplete recovery leading to residual palsy
2. Contracture of facial muscles
3. Aberrant regeneration:
   a. Fibres originally innervating *facial muscles* side-track and innervate *lacrimal gland*, leading to lacrimation during eating (gustatory lacrimation/ crocodile tear).
   b. Fibres originally innervating *orbicularis oculi* side-track and innervate *orbicularis oris*, leading to involuntary twitching of angle of mouth during attempted closure of eye.

CN8

Test	Normal	Conductive deafness	SN deafness
Rinne	AC > BC (Rinne positive)	BC > AC (Rinne negative)	AC > BC
Weber	Not lateralised	Lateralised to poorer ear	Lateralised to better ear

CN 9, 10, 11 & 12

These cranial nerves are also called 'bulbar cranial nerves' as the nuclei lies in the bulb (medulla).

Functions:

- CN9: Carries sensation from pharynx, palate, tonsillar region; taste sensation from post. 1/3rd of tongue.
- CN10: Supplies all muscles of pharynx (except stylopharyngeus, which is supplied by CN9), palate and intrinsic muscles of larynx.
- CN11: Supplies sternocleidomastoid (rotate the head towards the opposite side) and trapezius (elevation and retraction of shoulder).
- CN12: Supplies tongue muscles.

Reflexes mediated by bulbar cranial nerves:

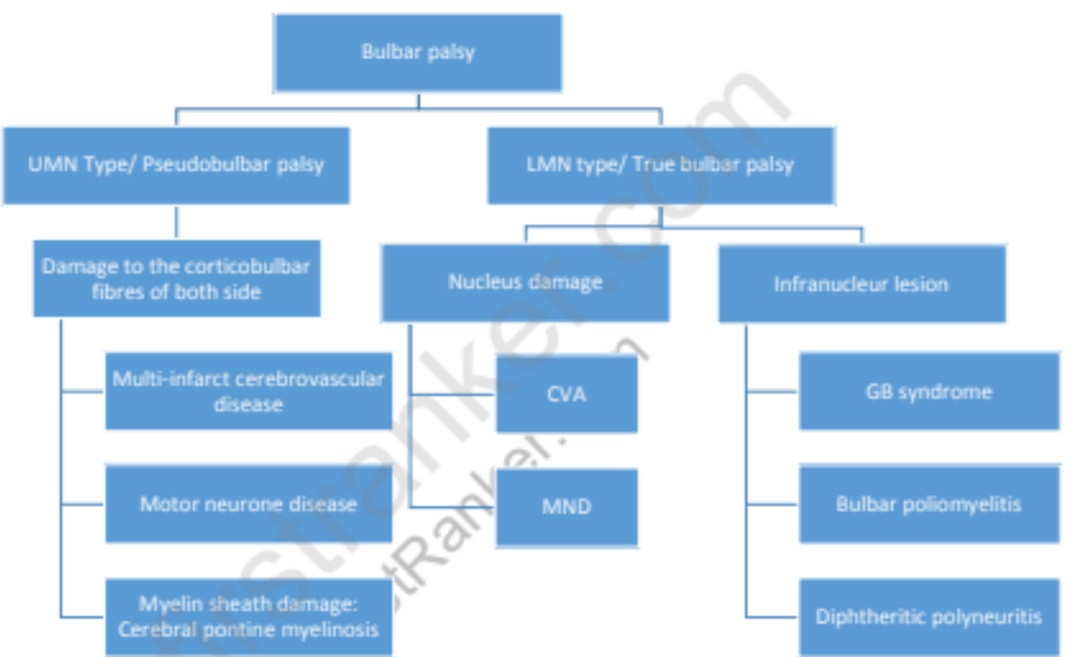
1. Pharyngeal/ Gag reflex:
   On stimulating the posterior pharyngeal wall/ palate with a swab stick, there is reflex contraction of pharyngeal/ palatal muscles, respectively.

The afferent pathway is formed by: CN9

The efferent pathway is formed by: CN10.

2. Jaw jerk:
   Patient is asked to open his mouth slightly and an area of chin just below the lower lip is tapped, which leads to reflex closure of mouth.
   In most normal individuals, response is non-elicitable and therefore, if the reflex deviation of lower jaw can be seen clearly, the jerk is considered to be brisk/ exaggerated.

Causes of bulbar palsy



Symptoms and signs of bulbar palsy (Pharyngo-laryngo-palatoglossal palsy)

Site	Symptom/ sign
Pharynx	Dysphagia Nasal regurgitation of food
Larynx	Dyspnea Recurrent episodes of aspiration pneumonia/ choking
Palate	Dysarthria Nasal intonation of voice

Differences between UMN type (pseudobulbar) and LMN type (true) bulbar palsy

Features	UMN type (Pseudo) bulbar palsy	LMN type (True)bulbar palsy
Dysarthria	Spastic dysarthria	Flaccid dysarthria
Wasting	No wasting	Wasting present
Fasciculation	No fasciculation	Fasciculation present
Jaw jerk	Brisk	Absent
Gag reflex	Lost	
Other feature	Emotionally unstable (laughs without any cause)	No such feature
Special features	As reflex movements are often spared, there features are not so prominent	As all movements are affected, nasal intonation of voice and nasal regurgitation of food are quite prominent

CN11 palsy

Clinical features

Sternocleidomastoid (SCM)

1. Paralysis/ weakness of SCM: Inability to rotate the head towards the opposite side
2. Wasting of SCM (in LMN type of lesion)
3. On attempted neck flexion, chin deviates towards the affected side.

Trapezius (TZ)

1. Inability to retract and elevate the shoulder
2. Wasting of TZ leading to loss of normal curvature of shoulder (in LMN type of lesion).

General topics

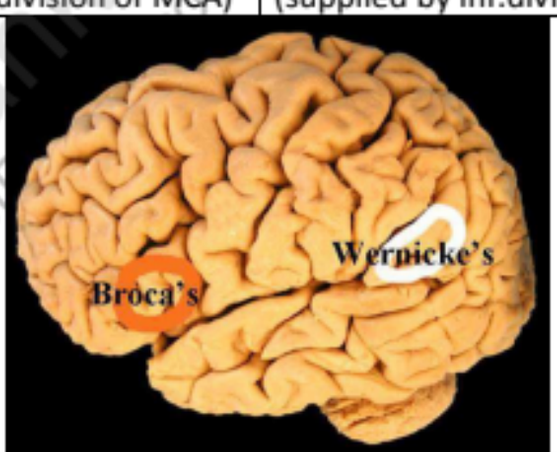
Topics to be discussed:

1. Higher function
2. Motor function: Tone, Power, Reflex, Atrophy
3. Sensory function
4. Sensory-motor coordination.

Higher function

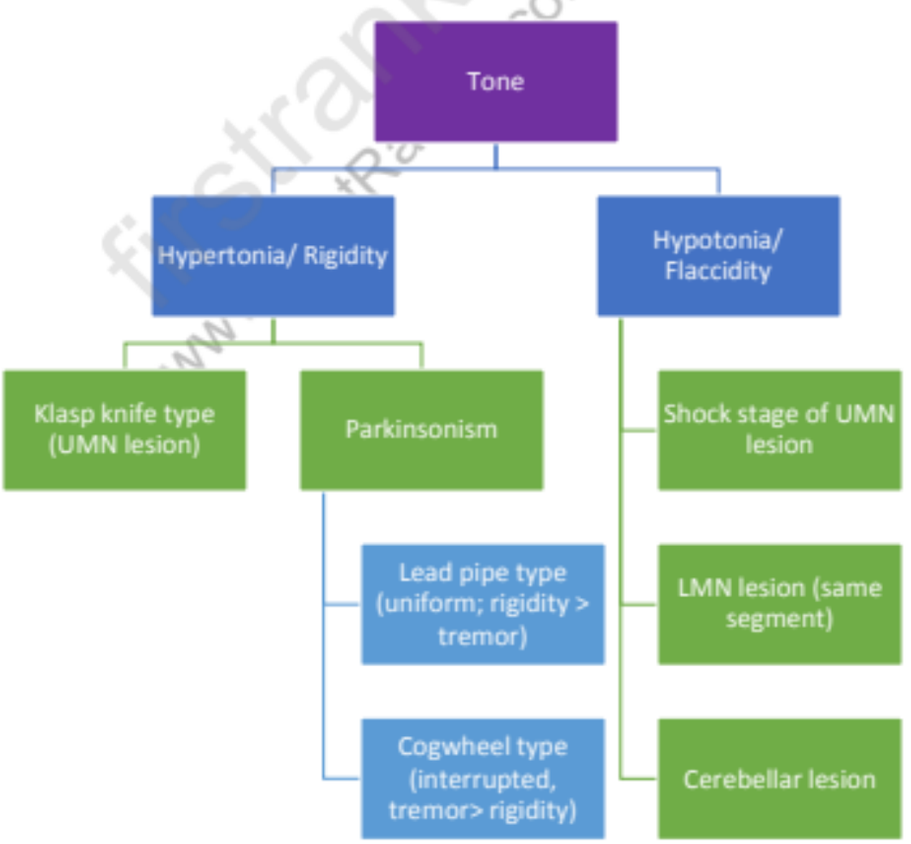
Speech:

a. Dysphasia
b. Aphasia:
   - ✓ Wernicke's
   - ✓ Broca's.

Features	Broca's aphasia	Wernicke's aphasia
Description	Due to lesion of inf.frontal gyrus (supplied by sup.division of MCA)	Due to lesion of sup. temporal gyrus (supplied by inf.division of MCA)
		
Comprehension	Intact	Impaired
Expression	• Impaired • Word finding difficulty • Short answers • Non-fluent aphasia/ telegraphic speech.	• Voluminous • Contains little information • Full of paraphasic errors • Neologism • Fluent aphasia/ Jargon speech

Motor function

Tone of muscles



Power of muscles

Power	Description
0	No movement
1	Flickering movement
2	Unable to move against gravity
3	Movement possible against gravity but not against resistance
4	Movement possible against resistance but with some weakness
5	Normal power

Reflex

Deep tendon reflex (DTR)

Reflex	***Spinal level***	***Peripheral nerve***
Biceps jerk	C5-C6	Musculocutaneous nerve
Supinator jerk	C5-C6	Radial nerve
Triceps jerk	C6-C7	Radial nerve
Finger flexion jerk	C7-C8	Median and ulnar nerve
Knee jerk	L2, L3, L4	Femoral nerve
Ankle jerk	L5, S1	Sciatic nerve

Abnormality of deep tendon reflexes:

1. ***Brisk/ exaggerated***:
   Cause: UMN lesion from non-neurological causes (anxiety/ thyrotoxicosis)
   Below the level of lesion, all reflexes will be brisk.
2. ***Clonus***:
   It is the external manifestation of an exaggerated DTR characterized by rhythmical repeated contraction of a muscle *in response to sudden sustained stretching of its tendon*, leading to oscillatory movement of a part of the body. It is best seen in the knee (patellar clonus) and in the ankle (ankle clonus).

Features	Pseudo-clonus/ Physiological clonus	True-clonus/ Pathological clonus
Extension	Ill sustained (<5-6)	Well sustained (≥7)
Signs of UMN lesion	Absent	Present
Cause	Anxiety	UMN lesion

3. ***Absent deep reflex***:

Causes:

- LMN lesion: Reflex(es) mediated by the particular damaged LMN pathway will be lost
- Spinal shock stage of UMN lesion.

Jendrassik's Maneuver

- It is a special maneuver performed while examining DTR when they seem to be absent/ diminished. It must be performed before concluding that the jerk is absent.
- In case of upper limb jerks, the patient is asked to clench his teeth tightly.
- In case of lower limb jerks, the patient is asked to hold his fingers tightly with each other and suddenly pull them up.
- Tapping of the tendon should coincide with this maneuver.
- Mechanism: This maneuver increases the excitability of reflex response by:
  a. By increasing the excitability of AHC
  b. By increasing the recruitment of γ-fusiform fibres of the muscle spindle.

Superficial reflex

Reflex	***Spinal level***
Abdominal reflex	T7-T12
Cremasteric reflex	L1-L2
Plantar reflex	S1
Anal reflex	S2-S4
Bulbocavernosus reflex	

Loss of superficial reflex:

- In UMN lesion: Below the level of lesion
- In LMN lesion: At the level of lesion.

Plantar reflex

Normal response:

1. Plantar flexion of the great toe
2. Plantar flexion and adduction of the other toes
3. Contraction of tensor fascia lata.

Why the lateral aspect of the sole is stroke?

- The reflexogenic area of the plantar reflex lies on the lateral aspect of sole
- If the medial aspect of sole is stroke, plantar reflex occurs due to grasp response.

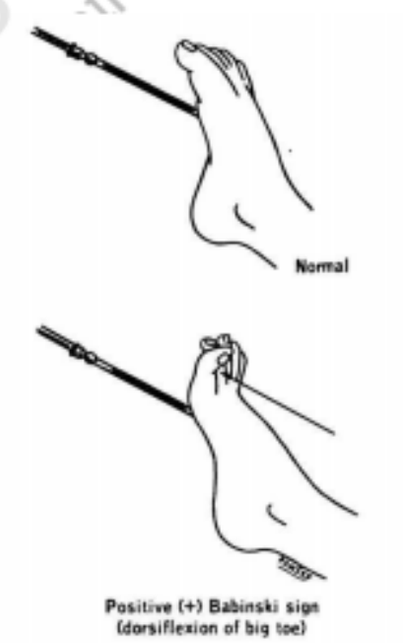
Why the ball of the great toe shouldn't be stroke?

If it is stroke, due to direct stimulation of muscle fibres; dorsiflexion of toe occurs giving an erroneous interpretation of extensor plantar.

Abnormality of plantar response:

Extensor plantar/ Babinski's sign/ Upgoing plantar

1. Dorsiflexion of great toe
2. Dorsiflexion and fanning out of the other toes
3. Triple flexion: Dorsiflexion at ankle + Flexion at knee + Flexion at hip.



Causes of Extensor plantar:

1. Structural damage of corticospinal/ pyramidal tract (usually bilateral extensor is seen)
2. Functional impairment of corticospinal tract:
   a. Newborn infant
   b. Any cause of deep unconsciousness/ coma
   c. Post-ictal stage.

Absence of plantar response:

1. LMN lesion of S1 segment
2. Thick sole
3. Anesthesia of sole.

Withdrawal response:

When the entire limb is withdrawn from nociceptive stimulation (usually occurs when patient is very sensitive to touch/ the sole is stroke forcefully).

Equivocal response:

Incomplete response. This is considered to be a part of extensor plantar response.

Ex: Great toe dorsiflexed but other toes plantiflexed.

Plantar equivalent:

In some patients with UMN lesion, the reflexogenic area of plantar reflex widens; leading to *extensor plantar response* when different parts of the lower limb are stimulated. Ex:

- *Gordon's sign*: When the calf muscle (Gastrocnemius) is squeezed
- *Chaddock's sign*: When the lateral malleolus is stroke in semicircular pattern
- *Oppenheim's sign*: When pressure is applied over the medial side of tibia in a downward direction.

Causes of extensor plantar with absent ankle and knee jerk

1. Spinal shock stage of UMN lesion

2. Compressive myelopathy where compression occurs in the fragment controlling the above jerk (S1)

Other uncommon causes:

1. Subacute combined degeneration of spinal cord
2. Fredrick's ataxia
3. Tabes dorsalis.

Muscle atrophy (Wasting)

Causes:

1. LMN atrophy
2. Myopathy.

Pseudo-hypertrophy:

The muscle bulk looks high due to abnormal accumulation of fibro-fatty tissues. It occurs typically in *Duchenne's muscular dystrophy* (which is a congenital myopathy) typically affecting the calf muscle.

Sensory function

1. Pain (mediated by lateral spinothalamic tract)
2. Temperature (-do-)
3. Touch (mediated by dorsal column)
4. Joint, position and vibration (-do-)
5. Cortical sensation.

Sensory-motor coordination

1. Finger nose test
2. Finger nose finger test
3. Keel knee test.

Spinal cord

Important structures of spinal cord:

Motor:

- Corticospinal tract ↓
- Anterior horn cell

Sensory:

- Lateral spinothalamic tract ↑
- Dorsal column ↑.

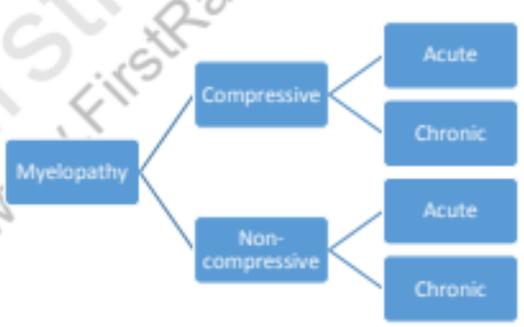
[↑: Ascending tract, ↓: Descending tract]

Myelopathy

Introduction:

Diseases affecting spinal cord.

Types:



Compressive myelopathy

Introduction:

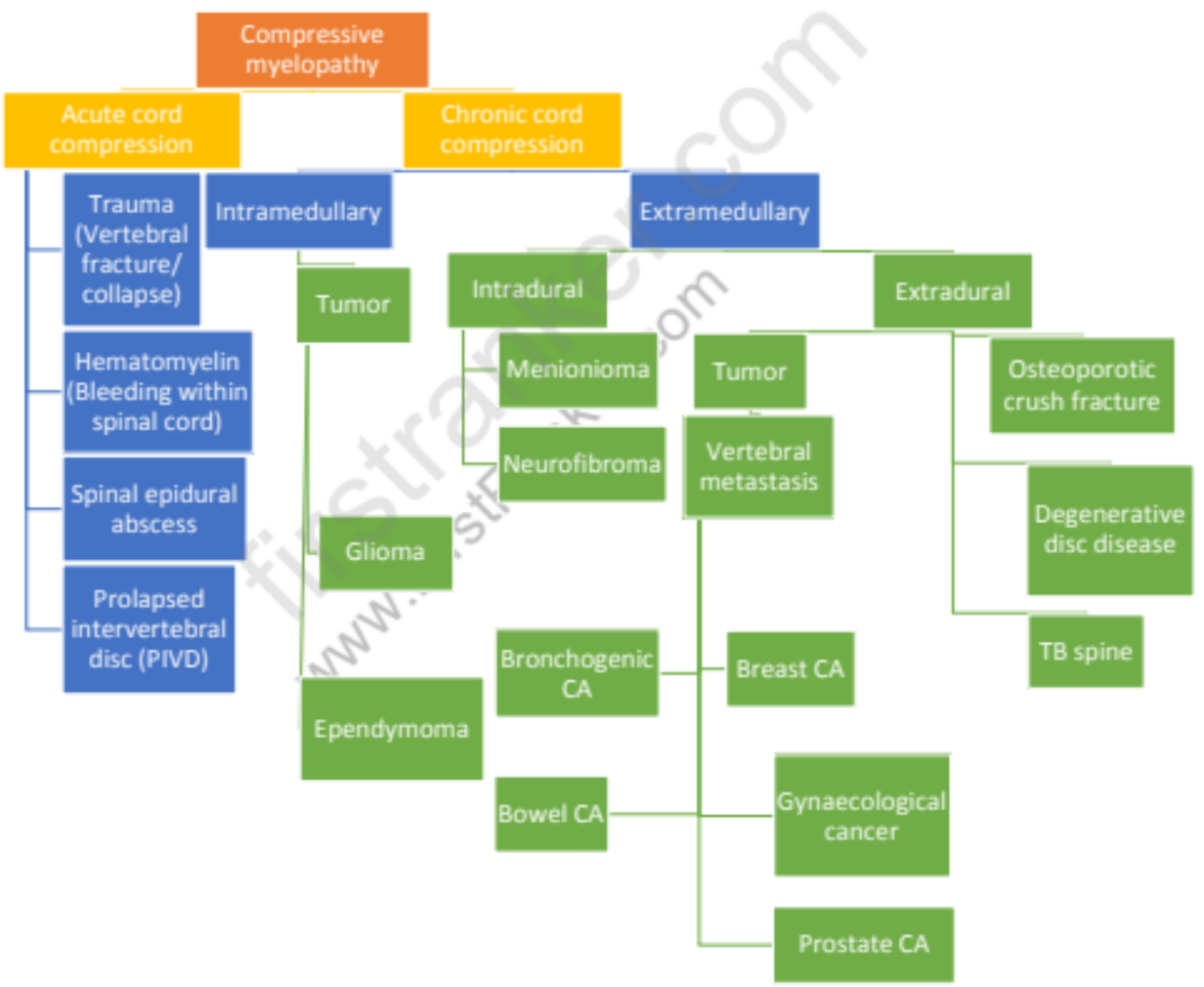
Compression of different structures within the spinal cord.

Structures which gets compressed in compressive myelopathy:

1. Anterior horn cell (AHC)

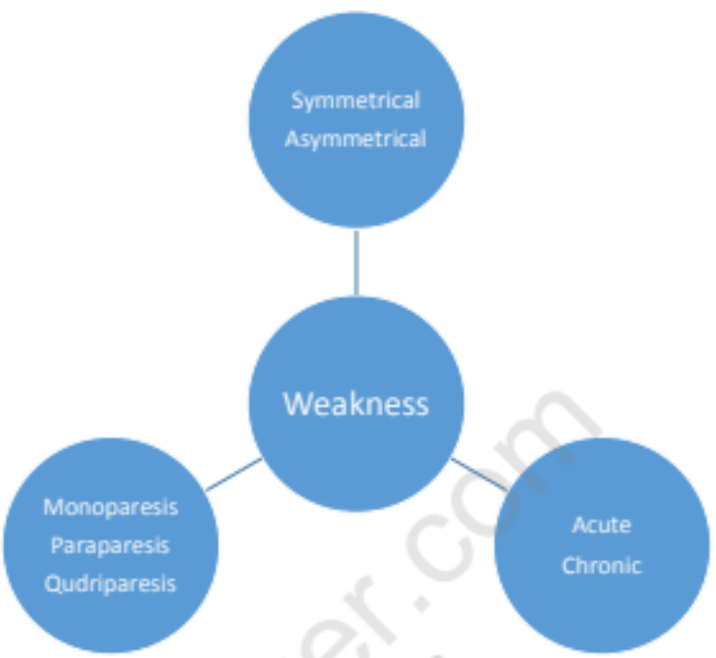
2. Corticospinal tract (CST)
3. Spinothalamic tract: Lateral and anterior
4. Dorsal column
5. Dorsal root
6. Fibres controlling sphincteric function.

Etiology:

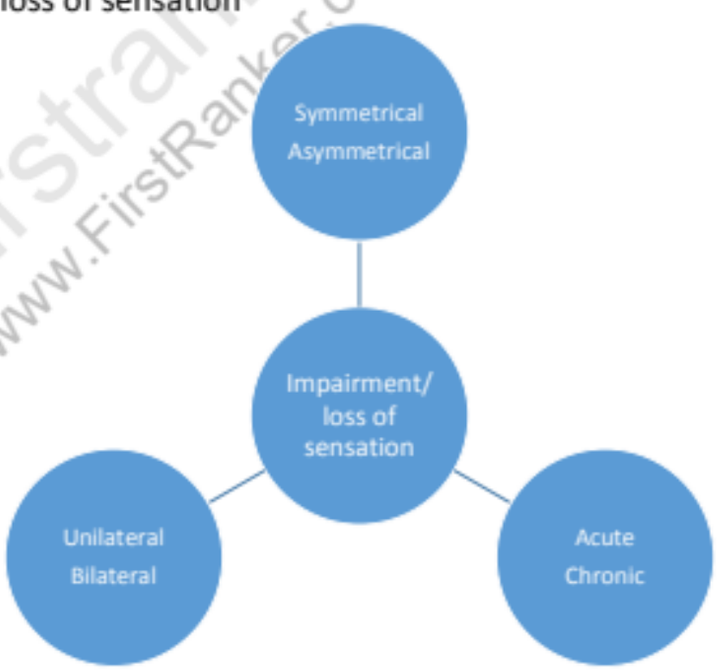


Symptoms:

1. Motor:



2. Sensory:
   a. Impairment/ loss of sensation



Often patient complain of a definite upper level below which sensations are impaired.

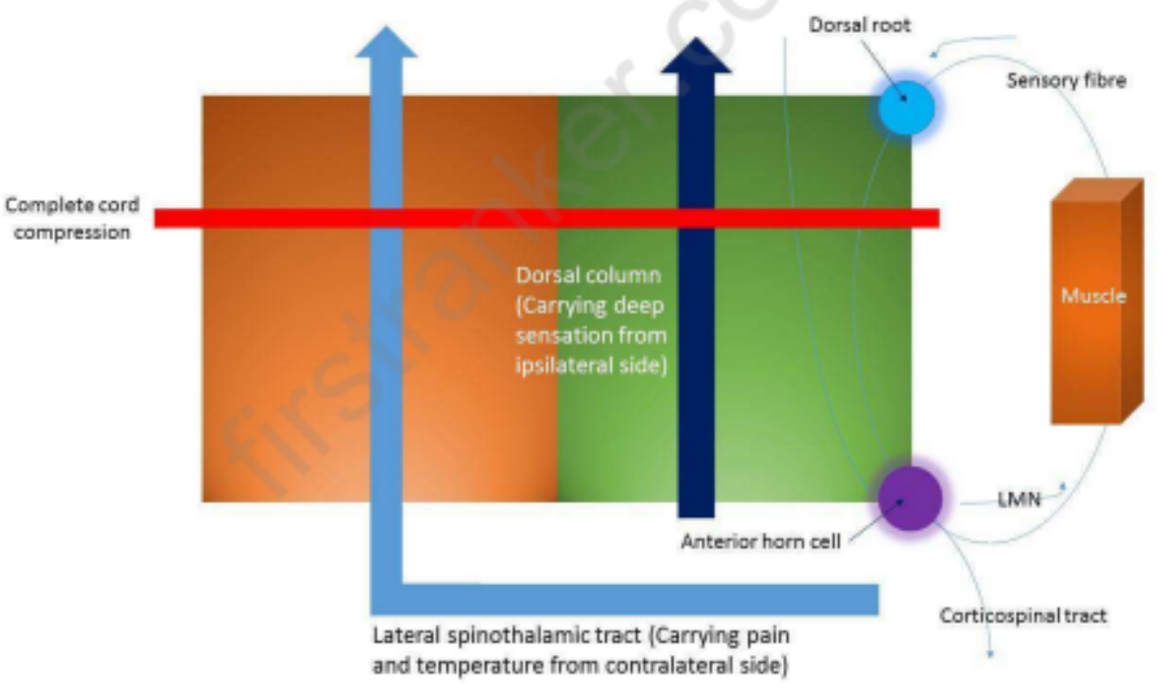
b. Root pain:
   - ✓ Site: In the dermatome of the compressed root
   - ✓ Character: Often severe, deep seated pain, may be aggravated with movements, coughing, sneezing.

c. Girdle sensation:
   A constricting sensation in a dermatome due to irritation of sensory fibres/ posterior root. Classically seen in thoracic cord compression.

3. Sphincteric disturbances:
   a. Bowel dysfunction: Incontinence
   b. Bladder dysfunction: Retention/ incontinence.

Signs:

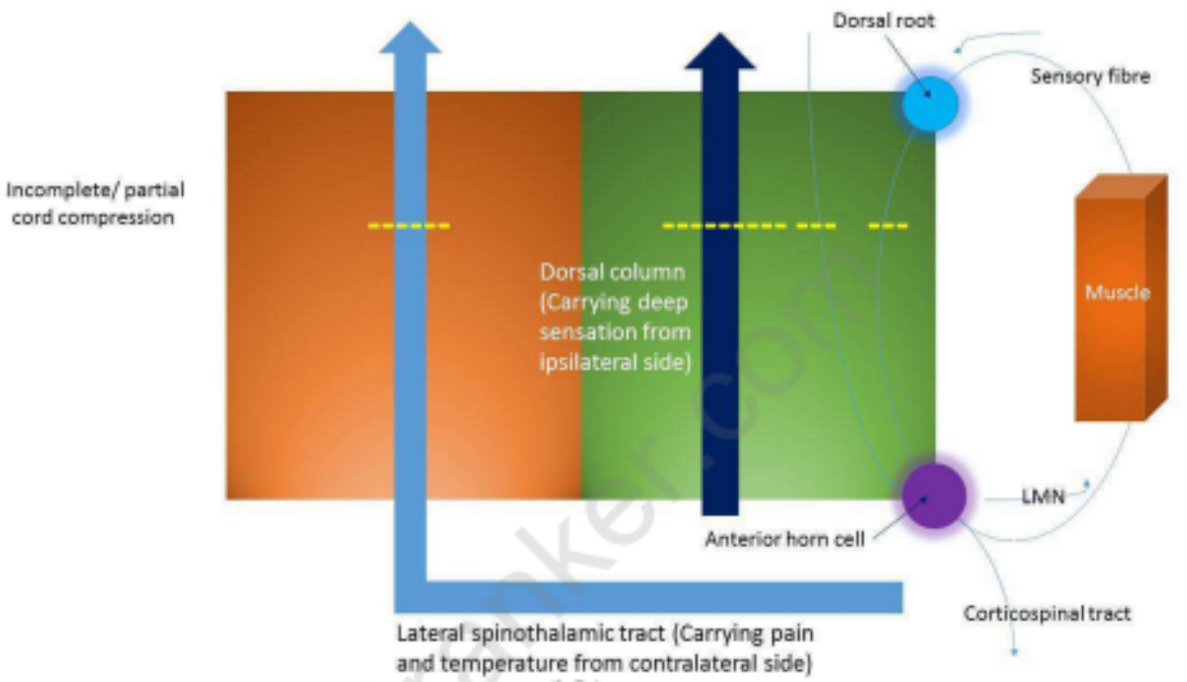
Complete cord compression/ transection



- At the level of compression:
  1. Bilateral LMN lesion (due to damage to AHC)
  2. Impairment of all sensations due to compression of dorsal root/ sensory fibres entering through it.
- Below the level of compression:
  1. Bilateral UMN lesion (due to damage to CST of both side)
  2. Impairment of all sensory modalities (superficial and deep) bilaterally.

- Therefore in complete transection of spinal cord, the upper level of sensory impairment corresponds to the dermatome of compressed segment.

Partial/ incomplete cord compression/ hemisection of spinal cord



- At the level of compression:
  1. Ipsilateral LMN sign (due to damage to AHC)
  2. Impairment of all sensations in the dermatome of compressed segment: Ipsilateral (damage to dorsal root).
- Below the level of compression:
  1. Ipsilateral LMN sign (due to damage to ipsilateral CST)
  2. Impairment of deep sensations of ipsilateral side
  3. Impaired pain and temperature sensation of the contralateral side below the level (due to damage of contralateral LSTT),
- Signs just below the level of compression:
  A zone of hyperalgesia may be present due to irritation of lowest viable segment by compressive lesion.

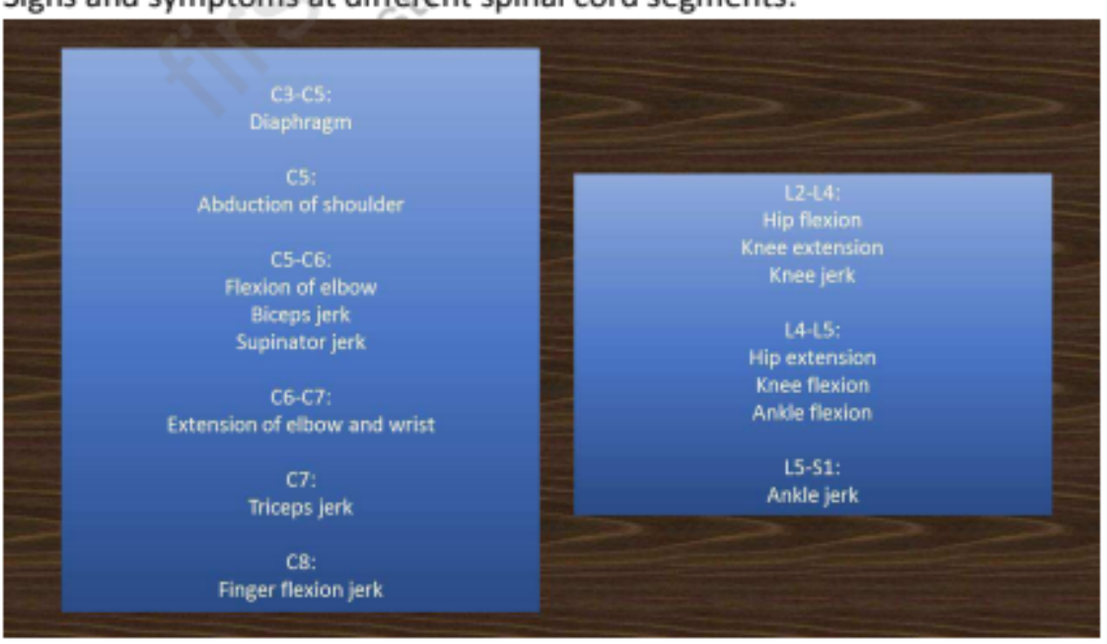
Localization of lesion

A probable localization of the level of spinal cord lesion can be assessed by noting some of the following features:

1. Distribution of weakness:
   - High cervical lesion (C1-C5): Quadriparesis
   - Low cervical lesion (C6-C8): Quadriparesis with proximal part of the upper limb spared
   - Thoracic/ lumber lesion: Paraplegia.
2. Distribution of LMN signs:
   Seen in the same segment.
3. Upper level of sensory loss
4. Vertebral tenderness/ deformity:

Level of vertebral tenderness/ deformity	Related spinal cord segment
Cervical	+1
T1-T6	+2
T7-T9	+3
T10	L1
T11	L2-L3
T12	L4-L5
L1	Sacral segments

5. Signs and symptoms at different spinal cord segments:



Affected nerve root	Signs and symptoms
C1-C4	• Vasomotor and respiratory dysfunction • Diaphragmatic palsy (as phrenic nerve emerges from C3-C5 spinal segments) If bilateral: Paradoxical respiration*.
C5-C6	• Normal shoulder movement • Weakness of elbow flexion • Biceps jerk/ Supinator jerk: Lost • Inverted (Biceps + Supinator) jerk^ may be present • Brisk triceps jerk.
C6-C7	• Weakness of (elbow + wrist) extension • Triceps jerk: Lost • Brisk finger flexion jerk.
C7-C8	Weakness of (wrist + finger) flexion jerk.
Thoracic cord (T1-T8)	• Paraparesis • Usually the upper level of sensory loss lies somewhere on the trunk. Ex: ➢ Nipple line: T4 dermatome ➢ Umbilical line: T10 dermatome.
T10-T12	Umbilicus is pooled upwards on attempting to raise the head as upper abdominal muscle contracts normally but lower abdominal muscle can't. It is called 'Beevor's sign'~.
L2-L4	• Weakness of hip flexion and knee extension • Loss of knee jerk (L2-L4) • Plantar: Bilateral extensor.
L4-L5	• Weakness of hip extension, knee and ankle flexion • Loss of ankle jerk (L5-S1).

[*Paradoxical respiration: It is a life-threatening medical condition that occurs when a segment of the rib cage breaks under extreme stress and becomes detached from the rest of the chest wall. It occurs when multiple adjacent ribs are broken in multiple places, separating a segment, so a part of the chest wall moves independently.

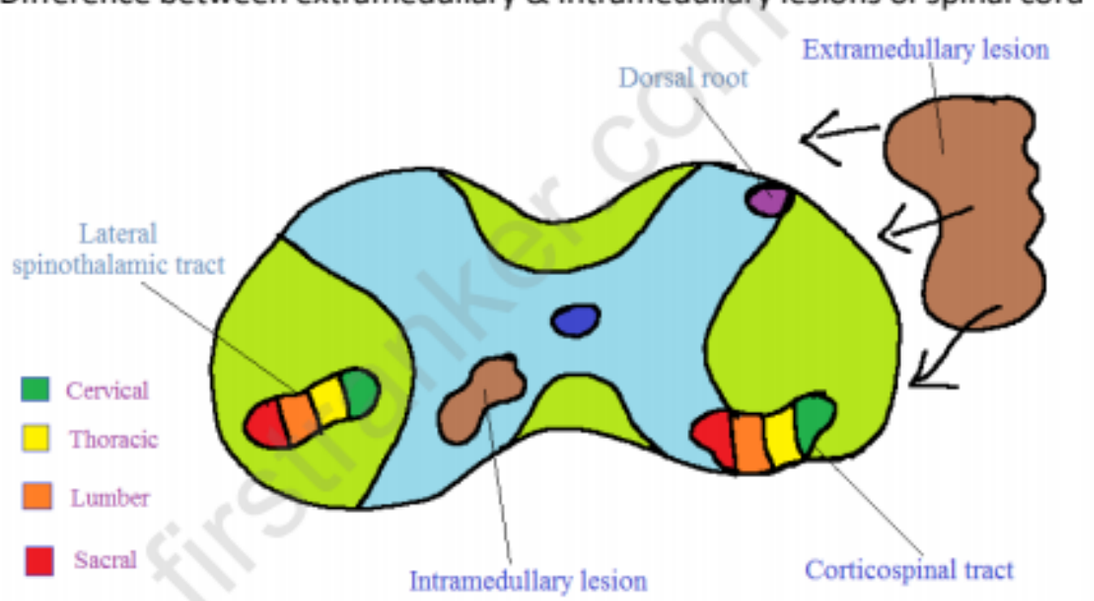
^Inverted biceps jerk: In some cases of compressive myelopathy of C3-C6 segment, a combination of biceps and supinator jerk elicits an elbow extension (C5-C6) and finger flexion response (C7-C8), respectively.

Compression of C5-C6 leads to exaggerated reflex response of C6-C7 and C7-C8 due to UMN lesion. However, in some cases, these segments become so hyperexcitable that attempted biceps and supinator jerk will lead to a response which is normally mediated by these hyperactive segments.

~ Beevor's sign:

The rectus abdominis muscle at the level of the umbilicus is supplied by the T10 nerve roots. Lesions of the spinal cord or roots between T10 and T12 will cause weakness of the lower part of the muscle, and thus a positive Beevor's sign.]

Difference between extramedullary & intramedullary lesions of spinal cord



Features	Extramedullary lesion	Intramedullary lesion
Root pain (Dorsal root compression)	Prominent an early manifestation	Often not prominent/ late manifestation
Pyramidal/ UMN sign (CST damage)	Early manifestation	Late manifestation, often spared
Sacral sensory loss	Early manifestation	Late manifestation
Progression of symptoms	Often rapid	Often slow

Difference between 'paraplegia in extension' & 'paraplegia in flexion'

Features	Paraplegia in extension	Paraplegia in flexion
Attitude of lower limb	Extended	Flexed

Plantar	Extensor	Extensor but may be accompanied by withdrawal response
Pathophysiology	Lesion of pyramidal tract	Lesion of pyramidal tract as well as extrapyramidal fibres

Some important etiology of chronic cord compression

1. Vertebral metastasis:
    - Common primary cancers:
        - ✓ Breast
        - ✓ Bronchogenic
        - ✓ Bowel
        - ✓ Gynaecological
        - ✓ Prostate.
    - Common site of metastasis: Thoraco-lumber vertebrae.
    - Symptoms:
        - ✓ Low back pain is a prominent symptom
        - ✓ Symptoms of primary malignancy may be present.
2. Caries spine/ TB spine/ vertebral TB:
    - It is due to hematogenous spread of bacilli
    - Symptoms of primary TB ***may/ may not*** be present
    - Causes of cord compression:
        - ✓ Collapse of the vertebra
        - ✓ Compression by paravertebral abscess
        - ✓ Tubercular myelitis
        - ✓ Tubercular vasculitis (of spinal cord blood vessels).
    - Vertebral tenderness may be present
    - Local deformity may be present:
        - ✓ Knuckle: prominence of 1 spinous process
        - ✓ Gibbus: prominence of >1 spinous processes.

Investigation of compressive myelopathy

1. Imaging:
    a. X Ray spine (to detect vertebral abnormality)

b. MRI confirms any cord compression, often tell about the underlying cause.

2. Investigations to assess the underlying cause:
   a. Preliminary investigations:
      - ✓ Hb, TC, DC, ESR (↑ in infection)
      - ✓ Na+ K+ Urea creatinine
      - ✓ Liver function tests (Alkaline phosphatase level↑ in metastasis)
      - ✓ Serum calcium (↑ in metastasis)
      - ✓ Chest X Ray.
   b. Special investigations:
      Nature of it depends on the definitive clinical diagnosis.
      Ex: Vertebral metastasis: Histopathology.

Treatment

Supportive

1. Immobilization of the spine (particularly in case of trauma)
2. Absolute bed rest
3. Bed sore prevention (in appropriate cases)
4. Catheterization (if required)
5. Dexamethasone (to reduce spinal cord edema)
6. DVT prophylaxis
7. Exercise and physiotherapy (when safe to start).

Definitive

1. Pharmacological:
   a. TB: Anti-tubercular drug
   b. Spinal abscess: IV antibiotic.
2. Surgical:
   a. Neurosurgical decompression
   b. Orthopedic fixation
   c. Tumor excision.
3. Radiotherapy:
   In case of metastasis.

Dx of cord compression

1. Acute cord compression (in spinal shock stage):
   a. Spinal shock stage of acute transverse myelitis — No H/O trauma
   b. Polio
   c. GB syndrome — No sensory disturbance and no bowel-bladder disturbance

2. Acute cord compression (spasticity has developed):
   Acute transverse myelitis — Do a MRI to confirm

3. Chronic cord compression:
   Other diseases causing paraparesis/ quadriparesis with spasticity may mimic chronic cord compression. These include non-compressive myelopathies:
   a. Motor neurone disease
   b. Multiple sclerosis
   c. Subacute combined degeneration of spinal cord.

- All these diseases have no upper border of sensory loss and bowel-bladder disturbance is not prominent in these diseases.

Non-compressive myelopathy

Acute transverse myelitis

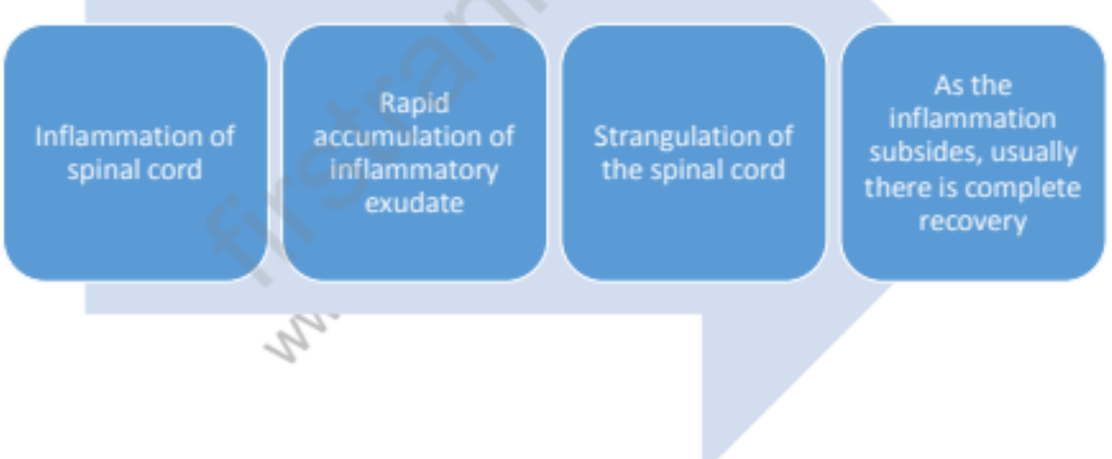
Introduction

It is a sudden onset, rapidly progressive inflammation of the spinal cord, where the inflammatory process can extend transversely as well as longitudinally for 1-2 segments.

Etiology

1. Most likely, the inflammation is an autoimmune response likely to be triggered off by different viruses (HSV. Measles etc.)
2. As a complication of old generation rabies vaccine.

Pathophysiology



Symptoms

1. *Prodromal symptoms:*
   Precedes the paralysis and characterized by fever, malaise, bodyache etc.
2. *Rapidly progressive weakness:*
   a. Usually starts with both lower limbs

b. Upper limbs are often spared as the inflammation commonly occurs in the thoracic segment
c. Rapidly progressive sensory impairment with a definite upper level above which sensations are normal.

3. *Prominent sphincteric disturbance:*
   a. Acute retention of urine
   b. Fecal incontinence.

- With proper treatment, symptoms start to improve after few days, leading to complete recovery, in some patients, a degree of residual weakness may persist.

Signs

1. Spinal shock stage:
   - Power: ↓
   - Tone: ↓ (Flaccid)
   - Deep tendon reflex (DTR): ↓/-
   - Plantar: Bilateral extensor (Up going: ↑↑)
   - Wasting/ fasciculation: Absent
   - Sensory: Impairment/ loss of all sensation with a definite upper level.
2. Stage of paralysis:
   - Power: ↓
   - Tone: ↑ (Spastic)
   - DTR: ↑↑ (clonus may be present)
   - Plantar: ↑↑
   - Sensory: Impaired.
3. Stage of recovery:
   - Power and sensation gradually start to return
   - UMN signs and sensory signs gradually disappears.

Investigations

1. MRI of spinal cord
2. Lumber puncture:
   a. WBC count (slightly ↑)
   b. CSF protein (Normal/ slightly ↑)

c. **NO** classical albuminocytological dissociation.

3. Routine investigations:
   Hb, TC, DC, CRP, Na+, K+, Urea, Creatinine.

Treatment

1. Supportive:
   A. Absolute bed rest
   B. Bed sore prevention
   C. Catheterization
   D. DVT prophylaxis
   E. Exercise, physiotherapy and walking aids.
2. Definitive:
   Systemic corticosteroid.

Dx of ATM

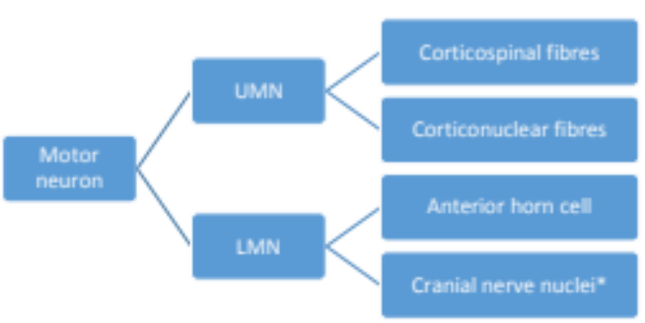
1. Acute compressive myelopathy in shock stage (H/O trauma is present)
2. GB/ Polio: No H/O sensory loss/ bowel-bladder disturbance
3. Compressive myelopathy (with spastic signs): Shows exactly same feature
4. Acute lesion in spinal cord (non-compressive): Ant. Spinal artery occlusion/ thrombosis.

Motor neuron disease (MND)

Introduction

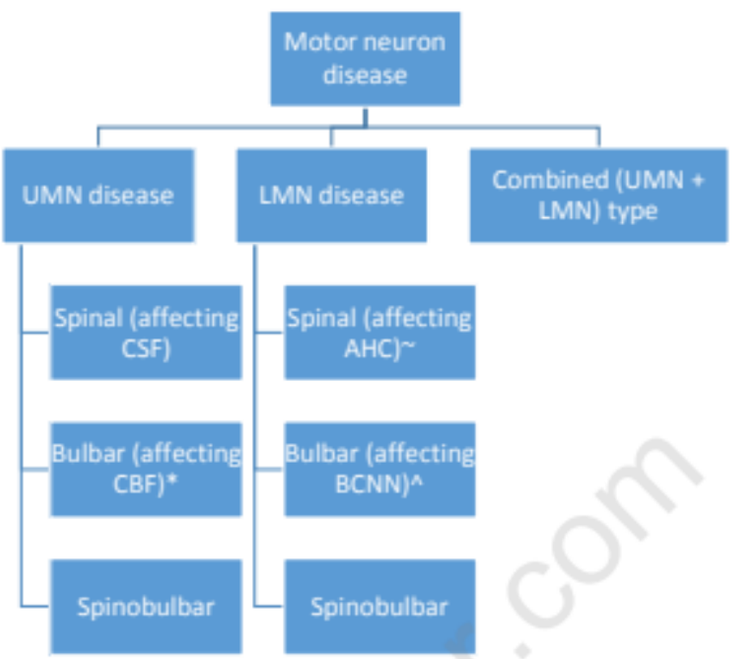
It is a chronic degenerative disease affecting motor neurons.

Affected motor neurons



*Predominantly bulbar cranial nerve nuclei are affected in MND.

Spectrum of motor neuron diseases



[CSF: Corticospinal fibres, CBF: Corticobulbar fibres, AHC: Anterior horn cell, BCNN: Bulbar cranial nerve nuclei]

Examples:

*Pseudobulbar palsy, ^True/ progressive bulbar palsy, ~Spinal muscular atrophy.

Note (incorporate with the picture below in signs and symptoms of AML):

In early stages of MND, CSF are damaged first in the upper limbs, leading to predominance of UMN signs. When it progresses, it gradually involves AHC, leading to mixed (UMN + LMN) signs. At the late stages, it progresses to predominance of LMN signs.

When we try to incorporate lower limbs in this picture, they show predominance of UMN signs until the late stages, when they also begin to show LMN signs.

Amyotrophic lateral sclerosis (ALS)

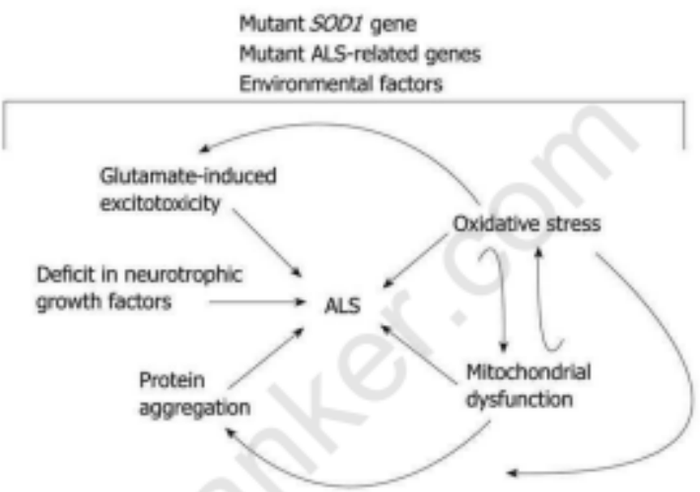
Introduction

It is the commonest variety of motor neuron disease characterized by combined (UMN + LMN) degeneration.

Etiopathogenesis

Exact cause is unclear, however there are few hypothesis:

1. Mutation of superoxide dismutase 1 (SOD1) gene leading to accumulation of abnormal proteins causing cytotoxic cell damage
2. Abnormal accumulation of excitatory neurotransmitter glutamate leading to cell apoptosis
3. Mutation of VEGF gene also plays some role.



Symptoms

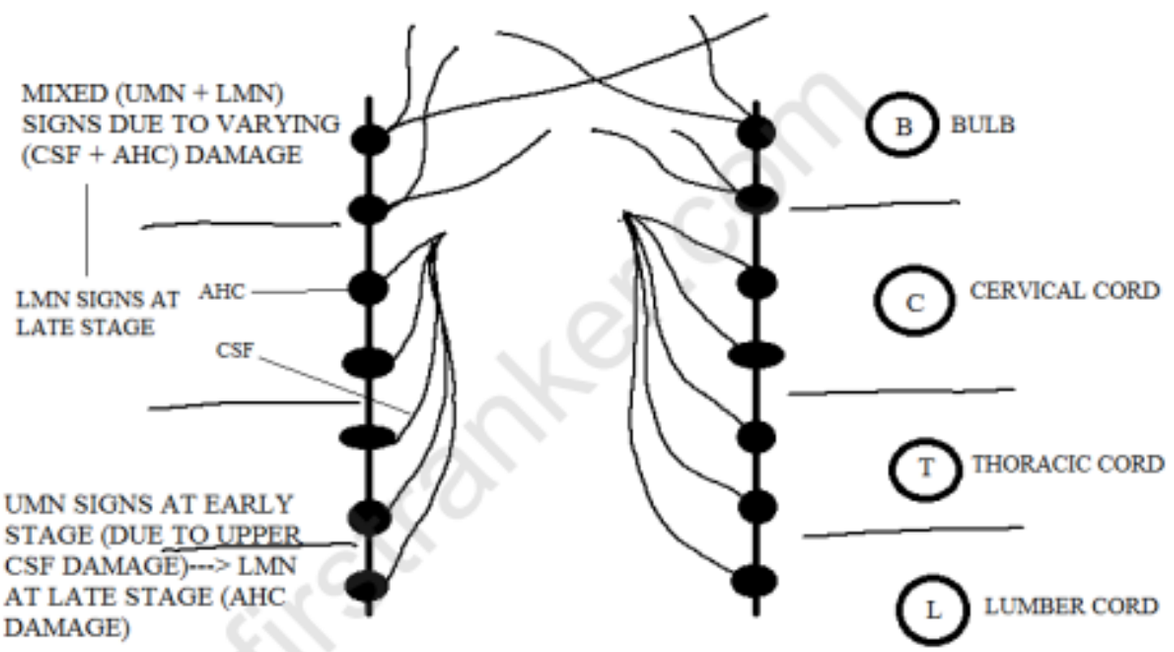
1. Bulbar:
   - ✓ Dysphagia
   - ✓ Dysarthria
   - ✓ Dysphonia
   - ✓ Nasal intonation of voice
   - ✓ Nasal regurgitation of food
   - ✓ Recurrent episodes of chocking/ aspiration leading to aspiration pneumonia.

   Tongue:
   - ✓ Tone: Spastic
   - ✓ Wasting and fasciculation: May be present
   - ✓ Pharyngeal reflex: Lost
   - ✓ Gag reflex: May/ may not be brisk

- ✓ Patient may be emotionally labile, leading to spontaneous spells of laughing/ crying (due to pseudobulbar palsy).

2. Spine:
   - ✓ Slowly progressive para/quadri-paresis
   - ✓ Wasting: Particularly of small muscles of hand, often very prominent
   - ✓ No sensory disturbances
   - ✓ No sphincteric disturbances.

Signs



1. Upper limb:
   - ✓ Combined (UMN + LMN) signs
   - ✓ Power: ↓
   - ✓ Tone: Spastic (due to UMN type of damage), but some muscles may be flaccid (due to LMN type of damage)
   - ✓ Wasting: Some muscles may be wasted
   - ✓ Fasciculation: May/ may not be present
   - ✓ Reflexes: Brisk/ absent in varying combinations.
2. Lower limb:
   - ✓ Usually spastic weakness (due to predominance of UMN signs)
   - ✓ Jerk: Brisk

- ✓ Wasting and fasciculation: Absent
- ✓ Plantar: Bilateral extensor
- ✓ As the disease progresses, lower limb will also show superadded LMN signs.

Functions which will remain unaffected in ALS:

1. Higher function
2. Function of extraocular muscles
3. ***Sensory function***
4. ***Sphincteric function normal*** (as nucleus of Onuf is usually spared).

Complications of ALS

1. Respiratory paralysis, leading to type respiratory failure
2. Recurrent episodes of chocking/ aspiration, leading to recurrent episodes of aspiration pneumonia.

Investigations

1. Nerve conduction velocity (NCV)/ Electromyography (EMG)
2. MRI spine: To rule out any structural compression of spinal cord.

Treatment

Supportive

B. Bed sore prevention
C. Catheterization (if required)
D. Diet:
   Maintenance of nutrition may be difficult due to swallowing difficulty, often patient requires *feeding jejunostomy* or *PEG* (Percutaneous endoscopic gastrostomy) *tube*.
E. Exercise and physiotherapy
F. Long term ventilatory support.

Definitive

Riluzole (Glutamate synthase inhibitor)

Dx of ALS

Patient with quadriparesis:

Cervical compressive myelopathy:
Here (sensory + sphincteric) disturbances are present

Patient with wasting of small muscles of hand:

1. Lower cord compression (C7-C8)
2. Plexopathy
3. Peripheral neuropathy
4. Pancoast's tumor (C8-T1 compression).

Cerebrovascular accident (CVA)

Part 1: Transient ischemic attack (TIA)

Introduction

TIA is characterized by ***reversible*** focal neurodeficit lasting usually <24 hours and is due to reversible occlusion of cerebral artery.

Risk factors

- Thrombosis (artery to artery embolism):
  The main cause is cerebral atherosclerosis resulting from:
  A. Abdominal obesity
  B. BP↑
  C. Cigarette smoking
  D. Diabetes/ Dyslipidemia
  E. Exercise↓.
- Cardioembolism:
  The causes are:
  1. *Atrial fibrillation*
  2. Mitral stenosis ± AF
  3. Infective endocarditis
  4. AMI (Mural thrombus)
  5. Paradoxical embolism (Dislodged deep vein thrombosis in a patient of patent foramen ovale, VSD, ASD etc.)

Clinical features of TIA

Clinically, TIA may be classified into 2 groups, each of which has some distinct clinical features, which may appear in varying combinations involving the particular areas affected:

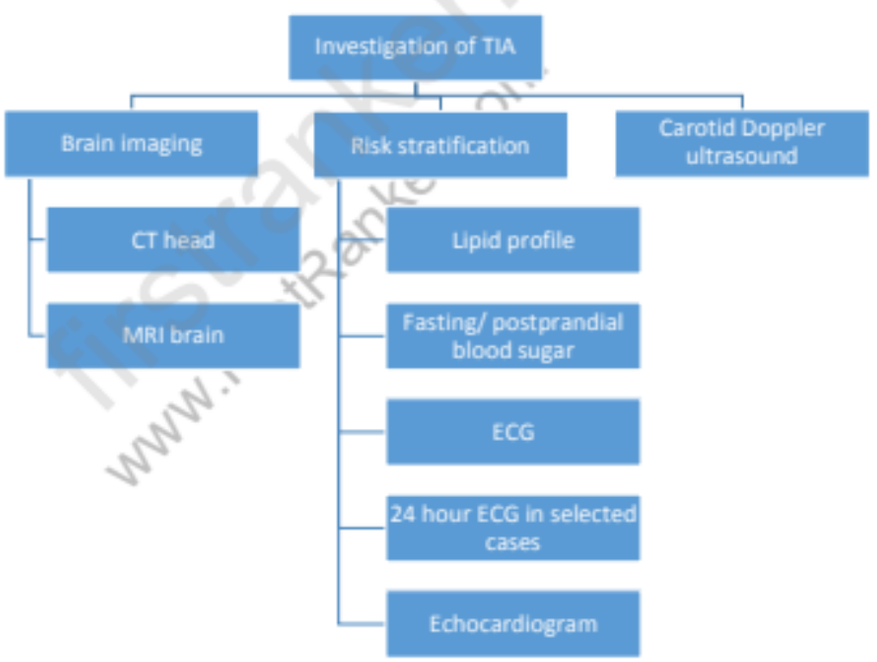
Carotid TIA	Vertebro-basilar TIA
Contralateral hemiplegia (CST)	Diplopia (CN 3,4,6)
Contralateral hemisensory loss (LSTT)	Dysarthria/ Dysphagia (Bulbar nuclei)
Contralateral homonymous hemianopia (Optic radiation)	Dys-equilibrium (Cerebellum)

Aphasia (Speech area)	Drop attack (sudden fall due to weakness and loss of tone of lower limb)
Amaurosis fugans (Transient loss of vision when the patient feels as if a curtain is coming down in front of the eye; cause: retinal artery occlusion)	Vertigo and vomiting
	Contralateral hemiparesis (CST)
	Contralateral hemisensory loss (LSTT)

All the patients of TIA should be assessed for:

1. Atherosclerotic risk stratification
2. Atrial fibrillation
3. Cardiac murmur
4. Carotid bruit.

Investigation



Treatment

1. Risk factor modification (write as per ABCDE in risk factor)
2. Pharmacotherapy:
    - Antiplatelet (Aspirin + Dipyridamol or Clopidogrel)
    - Anticoagulant (Long term Warfarin in case of cardiac embolism)

- Antilipidemic drug
- Antihypertensive drug
- Antidiabetic drug.

3. Surgical treatment:
   Carotid end-arterectomy
   Indication:
   >70% stenosis of internal carotid artery.

ABCD2 scoring and its application

This scoring system acts as a future stroke risk predictor in a patient of TIA:

A. Age> 60 years (1)
B. SBP> 140 mm Hg OR DBP> 90 mm Hg (1)
C. Clinical features:
   - ✓ Weakness ± Speech disturbance (2)
   - ✓ Only speech disturbance (1)

D1. Duration:

- ✓ >1 hour (2)
- ✓ 10-59 min (1)

D2. Diabetic (1).

Scoring	Implication
0-2	Low risk (does not require hospitalization)
3-5	Moderate risk (Hospitalization may be considered)
6-7	High risk (Hospitalization recommended)

PART 2: Stroke

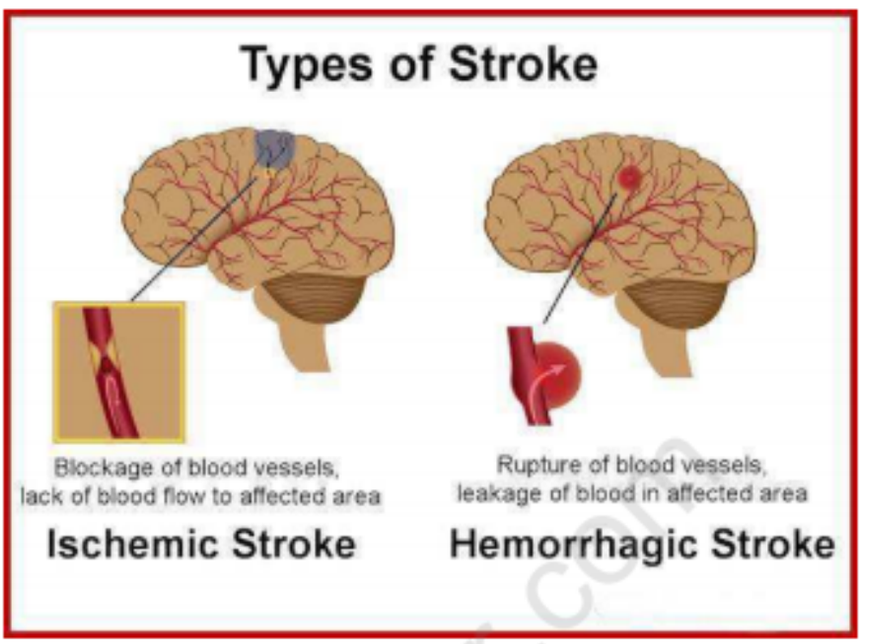
Introduction

Abrupt onset focal neurodeficit lasting for >24 hours due to a vascular event.

Type

1. Ischemic stroke:
   a. Thrombotic stroke
   b. Embolic stroke

2. Hemorrhagic stroke



Most common risk factors

1. Thrombotic stroke: Atherosclerosis
2. Embolic stroke: AF
3. Hemorrhagic stroke: Hypertension

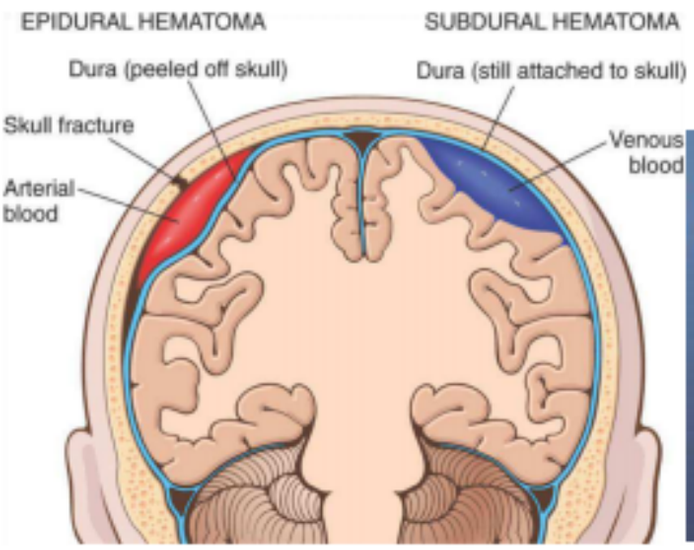
Hemorrhagic stroke

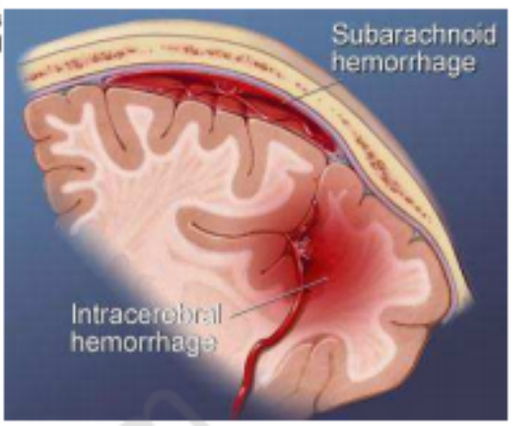
Risk factors:

- Arterial hypertension
- Aneurysm
- Arterio-venous malformation
- Side effects of antiplatelet/ anticoagulant therapy
- Accidental (head injury).

Locations of hemorrhage:

1. Epidural
2. Subdural
3. Subarachnoid
4. Intracerebral/ intraparenchymal.





Mechanism of clinical manifestations:

1. Due to damage to corticospinal and corticonuclear fibres innervating lower ½ of 7th cranial nerve nucleus
2. Damage to other structures: depending upon the site of lesion
   - These manifestations help us to ***localize*** the site of lesion
3. Due to underlying etiology/ risk factors
4. Due to raised ICT (resulting from compressive effect of blood/ intracerebral edema).

Case presentation of CVA

Note that, in long case of CVA, the type and localization of lesion has to be made roughly, and points have to be presented supporting your estimation.

History and examination should include thorough cerebrovascular risk assessment.

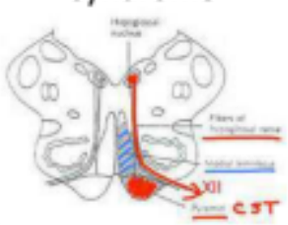
Clinical features which may help us to get an idea about the underlying type of stroke are as follows:

Features	Thrombotic stroke	Embolic stroke	Hemorrhagic stroke
Onset and evaluation	Sudden. At times, it progresses over few hours affecting one part after another	Sudden in onset Simultaneously affects all the parts	

Preceding symptoms	H/O a preceding TIA may be present		-
Associated symptoms	Rare		May be present (due to sudden rise of ICT)
History	H/O angina/ claudication may be present	H/O palpitation/ syncope may be present	H/O hypertension (poorly controlled) may be present
Pulse	-	Irregular pulse (if AF is present)	-
Murmur	-	Murmur of MS may be present	-

Clinical manifestations and localization of site of lesion

Site of lesion	Clinical features
Cortex	1. Contralateral hemiplegia (CST damage) 2. Contralateral lower facial weakness (CN7 palsy): Asymmetrical weakness/ monoparesis/ facial weakness only 3. Contralateral sensory loss 4. Contralateral homonymous hemianopia (Optic radiation damage) 5. Aphasia: a. Fluent (Wernicke's) b. Non-fluent (Broca's) • Usually seen in right sided lesion (as speech area is usually right sided) 6. Visuospatial neglect (non-dominant parietal lobe lesion): As the right lobe is dominant in majority of people, there is unawareness of the left side.
Internal capsule	1. Contralateral hemiplegia 2. Contralateral lower facial weakness.
Midbrain	1. Contralateral hemiplegia 2. Contralateral lower facial weakness 3. Ipsilateral CN3 palsy 4. Ipsilateral CN4 palsy (1+3= Weber's syndrome)

Pons	1. Contralateral hemiplegia 2. *Ipsilateral LMN type of CN7 palsy* 3. Ipsilateral CN6 palsy 4. Loss of touch sensation from ipsilateral ½ of face (Principal sensory nucleus of CN5 damage) 5. Ipsilateral Horner's syndrome (Sympathetic trunk damage) 6. Loss of pain and temperature sensation from contralateral ½ of body (LSTT damage)
Medulla	
Medial medullary syndrome 	1. Contralateral hemiplegia 2. Ipsilateral LMN type of CN12 palsy
Lateral medullary syndrome	1. Ipsilateral LMN type of CN 9, 10, 11 palsy 2. Loss of pain and temperature sensation from ipsilateral ½ of face (spinal nucleus of CN5 damage) 3. Ipsilateral Horner's syndrome (Sympathetic trunk damage) 4. Loss of pain and temperature sensation from contralateral ½ of body (LSTT damage) 5. Vertigo, vomiting, nystagmus (vestibular nucleus damage)

Common sites of hyperetnsive hemorrhage:

Site	Clinical features
Basal ganglia	Contralateral hemiplegia ± Contralateral lower facial weakness
Thalamus	• Contralateral hemiplegia ± Contralateral lower facial weakness • Contralateral sensory loss • Contralateral homonymous hemianopia • Conjugate movement abnormality • Chronic pain of the affected limbs.
Cerebellum	• Nystagmus

	• Ataxia • Incoordination of movement • Vertigo/ vomiting.
↑ICT	Dropping level of consciousness (low GCS score)

Investigation of stroke

1. Imaging
2. Risk factor diagnosis
3. Routine investigation
4. Special investigation

Imaging

1. CT head
2. MRI (in selected cases)
3. Cerebral angiography (in selected cases):
   a. MR angiogram
   b. CT angiogram

Risk factor diagnosis

1. Blood sugar (Fasting + Post-prandial)
2. Fasting lipid profile
3. ECG
4. Echocardiogram
5. 24 hour ECG monitoring (in selected cases: suspected paroxysmal arrhythmia)
6. Carotid Doppler ultrasound (in selected cases).

Routine investigation

1. Blood: Hb, TC, DC, ESR
2. Renal function: Na, K, Urea, Creatinine
3. Coagulation profile: BT, CT, PT, aPTT
4. Chest X Ray

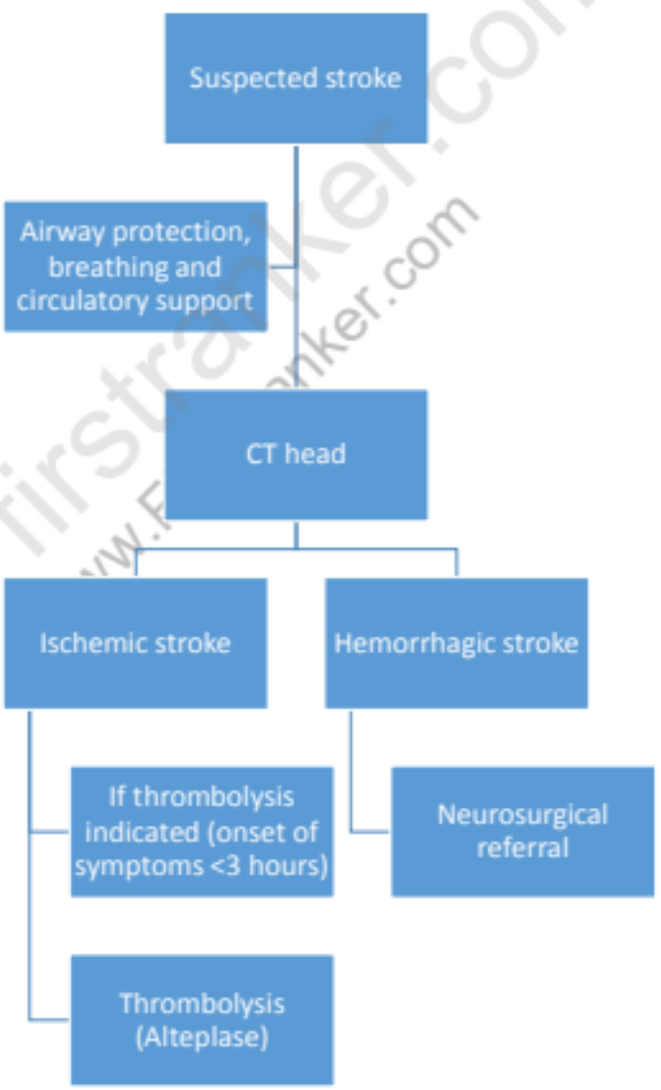
Special investigation

1. If vasculitis is suspected: Vasculitis screen (for detection of autoantibodies)
2. If hypercoagulable disorder is suspected: Tests to rule out hypercoagulable disorders
3. Id cerebral venous thrombosis is suspected: MR venogram.

Treatment

1. Immediate treatment
2. Risk factor modification
3. Supportive treatment.

Immediate treatment



Risk factor modification

1. Lifestyle modification:
   Control of ABCDE.
2. Pharmacotherapy:
   - Antiplatelet drugs (to prevent thrombus formation):
     - ✓ Aspirin 300 mg for 14 days
     - ✓ Clopidogrel for rest of the life.
   - Anticoagulant agents (in case of cardio-embolism):
     - ✓ Warfarin
   - Antihypertensive agents (during acute period):
     During acute period of stroke, BP should not be quickly reduced because rapid decline of BP may jeopardize cerebral autoregulation and may cause irreversible damage. So, this period is called "*ischemic penumbral zone*". During this period, the target BP should be (140-150) / (80-90).
     Long term target BP should be <140/80.
   - Antidiabetic drugs
   - Antilipidemic drugs:
     - ✓ Atorvastatin.

Supportive treatment

A. Airway protection (± oropharyngeal suction ± intubation, if required)
   Absolute bed rest
B. Breathing support (oxygen ± invasive ventilation)
   Bed sore prevention (frequent change of posture/ air or water mattress)
C. Circulatory support (IV fluid, if required)
   Catheterization, if required
   Cerebroprotective agents (Piracetam/ Citicoline): Have doubtful role
D. Diet:
   - Oral feeding (if safe)
   - Ryle's tube feeding
   - Long term feeding:
     - ✓ Feeding jejunostomy
     - ✓ Percutaneous endoscopic gastrostomy tube

Drugs:

- Anti-edema measures (IV mannitol followed by oral glycerol)
- Anticonvulsant (if there is post stroke seizure)
- Antibiotic (if any focal infection is suspected)

E. Exercise (Physiotherapy)

Differential diagnosis of stroke

1. *Dx of acute hemiparesis:*
   a. CVA/TIA
   b. Post-seizure hemiparesis (Todd's paralysis): Temporary manifestation
   c. Cerebral venous sinus thrombosis
   d. Multiple sclerosis.
2. *Dx of causes of stroke:*
   a. Ischemic
   b. Hemorrhagic.

Oxford classification of stroke

There are 4 categories in Oxford classification of stroke, which depends upon the presence of 1 or more of the following parameters:

1. Contralateral hemiplegia/ Contralateral hemisensory loss involving: Upper limb ± Lower limb ± Lower half of face
2. Contralateral homonymous hemianopia
3. Higher cortex dysfunction: Aphasia/ Visuospatial neglect
4. Cerebellar signs/ brainstem sign/ loss of consciousness/ isolated homonymous hemianopia

- 1+2+3: Total anterior circulation stroke (TACS)
- Any 2 of (1+2+3): Partial anterior circulation stroke (PACS)
- Only 1: Lacunar anterior circulation stroke (LACS)
- Any 1 component of 4: Posterior circulation stroke (POCS)

SN: Lacunar infarct

It is a *small vessel stroke* typically due to occlusion of a *deep penetrating branch* of a cerebral artery.

Risk factor: Athero-thrombotic occlusion

Clinical features:

1. Contralateral weakness: Upper limb/ lower limb/ lower half of face
2. Contralateral hemisensory loss.

Investigation:

1. Imaging
2. Risk factor stratification.

Treatment:

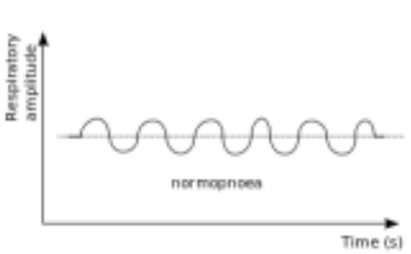
1. Immediate
2. Risk factor stratification
3. Supportive treatment
   - Same as general management of stroke.

Autopsy findings:

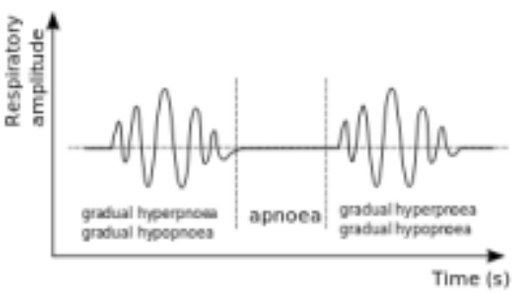
On autopsy, the affected region of the brain looks like a lake (small area filled with fluid), therefore the name "lacunar" is given.

SN: Brainstem signs

1. Contralateral weakness and contralateral hemisensory loss (LSTT damage)
2. Motor cranial nerve palsy
3. Vital center dysfunction:
   a. Respiration:
      - Respiratory depression
      - Cheyne Stoke's breathing (apnea followed by hyperpnoea)



Normal respiration



Cheyne-Stokes respiration
- Periodic breathing:
  Gradual hyperpnoea/hypopnoea and Apnoea
- Sleep/Hypoxemia/Drugs
- Hypoperfusion of the brain (respiratory center)

b. BP: Erratic
c. Pulse: Erratic

4. Loss of consciousness (Low GCS):

   Ocular abnormality:

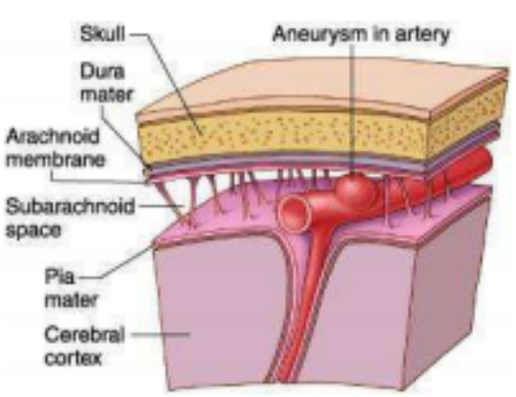
   I. Loss of Doll's eye/ Oculocephaic reflex
   II. Persistent conjugate deviation towards one side
   III. Pupillary abnormality:
      - Bilateral dilated non-reactive pupils (in brainstem death)
      - Pin point pupil (in pontine hemorrhage).

Subarachnoid hemorrhage (SAH)

Bleeding inside subarachnoid space.

Etiology

1. Rupture of a saccular aneurysm (Berry's aneurysm)
2. Intraventricular extension of intraparenchymal bleed.



Risk factors

A. Arterial hypertension
   AV malformation
   Anticoagulant/ antiplatelet therapy
   Aneurysm rupture
B. Bleeding disorder

Clinical features

1. Headache
   - Onset: Sudden
   - Site: Often occipital (due to vertebro-basillar artery aneurysm rupture)/ neck pain but may be generalized
   - Nature: Usually severe, often described by the patient as "worst headache of my life"
   - Some extra manifestations may be present due to meningeal irritation:
     a. Neck stiffness
     b. Photophobia.
2. Focal neurodeficit:
   - ***Falling level of consciousness (low GCS):*** Due to increased ICT secondary to obstructive hydrocephalus
   - ***Hemiparesis/ Aphasia etc.*** due to cerebral infarction resulting from severe vasospasm: Leaked blood irritates blood vessels
   - ***Neck rigidity*** may be present: Leaked blood irritates meninges
   - ***3rd cranial nerve palsy***: Due to an expanding aneurysm of posterior communicating artery
   - ***Homonymous hemianopia***: Due to an aneurysm of anterior cerebral artery (compression over optic tract).

Investigation

- Supportive/ routine:
  1. Blood: Hb, TC, DC, CRP/ESR
  2. Renal function: Na+ K+ Urea creatinine
  3. Coagulation profile: PT, aPTT

- Definitive:
  1. ***CT Head*** (sensitive to detect hemorrhage): It will show bleeding within sulcus/ cistern

  2. If CT is inconclusive: Do a ***Lumbar puncture***: It will reveal blood stained CSF, which may be of 2 origins and can be differentiated by following:
     - Continuous leak of blood stained CSF: Due to SAH
     - Gradually the blood staining gets clear: Due to traumatic tap.

     CSF RBC count:
     - In case of traumatic tap, there will be a significant drop in RBC count between sample 1 and sample 4
     - In case of SAH, there will be no such drop.

     LP should be avoided in case of suspected raised ICT.

     Xanthochromia:
     Yellowish discoloration of CSF due to presence of bilirubin/ other pigment.
     It usually takes 6-12 hours before Xanthochromia develops. Therefore, ideally LP should be performed before 12 hours of onset of symptoms.

  3. ***Cerebral angiogram***: 4 vessel DSA (Digital subtraction angiography).

Treatment

It has 2 components: supportive and interventional.

Supportive treatment:

A. Absolute bed rest for 48-72 hours (if patient prefers than in a dark room) Analgesic
B. Bowel: Laxatives (to avoid any straining during defecation)
C. Circulatory support: IV fluid (to maintain adequate hydration)
D. Drugs: ***Nimodipine*** (to counteract cerebral vasospasm).

Interventional treatment:

1. Neurosurgical: Clipping of the aneurysm
2. Radiological: Coiling
3. Ventriculostomy (if significant hydrocephalus develops).

Complications

1. Obstructive hydrocephalus
2. Vasospasm
3. Rebleeding
4. Cerebral salt wasting syndrome (hyponatremia).

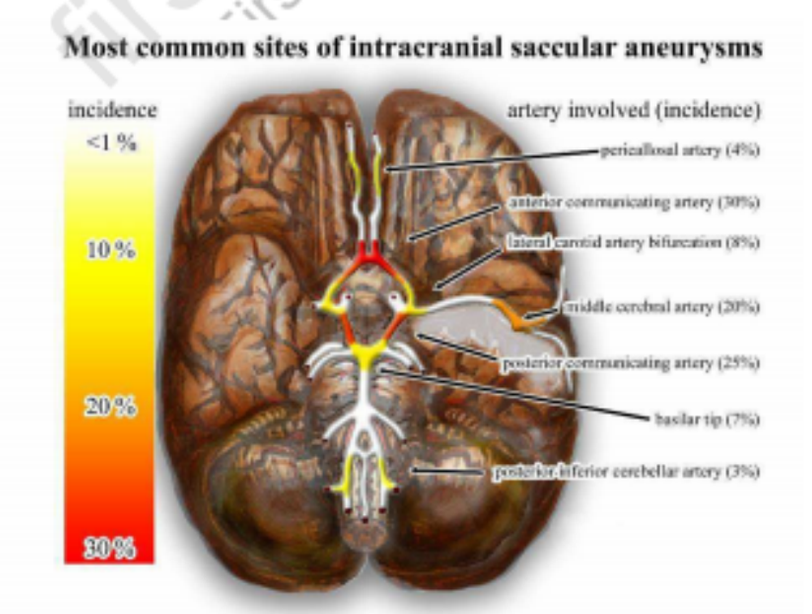
SN: Saccular aneurysm

Outpouching of an arterial wall of cerebral blood vessels usually due to congenital weakness of the wall.

Site:

Can occur anywhere in the Circle of Willis but common sites are:

1. Distal of ICA
2. Bifurcation of MCA
3. Top of Basillar artery.

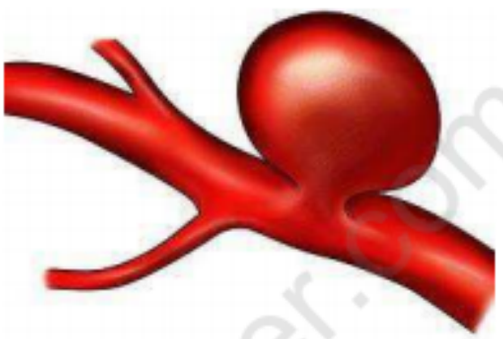


Associations:

1. Coarctation of aorta
2. Polycystic kidney disease

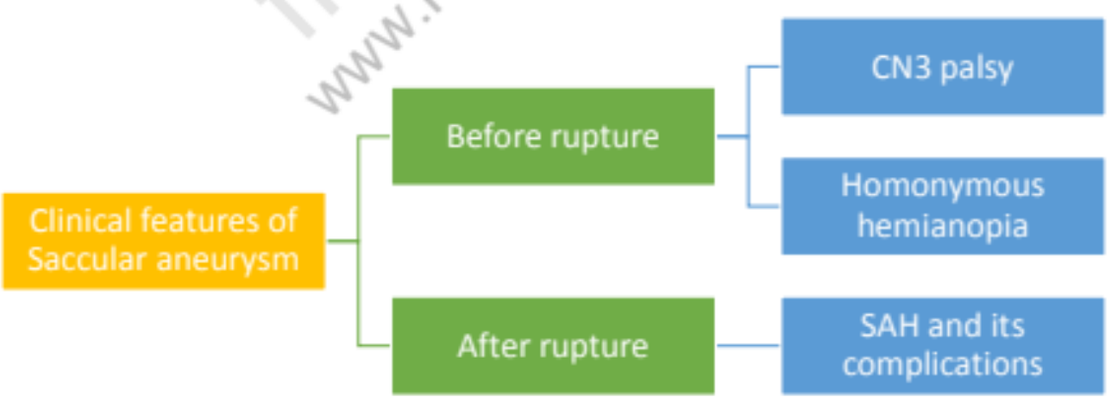
Description:

- A saccular aneurysm has two parts: a neck and an apex.
- Rupture usually occurs at the apex.
- Size of the aneurysm correlates poorly with risk of rupture.



Saccular Aneurysm

Clinical features:



Investigations:

1. CT
2. LP
3. DSA

Treatment:

It has 2 components: supportive and interventional (same as SAH).

Neuropathies

They are of 2 types:

1. Acute (Ex: Guillain-Barré Syndrome)
2. Chronic (Ex: Diabetes)

Guillain-Barré Syndrome (GB syndrome)/ Acute inflammatory demyelinating polyneuropathy (AIDP)

Introduction:

It is a disease characterized by rapidly progressing inflammatory demyelination of multiple, predominantly *motor peripheral nerves* (roots/ radicles may also be damaged).

Etiopathogenesis:

Most likely inflammatory reaction is triggered off by a recent infection: URTI/ GI infection.

Clinical features:

- Preceding history of an URTI (fever, sore throat, cough) or GI infection (diarrhea, abdominal pain etc.) may be present.
- Symptoms:
  1. *Rapidly progressive weakness*: Usually starts with lower limb, then ascends.
  2. Often *proximal weakness is more* than weakness in distal parts.
  3. *Sensory symptoms*: Patient often complains of a deep seated severe pain in the limbs.
  4. *Respiratory distress and drowsiness* due to type 2 respiratory failure (hypoxemia with hypercapnia).
  5. **Sphincteric disturbance is very rare.**
- Signs:
  Motor:
  1. Paraparesis/ Quadriparesis: Flaccid.
  2. Initially proximal weakness> distal weakness.
  3. **Symmetrical weakness usually present.**
  4. Deep tendon reflexes/ Jerk: ↓/ -

5. Planter: Lost/ unresponsive/ flexion (But never extensor)
   Wasting and fasciculation is absent in most of the cases due to rapidity of the progression of disease.

Sensory:

No objective sensory signs.

Cranial nerves:

Bilateral 7th cranial nerve LMN type of palsy may be present.

Autonomic:

1. Tachy/brady-arrhythmia: may be present.
2. Hyper/hypo-tension: may be present.

Paralysis of respiratory muscles:

Monitor by "Single breath: Number counting test".

1. Respiratory distress
2. Type 2 respiratory failure (hypoxemia with hypercapnia)
3. Tachypnea
4. Low GCS (Glasgow coma score): Due to drowsiness resulting from retention of CO_2.

Investigation

1. NCV:
   Confirms the diagnosis. In NCV, two types of lesion are seen:
   a. Demyelinating type
   b. Axonal damage type (bad recovery).
2. CSF/ Lumber puncture:
   a. Protein: Markedly increased
   b. WBC: Mildly increased
      - Therefore, *'albuminocytological dissociation'* is present.
   c. Bedside spirometry:
      FVC↓
3. Special test: Detection of anti GM1 antibody.

Treatment

1. Supportive treatment:
   - Bed rest
   - Bed sore prevention

- Catheterization
- DVT prophylaxis
- Exercise and physiotherapy.

2. Definitive treatment:
   - IV IgG for 5 days
   - Plasmapheresis.

Differential diagnoses

1. Polio
2. Shock stage of acute transverse myelitis
3. Shock stage of acute compressive myelopathy.

Miller Fisher Syndrome

It is an atypical variant of GB.

Clinical features:

All clinical features + ophthalmoplegia + ataxia.

Investigation:

1. NCV
2. LP
3. Detection of Anti Gq1b antibody.

Treatment:

As in GB syndrome.

Polio

Introduction

It is a disease characterized by rapid degeneration of anterior horn cell (AHC).

Causative agent

Poliovirus type 1, 2, 3.

Clinical features

There are 4 types of outcome in a polio patient:

a. Asymptomatic
b. Abortive
c. Non-paralytic polio (neck stiffness + meningeal signs)
d. Paralytic polio.

Course of paralytic polio (in comparison to GB)

1. Prodromal illness:
   - ✓ Fever
   - ✓ Malaise
   - ✓ Neck pain
   - ✓ Neck stiffness
   - ✓ Constipation.

GB: Usually symptoms of a preceding URTI/ GI infection is present

2. Asymptomatic latent period: Absent in polio

GB: May be present

3. Weakness:
   - ✓ Rapidly progressive
   - ✓ Para/quadri-paresis
   - ✓ Proximal weakness is more prominent throughout the course of illness
   - ✓ **Bilaterally asymmetrical involvement**
   - ✓ Tone of affected limb: ↓
   - ✓ Reflex: ↓/-
   - ✓ Plantar: Unresponsive/ flexor
   - ✓ **Fasciculation: Usually present**
   - ✓ **Wasting: Prominent**
   - ✓ Recovery: Very poor
   - ✓ Signs of LMN type bulbar palsy
   - ✓ Autonomic dysfunction: May occur
   - ✓ Respiratory paralysis with failure: May occur
   - ✓ **(Sensory + Sphincteric) disturbance: Absent.**

GB: Usually symmetrical

GB: Same features are seen

GB: Absent

GB: Often good recovery

GB: Facial palsy more common

Investigation

Detection of virus in stool sample

Treatment

- Established case: Supportive
- Primary prevention: Vaccination.

Peripheral neuropathy

Introduction

It is a condition characterized by:

- Mono-neuropathy or
- Poly-neuropathy.

Causes

A	• AIDP* • Alcoholic neuropathy
B	Vitamin B deficiency (B1, B6, B12) Vitamin B6 deficiency is commonly seen in INH therapy.
C	• CIDP^ • Chronic renal failure • Carcinomatous neuropathy (Paraneoplastic syndromes) • Connective tissue disease (vasculitis)
D	• **Diabetes** • Drug induced: ✓ Amiodarone ✓ Anticancer drugs ✓ Anti-retroviral drugs.
E	• Endocrinopathy (Hypothyroidism) • Exogenous toxins (Pb, As etc.)
F	Familial: Hereditary sensory motor neuropathy/ Charcot Merry Tooth disease
G	GB*
H	• HIV • Hansen's disease (Leprosy)

[* Both are same, ^IDP: Inflammatory demyelinating poly-neuropathy]

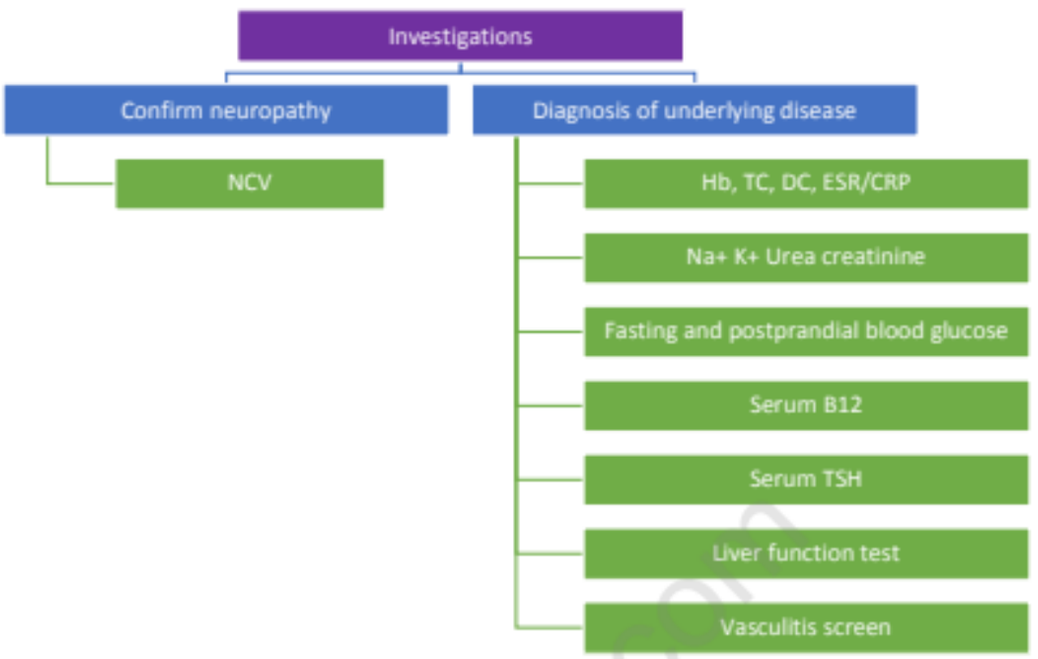
Clinical features

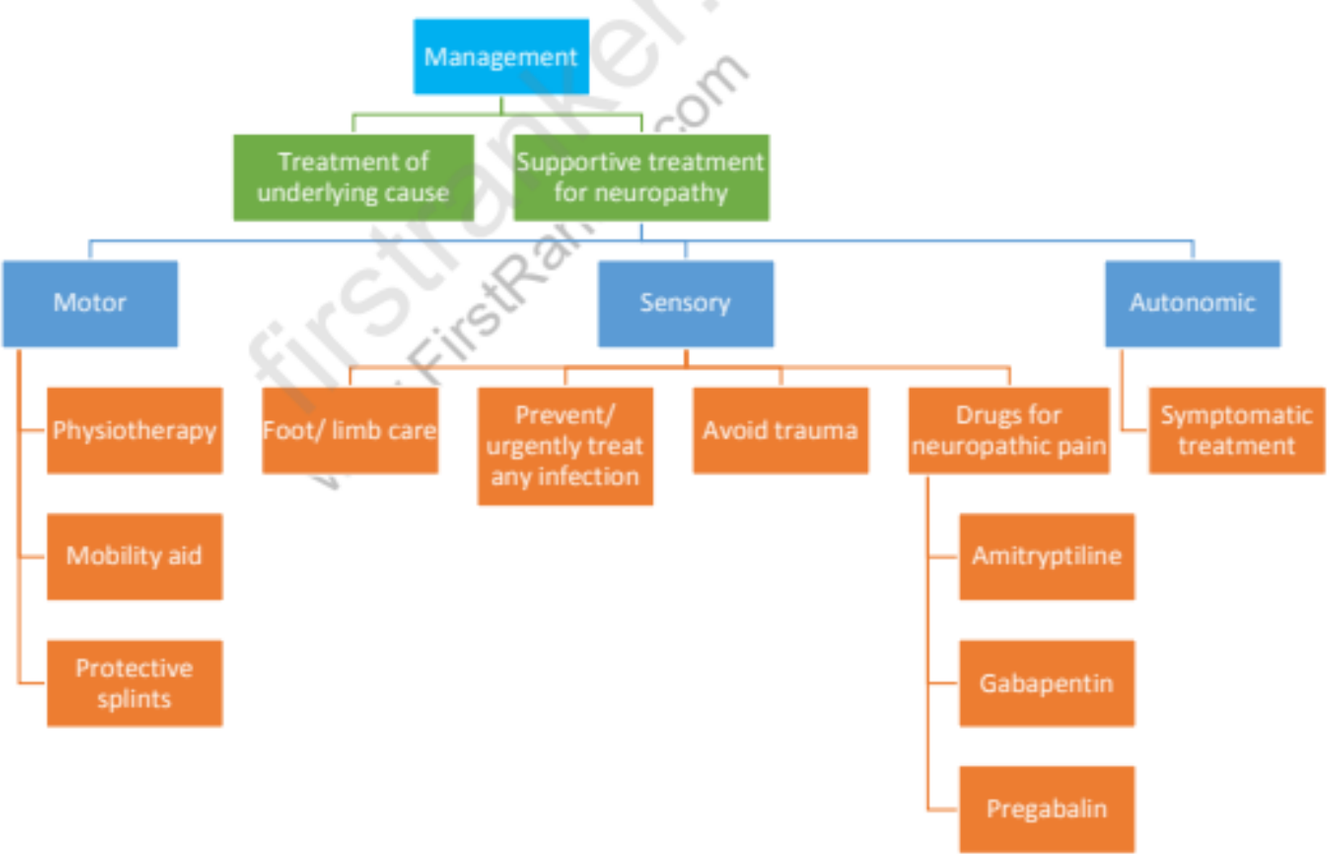
1. Due to the underlying disease
2. Due to neuropathy itself:
   Motor, sensory and autonomic manifestations may occur in an isolated manner/ in varying combinations.

A. Motor:
   - Weakness:
     - ✓ Uni/bi-lateral
     - ✓ Asymmetrical
     - ✓ Patchy in distribution: Involving only that part of the limb which is innervated by the affected nerve
     - ✓ Often starts with distal weakness.
   - LMN signs present in the affected part of the limbs:
     - ✓ Flaccidity
     - ✓ Reflex: ↓/-
     - ✓ Wasting: Present
     - ✓ Fasciculation: Absent
     - ✓ Plantar: Unresponsive/ flexor.

B. Sensory:
   - Sensory impairment:
     - ✓ Paresthesia: Tingling/ pins and needle sensation
     - ✓ In some cases (ex. diabetes): severe burning pain
     - ✓ Unilateral/ bilateral (asymmetrical).
   - Sensory loss:
     - ✓ Patchy in distribution: From the dermatome of the affected nerve
     - ✓ Distal part of limb is affected first (Glav and stocking distribution).
   - Neuropathic ulcer: Often present.

C. Autonomic:
   - Gastro-intestinal:
     Constipation due to hypomotility
   - Genito-urinary:
     Sphincteric disturbance
     Impotence
   - Postural hypotension.





Myasthenia Gravis

Introduction

It is an autoimmune disease characterized by fatigable weakness and is usually due to autoantibody mediated blocking of post-synaptic acetylcholine receptor.

Clinical association

1. Thymoma
2. Grave's disease
3. Addison's disease
4. Pernicious anemia.

Pathophysiology

1. There is auto-antibody mediated blocking of Ach receptor in the post-synaptic membrane resulting in too few functional Ach receptor.
2. Simultaneously, there is exaggeration of physiological rundown of Ach in the pre-synaptic membrane.
    - Both of these are responsible for impaired N-M transmission which gets further compromised on repeated contraction of muscle leading to fatigable weakness.

Clinical features

The type of weakness is fatigable; so ***signs and symptoms become more prominent as the muscle continues to work continuously***.

1. Ocular features:
    a. Drooping
    b. Extra-ocular muscle fatigue: Diplopia
    c. Extra-ocular muscle weakness: External ophthalmoplegia (distribution of weakness does not follow any specific cranial nerve pattern)
    d. Ptosis: Usually fatigable
    e. No pupillary abnormality.
2. Facial weakness
3. Weakness of bulbar muscles
4. Limbs:
    a. Fatigable weakness

b. Tone, reflex, jerk, plantar: Usually normal
c. Wasting, fasciculation: Absent.

5. Respiratory paralysis and type 2 respiratory failure may occur
   - Therefore, the patient should be regularly monitored for the following signs:
     - ✓ Unexplained shortness of breath (SOB)
     - ✓ Tachypnea
     - ✓ Signs of carbon di oxide retention
     - ✓ Single number breath counting test
     - ✓ Bedside spirometry.

Investigation

1. Edrophonium challenge test/ Tensilon test:
   Dramatic improvement of weakness
2. NCV (tests the nerve)/EMG (tests the muscle):
   Detrimental response on repetitive stimulation of muscle
3. Detection of Ach-R antibody
4. Antibody against muscle tyrosine kinase: Predictor of respiratory paralysis
5. CT chest: To look for thymoma.

Treatment

1. Stable stage:
   a. Neostigmine/ Pyridostigmine (anti-cholinesterase)
   b. Corticosteroid
   c. Immunomodulators (Micophenolate mofetil)
2. Myasthenia crisis (acute respiratory failure/ acute limb weakness):
   a. IV IgG/ Plasmapheresis
   b. Ventilatory support
   c. Thymectomy.

SN: Lambert Eaten Myasthenia Syndrome

Introduction:

It is a Paraneoplastic syndrome of small cell lung carcinoma. It is due to anti Ca++ channel antibodies in pre-synaptic membranes.

Clinical features:

Same as myasthenia gravis except:

1. ***Weakness improves with repeated muscle contraction***
2. Areflexia occurs
3. Autonomic disturbances occur.

Diagnosis:

EMG: Incremental response on repeated stimulation

Treatment:

Di-amino-pyridine

Note:

It may manifest even before lung CA appears radiologically. So a patient with LEMS must be periodically screened by X-Ray to detect any obvious mass on the earliest opportunity.

Miscellaneous topics

Subacute combined degeneration of spinal cord

Introduction:

It is one of the neurological complications of vitamin B12 deficiency.

Clinical features:

1. *Pyramidal tract damage:*
   Spastic weakness (UMN type).
2. *Dorsal column damage:*
   - Loss of joint, position and vibration sense (deep sensation).
   - ***Romberg's sign*** (Sowing to one side on eye closure): Positive.
   - ***Stamping gait***: Patient does not know where his foot is and so, while walking raises the foot high up and brings down on the ground forcefully like stamping.
3. *Peripheral neuropathy:*
   - Sensory: Impaired superficial sensation, patchy in distribution.
   - Motor: LMN signs.
4. *Cognitive impairment/ dementia.*

Investigation:

1. Nerve conduction velocity (NCV) study
2. MRI: To rule out any structural damage.
3. Blood: Hb↓, MCV↑, macrocytes present; pancytopenia may be present.
4. Estimation of serum vitamin B12 ± Folic acid.

Treatment:

Parenteral supplementation of vitamin B12 + Oral folic acid.

Fredrick's Ataxia

Inheritance: Autosomal recessive (AR).

It shows 'trinucleotide repeat sequence': manifestations occur earlier in successive generations.

Clinical features:

1. Pyramidal tract damage:
   Spastic weakness (UMN type).
2. Spino-cerebellar fibre damage:
   - Ataxia
   - Nystagmus
   - Past pointing (pointing beyond the finger in the finger-nose test)
   - Dysdiadochokinesia (impaired ability to perform rapid, alternating movements).
3. Peripheral neuropathy:
   - Sensory: Sensory loss
   - Motor: LMN type of weakness.
4. Optic atrophy
5. Cardiomyopathy
6. Skeletal:
   - Pes cavus
   - Pes equinovarus.

Investigation:

1. NCV
2. MRI
3. Genetic studies.

Treatment:

1. Supportive
2. Genetic counselling.

Tabes Dorsalis

Introduction:

It is a neurological complication of syphilis.

Clinical features:

D	Dorsal column lesion: Loss of deep sensation
O	Ocular manifestation: Argyll Robertson pupil*

R	Root damage: Severe electric shock like pain sensation along the distribution of affected root
S	Stamping gait
A	Autonomic disturbance: Acute abdominal pain (Tabetic crisis)
L	Loss of sphincteric control
I	Intact power of muscles (Pyramidal tract not affected)
S	Skeletal deformity: Charcot joint~

* Argyll Robertson pupils are bilateral small pupils that reduce in size when the patient focuses on a near object, but do not constrict when exposed to bright light. It is a highly specific sign of neurosyphilis.

~ Charcot joint is a progressive degenerative/ destructive joint disorder in patients with abnormal pain sensation and proprioception.

Investigation:

1. VDRL test
2. TPHA test.

Treatment:

Benzyl penicillin (IM)/ 3rd generation cephalosporins.

Multiple sclerosis (MS)

Introduction:

It is an autoimmune inflammatory demyelinating disease of the nervous system.

Etiopathogenesis:

In genetically predisposed individuals, an autoimmune response triggers off the demyelinating process. Environmental factors probably play some role.

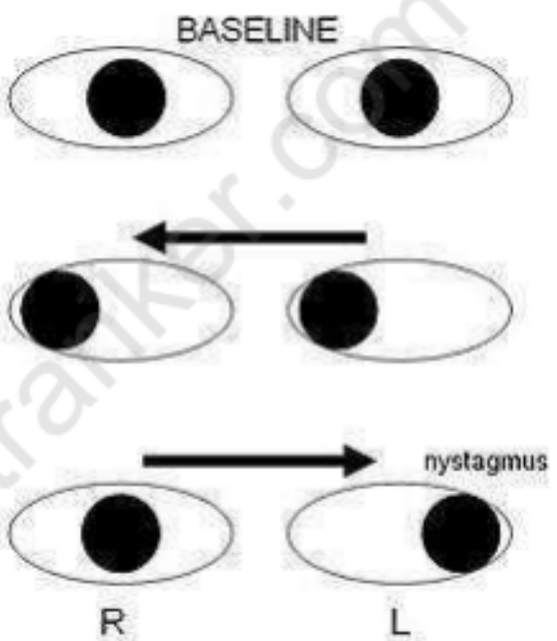
Clinical course:

Depending on the clinical course, MS is classified into 3 types:

1. Relapsing-remitting type
2. Primary progressive type
3. Secondary progressive type.

Clinical features:

1. Higher function:
   - Cognitive dysfunction
   - Depression.
2. Cranial nerves:
   - Optic neuritis
   - Trigeminal neuralgia
   - Internuclear ophthalmoplegia (On attempted lateral gaze, there is a failure of adduction and nystagmus of the abducting eye, cause: damage of median longitudinal fasciculus)



3. Motor:
   - Weakness:
     May affect one/ multiple limbs. There is marked spasticity with UMN signs.
4. Sensory:
   - Patient may complain of numbness, tingling, paresthesia of different parts of body.
   - Sensory impairment

- ***Lhermitte's sign***: Electric shock like sensation passes down the spine on attempted neck flexion.

5. Cerebellum:
   Cerebellar signs may be present.
6. Sphincter:
   Sphincteric disturbances are common (retention and incontinence).
7. Trophic changes:
   As many of these patients are completely bed ridden, they may develop bed sore, trophic ulcer etc.

Investigation:

1. MRI of brain and spine:
   Visualizes the demyelinating lesion (Plaque).
2. Lumber puncture:
   CSF shows markedly elevated IgG (Oligoclonal band of IgG).

Treatment:

1. Supportive:
   a. Bed sore prevention
   b. Catheterization
   c. Exercise and physiotherapy
   d. ***Daclofen***: For spasticity.
   e. Rehabilitation
   f. Mobility aid.
2. Drugs:
   a. Corticosteroid:
      - In acute flare: IV Methylprednisolone
      - Chronic phase: Oral corticosteroid.

   b. Immunomodulators:
      - ***Glatiramer acetate***
      - ***Natalizumab***
      - IV IgG/ Plasmapheresis may be effective during acute relapse.

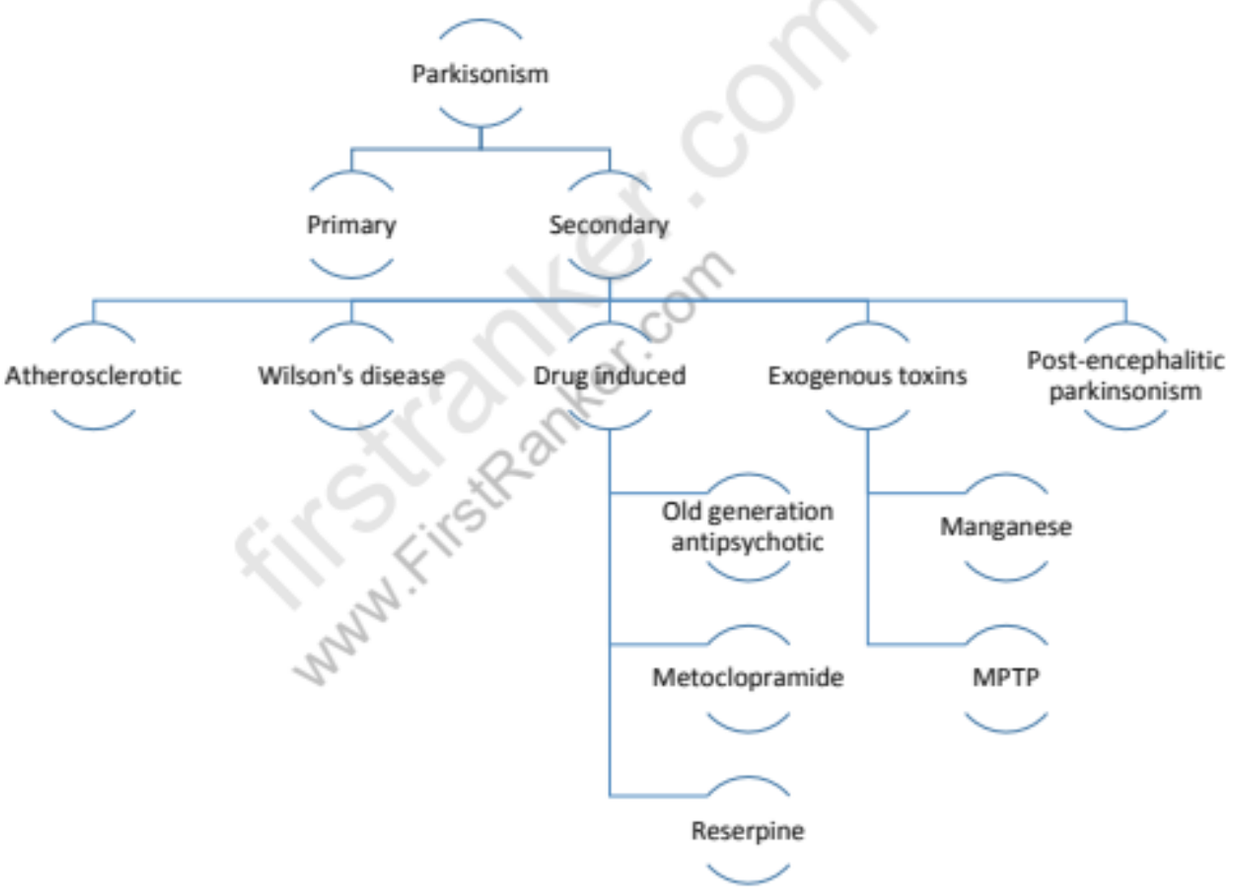
Parkinsonism

Introduction

It is a disease of extrapyramidal tract characterized by imbalance between dopamine and acetylcholine, leading to the triad of:

- Bradykinesia
- Rigidity and
- Tremor.

Etiology:



Pathogenesis:

Initiation:

1. Genetic mutation: *Parkin* and *α synuclein* genes are affected

2. Atherosclerosis
3. Toxins.

- Imbalance between dopamine (↓) and acetyl-choline (↑) in nigrastriatal pathway leads to clinical manifestations.

Clinical manifestations

1. Higher functions:
   - Cognitive dysfunction
   - Depression
   - Gambling tendency.
2. Movements:
   a. Voluntary:
      - Slow to initiate and carry out any activity (*bradykinesia*)
      - Finer movements (which requires precision) are impaired
        Ex: Handwriting becomes progressively illegible and small in size (micrographia)
      - Activities requiring postural control get impaired: there is a tendency to fall.

   b. Autonomic functions:
      Impaired/ lost.
      Ex: Loss of spontaneous arm swinging during walking.

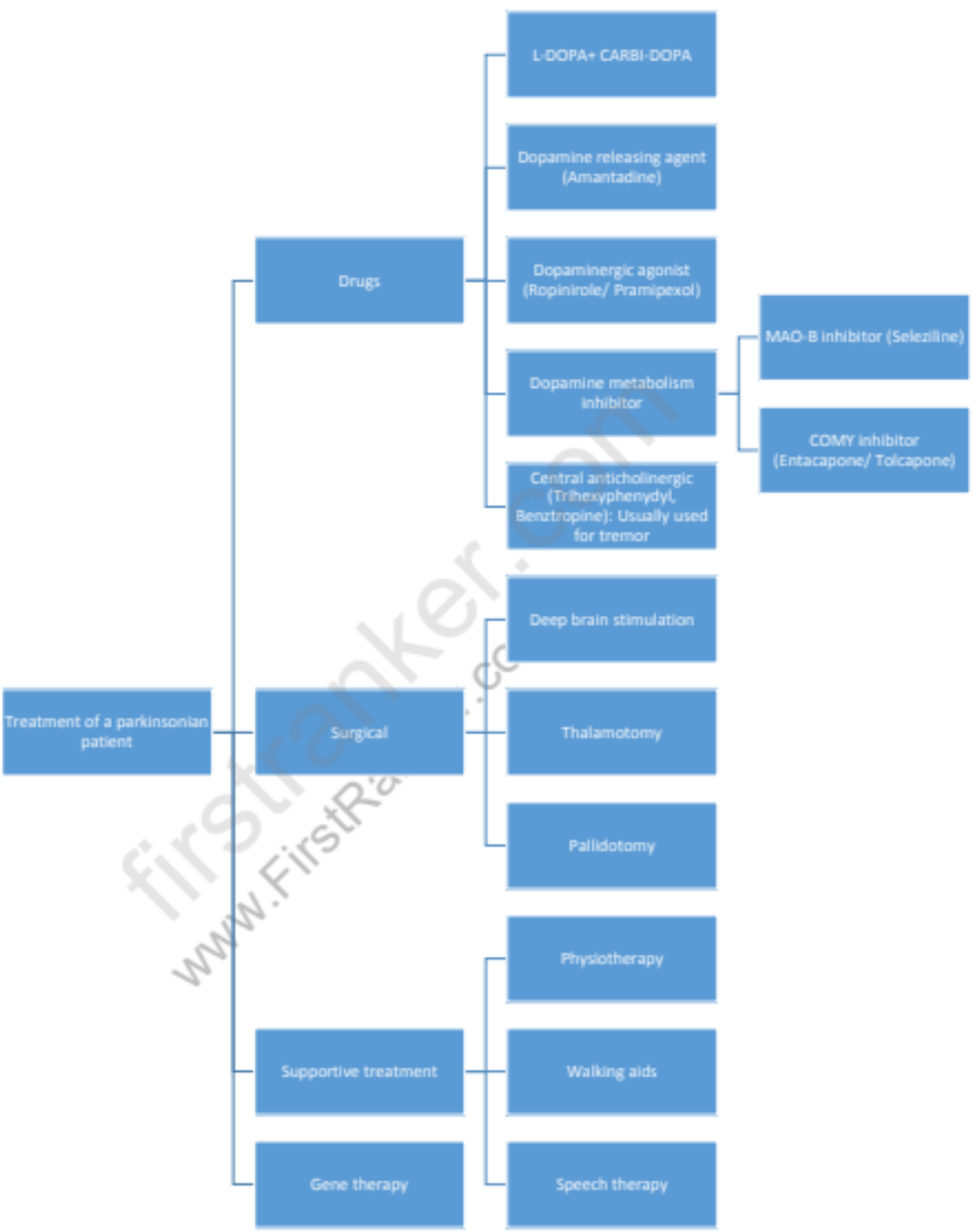
   c. Involuntary movement: *Tremor*:
      - Rest tremor:
        Rest tremor in the hand is often described as 'pill rolling'/ 'drum beating', the tremor is usually marked in the wrist and fingers, which display motion made in the act of rolling a pill.
        Rest tremor often also affects ankle and may be seen in the tongue and lower jaw.
      - Aggravates when the patient is emotionally upset/ excited
      - Decreases in relaxed state/ sleep/ when the affected part is thrown into action.
3. Power:
   Normal, provided that sufficient time is given to build it up.

4. Tone:
   Hypertonia/ rigidity
   - Types:
     a. Lead pipe rigidity (rigidity> tremor)
     b. Cogwheel rigidity (tremor> rigidity).
   - Usually the *flexors are more hypertonic* than the opposite group: this is responsible for the typical attitude/ posture of the patient
   - Rigidity of pharyngeal muscle:
     a. Swallowing difficulty
     b. Dribbling of saliva.
   - Rigidity of laryngeal muscle:
     Typically monotonous, slow voice without any modulation.
5. Wasting: Absent
6. Reflex: Usually normal
7. Sensory function: Usually normal
8. Sphincter function: Usually normal
9. Gait:
   - Difficult to initiate
   - Slow velocity: short shuffling gait
   - Impaired/ lost arm swing in time of walking
   - Tendency to fall and at times, to prevent this fall, patient tries to run towards the destination; leading to "*Festinating gait*"
   - Difficulty to stop
   - Difficulty to turn around (the whole body turn around like a statue).
10. Facies:
    Due to rigidity and bradykinesia of facial muscles, face becomes expressionless with infrequent blinking and staring look, often called "*Parkinsonian mask facies*".

Investigation

Clinical diagnosis, however a CT/ MRI of brain is often carried out to rule out any structural lesion.

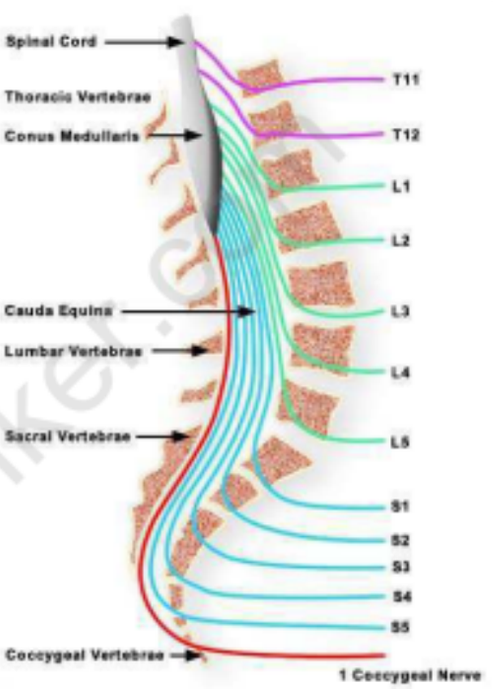
Treatment



Differential diagnosis

1. Diseases causing tremor:
   a. Parkinsonism
   b. Cerebellar disorders
   c. Benign familial tremor.
2. Parkinsonism plus syndrome:
   a. Progressive supranuclear palsy (PSP)
   b. Multisystem atrophy/ Shy-Drager syndrome
   c. Olivo-ponto-cerebellar degeneration.
3. Causes of parkinsonism (See in etiology).

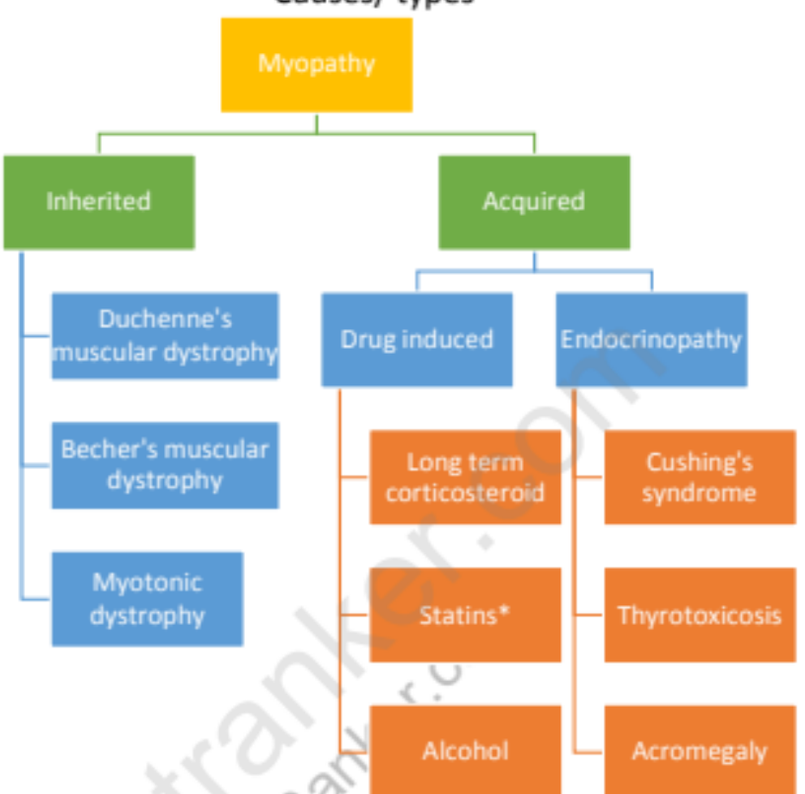
Conus medullaris and Cauda equina lesions

Points	Conus medullaris lesion	Cauda equina lesion
Definition	Conus medullaris is the terminal tapering end of spinal cord containing lower sacral and single coccygeal segment.	Cauda equina is the lower spinal nerves containing lower lumber, sacral and coccygeal nerves: all of which descend towards their exit through intervertebral foramina.
Graphical representation		
Clinical features		
Lower limb weakness	Absent	Present, usually variable and asymmetrical
Lower limb jerk	Normal	Asymmetrical hypo-reflexia/ a-reflexia
Sensory loss	From perianal and perineal region (Saddle back anesthesia)	Asymmetrical sensory loss in both lower limbs
Sphincteric disturbance	Very prominent	Not common
Etiology	Lumbosacral vertebral diseases	
Surgery	Often considered	Not commonly considered
Investigation	MRI spine	

Myopathy

Intrinsic diseases of muscles characterized by muscle weakness ± wasting.

Causes/ types



*Statins are more liable to cause myositis.

Symptoms and signs

1. Weakness: ***Proximal weakness***
2. In some cases, there may be ***severe muscle pain*** due to myositis
3. ***Muscle wasting***: Often present
4. Fasciculation: Absent
5. Tone/ jerks/ plantar: Usually normal.

Investigation

1. Serum CPK: May be elevated if there is myositis
2. EMG: Confirms the diagnosis
3. In selected cases, muscle biopsy may be required.

Treatment

1. Treat the underlying cause
2. Stop the offending drug
3. Rehabilitation.

Myotonic dystrophy

Clinical features

Organ/ area	Defect
Face	Frontal boldness
Eye	Bilateral ptosis Premature cataract Hollowing of the face (due to wasting of facial muscles)
Thyroid	Goitre
Heart	Conduction disturbance
Breast	Gynecomastia
Limbs	Myotonia (inability to relax muscles after sustained voluntary contraction, i.e. handshake)
Muscles	May be wasted, but *no gross weakness*
Testes	Testicular atrophy
Systemic	Diabetes

Investigation

Genetic testing

Treatment

- Genetic counseling and support.
- Often responds to:
  - Phenytoin
  - Procainamide
  - Quinine.

Duchenne's muscular dystrophy

General description:

- X linked recessive myopathy
- Usually occurs in male.

Clinical features:

1. ***Weakness***: Involves proximal muscles; so, more prominent in shoulder and pelvic girdle muscles
2. ***Wasting***: Affected muscles may get wasted
3. ***Pseudo-hypertrophy***: Particularly affecting Gastrocnemius muscle due to abnormal accumulation of fibro-fatty tissue
4. ***Wadling gait***: During walking, body slightly bend backwards and kid walks with wide based gait
5. ***Gower's sign***: If baby is asked to stand up from a lying down position, he first rolls on to one side then sits up supporting his elbow on the ground and then gradually stands up by climbing up his knees
6. ***Intellectual impairment***
7. ***Cardiomyopathy.***

Investigation:

Genetic testing: Mutation of "Dystrophin" gene.

Treatment:

Supportive. Usually these babies become completely bedbound by the age of 10-12 years.

Becker's muscular dystrophy

Clinical feature like Duchenne's muscular dystrophy but:

- May remain mobile upto the age of 15 years
- Intellectual impairment more common
- Cardiomyopathy less common.

Infective diseases

Meningitis

Inflammation of meninges.

Causes

1. Pyogenic organism: *N.meningitidis*, Pneumococcus, *Staph.aureus* (post neuro-surgical patients), Listeria
2. Tubercular
3. Viral: HSV, Mumps
4. Fungal (in HIV patients): Cryptococcus, Histoplasma
5. No cause is found in 50% of cases.

Pyogenic meningitis

Clinical features

1. Constitutional symptoms:
   - Acute febrile illness
   - Weakness, malaise, loss of appetite.
2. CNS symptoms:
   - Headache: Severe, usually gradual in onset and progressively increasing in nature
   - Neck pain ± neck stiffness
   - Photophobia: The patient often prefers to lie with eyes closed/ in a dark room
3. Features of increased ICT:
   A. Altered consciousness
   B. Behavioral abnormality
   C. Confusion, coma, convulsion
   D. Delirium
   Headache increases and vomiting may occur.
4. Cranial nerve palsy:
   CN 3, 4 and 6 palsy may occur.
5. Cutaneous manifestations:
   Meningococcemia: Meningococcal rash:

Typically starts as purpuric/ pin head spots which may enlarge. These rashes characteristically don't bleach on pressure (Glass test) due to small vessel vasculitis/ leakage.

Signs

1. Temperature: High
2. GCS: May be low
3. Meningeal signs:
   - Neck stiffness/ neck rigidity
   - Kernig sign
   - Brudzinski sign
4. Signs due to underlying focus of infection (ENT areas/ danger area of face).

Investigation

1. Blood: Hb, TC, DC, ESR/ CRP
2. Renal function: Na+ K+ Urea Creatinine
3. Blood culture and sensitivity
4. CT head: To rule out any radiological evidence of raised ICT (which is a relative contraindication of lumber puncture)
5. Lumber puncture:
   Description of CSF sample:
   I. Physical appearance: May be turbid
   II. Biochemistry:
      - Glucose:
        Normal: 40-85 mg/dL
        In pyogenic meningitis: ↓↓

        Normally $\frac{CSF\ Glucose}{Random\ blood\ glucose} > 0.6$
        In pyogenic meningitis, this ratio often goes <0.4

        Note:
        Paired CSF and random blood glucose is checked to rule out masking of CSF hypoglycemia due to hyperglycemic condition. (If a person is diabetic, his blood glucose level will be high and

consequently, his CSF glucose level will also be high. Even in the presence of pyogenic meningitis, his CSF glucose will be within the normal range. To avoid this fallacy, the ratio is taken.)

- Protein:
  Normal: 15-45 mg/dL
  In pyogenic meningitis: ↑↑

- Cytology:
  Normal WBC: 0-5/µL
  In pyogenic meningitis: Leukocytosis is seen, which is neutrophilic in nature. Often WBC level will go above >1000/µL.

- Microbiological:
  Gram stain + culture sensitivity.

Treatment

- Supportive treatment:
  A. Absolute bed rest
  B. Bed sore prevention
  C. Catheterization
     Circulation by IV fluid
  D. Drugs (supportive):
     - ✓ Antipyretic
     - ✓ Antiemetic
     - ✓ Anticonvulsant
     - ✓ Anti-edema (mannitol).

     DVT prophylaxis
     Diet.
- Definitive treatment:
  Empirical antibiotic (IV Ceftriaxone): Usually given for 10-14 days
  If Staph. infection is suspected, then add Vancomycin.

Complications

Mnemonic	Complications
S	Septicemia Subdural empyema Seizure
A	Abscess of brain Acute adrenocortical failure/ Waterhouse Friderichsen syndrome (due to adrenal insufficiency resulting from meningococcal vasculitis: patient suddenly goes into shock)
H	Hydrocephalus Hemiparesis (stroke due to cerebral vasculitis)

Tubercular meningitis

Clinical features

1. Constitutional symptoms:
   Gradual in onset, often may persist for few days to weeks before meningitis symptoms appear:
   - Low grade fever
   - Weight loss
   - Loss of appetite
   - Low grade headache.
2. CNS symptoms:
   - Headache: May increase
   - Vomiting
   - Photophobia.
3. Symptoms of raised ICT:
   ABCD as previous one
4. Symptoms of pulmonary TB may be present.

Signs

1. Temperature: Raised
2. GCS: May be low
3. Meningeal signs: Neck rigidity may be present
4. Cranial nerve palsy: CN 3, 4, 6 palsy (most common: CN6) may be present

5. Fundoscopy: Choroid tubercles may be present
6. Signs of active pulmonary TB may be present.

Investigations

1. Blood: Hb, TC, DC, CRP/ ESR
2. Renal function: Na+ K+ Urea Creatinine
3. Blood culture
4. Sputum AFB and mycobacterial culture (if active pulmonary TB is suspected)
5. Chest X Ray
6. CT head:
   - ✓ To rule out raised ICT
   - ✓ Tuberculoma may be present.
7. Lumbar puncture (LP):
   I. Physical appearance:
   'Cobweb coagulum' may be present
   II. Biochemistry:
   - Protein: ↑↑
   - Glucose: ↓↓
   - $\frac{CSF\ glucose}{venous\ glucose} = (0.4 - 0.6)$

   III. Cytology:
   - WBC count: ↑ (Usually 50-1000/µL)
   - Predominant cells: Lymphocyte

   IV. Microbiology:
   - AFB: May be present
   - Mycobacterial culture (BACTEC MGIT method): Often positive
   - Rapid detection method:
     X-PERT TB (Nucleic acid amplification)/ ***RIF*** (Rifampicin assay):
     - ✓ Promoted by WHO
     - ✓ Detect *M.tuberculosis*
     - ✓ Can detect rifampicin resistance which is a sensitive predictor of MDR-TB.

Treatment

Definitive treatment:

Anti-tubercular drug:

- Duration: HREZ for 2 months and HR for at least 7-10 months
- Dosage: H (5 mg/kg), R (10 mg/kg), E (15 mg/kg), Z (25 mg/kg)
- Vitamin B6 supplementation with H

Corticosteroid:

- Duration: 4-6 weeks
- Rationale: To prevent tubercular arachnoiditis and meningeal thickening so that chance of obstructive hydrocephalus and permanent neurological deficit is minimized.

Complications

1. Obstructive hydrocephalus
2. Stroke/ hemiparesis (resulting from tubercular arteritis).

Brain abscess

A localized area of suppurative infection within the brain.

Risk factor

Although infection may spread to brain from any primary focus of infection in the body, the common risk factors are:

- Ear infection
- Dangerous area of face
- Scalp laceration/ skull injury
- Post neurosurgical patients.

Common organism

Streptococcus/ staphylococcus/ anaerobes.

Clinical features

1. Systemic: Fever, malaise, weight loss
2. Features due to increased ICT
3. Primary focus of infection often present.

Investigation

CE-CT of brain

Routine: Hb, TC, DC, CRP, Blood culture

Treatment

Empirical antibiotic:

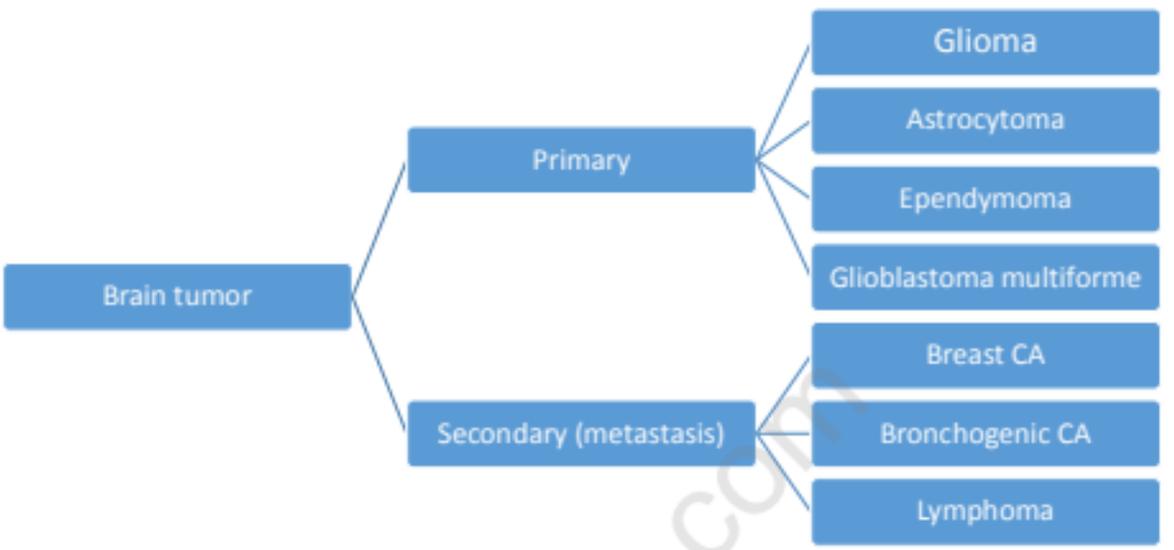
Ceftriaxone IV + Metronidazole

(+ Vancomycin if Staph. infection suspected)

Often IV antibiotics may be continued for 4-6 weeks after which they are changed to appropriate oral forms.

Brain tumor

Classification



Mechanisms of clinical manifestations

1. Raised ICT
2. Localizing symptoms and signs (focal manifestations)
3. False localizing symptoms
4. Herniation syndrome.

Clinical manifestations

Manifestations due to raised ICT

A. Altered consciousness
B. Behavioral disturbances
C. Confusion, coma, convulsion
D. Delirium
E. Etiology: Features due to underlying etiology
F. Focal neurodeficit
Fundoscopy: Papilledema
G. GI manifestations: Nausea, vomiting
GCS may be low
H. Headache
Herniation syndrome.

Localizing symptoms and signs (focal manifestations)

Area involved	Signs
Frontal lobe (Motor area)	• Contralateral pyramidal signs • Focal seizures (due to seizures) with motor manifestations • Aphasia (Broca's) • Anosmia (CN1) • Behavior and personality disturbances.
Parietal lobe (Sensory area)	• Contralateral hemisensory loss • Astereognosis (inability to recognize an object by its size, shape and texture) • Sensory inattention/ neglect (visual/ spatial) • Contralateral homonymous hemianopia (inf.quadrantic).
Temporal lobe	• Focal seizure with somatosensory manifestations • Psychiatric manifestations • Automatism • Aphasia (Wernicke's) • Contralateral homonymous hemianopia (sup.quadrantic).
Occipital lobe	• Contralateral homonymous hemianopia • Cortical blindness (in case of bilateral compression).

False localizing symptoms

A group of clinical manifestations occur in patients with raised ICT (particularly brain tumor) which gives an erroneous impression about the site of the lesion:

- CN6 palsy (as it has the longest intracranial course)
- CN3 palsy
- Bilateral extensor plantar (resulting from brainstem compression)
- Ipsilateral upgoing plantar (resulting from compression of cerebral peduncle against tentorium).

Herniation syndrome

If there is raised ICT, a part of the brain sometimes escape through a foramen towards a low pressure compartment. This commonly occurs with cerebellum,

which herniates through foramen magnum; leading to brainstem (particularly medullary) compression- causing:

1. Respiratory arrest
2. Circulatory collapse
3. Deep coma.

Investigation

1. CE-CT Brain
2. If CT is abnormal: MRI brain.

Treatment

Supportive treatment:

1. Propped up positioning
2. Anti-edema drugs:
    - IV mannitol (for 3-5 days) followed by oral glycerol
    - Steroid
3. Analgesic
4. Antiemetic
5. Anticonvulsant (if seizure occurs)
6. Surgical:
    - Ventriculo-peritoneal shunt (in case of obstructive hydrocephalus)
    - Craniotomy flap (in a stroke patient with severe edema).
7. Elective ventilation with permissive hyperventilation (hypocapnia will induce cerebral vasoconstriction and therefore ICT will be reduced).

Definitive treatment:

1. Surgical excision of brain tumor
2. Radiotherapy
3. Chemotherapy.

Pseudotumor cerebri/ Benign intracranial hypertension (BIH)

Introduction

It is a condition characterized by raised ICT where the underlying cause is innocuous.

Causes

1. Idiopathic (common in young obese females)
2. Drugs:
   a. Long term vitamin A derivatives (Ex: Retinoid)
   b. Tetracycline
   c. OCP.

Clinical features

- Often patients are obese
- H/O offending drug intake
- Signs and symptoms of raised ICT (ABCDEFGH as describe above)

Investigations

1. CE-CT Brain:
   Findings:
   - ✓ Signs of raised ICT
   - ✓ No structural lesion
   - ✓ Ventricular system not dilated.
2. LP (do very cautiously):
   High CSF pressure; other findings are normal.

Treatment

1. Encourage to lose weight
2. Stop offending drugs
3. Acetazolamide tablet (Carbonic anhydrase inhibitor)
4. Therapeutic lumbar puncture.

Seizure

Introduction

Definition of seizure:

Temporary cerebral dysfunction due to abnormal paroxysmal neuronal discharge.

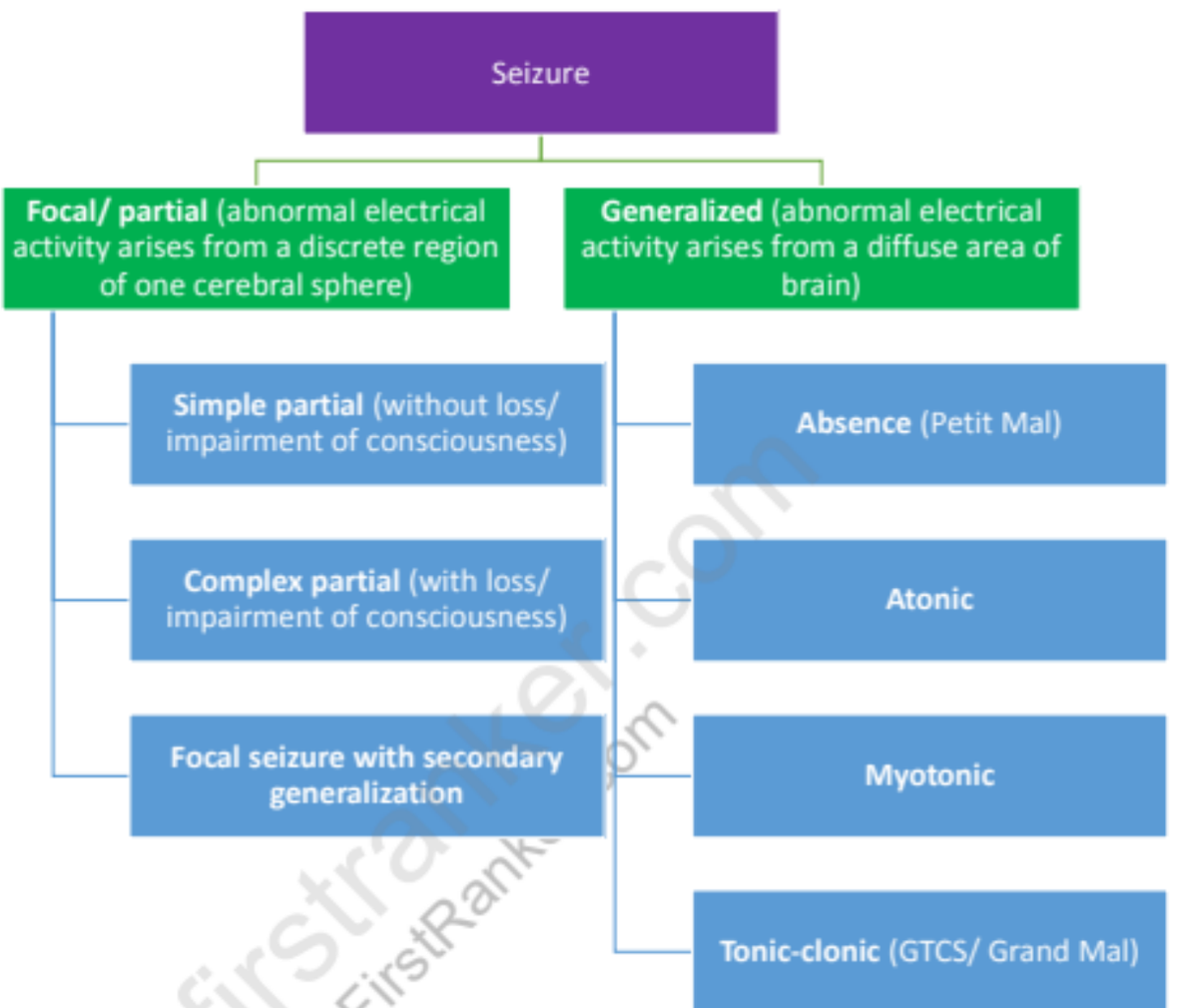
Definition of epilepsy:

It is a clinical syndrome characterized by *recurrent unprovoked seizures*.

Causes of seizure

A. Alcohol (intoxication/ withdrawal)
   Accident (Post traumatic seizures)
B. Biochemical abnormality:
   - Hypoglycemia/ hyperglycemia
   - Hyponatremia/ hypernatremia
   - Hypercalcemia
   - Metabolic encephalopathy (uremic/ hepatic/CO_2 narcosis)
   - Hypoxic/ ischemic encephalopathy.
C. Cerebral causes:
   - Vascular causes:
     - ✓ Ischemic stroke
     - ✓ Hemorrhagic stroke
   - Intracranial space occupying lesions (IC-SOL)
   - Infections:
     - ✓ Meningitis
     - ✓ Encephalitis
     - ✓ Tuberculoma
     - ✓ Neurocysticercosis
     - ✓ Brain abscess.
D. Drugs (side effect/ intoxication):
   - Fluoroquinolone
   - Carbapenem.
E. Exogenous toxins (Ex: Cocaine).
I. Idiopathic.

Types of seizure



Clinical features

Focal seizure:

Clinical features of this type of seizure depends on the location of focus of abnormal electrical activity.

Simple partial seizure

1. ***Motor:***
   - Twitching/ jerky movement of a part of the body
   - This may spread to other parts of boy (Jacksonian movement).
2. ***Somatosensory:***
   - Paresthesia (tingling/ pins and needle sensation)

- This can spread to other parts of the body.

3. ***Special sensory manifestations:***
   - Visual: Flashes of light
   - Auditory: Buzzing noise
   - Olfactory: Odd smell
   - Gustatory: Odd taste.
4. ***Autonomic:***
   - Epigastric fullness
   - Palpitation
   - Flushing/ sweating
   - Psychiatric manifestations:
     - Illusion
     - Hallucination
     - Deja-vu.

▪ The above manifestations usually occur for few seconds to minutes but there is ***no impairment of consciousness***.

Complex partial seizure

1. ***Aura:***
   Motor/ somatosensory/ special sensory/ autonomic/ psychiatric manifestations occur before loss of consciousness.
   However, these manifestations may accompany/ follow the loss of consciousness.
2. ***Loss of consciousness:***
   Typically lasts for few seconds to minutes, when the patient develops impairment/ loss of consciousness.
3. ***Automatism:***
   Abnormal voluntary activity like lip smacking, repeated swallowing effort, sudden running etc.
4. ***Recovery:***
   Usually, after recovery there is amnesia about impaired consciousness episode.

Focal seizure with secondary generalizations

This may be of 2 types:

1. Simple partial seizure followed by GTCS
2. Complex partial seizure followed by GTCS.

Generalized seizures:

Absence seizure

- Typically occurs in children (4-8 years) and usually ceases by the age of 20.
- Clinical features:
  - Sudden/ abrupt ***loss of external awareness***: this spell typically lasts for few seconds to minutes and followed by complete recovery.
  - Often occurs in the middle of a conversation when the patient misses a few words/ breaks off in missed sentence and restart the conversation from where he/she left off.
  - In some patients, it is accompanied by a tonic/ tonic-clonic movement.
  - Patients are not aware about these spells.

Atonic seizure

- ***Abrupt loss of postural tone*** leading to dropping of head/ sudden collapse/ fall. These patients are at risk of getting injured.

Myoclonic seizure/ Juvenile myoclonic seizures

- ***Sudden abnormal forceful contraction of muscles*** leading to violently disobedient limb.

Generalized tonic clonic seizure (GTCS)

Clinical stages:

1. ***Tonic stage:***
   Characterized by generalized rigidity/ spasm of different muscles leading to:
   - An abnormal posture (hyper-extended)
   - Eyes rolled upwards (ocular spasm)
   - Pooling of saliva and frothing (pharyngeal spasm)
   - Ictal cry/ a moaning (low pitch) sound (laryngeal spasm)
   - Tongue bite (Spasm of jaw)

- A brief spell of respiratory arrest (spasm of respiratory muscles).
  - Usually this tonic stage lasts for few seconds to minutes and followed by clonic stage.

2. ***Clonic stage:***
   - Abnormal jerky movement of different parts of the body which may be short lasting (seconds to minutes) or may go on for a while.
   - If this continues for a while ***without any intervening period of recovery of consciousness***, it is called "status epilepticus".
   - If this continues for a while with intervening periods of recovery of consciousness, it is called "serial seizures".
     - During tonic-clonic spells, patient usually becomes unconscious.
3. Post-ictal period:
   May last for few minutes to few hours and typically characterized by:
   - A state of confusion/ disorientation
   - Significant headache
   - Significant muscle pain
   - Patient may develop urinary/fecal incontinence. This is a usually a non-specific symptom.

Investigation

(Correlate with the causes of seizure)

1. Blood: Hb, TC, DC, ESR
2. Renal function: Na+ K+ Urea Creatinine
3. Liver function test
4. Serum Ca++
5. Blood glucose
6. CE-CT brain (to rule out a structural lesion); if CT is inconclusive, then an MRI may be required.
7. EEG: May be normal/ abnormal.

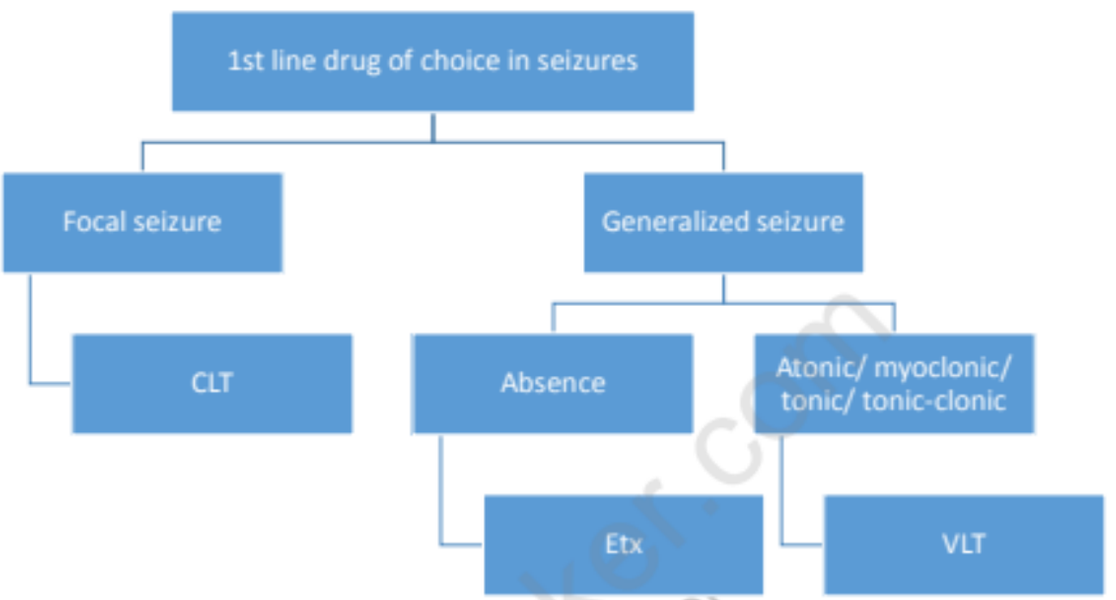
Treatment

General advice:

- Ensure enough sleep for at least 8 hours a day
- Avoid driving/ swimming/ working with heavy machinery

- Seek attention in case of any infection
- Monitor the side effects of your own medication(s).

Drug treatment:



C= Carbamazepine, L: Lamotrigine, T: Topiramate, V: Valproate, Etx: Ethosuximide

Intervention for medically refractory epilepsy:

1. Partial lobectomy
2. Lesionectomy
3. Vagus mediated stimulation.

Principles of drug treatment of seizures

1. When to start?
   a. Idiopathic/ unprovoked seizures:
      - Wait and watch policy
      - Long term antiepileptic drugs (AED).
   b. Secondary/ provoked seizures:
      - Treating the underlying cause is enough
      - AED on temporary basis
      - AED on long term basis.

2. What are the general principles of treatment?
   a. Start on monotherapy, then gradually increase the dose till the maximum dose is reached/ patient is seizure free
   b. Monitor for side effects/ toxicity
   c. If patient is not seizure free even after taking maximum tolerable dose, then consider starting another 1st line drug
   d. Gradually increase the dose of the 2nd drug and gradually taper off the dose of 1st drug
   e. If an add-on drug is required, then adjunctive drugs which are currently used are:
      - ✓ Gabapentin
      - ✓ Pregabalin
      - ✓ Levetiracetam
      - ✓ Lacosamide.
3. When to stop?
   AED can be stopped if the patient remains completely seizure free for consecutive 2 years (or 5 years).

Dx of epilepsy

1. Non-epileptic seizure/ pseudoseizure/ functional seizure:
   Points to differentiate:
   a. More common in young female
   b. Often witnessed
   c. Aura absent
   d. Convulsive movement often looks unusual
   e. Injury rare
   f. Tongue bite rare
   g. Incontinence rare
   h. Hyperprolactinemia absent.
2. Syncope:
   Points to differentiate:
   a. Pre-monitory/ warning symptoms are different from aura

b. Twitching/ jerky movement, if occurs, doesn't last for more than few seconds
c. No post-ictal symptoms
d. Tongue bite rare.

3. TIA:
   Points to differentiate:
   Negative symptoms predominant:
   a. Weakness (loss of power)
   b. Loss of sensation
   c. Loss of special sense.

Status Epilepticus

Introduction

Repeated tonic-clonic seizures for a while ***without any intervening period of recovery*** of consciousness.

Investigations

Once the seizure is controlled, relevant investigations are to be done to rule out any provocable cause of seizure.

Management

A. Airway: Must be secured. If required apply oropharyngeal suction and intubation.
B. Breathing: Free flow oxygen and ventilation, if required.
C. Circulation: Maintain circulatory volume by IV fluid
D. Drugs:

IV Lorazepam or IV/Per rectal Diazepam

↓

If fails

↓

IV phenytoin (Loading dose followed by maintenance dose)

↓

If fails

↓

IV phenobarbitone

↓

If fails

↓

Intubation + Ventilation + General anesthesia / Deep sedation

- Once seizure is under control, long term AED treatment should be considered.

Syncope

Introduction

Transient loss of consciousness (LOC) due to *global cerebral hypoperfusion*, where LOC is abrupt in onset, of short duration and followed by *spontaneous complete recovery*.

Causes

1. Neurally mediated/ reflex syncope:
   A. ***Vasovagal attack***:
      Pathogenesis:
      I. Vasodepressor response (loss of vasoconstrictor tone, particularly in upright position)
      II. Cardio-inhibitory response/ asystole.
   B. ***Situational syncope***:
      I. Forced micturition
      II. Forced defecation
      III. Cough syncope.
   C. ***Carotid sinus hypersensitivity***.
2. Cardiogenic causes:
   A. ***Arrhythmia***:
      I. Brady-arrhythmia:
         - Sinus node disease
         - AV junction block (Heart block)
      II. Tachy-arrhythmia:
         - Supraventricular tachycardia (SVT)
         - Ventricular tachycardia (VT).
   B. ***Structural heart disease***:
      I. Aortic stenosis (AS)
      II. Hyperobstructive cardiomyopathy (HOCM).
3. Postural/ orthostatic hypotension:
   Causes:
   A. Drugs: Vasodilators (antihypertensives)
   B. Autonomic neuropathy
   C. Volume depletion: Bleeding/ dehydration.

Clinical features

A. Prodromal/ warning/ pre-syncopal symptoms:

- Autonomic symptoms (suggestive of vasovagal attack):
  - ✓ Sweating
  - ✓ Nausea
  - ✓ Palpitation
  - ✓ Paleness of patient
  - ✓ Blurred vision.
- Postural symptoms (suggestive of orthostatic hypotension):
  - ✓ Dizziness
  - ✓ Light headacheness
  - ✓ Blurred vision.
- Usually no pre-syncopal symptoms (suggestive of transient arrhythmia).

- Some patients with warning symptoms can actually prevent the loss of consciousness by taking appropriate defensive mechanisms. Therefore, these patients often come with pre-syncopal symptoms.

B. Syncopal episode:

- Precipitating factors:
  - ✓ Prolonged standing (vasovagal)
  - ✓ Emotional stress:
    Bad news/ severe fear/ anxiety/ horrible sight/ smell etc.
  - ✓ Overcrowded place
  - ✓ Sudden head turning/ tight collar shirt/ shaving (carotid sinus hypersensitivity)
  - ✓ Sudden standing (postural hypotension).
- Actual episode:
  - ✓ Transient loss of consciousness usually lasting never for a period > (10-15) sec.
  - ✓ Often there is a gap in the memory
  - ✓ Jerky movements of the limbs (seizure) may occur but is usually *very brief* and usually occurs after TLOC
  - ✓ Momentary urinary incontinence may occur
  - ✓ Tongue bite *extremely rare*.

C. Recovery phase:

Usually recovery is complete and there is no post-syncopal confusion/ drowsiness.

Signs

- (Lying + standing) BP must be recorded if postural hypotension is suspected
- Ask the patient to stand for 2-3 minutes: A fall of SBP/DBP ≥ 20/10 mm Hg + postural symptoms is suggestive of postural hypotension
- Proper cardiovascular examination should be carried out to elicit any cardiac abnormality which may cause syncope (Arrhythmia/AS/HOCM).

Investigation

1. ECG:
   1. Resting ECG
   2. 24 hours continuous ECG (if transient arrhythmia is suspected and resting ECG is normal)
2. Echocardiogram (to rule out structural heart disease)
3. Carotid sinus massage (to confirm carotid sinus hypersensitivity)
   - To be avoided in patients with H/O carotid TIA/ if carotid Doppler shows atherosclerotic plaque.
4. Head tilt test/ Tilt table test (to diagnose neurally mediated syncope)
5. Electrophysiological study: Focus of tachyarrhythmia can be induced, located and ablated, if required.
6. CT head (to rule out any internal bleeding as a consequence of fall).

Treatment

1. Neurally mediated syncope:
   Advice the patient:
   a. To avoid any triggering/ precipitating factor
   b. To take a defensive mechanism during a pre-syncopal symptom
   c. Physical counter pressure maneuvers:
      I. Continuous limb movement while prolonged standing
      II. To sit with crossed legs
      III. Simple isometric exercise of the hand (hand-gripping)
   d. To drink plenty of fluid

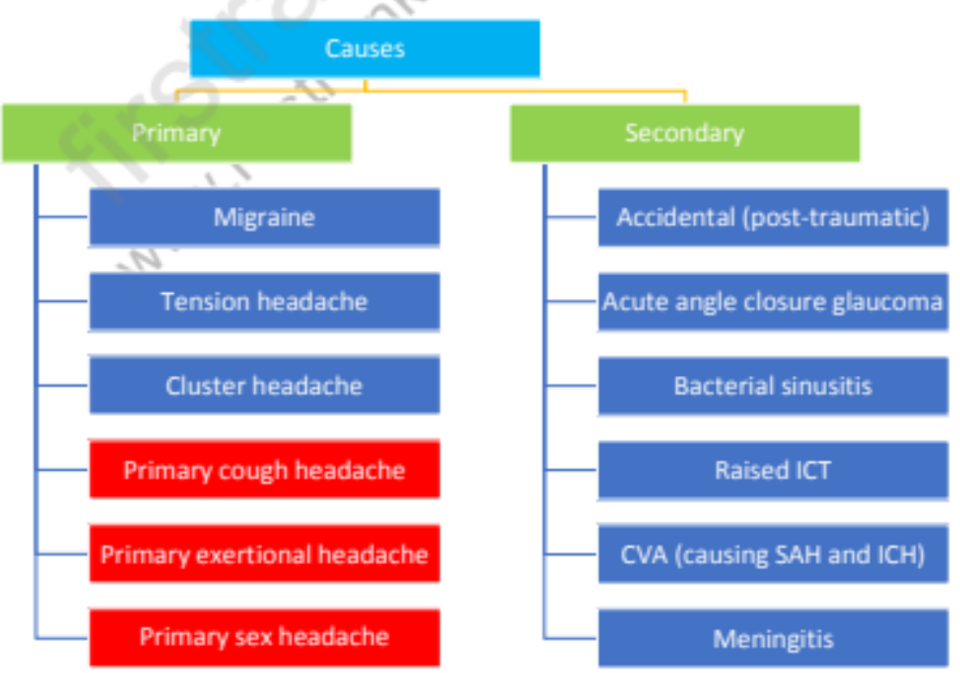
e. Midodrine (α-agonist).

2. Carotid sinus hypersensitivity:
   Permanent pacemaker
3. Cardiogenic syncope:
   a. Treat the underlying cause
   b. Implantable defibrillator in selected cases.
4. Orthostatic hypotension:
   a. Treat the underlying cause
   b. Avoid sudden standing.

Differential diagnosis

1. TLOC, but not due to global hypoperfusion:
   a. Hypoglycemia
   b. Primary seizure disorder
   c. Vertebro-basillar TIA
2. No TLOC, but transient impaired consciousness:
   a. Carotid TIA
   b. Cataplexy.

Headache



*Red colored causes should be ruled out first.

Migraine

It is a condition characterized by recurrent severe headache resulting from inappropriate vasodilation of cerebral blood vessels.

Pathogenesis

There is intermittent vasodilation of cerebral blood vessels which is mediated by different neurotransmitters.

Clinical features

1. Triggering factors:
   - Bright light
   - Loud sound
   - Chocolate/ Caffeine
   - Oral contraceptive pills
2. Aura:
   These are the symptoms which precede/ accompany the headache:
   - Visual symptoms:
     - Flashes of light
     - Zigzag lines
     - Tunnel vision.
3. Headache:
   - Site: Classically the hemicranium, but may be generalized
   - Character: Throbbing
   - Intensity: Moderate to severe
   - Duration: For several hours
   - Accompanied by: Nausea, vomiting
   - Patient prefers to lie in closed eye/ in a dark quiet room.

Atypical varieties of migraine

1. Basillar artery migraine (posterior circulation migraine):
   Headache + focal neurodeficit (Ex: Dys-equilibrium, Dysarthria, Temporary drowsiness)
2. Ophthalmoplegic migraine:
   Diplopia + Weakness of extraocular muscles.

Investigation

It is essentially a clinical diagnosis. In severe recurrent headache, a CT head is required to rule out any structural lesion.

Treatment

1. Avoid any triggering factor
2. Acute attack: Analgesic (Paracetamol/ NSAIDS) + Antiemetic
   Drug of choice is Sumatriptan.
3. Prophylactic/ preventive:
   A. Antiepileptic: Topiramate
      Amitriptyline
      - These 2 drugs are drug of choice.
   B. β-blocker: Propranolol
   C. Calcium channel blockers: Flunarizine.

Tension headache

Severe recurrent headache often precipitated by stress.

Pattern of headache:

- Site: generalized
- Nature: Band like sensation/ constricting sensation
- Severity: Moderate to severe
- Usually no aura.

Investigation:

CT head, if required

Treatment:

- Acute: Analgesic
- Prophylactic: ABC (as above).

Cluster headache

Typically occurs in middle aged males.

Pattern of headache:

- Site: Orbital, retro-orbital and temple regions
- Character: Moderate to severe
- Nasal symptoms:
  Running/ blocked nose
- Ocular symptoms:
  Watering from eyes/ ptosis/ temporary Horner's syndrome
- Why the name "cluster"?
  It occurs in spells when the patient develops severe headache which may vary in severity in different parts of the day and such spells recur almost every day for few days to weeks; then there is a symptom free period of few weeks after which the next cluster comes.

Investigation:

Ct head to rule out any structural lesion.

Treatment:

- Acute: Analgesic + High flow oxygen + Sumatriptan
- Prophylactic: Verapamil + Lithium.

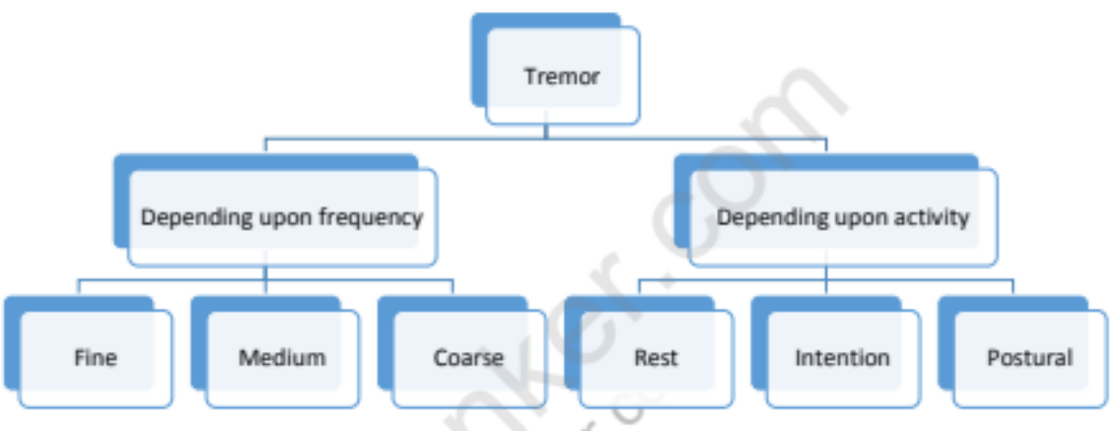
Extras

Involuntary movements

Tremor

It is a *rhythmic oscillatory movement* of different parts of the body due to repeated contractions and relaxations of a group of muscles.

Classification/ types



Postural tremor: Usually occurs when tremulous part of the body is kept in a sustained posture.

Intention tremor: Tremor increases while attempting to execute a voluntary activity.

Causes

Type	Causes
Fine/ intention/ postural	A. Alcohol excess/ withdrawal (chronic) B. Benign essential tremor C. Cigarette smoking D. Drugs: β2 agonist E. Endocrinopathy: Thyrotoxicosis.
Medium/ coarse/ rest	• Parkinsonism • Cerebellar lesion.

Clinical features

- Affected part of the body shows tremor: type and aggravating factor of which depends on the underlying disease
- Specific features of the underlying disease.

Investigation

1. TSH
2. Relevant investigations to assess the underlying disease
3. Stopping of any triggering factor.

Special note: Benign essential tremor

History:

Family history of tremor usually present.

Clinical features:

Description of tremor:

- Bilaterally symmetrical
- Fine tremor
- Usually affects upper limbs, head, tongue but usually spares trunk and lower limbs
- Aggravated by stress
- Temporarily relieved by alcohol.

Treatment:

- Drug of choice: Propranolol
- If fails: Primidone.

Chorea

It is an abnormal involuntary movement characterized by *non-repetitive jerky movement* particularly affecting the extremities.

Site of lesion:

Caudate nucleus

Causes:

1. Sydenham's chorea
2. Huntington's disease
3. Wilson's disease
4. Chorea gravidarum.

Athetosis

It is an abnormal movement characterized by *slow sinuous, often fine movement* particularly affecting the extremities.

Site of lesion: Putamen

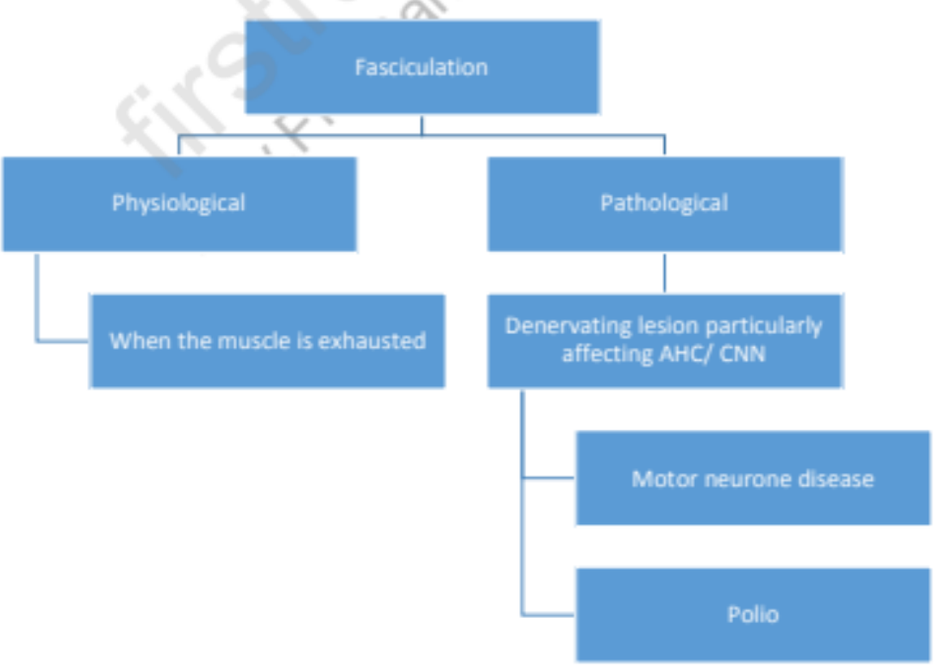
Causes:

1. Wilson's disease
2. Perinatal hypoxia
3. Kernicterus.

Fasciculation

Abnormal twitching movement involving few muscle fibres.

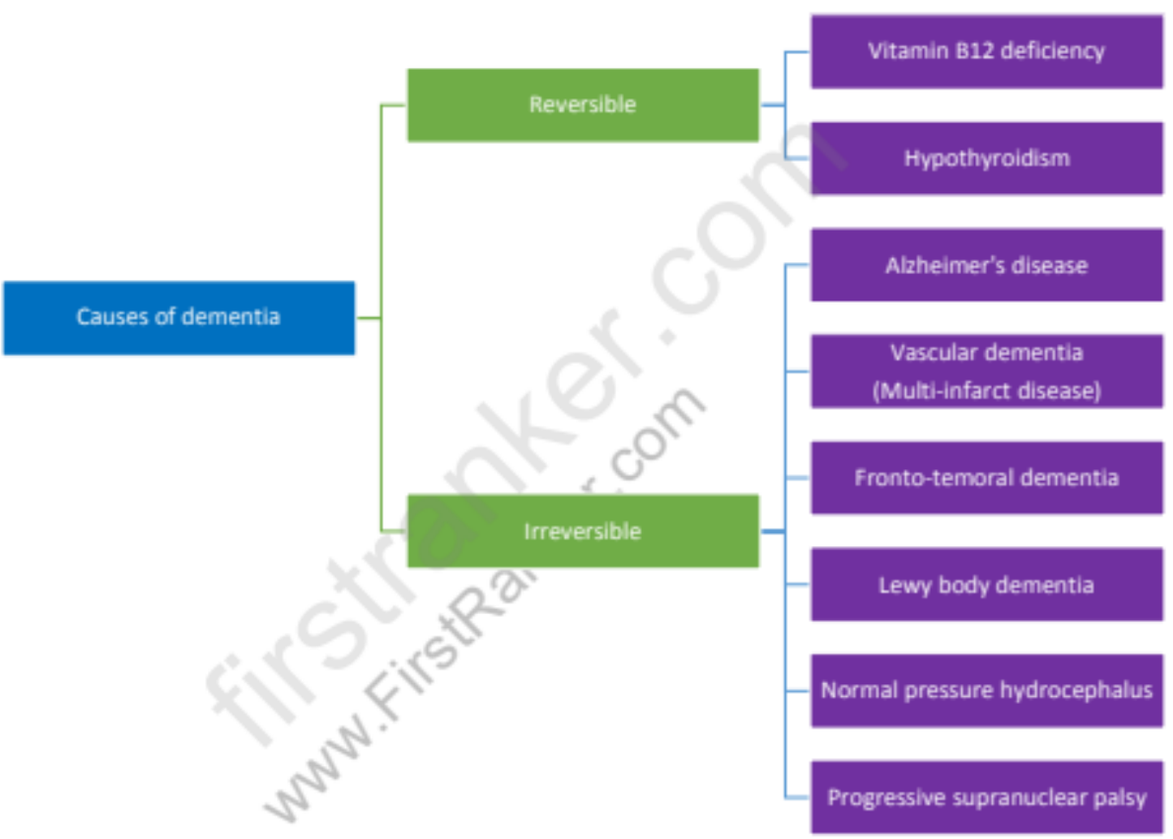
Types



Dementia

It is a state of cognitive impairment which is usually slowly progressive and often causes social and occupational dysfunction.

Causes



Clinical features

1. Short time memory loss
2. Inability to name an object (nominal aphasia)
3. Visuo-spatial dysfunction:
   a. Inability to navigate
   b. Gets lost in a known surrounding
4. Executive dysfunction:

a. Easy distractibility
b. Lack of concentration

5. Apraxia (inability to execute a motor activity in spite of normal power)
6. Along with these manifestations, patients often become delirious
7. Patient often develops apathy (indifference to others)
8. Specific signs and symptoms of the underlying disease
9. "Mini-mental state examination" score <20.

Investigation

1. Dementia screen:
   a. Blood: Hb, TC, DC, CRP/ESR
   b. Serum vitamin B12
   c. Serum TSH.
2. CT head.

Treatment

1. Non-pharmacological management:
   Family and social support
2. Pharmacological management:
   a. Treatment of any reversible cause
   b. For dementia:
      Anticholinesterase:
      - ✓ Donepezil
      - ✓ Rivastigmine

      NMDA receptor blocker:
      Memantine.