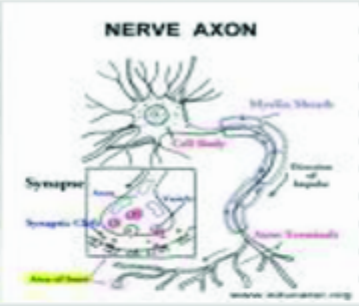

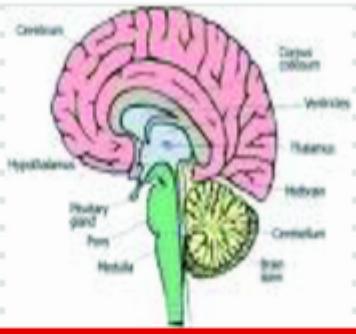

SPOTLIGHTS ON NEUROLOGY

BY

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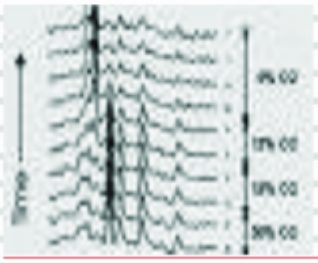
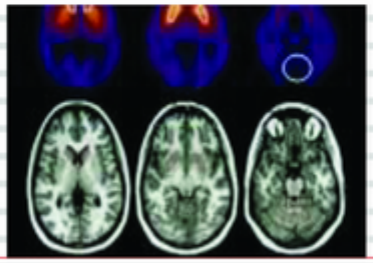




General & Anatomical features of the nervous system

-**Neuroanatomy**: is the study of the form of the nervous system

-**Neurophysiology** : is the study of the function of the nervous system

-**Neuropathology**: is the study of the nature & extent of the lesions

-**Neuroradiology** :is the study of the various modes of investigation used in nervous system

-**Neuropediatrics**: is the study of the diseases of the nervous system in pediatrics

-**Neuro ophthalmology:** is the study of the visual system & ocular nerves disorders in relation to the nervous system

Neurology

is the science & technique of studying the site ,causes & nature of the nervous system diseases with their diagnosis & therapy.

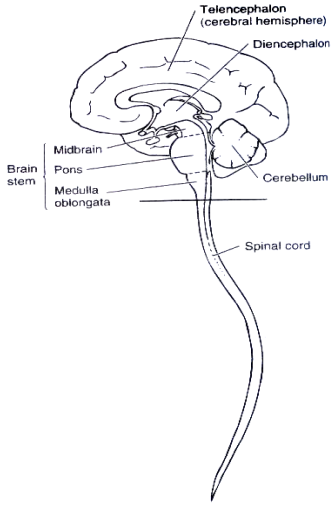
Anatomical features of the nervous system

There are **two main divisions** of the nervous system.

(1)-Central Nervous System(CNS) : Fig(1)

- Brain
- Spinal Cord

Both of them are enclosed & protected in bone (skull &vertebral column), protective coverings (meninges) & fluid filled spaces (CSF).



Fig(1)

(2)-Peripheral nervous system:

- Cranial nerves & their nuclei
- Roots-- Spinal nerves…,…till the muscles

The Central Nervous System(CNS)

Structural units of the CNS:

The Brain

- Is nearly about 2% of the total body weight (1.5-2 kg in adult) ,contains billions of neurons (nerve cells).

Nerve cells with common form ,function & connections within the CNS are grouped together to form **neuclei**, outside the CNS are grouped together to form **ganglia.**

- Brain is formed of:

(A)cerebrum (B) brain stem (C) cerebellum

(A) The cerebrum

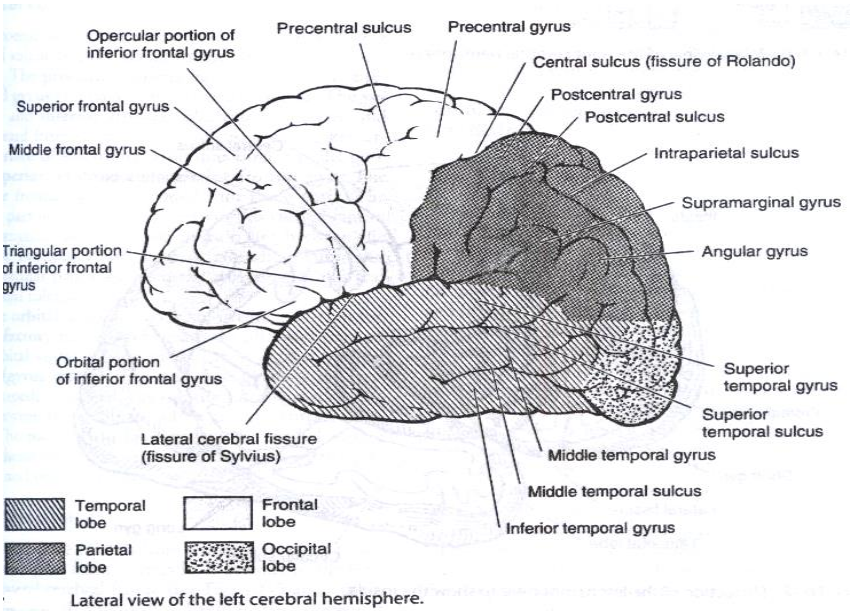
Is composed of two cerebral hemispheres with outer grey matter (cerebral cortex) composed of nerve cells , and inner sub cortical white matter composed of nerve fibers.

The two cerebral hemispheres are connected to each other by the corpus callosum.

the interconnections between various parts of the brain are through horizontal connections called commissures , and through bundles or tracts

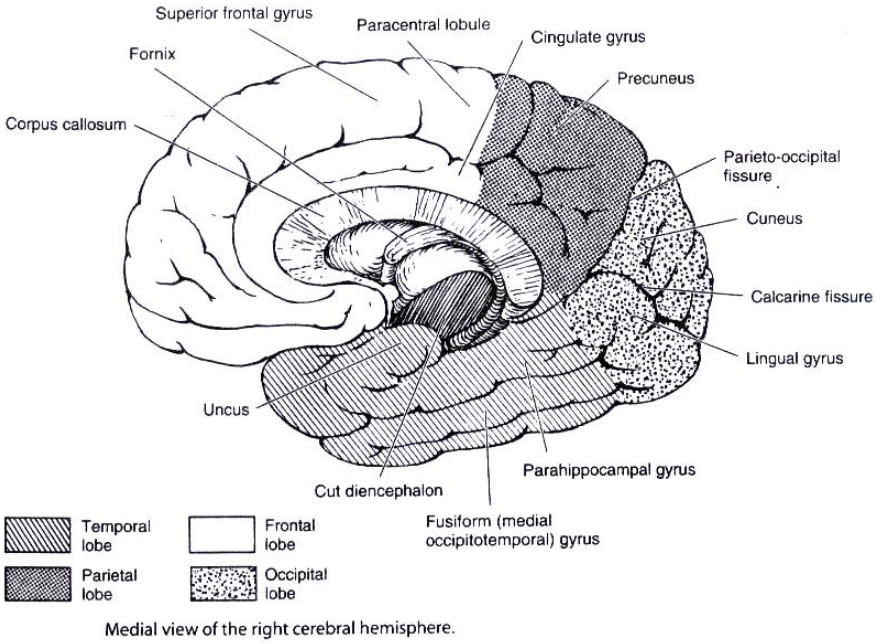
-**The Cerebral cortex** : Fig(2) ,(3) and (4)

The cortex is divided functionally & anatomically into lobes &areas of specific function, with thickness of 2- 4.5 cm



Lateral view of the left cerebral hemisphere.

Fig(2)

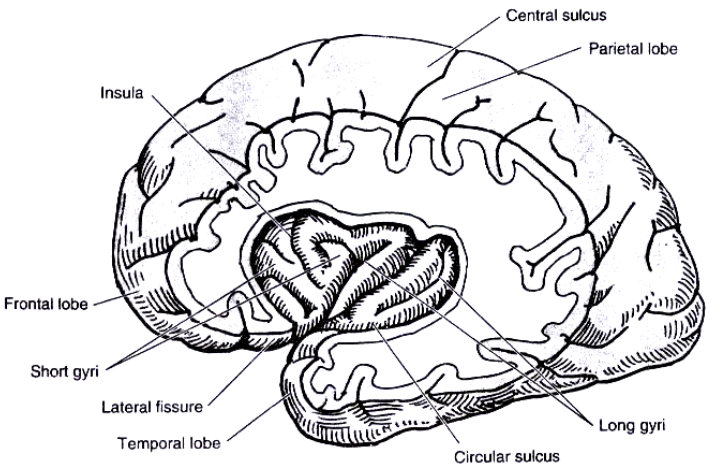


Medial view of the right cerebral hemisphere.

Fig(3)

(1)-**The Frontal lobe** :

- Concerned mainly with control of movements.
- Lies anterior to the central sulcus & contains



Dissection of the left hemisphere to show the insula.

Fig(4)

a- Motor area (4):

In the floor of the central sulcus ,it is concerned with initiation of the voluntary motor activity of the opposite half of the body.

b- Premotor area (6):

In the anterior part of the precentral gyrus,it shares the function of area 4

c- Motor speech center: (Broca's area 44):

In the middle part of the inferior frontal gyrus of the dominant hemisphere

d- Writing center (Exner's area 45):

Lies in the posterior part of the middle frontal gyrus

e- Area 8 of voluntary conjugate eye movement

Lies in the posterior part of the middle frontal gyrus ,it causes conjugate eye movements to the opposite side of stimulaton

f- Prefrontal area:

Occupies the anterior pole ,it is concerned with higher mental functions (memory ,orientation ,thinking, intelligence & personality.)

(2)-**The Parietal lobe:**

Lies behind the central sulcus & concerned mainly with sensation.

Areas of particular importance include;

a. Cortical sensory areas (1,2,3):

In the posterior central gyrus ,concerned with perception of cortical sensations from the opposite half of the body

b. Visual psychic area (39) of speech :

In the dominant hemisphere , concerned with recognition of the written speech

(3)-**The temporal lobe** :

Lies below the lateral sulcus (fissure of sylvius),includes the following areas,

A .Auditory sensory area (41,42):

lies in the superior temporal gyrus, concerned with hearing

B .Auditory psychic area(22):

lies posterior to the auditory sensory area in the dominant hemisphere,it is concerned with recognition of sounds

C .Uncus:

Lies in the medial temporal lobe ,concerned with smell & memory

(4)- **The occipital lobe:**

Lies behind the parito-occipital sulcus ,concerned mainly with vision, & contains, .

a) *Visual sensory area (17):* concerned with reception of vision

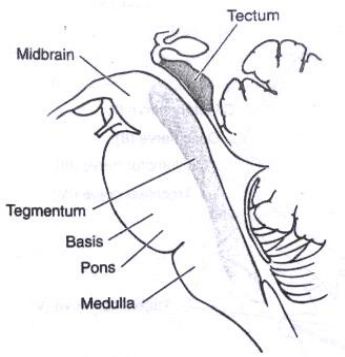
b) *Visual psychic area (18 & 19) :* Concerned with recognition of what is seen by area 17

N.B

‣In the depth of each cerebral hemisphere ,there are certain principal grey nuclei:

-Thalamus , hypothalamus

-Basal ganglia (caudate nucleus ,lentiform nucleus ,globus pallidus & putamen)



(B) The brain stem Fig(5)

Consists of three parts from above –below , Mid brain, Pons & Medulla.

‣ Is largely made up of ascending , descending & decussating tracts which join the different parts of the brain and spinal cord. together with the brain stem , and the brain stem is connected with the cerebellum which lies behind it through three cerebellar peduncles.

The brain stem in addition **contains important structures such as : Fig(5)

(1) The cranial nerve nuclei which are :

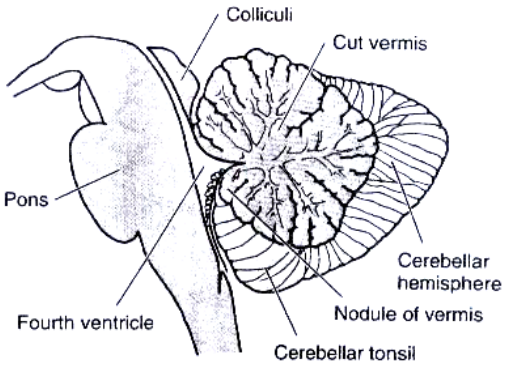
-Nuclei of cranial nerves (3) oculomotor & (4) trochlear in the *mid brain* .

-Nuclei of cranial nerves (5) trigeminal ,(6) abducens. (7) facial & (8) vestibulo cochlear in the *pons.*

-Nuclei of cranial nerves (9) glossopharyngeal ,(10) vagus, (11) spinal accessory & (12) hypoglossal are situated in the *medulla.*

(2) The Reticular formation which is related to sleep, consciousness , behaviour & memory.

(3)Respiratory ,Cardiac &Vomiting centers .



(C) The cerebellum ; Fig(6)→

Anatomically, consists of two cerebellar hemispheres and three cerebellar peduncles that connect it with the brain stem

Functionally, it can be divided into- 3- divisions:

- Archicerebellum → concerned with equilibrium.

- Paleocerebellum → concerned with muscle tone & automatic movements.
- Neocerebellum → concerned with coordination of fine movements.

The spinal cord " (myelon , medulla spinalis) " Fig(7)

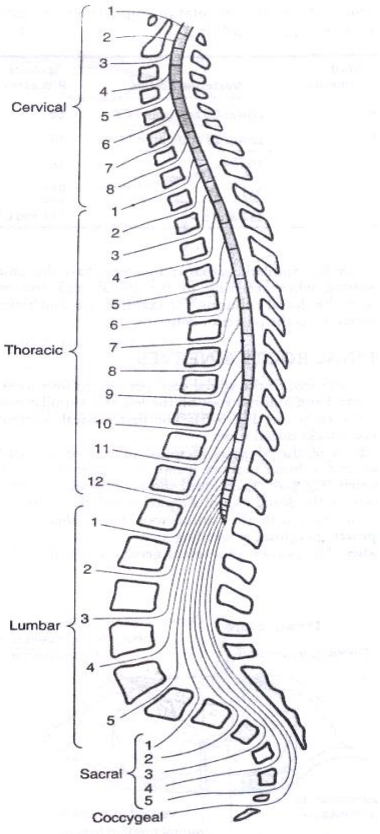
* Is an elongated structure , about 42-45 cm long in adults, its upper end continuous with the brain stem at the foramen magnum & extends to the lower border of the 1st lumber vertebra.
* The rest of the spinal canal is filled with filum terminale "extension of pia matter ".
* The lower tapering border of the spinal cord is termed conus medullaris.
* The cord is divided into 31 segments :

(8)cervical , (12)thoracic, (5) lumbar, (5)sacral & (1) coccygeal ,

* There are tow enlargements; cervical & lumbar enlargements with the brachial plexus & lumbar plexus originate from those enlargements in order.
* In contrast to the brain ,the cord consists of inner H shaped grey matter surrounded by the white matter.

-The grey matter consists of 2 posterior horns "sensory" & 2 anterior horns ":motor" and intermediolateral horns in the thoracic & sacral regions (for sympathetic & parasympathetic efferents).

-The white matter contains many ascending & descending tracts.



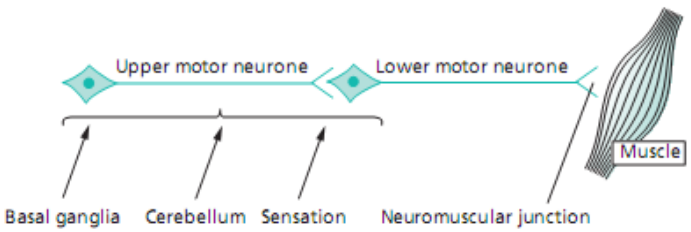
Fig(7)

The peripheral nervous system

Consisting of the cranial nerves with their nuclei , anterior horn cells(AHC) , spinal nerves peripheral nerves, neuromuscular junctions & the muscles .

N.B:

- From the cerebral cortex to the spinal cord this is called upper motor neurons
- From the AHC till the muscles this is called lower motor neurons , both of them has characteristic clinical features. Fig(8)→



NB: Organizations of movements in the motor system

For normal voluntary motor activity , the following parts of the nervous system must be normal and cooperate :

1) The motor areas, corticospinal tract "pyramidal tract ;upper motor neurons" Fig(9).

2) The basal ganglia .

3)The cerebellum .

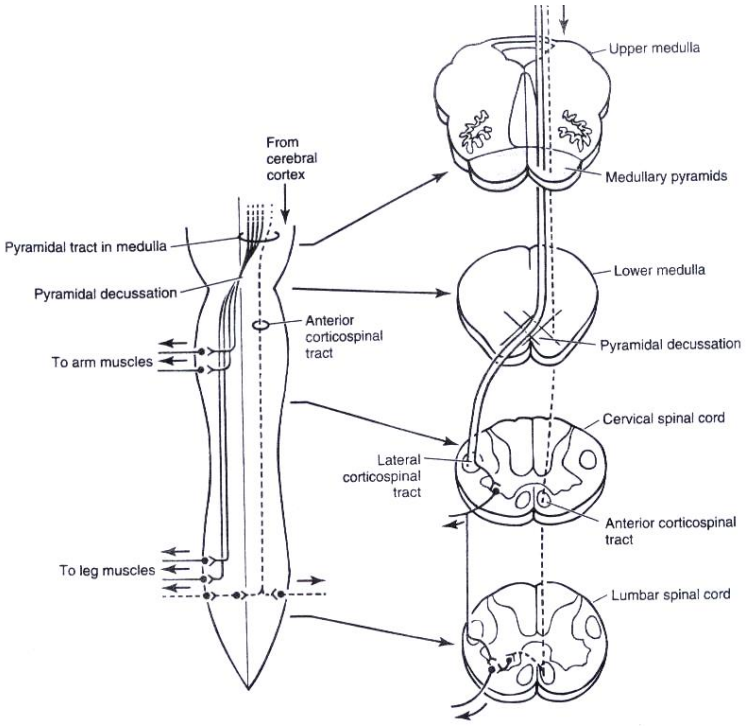
4) The lower motor neurons .

The components of the motor system:

The pyramidal tract :

Constitutes the descending pathway which arise from the giant cortical Betz cells of the motor & pre motor areas . There is a wide presentation of the body in the cortex & it is presented upside down . The nerve fibers descend in the cerebral hemisphere in a collecting manner

-The pyramidal tract in human contains about 1 million fibers , 94% of which are myelinated →the descending fibers



Fig(9)

comes together in **the corona radiate** → then to be collected more in the **Internal capsule** (lies deep in the cerebral hemisphere) where it occupies the post 1/3 of the anterior limb ,the genu & the anterior 2/3 of the posterior limb ,from the internal capsule →The tract descends through the middle 3/5 of **the cerebral peduncle** to enter **the mid brain** →Then to **the pons** where it is broken into bundles by the transverse pontine fibers → Then to the medulla where it occupies an anterior prominence **(pyramid)**. Throughout the brain stem the pyramidal tract gives off the corticobulbar tract which supply the nuclei of the motor cranial nerves of the opposite side

In the lower part of the medulla > 80% of the pyramidal tract **decussate**s (cross to the other side) to form **the crossed tract** which descends in the lateral column of the spinal cord on the opposite

side , the remaining **uncrossed fibers** descend on the same side in the anterior column forming the direct pyramidal tract.

> ***NB:*** All the cranial nerve nuclei receive bilateral cortical supply except the lower half of the facial & the hypoglossal nuclei which receive only contralateral cortical supply

The basal ganglia & the cerebellum

Informations for normal motor activities before passing to the spinal cord & the final organized station "the lower motor neurons" must pass at first through the basal ganglia and the cerebellum through the different connections between the cerebral cortex-brain stem-spinal cord.

* ***The basal ganglia-*** has the following influence on the motor system:
  - Regulation of posture.
  - Plays a vital role in initiating movements "lesions → akinisia or bradykinisia.
  - Gives off smooth coordinating voluntary motor activity.
  - Regulation of muscle tone and voluntary movements " lesions → rigidity & tremors".
* ***The cerebellum*** - shares in the organization of the motor system activity through:
  - Regulation of postural reflexes and equilibrium " lesions → unsteadiness"
  - Regulation of muscle tone " lesions → hypotonia"
  - Regulation and smooth coordination of limb movements" lesions → ataxic movements"

The lower motor neurons:

Forming the final process in organizing motor activity, it constitutes the following:

- Nuclei of motor cranial nerves.
- Anterior horn cells of the spinal cord → giving off the anterior roots which form with the afferent posterior roots the spinal nerves → which will form the peripheral nerves→ neuromuscular junction → into the muscles.

IN CONCLUSION:

In response to the afferent stimuli or desire to do voluntary activity, the stimuli will travel through the afferents up to the cortex, passing through the spinal cord, then, from the cortical motor area→ efferent stimuli will descend→ corona radiata→ internal capsule→ spinal cord→ lower motor neurons to initiate the desired movement, through activation of specific AHC and the motor units they supply , at the same time, information about the required movement reach the basal ganglia and the cerebellum to regulate, coordinate, smooth, & counteract any undesired movements, simultaneously, as the required muscles "agonist" are being stimulated to contract, the synergists "helper" contract to assist, the fixators contract to fix and the antagonists relax. Once the required movement have begun, it is then continuously modified and maintained if needed through continuous afferent sensory impulses arriving the motor system components from the "deep sensory receptors" eyes, and labyrinths. Therefore, lesions in any of the above

parts involved in initiating and controlling motor activity will result in disorganized, abnormal movements.

Evaluation of neurological patient

Neurology sheet.

A-HISTORY

I-Personal history:

-Name -Age

-Sex -Marital status:

-Occupation -Residence:

-Special habits -Handedness:

II-Complaint:

III-Present history:

1-Analysis of the complaint: ***Onset*** (sudden,acute,subacute,gradual)

Course (progressive,regressive,intermittent,remittent)

Duration.

Precipitating ,or relieving factors.

2-Symptoms suggesting cranial nerves affection:

Olfactory N: Smell disoders either quantity or quality.

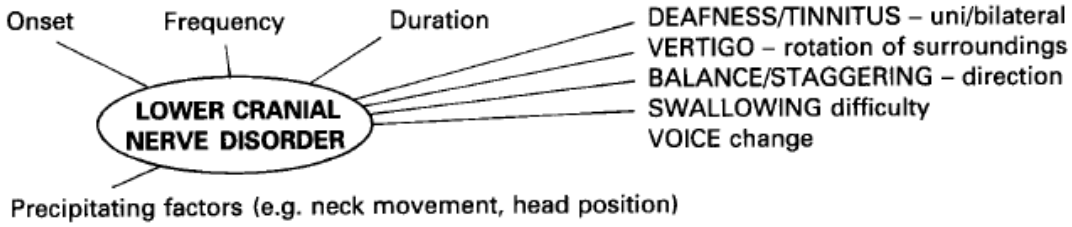
Optic N : Disorders of visual acuity,visual fields,blurred vision.

Ocular nerves: Diplopia,squint.ptosis

Trigeminal N : Disorders of mastication ,facial sensations.

Facial N: Accumulation of food,drippling of saliva,

Lower cranial nerves ,see Fig(10)



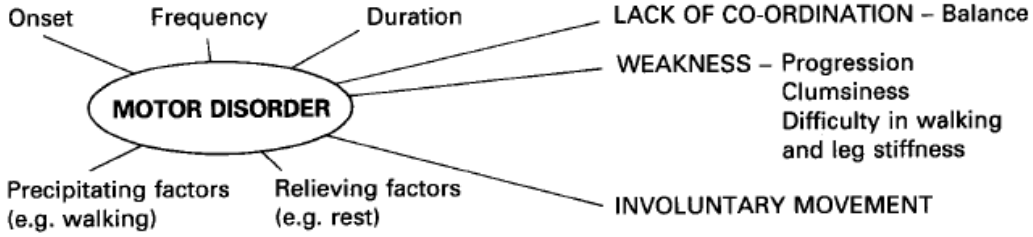
Fig(10)

3- Symptoms suggesting motor system affection Fig(11)

Unilateral or bilateral

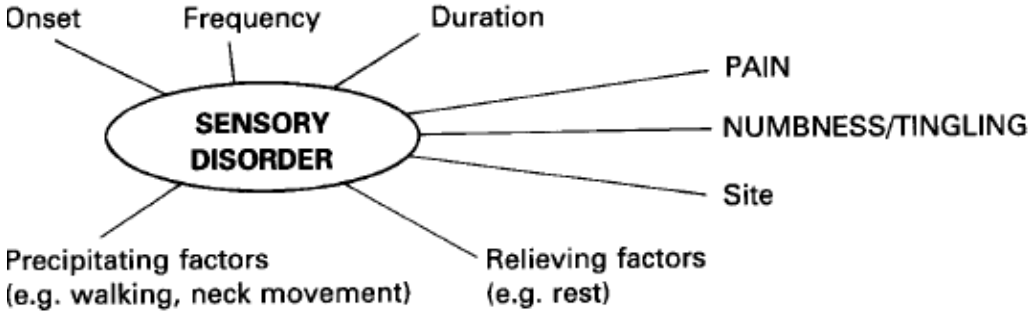
symmetrical or asymmetrical

distal or proximal,diurnal variation



Fig(11)

4- Symptoms suggesting sensory system affection Fig(12)



Fig(12)

5- Symptoms suggesting ↑ ICT

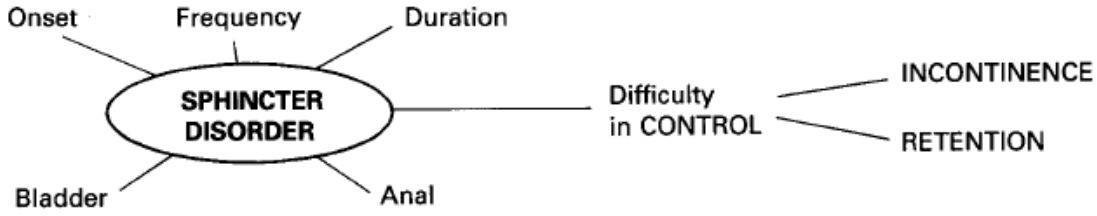
Headache (characters)

Vomiting (characters)

Blurring of vision

6- symptoms suggesting sphincteric disturbances Fig(13)

Incontinence,precipitancy,urgency,hesitancy,stress incontinence and retention



Fig(13)

IV-Past history:

Similar conditions, truma,road traffic accidents,operations,drug intake of neurologic importance,fever of neurologic importance,cardiac diseases,T.B,diabetes and hypertension

V-Family history:

Simillar conditions,consanguinity,health of the partener.

B-EXAMINATION:

I-General examination

-General appearance

-Pulse - B.P -Temperature -Respiratory rate -Chest ,Heart ,Abdomen

II-Neurological Examination

1-Mental state examination

-State of consciousness

-Orientation for time ,place ,person

-Memory (immediate , recent, remote)

-Mood, affect,behavior & thought content.

-Intelligence

2-Speech examination: *Examine comprehension, fluency, repetition.naming, word finding*

-Aphasia: sensory, motor, mixed….etc

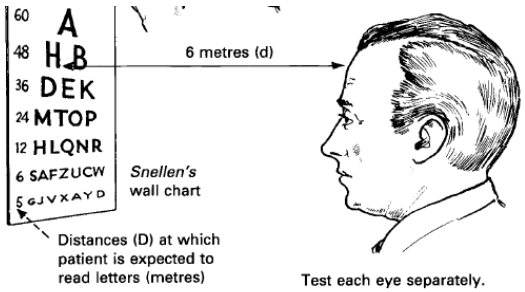
-Dysarthria: slurred , staccato , monotonous …etc.(see page ,30)

3-Cranial nerve examination:

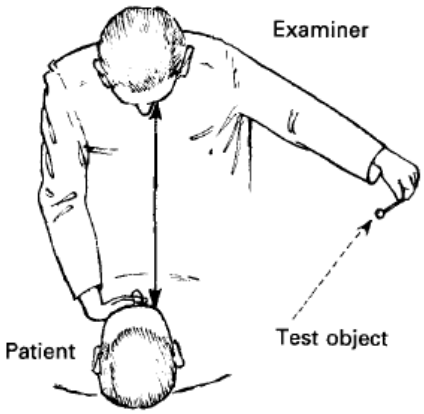
-(1) Olfactory nerve. Test each nostril for smell disorders either in quantity or quality.

-(2) Optic nerve: Fig(14,15)

-Visual acuity -Visual field-Fundus examination



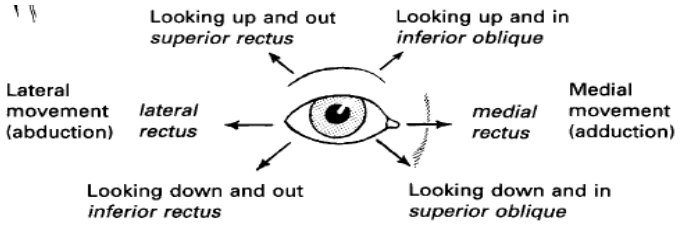
Fig(14)



Fig(15)

-(3) 3,4,6 ocular nerves: Fig(16)

-Extra ocular movements, -Pupils,-Ptosis



Fig(16)

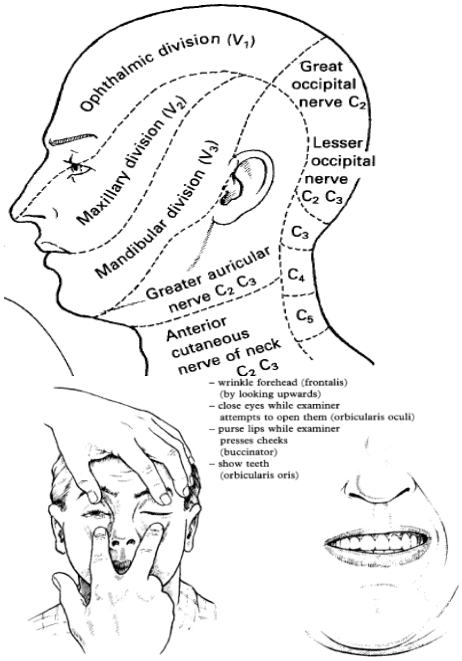
-(5) Trigeminal nerve: → Fig(17)

-Motor: temporalis,masseter,pterygoids.

-Sensory: over the face.

-Reflexes:jaw,corneal, conjunctival.

-(7) Facial nerve: Fig(18) →



-Motor:frontalis,orbicularis occuli,

orbicularis oris, buccinator, retractor anguli.

-Sensory: taste over Ant. 2/3. of tongue .

-Reflexes: glabellar.corneal

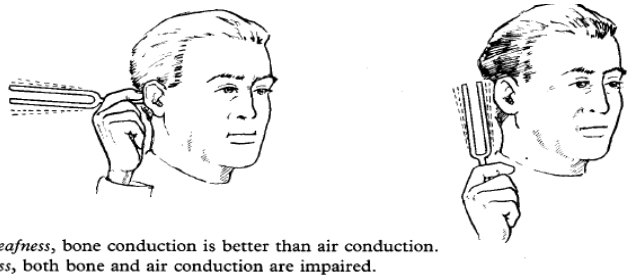
-(8) Cochleo-vestibular: Fig(19,20)

a) Cochlear part:

-Watch test , whispering sound ,

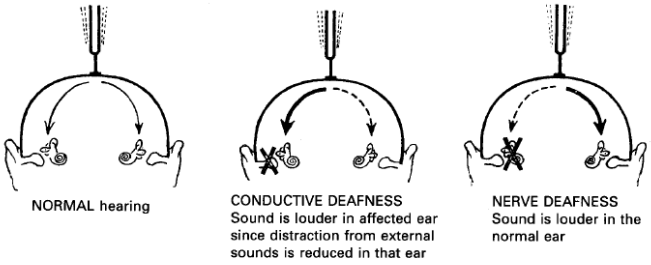
-Rinne's test Fig (19)→

-Weber's test Fig(20) ↓



In *conductive deafness*, bone conduction is better than air conduction.
In *nerve deafness*, both bone and air conduction are impaired.

Fig(19)



Fig(20)

b) Vestibular part:

-Caloric test , hallpick test ,rotating chair test

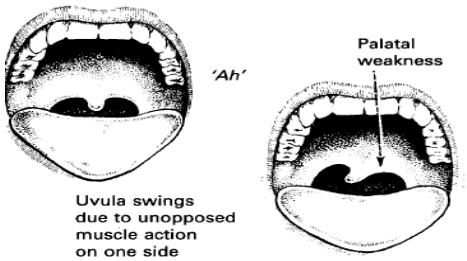
-Electronystagmography

-(9) & 10) Glossopharyngeal & vagus vagus nerves ----→ Fig(21)

-Position of uvula

-Palatal reflex

-gag reflex



-(11) Accessory nerve Fig(22,23) ↓

a) Spinal part:

←-Sternomastoid

-Trapezius →

b) Intracranial part - joins the vagus

-(12) Hypoglossal nerve: Fig(24)

-Tongue movement , wasting, deviation , fasiculations, abnormal movements.

Fig(24) →

4-Motor system examination:

1) Inspection: to detect:

-Wasting -Hypertrophy -trophic changes

-Fasiculations -Abnormal movements

-Skeletal deformities -Abnormal position

2) Muscle tone: Methods:

-Shaking method

-Passive flexion & extension

-Gower's method -lifting methode -

Abnormal muscle tone-(distal or proximal,flexors , extensors,adductors or abductors)

-Hypotonia

-Hypertonia → Spasticity or rigidity

3) Muscle power:

- Examine ULs & LLs. - Distal & proximal muscles. - Detect grade of muscle weakness:

grade 0→ No movment

grade 1→ flickering

grade 2→ movement with elimination of gravity

grade 3→ movement against gravity

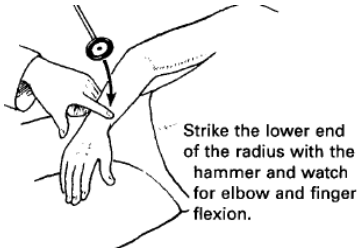
grade 4→ movement against resistance

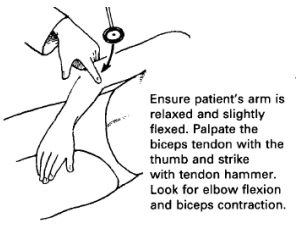
grade 5 → normal power

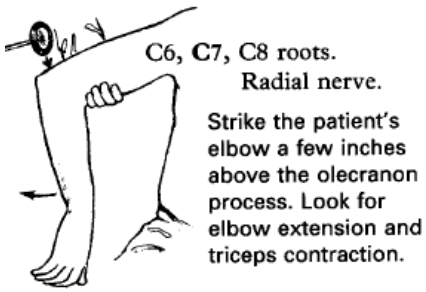
4) Reflexes: a) Deep reflexes:

●**Upper limb:** Fig(25) →-Brachioradialis reflex (C5,6) (a)

-Biceps reflex (C5,6) (b)







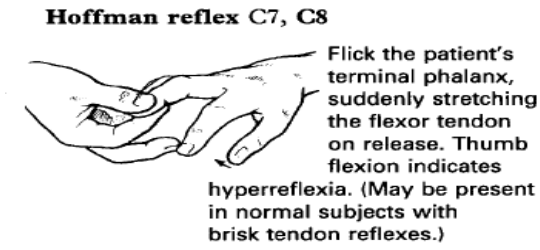
Fig(25)

(a) (b) (c)

-Triceps reflex (C6,7) (c)

-Pathological reflexes e.g inverted supinator ,

Hoffman's reflex. Fig(26)



Fig(26)

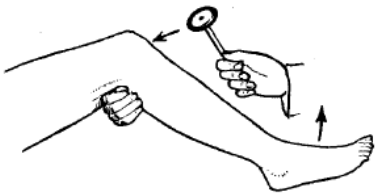
●**Lower limb:** Fig(27)

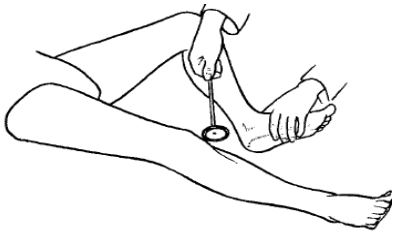
-knee reflex (L3,4) (a)

-Ankle reflex (S1) (b)

-Pathological reflexes e.g adductor,patellar,

- clonus





Fig(27) (a) (b)

b) Superficial reflexes:

●Abdominal reflexes (T6→T12)

●Cremasteric reflex (L1)

●Gluteal reflex (L4,5)

●Anal reflex (S3,4,5)

●Planter reflex (S1).Fig(28)

by one of the following methods:

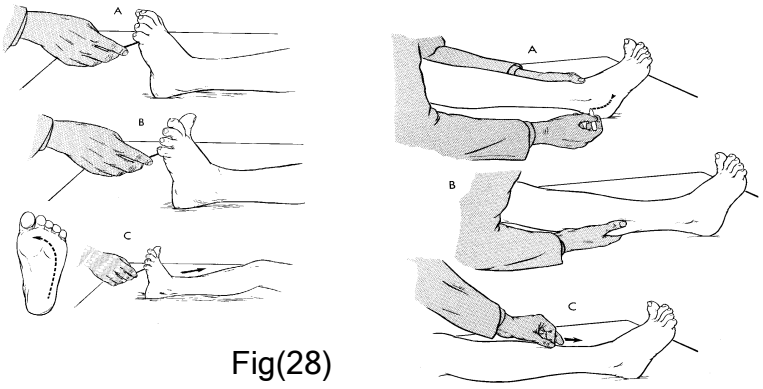
1) Babinski method

2) Shaddok's method

3) Schaefer's method

3) Gordon's method

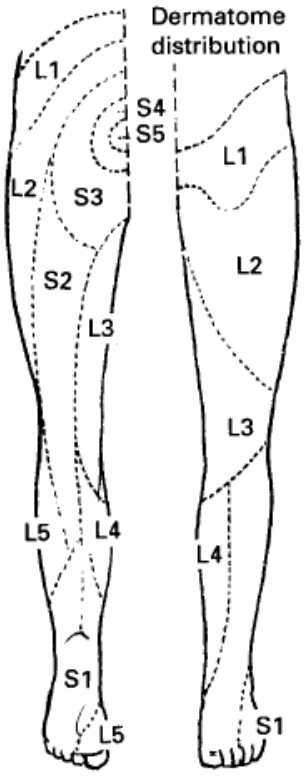
4) Oppenheim's method

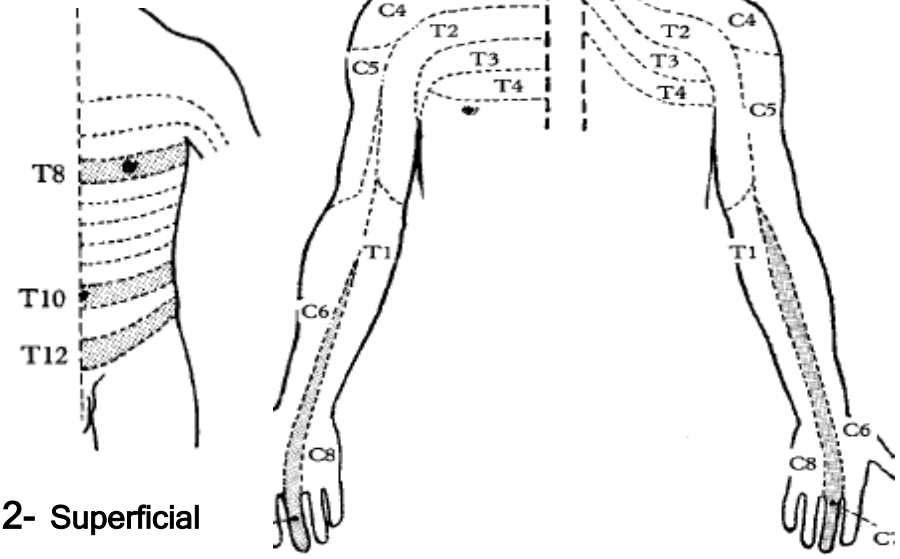


Fig(28)

5-Examination of the sensory system:

1- **Sensory supply of the body**. Fig(29)





Fig(29)

2- Superficial sensations:

including pain , temperature and touch.see patern of sensory loss,table (1)

3- Deep sensation include :

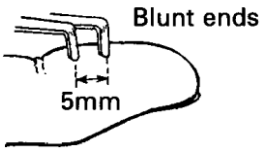
a) Vibration sense. - b) Nerve sense -c) Muscle sense (by pinching the calf)

d)-Joint sense(sense of position &movement) (fig 30, a and b) ↓

4- Cortical sensation:

a) Tactile localization

b) Two points discrimination …Fig(31)→



c) Stereognosis

d) Graphosthesia

e) Perceptual rivalry.

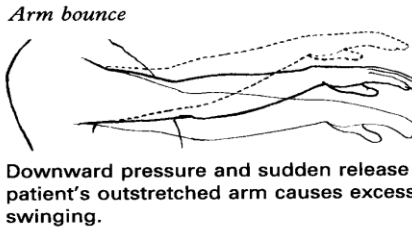
Patterns of sensory loss. Table (1)

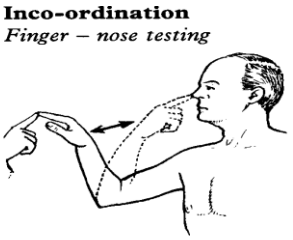
Pattern of sensory loss	Site of lesion
1- glove &stock hypothesia	Peripheral nerve
2-radicualr sensory loss	Root
3-saddle area loss	Conus
4-dissociated sensory loss	Unilateral cord lesion
5- sensory level	Extramedullary lesion
6-jacket sensory loss	Intramedullary lesion
7-crossed hemihypothesia	Lateral medullary syndrome.
8-hemihypothesia	Capsular &brain stem

6-Examination of coordination:

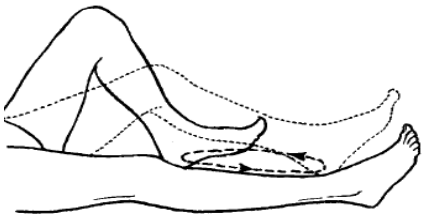
A- In the upper limb Fig(32 a)

1- Finger to nose test.
2- Finger to finger test.
3- Finger to doctor's finger test.
4- Adiadokokinesia or dysdiadokokinesia.
5- Rebound phenomenon.





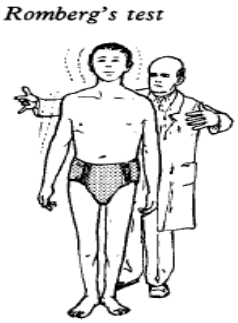
Fig(32a.)



Fig(32,b)

B- **In the lower limb:** Fig(32, b)

1- Heel to knee test. ,Walking .
2- Romberg test. Fig(32,c)



Fig(32,c)

7-Examination of the back &spine :

Examine for : - Tenderness, Swelling ,

- Deformity,Abnormal pigmentation

♣ Examination of cranium:

Size , shape , sutures &fontanelles.

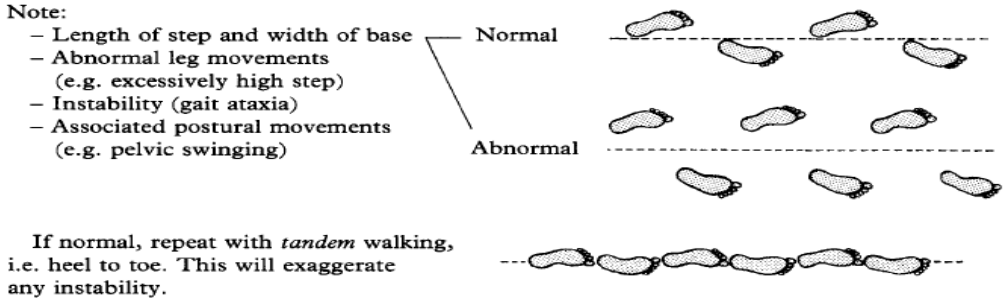
1- Bony bosses & tenderness.

2- Dilated veins , bruit & naevi.

♣ Examination of the neck:

Examine for : signs of meningeal irritation.

♣ Examination of the gait . Fig(33)→



INVESTIGATION

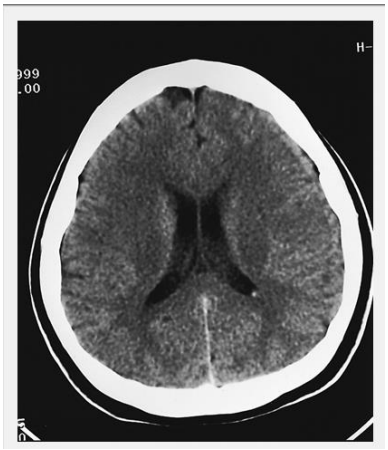
A-Imaging of the nervous system:

1-Computed tomography scanning (CT scan) Fig(34)

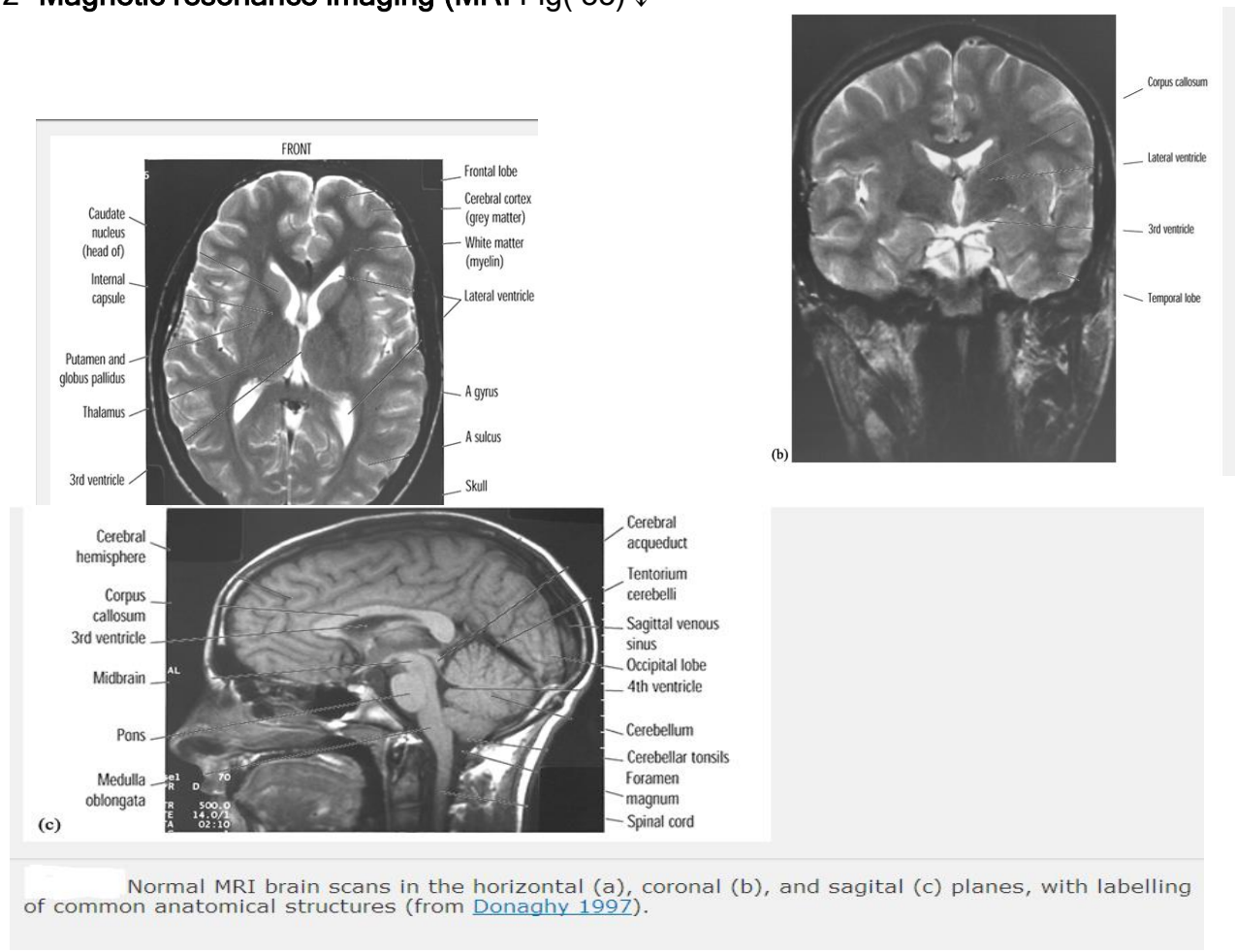
- Bone, calcification, or acute haematomas, are seen as bright areas (hyperdence)

Fig(34)

-Cerebrospinal fluid is dense dark,infarction less dark(hypodence)



2- **Magnetic resonance imaging (MRI** Fig(35) ↓



Normal MRI brain scans in the horizontal (a), coronal (b), and sagital (c) planes, with labelling of common anatomical structures (from Donaghy 1997).

MRI is more sensitive in small,deeply seated lesions.in demylinating & inflammatory lesions

3-Positron emission tomography(PET)

This imaging technique employs radioactive isotopes and can study a variety of functional or chemical features in normal and pathological brain which are inaccessible to other imaging modalities.

4- Imaging the cerebral circulation

-Transcranial Doppler sonography

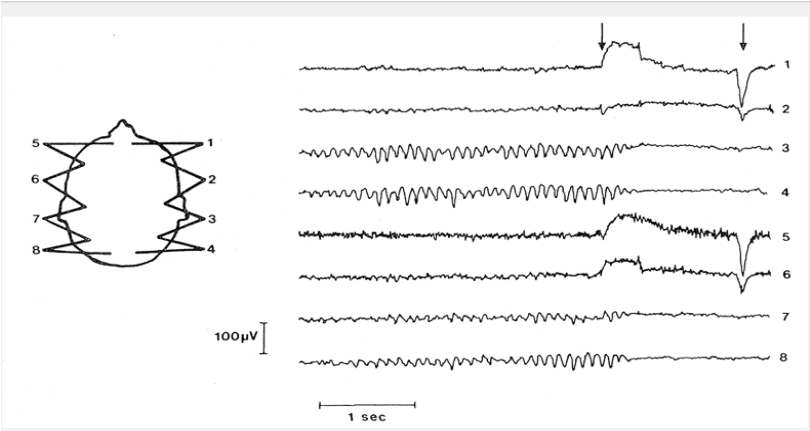
-MR angiography

-CT angiography .

B-Neurophysiology:

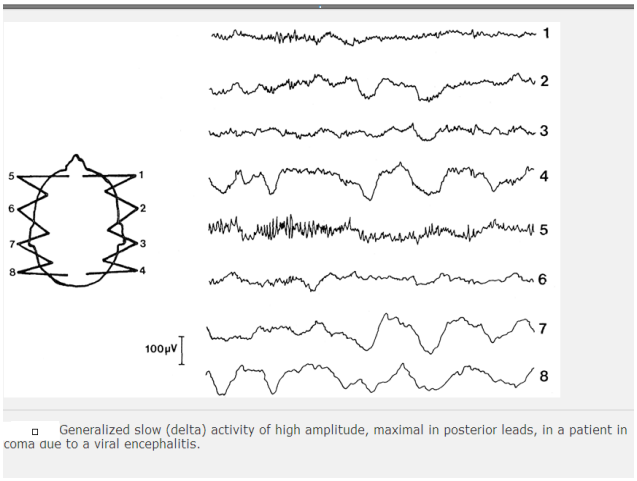
(1)-Electroencephalogram(EEG):

Fig(36)→

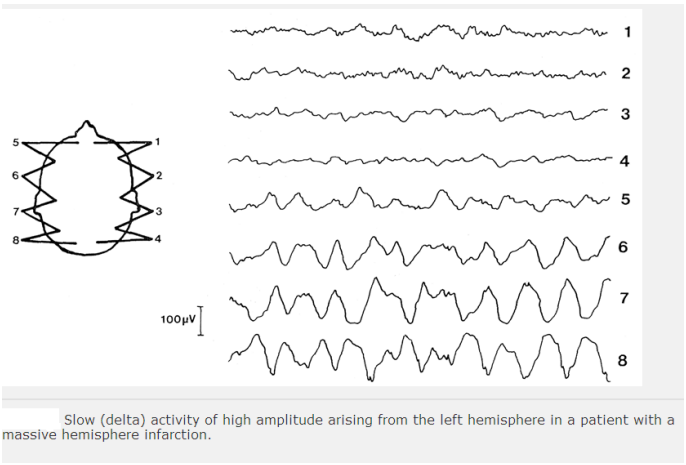


A normal EEG with eye-movement artefact marked by arrows.

Fig(37)



Generalized slow (delta) activity of high amplitude, maximal in posterior leads, in a patient in coma due to a viral encephalitis.



Slow (delta) activity of high amplitude arising from the left hemisphere in a patient with a massive hemisphere infarction.

-Spike discharges or sharp waves ..Fig(38 b)

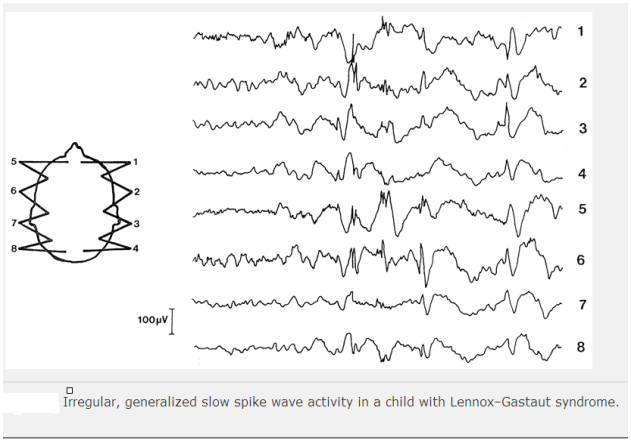
Are the characteristic interictal abnormality found in epileptic patients.

Sharp waves may be induced by drugs and alcohol withdrawal.

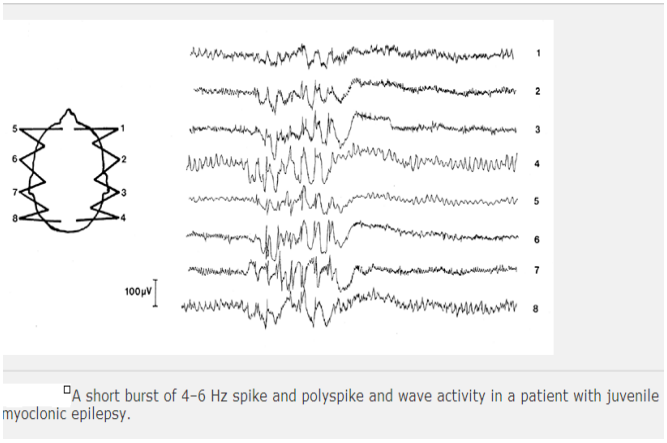
 # The EEG in *childhood absence epilepsy;*

an EEG that shows 3/second spike wave activity during a period of hyperventilation , hyperventilation being strongly provocative of spike wave activity in this and other generalized epilepsies.

Atypical absences are more commonly associated with slower, more irregular spike wave activity associated with a more abnormal EEG background Fig.(38 a)



Irregular, generalized slow spike wave activity in a child with Lennox–Gastaut syndrome.



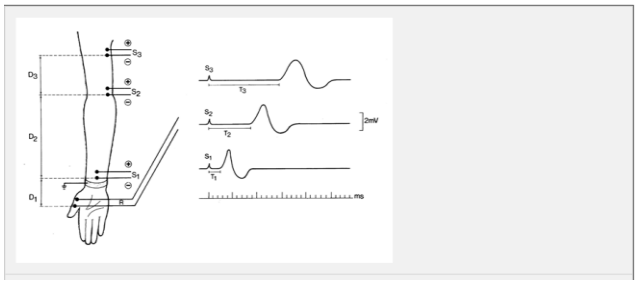
A short burst of 4–6 Hz spike and polyspike and wave activity in a patient with juvenile myoclonic epilepsy.

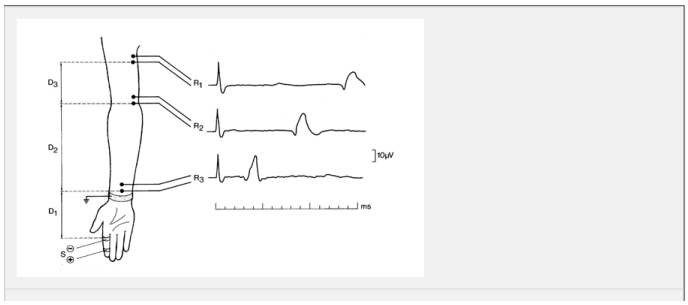
Fig(38) a b

(2)-Nerve conduction studies

Nerve conduction studies allow localization of compressive focal neuropathy and detection of polyneuropathy, with distinction between demyelinating, axonal degeneration and conduction block neuropathies.

-Motor conduction studies Fig(39)a



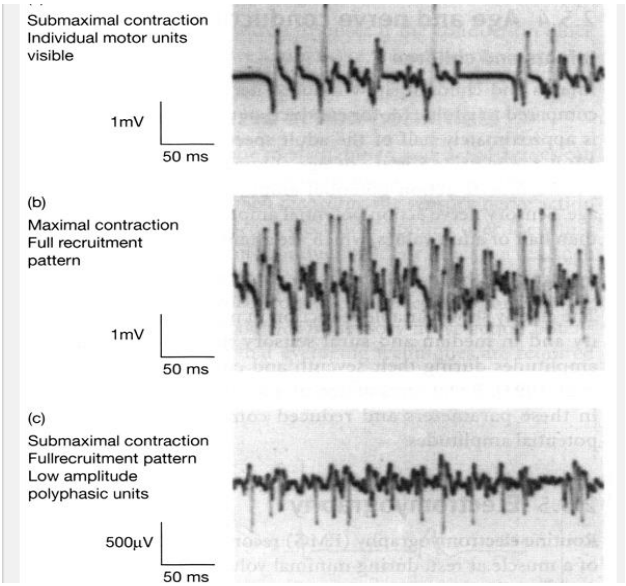


Fig(39) a → motor b →sensory

-Sensory nerve action potentials Fig(39)*b*

(3)-Electromyography(EMG): Fig(40)➵

Electromyography detects denervation of muscle, helps to distinguish between myopathic and neuropathic weakness, and is diagnostic of myotonias and neuromyotonias



(4)- Visual Evoked potentials:

For detection of various causes of optic neuropathy ,and visual pathways disorders.

(5)-Somatosensory evoked potentials:

The somatosensory evoked potential (SEP) is reduced or absent in the acute phase of demyelinating myelopathy involving the dorsal part of the cord, and this abnormality often persists after clinical recovery

(6) -Brainstem auditory evoked potentials:

(7) -Central motor conduction study:

(8) -Event-related potentials p300:

Event-related potentials reflect the summation of cognitive processes, including attention, memory, language, and responses

C-Cerebrospinal fluid:

Indications for lumbar puncture; →Table (2)

Broadly speaking, spinal (usually lumbar, as noted above) puncture is performed for either diagnostic or therapeutic purposes -- ↓ table (2)

Diagnostic	*Therapeutic*
Emergency 1-Suspected infection Meningitis (viral, bacterial, fungal) Encephalitis 2-Suspected subarachnoid haemorrhage 3-Unexplained confusional state	1-Reduction of CSF pressure 2-Instillation of drugs and contrast media 3-Spinal anaesthesia
Non-emergency 1-Multiple sclerosis 2-Malignancy -Carcinomatous meningitis - Lymphoma 3-Sarcoidosis 4-Demyelinating polyneuropathy 5-Infection (syphilis, chronic fungal) 6-CNS vasculitis 7-Benign intracranial hypertension	

Table -2

-Contraindications of lumbar puncture;

1-Raised CSF pressure due to an intracranial space occupying lesion

2-Local lumbar skin sepsis

3-Warfarin or heparin therapy

4-Coagulation defect

5-Spinal block

Headache

Defintion:

■ Headache means pain in the head, It is a common symptom in clinical practice in 93% of males & 99% of females.

■ It is a symptom of a disease which is in need for searching for the causative disease which may be serious.

■ Every Patient with headache requires firstly history in detail with attention to the following points:

1-Duration of suffering from headache.

2-Is it increasing in severity, constant or paroxysmal?.

3-Side and site of headache.

4-Duration of each attack.

5-Precipitating factors e.g. cough, straining, psychic stress…etc

6-Relieving factors e.g. vomiting, analgesics,….etc

7-Character of headache eg; pulsating, throbbing, tingling, explosive or vague character …. ect

8- Associating symptoms eg; nausea, vomiting, blurring of vision, vertigo, insomnia,..ect

9- History of head trauma, nasal obstruction or discharge, visual disorders, teeth problems ,..ect.

10- Is the patient anxious, tense or depressed ?

11- Presence of signs of systemic disease e.g, fever, anaemia

The Mode of production of Headache:

- All the tissues covering the cranium are pain sensitive eg; arteries, muscles and pericranium
- Skull bone itself is insensitive to pain.
- Intracranial pain sensitive structures:
  - Venous sinuses - Dura matter - Cerebral arteries
  - II, III, V, IX & X cranial nerves

The Main factors which can cause headache:

1. Inflammation in pain sensitive structure
2. Referred pain (e.g; sinuses, cerevical spondylosis)
3. Meningeal irritation (meningism, meningitis)
4. Traction on or dilatation of blood vessels e.g; anaemia
5. Pressure upon or distortion of pain sensitive structure e.g; tumors or other SOL.
6. Psychological causes,the most common nowadays

Classification of Headache:

Acording to headache classification committee of the international headache society, headache was classified as the following:

1. **Migraine**
   1. Migraine without aura
   2. Migraine with aura
   3. Complicated migraine
2. **Tension type headache**
3. **Cluster headache(Histamine cephalgia, chronic paroxysmal hemicrania)**
4. Headache associated with **vascular disorders** eg; CVS, AVM, arteritis, anemia, venous thrombosis and hypertension…
5. Headache associated with **head trauma**
   1. Acute post-traumatic headache

2. Chronic post-traumatic headache

6. Headache associated with **non vascular-intracranial disorders** eg; benign increased ICT, hydrocephalus, infection, neoplasm…
7. Headache associated with **non cephalic infection (viral, bacterial, systemic or focal)**
8. Headache associated with **metabolic disorders** eg; hypoxia, hypoglycemia, hypercapenia, hyperthyroidism, dialysis…
9. Headache **Referred** from diseases of

- skull bone
- Neck eg; cervical spondylosis
- Eyes eg; glucoma,refractive errors
- Ears eg; otitis media , otitis externa
- Nose and sinuses; sinusitis
- Teeth, jaw and related structure disorders
- Tempromandibular joint diseases

10. **Cranial neuralgia**
    1. Trigeminal neuralgia
    2. Glossopharyngeal neuralgia
    3. Occiptal neuralgia
    4. Optic and retrobulbar neuritis

Migraine

- Migraine; common in females(15-20%) than males(5-10%)
- Strong family history is present
- Common in urban residents
- Onset: at /or shortly after puberty
  - Less common before puberty
- Frequency; - may occure 2-3 times/week
  - others may only have 1-2 attacks in lifetime
  - frequency decrease with age
  - more frequent at time of menstruation and less frequent during pregnancy.

Precipitating factors of migraine attacks :

1) Excitation or excessive work
2) Menses
3) Bright light
4) Extra sleep
5) Strong smells
6) Minor trauma
7) Certain diets e.g.; cheese, chocolate, caffeine, alcohol, nuts, milk or missing meals.

* usually headache starts gradually either preceded by aura or not, in one side then spreads to involve the whole side, some time may be bilaterally

* usually one side is commonly affected than the other

Phases of migraine:

Migraine usallay passes through 4 phases

1- Prodromal phase:
2- Aura phase:
3- Attack phase:
4- Post ictal phase:

Diagnostic criteria of migraine:

I- Migraine without aura.

A. At least 5 attacks fulfilling B – D criteria
B. Headache attacks lasting 7-48 Hs.
C. Headache has at least two of the following:
    1. Unilateral ., Pulsating
    2. Moderate / severe
    3. Aggravated by walking or any routine physical stress
D. During headache at least one of the following:
    1. Nausea &/ or vomiting
    2. Photophobia and phonophobia

II- Migraine with aura.

- Usually headache attacks as migraine above beside an aura symptoms indicating focal cortical or brain stem dysfunction
- Aura symptoms develops gradually over more than 4 minutes- lasting not more than 60 minutes.
- Headache follows aura with free interval of less than 60 minutes.

Management of Migraine

I- General measures

- Avoidance of -Stressful situations,
  - Mental and physical fatigue
  - Precipitating diet elements
- Dealing with other ppt. factors

II- Medical measures

- During the attack:
  - Strong analgesics eg aspirin / paracetamol
  - Ergot preparation (e.g Migranil) oral, IM or suppository

NB: ergot itself can cause headache in over dose and a weekly dose of 2-4 mg shouldn't be exceeded

- 5HT- receptor antagonist (Sumatriptan) given in 6mg SC injection
- Prostaglandin inhibitors eg. Flufenamic acid

- Prophylactic measures:
  - Ergot preparations(in closely related attacks) must be tacken before the headache (in the aura)
  - Aspirin – twice daily
  - B-blockers eg; inderal 30-120 mg/day
  - Ca+ channel blockers (flunarzine)
  - Tri-cyclic antidepressants. - Antihistaminic.

Tension type Headache

- Frequently associated with anxiety, depressive disorders and insomnia
- The commenest in 68% of males & 88% of females
- Each attack lasts from 30 minutes up to 7 days
- At least two of the following:
  - Pressing / tightening (non-pulsating) quality.
  - Mild or moderate.
  - Bilateral location
  - No aggravation as in migrain
- No nausea or vomiting
- No phonophobia or photophobia
- May be Associated with spasm of / or tender pericranial muscles.
- Usually precipitated by stressful situations, associated with insomnia, lost appetite.
- Considered chronic if persists for →
  - more than 15 days / month or more than 6 months / year.

■Treatment of tension headache :-

- Analgesics -Muscle relaxants
- Antidepressants -Anxiolytics

Cluster Headache

- Severe unilateral orbital, supra-orbital and / or temporal pain.
- Lasting 15-180 minutes
- Recurrent attacks over the day and may awaken the patient from sleep
- Headache is associated with at least one of the following on the pain side
  - * Conjunctival injection * Lacrimation
  - * Nasal congestion * Rhinorrhhea

*Miosis * Ptosis

*Eyelid edema * Forehead and facial sweating.

- **Treatment:**
  - Strong analgesics -Ergot preparations -Antihistaminics
  - Non steroidal… Especially ''endomethacine'

Speech Disorders

Speech is one of the important aspects to be assessed in neurology. Many neurological diseases may be reflected through specific disorders of speech .

Speech process is formed of two important aspects :

(1) Formulation of speech:

This is the function of higher centers, located mainly in the dominant hemisphere, through which ideas, feelings stimuli and thoughts are received, constructed, formulated and ready to be expressed in either spoken or written speech , -disorders here are called aphasia or dysphasias.

(2) Articulation of speech:

This is the process through which speech is expressed, conducted and produced as words spoken or written – disorders here are called dysartheria.

Anatomical and physiological considerations

-In the process of speech, there are two main mechanisms ;

1- Receiving (sensory) mechanism.

2- Expressing (motor) mechanism.

-In assessment of speech, both spoken and written speech are looked for;

-In spoken speech:

-One have to hear sounds and words at first which are received in the auditory sensory area in the temporal lobe as meaningless words.

-One must be able to understand the meaning of what is heard, this is the function of the auditory psychic area.

-In written speech:

-One have to see at first the written stimuli which are received as meaningless stimuli in the visual sensory area.

-Then, understanding the meaning of these stimuli is the function of the visual psychic area.

-After hearing and seeing the sounds, words and then → recognizing and understanding their meanings in the ***Wernick's*** and ***concept areas*** respectively,then → this is conveyed to the motor speech center (***Broca's area***) in case of spoken speech, and to the writting speech area (***Exner's area***) in case of written speech, this connection is through commissures and associative fibers which connect the auditory sensory and psychic speech centers, the visual sensory and psychic speech centers → Wernick's area → Concept areas → the Broca's and Exner's areas.

→ In the motor speech centers (Broca's and Exner's areas) the process of speech is constructed, generated and can be expressed now in words either spoken or written, this process of expression to be completed is in need for intact motor areas (4&6), corticobulbar tracts, intact cranial nerves involved in the process of speech (5,7,9,10,11&12) for spoken speech, also intact corticospinal tract, A.H.C., nerves and muscles of hands for written speech.

→ The process of conveying this expression through the corticobulbar and corticospinal tracts down to the required muscles is termed Articulation.

→ Articulation to be completed is in need for healthy basal ganglia and cerebellum for good cooperation, smooth coordination of various muscles in the process of articulation.

Speech disorders can be classified into:

**Disorders of formulation - Aphasia.*

**Disorders of articulation - Dysarthria.*

Aphasia

Types of Aphasia:

(1) Broca's aphasia (dominant hemisphere):

with lesions in the motor speech area. The patient can see, hear, understand but can't express his emotions, ideas in spoken speech. i.e. comprehension is intact, but quantity and flow of speech is impaired "non fluent".Lesions here are usually due to stroke, tumors, degenerations.

(2) Wernick's Aphasia:

Due to lesions in the dominant hemisphere associative areas and fibers in the left posterior superior temporal gyrus. Speech is fluent, but with paraphasic errors, meaningless, impaired comprehension.

(3)Transcortical motor aphasia:

(4) Transcortical sensory aphasia :

(5) Conductive aphasia:

(6) Nominal → left angular gyrus (temp. lobe):

- ***Clinical Criteria of the dysphasic disorders:*** Table (3) ↓

	Comprehension	*Fluency*	*Repeatition*
Wernick's Aphasia:	No	Ok	No
Broca's aphasia	Ok	No	No
Conductive Aphasia	Ok	Ok	No
Transcortical motor Aphasia	Ok	No	Ok
Transcortical sensory Aphasia	No	Ok	Ok
Global Aphasia	No	No	No

NB -Visual agnosia: with lesions in the visual psychic area (18, 19), the patient can see, but (can't recognize) does not understand what he sees.

- Alexia: due to lesions in the visual sensory area 39 where the patient can see, but can't read, can write but can't read what he writes.

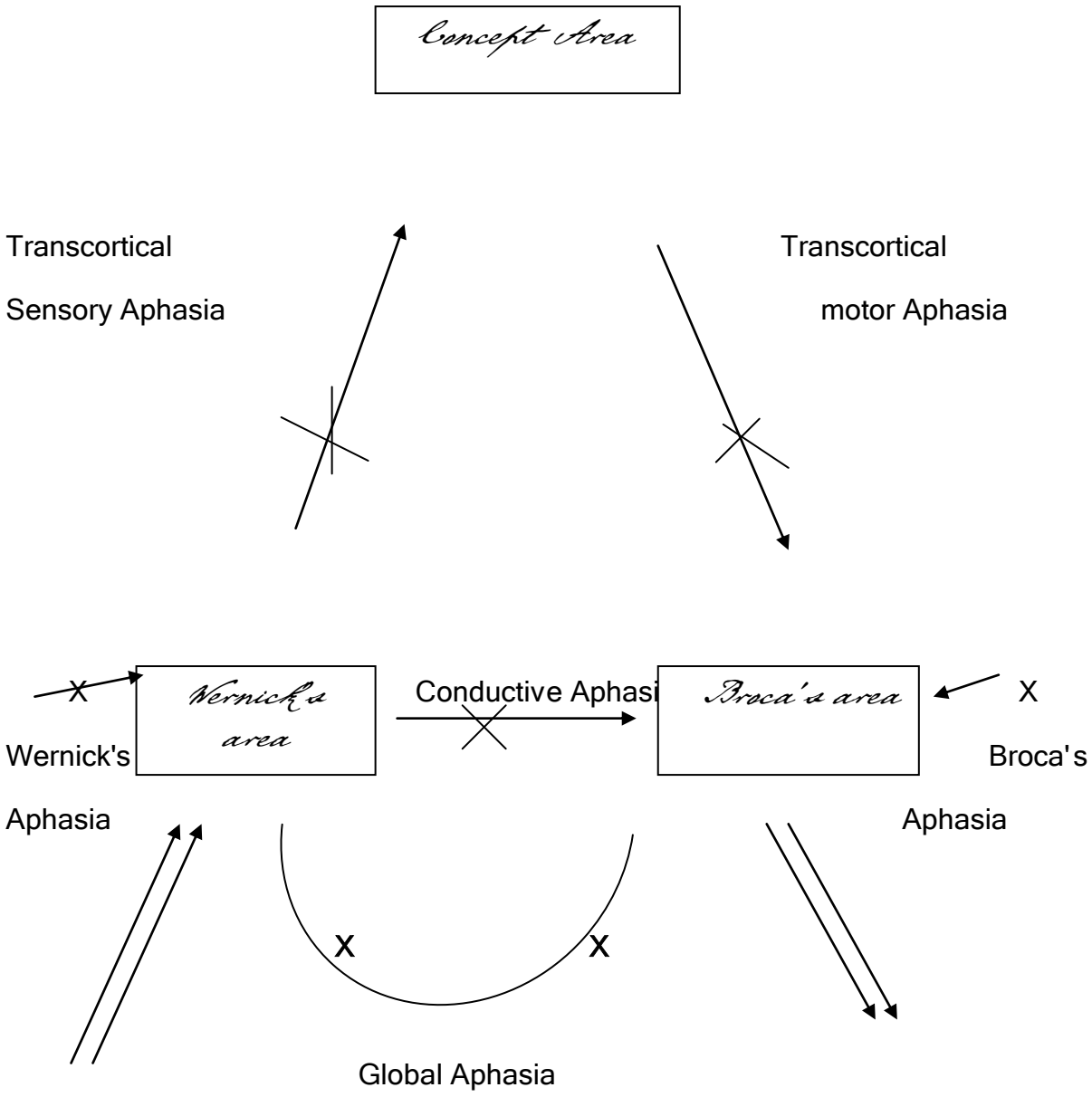
-Auditory agnosia: due to lesions area 22 auditory sensory area.the patient can't recognize the sounds.

-Agraphia: due to localized lesions in the motor writing center "Exner's area". Comprehension is intact but the patient can not express his ideas in written speech

Sites of lesion in aphasia: Fig(41) and diagram(42)

1. **Global Aphasia:** lesion in the dominant hemisphere affecting both Broca's and Wernicke's .
2. **Wernick's Aphasia:** lesion in the Wernicke's area in the supramarginal gyrus of the parietal lobe and upper part of the temporal lobe, may be associated with field defect.
3. **Broca's aphasia:** lesion in the dominant hemisphere in the Broca's area .
4. **Conductive Aphasia:** lesion in the arcuate fasciculus.
5. **Trancortical sensory Aphasia:** lesion in the posterior parietooccipital area.

6. **Transcortical motor Aphasia:** lesion incomplete in Broca's area.
7. **Nominal aphasia:** lesion in angular gyrus.



(Fig .41)

Diagram of a simple bed side test for Dysphasic Speech Disorders

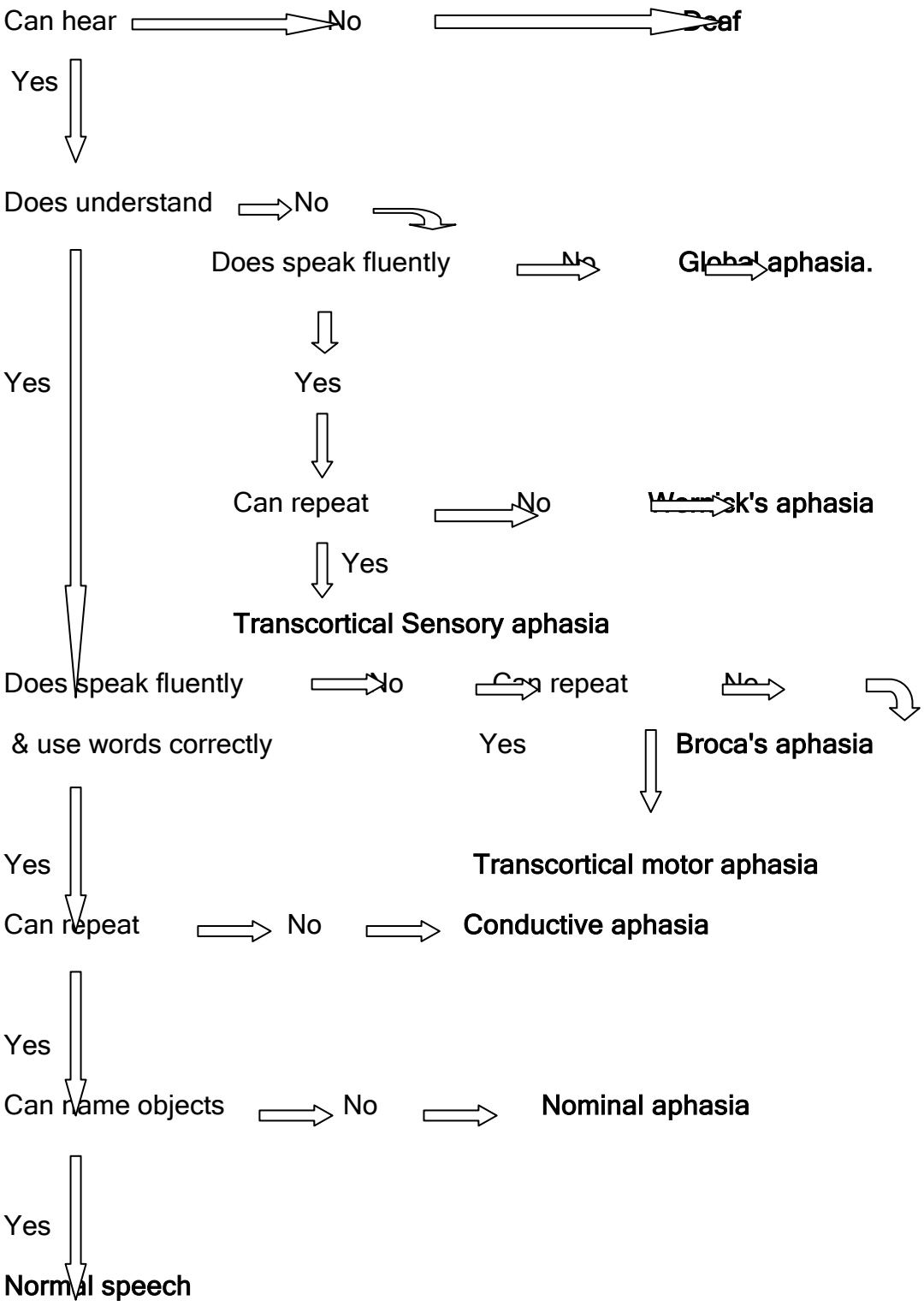


Diagram → (42)

Dysarthria

Causes:

(1) Upper motor neuron lesions:

Muscles of articulations and their cranial nerves receive bilateral pyramidal tract supply, therefore unilateral UMNL rare to cause dysarthria except transient one or if the lesion is severe. In bilateral corticospinal tract lesions as in stroke, diplegia, degenerations and tumours, speech is slurred (mainly labials and dental consonants) spastic, sometimes explosive, hardly produced.

(2) Extrapyramidal lesions.

As in parkinsonism, and as a result of muscular rigidity. Speech is slow, slurred, monotonus and of low pitch, low volume.

(3) cerebellar lesions:

As in ataxias, M.S. due to defects in the co-ordination, co-operations of action of muscles of articulation, speech is explosive, slurred, with wide separation of syllables→ Scanning and staccato.

(4) Lower motor neurone lesions:

In LMNL weakness, wasting of muscles of articulation occurs "bulbar palsy" as in motor neuron disease, syringobulbia, bulbar poliomyelitis, polyneuritis cranialis.

Speech here of low volume, hypotonic, labials consonants suffer early then dentals and gutturals, nasal quality due to weakness of the soft palate with dysphonia. Also in myopathies → speech disorders as in LMNL occurs.

Other rare speech disorders

-Palilalia: Compulsive repetition of last word or phrase of the patient own speech with increasing rapidity and decreasing volume finally into aphonic speech as in:

- postencephalitic Parkinsonism. - general paresis.

-Echolalia: the patient repeats words and phrases that he hears in a parrot like attitude as in dementia, stroke .

-Aphonia: lost phonation which may be:*organic -Laryngitis "severe form"

-Paralysis of adductors of vocal cords.

*Hysterical –common→ ask the patient to cough if can→ Hysterical.

-Mutism: Complete loss of speech (phonation & articulation) in a conscious patient as in congenital Deaf-mute.

-Coprolalia: uttering of swear words as in complex tics.

-Stammering or stuttering: sudden stop of speech flow with forcible repeatition of the sounds or syllables associated with various facial and other body muscle contractions. It is usually psychogenic or familial.

How to test speech

1-Test understanding→ ask simple questions then complicated ones.

2-Test spontaneous speech and look for:

-Flow of speech ,fluent or not.

- Content of speech.

-If there is paraphasic errors or not.

- Slurring or other productive speech disorders

3- Test repetition.

4- Test naming, and word finding.

5-Test reading.→ disorder → dyslexia.

6- Test writing → disorder → dysgraphia.

Disorders of Stance and Gait

Detection of disorders of stance and gait is considered one of the important corner stones in the diagnosis of a neurological case where there are many diseases in neurology which have characteristic gait disorders.

To understand stance and gait disorders, we have to know the normal stance and gait and their requirements

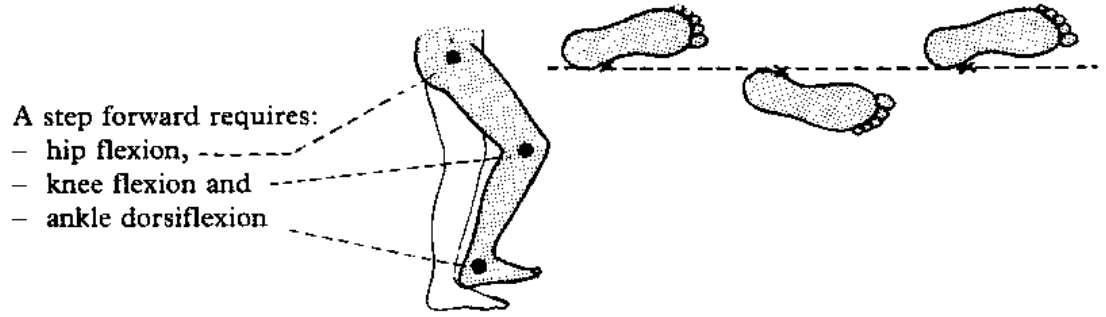
Normal Gait : Fig(43)

It is characterized by:

- Erect posture of the body
- Moderately sized steps
- Medial malleoli of both tibia tracing a straight line

- On walking, hip flexion, knee flexion & ankle dorsiflexion should occur
- Coordination between all the previous events.

The normal gait is characterised by an erect posture, moderately sized steps and the medial malleoli of the tibia 'tracing' a straight line.

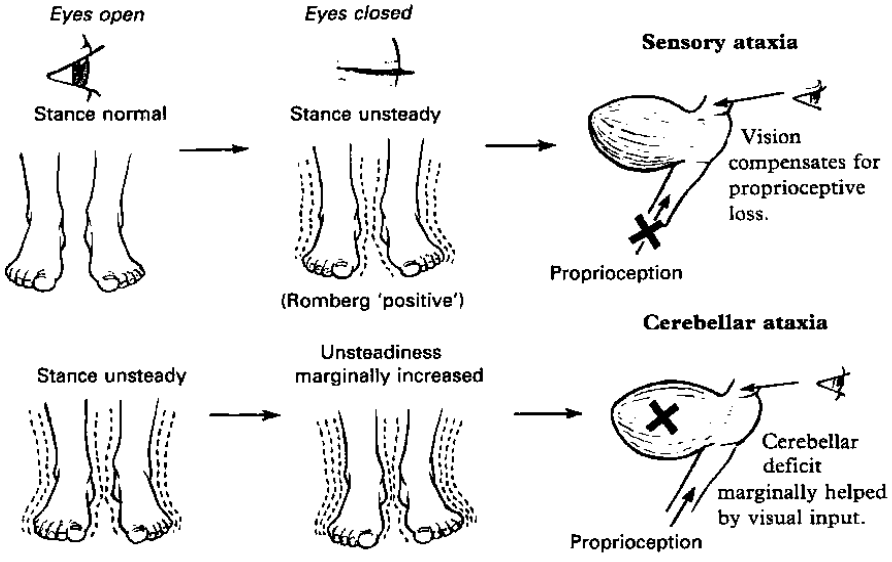


Co-ordination ensures fluidity of movement.
Antigravity reflexes maintain the erect posture. They depend upon spinal cord and brain stem connections to produce extension.

Fig(43)

How to assess stance and gait ? Fig(44)

- Observe during:
  - Standing - Walking
  - Tandum gait (heal to toe walking)
  - Standing with heels together with
    - Eyes opened - Eyes closed



Fig(44)

Specific disorders of stance & gait:

1. Cerebellar ataxic gait.
2. Sensory ataxic gait.
3. Parkinsonian gait.
4. Hemiplegic gait.
5. Paraplegic gait.
6. Myopathic gait.

7. Neuropathic gait.
8. Hysterical gait.

→Cerebellar ataxic gait (Wide base gait, drunken gait, staggering gait)

- Feet are widely separated.
- Steps vary in size.
- Steps are jerky, unsure.
- Swaying of the trunk, Unsteady with tendency to Fall to one or either side.
- In mild cases…. Impairment of tandem gait with falling down.

→Sensory ataxic gait

- Due to defects in the deep sensation (conscious or unconscious) in the posterior column .
- Gait is stamping: The patient need to stamp (firmly catch) the ground with feet to ensure that his feet are in their normal position in the ground, with the" Rombergism"on closure of the eyes.

■ Absence of visual compensation → swaying & falling.

→Parkinsonian gait (Festinant gait, short steppage, shuffling gait) Fig(45)

■ There is generalized rigidity; therefore the Patient adopts a flexed stooping posture.

■ There is difficulty in initiating walking "Bradykinesia"

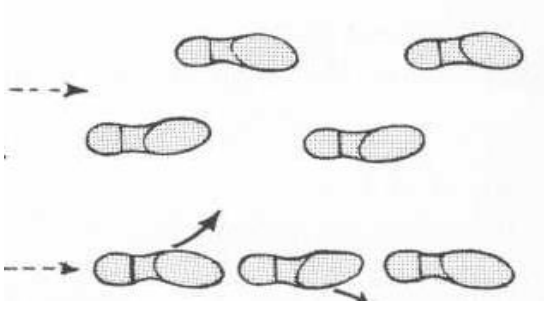
■ The patient feels as his feets are freezing to the ground; his steps are short, small, hurry to avoid falling from his forward stooping

Fig(45)

→Hemiplegic gait (Circumduction gait) Fig(46)

- In hemiplegia, there is Spasticity in extensors & abductors of the LL. & in the flexors and adductors of the UL.
- During walking; the patient needs to bring his extended & abducted diseased LL. To meet the other limb to support his body
- On walking; this will make circumduction form, the UL is held in flexed, adducted position (with limping on the weak LL on walking.)

Hemiplegic gait



→Paraplegic gait (Scissoring gait)

- Due to spasticity of both LL., the gait is spastic, Scissoring like

→Myopathic gait (Waddling gait)

■ Due to Proximal pelvic muscles weakness, on walking and with the hip flexion the other pelvic weak muscles can't support the pelvis→the pelvis will tilt from side to side(**waddling gait)**

■This is associated with lumbar lordosis & pot-belly abdomen

→Neuropathic gait (High steppage gait) Fig(47)

- In peripheral neuropathy, there is Weakness of distal muscles, with foot drop.
- On walking the patient needs to left the dropped foot from the ground to be able to walk forming horse like gait.(high steppage gait)

Fig(47)

→Hysterical gait.

- Bizarre in nature with no characteristic form.
- The hallmark is absence of neurological signs.

The cranial nerves

There are 12 pairs of cranial nerves, the olfactory (I) and the optic nerves (II) are not true nerves but fiber tracts of the brain, also the spinal accessory nerves (XI) is derived from the upper cervical cord segments.

The remaining (9) nerves relate to the brain stem.

1) Olfactory Nerve and smell:-

The olfactory receptors “bipolar nerve cells” are situated in the upper part of the nasal mucosa. Inhaled gases given off by odorous materials become dissolved in the nasal secretion and the nerve cells → axons of the receptor nerve cells form the olfactory bundles → pass through the cribriform plate of the ethmoid bone → to the olfactory bulb lying beneath the ipsilateral frontal lobe → to the olfactory tract → to the olfactory cortical areas “uncus, amygdala (near the temporal lobe).

Olfactory nerve examination Fig(48)

-Test sense of smell (Quantity & Quality) by using familiar, non irritating material to avoid stimulation of the trigeminal nerve fibers in the nasal mucosa.

-Ask the patient to close his eyes to avoid visual identification of the tested material, examine each nostril on obstructing the other.

OLFACTORY NERVE (I)

Test both perception and identification using aromatic non-irritant materials that avoid stimulation of trigeminal nerve fibres in the nasal mucosa, e.g. soap, tobacco.

One nostril is closed while the patient sniffs with the other.

Fig(48)

→Common disorders:-

- Anosmia – hyposomia: (in Quantity)

-Unilateral –most common with neurological disorders.

- Bilateral -most common with ENT disorders.

- Olfactory hallucination:- In psychosis, uncinate seizures.

- Parosmia (in Quality) unpleasant smells:- In head injury, nasal infection, depression.

2) The optic nerve and vision Fig(49)

Starting from the retina. there are two types of retinal receptors “rods & cones “, Rods → responsible for night vision & cones → responsible for color , daylight , high acuity vision → axons of rods and cones form the optic nerve fibers (nearly 1 million fibers)→ optic nerves pass backwards and inwards through the optic foramina to the optic chiasma - in the optic chiasm the two optic nerves unit where the nasal fibers decussate to pass to the optic tract of the opposite side → the optic tract pass between the cerebral peduncles to terminate in the superior colliculus (concerned with reflex activity)and the lateral geniculate body

(concerned with vision)→ from the lateral geniculate body the visual fibers enter the posterior limb of the internal capsule → emerge from the internal capsule as the optic radiation → to the visual occipital cortex .

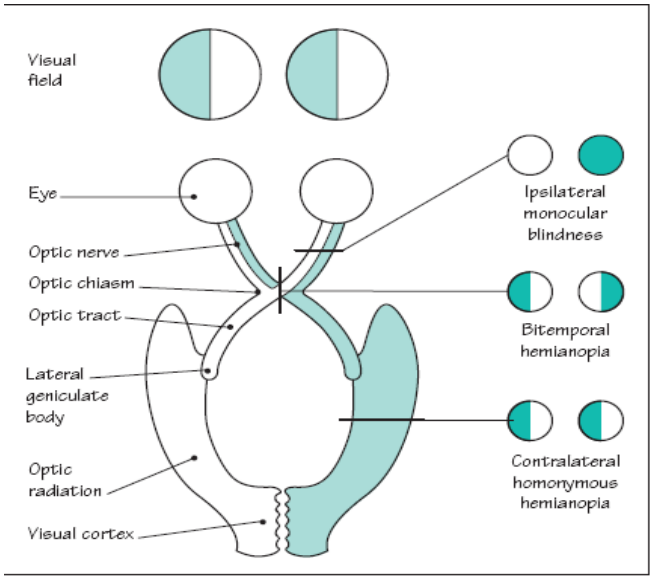


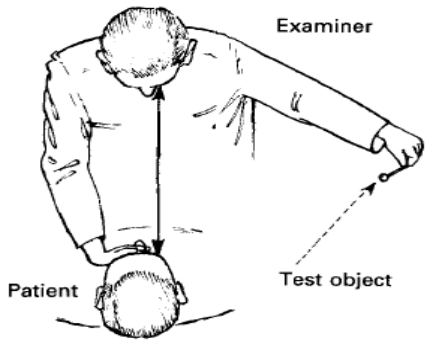
Fig. The anatomy of the visual pathways, and the three common types of lesion occurring therein.

Fig(49)

The optic nerve examination:-

Test for (acuity of vision, field of vision, and fundus examination)

1- **Acuity of vision** : using the snellen 's chart , if not available , use counting fingers “6” meters away from the patient, test each eye separately . if the patient cannot count your finger at this distance , near the distance till 30 cm , if the patient still fails → test for hand movements → if failed → test for light perception



2- **Field of vision:-** → Fig(50)

“Confrontation test “the physcian stands or sits in front of the patient and at the same level 60 cm – 100 cm away from him. Ask the patient to close one eye with his hand and you have to close the opposite eye , ask the patient to fix his other eye gaze upon the bridge of the physician's nose →the physician moves one of his fingers or any moving objects e.g. pencil,

in the six cardinal directions (midway between the patient & physician) of the visual fields

The patient on fixing his other eye gaze is asked to say when he first sees the moving finger or object, and then the physiation can determine the extent of the patient's visual fields.

■Abnormalities of the visual fields:-

- Hemianopia: means loss of vision in ½ of the visual field
- Homonymous hemianopia: if both right and left halves are affected.
- Bitemporal, binasal hemianopia: if both temporal or both nasal halves are affected.
- Quadrantic hemianopia: field defect limited to one quadrant.

N.B:-

In young children or uncooperative patient the Menase reflex can be used for visual field examination. By moving an object or hand from periphery in various directions and noticing whether the patient blinks in response to seeing the object.

3- **fundus examination :-**

By the ophthalmoscope, the fundus must be examined for:-

- Signs of papilloedema,
- State of retinal vessels,
- Retinal hge or exudates.

Some lesions of the optic nerve:-

- **papilloedema** → means oedema of the optic disc .mostly due to various causes of increased intracranial pressure as tumors , Hge , infarction , infections of C.N.S, inflammations of the optic nerves
- **optic neuritis** and retrobulbar neuritis → means inflammation of the optic nerve which may be due to local spread of infection , acute demyelination , vascular occlusion , toxins , drugs , metabolic or hereditary causes.
- **Optic atrophy** → which may be the end result of most of optic nerve lesions "2nd optic atrophy "with slow progressive visual failure.

3) The ocular nerves(3rd , 4th, 6th) and ocular movements .

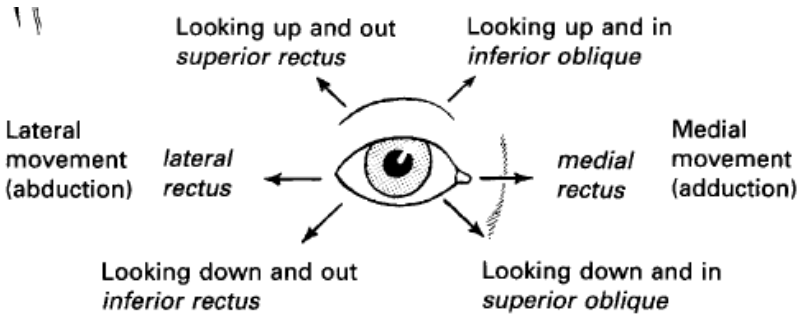
Ocular movements:-

1- Individual ocular movements mediated and controlled by ocular nerves and their nuclei in the brain stem.
2- Conjugate (version) ocular movements either saccadic or smooth pursuit, both horizontal and vertical are controlled by the supranuclear centers mainly in the frontal cortex (area 8) & posterior parietal area.
3- Dysconjugate (vergence) either convergence or divergence also controlled by the supra nuclear centers mainly in the posterior callosal and anterior occipital regions.

.N.B: All the supra nuclear centers feed into the medial longitudinal fasciculus(M.L.F) in the brain stem (posterior longitudinal bundle) which also link the 3rd , 4th and 6th nerves together for coordination of ocular movements , also MLF receives extensive inputs from the vestibular , cerebellar and other nuclei concerned with reflex ocular movements .

The extrinsic ocular muscles are: Fig(51)

- *4 Recti* → - Superior rectus , action ⟶ elevation of the abducted eye
  - Inferior rectus , action ⟶ downward movements of the abducted eye.
  - Medial rectus , action ⟶ adduction
  - Lateral rectus , action ⟶ abduction.
- *2 obliques*:- - Superior oblique , action ⟶ downward movements where the eye is adducted.
  - Inferior oblique , action ⟶ elevation where the eye is adducted.



Fig(51)

All the extrinsic ocular muscles are supplied by the 3rd oculomotor nerve except: -

- the lateral rectus → supplied by the abducens nerve
- the superior oblique → supplied by the trochlear nerve.

The 3rd & 4th cranial nerves are located in the midbrain just anterior to the cerebral aqueduct while the 6th nerve nuclei lie in the pons beneath the floor of the 4th ventricle.

The 3rd nerve pathway:-

The oculomotor nerve emerge and leaves the brain on the medial side of the cerebral peduncle → then passes in the lateral wall of the cavernous sinus →then leaves the cranial cavity through the superior orbital fissure to supply its muscles .

The 4th nerve pathway:-

This is the only crossed cranial nerve, it originates in the lower midbrain and cross to the opposite side → then passes → to the lateral wall of the cavernous sinus → enters the orbit through the superior orbital fissure to supply its muscle → superior oblique.

The 6th nerve pathway :-(longest intracranial pathway)

From the 6th nerve nuclei which lie in the pons , the nerve emerge at the ponto medullary fissure → passes through the base of the brain and over the apex of the petrous temporal bone → then passes through the lateral wall of the cavernous sinus → enters the orbit through the superior orbital fissure to terminate in the lateral rectus muscle.

How to test ocular nerves?

Examine for - eye lid for ptosis.

- Pupil.
- Extrinsic individual ocular movements & squint.
- Nystagmus.

The pupils, eyelids and pupillary reactions:-

■ *The size of the pupil is under the control of two muscles:*

1) Sphincter "circular "pupillae muscle which is supplied by the 3rd cranial nerve → its contraction causes miosis.
2) Dilator pupillae muscle which is supplied by the cervical sympathetic "C8-T1"which arise from the hypothalamus →its contraction causes mydriasis.

N.B

Size of the pupil is 2-4 mm.

Paralysis of the dilator pupillae - - sympathetic paralysis :-

The pupil will be constricted owing to the unantagonized action of the sphincter pupillae muscle

Lesions of the ocular cervical sympathetic fibers will lead to what is called "Horner' s syndrome " where other ocular sympathtetic fibers lie near that of the dilator pupillae , therefore in **Horner's syndrome** there is :

-Miosis - Enophthalmos .

- Slight ptosis - Impaired sweating "anhydrosis"

■ *The eye lid muscles innervation:-*

Two muscles elevate the upper eyelid:

- Levator palpebrae superioris → supplied by the 3rd nerve .
- Muller's palpebrea muscle → supplied by the cervical sympathetic fibers .

■*The light reflex:-*

On exposure to light , → to the 3rd nerve → to both sphincter pupillae muscles to cause constriction .

To test light reflex , one eye is closed , and the patient is asked to look at a distant object to eliminate the effect of accommodation , the eye is exposed to the light source , the stimulated eye pupil will be constricted (direct reaction) and the opposite pupil is also constricted (consensual reaction)

Test of ocular movements:-

- for individual eye movement → each eye is tested alone by asking the patient to look at any object located in the six cardinal directions and the physician should notice any limitation of muscle movements or if the patient complains from double vision in certain direction .

Oculomotor nerve lesions:-

a) in complete lesions of the 3rd nerve , the following will occur:

- weakness of the four external ocular muscles (superior , inferior , medial recti ,and the inferior oblique muscles), therefore the eye will be directed down and out by the unantagonized action of the superior oblique and lateral rectus muscles

- Unilateral complete ptosis → due to paralysis of the levator palpebrea superioris .

- lost accommodation.

- fixed dilated pupil → due to paralysis of the sphincter (constrictor) pupillae muscles

b) In partial lesion. Some nerve fibers may escape mainly those of the constrictor pupillae muscles .

c) common causes of the 3rd nerve lesions :

- Infarction, Hge, tumors, demyelination of the mid brain.

- Posterior communicating artery aneurysm.

- Trauma, arteritis, infection.

Trochlear nerve lesions:-

Will result in paralysis of the superior oblique muscle with weakness of downward and inward movement and with double vision when the patient looks downwards to the affected side.

N.B:- In intracerebral lesions before decussation, the paralysis of the superior oblique will be on the opposite side, while in extracerebral lesion after decussation, the paralysis will be ipsilateral.

Causes of the 4th nerve lesions:-

Infarction, Hge, tumors in the mid brain, trauma, mastoiditis, meningitis.

N.B: Isolated nerve lesion is rare.

Abducens nerve lesions:-

- will result in paralysis of the lateral rectus muscle with limitation of the abduction of the eye. With its deviation inwards by the unantagonized medial rectus muscle, with diplopia on looking laterally on the same affected side.

- *causes* :infarction , Hge , tumors ,inflammation , demyelination of pons , trauma , mastoiditis

Combined ocular motor nerve palsies "3rd, 4th, 6th" Fig(52)

1. in the brain stem : due to stroke , tumours , inflammations, associated with brain stem signs.
2. in the cavernous sinus with involvement of the ophthalamic division of trigmenal nerve
3. in the superior orbital fissure .
4. in the orbit → due to tumours , trauma , orbital cellulitis .

Nystagmus:

Definition:

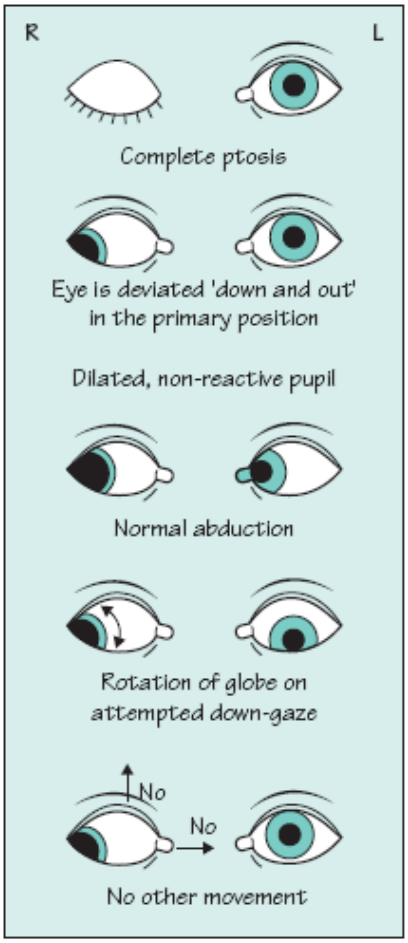
Nystagmus is a disturbance of ocular posture characterized by involuntary rhythmical oscillation of the eyes.

Types & causes:-

1) pendular nystagmus → - the same rate in both directions.
2) Jerky nystagmus →quicker in one direction than the other and the quick phase is taken to indicate the direction of the nystagmus
3) Resting nystagmus: occurs when the eyes are on rest.
4) Positional nystagmus: occurs only when the head is in certain position.

-Nystagmus movements may be - horizontal - vertical → on one plane.

- Rotatory.

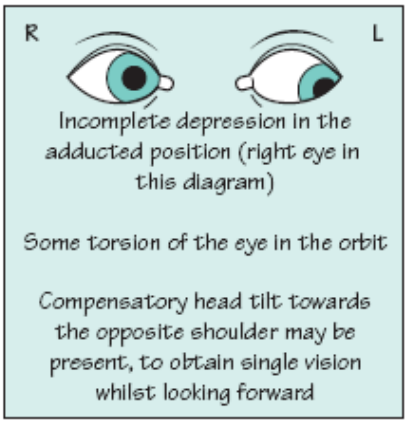


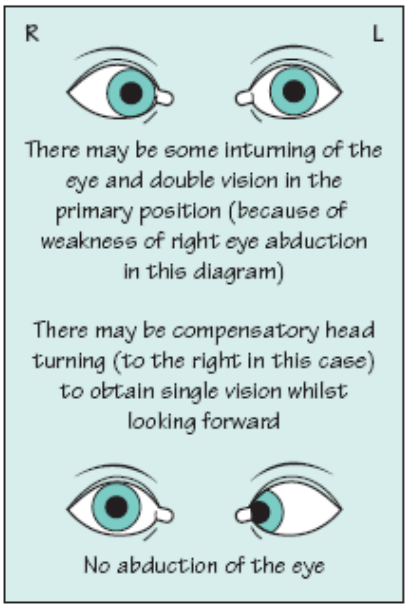
Third nerve palsy

- Common.
- Common causes:
  posterior communicating artery aneurysm (painful);
  mononeuritis in diabetes (pupil usually normal);
  pathology beside the cavernous sinus, in the superior orbital fissure or in the orbit (adjacent nerves commonly involved, e.g. 4, 6, 5a, and 2 if in the orbit).
- The parasympathetic innervation of the eye is supplied by the 3rd nerve.
- The diagram shows a complete right 3rd nerve palsy. The lesion can be incomplete of course, in terms of ptosis, pupil dilatation or weakness of eye movement.

Fourth nerve palsy

- Uncommon.
- Common cause; trauma affecting the orbit.





Sixth nerve palsy

- Common.
- Common causes:
  as a false localizing sign in patients with raised intracranial pressure;
  multiple sclerosis and small cerebrovascular lesions within the pons;
  pathology beside the cavernous sinus, in the superior orbital fissure or orbit (adjacent nerves commonly involved, e.g. 3, 4, 5a, and 2 if in the orbit).

Fig(52)

- Nystagmus may be - 1st degree → occurs on lateral gaze to one side

- 2nd degree →occurs on looking ahead but more on looking to one side

- 3rd degree→ present on fixation, looking to both sides but more to one side.

- Nystagmus may be - peripheral (due to retinal or labyrinthine causes) or
- central due to (spinal cord, cerebellum, and brain stem lesions)

- Nystagmus may be physiological(optokinetic nystagmus) → on looking to a series of moving objects pass before the eye e.g on looking out of the window of a moving train .

-congenital & familial nystagmus

- Voluntary nystagmus which can be initiated by persons

-Toxic nystagmus.

<u>The fifth (trigeminal) nerve</u> Fig(53)

Anatomy & pathway

Trigeminal nerve is the principle sensory cranial nerve, mixed nerve composed of two roots → long,large sensory & small motor roots arising from the nuclei which are located in the inferior lateral surface of the pons.

1- **Sensory nuclei :- two**
   1- Large principle sensory nucleus which receives tactile and postural sensation from the face.
   2- Spinal nucleus and its tract which ends in the upper cervical cord C2 and carry pain and thermal sensation from the face.

   The two nuclei join the spinothalamic tract.

2- **The motor nucleus**: -

   Single, and is located in the lateral part of the pontine tegmentum .

   ---There are three large nerve trunks or divisions namely the **ophthalmic** , the **maxillary** , and the **mandibular** .

   The motor part unites with the 3rd mandibular sensory division to form the mandibular nerve.

a- *The ophthalmic division* : → passes to the lateral wall of the cavcernous sinus with the 3rd , 4th, 6th cranial nerves → enters the orbit through the superior orbital fissure → face where it supplies an area surrounded by two lines → upper one passing from the tragus of one ear to the other passing through the lambdoidal suture and the lower one passing from the tragus of each ear to the lateral epicanthus of each eye .this area includes : the nose , forehead , scalp , upper eyelid , conjunctiva , cornea, iris , and the mucous membrane of the frontal sinuses and upper nasal cavities .

b- *The maxillary division* → passes through the inferior part of the cavernous sinus → enters the orbit through the inferior orbital fissure → to the infraorbital formina → to the face to supply an area surrounded by an upper line from each ear tragus to the lateral epicanthus of one eye and a lower one connecting each tragus to the lateral angle of the mouth on each side.

This area includes – skin of the cheek. - Skin of the upper lip.

_ Mucous memberne of maxillary sinus, lower nasal cavity, hard & soft palate, teeth of the upper jaw.

c- *The mandibular division:-*

(1)- The sensory part : passes out of the skull with the motor part through the foramen ovale to the face to supply an area lying below a line from tragus of each ear to the lateral angle of the mouth except that of the angle of the mandible which is supplied by C2, the area supplied by the mandibular division includes also :-

- Tympanic membrane, external auditory meatus.
- The tragus ,ear pinna"upper part"
- Lower jaw , teeth , floor of the mouth , ant 2/3 of the tongue .

(2)- The motor part : The motor part follows the same pathway with the mandibular division where they unit to form the mandibular nerve , the motor part supplies :

- Temporalis .- Masseter.
- Medial & lateral pterygoids.
- Anterior belly of the diagastric , mylohyoid , and the tensor palati → if weak → flattening of mouth floor.

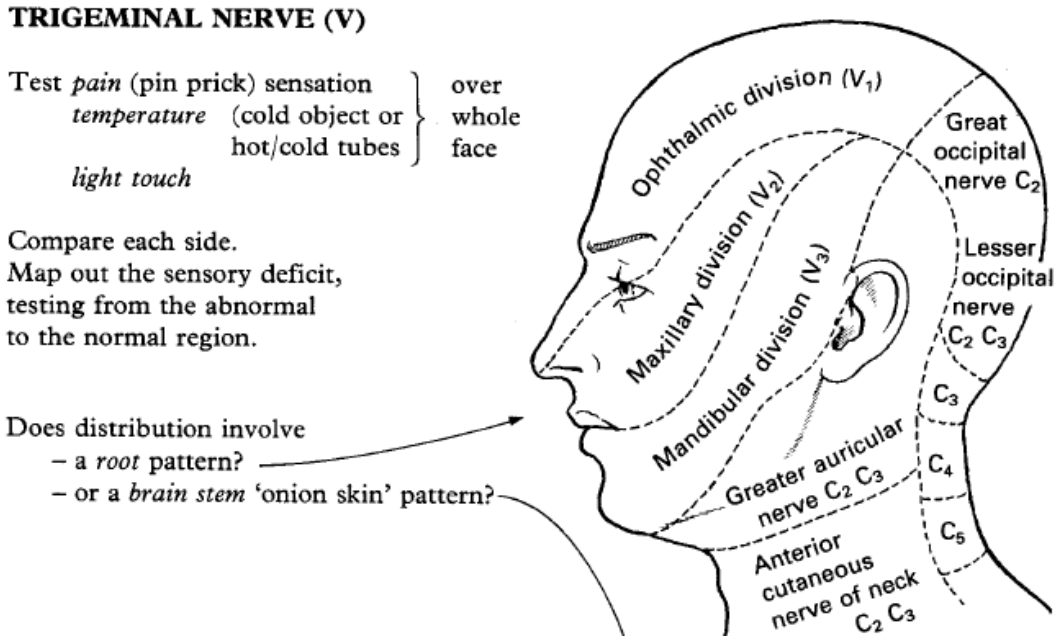
How to examine the trigeminal nerve ?(sensory , motor , reflexes)

1) Sensory :

- Define the land marks of the three divisions of the trigeminal nerve .
- Ask the patient to close his eyes.
- Test pin prick , touch or thermal sensation in the following manner –side by side , division by division , central and peripheral part of each division .

2) Motor :-

- Ask the patient to clench his teeth and compare the contraction of both temporalis and masseters on both sides with palpating these muscles
- For the pterygoids , ask the patient to open his jaw to notice any deviation (muscles here are pushing in action & the deviation will be towards the paralysed side , then ask the patient to open his jaw from side to side against resistance.



Fig(53)

3) Reflrxes :-

1-Corneal reflex :(afferent 5th cranial nerve "1st division " & efferent 7th cranial nerve & center in pons).

The reflex is tested by applying a piece of cotton to the cornea on asking the patient to look to one side and the examiner standing on the other side and behind the patient , avoid not to touch the conjuctiva (common mistake with false response). → the response will be blinking of both eyes

2 - The jaw reflex is tested by indirect taping on the partly opened jaw , afferent & efferent are through the 3rd division of the trigeminal nerve with the center in the pons

-Normally → no response will occur (or very slight) Fig(54)

-Brisky jaw reflex "brisk closure of the jaw "→ occurs in UMNL above trigeminal nerve nuclei in pons.

3- Glabellar tap reflex :

Frequent direct taping on the root of the nose will induce bilateral frequent blinking of both eyes which will be lost (accommodation) after few taps in normal individuals , this accommodation is lost with continous blinking on frequent taping in patients with Parkinson' s disease .

-Afferent → 5th cranial nerve. - Efferent → 7th cranial nerve.

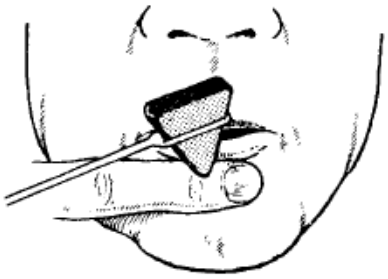
TRIGEMINAL NERVE (V) *(contd)*

Jaw jerk

Ask patient to relax jaw. Place finger on the chin and tap with hammer:

Slight jerk – normal

Increased jerk – bilateral upper neuron lesion.



Fig(54)

Some lesions of the trigeminal nerve:-

1- **Pontine lesions** : which will involve the trigeminal nerve nuclei beside involvement of the pyramidal tract and spinothalamic tracts as in tumors , stroke , demyelination , syringobulbia , will result in :

- Ipsilateral weakness of muscles of mastication (temporalis , masseters, and pterygoids)
- Ipsilateral loss of facial sensation .
- Contralateral hemihypothesia &hemiplegia.

2- **Trigeminal neuralgia : (tic douloureux)**

-Characterized by paroxysmal , brief attacks of severe pain (lancinating, electric like) in one or more division of the trigeminal nerve (the mandibular and the maxillary parts are most commonly involved)

- Women are more affected with peak incidence in 5^{th} - 6^{th} decades of life .

- **Causes** :-→ - may be idiopathic

- or symptomatic as in multiple sclerosis which is considered the commonest cause , tumors , trauma , vascular causes .

- Diagnosis of trigeminal neuralgia :

1- Clinical diagnosis by the characteristic symptoms of :

- Severe lancinating , electric , stabbing pain as hot needle in one or more divisions
- Duration not more than 1 or 2 minutes .
- Very severe pain which evokes reflex spasm of the facial muscles and let the patient in agony and depression .
- ± Associated with flushing of skin , lacrimation & salivation .
- Precipitated by touching the face in shaving or washing , by talking , mastication and swallowing .
- Many patients describe areas – touching of which excites an attack → trigger zone .
- The pain may be radiated to other areas of trigeminal nerve divisions.
- Sensation is usually intact except in long standing & symptomatic cases.

2- Differential diagnosis :

- Migraine .
- Cluster headache .
- Referred pain from sinusitis , tooth ache , eye disease .
- Psychogenic pain and tension headache .
- Other causes of headache .

Treatment of trigeminal neuralgia :

- Carbamazepine (tegretol)400- 1600 mg / day on two or three divided doses .or gabapentin
- Anti depressants .
- On failure of medical treatment , local injection of alcohol or phenol around the supraorbital or infraorbital or to the trigeminal ganglion may be effective.

The 7th (facial)nerve:

Anatomy

The 7th facial nerve is a mixed nerve "motor , sensory , autonomic consists mainly of motor fibers . its upper part is bilaterally supplied from corticobulbar tract of both sides , while its lower part is unilaterally supplied from corticobulbar tract of the opposite side . The fibers of the motor nucleus form a loop around the 6th nerve nucleus before turning forwards to emerge from the lower part of the pons on the medial side of the 8th cranial nerve from which the 7th nerve is separated by the nervous intermedius .

The three nerves (7th, 8th, and the nervous intermedius)pass together from the pons through the subarachinoid space of the cerebellopontine angle to the internal auditory meatus where the facial nerve passes to the facial canal which leaves it from the stylomastoid foremen into the face to supply its muscles .

In the facial canal , the geniculate ganglion sends peripheral fibers (chorda tympani nerve) which supply the sublingual and submaxillary glands and receives taste sensations from the anterior 2/3 of the tongue .

Supply of the facial nerve :

1- Motor :

- Frontalis .
- Orbicularis occuli .
- Buccinator
- Orbicularis oris

} after leaving the stylomastoid foramen

- Retractor anguli .

- Platysama .
- Stylomastoid } after leaving the stylomastoid foremen
- Posterior belly of the diagastric .
- Occipital belly of occipitofrontalis
- Stapidius→its fibers arise within the facial canal .

2- Sensory : taste sensations from the anterior 2/3of the tongue through the chorda tympani

3- Autonomic :

- To the otic & sphenopalatine ganglia .
- To the sublingual & submaxillary glands .

How to examine the facial nerve (motor, sensory , reflexes)? Fig(55)

1-Motor :-

a- ***Upper face*** :-

- Test for *frontalis* → asking the patient to raise his eye brows → look for corrugation of the forehead on both sides .
- Test for *orbicularis oculi* → asking the patient to close his eyes → look for the opened unclosed globe or for the skin folds around the globe in partial weakness .

b- ***Lower face*** :-

- Test for *buccinator* → by asking the patient to blow his cheek .
- Test for *orbicularis oris* → by asking the patient to whistle .
- Test for *retractor anguli* → by asking the patient to show his teeth .

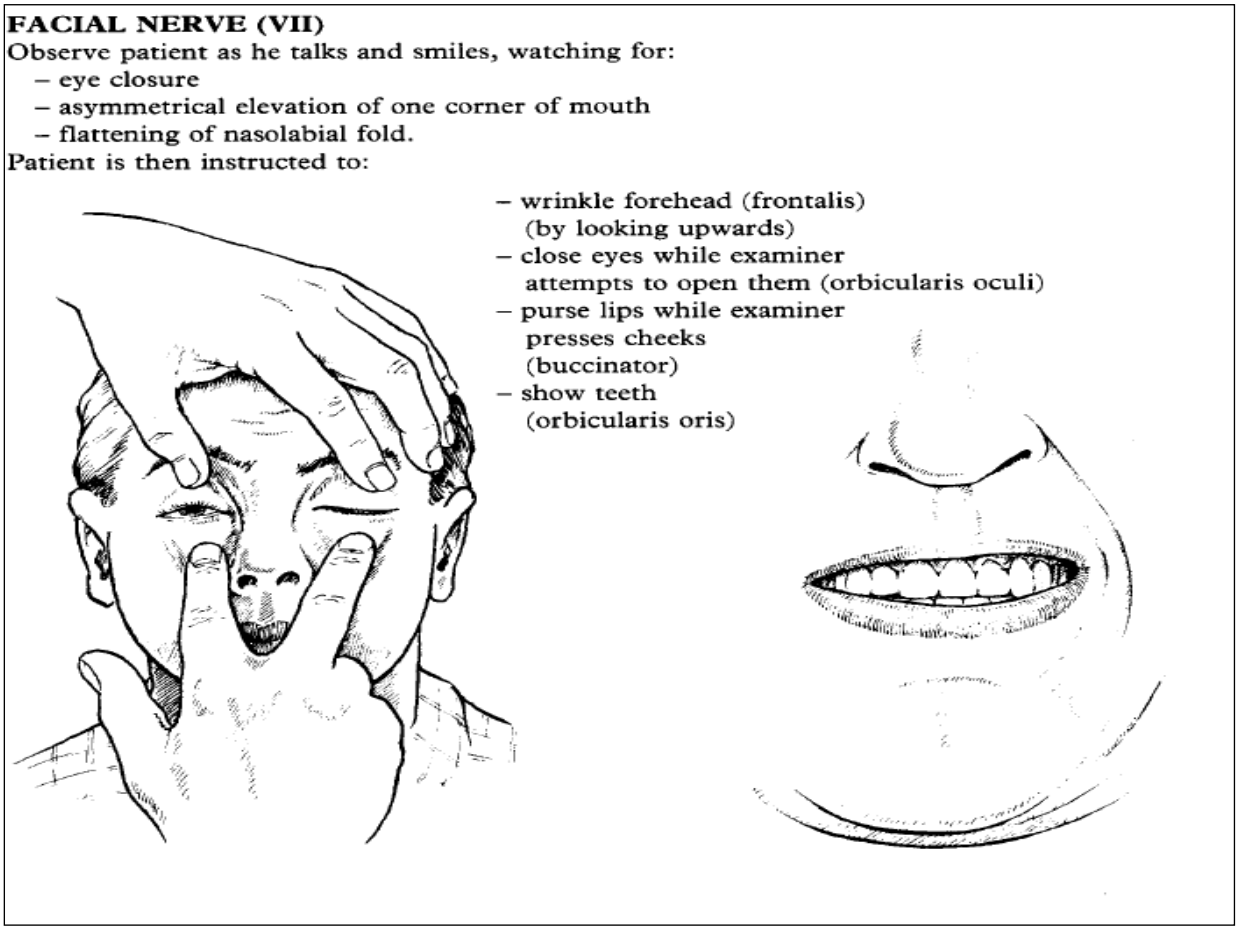
N.B :

Emotional facial movements have a distant pathway originating in the frontal lobe with different route.

2- Sensory :-

Test for taste sensation over the anterior 2/3 of the tongue → dry the patient tongue to avoid transmitting of the tested material by saliva to other parts .

- Ask the patient to close his eyes to avoid visual recognition
- Put the tested material and see if the patient can recognize .



Fig(55)

3- Reflexes :- - Corneal reflex - Glabellar reflex.

Lesions of the facial nerve

1- Supranuclear lesions :

- UMNL involving the corticobulbar fibers concerned with the voluntary facial movement

- The muscles of the lower face of the opposite side will be affected more .

- Facial paralysis will spare the facial emotional movements .

- Will be associated with hemiplegia on the same side of facial weakness.

- Causes are usually are those of stroke .

2- Supranuclear lesions involving only the fibers concerned with emotional movements - Mimic paralysis on the opposite side leaving the voluntary facial movements unimpaired

3- LMNL involving nuclear , infranuclear lesions or lesions of the nerve itself .

The lower motor facial nerve lesions may be at the following sites:

- The pons

- Within the posterior fossa “cerebellopontine angle”

- Within the temporal bone . - Outside the skull.

i. **Nuclear lesions (pontine lesions):**

- Parslysis will occur to the whole facial muscles on the same side of the lesions

-Other cranial nerves may be affected especially the 5th & the 6th .

- Pyramidal tract involvement with hemiplegia on the opposite side "crossed hemiplegia "

- Taste sensations from the tongue , salivation & lacrimation are impaired.

- There may be also sensory loss due to involvement of the trigeminal nucleus and of the spinothalamic tracts .

Causes : -

Vascular (stroke) , inflammations, M.S , tumours, demyelination , syringobulbia, poliomyelitis .

ii. **Within the posterior fossa :-**

- 8th nerve lesions with deafness

- loss of taste sensations on the anterior 2/3 of the tongue (nervus intermedius lesions)

- LMN facial of all the facial muscles on the same side .

Causes : - Acoustic neuroma . - Cerebellpontine angle tumours

- Aneurysm . - Meningitis .

iii. **Within the temporal bone & facial canal** : e.g Bell's palsy .

- LMN facial palsy on the same side .

Causes : - OM , mastoditis , head injury , ear operation , herpes zoster .

iv. **Outside the skull :**

LMN facial palsy on the same side.

Causes : - Tumours , lesions or suppuration of the parotid gland

- Sarcoidosis .
- Infective mononucleosis.

Bell's palsy

1- **Definition :**

LMN facial paralysis of acute onset , due to non suppurative inflammation,of the facial nerve within facial canal .

2- **Aetiology & Pathology :-**

- There is acute inflammation with oedema involving the nerve within the facial canal .

- Can occur at any age – from infancy to old age .

- Oedema , vascular ischemia & inflammation of the nerve with consequent compressive /or ischemic peripheral neuropathy may be the main pathogenesis.

- causes : - Diabetes mellitus .

- ↑ incidence of Bell's palsy in hypertention , pregnancy .
- Exposure to cold air drafts → autoimmune hypothesis .
- Viral infections "herpes zoster→ Rumsy Hunt syndrome → rare causes.

3- Symptoms & signs:-

- Onset may be preceeded by pain in the ear or peri auricular .
- The onset is acute and mostly observed by the patient 's family or friends
- The weakness is partial in30 % and complete in 70% of patients .
- The weakness will involve the facial muscles of both the lower and upper parts of the face equally with involvement of the emotionally and associative movements.

- the patient will complain of :

- Drooping of the eye brow
- Difficult flowing & raising of the eye brow

} weakness of the **frontalis** .

- Wider palpebral fissure
- Impossible closure of the eye
- Continous tearing .
- When the patient tries to close the eye , the globe rolls upwards and inwards (Bell's phenomena)

} weakness of the **orbicularis oculi**

- The patient can not retract the angle of the mouth of the diseased side.
- The nasolabial fold is smoothed and obliterated on the diseased side

} → ------ weakness of **retractor anguli**.

- The patient can not purse the lips as in whistling .
- The cheek puffed out in respiration
- Food accumulates behind the cheek .

} weakness of the **buccinator**

- Intensification of the loud noise in the affected ear "hyper acusis "→stapidius weakness

4- differential diagnosis :

Bell's palsy must be distinguished from facial weakness due to :

- Pontine lesions
- Posterior fossa
- 2ry to OM , mastoditis , M.S

5- Treatment :

1- Medical treatment :-

a- Prednisone :60- 80 mg/day for 5 days followed by gradual decrease of the dose

b- other medication may be needed "not essential "

- Analgesic – Vitamins - - Corneal protectors ,Vasodilators

2- Physiotherapy :-

- Massage of the facial muscles .
- Hot fomentations of facial muscles
- Muscular exercise of the weakned muscles .
- Direct electrical stimulation may predispose to contractures → not widely used

The 8th vestibulcochlear nerve:

Anatomically : The 8th nerve contains two groups of fibers:

1- The *cochlear fibers* which supply the cochlea→ concerned with hearing .

2- The *vestibular fibers* which supply the semicircular canals , the utricle & the saccule→ concerned with equilibrium.

Anatomical and physiological consideration of the cochlear nerve and hearing :-

- Sound waves transmit in the surrounding medium as pressure waves→ collected and perceived by the *ear pinna* (auricle)→ into the *external auditory canal* →to the *tympanic membrane* where the recorded sound vibration are more modified and filterd and will be transmitted→ to the middle ear to pass through the *chain of auditory ossicles*(malleus, uncus & stapes) for more modifications , in turn transmit the sound vibrations across the middle ear cavity → to *the oval window* of the cochlea

- This pathway is affected by the tensor tympani muscle(supplied by the trigeminal nerve) and by the stapedius muscle (supplied by the facial nerve), the two muscles protect the ear against high intensity sounds .

- The sound vibrations after passing from the oval window , → finally stimulate→ *the cochlear sensory cells* whose fibers form the *cochlear nerve* .

→ The cochlear nerve pass from the inner ear through the internal auditory meatus and canal to join the vestibular nerve at its entry to the brain stem passing through the cerebellopontine angle

- In the brain stem the cochlear nerve → relay in the *cochlear nuclei in the lower pons* .

- From the cochlear nuclei → fibers ascends in both sides , →finally to the *auditory cortex* in the superior temporal gyrus .

Anatomical & physiological consideration of vestibular nerve and equilibrium.

- In the inner ear , the semicircular canals and the vestibule (utricle& saccule) are stimulated through their receptors by any change of body position regarding the surrounding space to maintain its steadiness and equilibrium .

→ The fibers of these receptors form the vestibular nerve which pass through→ the internal auditory meatus and canal with the cochlear nerve to emerge from the inner ear to→ the brain stem after passing through the cerebellopontine angle.

- In the brain stem , the fibers relay in → the vestibular nuclei in the lateral pons and medulla while some fibers enter the cerebellar flocculonodular lobe .

- The vestibular nuclei in the brain stem are connected to→ the :-Spinal cord ,

-3rd , 4th , 6th cranial nerves, -Medial longitudinal fasiculus , -Cerebellum, the cerebrum.

N.B: Lesions of the cochlear nerve → Tinnitus & Deafness.

Lesions of the vestibular nerve → Vertigo.

How to test the function of the vestibulocochlear nerve

A -Testing the cochlear nerve (hearing):

Test for acuity of hearing and type of deafness if presented by :

1- **Watch test or whisepering sound** → (to define the side of deafness) see if the patient can hear them or not → if not do the following :

2- **Weber's test :** a vibrating tunning fork (c=256)is applied to the forehead or vertex in the midline and the patient is asked where the sound is heard ?

-Normal → in the midline

- Nerve deafness → in the normal ear .

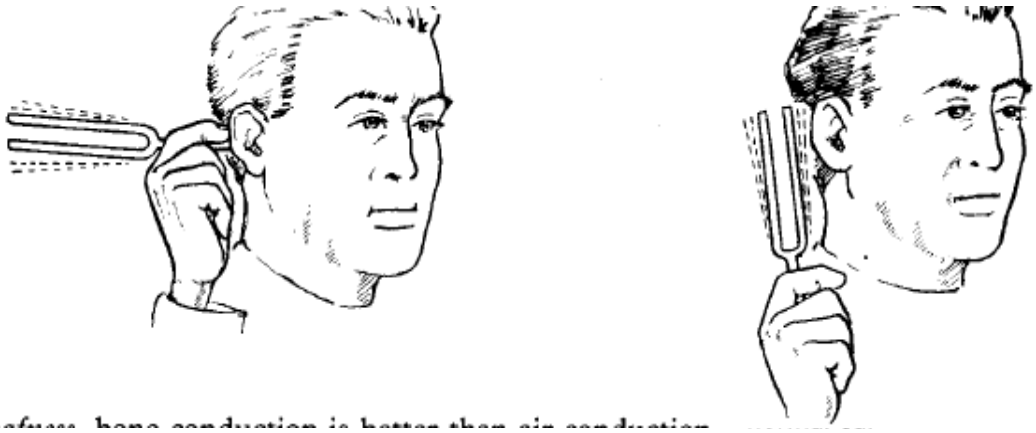
- Conductive deafness → in the affected ear.

3- **Rinne 's test** : Place the vibrating tunning fork on the patient mastoid process (for bone conduction)while the patient 's ear being occluded by the examiner's fingers , then the patient is asked when he ceases to hear the sound → then the fork is held in front of the ear (for air conduction)→ the patient is asked if he still hears the sound .

- Normally → air conduction is better than bone conduction .

- In conductive deafness → bone conduction is better than that of air .

- In nerve deafness → both air and bone conduction are impaired.



In conductive deafness, bone conduction is better than air conduction

sounds is reduced in that ear

Fig(56)

4- **Audiometry** : A quantitative method of testing hearing and should be a routine procedure in all patients with suspected hearing loss.

5- **Auditory evoked potentials :** by recording the electrical potentials originating from the inner ear , brain stem , and the auditory cortex in response to clicking sound.

B- Testing the vestibular nerve :

- Gait →unsteady in vestibular lesion .

- Nystagmus→ mainly positional nystagmus .

- Caloric test→ with head raised 30 , irrigate each ear in turn with 240 ml water at 30c and 44c consequently , normally nystagmus will occur towards the stimulated ear with warm water & away from stimulated ear with cold water

- Positional tests →which will evoke severe vertigo and nystagmus

Vertigo

Definition : True vertigo can be defined as a sense of rotation of the body in relation to the surroundings or the reverse .

-Dizziness or giddness may be used for cases without true vertigo

-Vertigo is usually accompanied with motor and autonomic manifestations e.g :

falling, nystagmus , diplopia, unsteady gait .pallor , sweating , nausea & vomiting .

Causes: disturbance of function at different levels may be involved in aetiology of vertigo :

1- Ocular causes: due to improper visual perceptions or ocular nerve lesions
2- Aural causes : OM , excess wax.
3- 8th nerve lesions
4- Cervical causes : cervical cord lesions , cervical disc prolapse , cervical osteophytes, which interfere either with cerebral blood flow or with afferent impulses originating from the different proprioceptors
5- Cortical causes : in temporal lobe lesions , ↑ ICP.
6- Cerebellar causes : mainly in flocculonodular lobe .
7- Brain stem lesions
8- Psychogenic causes : in depression , panic attacks , anexiety states.

The ninth (glossopharyngeal nerve)

***Anatomy :**

It is a mixed nerve containing motor, sensory, and autonomic fibers, originating from the medulla between vagus and accessory nerve → crosses the posterior fossa → passes out through the jugular foramen to the neck → passes downwords and forwards between the internal carotid and the external carotid arteries to the side of the pharynx where it divides to its terminal branches.

***Branches & supply:**

- Motor : only to the stylopharyngeus muscle , its weakness may lead to slight lowering of the palatal arch.
- Autonomic :To the parotid gland ,
  To the otic ganglion to exit salivary secretion.
- Sensory to : Tympanic cavity →through the tympanic nerve.
  Tonsil , lower and posterior surface of the soft palate , pharynx.
  Taste sensation from the posterior 1/3 of the tongue.

N.B :

Isolated lesions of the glosopharyngeal nerve is seldom occurred and is usually associated with that of vagus nerve , therefore, they are examined togther .

The tenth " vagus nerve"

***Anatomy:**

- It is a mixed nerve carrying motor , autonomic and sensory fibers originating in the medulla from different nuclei which give rise the fibers of both the 9^{th} and 10^{th} , → motor fibers , sensory fibers , & autonomic fibers.)
- The vagus nerve leaves the medulla by passing between the glossopharyngeal above and the accessory below → leaves the skull through the jugular foramen → in the neck it lies in the carotid sheath → enters the thorax behind the large veins.

***Branches and supply :**

1- Motor to → the soft palate , pharynx and larynx .

2- Sensory → dura matter of the posterior fossa through the meningeal branch , auricle and external auditory meatus through the auricular branch , pharynx , larynx , trachea , oesophagus ,thoracic and abdominal viscera → through the superior laryngeal and recurrant laryngeal nerves.

3- Autonomic : Cardiac → decrease the heart rate .
GIT and respiratory rate.

Symptoms & signs of 9^{th}, 10^{th} cranial nerve lesions :

1- Paralysis of the palate :

- ***Unilateral lesions*** : uvula is deviated to the normal side (muscles is pulling in action), few symptoms in the form of :post nasal disharge , snoring , slight changes in phonation , slight changes in speach .

- ***Bilateral lesions*** :

Symptoms :
- Nasal regurg. of food ,
- Nasal tone of voice ,
- Mouth breathing and snoring at night .

Signs:
- Uvula is immobile ,
- Lost palatal and pharyngeal reflexes.

2- Paralysis of the pharynx:

- Unilateral lesions : - Drooping the pharyngeal wall on the affected side .
  - Ipsilateral loss of the pharyngeal reflex .
  - Collection of frothy mucus above the oesophagus opening .
  - Difficut clearing of voice throat and coughing .
  - Difficult swallowing .

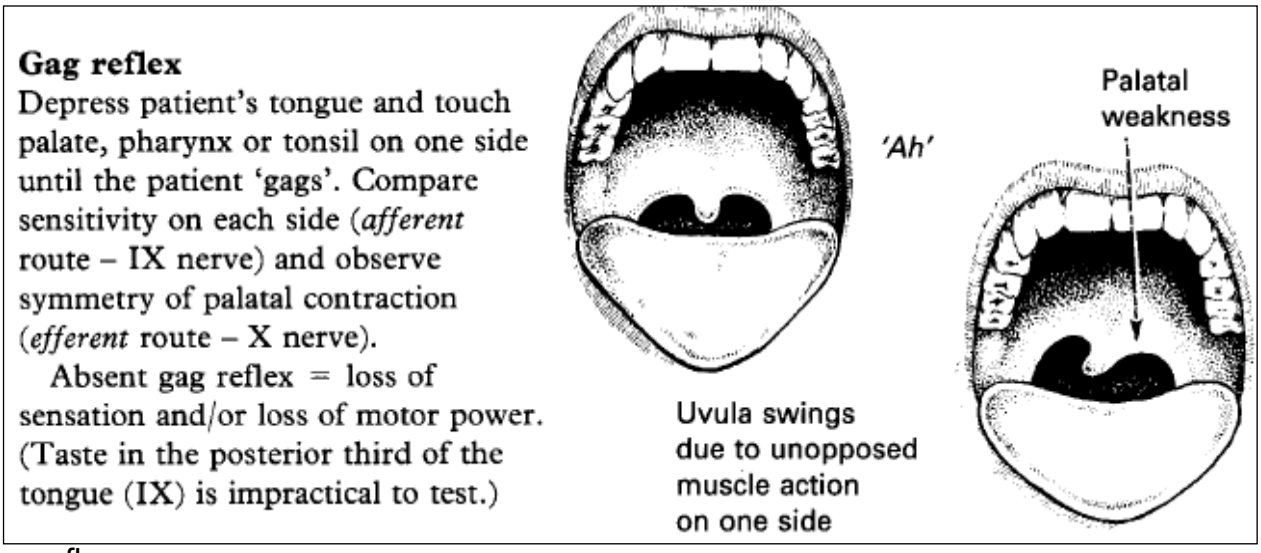
- Bilateral lesions : - Marked dysphagia . - Bilaleral loss of the pharyngeal reflex .

3- Paralysis of the larynx:

- Unilateral lesions : - Slight horseness of voice ,
  - Ipsilateral paralysed vocal cord ,
  - Slight difficult swallwing of fluids .
- Bilateral lesions : - Bilaterally paralysed vocal cords which are held close togther .
  - Weak voice , - Dyspnea on exertion ,
  - Inspiratory stridor , - Difficult swallowing , coughing .

N.B:-

- Symptoms resulting from 9th and 10th cranial nerves are collectively termed bulbar symptoms (dysphagia , nasal tone of voice, nasal regurgitation of food , hoarseness of voice,chocking)
- If the cause of bulbar symptoms is UMNL (bilateral corticobulbar lesions) → ***pseudobulbar palsy*** →associated with brisky palatal and pharyngeal reflexes and other features of pyramidal tract lesions .
- if the cause of bulbar symptoms is of lower motor neurone lesions (nuclear , infranuclear, nerve lesions) → ***true bulbar palsy*** with lost palatal and pharyngeal reflexes.



Fig(57)

****How to test the 9th, 10th cranial nerves?***

1- Ask the patient to open his mouth and to say ah and note the movement of the uvula ,

Normally , it is mobile and centralized .

Unilateral lesions → deviated to the healthy side,

Bilateral lesions → immobile

2- Palatal reflex (afferent 5th and 9th cranial nerves& efferent 10th cranial nerves):

- Ask the patient to open his mouth , touch the soft palate with tongue depressor with depression (fixation of the tongue) with another depressor → normally the soft palate will elevate , In UMNL→ exaggerated ,

In LMNL → lost .

3- Pharyngeal reflex (afferent 9th cranial nerve , efferent 10th cranial nerves)gag reflex : Fig(57)

- with opening the patient 's mouth , and as done with palatal reflex , touch the posterior pharyngeal wall → normal gaging will occur with closure of the mouth ,

In LMNL → lost .

InUMNL→exaggerated.

4- Taste sensations over the posterior 1/3 of the tongue → difficult to be tested .

<u>The eleventh "accessory nerve</u> → I t is a pure motor nerve , has two portions :

-Cranial part → arising in the medulla , joining the vagus nerve , leaves the skull through the jugular foramen to the pharynx and larynx .

-Spinal part → arises from the upper cervical cord segments C1-C3 , these roots unit together and ascend in the spinal subdural space to join the cranial part in the foremen magnum with the vagus nerve .

The spinal part enters the neck by passing downwards to supply →the sternomastoid & the trapezius.

***How to test the 11th cranial nerve ?**

Fig(58) ⟶

ACCESSORY NERVE (XI)

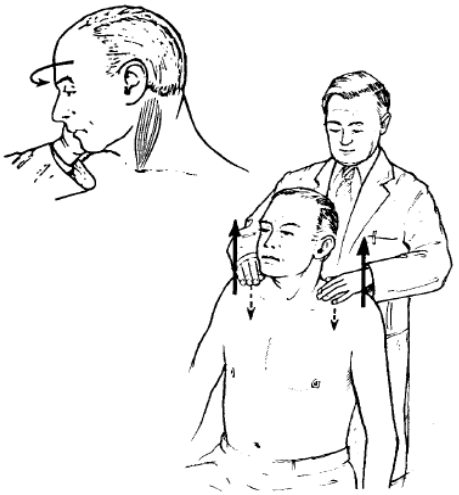
Sternomastoid

Ask patient to rotate head against resistance. Compare power and muscle bulk on each side. Also compare each side with the patient pulling head forward against resistance.

N.B. The left sternomastoid turns the head to the right and *vice versa*.

Trapezius

Ask patient to 'shrug' shoulders and to hold them in this position against resistance. Compare power on each side. Patient should manage to resist any effort to depress shoulders.



1- the cranial part examined with 9th, 10th nerve.

2- the spinal part → ask the patient to rise his shoulder against resistance → to test trapezius .

In paralysis of trapezius → the shoulder is lowered on the affected side

→ winging of the scapula ,its lower border being nearer to the midline than the upper border

→ for sternomastoid : Ask the patient to turn his neck to each side in turn against resistance .

N.B:

1-In bilateral lesions of sternomastoid → weak neck flextion , the head tends to fall forwards,.

- In bilateral lesions of trapezius → weak neck extension → head tends to fall backwards .

2- In UMNL : the cerebral hemisphere supply the ipsilateral sternomastoid and the contralateral trapzius → giving signs of both sides .

The twelfth "hypoglossal nerve"

***Anatomy:**

It is a pure motor nerve , originating in the medulla , near the midline , the fibers emerge from the medulla the posterior fossa → hypoglossal canal → leaves the skull → towards the hyoid bone → over the two carotids → to reach the tongue

***How to test the hypoglossal nerve** Fig(59)

Ask the patient to protrude his tongue , to push his check by his tongue from inside against resistance, and inspect the tongue .

***In hypoglossal nerve lesions:**

- In unilateral lesions (UMNL or LMNL)→ the tongue is deviated to the diseased side where its muscles are pushing in action Fig(51)
- In bilateral nerve lesions (UMNL orLMNL)→ the tongue can not be protruded
- In LMNL→ the tongue is wasted
- In UMNL→ the tongue is spastic.

HYPOGLOSSAL NERVE (XII)
Ask patient to open mouth; inspect tongue.
Look for – evidence of atrophy (increased folds, wasting)
– fibrillation (small wriggling movements).

Ask patient to protrude tongue. Note any difficulty or deviation. (N.B. apparent deviation may occur with facial weakness – if present, assess tongue in relation to teeth.)
Protruded tongue deviates towards side of weakness.
Non protruded tongue cannot move to the opposite side.
Dysarthria and dysphagia are minimal.

Fig(59)

unilateral paralysis of the hypoglossal muscles usually doesn't affect the articulation .

- bilateral lesions cause dysarthria , with slight dysphagia.

Peripheral Neuropathy

Definition: Means inflammation or degenerative diseases affecting peripheral or cranial nerves leading to defective motor and /or sensory and /or autonomic functions of the affected nerves.

Peripheral neuropathy may be

- Mononeuropathy: single affected nerve.
- Mononeuropathy multiplex: more than one nerve is affected in one limb.
- Polyneuropathy: affection of peripheral nerves in the four limbs.

Aetiology of peripheral neuropathy

I- Inherited polyneuropathy; eg

- Peroneal muscular atrophy.(Charcot Marie Tooth disease).
- Refsum disease.
- Porphyric neuropathy.
- Metachromatic leukodystrophy.

II- Acute idiopathic polyneuropathy:eg

- Guillian- Barre syndrome.
- Miller Fisher syndrome.

III- Chronic idiopathic polyneuropathy.eg

- Chronic inflammatory demylinating polyneuropathy (CIDP)

IV- Polyneuropathy with infections.eg

- Leprosy
- Diphtheria
- HIV infections
- Lyme disease

V- Neuropathy due to systemic medical disorders.eg

- Diabetes mellitus
- Acromegaly
- Chronic renal failure
- SLE
- Hypothyroidism
- Primary biliary cirrhosis.

VI- Carcinomitous neuropathy - paraneoplastic neuropathy

VII- Drug induced neuropathy.eg

- Alcohol
- Phenytoin
- Corticosteroids
- Dapsone
- Vincristine
- Chloroquine
- INH
- Metronidazole
- Colchicine

VIII- Metal poisoning (toxic) neuropathy.eg

- Lead
- Mercury
- Irradiation
- Arsenic
- Organophosphorus

IX- Vascular causes: eg

- Systemic vasculitis
- Major arterial occlusion
- Collagen diseases

X- Vitamin deficiency neuropathy.eg

- B1
- B6
- B12
- Vit E.

Clinical diagnosis of polyneuropathy:

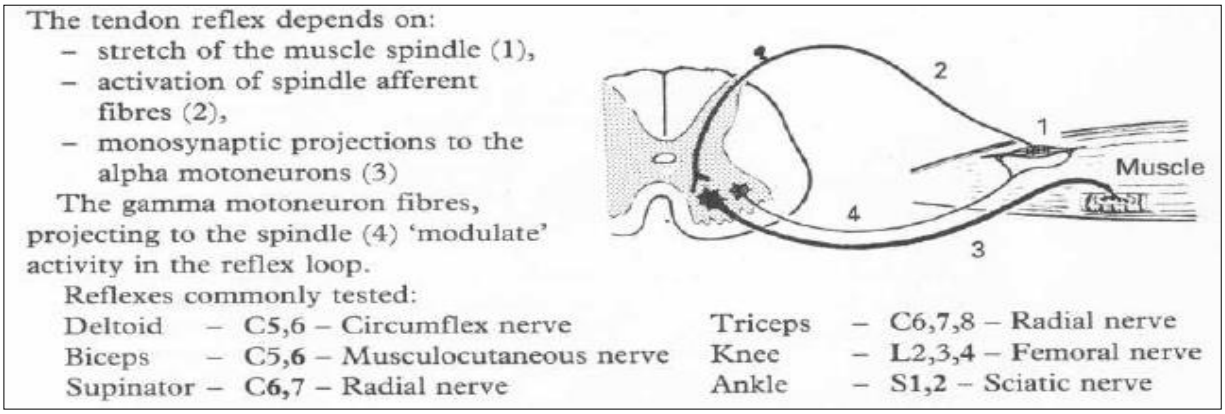
In general, clinical presentation of peripheral neuropathy may be in the form of at least one or all of the following;

I- Motor:

A- Weakness: of LMN features" characterized with weakness affecting

- Both UL and LL ,more in LL
- Bilateral symmetrical
- Distal more than proximal usually leading to foot drop and wrist drop.
- Extensors are more affected than flexors

B- Hypotonia, hyporeflexia usually affecting distal limb parts. Fig(60)



Fig(60): (Commonly tested deep reflexes)

C- Wasting is usually distal.

D- Gait is usually high steppage.

Subjectively the patient will complain of

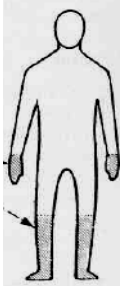
- Easy triping on waking
- Difficult and high steppage gait.
- Falling of shoes “Can’t keep it"
- Difficult down stairs.
- Falling of objects from hands.
- Weakness of hand grip and catching objects.

II- Sensory:

- Impairment of superficial sensations leading to glove and stock hypothesia .Fig(61)
- Impairment of deep sensation.

Subjectively the patient will complain of

- Pain and parathesias in the form of numbness, hotness in the distal limb parts
- Falling of shoes unconsciously.
- Falling of objects unconsciously.



Fig(61)

III- Autonomic:

- Coldness and cyanosis of the affected parts.
- Loss of hair, oedema, brittle nails, trophic ulcers.

Important clinical hints in diagnosis of peripheral neuropathy.

The clinical presentation may be

1- Mainly pure motor as in;
   - lead neuropathy
   - Porphyric neuropathy
   - dapsone neuropathy
   - Beri-Beri.
   - G.B. syndrome.
   - Organophosphorus.
2- Mainly pure sensory as in;
   - Vitamin E, B12 deficiencies neuropathy.
   - Diabetic neuropathy "some types"
   - Leprosy.
3- The weakness may be proximal as in;
   - Porphyric neuropathy
   - Demyelinating neuropathies.
   - G.B. syndrome.
   - Diabetic neuropathy(some types).
4- The autonomic features may be prominent as in;
   - DM.(impotence,Neurogenic bladder)
   - Hereditary sensory autonomic neuropathy.
5- In focal neuropathy, the wasted weak muscles, the lost reflex arc, and the sensory disturbances are restricted to the distribution of the affected nerve(s) or root(s).
   - Causes of focal neuropathy includes; irradiation, compression, leprosy, trauma, D.M., vasculitis, and tumors (Local infiltration).
6- Demyelinating neuropathy is suggested in the presence of
   - Proximal muscle weakness.
   - Relative lack of muscle wasting.
   - Lost deep sensations compared to relative preservation of superficial sensation.
7- Inherited neuropathy is suggested in the presence of the following;
   - Positive family history of neuropathy or parental consanguinity.
   - Absence of positive sensory symptoms.
   - Often other system affection.

Specific types of neuropathy:

1- Peroneal muscular atrophy:

-It is a hereditary motor and sensory neuropathy usually associated with autosomal dominant mode of inheritance.

- The age of onset of this type is usually in the 1st or 2nd decades of life,

- It is a form of demylinating neuropathy with onion bulb changes on nerve biopsy and palpable nerve thickening

- Of gradual onset and progressive course.

▸ It is presented clinically with

- Distal leg muscle wasting and weakness up to the lower third of the thigh giving the leg picture of inverted bottle appearance.
- Frequent skeletal deformities especially pes cavus.
- There is discrepancy between wasting “marked” and weakness “less evident ".
- Lost ankle jerks and total areflexia occur in half of the patients.
- All modalities of sensations may be impaired distally. NB- Positive sensory symptoms (e.g. parathesias) →cast doubt on diagnosis.
- May be associated with scoliosis, tremors, ataxia, pupil changes.

2- Guillian Barre syndrome:

Synonyms: Acute post infective polyneuritis, acute inflammatory demylinating polyneuropathy.

It is a symmetrical, predominantly motor neuropathy, frequently involving facial and other cranial motor nerves and commonly preceded by viral or other infections.

***Aetiology of Guillian Barre syndrome:**

- Genetic factors may have a role, more common in females.
- About half of patients were found to be preceded by infections days.up to 1 month before the disease e.g., URTI or GIT infections.
- High incidence of preceded viral infections.
- May be associated with lymphomas, follow vaccination, steroid therapy, blood transfusion, or organ transplantation.

***Clinical presentation:**

- An aura of URTI or GIT infections is usually reported days before the disease with backache.
- Paraesthia is often the 1st heralding symptom for the disease and LL are more commonly affected.
- Hours or days later, acute or subacute onset and progressive course of weakness or paralysis starting in both LL, usually ascends to involve trunk, respiratory, and both upper limbs muscles.
- Weakness is usually symmetrical, more proximal of LMN features with areflexia, hypotonia, or flaccidity.
- Progress of weakness takes usually takes1-4 weeks for maximum.
- Sensory affection is mainly subjective, e.g. parathesias, pain in calf muscles, but objective sensory changes as mild glove and stoke hypothesia may be present. The presence of sensory level will cast doubt on the diagnosis.
- No definite or persistant sphincteric disturbance can be detected, except in minority of cases and usually transient, but persistent &definite sphincter disturbance will rule out the diagnosis.
- Cranial nerves affection is common e.g. III, VII, X.
- Involvment of UL and trunk muscles may herald the occurrence of chest infection and respiratory failure and ventilation may be required.
- Hyponatremia, papillodema, and autonomic disturbances, may develop .
- Prognosis is usually good, and mortality rate is about 5%.

***Investigations:**

1-CSF analysis: CSF shows cytoalbuminous dissociation and the CSF proteins increase up to several gms /litre and the cells are within normal.

2-NCS show marked slowing, conduction blocks.

***Management:**

1- Complete bed rest.&physiotherapy
2- Care of bed ridden patients as usual.
3- Care of respiration and bulbar muscles.
4- Corticosteroids, e,g. oral prednisolone 60-120 mg/day, but now most literatures suggest that steroids have no role in management of GBS.
5- Plasma exchange is highly needed within the 1st week for 4-5 sessions.
6- High dose IV immunoglobulin may be superior to plasma exchange 0.4 gm /kg for five days.

1- Diabetic neuropathy:

D.M., is one of the most common causes of disabling polyneuropathy, different types may be present e.g.

- Diabetic sensori-motor neuropathy.
- Diabetic autonomic neuropathy.
- Diabetic proximal neuropathy.
- Diabetic mononeuropathy (cranial or peripheral)
- Diabetic trunkal neuropathy.

The same patient may develop two or more of these types.

Pathogenesis:

Multiple pathogenic mechanisms are involved in development of diabetic neuropathy.

- Multifocal ischemia.
- Nutritional factors mainly vitamin deficiencies.
- Effect of toxic products.
- Persistant hyperglycemia , causing sorbitol accumulation as a result of enhancing aldose reductase activity.

A- Diabetic sensori-motor neuropathy;

- Common in IDDM.
- Sensory symptoms predominate as parathethesias , burning and lancinating pains and sensory ataxia with positive Rombergism are common especially in the LL.
- Hands are rarely involved.
- Lost ankle reflex.
- Autonomic features may develop in the form of
  - Impairment of sweating.• Warm dry foot.
  - Foot ulceration. • Neuropathic joints.
- Distal muscle weakness and wasting may develop in long standing cases.

N.B: Neuropathy may be the 1st symptom of D.M.

B- Diabetic autonomic neuropathy;

- Common in IDDM.
- Presented with;
  - Abnormal sweating. • Vomiting
  - Diarrhea. • Postural hypotension.
  - Bladder dysfunction (precipitancy or urgency).
  - Cardiac arrhythmias
  - Impotence.

C- Diabetic mononeuropathy;

- Affects peripheral or cranial nerves in diabetics.
- Ocular nerves are common to be involved "mononeuritis cranialis".
- Peripheral nerves liable to compression are usually affected e.g.
    - Radial nerve around humerus.
    - Median nerve in carpal tunnel.
    - Ulnar nerve in cubital groove.
    - Lateral cutaneous nerve of the thigh at inguinal region.

Management:

1- Strict control of blood sugar.
2- Aldose reductase inhibitors " sorbinol"
3- Vasodilators and tonics.
4- Carbamazepine, phenytoin, amitriptyline, may help in sensory discomfort.
5- Skin care is very essential.

4- <u>Vitamin deficiency neuropathy:</u>

A- Vitamin B1 deficiency "Beri-Beri"(thiamine deficiency)

- Common in malnourished and alcoholic patients.

* Dry Beri-Beri is associated with
    - Sensory-motor neuropathy mainly motor.
    - Distal leg muscle weakness, wasting and leg oedema occurs,
    -Neuropathic limb pains or paraethesias may develop.
* Wet Beri-Beri is associated with symptoms and signs of congestive heart failure and in chronic cases the picture of polyneuropathy with H.F. develops.

- In alcoholics, thiamine deficiency is encountered in Wernicke's encephalopathy.

B- Vitamin B6 deficiency " Pyridoxine deficiency "

-**D**ermatitis, red brown hyperkeratotic rashes in the sun exposed areas and bony prominences.

-**D**iarrhoea, with gasterointestinal symptoms, dyspepsia, colics, vomiting.

-**D**epression, fear, suicidal thoughts .

-**D**ementia.

-**S**ensori-motor neuropathy mainly sensory with paraethesias, pain and tender distal leg muscles.

C- Pellagra:

A disease caused by multiple vitamin deficiencies chiefly nicotinic acid and B6 and characterized by

- Cutaneous lesions
- Mental changes.
- Glossitis and GIT lesions.
- Degeneration of the brain, spinal cord, peripheral nerves.

Clinically characterized by(Ds syndrome)

- Dementia. - Diarrhoea - Dermatitis
- Depresson - Diplopia - Dysphagia
- Dytharthia - Degeneration of spinal cord with spastic paraplegia.
- Degeneration of the peripheral nerves with polyneuropathy.

D- Vitamin B12 deficiency (pernicious anemia, subacute combined degeneration of the cord)

***Pernicious anaemia**:

- Peripheral nerves>> neuropathy; mainly sensory with paraethesias in the feet, distal loss of all sensations(glove & stoke), lost ankle jerks.

***Subacute Combined degeneration.**

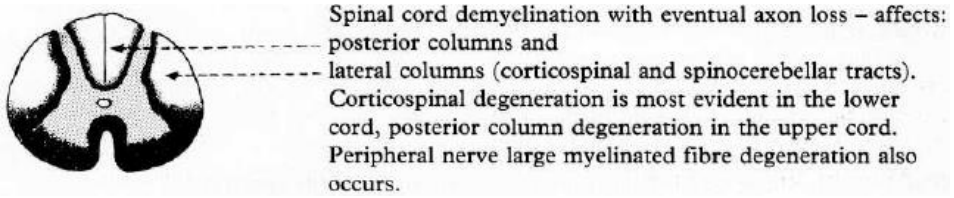
-Degeneration of → Fig(62)

-Spinal cord leading to paraparesis, spasticity, sensory ataxia with late sphincteric disturbances.

- Optic nerves >>bilateral optic atrophy.

-Peripheral nerves >> mainly sensory neuropathy with glove stoke hypothesia

- Brain >> leading to dementia, impaired memory, confusion, and psychiatric disorders.



Fig(62)

E- Vitamin E deficiency;

- Pure sensory neuropathy - Prominent cerebellar ataxia
- Pigmentary retinopathy. -Impotence, lost libido

5- Leprotic neuropathy:

Leprosy is a skin disease caused by mycobacterium leprae, an acid fast bacillus with infection of the skin, mucous membranes and peripheral nerves. The combination of skin lesions and peripheral neuropathy is the hallmark of leprosy.

There are two clinical forms of leprosy

1- Tubercloid leprosy:

- Sharply demarcated hairless, anaesthetic, erythematous plaques
- Sensory or motor loss in the distribution of one or two damaged nerves, which are often palpable, the greater auricular, and superficial peroneal nerves are commonly affected.

2- Lepromatous leprosy:

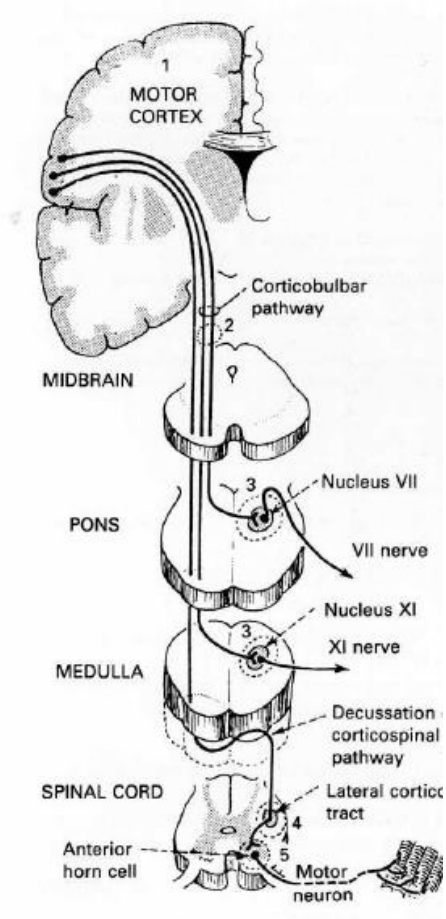
- Skin thickening with characterstic leonine facies.
- Thickening of the nose, ear lobes, with perforation of the nasal septum may occur.
- Nerves are diffusely and progressively involved leading to mononeuritis multiplex.

Motor neuron diseases

These disorders are characterized by selective loss of function of the Lower and\or upper motor neurons due to selective degenerative changes which involve;

→ The ***anterior horn cells*** of the spinal cord,

→ The ***corticospinal tracts*** and the ***motor nuclei of the brain stem***. Fig(63)



Classification of Motor neuron diseases

1-***Pure upper motor neuron involvement***

1. Hereditary spastic paraplegia
2. 1ry lateral sclerosis
3. Pseudobulbar palsy

2-***Pure lower motor neuron involvement***

1. Progressive muscular atrophy
2. Progressive bulbar palsy (hereditary bulbar palsy)
3. Spinal muscular atrophies

3-***Combind upper and lower neuron involvement***

*(***Amyotrophic lateral sclerosis***)

Fig(63) ⟶

1-Hereditary spastic paraplegia

*Characters:

-Autosomal dominant or recessive inheritance

-Age of onset may be early before 20 years but not later than 40 years.

-Gradual onset, progressive course

-There is degeneration of the crossed pyramidal tract

-The dorsal root ganglia, posterior roots & the peripheral nerves are spared

*Clinically:

-Weakness of both LL of UMN features

-Difficult spastic gait, tendency to trip.

-Hyperreflexia, clonus in both LL

-Stiffness and spasticity of both legs

-Bilateral extensor planter reflex

-Upper limbs: may be slightly effected in the form of

*brisky reflexes **mild weakness, *** Stiff finger

-No sensory level,No sphinecteric disturbances

NB -Deep sensory impairment (vibration, position) and urgency may be present only in 10% of patients.

2-1ry lateral sclerosis

Spasticity , clonus,hyperflexia,extensor planters and pyramidal patterns of weakness in both LL (mainly) .

-Bladder,bowel,sensory and sexual functions are usually normal

3-Progressive muscular atrophy

-Developed first in the hands then the forearm muscles ,usually asymmetrical, and flexors usually before extensors

-Early proximal presentation is rare

- Weakness, hypotonia, lost reflexs, wasting and fasciculation are the cardinal features with progressive course and finally to involve the LL. muscles and the respiratory muscles

- Bladder, bowel, sexual and sensory functions are normal

4-Progressive bulbar palsy

-Involvement of motor cranial nerve nuclei of the brain stem will lead to

→ Tongue 1st to be affected leading to wasting, fibrillation and dysarthria.

→ Next ..Bulbar nerves giving rise to bulbar manifestation

-Dysartheria, dysphonia, dysphagia, -Nasal regurgitation of fluids

-Chocking - lost palatal &pharyngeal reflexes

→Weakness of extensor muscles of the neck giving rise to difficulties in maintaining head posture.

→Weakness of orbiculars oculi and jaw muscles.

5-Amyotrophic lateral sclerosis

***Epidemiology & Aetiology :**

-Disease of late middle life with onset between age of 50 and 70years.

-Male predominance

-Genetic inheritances are common with autosomal dominant inheritance –chromosome 21

-Other aetiological factors may play role e.g

* Post viral infection. * Immunological role

* Premature aging process of the motor neurons.

. *Exposure to toxic elements e.g

- Heavy metals, -Solvents -Smoking

-Cycad plant(neurotoxin ,cycasin derived from the plant cycas)

. ***Clinical features:**

NB. An essential feature to diagnose ALS is the presence of both upper and lower motor neuron sings within muscles supplied by the same spinal segment i.e the same muscles of the limb e.g exaggerated deep reflexes in the upper limbs in the presence of wasting &weakness of the same limb .→ Usually the following clinical features are reported.

* Weakness, wasting of UL muscles mainly distal with brisky reflexes, hypotonia
* Widespread fasciculations
* Spastic paraparesis with exaggerated deep reflexes and bilateral extensor planter response
* Abdominal reflexes are usually normal
* Bladder, bowel, sexual and sensory functions are usually normal
* Pseudobulbar palsy may occur with progression of the disease leading to bulbar manifestations and emotional liability
*Dementia of frontal lobe type with mild cognitive functions disorder may occur

***** Investigations:**

*Motor nerve conduction , and Sensory nerve conduction studies are usually normal
* Fasciculation and fibrillation potentials on EMG.
*Giant motor unit potentials. * Mild to moderate increase of CK.
* Muscle biopsy shows features of denervation
* Increased CSF protien

*****Differential diagnosis;**

1-Cord lesions eg; cervical spondylosis, syringomyelia, cord tumors
2-1ry muscle disease
3- Cerebrovascular lesions leading to bulbar palsy (corticobulbar tract lesion)
4-Lower motor causes of true bulbar palsy eg. Myasthenia gravis, diseases of base of the skull

*****Prognosis;**

* Patients with later onset or those with bulbar palsy have the worst prognosis;
* Inhalation pneumonia, respiratory failure and malnutrition are common sequale

*****Management;**

- Unfortunatly the disease is incurable till now,but the followings can be used,
- Thyrotrophin releasing hormone, Anticholineastrase, Corticosteroids
- Antiglutamate medications

Muscle diseases

(myopathies)

**Definition:

Myopathy is used to define any primary muscle disease in which the patient's symptoms and/or signs are related to the muscular system ,and are attributed to changes which occur in the muscles themselves.

**Types of muscle diseases:

1. Congenital muscular dystrophies.
2. Myotonic myopathies & disorders.
3. Inflamatory muscle diseases.
4. Myasthenic muscle diseases.
5. Metabolic myopathies.
6. Endocrinal myopathies.
7. Congenital myopathies.

I) Congenital muscular dystrophies:

Genetically determined

-X-linked -Autosomal.Recessive -Autosomal.Dominant

II) Myotonic myopathies & disorders:

1. Myotonia congenita.
2. Myotonia dystrophica.
3. Paramyotonia.
4. Drug induced & symptomatic myotonia.

III) Inflammatory muscle diseases:

may be due to

1-Infective agents

(viral, bacterial, fungal, protozoal)

2-Idiopathic *

Polymyositis * Dermatomyositis *Inclusion body myositis

3-Drug induced

4-Collagen disease, immune deficiency states

5-Malignancies

IV) Myasthenic muscle diseases:

1. Myasthenia gravis
2. Congenital myasthenia
3. Neonatal myasthenia
4. Drug induced myasthenia
5. Symptomatic myasthenia

V) Metabolic myopathies:

1. Metabolic bone diseases
2. Chronic renal.failure
3. Glycogen storage disease
4. Lipid storage disease
5. Periodic paralysis syndrome
6. Drugs & toxins
7. Alcoholic myopathies
8. Malignant hyperpyrexia
9. Malignant neuroleptic syndrome
10. Nutritional myopathies

VI) Endocranial myopathies:

A-Thyroid gland diseases. - Hyper,& hypothyroidism

B-Pituitary gland diseases.

-Addison's disease -Conn's syndrome

C-Cushing disease

D-Steroid myopathy

VII) Congenital myopathies.

Clinical features of muscle disease in general:

From the history we can elicit the following :

- Proximal leg weakness→ difficulty in getting up from the ground, getting off of toilet, getting out of car.
- Proximal arm weakness → difficulty in lifting or carrying objects, combing hair, wearing clothes.
- Symmetrical weakness→ **N.B** weakness restricted to one side or one limb is rare to be caused by myopathy.
- Intact sensations → **N.B** actual sensory loss or changes should not occur with myopathy.

  N.B: There are other symptoms which suggest muscle disease (e.g. fatigue, cramps, and stiffness)
- On examination , the followings can be detected:
  1) Power → symmetrical, bilateral, proximal weakness.
  2) Reflexes → decreased.
  3) Tone → decreased.
  4) Muscle state → may be normal, pseudo hypertrophy[Fig(68)], or wasted.
  5) Sensations are intact.
  6) Weakness of pelvic girdle muscles → waddling gait and/or Gower's sign.
  7) Weakness of shoulder girdle muscles → slopping of the shoulder, winging of the scapula.
  8) CPK is usually elevated in most types.
  9) EMG shows features of myopathy

Muscular dystrophies

** *Types*:

(1)-*X- linked muscular dystrophy* :

- Severe ; Duchenne type.
- Benign ; Becker type.

(2)-*Autosomal recessive dystrophy* :

- Limb girdle type.
- Distal type.
- Childhood type .
- Congenital type

(3)-*Autosomal dominant dystrophy* :

- Distal type.
- Ocular type.
- Occulopharyngeal type.
- Fascioscapulohumeral type

**Clinical characters and features of muscular dystrophy in general :

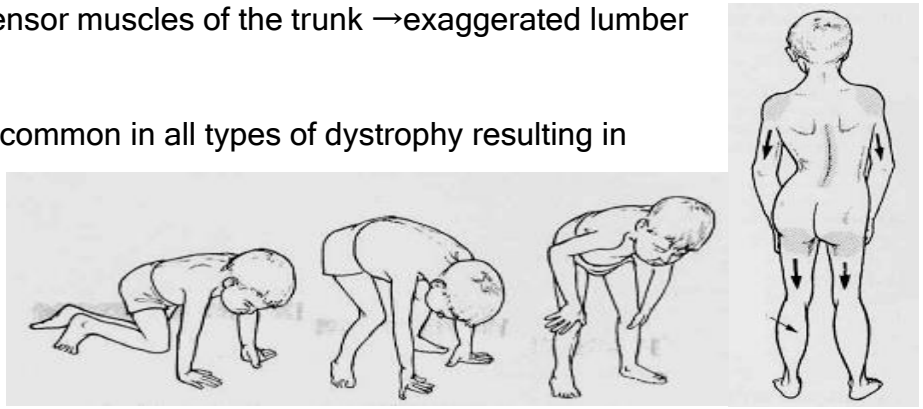
1. Bilateral, proximal and symmetrical weakness of both upper limbs (shoulder girdle muscles) and both lower limbs (pelvic girdle muscles), associated with LMN features → hypotonia, hyporeflexia and wasting.
2. There is selectivity of the involved muscles e.g. :
   - In the upper limbs → deltoid and triceps are spared.
   - In the lower limbs → calf muscles are spared.

   NB. The selectivity pattern is highly suggestive of muscular dystrophy

- Weakness of abdominal muscles→ pot belly abdomen.

  ↓
- Weakness of extensor muscles of the trunk →exaggerated lumber lordosis.
- Contractures are common in all types of dystrophy resulting in skeletal deformities.
- Gower's sign Fig(64)

  → ●Waddling gait



X- linked muscular dystrophy .

- Restricted in males , very rarely to occur in females (genetic mutations)
- Marked rise of serum CPK. ■ Early age of onset.
- Mainly due to deficiency or missing of gene product protein which is termed dystrophin which is essential component of muscle fiber protein.

Comparison between both types : Table (4)

Data	Duchenne	Becker
Age of onset	First half of the first decade of life (3-5years)	5-10 years
Severity	Severe	Less severe
Skeletal deformities	Common and severe	Less common and late
Course	Progressive → bed ridden by the age of 10 years usually and death may occur 10-20 years later.	Less progressive and bed ridden at age20 -30 years
Skeletal atrophy	Marked	Less marked
Macrglossia	Common	Less
Cardiac involvement	Common	Less
Speech and intellectual function affection	Common	Less
Muscle hypertrophy	Common in calf muscles, less in quadriceps and deltoid. Occur in 90 % of cases. Early it is true hypertrophy, and then becomes pseudo.	Less common
CPK	Markedly elevated 20-200 times than normal , decreases by 20% / year till it becomes normal with fibrosis of muscles	Moderately elevated
Respiratory insufficiency	Common	Less common

** Some clinical hints of other types of muscular dystrophy:*

- **Limb girdle dystrophy:**
  - Age of onset 20-30 years.
  - May start scapulohumeral or pelvifemoral.
  - Disability is severe within 20 years of onset (or up to 70 years of age).
  - CPK is moderately elevated or normal in chronic cases.
- **Childhood muscular dystrophy:**
  - Age of onset 2-14 years.
  - Common in North Africa and Middle East.
  - Slowly progressive.
  - Disability usually at the age of 40 years.
- **Distal types:**
  - Older age of onset (40-60 years).
  - Weakness begins in small muscles of the hands and feet → finally spreads to involve proximal muscles.
- **Ocular , pharyngeal and facial myopathy :**
  - Are involved in certain types .

*** Investigations of a case of muscular dystrophy:**

1) Increased CPK level in serum.
2) Muscle biopsy:
   a. Marked variation of myofiber size.
   b. Degeneration of muscle fibers.
   c. Fibrosis and fibro adipose replacement of skeletal muscle fibers.
   d. lack of dystrophin in muscle fibers.
3) EMG study:
   a. Increased polyphasic action potentials.
   b. Short duration of action potentials.
   c. Decreased amplitude and duration of mean action potential.
4) DNA studies and genetic counseling.

*** Treatment:**

- No specific drug treatment is recommended at present .
- Treatment includes :
    1. Supportive treatment may be of value e.g. :
        a) Some vitamins (e.g. vitamin E).
        b) Physiotherapy is of value in delaying :the march of weakness.&onset of contractures and skeletal deformity.
        c) Wearing light spinal supports and specific shoes are of value in delaying skeletal deformities.
        d) Ventilatory support “assisted respiration “is needed in late terminal stages when respiratory failure worsen the course of the disease
        e) Psychological support.
    2. Infrequent course of methyl predinsolone “1mg/kg/day”in short periods may be of value in transient improvement of power.
    3. Dealing with complications e.g. bed sores pneumonia.
    4. Genetic counseling before marriage and limitation of intermarriage between families of closed relations to minimize born of further affected siblings

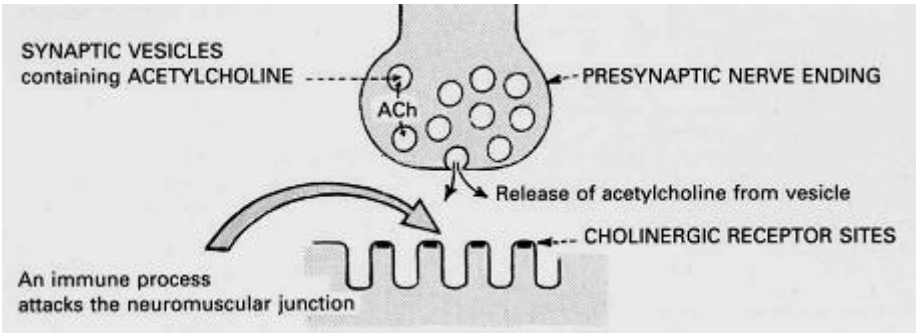
Myasthenia gravis (MG)

****Definition:**

A chronic disease due to disorder of transmission at the myoneural junction as a result of formation of circulating auto antibodies “autoimmune” against acetyl choline (Ach) receptors , clinically characterized by muscular fatigability , which may be restricted to a group of muscles , with descending march course and tendency to relapse and remission , and in later stages associated with permenant weakness.

****Aetiology:**

- Production of auto antibodies against acetylcholine receptors by thymus gland is the main cause . Fig(65)
- There is a close relation between Myasthenia gravis and other autoimmune diseases e.g. thyrotoxicosis, SLE, MS and rheumatoid arthritis.
- Myasthenic features can be produced by some drugs e.g. phenytoin, magnesium, streptomycin, pencillamin.
- Transient myasthenia may occur in infants born of myasthenic mothers.



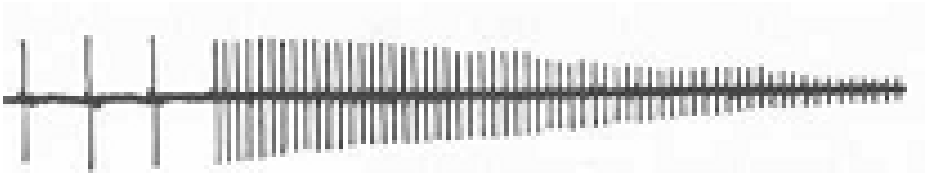
(Fig(65)

**Clinical features:

- At any age, in both sexes, but more in females and middle age .
- The main complaint is fatigability which usually starts and may be restricted to ocular, facial and bulbar muscles.
- The fatigability appears on repeated or sustained movements e.g. up gaze, talking or counting.
- The course is usually fluctuating with periods of relapse and remission, may be stationary , grave or progressive.
- Usually the fatigability on spreading, take a descending march course.
- In late stages, the involved muscles become weak.
- Involvement of the ocular muscles will lead to → diplopia, dropping of the eye lids.
- Involvement of the facial and bulbar muscles will lead to → dysphonia, dysarthria and dysphagia. & nasality of voice.
- In case of involvement of limb muscles→ proximal muscles are more involved.
- Usually the thymus gland is hyper plastic or site of neoplasm.
- There is no wasting of muscles and reflexes are normal or brisky.

**Diagnosis:

1. Clinical diagnosis→ by asking the patient to count up to 100 , to talk continuously for 5 minutes or to do up gaze for 5 minutes → fatigability .
2. Therapeutic test: tensilon test (anti choline esterase): 2 mg of tensilon are injected IV→ if there is no allergic reaction→ inject 8 mg→ rapid improvement of fatigue.
3. Detection of serum anti Ach receptors antibodies by radioimmunoassay (RIA).
4. X ray chest, CT chest to detect thymoma.
5. T_3, T_4 for thyrotoxicosis.
6. EMG study: decreased amplitude and decrementing response in the amplitude of CMAP in response to repeated stimulation. Fig(66)



Fig(66) **EMG in MG**

decrementing response in the amplitude of CMAP in response to repeated stimulation.

****Treatment:**

1. Anticholine esterase :
   a- Neostgmine 15- 30 mg/6hours. B-Mestinon 60 mg / 12 hours.
2. Prednisolone 40- 60 mg /day or 120 mg every other day.
3. Immunosuppressive drugs e.g. azathioprine 2- 2.5 mg /kg/ day orally.
4. Thymectomy must be done in all patients with thymoma.
5. In severe and intractable cases :-IV immunoglobulins -Plasma exchange.

Myotonic muscle disorders

Definition:

- Myotonic disorders are primary muscles diseases characterized by myotonic phenomena which is in the form of delayed relaxation of a skeletal muscle after its contraction either voluntary , electrically or mechanically and associated with electrical after discharge in the EMG .
- Clinically it may be demonstrated as a slowness in relaxation of the hand grip or by a persisting dimpling after sharp blow on a muscle belly e.g. in the tongue, thenar muscles or deltoid muscle.

Hereditary syndromes of myotonia:

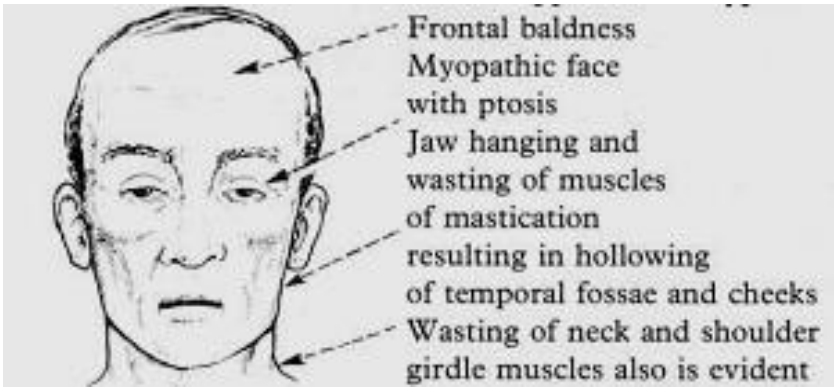
- Genetically determined, autosmal dominant inheritance,may be recessive.
- Alteration of K^+ and Na^+ channels are involved in the pathogenesis.
- There are three different types :

1- Myotonia congenita:

a. Early age of onset, usually since birth.
b. Myotonia is generalized giving painless stiffness of muscles, feeding difficulty, typical strangled cry.
c. Myotonic features are exaggerated by rest and cold and relived by warm and exercise.
d. Diffuse muscle hypertrophy is common and persist through out life giving the patient Herculean character.
e. Associated psychosis may occur.

2- Myotonia dystrophica :

a. Late age of onset 20- 50 years.
b. clinically presented with : Fig(67)
   - Distal muscular dystrophy.
   - Trophic changes e.g.
     1- Cataract, frontal baldness, poor vision. 2- Gonadal atrophy, lost libido.
     3- Cardiomypathy. 4- Dementia, 5 -Hypersomnia.
   - There is characteristic facial appearance:
     1. Sady faces (expressionless face – sunken cheeks).
     2. Scraggy neck (swan neck), eyelid drooping.



Fig(67) Myotonic features
(facial appearance)

3- Paramyotonia congenita:

- characterized by myotonia and attacks of generalized muscular weakness only on exposure to cold.

N.B: - Drug induced myotonia→ as with clofibrate, beta blockers.
- Symptomatic myotonia →as with polyneuropathy & polymyosites

@Diagnosis:

- Clinical diagnosis.
- EMG: - electric after discharge. – Dive bombers sounds on loud speaker on EMG.
- Muscle biopsy: aggregation of small nuclei within muscle fibers, abnormal muscle spindles.

@Treatment:

- Phenytion 100 mg tds→ effective.
- Acetazolamide →also effective.
- Quinine, steroids, procainamide → may be effective.
- Tocainide 400- 1200 mg /day → also effective.

Inflammatory muscles diseases (myositis)

 # Definition:

Inflammatory muscle diseases may be acquired or idiopathic, clinically characterized by muscle pain and tenderness associated with muscle weakness and wasting, histopathologically characterized by muscles fiber necrosis , regeneration and interstitial inflammatory cell infiltration .

 # Types & causes of myositis:

1) Infective agents :
    a. Viral myositis e.g. with influenza virus, HIV, HSV.
    b. Bacterial: acute suppurative myositis as after crush injuries, complicating bed sores, osteomyelitis and abscess.
    c. Parasitic myositis as with toxoplasmosis, cysticercosis.
2) Connective tissue diseases:" collagen diseases " as in :
    a. Rheumatoid arthritis, sarcoidoisis, SLE.
    b. Polyarteritis nodosa, progressive systemic sclerosis.
3) Idiopathic polymyositis and dermatomyositis :
    a. May be autoimmune. -Common in females. -More common in adult life.
    b. clinically :
        I. Onset is subacute, rarely to be acute.
        II. Muscle pain & tenderness in > 90 % of cases.
        III. Subacute proximal weakness.
        IV. Skin manifestations in cases of dermatomyositis , in the form of :
        -butterfly erythema in the face and exposed area of the chest (V shaped area)
        - Periorbital oedema. - Congestion of the nail bed.
        -Ulceration over bony prominence.
        -Thick, unelastic skin of fingers and face.
        V. Joint pain & stiffness in >25% of cases.
        VI. Involvement of neck muscles is common.
        VII. Involvement of facial muscles , extra ocular muscles and distal limb muscles may occur (rarely)
        VIII. The deep tendon reflexes may be depressed in the affected muscles but commonly to be found brisky despite of severe weakness.
        IX. Respiratory disorders and myocardial damage and pericarditis may occur.
        X. The course of the disease is variable, may arrest spontaneously, may deteriorate seriously, and may fluctuate.
        XI. Associated malignancies in > 20% of patients of dermatomyositis.

 # Diagnosis:

- Clinical diagnosis.
- EMG study :
  - Decreased duration and amplitude of action potentials.
  - Increased incidence of polyphasic potentials.
  - Detection of fibrillation potentials.
- Histopathologically :
  - Muscle fiber necrosis, phagocytosis and regeneration.
  - Interstitial, perivascular and perifasicular inflammatory cell infiltration.
- Laboratory : increased levels of :- CPK. - Aldolase. - Transaminase.

 # Treatment:

- Prednisolone orally 60mg/day or 120 mg/ every other day.

- On clinical improvement ..decrease the dose to maintenance dose upto 1-2 years .

- Immune suppression drugs (azathioprine 2- 2, 5 mg/ kg/ day) combined with prednisolone is the commonest combination for 1-2 years.
- IV methyl prednisolone is better in severe cases.
- IV immunoglobulin and plasma pharesis

Paraplegia and spinal cord lesions

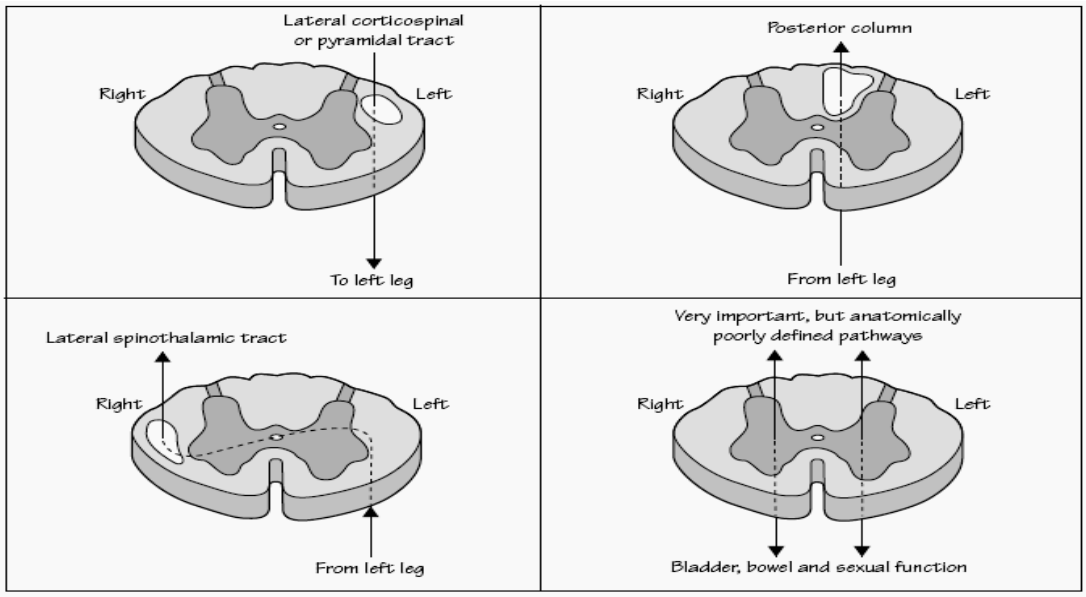
Anatomical consideration

-The spinal cord lies within the vertebral canal, it extends from the lower part of the medulla at the level of the foramen magnum "level of atlas" down to the level of L1 or L2.

- It 's oval in shape with two enlargements (cervical and lumbar) from which outflow of nerves to the four limbs occurs , the cord is formed of segments ***,(8) cervical , (12) thoracic or dorsal ,(5) lumbar,(5) sacral , (1) coccygeal*** ,from each side of each segment two pairs of roots emerge → posterior dorsal root (sensory) & anterior ventral root (motor), one ventral and the corresponding dorsal root join together to form→ a spinal nerve.

-The cord like the brain is surrounded by three meninges from inwards to outwards →the pia matter,--the arachinoid matter which is separated from the pia matter by the subarachinoid space which contains CSF then the outermost dura matter which lines the vertebral canal.

-On transverse section , the cord is formed of central "H" shaped grey matter of ganglion cells and nerve fibers and peripheral white matter of nerve fibers and myelin sheaths , the central grey matter is composed of anterior motor horns and posterior sensory horns on each side with the central canal in the middle.

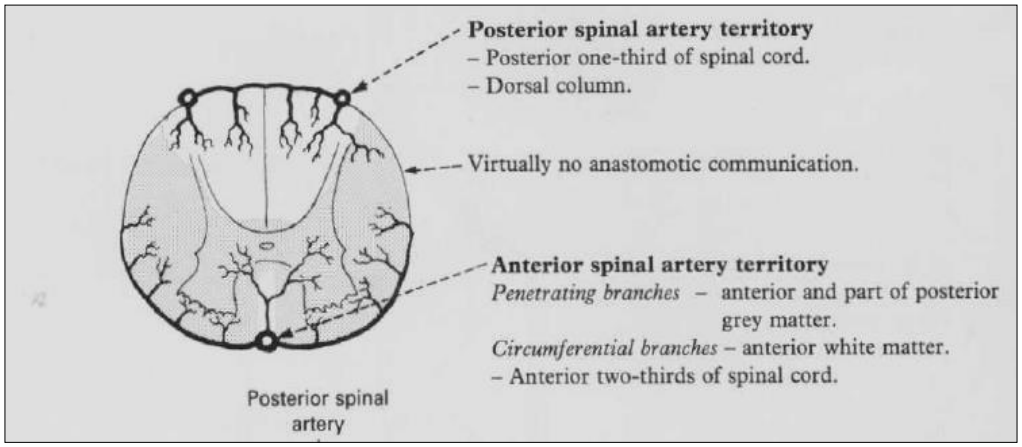


Fig(69)Diagram to show the spinal cord, the important tracts and their relationship to the left leg.

Blood supply: Fig(70)

- It is mainly from **single anterior spinal artery (ASA**) branch from the vertebral artery
- **Two posterior spinal arteries (PSA)** arising from the vertebral or the posterior inferior cerebellar arteries.
- Segmental arteries .the most important of which is the **artery of Adamkiewicz** which enters the cord between T5&T8

* Within the cord, the A.S.A supplies all the cord except the posterior columns and the posterior horns which are supplied by P.S.A.



Fig(70): Blood supply of the spinal cord

N.B:

- ***Cauda equina*** :is formed from the lumbosacral roots which occupies the rest of the spinal canal below L1-L2(lower motor neuron)
- ***Conus medullaris***: is formed from the lower most three segments of the spinal cord S3,4, 5.
- ***Epiconus***: is formed from L4, 5 & S1, 2 cord segments.

Paraplegia

■**Definition:** weakness or paralysis of both LL due to bilateral corticospinal tract lesions

■**Causes :**

- Cerebral → rare causes
- Brain stem ---rare causes
- Spinal →the commonest

(1*) Cerebral causes:*

Paraplegia to occur here, the lesion must involve leg areas of both cerebral hemispheres

Cerebral causes of paraplegia

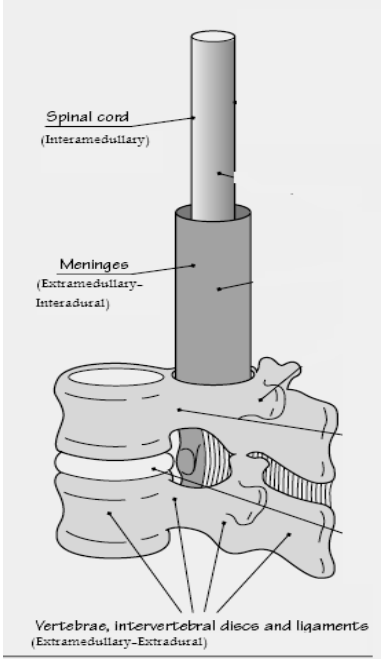
"both Para central lobules" as in:

- Depressed fracture over the vault of the skull (subdural heamatoma)
- Tumors.
- Superior sagittal sinus thrombosis.
- Encephalitis.
- Cerebral palsy.

Paraplegia here is associated with higher cerebral functions disorders

(2) *Brain stem causes*:

Must involve the midline structures to affect both corticospinal tracts, here with paraplegia, cranial nerves affection are present------ e.g. with brain stem tumors, vascular lesions, syringobulbia.



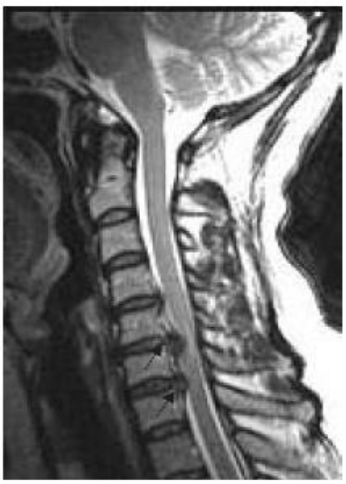
(3) *Spinal causes*: Fig(71)

- Focal →the commonest
  Fig(71)
- Systemic → rare causes
- disseminated→ rare causes

■**Focal spinal paraplegia :**

- *Extra medullary causes* → Extradural(vertebral)
  → Intradural (meningeal)
- *Intra medullary*

1- Vertebral causes(extradural) :-



Low cervical cord compression due to prolapsed discs (arrows).

1- Atlanto axial dislocation

2- Cervical myelopathy

3- Central disc prolapse →direct compression Fig(72) →

4- Cervical spondylosis →spondylotic myelopathy (through direct compression & interference with blood supply),

5- Fracture, fracture dislocation

6 - Congenital marked deformity of the spines e.g. kyphoscoliosis

7- Tuberclous spinal osteitis:

- Common in young adults
- Common in dorsal cord
- Paraplegia may be acute or chronic
- Acute paraplegia results from sudden collapse of the diseased vertebrae with angular deformity.
- Chronic paraplegia results from Pott"s disease "cold abscess" which causes direct cord compression., cord ischemia,TB endarteritis
- Management is mainely conservative with antiTB drugs even in sever cases

8-Syphilitic spinal osteitis:

It is now a very rare cause of spinal paraplegia, in Paget's osteitis deformans, softening & collapse of vertebrae occur without abscess formation

9- Neoplasm of the vertebral column which may be:

- Primary as :- Sarcoma -Cavernous haemangioma -Osteoma - Myeloma
- Secondary →the commonest, and rare before age of 35 years, usually the primary in the lung , breast , thyroid or prostate .

10- Spinal extra dural abscess arising either through blood born infections or from vertebral osteomyelitis .

2- Meningeal (intradural) causes:

1- Inflammatory causes as.

- $, TB, post traumatic, sarcoidosis, meningitis, cystic arachnoiditis

2- Neoplasm: as

- Meningioma → common in dorsal region, affect females more, arise from arachnoids covering the root.
- Neurofibroma → arise from spinal root.
- Lipoma, chondroma, dermoid cyst.

3- Arachinoidal cyst → usually of developmental origin.

4- Parasitic cyst e.g. hydatid cyst or cysticercosis cyst.

3- Intramedullary causes:

1- Inflammatory & infective causes e.g.

- Transverse myelitis (due to viral, fungal, bacterial, parasitic,$,T.B,…) -
- Myeloradiculitis - Post infectious, post vaccine.
- Myelomeningitis - M.S - Schistosomiasis -Spinal-cord abscess

2- Neoplasm of the cord e.g.

- Ependymomas - Oligodendrogliomas
- Medulloblastomas - Intramedullary metastasis
- Angiomatous malformations
- Leukaemic deposits

3- Degenerative e.g. - Syringomyelia - Neural tube defects

4- Vascular e.g. - Anterior spinal artery occlusion. - Dissecting aortic aneurism

5- Radiation myelopathy (radiotherapy)

6- Electrical current injury of the cord

7- Toxic myelopathy e.g. nitrous oxide, cycads "cassava roots"(plants consumed as a food in some areas in Africa)

8- Hemorrhage into the spinal cord "hematomyelia"→may be 2 ry to trauma or 1ry from bleeding diseases , vascular malformations or anticoagulants.

9- Decompression sickness “Caisson disease"→manifests in underwater divers; with decompression, nitrogen bubbles can form in spinal vessels, especially in the upper thoracic cord. Symptoms may be mild and transient or may be devastating with a complete transverse myelopathy. The patients should be immediately treated with recompression in a hyperbaric chamber.

4- Other causes of spinal paraplegia (systemic & disseminated causes)e.g.

- Hereditary spastic paraplegia.
- Cerebral palsy
- Heridofamilial ataxias
- Subacute combined degeneration
- Motor neuron diseases
- Demyelinating diseases
- Collagen vascular diseases
- AIDS vascular myelopathy
- Tropical spastic Paraparesis with HTL V-1

N.B :

♥ Spinal tumors are most commonly situated extramedullary 50-65%, Intra medullary 14-26%, followed by extradural sites 10- 18%

♥ The thoracic cord is the commonest site of extra dural and extramedullary tumors

♥ Intra medullary tumors are common in childhood.

N.B: Regarding onset of paraplegia in relation to aetiology:

♠ Acute onset paraplegia are mostly common with

- Inflammatory causes
- Traumatic causes
- Vascular causes
- Acute demyelinating exacerbation
- Para neoplastic myelitis
- Acute toxic causes

♠ Chronic paraplegia are encountered with :

- Most of vertebral causes
- Hereditary causes
- Neoplasm
- Motor neuron disease

Clinical presentation of paraplegia :Figs (73)

- The clinical picture of well established paraplegia is the same in all causes of paraplegia either extramedullary or intramedullary but with some features characteristic for each . A paraplegic patient will be presented with the following characteristic triad:

1- Weakness or paralysis of both LL of UMNL features

2- Sensory level below which sensations are lost or impaired

3- Sphincteric disturbances

1- Mode of onset

a) *In extra medullary causes* , the onset is painful, gradual onset with long course but may be rapid in tumors ,or acute in trauma with the following added features at the level of the lesion :
   - Localized back pain , tenderness ± swelling , ±deformity in cases with vertebral causes
   - With the back pain , there is root pain radiated to the same area supplied by the affected root with all characters of root pain
   - Impaired sensation in the dermatome supplied by the affected root
   - LMN features of the muscles supplied by the affected root (weakness , wasting , hypotonia , lost reflexes)

b) *In intra medullary causes* of paraplegia , the onset is usually not painful, more rapid , with short course

c) *In acute causes* where the onset is acute , the patient will pass through the shock stage "2-6 weeks" , during which there is sudden paralysis of both LL , with complete loss of tone , and reflexes and retention of urine. The shock stage is due to sudden interruption of the descending motor tract.

2- Motor symptoms and signs in the LL:

Weakness or paralysis of both LL will be presented as follow:

* **Weakness will be in both LL** , nearly symmetrical in cases of intra medullary paraplegia and nearly asymmetrical in cases of extra medullary causes

***Weakness will be**: -Distal more than proximal.
- Adductors more than abductors.
- Affect group of muscles.
- Flexors more than extensors.

***Deep reflexes :**

- Exaggerated deep tendon reflexes ,knee& ankle (After the shock stage)
- Clonus (patellar& ankle) is usually present with hyperreflexia.

***Hypertonia**: both LL will be spastic, more in extensors and the LL are held in the extended position (after the shock stage).

***Superficial reflexes**:

- Lost or diminished abdominal reflexes. (according to the level of the lesion)
- lost or diminished cremasteric reflexes.

- Bilateral + ve babiniski sign

***Gait**: if the patient can still walk. His gait is spastic and scissor like gait.

N.B:

♦ With progression of the lesion, the extra pyramidal fibers are involved (complete cord affection), and the flexors will be more hypertonic, and the lower limbs in this case will be held in the flexion position (paraplegia in flexion)which is considered the last stage of paraplegia ,here the mass reflex can be elicited →by scratching the skin over the medial aspect of the thigh →spontaneous urination , defecation , even erection and ejaculation.

♦ Also in progression of paraplegia to the last stage, pierre Marie Foix test can be elicited through firm passive planter flexion of the toes → result in spontaneous flexion of the hip, knee, dorsiflexion of the ankle (withdrawal reflex)

3- Sensory manifestation in the LL:

***The patient will complain** of impaired sensations in both LL with certain level according to the level of the diseased segment where the ascending sensory tracts (spinothalamic and posterior columns)will be involved.

***The sensory level** may be defined by the patient as a sense of belt , band or tightness , encircle his trunk , below which there is impaired sensations

***In cases of extra medullary lesions**, all types of sensation are impaired with early loss of sensation in the saddle area.

***In cases of intra medullary lesions** , there will be jacket sensory loss of dissociative nature (loss of pain , temperature , with preservation of touch) in both lower limbs below the sensory level with sparing of the saddle area (or late affection)

***Lhermitt's sign** : in cases of cervical cord lesions either extra- medullary or intra- medullary , the patient will experience pain , numbness , electric shock like pain in the back and limbs on flexing or extending his neck.

N.B: In slow spinal compression, the pyramidal tract (corticospinal tract) is affected first, then the posterior column, and the spinothalamic tract last.

To detect the sensory level follow the following: V (vertebrae) + X(constant) = S (segment).

X .. - For last cervical vertebrae → add 1

- For thoracic vertebrae 1-6 → add 2
- For thoracic vertebrae 7-9 → add 3
- Thoracic 10 vertebrae overlies →L1-2 segments
- Thoracic 11 vertebrae over lies →L3-4 segments
- Thoracic 12 vertebrae overlies →L5 segment
- L1 vertebra overlies →sacral & coccygeal segments.

4- Sphincteric disturbances:

***In acute lesions** → early affected, there is retention of urine in the shock stage.

***In gradual lesions** → sphincters are not usually early affected, especially in extra medullary lesions, but there is urgency, precipitancy, or hesitancy of micturation and later retention may occur or automatic bladder.

***Constipation** is common, but fecal incontinence may occur in severe paraplegia.

***Sphincters** are affected early and severely in cauda equina and conus medullaris lesions than higher cord level lesions.

5- Other manifestations:

- Autonomic symptoms in the form of excessive sweating ,cyanosis , edema & coldness' of LL due to interruption of ascending sympathetic flow.
- Papilloedema can occur as a complication of spinal tumors as result of ↑ CSF protein.

Differential diagnosis of a paraplegic case (spinal paraplegia):

1- Cerebral causes → with higher cerebral functions disorders
2- Brain stem causes → with cranial nerves affection
3- Gillian Barrie syndrome → LMN lesion ,no sensory level , no sphincteric disturbances, +ve NCS findings,…
4- Motor neuron diseases → no sensory changes , no sphincteric disturbances,….etc
5- Hereditary spastic paraplegia → no sensory changes , no sphincteric disturbances,
6- Poliomyelitis → pure motor, young age, no sphincteric disturbances, +ve EMG findings,..
7- Peroneal muscle atrophy → marked distal LL wasting , distal sensory loss , no sphincteric disturbances, NCS & EMG findings,….
8- Hysterical causes.

How to investigate a case of paraplegia:

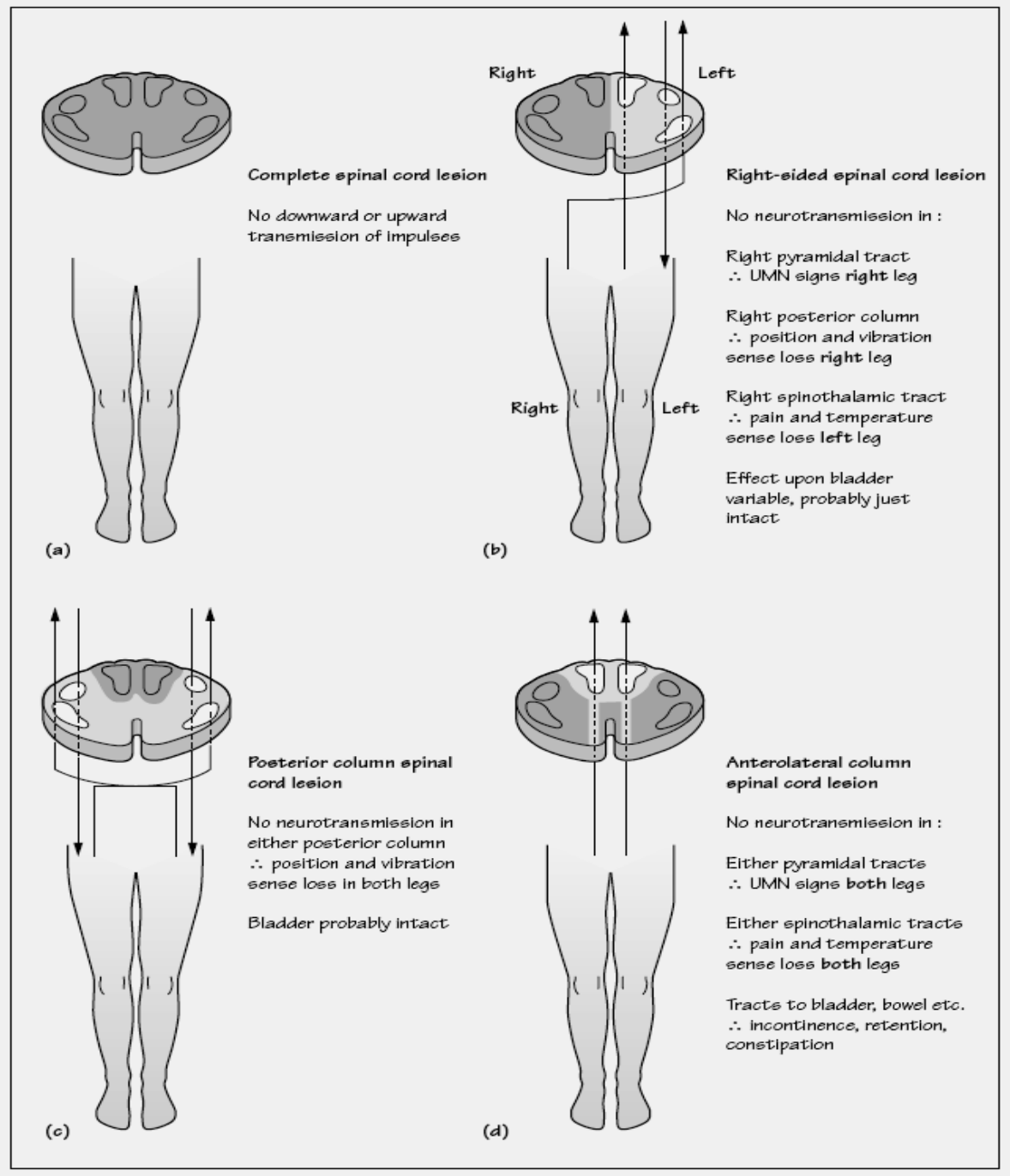
1) *Plain radiography* :-

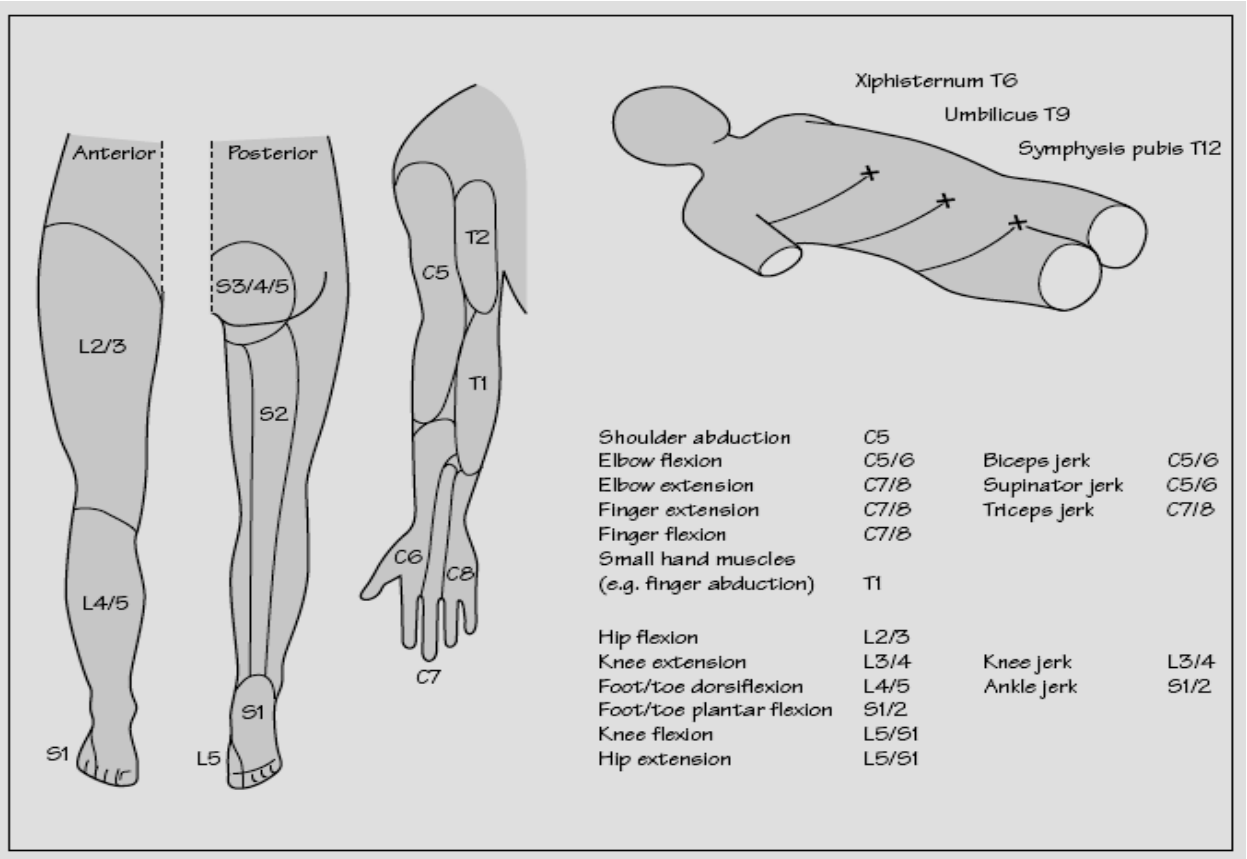
Is obligatory in all cases of paraplegia, in vertebral lesions it is highly informative, you have to look for:-

- Vertebral destruction in cases of T.B, $ osteitis, tumors
- Vertebral fractures, dislocation
- Degenerative changes in the spine e.g.
  - Spondylosis
  - spondylolisthesis
- Narrow disc space in chronic disc prolapse

Fig(73)

Various spinal cord lesions and their tract signs :





Fig(74)The important dermatomes, myotomes and reflex arc segmental values, with which a student should be conversant.

2) *C.T scanning of the spine* : -(plain& with contrast myelograghy)which can demonstrate

- Metastasis
- Soft tissues swellings
- Prolapsed discs
- Cord tumors
- Fractures
- Spinal cord stenosis

3) *MRI of the spine* :-

Is superior to other investigations in identifying soft tissue masses, tumors, hge, intrinsic & cystic lesions

4) *C.S.F study* :-

May be of diagnostic value in some inflammatory & demyelinating causes where CSF proteins are increased with increased mononuclear cells in cases of extra medullary cord compression , there is also xanthochromia (yellowish discoloration of the CSF) and the CSF may coagulate spontaneously

N.B:-

- In cases of extramedullary abscess, lumbar puncture should not be done at or near the site of spinal pain or tenderness.
- With strong clinical suspicion of spinal compression or papilloedema →C.T or MRI spine should be done & better to avoid lumbar puncture

5) Electrophysiological studies :-

Recording spinal & cortical sensory evoked potentials may be of value in distinguishing organic from hysterical cases. EMG & NCS may help in diagnosis of the cause of muscle wasting and radiculopathy

6) *Spinal cord angiography :-* Is of value in diagnosis spinal vascular malformations

▲Transverse myelitis:

■Definition:

Acute inflammatory lesions (infective or non infective) of spinal cord segments, common in adults & women .

■Aetiology:

Different pathologies may cause myelitis include:-

1-Myelitis due to viruses .

- Polio virus - AIDS - Herpes zoster, herpes simplex

2-Myelitis due to bacterial, fungal, and parasitic diseases.

- Syphilitic myelitis
- Lyme disease
- Tuberculous myelitis
- Pyogenic (suppurative) myelitis
- Parasitic or fungal infection of the meninges

3-Myelitis due to non infectious inflammatory causes .

- Post infectious - Post vaccinal
- Acute relapsing multiple sclerosis
- Radiation myelitis (myelopathy)
- Acute or subacute necrotizing myelitis
- Vasculitis- collagen vascular disorders
- Device's disease (neuromyelitis optica)
- Paraneoplastic myelitis

■Clinical picture:

- Preceding paraplegia in >1/3 of patients may report upper respiratory tract infections, GIT infections, localized LBP, days before onset of paraplegia.
- Acute onset of flaccid paralysis , complete loss of tone , power , reflexes,and sensation with retention of urine (shock stage)
- Spastic paraplegia will evolve 2-6 weeks later with its characteristic clinical picture

■Investigations:

- Patients must undergo radiological investigations at first to exclude acute, subacute , cord compression injury ,(C.T or MRI) which may show mild cord swelling.
- Lumbar puncture is then important to be done for CSF examination which will reveal:
  - ↑ mononuclear cells
  - ↑ proteins
- In about 40 % of cases → no specific aetiology can be detected

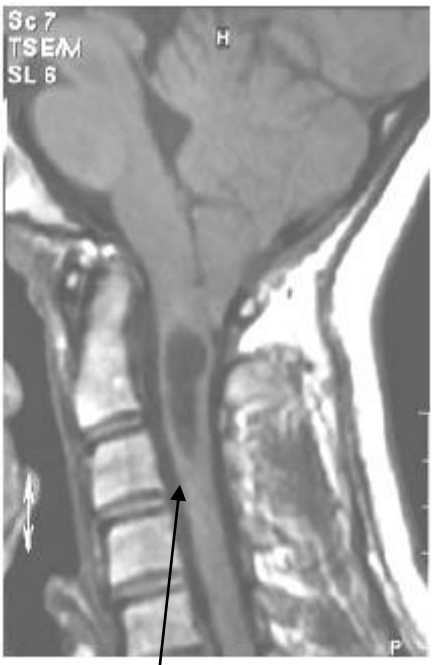
■**Differential diagnosis:**

- Traumatic causes
- Anterior spinal artery occlusion
- Haematomyelia

■**Management:**

1- Care of paraplegic patient in general (will be discussed later)

2- In non infectious inflammatory myelitis high dose of intra venous methyl predinsolone(1gm/day for 3 days) may be highly effective. ± plasma exchange and intravenous immunoglobulin,



Low signal cavity within the upper cervical cord, with mild Arnold–Chiari malformation.

▲**Syringomyelia:** Fig (75) →

****Definition:**

Chronic disease characterized by the presence of long cavities, surrounded by gliosis, situated in the central part of the spinal cord, often extending to the medulla (syringobulbia).

****Aetiology:**

1) non communicating →less common causes →spinal trauma, tumors, arachinoditis

2) communicating →common variety: there are different causes which can contribute to development of diseases e.g.

‣Chiari type 1 anomaly (congenital extension of the cerebellar tonsils below the foreman magnum)

‣ Craniovertebral developmental anomalies ,eg

Dandy Walker syndrome →closure of the foremen of the magendi , preventing intermittently the cross of CSF from the 4th ventricle into the subarachinoid space , pressure of CSF will be forced down into the central canal of the cord →become dilated

‣ Basal arachinoditis (post traumatic , post meningitic ,S.A.H)

‣Spinal cord injury & tumors

**Clinical picture:

- the disease is common in males than females , usually appears between the age of 25 – 40 years
- The disease process is most frequently situated in lower cervical, and upper dorsal segments, may extend up to medulla and pons, even as high as the internal capsule.
- The onset is usually gradual with :
  - Wasting, weakness of LMN features of small muscles of the hands (ulnar side) →are early symptoms-
  - Cutanous analgesia, lost feeling, trophic changes, in the UL (hands) →this is due to destructions of the AHC and the decussating sensory fibers
- Compression of the corticospinal tract in the spinal cord leads to weakness of both LL of UMN features (paraplegia)
- Compression of the lateral spinothalamic tract on one or both sides leads to diminished or lost superficial sensations in lower limbs with jacket sensory loss with sometimes an area of normal sensation over the abdomen.
- The posterior columns are usually the last to be affected , therefore early in the disease , the lost or diminished sensations are of dissociative nature,
- Different skeletal deformities may be associated
- Trophic symptoms & signs e.g. skin →ulcers , cyanosis , hyperkeratosis,Bone → atrophy, decalcification, charcot's joint, bone necrosis

Morivan's syndrome →progressive loss of pain, ulceration, loss of soft tissues resorpition of phalanges.

**Investigation: -MRI is the most diagnostic investigation
- CT Myelography

**Treatment: -physiotherapy ….for the weakness & spasticity
- Analgesic & muscle relaxants
- Surgical decompression of the cavities by laminectomy may be needed

**Differential Diagnosis:

- Other causes of spinal paraplegia (intramedullary & extramedullary)
- Cervical spondylosis -MND -Cervical.rib -Peroneal muscle atrophy

Neurogenic bladder

 # Afferent supply of the bladder:-

-From the urethra & external urethral sphincter→ to the *pudendal nerve →sacral cord segments* S 2, 3, 4

- From the bladder ▸ Sense of filling & desire to micturate

▸Touch & pressure sense

▸Chemical sense

All pass through the pelvic nerve → dorsal root segments S2,3,4→ spinal cord (sacral segments) → to the *spinothalamic tract* regarding sense of filling & → to the *posterior column* regarding touch , pressure , chemical sensations.

Then from the *sacral cord segments →thoracic lumbar segments →supra spinal centers* which control micturation

- Bladder cortical centers - Brain stem centers (ponto mesencephalic)
- Basal ganglia - Cerebellum

 # Efferent supply of the bladder:-

1. Parasympathetic " stimulatory":-

From the sacral segments S2, 3, 4 (through the pudendal nerve& pelvic nerve) to supply

- Longitudinal fibers of detrusor muscle →contraction →open the sphincter
- Circular muscle of the bladder wall →contraction →evacuation

2. Sympathetic "inhibitory":-

From mainly L1-2 with contribution from T 10, 11, 12 & L3, 4→the fibers unite to form the superior hypogasteric plexus →presacral nerve in front of aortic bifurcation to form two hypogastric nerves →to the vesical plexus in the lateral aspect of the bladder wall.

▸ There are two sympathetic fibers:-

- Alpha sympathetic fibers →in the bladder neck & internal sphincter → on contraction.→.clousre of the sphincter
- Beta sympathetic fibers → in the bladder wall → relaxation → filling of the bladder

Nervous mechanism which control bladder function:-

There are three different nervous mechanisms:-

1) Sacral reflex arc →which cause reflex evacuation of the bladder on filling through sacral segments (through its afferent & efferent) → present in infancy
2) Sympathetic inhibition over this reflex
3) Voluntary control over this inhibition &voluntary initiation of micturation through supraspinal centers
   This volantry control is supported by the action of pelvic floor muscles & abdominal muscles.

Neurogenic bladder disorders:

1-lesions of the sacral reflex arc (LMN lesions):-

a) Afferent lesions "sensory "

as in D.M. ,T.dorsalis , perncious anemia, dorsal root ganglioectomy ... resulting in -sensory atonic bladder

clinically – Absence of sense of fullness & desire to micturate

- Gradual over distension of the bladder which may reach huge size
- Lack of reflex events of micturation
-Patients will tend to leak urine at very high bladder volume, and usually those patients become associated with urethral dilatation & vesico urethral reflux.

b) Efferent lesions "motor"

Isolated efferent lesions are uncommon as in:-

-Poliomyelitis (isolated AHC. Lesions)

- Isolated sympathetic lesions in pelvic surgery

This lead to motor atonic bladder, preserved sensations,hypertonic bladder,painful retention, inability to evacuate→ it must be catheterized.

c) Combined afferent & efferent lesions:-

Most common as in conus medullaris lesions, cauda equina lesions, here there is autonomic bladder: incomplete, irregular, involuntary, evacuation.

2-UMN lesions of the spinal cord or supra spinal centers

Complete cord lesions -retention

Incomplete lesions of the descending sympathetic concerned with inhibition -difficulty in holding urine -precipitancy ,- urgency e.g. in D.S

-Lesions of the descending fibers concerned with voluntary initiation of micturation -retention

-Lesions of the posterior column which carry touch & pressure sensations -hesitancy.

-Cerebral lesions : as in C.V.A, tumors, dementia, fibers concerned with voluntary initiation of micturation can be involved leading to retention, incontinence or precipitancy.

NB:- Stress incontinence as in frequent pregnancies, post labor, & as result of weak pelvic floor muscles (which help in voluntary inhibition ofmicturation) the patient can pass urine on situations

with increased intra pelvic pressure & intra abdominal pressure as during cough, straining, sneezing, laughing.

- Nocturnal enuresis: as in children with psychological factors will acquire abnormal conditioned reflex with continuous bladder evacuation during sleep.

Management of neurogenic bladder disorders:

1) Retention "sympathetic over activity" :-

Treated by-

- Sympatholytics or parasympathomimetics e.g. -distigmine 0.5 mg injection or 5ml oral
-Phenoxybenzamine 10 - 20 mg /day
-Bethancol 10 -100 mg /day

In chronic cases -division of internal sphincter (bladder neck resection) or by resection of the presacral nerves

2)Urgency, precipitancy, frequency, incontinence "parasympathetic over activity" - Parasympatholytics are used eg :-

- Anticholinergic drugs - Propantheline 15-30 mg/day -Clomipramine (tofranil)

Spinal degenerative diseases

NB.Some anatomical consideration :

@ Each spinal nerve is composed of dorsal root "*sensory* "and ventral root "*motor*" the two roots unit → to form the spinal nerves which travel through the intervertebral foramen.

@ The intervertebral disc consists of central semifluid portion "*nucleus pulposus* which is surounded by strong fibro cartilaginous band "*annulur fibrosus* ".On degenerative spinal diseases with age, trauma, the annular fibrosis will become weak and the central nuclear pulposis will herniate through the weak point and compress either the central spinal canal causing spinal canal stenosis ,or the cord, roots, or both.

@ with advanced age ,frequent trauma,some occupations(porters), sporters ,the frequent spinal movements will degenerate the vertebrae with new bone formations called osteophytes "spondylosis" these osteophytes may cause narrowing of the intervertebral foramen causing radiculopathy or compress the cord causing spondylotic myelopathy.

@ Cervical & lumber spines are the commonest sites of spinal degenerative diseases due to their higher mobility.

Clinical picture of radiculopathy in general:

In radiculopathy due to disc lesions, spondylosis or due to other spinal diseases, there are three main clinical corners to be discussed .See Fig (29)

1) Sensory symptoms :

Lesions of the dorsal root will cause radicular or root pain which has the following characters:

-Lancinating , electric , burning.
-Abrupt, sharp , well localized. ↑ at night

-Referred to a spicific dermatome .

-Precipitated or ↑ by ; coughing, sneezing,straining.spine movement

2) Motor symptoms & signs:

Due to ventral root lesions , there is weakness & wasting of the muscles supplied by the affected root, fasciculations may be present in the affected muscle

3) Reflex signs:

Lesions of the dorsal or ventral roots may interrupt the afferent or efferents, causing diminshed or lost reflexes of the muscles supplied by the same root.

Cervical radiculopathy & myelopathy:

- In cervical spondylosis

Usually the symptoms are subacute or insidous with radicular pain in the neck, radiated to the dermatome supplied by the affected segment ,with the different characters of radicular pain as mentioned above.

-There may be localized area of tenderness in the corresponding paraspinal muscle.

-In case of myelopathy

There may be inverted biceps or triceps reflexes according to the level of the affected segment with features of bilateral pyramidal tract lesions in both lower limbs which are usually asymmetrical and with certain course of spread of lesion which are usually affect the lower limb on the same side of the affected previous upper limb, then the other lower limb.

-In radiculopathy ,the symptoms may be mainly sensory on lesions of the dorsal root or mainly motor on lesions of ventral root or both.

-N.B The inverted reflexes mean that ,at the level of the lesions ,the reflexes mediated by the affected segment will be lost or diminshed , and due to the cord lesions "myelopathy" , the segments below the level of the lesion will be hyperexcitable with UMN features. and where the stimulus carried by the afferent spread little to the segments below the desired one ,therefore the reflexes mediated by the segments below will appear e.g- at the lesions of C5,the biceps reflex will be lost but with exaggerated triceps or finger flexor jerk.

▲Compression by intervertebral disc prolapse

This is most common in the cervical region and, if centrally located, can result in acute or subacute cord compression. Much more commonly, cervical disc prolapses occur laterally and result in upper limb pain and signs of a cervical radiculopathy. Thoracic disc protrusions, although less common, are a well recognized cause of subacute or chronic cord syndromes, with paraparesis or a Brown-Séquard syndrome when the compression is asymmetrical. A clear-cut sensory level is usual. Sometimes, the neurological symptoms fluctuate over time. MRI readily demonstrates cord compression due to disc prolapse. Occasionally, marked cord compression may be found radiologically in the absence of clinical deficits. Acute central disc protrusion of a cervical intervertebral disc should be treated immediately by immobilizing the neck in a plastic collar; if cord compression is severe and is resulting in a significant clinical deficit, surgical decompression may be needed

EPILEPSY

Important Definitions:

→Epileptic seizures: Can be defined as an intermittent ,transient,recurrent,stereotyped paroxysmal attacks of motor,sensory,behavior or emotions disturbances usually but not always accompanied by loss of consciousness resulting from uncontrolled paroxysms of neuronal cortical discharge N.B .7-10% of the population may have a seizure during their life.

→ The prodroma: Refers to mood or behavioral changes that the patient experienced preceding the seizures by some hours .if well known ,the patient can protect himself.

→The aura: Refers to the symptoms that occur immediately before loss of consciousness ,which results from the beginning of the abnormal neuronal discharge and will localize the focus ,the patient usually cannot take care (no time to protect himself).

→ The ictus: Refers to the seizure itself.

→The postictal: Refers to the period after the attack,which results from the neuronal exhaustion,during which the patient may be confused,disoriented,comatosed,suffering from headache,automatic behavior,feel sleepy or postictal focal neurological deficits.

Classification of epilepsy:

A) Partial seizures:

-Simple partial :(no disturbance of consciousness)

*with motor symptoms.

*with sensory symptoms:somatosensory or special sensory.

*with autonomic symptoms.

*with psychic symptoms.

-Complex partial (with impairement of consciousness)

-Partial seizures secondarily generalized.

B) Generalized seizures:

-Tonic-clonic. seizures (grand mal).

-Absense seizures(petit mal).

-Myoclonic seizures.

-Clonic seizures. -Tonic seizures. -Atonic seizures.

Aetiology of seisures (Symptomatic & Idiopathic)

A) Symptomatic causes of epilepsy (acute and remote causes):

***Acute symptomatic seizures:**

1) Fluid and electrolyte disturbances:eg

-Hypernatremia.(> 150mmol/L)as in fever,sweating,diabetes.

- Hyponatremia(< 125mmol/L)as in liver diseases,nephrotic syndrome,

-Hypocalcemia(<7mg):as in vit.D.deficiency,acute pancreatitis

-Hypercalcemia(>12mg):as in hyper parathyroidism,malignancy,bone metastasis.

2) Metabolic disorders:

-Non ketotic hyperosmolar diabetic coma:Seizures are common(in 25% of patients)

-Ketoacedotic coma:rare

-Hypoglycemic coma:in up to 7% of patients

- Hypothyroidism:common up to 25% of patients

- Hyperthyroidism:seizures are rare

-Porphyria:in 15% of patients

-Acute hepatic failure:seizures may be focal,myoclonic or generalized

-Renal failure:in acute uraemic encephalopathy,dialysis encephalopathy

-Hyperpyrexia - Alkalosis -Pyridoxine deficiency

3) Drug induced seizures:

***Antibiotics:** -penicillin. -ampicillin. -I.N.H.

***Antidepressants:** -Amitryptaline. - Imipramine.

***Antipsychotics:** -Chlorpromazines. - Lithium. -Phenothiazines

***Anaesthetics**: - Ether. -Halothane.

***Antidysrrhythmic**: Lignocaine.

***Radiographic contrast media**.

***Antiepileptics**:In overdose.

***Others:** - D.penicillamine. -Folate. -Alcohol. -Cocaine - Amphetamine.
-Hypoglycemic drugs. --Immunosupressants. -Withdrowal seizures(addict

N.B : patients with hepatic or renal failure,those with previous history of epilepsy,or those with preexisting brain diseases are usually at risk of developing seizures with these drugs.

4) Cerebrovascular accident (C.V.A.): Common to occur early with subarachnoid haemorrhage,intracerebral haemorrhage,less common to occur early with cerebral infarction but common to occur late.

5) Head injury:Seizures may occur early with the trauma or late up to years after trauma.

6) C.N.S.infections:As encephalitis where seizures usually occur with the onset with fever,cloudness of consciousness,and focal neurological deficit.

*Remote causes of symptomatic epilepsy:

1) Head inujry:Common with those with depressed fractures,long posttraumatic amnesia, ,intracerebral haemorrhage,early seizures, lost consciousness.

2) post craniotomy:In patients undergoing brain surgery.

3) Brain tumours: Mainly in tumours of frontal, parietal or occipital regions ,seizures are usually focal ,associated with focal neurological signs and with focal EEG changes .

4) C.V.A: Late seizures also occur with stroke mainly with infarction.

5) Hypoxic Ischemic encephalopathy: -CO,-CO2,- asphyxia. -profound anermia

6) C.N.S.infection and infestation: -Encephalitis. -Meningitis.
-parasitic infestation. -slow virus infection.

7) Demyelinating diseases

8) Neurodegenerative,congenital and inborn errors of metabolism.

B) Idiopathic Epilepsy:

-When no local or generalized causes can be detected.

- ?Most cases here are heredofamilial.

*_Factors that precipitate seizures in epileptics or in those with liability to develope seizures:_

1) Reflexly induced:

1 -Visually induced:as in T.V.,video games,computers.

2 -Primary reading. 3 -Musicogenic:auditory evoked.

4 -Writing. 5-Eating.

2)Non specific precipitating factors:

1-Sleep and sleep deprivation. 2-Psycological stress.

3-Before menstruation 4 -Oestrogen medication

5-Hypocarbia,H2O retension 6-Hypoglycemia.

7-Pyridoxine deficiency. 8-Psychotropic drugs.

9 -Electrolyte imbalance.

*_Factors predicting poor outcome of epilepsy (intractability):_

1) Early age of onset of epilepsy.

2) Organic neurological diseases with mental or focal neurological deficits.

3) History of early status epilepticus.

4) History of polyfits (polytype),recurrent and frequent fits.

5) Persistent interictal EEG changes.

6) Certain seizure types:eg, Temporal lobe epilepsy,Lennox Gastaut Syndrome.

***Clinical features of epilepsy:**

#1- *Simple partial seizures:*

a) Simple motor:

In the form of tonic or clonic movement of any part of the body usually involve the face ,hand,arm,or leg.The focus is usually in the frontal motor cortex.It is termed Jacksonian if the convulsions take a march course of spread from one muscle group to the next.usually after simple motor seizures,the convulsed part may remain weak or paralyzed for sometimes (Todd 's paralysis).When the patient eye and heads turn away from the site of the focal discharge,it is termed adversive seizures.

b) Simple somatosensory:

The focus lies in the sensory cortex,seizures in the form of transient attacks of numbness,tingling,or parasthesia affect the face or hand and may spread as do Jacksonian seizures .Disturbing or painfull sensations are rare to occur.postictal numbness as in Todd 's paralysis occurs.Motor symptoms occur due to the close association between sensory and motor cortex.

N.B :- Motor and sensory seizures usually indicate organic brain disease ,the focal onset localizing the site of the lesion ,full investigation are needed.

-Fits of visual symptoms including spots,flashes of light may occur with occipital seizures.

-Fits of buzzing,hissing,whistling and ringing noises can occur with seizures originating in the lateral temporal parts .

-Fits of visceral or autonomic symptoms as vague epigastric sensation , nausea vomiting,bleching,pallor,flushing,sweating,palpitation,intense desire to micturate,all may occur in cases of abdominal epilepsy usually originating from the temporal or frontal lobes(limbic structures

-Fits of olfactory and gustatory symptoms in the form of unpleasant smells,tastes(uncinate fits),in seizures originating from the uncus.

-Complicated psychic symptoms in the form of dreamy like state,feeling of unreality,depersonalization,illusions,hallucinations,intense fear,sense of familiarity(déjà vu),or strangeness(jamais vu),usually in seizures originating from the medial temporal or frontal limbic structures.

#2-*Complex partial seizures:(Psychomotor epilepsy,Temporal lobe epilepsy):*

-Usually arise in the medial temporal lobe or less common in the fronto-orbital regions.

-Must include some impairement of consciousness.

-May or may not be associated with simple partial seizures.

-The psychic manifestations of complex partial seizures include:eye staring,motionless star,arrest of activity followed by stereotyped automatism,e.g:**motor automatism** in the form of lip smacking,chewing,swallowing,walking or running or **verbal automatism** ,usually followed by confusion.if prolonged,will be secondarily generalized.

N.B:

-The sequence of events is usually stereotyped for the same patient.

-Common type of seizure. -One of the seizures intractable to treatment.

-Additional psychological or psychiatric handicap are common.

#3-*Partial seizures progressing to generalized seizures*:

-When the seizures discharge spread from its point of origin and exite other structures in the subcortical regions (**Thalamus,Reticular Formation**),their excitation releases a discharge which spreads back to excite the whole cerebral cortex of both hemispheres,resulting in tonic clonic epilepsy .

-More common in children than adults.

-Susceptible to antiepileptic drugs.

#4-*Generalized seizures*

1) *Petit mal :-*
   - Onset in childhood (4-12years).
   - +ve family history in 40 % of patients.
   - Duration of each attack 5-15sec
   - Frequency -many times/day up to100 times.
   - Clinically -- the child stars, stop what he is doing for seconds , then can complete his act , rare to find myoclonic jerks or drop attack , in those with atypical presentation and where fits persist into adult life , those patient are at high risk to develop grand mal seizures.
   - The fits are usually induced by hyperventilation.
   - The typical fits are easily controlled by antiepileptic.
   - EEG is characteristic with 3cycles/second spike wave complex.

2) *Grand mal* :-
   - Usually starts <20 years.
   - Prodroma may occur before the fits in the form of headache, irritability, depression, there is no focal onsets which distinguish this primary generalized seizures from secondary generalized.
   - Classical epileptic fit starts with **tonic phase**, followed by the **clonic phase**. In the **tonic phase** (15-30sec) epileptic cry occurs resulting from tonic contraction of respiratory muscles with partial closure of vocal cords, jaw muscles contract producing biting of the tongue, the patient falls to the ground, cyanosed & stiff, the **clonic phase** (contraction & relaxation) starts when the neuronal discharge slows , frothing from the mouth , spontaneous micturation occurs, the violent convulsive movements gradually subside leaving the patient in coma or confusion .

3) *Tonic seizures* :-

Sudden sustained muscular contraction with immediate loss of consciousness. Common in children with anoxic attack.

4) *Atonic seizures*:-

Sudden falling to the ground flaccid associated with cloudiness consciousness

5) *Myclonic seizures*:-

Shock like muscle contraction, may be associated with petit mal, tonic – clonic seizures.

6) *Febrile convulsions*:-

- In children below age 5 years with fever.
- +ve family history of febrile convulsion is common
- Duration –brief <15 seconds

- - Generalized or focal tonic-clonic, usually subside with growing, but if repetitive, status, focal or associated with delayed milestones or structural brain damage, later epilepsy will develop.
- **Risk of a second febrile seizure** is 50% with age less than 1 year and 33% if greater than 1 year of age
- **A second febrile seizure will occur** within 6 months in 50% of cases, 1 year in 75% of cases and within 2 years in 90% of cases
- **Rarely** more than 2 episodes of febrile convulsions will occur.
- **Lumbar puncture** should be strongly considered in children less than 1 year of age, considered in children less than 18 months of age and strongly considered in children who received prior antibiotic treatment. Lumbar puncture is recommended in all children with clinical signs of meningeal irritation or intracranial infection .

7) *Infantile spasm (west's syndrome):-*

- Occurs in the 1st few months of life.
- Repetitive shock like flexion of neck and trunk with flexion of knees (salaam attack).

-Associated with progressive mental regression.

-Spasms occur as **clusters** soon after awakening

- **Etiologies** - strokes, tuberous sclerosis, hypoxia, cerebral hemorrhage, brain infections, brain malformations, inborn errors of metabolism
- The exact etiology may not be determined (cryptogenic epilepsy)
- **Poor prognosis** 67% of children develop severe impairment

Differential diagnosis of epileptic fit:

1) **Syncope**
   - Transient impairment of consciousness resulting from decrease cerebral blood flow
   - Occurs mostly in the upright position ,
   - Precipitated by heat , hunger , dehydration , alcohol excess , long standing , unpleasant sounds or sights
   - Associated with pallor, sweating, cold extremities, and palpitation.
   - Gradual onset with no injury or convulsions.
   - With the onset, there is feeling of warmth, dry mouth ,desire for fresh air.
   - Recovery is rapid, with no post icteal sequalae.

2) **Pseudoseizures (hysterical or malingering)**
   - The patient rare to injure himself & bite his tongue.
   - No urination, pupil is unchanged (dilated during epileptic fits).
   - Blood pressure & heart rate do not change (↑ in epileptic seizures).
   - No stereotyped or typical presentation, but vague, of long duration, never occur alone or during sleep, but in front of others to attract the attention.

3) **Other differential diagnosis**
   - Sleep disorders - Migraine. TIA. - Hypoglycemia -Vertigo.- Psychogenic disorders

Management of epilepsy:

1) Diagnosis:

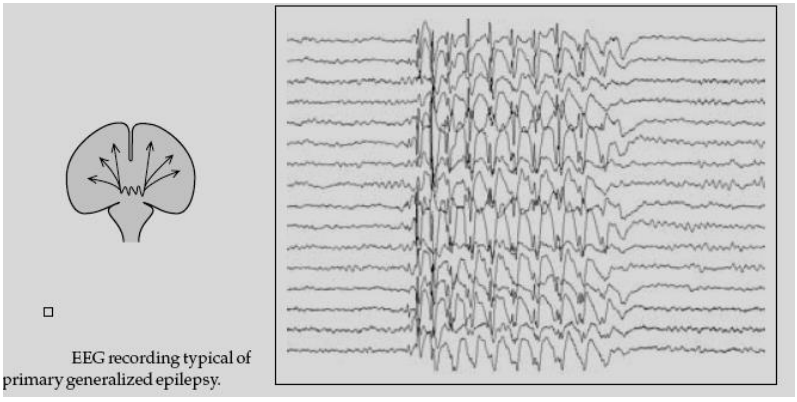
- Characteristic clinical features.
- Investigations:

-EEG (electroencephalogram)

- C.T

- MRI

- Lab investigation.



EEG recording typical of primary generalized epilepsy.

2) General measures :

- Avoidance of precipitating factors
- Assurance of the family, the patient &the surroundings

3)Strategy of medical treatment of epilepsy:

1- starting therapy :-

- one fit is not in need for treatment except if prolonged , status , focal , abnormal EEG , +ve family history , neurological diseases
- two or more unprovoked fits→ start

2- choice of drugs :-

- Mono therapy is better than poly therapy
- Start with one drug with small dose and ↑ the dose gradually over weeks until controlling of fits or appearance of side effects.
- Drugs of choice for :
  - Petit mal → zarontin, followed by Na. Valporate
  - Partial seizures → carbamazepine
  - Grand mal → carbamazepine, Na .valporate, phenytoin.
  - Myoclonic →Na.valporate, colonazepam, piracetam (poly therapy is indicated)
- Dose :

  Tegretol (carmbamzepine):400-1800mg/day divided in2-3 times (20-40 mg/kg)

  Depakine (Na valporate): 600- 2000mg /day divided in2-3 times (20- 50mg/kg)

  Epanutin (phenytoin) : 200-600 mg/day divided in 1-2 times (5-7 mg/kg)

  Rivotril (clonazepam): 5- 4mg/day in 2-3 times (0, 1- 0, 2 mg/kg)

  Phenobarpitone: up to 200 mg/day in 2-3 times (4-5mg/kg/day)

  Zarontin : 1-2 gm /day in 2-3 times (20- 40 mg/kg/ day)
- Duration of treatment :

Gradual drug withdrawal must be considered after 2-5 years fit free with medications, unfortunately, 20% of patients may develop recurrent fits.

N.B:

- Vitamins mainly pyridoxine better to be given with treatment
- You have to treat the patient, not to treat EEG or patient's serum level.
- In pregnancy do not withdraw antiepileptic drugs to avoid occurrence of status epileptics
- Teratogenicity is higher with poly therapy than monotherapy .
- Folic acid during the first trimester can protect from the teratogenic effects of antiepileptic drug

Status epilepticus (convulsive status -tonic- clonic status):

- This is a medical emergency with high mortality rate and morbidity if untreated.
- It is defined as continuous seizures with no recovery of consciousness lasting for longer than 30 minutes.

- Usually associated with hyperpyrexia , hypotension , acidosis , hypoglycemia , cardiac dysrhythemia , shock ,reflex pulmonary edema , aspiration pneumonia , neuronal damage which may be irreversible.

- Causes: usually organic e.g. metabolic, infection, stroke, trauma, tumors,sudden withdrawal of AEDs

- Treatment of status epilepticus :-

- On admission: - care of vital functions; BP, temp, respiration, heart rate, intubations may be required and low flow oxygen inhalation.
- Start an IV infusion of glucose 5% and 0.9 saline 500ml, 100mg thiamine.
- Draw blood sample for – Antiepileptic serum level - Glucose level, electrolytes
  - Renal and hepatic function tests. - Blood gases.
- At the same time a second IV line should ready for :-IV diazepam 10-20 mg over 2minutes
  - Repeat after 5minutes if still fits.
  - Phenytoin (1,000-2,000 mg) IV diluted in saline and infuses at a rate of 50 mg/minute., not dilute in glucose → precipitated.

→ if still fits → phenobarbitol 4000- 2000 mg dilute in saline , infuse at a rate of 100 mg /minute.

→ if still fits →barbiturate anesthesia .

■*Surgical treatment of epilepsy:*

- Common to be used in temporal lobe, intractable seizures, (antiepileptic mostly for ever are used here after surgery)
- Surgical procedures in the form of resection (removal) or division.
- Patients 'age usually should between 12- 30 years, C.T or MRI should detect structural brain lesions.
- IQ must not be less than 70%.

■ *Prognosis of epilepsy:*

-Seizures within the first year of life → carries bad prognosis if symptomatic.

- Frequent partial and tonic - clonic seizures prior to treatment → has bad prognosis.

- Poly fits in the same patient → of bad prognosis.

- Seizures with focal neurological deficits or psychiatric handicap → of bad prognosis

- Complex partial seizures → no good prognosis.

- Patients with idiopathic seizures , febrile convulsions , typical petit mal , age <16 years , infrequent fits , normal EEG or EEG without generalized spike – wave discharge → all carry good prognosis .

- Patients with reversible metabolic disorders → of good prognosis.

■*Alternative non AED treatment of refractory epilepsy:*

-Vagal nerve stimulation. -Ketogenig diet

-IV immunoglobulins. – Hormonal therapy

Cerebro vascular accidents (C.V.A)

(Stroke)

Blood supply of the brain:-

-The brain is about **2%** of the total body weight, receives **15%** of the cardiac output and **25%** of the total inspired air O_2.

-The cerebral blood flow (C.B.F) is about **50-60 ml blood /100gm brain/minute** in the vital **grey matter** and about **40ml blood /100g brain /minute in the white matter.**

- If the cerebral blood flow is below **10ml/100gm/minute infarction will occur** and even if flow is restored, the function does not recur.

-The rich blood supply is carried to the brain through two internal carotid arteries and two vertebral arteries which anastamose at the base of the brain forming the circle of willis.

- **The carotid system (the anterior circulation):-** Fig(76)

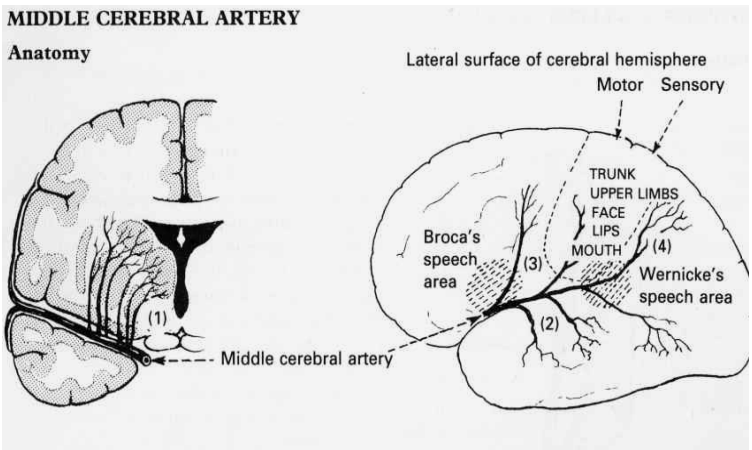
  Each internal carotid artery (I.C.A) starts as a branch of the common carotid artery at the level of the upper border of the thyroid cartilage ,enters the base of the skull through the carotid canal , it then runs through the cavernous sinus where it gives the small branches :-

- Finally, the I.C.A terminates by dividing into its two terminal branches:-

1- The anterior cerebral artery: → pass on the medial surface of the cerebral hemisphere and anastomse with its counter part of the opposite side via the anterior communicating artery.

→This artery supplies:-

-Anterior and medial parts of the cerebral hemisphere.



The middle cerebral artery is the largest branch of the internal carotid artery. It gives off (1) deep branches (perforating vessels – lenticulostriate) which supply the anterior limb of the internal capsule and part of the basal nuclei. It then passes out to the lateral surface of the cerebral hemisphere at the insula of the lateral sulcus. Here it gives off cortical branches (2) temporal, (3) frontal, (4) parietal.

-Anterior basal ganglia and the internal capsule (anterior limb &genu).

- Parts of the optic nerve ,chiasma & hypothalamus.

Fig;(76)

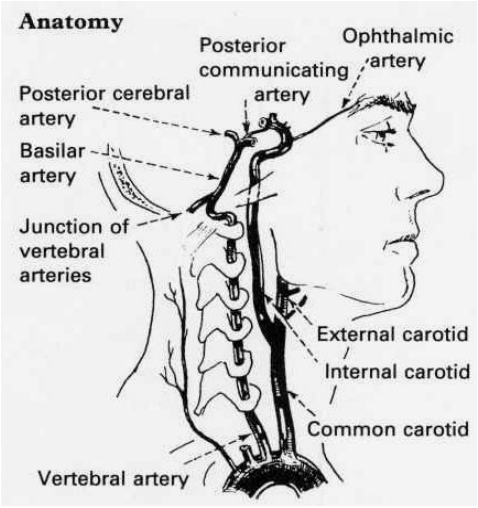
N.B:- ■ The external carotid artery which is the other branch of the common carotid artery, supplies the jaw ,face ,neck , meninges , mainly through the branch of the middle meningeal artery and also , supply the scalp through the superficial temporal , occipital , and the posterior auricular arteries , the external carotid artery does not share in the circle of Willis.

2- The middle cerebral artery:-

Runs on the lateral surface of the cerebral hemisphere to supply:-

■Most of the lateral parts of the cerebral hemisphere.

■Lenticulostriate arteries and arterioles to supply the basal ganglia and the ant.2/3 of the post.limb of the internal capsule .



Fig(77)

☺**The vertebro basilar system (posterior circulation):-fig (77.78)**

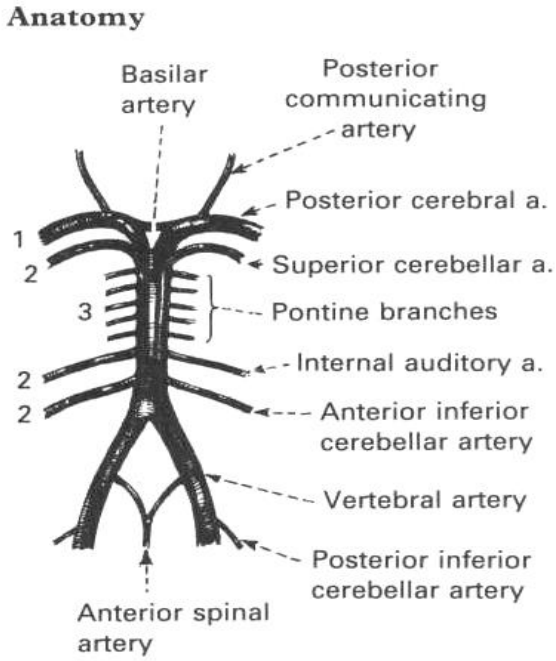
-Formed of two vertebral arteries , each arise from the 1st part of the subclavian artery and ascends through the transverse foramina of the first six cervical vertebrae giving off small muscular branches →then enters the skull through the foreman magnum , unites with the opposite one at the ponto- medullary junction to form the basilar artery which ascends upwards to the ponto- midbrain junction where it divides into its two terminal posterior cerebral arteries .

***Branches of the vertebral artery :-**

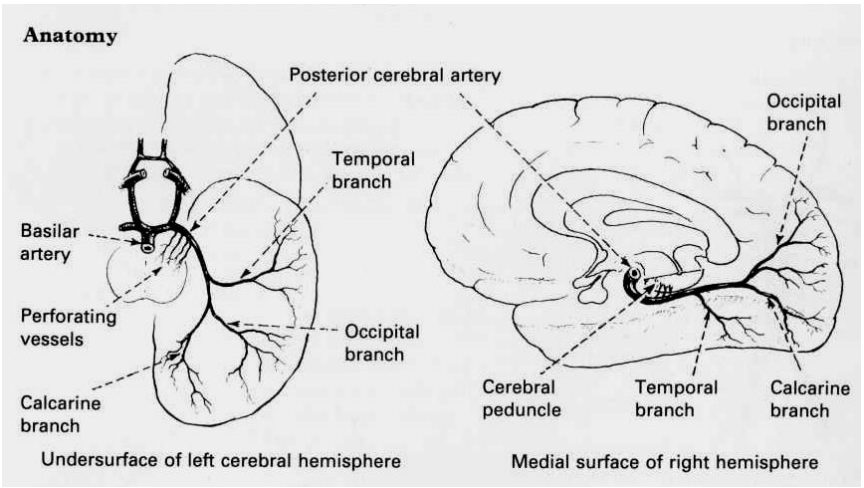
- To the meninges
- *Anterior & posterior spinal arteries to the spinal cord .*
- Posterior inferior cerebellar artery to the cerebellum .
- Small penetrating arteries to the medulla.

***Branches of the basilar artery :** Fig(78)⟶

- The anterior inferior cerebellar artery .
- The superior cerebellar artery.
- The internal auditory artery .
- Small penetrating branches to the brain stem .



***The posterior cerebral artery :**
Encircles the mid brain
and Fig (79) ⟶



supplies the:-

- Inferior part of the temporal lobe.
- The occipital lobe.
- Midbrain , thalamus , hypothalamus , and geniculate bodies .
- Posterior 3rd of the posterior limb of the internal capsule

Each posterior cerebral artery communicates with the ipsilateral middle cerebral artery through the posterior communicating artery.

Regulation of the cerebral blood flow (CBF)

CBF is affected by the following factors:-

1- **Arterial CO_2 tension** →arise of Pa CO_2 of 1mmhg →causes an immediate increase in CBF of 5%.

2- **Arterial O2 tension** →its effect is less than that of CO_2.

3- **Blood viscosity (haematocrite)** →CBF is inversely related to the whole blood viscosity.

4- **Arterial blood pressure** : CBF remains constant when the mean systemic blood pressure is between 60 -160mmhg which is known as auto regulation .

- Decrease systemic arterial blood pressure → vasodilatation → increase CBF →until exhaustion occurs → oligemia & ischemia.
- Increase systemic blood pressure → vasoconstriction →decrease CBF → until exhaustion occurs→ hyperemia &ischemia.

In chronic hypertensive patients, the auto regulation range is shifted upwards.

- Auto regulation is impaired in:
  - Previously damaged brain (e.g. old stroke, trauma …)
  - Elderly. - Organ failure.
  - Metabolic impairment.

Stroke and hemiplegia

Definition :-

■**Stroke:-** Is a rapidly developing clinical symptoms and or signs of focal or global loss of cerebral function, lasting more than 24 hours or leading to death, with no apparent cause other than that of vascular origin.

■**Transient ischemic attacks (TIA):**

Transient focal neurological deficits lasting for less than 24 hours, usually minutes of vascular ischemic origin with complete recovery.

■**Reversible or focal ischemic neurologic deficit (RINDS or FINDS) :**

Transient focal ischemic neurologic deficits lasting >24 hours with mild ischemic strokes with no persisting neurologic disability.

NB-*Cerebral infarction is responsible for nearly 80% of all first strokes,*

-Hemorrhage for 10%,

-Subarachnoid hemorrhage for 15%,

-Uncertain causes 5%.

Risk factors for stroke:

1) **Age:** is the strongest and inevitable risk factors of stroke, stroke in people aged 75-84 years is 25 times the risk in people aged 45-54 years.
2) **Sex:** there is small male excess of strokes mainly in middle age most probably due to prophylactic role of endogenous sex hormone in females (estrogen).
3) **Hypertension:** either elevated diastolic or systolic blood pressure is associated with increased risk, with each 7, 5 mmhg increase in diastolic blood pressure stroke risk doubles.

-Hypertension causes stroke (infarction) through its continuous friction power with increasing the extent and severity of atheroma, or with sudden rise → rupture of vascular malformations causing hemorrhage.

NB-Hypotension also in aged atheromatous cerebral vessels causes strokes (infarction).

4) **Diabetes mellitus:** doubles the risk of stroke compared with non-diabetics.
5) **Cardiac diseases:** e.g. coronary heart diseases, heart failure, A.F, valvular diseases, arrhythmias, all are important risk factors for embolic infarction.
6) **Dyslipidemia:** with high levels of low density lipoprotein (LDL) and decreasing levels of high density lipoprotein (HDL) is considered a highly risk factor for stroke & coronary heart diseases.
   →A reduction of plasma cholesterol by 10% reduces risk of coronary by 20%.
7) **Hyper coagulability:** with increase plasma fibrinogen, increase haematocrite, risk of stroke is increased (polycythemia), the risk is attenuated by cigarette smoking.
8) **Alcohol consumption:** increases the blood pressure & blood lipids and also is usually associated with AF, cardiomypathy consequently increase the risk of stroke.
9) **Coffee :** Consumption of boiled unfiltrated coffee has a small hyper lipaedemic effect.
10) **Cigarettes smoking:** is an important risk factor for stroke either infarction or hge (doubles stroke).Recently reported to be one of the major 5 risk factors of stroke
11) **Dietary habits:** diet rich in fatty fish, with moderate salt intake, high k, and rich in fresh fruits, and vegetables, vitamin E, C, folic acid & selenium have a protective value.
12) **Drug induced stroke:**
    - Hypotensive drugs.
    - Cytotoxic drugs → thromboembolic strokes.
    -Aspirin, anticoagulants, thrombolytic therapy → Hge.

13) Contraceptive pills: with high doses of estrogen >50 ug → triple the risk of stroke while lower doses of estrogen lowers the risk , therefore post menopausal estrogen replacement may have some protective effect.

14) Past history of TIA or mild stroke.

15) Family history of stroke.

16) Lower social class.

17) Body built. Obesity mainely truncal obesity, ↑ body mass index, → ↑ risk of stroke

18) Physical inactivity.

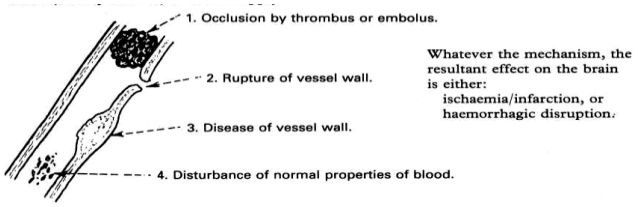
19) Chronic stress.

The causes of stroke: Fig(80)

Stroke may be - Ischemic → thrombotic
Embolic
} 80%

-Hemorrhage → cerebral hemorrhage 10%
Subarachinoid hge 5%

-Uncertain causes 5%



Fig(80)

1-Cerebral ischemia (infarction)

Etiology:

a) Arterial wall diseases

- Atherothromboembolism →50 % of causes Fig(88)
- Intracranial small vessel diseases →25 % of causes

Fig(81)

b) **Embolism from the heart →20% of causes**

1- Paradoxical embolism : - Atrial septal defect
- Ventricular septal defect
- Patent foramen oval

2- Left atrium: - AF - Myxoma

3- Mitral valve: -Rheumatic stenosis or incompetence
- Infective endocarditis - Prosthetic valve
- Mitral valve prolapse - Mitral valve calcification.

4- Left ventricle:
- Myocardial infarction. - Cardiomypathy.
- Myxoma. - Aneurysm.

5- Aortic valve:
- Rheumatic stenosis or incompetence. - Infective endocarditis.
- Prosthetic valve. - Calcification.

6- Congenital cardiac diseases.

7- Cardiac surgery, catheterization, angiography.

c) **Hematological disorders:-**

- Polycythemia - Thrombocytopenia
- Leukemia - Sickle cell disease
- Iron deficiency anemia. - Thrombotic thrombocytopenic purpura.
- Disseminated intravascular coagulation.(DIC) - Hypercoagulability.

2) Intracranial hemorrhage

This may be-

1. Subdural, extradural → mostly traumatic.
2. Subarachinoid hemorrhage.
3. Intraventricular hemorrhage. 4-Cerebral hemorrhage. 5-Cerebellar hemorrhage.

Causes of spontaneous intracranial hemorrhage:-

1. Hypertension 5- Vascular malformation.
2. Aneurysms. 6-Hemorrhagic blood diseases.
3. Vascular tumors. 7 Anti platelets & anticoagulant, thrombolytic therapy.
4. Intracranial venous thrombosis. 8- Drug abuse.

N.B: the vascular causes are the most common in hemplegia.

NB- Other causes of hemiplegia

1. Space occupying lesions: e.g. abscess, tumors.
2. Infection: encephalitis, meningoencephalitis.
3. Demyelinating: disseminated sclerosis, disseminated encephalomyelitis.
4. Congenital cerebral palsy.
5. Hysterical.

***Clinical presentation and localization of a case of hemiplegia*.**

The clinical presentation of hemiplegia varies much according to-

- The site of the lesions in the nervous system.
- Etiology of hemiplegia.

General clinical features:-

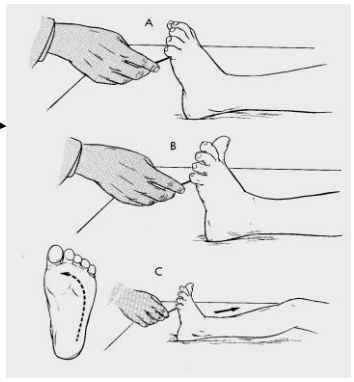
1) **Onset:** is usually acute in hemorrhage, may be subacute in thrombosis, sudden in embolic, gradual in neoplasm, intermittent in D.S.
2) **Cause:** may be regressive in inflammatory causes, vascular, traumatic causes, Progressive in neoplasm.
3) **Weakness:** is usually affecting one half of the body UL ,LL in equal degree or one may be affected than the other , weakness affects group of muscles ,affecting fine movements more , distal muscles are more affected , progravity muscles are more affected than antigravity - i.e. In UL→ Extensors are weaker - In LL→ Flexors are weaker
4) **Muscles tone :** -In acute lesions there is a shock stage lasting for 2-6 weeks , during which there is a complete loss of tone of the paralyzed side , after this stage tone gradually returns and spasticity appears

- In gradual lesions: spasticity develops from the start and affects the antigravity muscles more than progravity muscles
- i.e. UL – flexors are more spastic - LL – extensors are more spastic & adductors are more affected than abductors.

5) **Reflexes** :

-Deep tendon reflexes in the affected limbs are exaggerated, pathological reflexes and clonus may be elicited. (**N.B**: in the shock stage deep reflexes are lost or diminished)

- Superficial reflexes:-
  - +ve babinski sign. Fig(82)
  - lost abdominal and cremasteric on the affected side.



5) **Gait**: is usually circumduction.
6) **Other features** of sensory impairment, cranial nerves affection dependant on site of the lesions as shown later in discussion of localization.

Clinical approach in localization of the site of the lesion in hemiplegia

Hemiplegia occurs in lesions affecting the pyramidal tract along its course from:-

1) Cerebral cortex (cortical lesions)
2) Sub cortical lesions
3) Capsular
4) Brain stem, (mid brain, pons, medulla).
5) Spinal cord

1) Cortical lesions : -

- Usually hemiplegia is not complete, monoplegia is more encountered due to the wide origin of pyramidal tract, and weakness is contra lateral.
- Cloudiness of consciousness is common.
- Contra lateral cortical sensory loss in involvement of the parietal lobe .
- Convulsions, which may be focal or secondary generalized, in cases of irritative or extensive lesions.
- Higher mental functions disorders e.g. aphasia, agraphia, and agnosia, in lesions affected specific lobes, centers, or sites.

2) Subcortical lesions:-

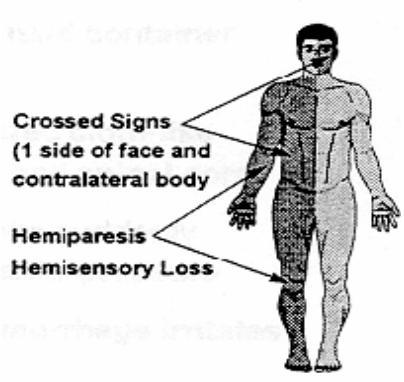
As in cortical but weakness is more extensive.

3) Capsular : the commonest

- Hemiplegia is complete
- Hemi hypothesia on the paralyzed side.(common)
- UMN facial and hypoglossal on the same side of paralysis(common).

Brain stem :- Fig(83)

Hemiplegia in brain stem lesions is termed crossed hemiplegia ,characterized with→hemiplegia on the opposite of the lesion with LMN cranial nerves affection on the same side of the lesions (opposite to the hemiplegia). Fig(83)



a) Mid brain syndromes:-eg

Weber's syndrome :

- Ipsilateral 3rd cranial nerve lesion
- Crossed hemiplegia (contra lateral).

b) **Pons syndromes :eg**

Millard Gubler syndrome :

- Ipsilateral 6th , 7th cranial nerves lesion
- Crossed hemplegia.

c) **Medullary syndromes :eg**

Wallenberg syndrome :

-Ipsilateral -Horner's syndrome

-Ataxia

-9, 10, 11th cranial nerves

- Decrease sensation over the face

- Contra lateral hemianathesia

4) **Spinal cord lesions (Brown Sequard's Syndrome):-** Fig(84)

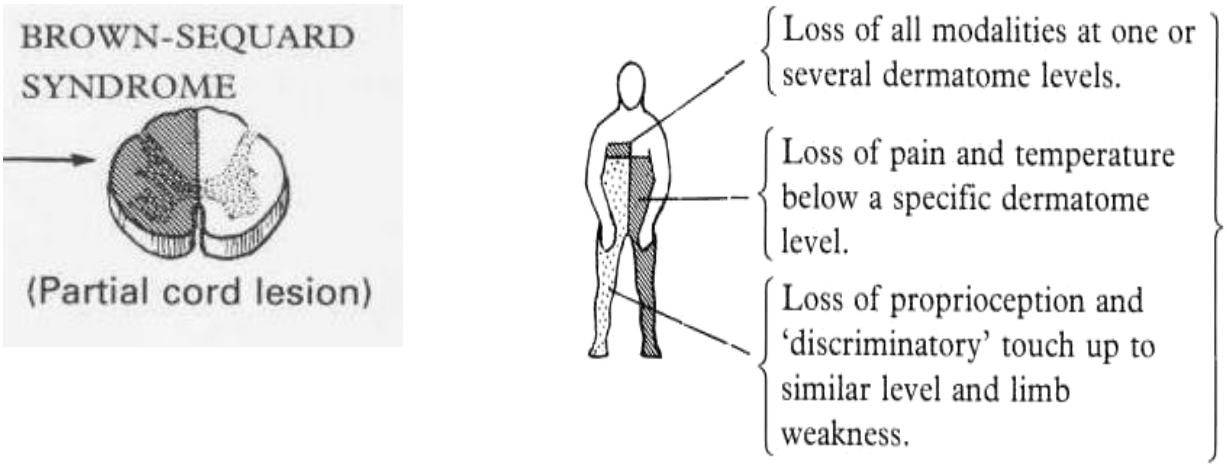
In hemi lesion of the spinal cord in upper cervical segments, →hemiplegia may occur with the following features:-

1- **At the level of the lesions**:

- Ipsilateral LMN weakness of muscles supplied by the affected segments
- Loss of reflexes mediated by the interrupted segments
- Loss of sensation (radicular) in the area supplied by the diseased segments

2- below the level of the lesion :

- Ipsilateral hemiplegia
- Ipsilateral deep sensory loss
- Contralateral superficial sensory loss.



Fig(84)

Differential diagnosis of stroke:

The diagnosis of stroke versus non stroke is straight forward if there is a history of acute or sudden onset of focal neurologic deficits of vascular origin , if the history is not clear C.T will solve the problem , but clinically the followings must be differentiated :-

1) **Brain tumors:** history of headache, papilloedema, seizures, blurring of vision, progressive course, with gradual onset.
2) **Chronic subdural hematoma:** history of head injury, weeks up to years before, more drowsiness, confusion & headache than the focal deficits.
3) **Encephalitis:** acute or subacute onset of fever, cloudiness of consciousness, convulsions, signs of meningeal irritation, higher mental function disorders.
4) **Brain abscess:** history of fever, headache with signs & symptoms of increased ICT, focal deficits, with source of infections e.g. mastoditis, OM, congenital heart diseases.
5) **Todd's paralysis:** history of epileptic seizures of focal onset.
6) **Hysterical hemiplegia:** usually occurs in young age, with no +ve data on examination.

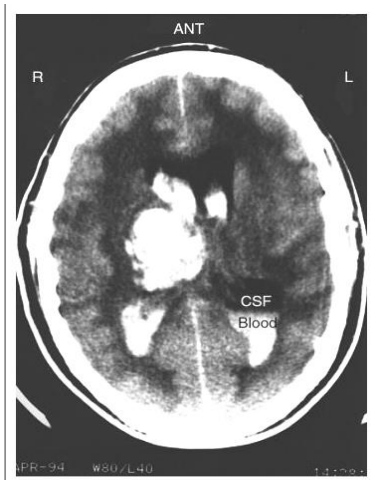
Investigation of a case of stroke:

1) **Specific investigations:-**
    1. **C.T scanning :**-The next step after clinical diagnosis of stroke is to do C.T to diagnose the type and site of stroke

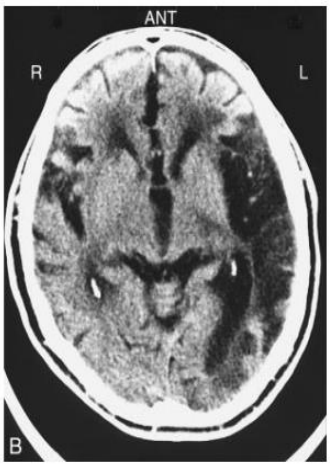
- Hemorrhage appears once on C.T as well-demarcated hyper dense area (white).

See Fig(85)

-Infarction appears as hypo dense area(black).Fig(86),may take up to 48 hrs to appear on the CT



Fig(85) cereberal haemorrhage



Fig(86) cereberal infarction

2. **M.R.I:**

   More sensitive, can show early infarctions not seen on C.T, can shows small or deeply seated infarction.

3. **Angiography :**

   Imaging the cerebral circulation may show stenotic, dissecting vessels, vascular malformations, M.R.I angiography is more sensitive.

2) **General investigations :-**

- ECG 24 hours monitoring →ischemic heart diseases & arrhythmia.
- Renal function tests → renal impairment in hypertension
- Lipid profile → atherosclerosis
- Blood sugar → diabetes mellitus
- Blood electrolytes → electrolyte imbalance
- Echocardiography →cardiac source of emboli.
- X ray chest → cardiomyopathy & calcified valves.
- CBC & coagulation profiles → hematological disorders.

Treatment of acute stroke:

(1)-General measures:

A- **Maintenance of vital signs:**

a- **Pulmonary functions**, open air way, observe type and rate of respiration, blood gases, O2 supply if there is hypoxia.

b- **Cardiovascular**:

-Blood pressure (normal arm) not deal with blood pressure up to 200/120 withen the 1st days, if >200/120→ gradual decrease of blood pressure is needed , urgent deal only in cases of hypertensive encephalopathy

- Heart rate. -ECG

c- **Blood electrolytes** and fluid intake versus output.

d- **Temperature (**normal axilla)

e- **Blood glucose**.

B- **Nursing care :-**

- Turn the patient on bed regularly at least every two hours.
- Frequent massage & washing the body with alcohol & talc powder, the patient must be kept clean and dry with regular change of the bed sheets.
- Catheterization if there is retention or incontinence with measuring urine volume/ 24 hours.

-if the patient is comatosed, or if there is shocking, nasogasteric tube must be inserted for oral feeding with IV fluids (from 1, 5 litre -3litre /day).

N.B: Routine monitoring in acute stroke:

▸Vital signs:-

- Respiratory rate, rhythm, blood gases.
- Heart rate, rhythm, ± ECG monitors
- Blood pressure
- Temperature.

▸ Neurological:-

- Conscious level.
- Weakness.
- Pupils.
- Seizures

▸ General:-

- Fluid intake.
- Electrolytes & urea
- Blood glucose.

(2)-specific measures:

1) **Dealing with the occluded vessels (thrombus , embolus in infarction only)**

1. **Anticoagulants (heparin) (??doubtful)may be of value in :**
   - TIA , progressive stuttering stroke
   - Small infarction.
   - Cardiac source of embolism.
   - Young age patient.

   Of doubtful value (better to be avoided) in:-
   - Hypertensive(resistant) patient. ,Complete hemiplegia
   - Elderly patients. , Massive infarctions
   - History of recent bleeding peptic ulcer.
   - Haemorrhagic tendencies.

   Dose 20000-30000 IU infusion/day guided by PTT lap.adjustment
2. **Antiplatelets :-**

   - **Aspirin** in dose of 300 mg once for about 2 weeks to be reduced into maintaince dose of 75 -150 mg/day forever.-**Clopedigri**l 75 mg daily(in aspirin contraindications)
3. **Thrombolytic therapy :-**

   -Streptokinase, urokinase → of doubtful effect.

   -Tissues plasminogen activator(tPA) now is widely used ,highly effective if used within the therapeutic window (3-6 hours of the onset),with dose IV 0.9 mg/kg/day

   Exclusion criteria:

   - Patients older than 80 years
   - All patients taking oral anticoagulants regardless of international normalized ratio (INR)
   - Patients with a history of stroke and diabetes

2) **Improve cerebral blood flow :-**

- Hypertensive agents in hypotension.
- Vasodilators e.g. CO_2 inhalation, papaverine.
- Haemodilution (dextran) → ↓ haematocrite.

3) **Dehydrating measures to relieve brain edema :-**

-Manitol 1, 5 – 2 gm/ kg once, then to be divided /4hours.
- Glycerol 30 ml /4-6 hours.
- Corticosteroids 4-6 mg / 4-6 hours.

} For up to one weak& gradual withdrawal.

4) **Neuronal protection :-**

- Barbiturates. - Phenytoin.
- Hypothermia - Ca^+ channel blockers.
- Vit E, C, selenium. - Free radical scavengers. - Hyperparic O2 .

5) **Dealing with systemic complications which may occur and cause neurological deterioration as:-**

1. - Pneumonia
2. - Cardiac failure.
3. - Dehydration.
4. - Bedsores.
5. - Bleeding stress ulcer.
6. -D.V.T.
7. - Pulmonary embolism.
8. – Cardiac arrhythmia.
9. – Electrolyte imbalance.
10. – Urinary tract infection.
11. – Septicemia.

6) **Dealing with neurological causes of deterioration as:-**

1. Haemorrhagic transformation of infarction.
2. Brain shift.
3. Recurrent haemorrhage.
4. Seizures.
5. Cerebral edema.
6. Recurrent embolism.
7. Propagation of the thrombus.

7) **Rehabilitation after the 1st few days by the physiotherapist.**

8) **Starting prevention :-**

- **Control of the risk factors** mainly DM, hypertension, cardiac causes, hyperlipedemia, smoking, etc.
- **Anticoagulants ,** in selected cases as discussed before with oral anticoagulants as warfarine ,(do not forget to adjust the dose with frequent INR lab.assesment).
- **Antiplatelets** are given mainly to those:
- With TIA.

- Mild ischemic stroke.
- With high vascular risk factors.
- Carotid endarterectomy in symptomatic patients (TIA, mild ischemic stroke) with internal carotid artery stenosis.

N.B:-

Well organized care from professionals who are familiar with stroke should lead to a better outcome, this may explain why the prognosis is better in stroke unite than general medical unites where the patients are investigated & treated by the neurologist.

Subarachinoid haemorrhage (SAH)

Definition:

Haemorrhage in the sub arachnoids space.

Causes:

May be ***primary*** due to:

- Hypertension.
- Rupture of intracranial aneurysm.
- Rupture of angiomatous malformations
- Blood diseases.

Secondary due to:

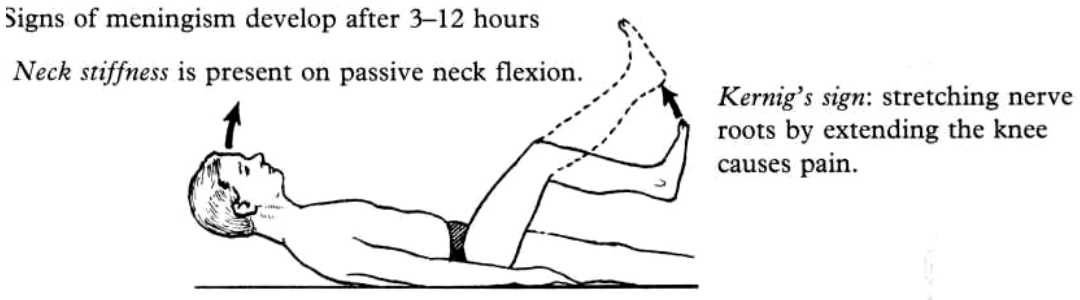
- Anticoagulants & antiplatelets.
- Haemorrhage into brain tumors
- Head trauma.
- Inflammatory vessel wall diseases
- Haemostatic failure.
- Vascular tumors.
- Intracranial venous thrombosis.

Clinical features:

Usually precipitated by prolonged straining, coughing, or sexual intercourse.

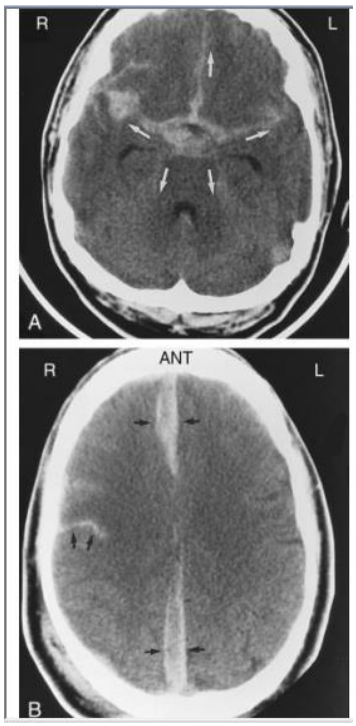
- **Sudden onset of headache:** generalized more in the occipit and neck, ↑ by flexion of the neck with radiation to the back, shoulders & legs.
- **Nausea, vomiting, fever, and vertigo.**
- **Signs & symptoms of meningeal irritation** Fig(87)

As rigid & stiff neck , painful back , painful restricted straight leg rising sign , +ve kerning's sign & brudzinski signs.



Fig(87)

- Patients are usually **irritable, photophobic** for several days later.
- **Neurological symptoms :**
  - **Seizures**.
  - **Loss of consciousness** may be brief in half of patients.
  - Focal S&S may suggest **intracerebral haemorrhage or local pressure**.- **Cranial nerves palsy e.g. (3rd, 4th, and 6th)** **Diagnosis & investigations:**



1) **C.T :** Fig(88)⟶

Enhanced C.T scan in >90% of patients show intracranial blood in the subarachinoid space, also may show associated intracerebral, ventricular hge or complicating hydrocephalus , also the scan may show calcification in the rim of bleeding aneurysm.

2) **Lumber puncture ,** if the C.T is unavailable , the CSF will be :

- Under tension. - Bloody. - ↑ protein content. - Xanthochromia.

N.B:

-It is important to remember that, blood and xanthochromia may not appear up to 12 hours after the insult.

-Lumber puncture may precipitate cerebellar coning or transtentorial herniation in presence of cerebral edema, cerebral or cerebellar haemorrhage, therefore C.T and fundus examination must be made first.

-Bloody CSF may be traumatic from the spinal needle and must be differentiated from the bloody CSF of SAH by collecting CSF in three consecutive tubes , where we will observe clearance of the CSF of blood in cases of traumatic bloody CSF but not in cases of SAH.

3) **Cerebral angiography :**

On recovery, angiography must be done to search for the causative aneurysm , vascular malformations for possibility of surgical interference, where recurrences are common in cases of SAH with increasing rates of mortality and morbidity on recurrent bleeding.

Treatment:

1. The patient should be **nursed in a quiet, dark room.**
2. Headache , nausea , vomiting , irritability , seizures , cough , constipation , all if present are in need for **symptomatic treatment .**
3. **Dehydrating measures** as manitol 25% in presence of cerebral edema.
4. **Dealing with the complications with the onset as:-**

- Hydrocephalus which occurs in >20% of cases due to obstruction of the CSF flow by blood , → draining may be urgently needed or frequent lumber puncture ,

- Cerebral edema →dehydrating measures.

- **Delayed cerebral ischemia** occurs in 25% of patients 4-14 days after the onset → give nimodipine 60 mg/ 4hours orally for 21days from the onset.

- **Hyponatremia,** occurs in about 1/3 of patients 1-2 weeks after SAH → give glucose – saline or plasma but not water restriction.

- Rebleeding which may occur and cause sudden neurological deterioration , → prophylactic use of antifibrinolytic drugs is advised.

-**Surgical interference** for the aneurysm or the vascular malformations.

Cerebellar disorders(Ataxia)

***Cerebellum** acts as a modulator of motor activity originating in other brain centers , mainly it excitates automatically antagonist muscles at the end of movement , with simultaneous inhibition of the agonist muscles that initiated the movement . It coordinates movements, equilibrium & muscle tone.

***Anatomically.**

It is formed of two cerebellar hemispheres separated by the vermis , composed of three lobes anterior, posterior & floculonodular lobes.

***Embryogenically & functionally**

It is formed of three lobes:

***Archicerebellum** (vestibulo-cerebellum) → equilibrium

***Paleocerebellum** (spino-cerebellum) → muscle tone

***Neocerebellum** (ponto-cerebellum) → voluntary muscle activity

***N.B** - Recently it showed that cerebellum shares in motor learning & cognitive functions

***The cerebellum** is connected to the brain stem by three large cerebellar peduncles: superior, middle & inferior

***Blood supply of the cerebellum >>three arteries :**

-Posterior inferior cerebellar artery… from the vertebral artery.

-Anterior inferior cerebellar artery…from the basilar artery.

-Superior cerebellar artery……from the basilar artery.

***Clinical manifestation of cerebellar dysfunction(in general):**

1- **Head ;** Nodding, abnormal postures(tilting)

2-**Eye;** Nystagmus , ocular dysmetria, skew deviation

3-**Speech** ; Dysarthria in the form of staccato, scanning, explosive speech

4**- U.L;**

Intentional kinetic tremors (tremors on reaching the target).

Decomposition of movements.

Dysmetria**,** dysdiadocokinesia ,jerkiness of movement.

Hypotonia, rebound phenomenon.

5-Trunk;

Titubation , trunkal instability(tested by asking the patient to sit without back support, with extended arms ,then ask the patient to sit or arise from the chair without use of arms)

6-L.L; as in U.L

7-Gait; -Ataxic gait ;(wide base, drunken) , deviated to one side (in unilateral cerebellar lesions)

-Can be enhanced by tandem gait, heel or toe walking, walking backward

8-Hypotonia

Generalized , more in U.L , ipsilateral .....(.pendular reflex-cerebellar drift ,rebound phenomenon)

***Other unusual features;**

-Asthenia & fatigability ,barognosia(misperception of weight)

-Macrographia , -Vertigo

-Catalepsy (sudden collapse due to loss of postural tone)

-Cerebellar mutism(transient loss of speech in children after posterior fossa surgery)

-Cerebellar fits (episodes of decerebrate rigidity that seen with large midline cerebellar masses

***N.B**

Cerebellar features can be summarized & remembered by the term

HANDS TREMORS

H-Hypotonia , A-Asynergy, N-Nystagmus(ocular features), D-Dysartheria(speech),S-Stance,gait

Atiology of cerebellar ataxia:

1-*Congenital* e.g. Ataxic cerebral palsy

2-*Inherited*

--A) With known metabolic defect.

--B) Without known metabolic defect

-1- Early onset -2- Late onset

3-*Acquired causes*

-Mostly reversible

-Frequently present as acute. often treatable if recognized early.

****Causes of acquired cerebellar ataxia**

1) Vascular Infarction ,Hge, TIA, basilar migraine ,vascular malformation, SLE

2) Neoplasm Medulloblastoma, astrocytoma ,metastasis

3) Infection T.B ,abscess ,cysticercosis.

4) Inflammatory&autoimmune

M.S ,acute postinfective cerebellitis ,paraneoplastic , Miller Fisher syndrome

5) Metabolic

-Hypothyroidism -Hyponatremia -Hypoxia -Hypoglycemia - Hypo-hyperthermia

-Hepatic encephalopathy

-Deficiency of ,Vit. E,C ,zinc ,essential amino acids,niacin ,thiamine

6) Drugs & toxins

-Anticonvulsants (phenytoin,carbamazepine ,barbiturates ,valporate)

-Chemotherapy ,alcohol ,organophosphorus

-Heavy metals ,volatile agents , lithium

7) Developmental

-Dandy Walker syndrome.>obstruction of foramina of the 4th ventricle.

-Chiari malformation> congenital displacement of the cerebellar tonsils downward into the cervical canal.

-Hydrocephalus.

-Foramen magnum compression.

8) Post traumatic

-Post concussion

-Contusion ,haematoma

****Inhireted ataxia with known metabolic defects**: e.g

Ataxia telangectasia(ataxia with defective DNA repair)→clinically characterized by:

.Cerebellar ataxia .Physical & mental growth retardation

.Dystonia ,chorea .Peripheral neuropathy

.Cutaneous telangiectasiae

****Inherited ataxia of early onset with unknown aetiology:**

-All are autosomal recessive & age of onset <25 years

1-Friedreich ataxia;

-The commonest of early onset ataxias & form >50% of early onset ataxias.

***Clinical picture of Friedreich ataxia**

-Age of onset between 8-15years

-Autosomal recessive

-The clinical picture reflects the pathological features which are situated in the following sites:

i) Spinocerebelar tracts &Purkinje cells of the cerebellum ; resulting in

-Gait ataxia which considered the most frequent presenting symptom

-Limb ataxia -Ataxic speech

-Nystagmus ,ocular dysmetria

ii) Pyramidal tracts; resulting in

-Pyramidal weakness of legs

-Extensor planter responses

iii) Peripheral nerves; resulting in

-Absent tendon reflexes

-Distal wasting+ glove &stock hypothesia

iv) Posterior column; resulting in

-Loss of position &vibration sense

-Positive Rombergism

v) Optic tract; resulting in

-Optic atrophy

-Reduced V/A

vi) Other associative features of Friedreich ataxia

-Nerve deafness in >10 % of cases

-Scoliosis, pes cavus & equinovarus

-Cardiomyopathy in > 2/3 of patients,

-Abnormal ECG

-Dementia>> mild degree

-D.M in >10 % of cases

-Sphincteric dysfunction in the form of urgency

NB. Most patients become unable to walk 15 years after the onset of the disease

2-Other examples of early onset ataxias

*** Holmes ataxia**

-Cerebellar ataxia -Mental retardation , dementia

-Hypogonadotrophic hypogonadism -Choreoathetosis

-Retinopathy - Deafness

***Ramzy Hunt syndrome**

-Ataxia -Myoclonus -Tonic-clonic seizures

***Behr syndrome**

-Ataxia -Optic atrophy

- Spasticity -Mental retardation

****Late onset inherited ataxias of unknown aetiology(familial-spino cerebellar ataxia)**

-All are autosomal dominant

-Age of onset usually>25 years

Examples of autosomal dominant ataxias:eg

*** Machado-Josef disease**

-Age of onset 30-40 years up to 65 years

-Gait ataxia is the most frequent presenting feature

-Optic atrophy -Distal wasting -Ophthalmoplegia -Dementia

-Extra pyramidal features -Speech disorders of cerebellar &pseudobulbar features

***Idiopathic late onset ataxia e.g Maries atxia**

-Mean age of onset 55 years

-Progressive gait ataxia is the prominent feature

-Relative preserved coordination of the U.L

-Mild dysartheria,Dementia

Investigation of a case of cerebellar ataxia;

1- Neuroanatomical imaging eg CT, MRI

-Will reveal cerebellar & brain stem atrophy, sometimes with cerebral atrophy in most of inherited ataxias

-In acquired ataxias, the underlying pathology may be detected e.g. vascular lesions, infections, demyelination,SOL,…

2-Neurophysiological studies;

-EEG; May yield epileptic discharges in ataxic patients presented with seizures

-Visual evoked potentials; Abnormal in Friedreich ataxia & some other inherited ataxias

-Auditory evoked potentials; Abnormal also in some inherited ataxia

-Somatosensory evoked potential; May be abnormal also

-NCS & EMG; May yield neuropathic features in most of ataxias

3- Neurophysiological imaging; e.g PET ,SPECT

4- Metabolic screen; To search for the etiology of acquired ataxias.

*Management of cerebellar ataxia;

1-In acquired ataxias.

-Management is more obvious &easy on recognizing the cause

2-In inherited ataxias.

There is no definite therapeutic measures , but in general the following measures are needed.

i)- **Physiotherapy &correction of skeletal deformities**

ii)- Management of associative medical disorders

e.g D.M, heart disease

iii)-Supportive &symptomatic measures

iv)-Recent studies revealed some benefits from the following medications:

-isoniazide. -thyrotropin releasing hormone

-physiostigmine. -propranolol. -Vit. E

-amantadine -clonazepam

Movement disorders

Neuroanatomical consideration;

Many movement disorders are attributed to diseases in the basal ganglia which consists of group of ganglia & nuclei situated deeply in the brain & brain stem ,the term basal ganglia refers to :

A- **Caudate nucleus**

B- **Putamen** } A&B → both form the corpus striatum

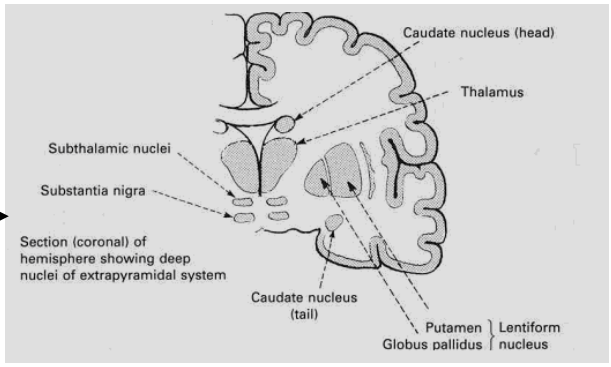
B&C → Both form the lentiform nucleus

C **Globus pallidus**

D- **Substantia nigra**

E - **Subthalamic nucleus** Fig(89)

Fig(89) →



Neurophysiological consideration;

The different components of the basal ganglia comprise many connections to(efferent),from(afferent)& within(internuclear).

***It receives afferents from**;

the cerebral cortex, thalamus ,brain stem &amygdala.

***Sends efferents to** ;

the cerebral cortex, thalamus ,brain stem & reticular formation.

-These different connections consume many different neurotransmitters, the most important of which is dopamine, other neurotransmitters include; glutamate ,acetyl choline ,GABA, serotonin, enkephalin, neurotensin & substance p,

-Through these different connections the basal ganglia exert its role in controlling:

****Voluntary motor activity**→planning & programming of voluntary movements & control of subconscious automatic movements

****Muscle tone** →Inhibition of muscle tone

****Cognitive function** →Through connections with frontal , prefrontal & limbic structures.

In parkinsonism,

There's a depletion up to 80% in striatal dopamine, also deficiency occurs to:

- Tyrosine hydroxylate -Melanin
-Serotonin -GABA

while increase in histones,& Acetyl choline occurs.

*Neuropathological changes;

→ With age, some aging degenerative processes occurs in the whole brain, including the basal ganglia which include;

-Depletion of dopamine

-Decrease in the fresh volume of Substantia nigra with loss of nigral neurons

→In compensated cases:

- Increase in the activity of the remaining intact dopaminergic neurons will occur, associated with presynaptic feed back regulation ,and super sensitivity of synaptic receptors .

→ In decompensated cases ,& for the symptoms & signs of Parkinson's disease to appear the following must happen;

-Up to 25% decrease in the fresh volume of Substantia nigra

-Up to 80 % loss of pigment neurons

-Up to 80 % depletion of dopamine

Classification of movement disorders;

(A)-Akinetic rigid syndromes (parkinsonism);

i- Idiopathic Parkinson's disease

ii- Arteriosclerotic parkinsonism

iii- Postencphalitic parkinsonism

iv- Symptomatic parkinsonism;

1-Drugs(metoclopromid,methyldopa,captopril & phenothiazines)

2-Toxins & poisons- co, manganese,

MPTP(methyl phenyl tetrahydropyridine),TIQ(tetra hydroiso quinoline)

3-Repetitive head trauma (punch drunk syndrome)

4-CVA

5- Cerebral anoxia

6- Brain tumors

v-Degenerative causes(Parkinsonism plus syndromes);eg

1-Progressive supranuclear palsy

2-Multiple system atrophy (shy dragger syndrome)

3-Wilson's disease

(B)-Dyskinesias

*Chorea *Dystonia *Athetosis -

*Hemiballismus *Tics *Myoclonus

Parkinson's disease(shaking palsy):

■**Aetiology:**

-Natural aging process in autoimmune predisposed patients

-Exposure to endogenous toxins;

. MPTP which is converted by MAO- B to the toxic MPP(methylphenylpyridinium) which will destroy nigral neurons

.TIQ (endogenous or exogenous toxin which may destroy dopaminergic neurons)

-May be genetically determined

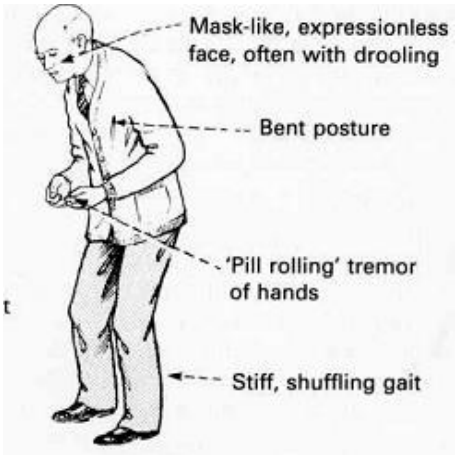
-Defective anti- oxidant system

-Other risk factors ? rural residence, exposure to well water, pesticides and herbicides, farming, wood pulp mills, and iron and steel production

■**Neuropathological & Neurophysiological changes;**

discussed before.

■**Clinical features;** Fig(90)



(1) **-Rigidity;**

-Present in all cases with onset,and presented as:

- Clumsiness of movement,
- Stiffness,
- Difficulty in turning over the bed , Fig(90)
- Difficulty in changing position,
- Difficulty &fatigue in waking

-It is generalized in all muscle groups, more in axial, flexors & proximal muscles giving the body flexion attitude and stooping forward (gorilla like attitude)

-May be equal in the same degree all through the movement "lead pipe" or interrupted by the tremors" cog wheel"

(2)-Tremors;

-Present in at least 80 % of patients

-Static, may be confined to one upper limb,or one side

-Regular ,rhythmic with rate of 4-6/sec

-In the form of pill rolling ,counting money or drum tapping

-Increased by emotions ,fatigue & rest &decreased on movement & on sleep →disappear

-Marked in wrist joint & fingers ,less in ankle , head ,tongue &mandible

(3)-Disorders of voluntary movements.

-Slowness of movements “bradykinesia”up to akinesia.

-Reduced amplitude of movements ”hypokinesia” evident in hand writing

-Reduced or lost arm swinging

-Clumsiness &difficulty of hand & fine movements

- Power , reflexes & planter responses, all are normal

(4)-Postural instability: Propulsion –retropulsion When the patient is pushed forward or backward ,the patient cannot keep erect posture & fall down

(5)-Speech. Soft monotonous voice.

(6)- Gait Short steppage ,festinant, rapid small steps, shuffling(friction with ground).

(7) -Facial features. Immobile ,mask face,reduced facial expressions & blinking.

(8)- Other features ; “not in all patients”

-Akathesia ; discomfortable restlessness sensation ,requiring continuous desire to change position

-Autonomic features ; Constipation ,sweating ,greasy appearance ,flushing, hypothermic episodes ,excessive salivation urinary frequency or incontinence

-Mild eye movement disorders ; . limitation of upgaze

. Paralysis of convergence

. Pursuit eye movement is replaced by saccadic movements

-Cognitive functions ; Normal in early stages , later on ,depression psychotic episodes

N.B ; rigidity & tremors usually are confined to one limb or to one side months or years before generalization to the other limbs

Diagnosis & differential diagnosis of parkinsonism:

■ **Parkinson’s disease characters**

- Age between 55-70.years

- C/P as described above .

- Asymmetrical presentation with gradual onset & slowly progressive course.

- Good response to levodopa.

-Absence of symptomatic causes.

■ **Idiopathic Parkinson’s disease must be differentially diagnosed with** ⟶

i) Drug induced parkinsonism;

-History of taking neuroleptics in psychotic patient.

-Taking antiemetics or vestibular sedatives.

-Of acute or sub acute onset.

-Prominent extra pyramidal rigidity with tongue protrusion

ii) Other symptomatic cases ; from the history

iii) Post encephalitic parkinsonism;

-History of encephalitis “fever ,convulsions ,neurological deficits months or years before.

-Any age .

-Prominent oculogyric crisis (spasm of up gaze).

-Prominent behavioral changes.

-Change in sleep rhythm.

-Respiratory disorders.

-Dyskinetic movements as chorea ,dystonia, myoclonus.

-Rigidity more common.

-Pyramidal signs.

iv) Arteriosclerotic parkinsonism;

-Older age.

-Presence of vascular risk factors.

. Hyper or dyslipidaemia. .Hypertension.

.D.M .Cardiac disorders

-Pyramidal signs.

-The complete C/P comes after frequent vascular insults of acute onset.

-Signs & symptoms of pseudo bulbar palsy.

v) Wilson’s disease (Hepatolenticular degeneration).

-Age under 50 years. - Heredofamilial(Autosomal recessive) .

-There is deficiency of ceruloplasmin (enzyme binding copper) leading to deposition of copper in → **lentiform nucleus** , **liver** ,**cornea …..clinically →**

■Behavioral & psychotic disorders .

■Hepatic disorders .

■Presence of Kayser Fleisher ring in the cornea (by slit lamp)

■Other Dyskinetic or pseudocerebellar disorders.

■Positive lab. Investigations-as

. Low ceruloplasmin in serum.

. Liver biopsy → increased copper concentration.

- **Teatment**: D.penicillamine(copper chelating drug) 250 mg 4 times daily before meal

Oral zinc reduces copper absorbition in early stagrs

vi) Progressive supranuclear palsy.

-Middle or older age

-Minimal or no levodopa response

-Symmetrical presentation

-Frontal lobe signs with prominent cognitive disorders

-Supranuclear ophthalmoplegia

-Postural instability

-Pyramidal tract dysfunctions, dementia, dysphagia, dysarthria

vii) Multiple system atrophy "shy drager syndrome.

-Age under that of parkinsonism 35-60 yrs.

-Akinetic rigid syndrome characteristic with;

.Classical resting tremors is unusual

.Little or no response to levodopa

.Early instability & falls

.More progressive course

.Other Dyskinetic disorders

- Cerebellar syndrome.
- Symptoms of autonomic dysfunction e.g

.Orthostatic hypotension .Urinary incontinence

.Loss of sweating .Sleep apnea & respiratory stridor

viii) Benign essential tremor is often misdiagnosed as Parkinson's disease. The postural tremor, length of the history (often many years by which time Parkinson's disease would be very severe), lack of additional neurological impairment, and family history should all help to avoid this error

Treatment of Parkinson's disease:

A -Medical treatment;

i) **Anticholinergic drugs**

-May be used as initial treatment in mild cases or with levodopa

-Helpful in reducing tremors & rigidity , have little effect on bradykinesia

→ Dose; cogentine 2 mg tablet 1 tab t.d.s

parkinol 5mg tablet 1 Tab t.d.s

→Side effects:- Blurring of vision - Dryness of mouth

-Confusional state - Constipation& retension of urine especially in elderly

→Contraindication: Cardiac ,hepatic patients, glaucoma

ii) Levodopa ;

■Converts to dopamine by dopa decarboxylase .where deficiency of dopamine is the main causative factor in Parkinson's disease

levodopa therapy is the corner stone in treatment of Parkinson' s disease

■Levodopa is given in combination with dopa decarboxylase inhibitor e.g carbidopa to prevent peripheral effects of dopamine(ratio 10-1)

-It s available as sinemet 275 mg tab

■Dose- must be introduced in small doses with gradual increase over a period of weeks or months up to ;2gm(4-8 tab)in 4-6 divided doses

■Side effects

-Nausea, vomiting -Postural hypotension

-Cardiac arrhythmias

-Psychiatric disturbances as confusion, agitation and hallucination

. -Movement disorders as end of dose akinesia or dyskinesia, on off effect

iii) Monoamine oxidase B inhibitor

deprenyl (jumex) prolongs levodopa action, dose ;5 mg tab t.d.s up to 30 mg /day.

iv) Dopamine agonists(as bromocriptine)

Dose 2.5mg tab with gradual increase up to 20-40 mg/day (8-16tab)

(B) Surgical treatment:

Intracerebral implantation of catecholamine producing cells (dopamine precursors)from adrenal medulla or fetal mesencephalon

-Not in common use, has limited value & spread

(C) Recent concepts in treatment of parkinsonism;

-*Controlled release of levodopa – carbidopa*

Guard against side effects (motor problems) of levodopa especially which occur in the morning due to the long period between the last night dose & the next morning dose

-Early use of levodopa carbidopa & deprenyl has a neuroprotective effect

-New dopamine agonist

As pergolid (permax has a neuroprotective effect)

-*Protein restriction* may enhance the efficacy of levodopa particularly the evening meal

-COMT(catechol amine o methyl transferase) inhibitors

which inhibit peripheral & central catabolism of dopamine (sparing dopamine)

-Neuroprotection of neurons of Substantia nigra

with the use of free radical scavengers & antioxidants

Chorea

Involuntary, static, sudden, jerky, dysrhythmic, pseudopurposeful movements due to lesion mainly in the caudate nucleus

Causes;

i) Genetically determined;

-Huntington chorea

-Benign hereditary chorea

-Neuroacanthocytosis.

ii) Symptomatic chorea;

1-Rheumatic chorea(Sydenham's chorea) 2 -chorea gravidarum

3-Thyrotoxicosis 4 -SLE

5-Postencephalitic 6 -Hyper-hypo-glycemia

7-Hypernatremia 8 -hypoparathyroidism

9-Polycythemia 10-hypo-hyper-calcemia

iii) Drug induced chorea;

1-Neuroleptics 2-Levodopa

3-Dopamine agonists 4-Anticholinergics

5-Contraceptive pills 6-Phenytoin

7-Antihistaminic 8-Cimitidine(H2 antagonist)

iv) Hemi chorea & Hemi ballismus.

-stroke, tumour, trauma & multiple sclerosis.

Rheumatic chorea:

-One of the major criteria of rheumatic fever & it s related to streptococcus group A infection but other streptococcal infection, scarlet fever & diphtheria can cause chorea

-Age of onset 7-12 yrs, in females more than males(2/3 of patients)

■Clinical picture:

-The initial symptoms are often irritability, behavioral & emotional disturbances

-Chorea may develop suddenly or gradually

-Chorea may develop generalized or may be unilateral in 20% of patients

-In rheumatic patients, about 1/3 of patients have evidence of carditis

■Movements; .

.In the tongue, can not keep it protruded& must be supported by his teeth

.In the face, frequent facial grimacing & impression with sudden jerky facial muscle movements

.In the arm, sudden jerky movements with dropping of objects,

can not keep arm quiet behind, jerkiness of shoulder.

-Movements are usually more proximal.

■Hypotonia;

The affected limbs are hypotonic, on outstretching the arms there is flexion of the elbow , extension of metacarpophalangeal & interphalangeal joints giving the hand boat shaped appearance.

■Behavioral disorders;

Irritability, confusional state, emotional instability(sudden crying or laughing).

■Treatment:

1 -Complete rest in bed

2 -Mild sedation with diazepam is recommended 2-4mg/t.d.s

3 -Dopamine antagonists

-Halopredol 3-10 mg t.d.s/day

-Chlorpromazine10 mg t.d.s up to 75 mg

-Na valporate may be used in resistant cases.

4 - Course of penicillin is needed with continous long acting penicillin up to the age of 20 years in rheumatic patients

Differential diagnosis of choreic movements;

1-Rheumatic chorea -discussed above

2-Huntington chorea ;

-Heredofamilial autosomal dominant

-Older age of onset 20 -55 years

-Clinically; .- Generalized chorea

-.Early, evident psychiatric disturbances in the form of personality changes, paranoid psychosis, emotional blunting, irritability

-Prominent gait abnormalities -Dementia

-Elements of Parkinsonism, dystonia & myoclonus

3-Neuroacanthocytosis;

-Age of onset 8-62 years - Heredofamilial

-Acanthocytosis; in blood film,(spiky appearance of RBCs)

-Cognitive dysfunction .Axonal neuropathy

.Akinetic rigid syndrome & seizures may be present.

4- Chorea gravidarum ;

-Common in primigravida

-Common in those with history of recurrent abortion with history of venous thrombosis

-May be associated with lupus anticoagulant

-Older age than that of rheumatic chorea

Dystonia

Static, intermittent or repetitive ,slow ,sustained muscle spasm, including dystonic spasm of neck muscles causing torticollis,retrocollis, anticollis ,spasm of back muscles causing lordotic or scoliotic postures, planter flexion & inversion of foot ,dystonic writers cramps

■Causes of dystonic syndromes ;

NB. Dystonia usually arise due to lesion in the putamen

(A)- Idiopathic dystonia ,genetically determined

(B)- Symptomatic dystonia

1-Drug induced (neuroleptics, metoclopromide) 2 -Postencephalitic

3-Toxic (CO ,cyanide ,methanol) 4-Tumours

5-Trauma 6 -Stroke

7-Post anoxic 8 -Sickle cell disease

(C)-Other genetically determined & metabolic disease e.g

*Dopa responsive dystonia *Paroxysmal dystonia

*Wilson s disease *Degenerative ataxias

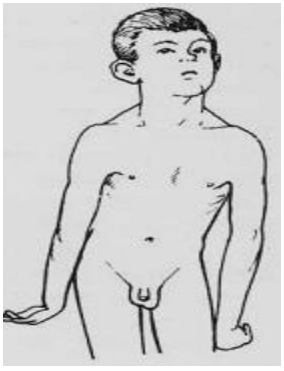
■Clinical picture of idiopathic torsion dystonia;(fig 91)

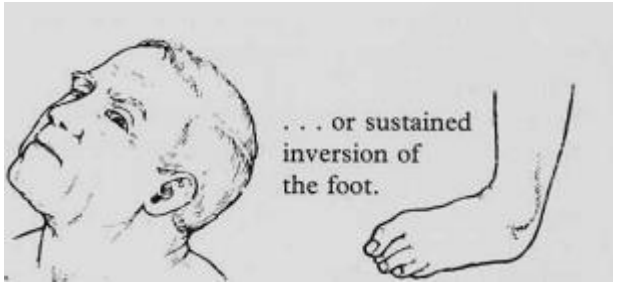
Dystonia may be focal ,segmental or generalized

1-Focal dystonia ;

.In adult life

- Axial dystonia in the trunk , in the neck→ spasmodic torticollis retrocollis, antecollis
- Blepharospasm (contraction of orbicularis oculi)
- Oromandibular dystonia, spasm of the jaw,mouth,tongue pharynx & larynx causing lip protrusion, jaw closure or opening , facial grimacing, dysphagia dysphonia
- Dystonic writers cramps affecting small muscles of the hand.





Fig(91)

2-Generalized dystonia; . Before the age of 20 years

. Dystonic features appear in the arms ,trunk, neck& legs causing walking difficulties.

3-Segmental dystonia;

-Has a wider range of age of onset

-Affect adjacent parts of the dody e.g arms & neck.

■Treatment lines of dystonia.

A) In generalized dystonia;

- Line 1→ all patients should receive 2-3 months trial of levodopa (250 mg tds) to exclude dopa responsive dystonia →on failure

-Line 2→ Anticholinergics up to 120 mg /day→ on failure

-Line 3→Benzodiazepines in large doses

→ Tetrabenazines, phenothiazines → on failure

-Line 4 → Unilateral or bilateral thalamotomy

B) In focal dystonia;

1- Anticholinergics, benzodiazepines

2-Local injection of botulinum toxin type (A) in the affected muscle, which will produce temporary paresis of the dystonic muscle for about 3 months, to be repeated , injection is done under EMG control

-This treatment has changed dramatically the management of focal dystonia in the last few years

Hemiballismus

-Severe ,static, involuntary , wide flinging movements of large amplitude due to lesions in the sub-thalamus ,usually cause skin abrasions ,macerations ,exhaustions, even bone fractures, most frequently in elderly

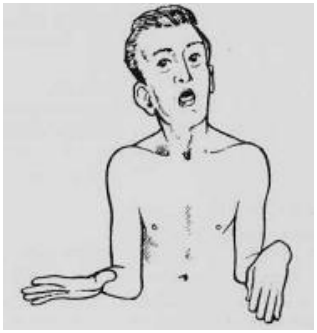
-Vascular causes e.g infarction or hemorrhage being the commonest causes

- Many patients may resolves into hemi chorea after Hemiballismus

- Good response is usually achieved with tetrabenazines

Athetosis

-Distal movements ,in frequency lies between chorea (more slow) & dystonia (more rapid) , it is static, involuntary, snake like movement, eg ;oriental dancing movement →



Fig(92)

- Usually accompanied clinically with chorea, choreo-athetosis.

- Treatment with anticholinergic & dopamine antagonists.

Tics

- Involuntary, repetitive ,stereotyped motor acts that resemble normal pattern of muscle contractions
- Common in childhood, common in hands ,neck & eye muscles
- **Motor tics** may be - simple as blinking ,head nodding, blepharospasm
  -complex(as copropraxia, echopraxia ,jumping, hitting)
- **Vocal tics** may be- simple as blowing ,sneezing , suckling, nasal clearing
  -complex as palilalia ,coprolalia ,echolalia

-Complex tics multiple motor & vocal ones ,with behavioral &learning problems(Gilles de la Tourette syndrome)

- ***Treatment***; Haloperidol can control the tics

Tremors

-Tremors are defined as a rhythmical , sinusoidal , oscillatory movements of a body part caused by regular ,rhythmical muscular contraction of both agonist & antagonist muscles

Types &causes;

(1)-Rest tremors ; tremors which are present only during rest ,disappear during movement

e.g parkinsonian tremors -Parkinson's disease
- Postencephalitic parkinsonism
-Drug induced parkinsonism

(2) -Postural tremors ;

which are present on maintaining postures e.g holding the arms out stretched, catching objects

1. Physiological tremors which may appear in states of nervousness,anxiety,fatigue, emotions,fevers
2. Thyrotoxicosis -fine rapid with other S& S of hyperthyroidism
3. Hypoglycemia
4. Drug induced- - tricyclic antidepressants, -sympathomimitics, - bronchodilators, - Na- valporate, - caffeine
5. Alcohol withdrawal

(3)-Essential tremors;

-Autosomal dominant trait present at any age, more in old age where it is termed senile tremors

-Frequency of tremors 5-8 /sec

-Associated with tremors of the jaw, head nodding, tongue tremors

(4)-toxic tremors ;

In renal , hepatic failure (flapping tremors-Asterexis)

(5)- Hysterical tremors;

Irregular ,of haphazard distribution ,varying in severity , frequency & location from time to time

(6)-Demyelinating neuropathies

(7)-Neurosyphilis;

(8)-kinetic tremors

which appear during limb movement ,if tremors increased at the end of movement on reaching the target they are termed *intention tremors*

Usually associated with brain stem or cerebellar diseases as cerebellar ataxia & multiple sclerosis

■Treatment;

-Deal with the cause

-In cases of anxiety tremors,→ B – blockers are of value

-In cases of essential , senile tremors

Higher doses of B- adrenergic antagonist "propranolol" are needed

start with 40 mg /day up to 300 mg, on failure or in presence of contraindications or with much side effects of B- blockers ,use primidone (in a dose up tp 500 mg /day)

-In cases of kinetic tremors with cerebellar ataxia or M.S

INH, tregretol & glutathimide are of value.

CNS Infections

- **Include:**
  - **Meningitis**
  - **Encephalitis**
  - **Local suppurations**
- Usually one syndrome predominant, but two of them may coexist in the same patient.
- **Common shared clinical features:**
  - Pyrexia
  - Headache
  - Drowsiness
  - Disturbed conscious level
  - Focal neurological deficits
  - Features of increased intracranial tension

Meningitis

- **Definition:** inflammation of the meningeal coverings of the brain and/or spinal cord,
- may be;

  -Acute – Subacute - Chronic
- **Causes:**
  - ***Inflammatory Infective eg***;
    - Bacterial "septic meningitis" >>> polymorphnuclear cells
    - Viral "aseptic meningitis" >>> lymphocytes are the predominant cells in CSF
    - Fungal, parasitic, TB,…
  - ***Inflammatory Non infective*** eg toxic,demyelinating
- **Pathogenesis**:
  - Route of infection
    - Blood stream >>> blood born organisms cross BBB.
    - Direct spread of infection

      .From local para-meningeal suppurations

      .Direct introduction; neurosurgery or trauma
- **Clinical Feature of meningitis:**

  *Age incidence:

  - Children...▸young adult…▸ elderly and immune suppressed

  * Clinical picture is common to all types, easy to be recognized in more than 80% of cases and includes:-

1) **General features:**

* Fever, malaise, vomiting, headache, photophobia,neck stiffness,…..
* **NB**. Absence of fever and or neck stiffness not exclude meningitis mainly in children, elderly, immunosuppressed patients and severe infection
* Headache is more common with viral meningitis
* Disturbances in consciousness is more common with bacterial meningitis
* TB, fungal and parasitic >>>> chronic presentation
* Viral and bacterial >>>>>>>> acute presentation

2) **Seizure:**

- Occurs in up to 40% of children & also in old patients
- Early evoked by fever and infection
- If prolonged ,CSF must be examined for encephalitis

3) **Systemic infection:**

Search for

- Chest infection -Septic arthritis
- Endocarditis -Skin rashes

4) **Focal signs**: (present in more than 15% of bacterial meningitis)

May be duo to :

- Arteritis of intracranial arteries >>> infarction
- Cerebral venous thrombosis >>> focal signs, seizures
- Inflammation of basal meninges >>> cranial nerve palsies
- Abscess formation

5) **Electrolyte disturbances:** More in children

The syndrome of inappropriate secretion of ADH >>> hyponatraemia

Viral Meningitis;

- Forms > 70% of acute aseptic meningitis
- More in children and young adults
- More in summer & autumn
- Usually benign and self limiting
- Commonest causative viurs ; Enterovirus (echo.., polio..), Arbovirus ,H. simplex ,Varicella zoster,Mumps and HIV

Clinical Features:

- Is Common to all viruses
- Common to find Signs of specific infections eg; mumps,parotitis…
- Sudden onset of fever, headache, neck stiffness

- URTI, nausea, irritability, myalgia, drowsiness
- Appearance of focal neurological deficit→ suggest Encephalitis
- **Prognosis & outcome :**
  - May recover spontaneously in few days, rarely weeks
  - Recovery is complete in most cases

NB * Headache is common *Consciousness is not impaired

Bacterial Meningitis;

- Common in children < 5 ys.
- Boys > girls
- **Transmission**:
  - Personal contact
  - Inhalation of airborne infected droplets
- Common **organisms in children**

  * E. coli * Streptococci * H. Influenza
- Common **organisms in adults**

  * Meningococci (commonest) * Pneumococci
  - Common **organisms in elderly:** * G –ve bacilli
- **Clinical picture**

  Common in closed communities (schools, military camps,…)
- **Incubation Period** < 10 days
- **Suggestive features of meningococcal meningitis.**
  - Typical rash appears on the trunk, legs, palms, soles, conjunctiva and mucous membrane ranging from purpura and extend to become bullous lesions
  - DIC (in severe cases)
  - Adrenal Hemorrhage may result from severe meningococcal septicemia & cause cardiovascular collapse
  - Myocarditis, pericarditis and arthritis

TB Meningitis:

- The incidence is decreasing due to :

1- Improvement in sanitation and social circumstances

2- Availability of BCG.

3- Immunization.

4- Development of anti T.B.drugs

NB * In the developing countries, the incidence is still high.

* Usually T.B. meningitis occurs secondary to T.B. infections else where, commonly pulmonary T.B. (Haematogenous spread)

- Infection is more severe in & around.

1- Basal meninges

2- Foraminae of the 4th ventricle

3- Meningeal arteries

4- Cerebral arteries

- Leading to the **complications of T.B.** meningitis as hydrocephalus, increased ICT, cranial nerve palsies, focal deficit

Clinical Features :

- Appears slowly over 3-4 weeks in adults and shorter in children
- Symptoms and signs are nonspecific:

1- Nausea and vomiting

2- Malaise and apathy

3- Anorexia, irritability, depression, confusion, disturbed behavior

- Headache & fever…..not common
- Signs of meningeal irritation, epileptic seziure, focal neurological deficits, Cranial nerve palsies, hydrocephalus, cerebeller tuberculoma, spinal TB.

Diagnosis and investigations of meningitis:

- **Clinical picture:**

Diagnosis of meningitis must be considered in any patient with :-

- Fever, malaise, headache, neck stiffness, irritability ± confusion.
- N.B. in up to 20% of cases this is not the role
- Once you suspect → start antibiotics

- **CSF:**

* CSF must be examined for,

- ↑ Pressure
- Aspect (cloudy or turbid)
- Cytology : Increase cells > 5 -100 cells/cubic ml lymphocytes ? viral / > 100 -1000cs/ml PNL or more ? bacterial,)
- Colour
- ↓ CSF glucose (<45 mg/ dl)
- ↑ CSF protein (30-40 mg/dl)

- **Blood culture and blood tests:**
  - In > 50% of cases, organism can be detected.
  - Increased ESR, blood electrolytes, CBC.
- **Brain imaging**

MRI is better than CT.

Differential Diagnosis:

1- Infective causes:

Encephalitis ,Brain abscess, Septicemia & Subdural empyema

2- Non-infective causes:

SAH, Drugs, Chemical irritation , Brain tumour, Collagen diseases eg;SLE

Treatment of meningitis:

- Treatment should be Started as soon as possible once the diagnosis has been considered
- Treatment of brain odema and increased ICT
  - Dexamethasone:- should be used early, even before antibiotics
    - dose: 0.15 mg / kg / 6 hourly IV for no more than 4 days
  - Mannitol 25% 1-2 gm / kg /day / to be divided 4-6 hourly
  - Hyperventillation,
  - Raising the patient head to an angle of 30° to improve venous drainage from the brain
- Fluid intake and serum electrolytes →.to be looked for & controlled
- Epileptic seizures (Phenytoin is preferred).
- Antibiotics and antiviral agents

  - Should be administered IVas early as possible.

  -3rd generation cephalosporins ± penicillins , ampicillin, vancomycin(in sever cases) should be used empirically before culture and in all age groups.

  # For viral infection :-usually self limited

  - Symptomatic treatment
  - Short course of penicillin. - Acyclovir I.V.

  # For T.B meningitis :

  Combine 4-5 anti T.B drugs (rifampicin, pyrazinamide, INH, Ethambutol & streptomycin) for at least 6 months

Encephalitis

-The term encephalitis means acute inflammatory changes affecting the brain parenchyma

-Viral infections are the commonest ,but protozoa ,bacteria ,fungi can also cause encephalitis

▲Viral encephalitis :

* usually occurs as a complication of systemic viral infection.

▲ Modes of transmission:

- Arthropode vector
- Faecal-oral route.
- Respiratory route.
- Direct contact
- Laboratory transmission

▲ Types of viral Encephalitis : (Regarding pathogenesis)

1- Acute viral encephalitis (infective as direct viral invasion)

2- Postinfectious encephalitis (Para infective as a remote effect)

3- Slow virus infections (Prion diseases)

- **The most common viruses** that cause encephalitis are, herpes simplex, herpes zoster, CMV, mumps, measles, HIV, Arbovirus, West-Nile virus)

▲ Clinical Features of Encephalitis:

- Common to all viruses
- Prodrome of nonspecific illness, may be present days before the onset (fever, malaise, fatigue, myalgia)
- Symptoms and signs occurs acutely within hours in the from of a triad of .

1- Fever, headache, and signs of meningeal irritation.

2- Disturbed conscious level.

3- Focal neurological signs and higher mental function affection (epileptic seizure, hemiparesis, sensory loss, cerebellar features, speech disturbances, hallucinations, memory loss, behavioural changes, stereotyped automatisms)

▲ Differential diagnosis:

- Meningitis
- Brain abscess
- Metastatic brain tumours
- Severe forms of MS
- CVA especially cerebral heamorrhage.
- Metabolic encephalopathy.

▲ **Investigations:**

- Virology (viral culture- of high cost)
- CSF examination:
  * Increased pressure.
  * Increased cells from 10 up to 1000 cells/mm^3 mostly lymphocyte
  * Increased protein content
  * Normal glucose

  N.B. CSF changes may be minimal or even within normal criteria
- EEG: diffuse slow waves, focal temporal changes in H.S encephalitis.
- Imaging … CT, MRI
  * MRI is superior to CT
  * MRI can demonstrate Brain odema, white matter disturbances, B.B.B disruptions & contrast enhancement
  * Changes are usually subtle or not evident in the first few days

▲Treatment:

- **General measures:**
  - Maintain nutrition, hydration and ventilation
  - Deal with epileptic seizures
  - Deal with increased ICP & brain edema >>>> mainly manitol except in severe fulminating cases>>>> dexamethasone
  - Neuroprotective agents
- **Antiviral drugs**

  Acyclovir
  - Reduce mortality rate below 70% in cases of HS encephalitis,
  - Also useful in varicella zoster

Brain edema

General consideration:

- **Definition.:** increased intracranial fluid content
- Associated with many brain pathologies
- Contributes to the resultant morbidity and mortality and plays a major role in prognosis of many brain conditions eg; head injuries, infarction, hemorrhage, tumors, encephalitis, meningitis, organic encephalopathy; lead encephalopathy, ketoacidosis, hydrocephalus…
- Can be measured now by MRI-diffusion-weighted imaging.

Classification of brain edema

- Vasogenic brain edema
- Cytotoxic brain edema
- Interstitial brain edema

I- Vasogenic brain edema "open barrier edema"

- Associated with increased capillary permeability and open B.B.B.(blood brain barrier)
- The most commonly edema seen in clinical practice
- Fluids accumulate in interstitial spaces
- Commonly seen with tumors, hemorrhage., infarction, abscess, contusions, meningitis
- Usually localized around the 1ry lesion
- Produce focal symptoms and signs that are often due to the edema rather than the primary lesion.
- Respond good to dexamethasone.

II- Cytotoxic brain edema "closed barrier edema"

Involves swelling of all the cellular elements of the brain with fluid e.g.neurons,glia,endothelial cells ,with reduction of extracellular fluid and intact B.B.B.

- Clinically encountered in conditions with diffuse brain pathology e.g; encephalitis, Diabetic ketoacidosis, haemodyalysis, hyponatremia.
- Symptoms & signs are more generalized: confusion, coma, drowsiness
- Respond more to osmotherapy as manitol ,furesemide.

III- Interstitial brain edema "hydrocephalic edema"

Seen in hydrocephalus

- Fluids accumulate in the periventricular area
- Seen in CT-Scan as periventricular hypodensity
- Respond well to osmotherapy and diuretics

Brain tumours

Brain tumours are space-occupying lesions

CLASSIFICATION:

1. Tumours arising from the meninges:

Meningiomas

2.Tumours arising from the brain tissue:

Gliomas (astrocytoma, glioblastoma multiformis, medulloblastoma).

3.Tumours of the blood vessels: * Haemangioma. * Hamangioblastoma.

4.Tumours of the cranial nerves:

Acoustic neuroma of the 8th nerve.

5. Pituitary tumours: * Suprasellar: craniopharyngioma.
* Intrasellar: acidophil, basophil and chromophobe adenomas.

6. **Secondary**: * Metastatic from lungs, breast, G.I.T., kidney & prostate.
* Invasion of the brain by a nasopharyngeal tumour.

7-Congenital tumours:

Teratoma, cholesteatoma.

Clinical picture:

Brain tumours manifest clinically by:

a) General symptoms and signs of increased intracranial tension (↑ ICT)

b) False localizing symptoms & signs regardless to the site of the tumour ,mostly due to increased I.C.T.

c) Specific symptoms and signs according to the site of the tumour (true localizing signs).

(1) General signs & symptoms of ↑ I.C.T.:

The increased I.C.T. is due to:

1. The tumour itself, which is a space-occupying lesion.
2. Obstruction of the venous return from the brain.
3. Obstruction of the C.S.F. drainage.
4. Hemorrhage into or degeneration of the tumour.

The increased I.C.T. presented with classical triad of:

I.Headache

1. It is due to stretch of the meninges by the space occupying lesion.
2. It is dull, aching, bursting in nature or throbbing.
3. Intermittent at first, more in the morning and is alleviated as the day passes, later become permanent.
4. It is exaggerated by conditions which increase the intracranial tension as coughing, straining, sneezing and stooping.
5. It has no significant value in the localization of the site of the tumour; however, generally, supratentorial tumours present with frontal headache, (which is of a late onset) while infra-tentorial tumours present with occipital headache (which is of an early onset and radiating to the neck),later become diffuse.

II. Vomiting

1. It is due to stimulation of the vomiting centre in the medulla.
2. It is more frequent in the morning.
3. It is not related to meals, and is not preceded by nausea, projectile.
4. It usually accompanies the headache & may temporarily relieve it.
5. It is more frequent in infra than in supra-tentorial tumours.

III. Papilloedema

1. This is a reliable sign of increased I.C.T.
2. The patient complains first of blurring of vision followed by gradual diminution of vision: failure of vision follows when post-papilloedemic optic atrophy occurs.
3. The field of vision shows peripheral concentric contraction.
4. Fundus examination .: there is:
   a. Congestion and tortuosity of the retinal veins.
   b. Haziness and blurring of the disc margins.
   c. Filling of the optic cup.
   d. Thinning and attenuation of the retinal arteries.
   e. Retinal haemorrhages and exudates.
   f. Post-papilloedemic optic atrophy may supervene.

(2) False localising signs:

These are signs which may be present regardless of the site of the tumour & are due to increased I.C.T.:

1. **Lateral ventricle dilatation:**

. Mental confusion.

2. **3rd ventricle dilatation:**

- Bitemporal hemianopia (compression of optic chiasma).
-Hypopituitarism (compression of pituitary gland).
-Hypothalamic manifestations (compression of hypothalamus).

3. **4th ventricle dilatation:**

-Vomiting (irritation of vomiting centre).
-Hypertension, bradycardia (irritation of vasomotor centre).

4. **Cranial nerve palsies**

. -Specially the 6th nerve as it has the longest intracranial pathway so, easy to be involved.

5. **Herniation syndromes**:

(a) Tentorial (uncal) herniation:

. -Supratentorial tumour leads to herniation of the uncus of temporal lobe into the tentorial hiatus resulting in:

* Compression of the reticular formation in the mid brain → impairment of consciousness.
* Compression of the 3rd nerve & its nucleus in the mid brain → dilated fixed pupil.

(b) Tonsillar herniation:

. -Subtentorial tumour leads to herniation of the cerebellar tonsils into the foramen magnum resulting in:

* Compression of the medulla → respiratory irregularities & impairment of consciousness.
* Tonsillar impaction in the foramen magnum→ neck stiffness & head tilt.

(3) True localizing signs:

These signs depend on the site of the tumour.

I) Frontal lobe tumours:

A. Destructive lesion:

one or more of the following:

1.Mentality changes: with tumours of prefrontal areas

2. Progressive contralateral hemiplegia.

3. Motor aphasia and agraphia (if in the dominant hemisphere)

4. Paralysis of conjugate movements of the eyes, to the opposite side.

5. Forced grasp reflex.

6. Anosmia with tumours of inferior surface.

B. Irritative lesion:

Focal or Jacksonian contralateral convulsions.

N.B.:

Tumours of the orbital surface of the frontal lobe near the optic nerves manifest by the **(Foster-Kennedy syndrome)** [ipsilateral primary optic atrophy & contralateral papillaedema]

II) Parietal lobe tumours;

A. Destructive lesions:

one or more of the following:

1. Contralateral cortical sensory loss.
2. Lower quadrantic homonymous hemianopia.
3. Apraxia, alexia, jargon's aphasia (if in the dominant hemisphere).

B. Irritative lesion:

Focal or Jacksonian contralateral sensory fits.

III) Temporal lobe tumours:

A. Destructive lesion:

1. Auditory agnosia.
2. Mentality changes.
3. Upper quadrantic homonymous hemianopia.
4. Contralateral motor weakness(in deep lesions)

B. Irritative lesion:

1. Uncinate fits.

2.Temporal lobe epilepsy.

IV) Occipital lobe tumours:

A. Destructive lesion:

1. Contralateral homonymous hemianopia.
2. Visual agnosia.

B. Irritative lesion:

Unformed visual hallucinations.

V) Pituitary tumours: They may present with;

A. Hormonal Manifestations:

- Chromophobe adenoma → Hypopituitarism.

-Acidophil adenoma → Gigantism or acromegaly.

- Basophil adenoma → Cushing's syndrome.

B. Neurological manifestations:

2ry to compression of neighbouring structures.

1. Anteriorly:

- Compression of the optic chiasma → bitemporal hemianopia.
- -Compression of the optic nerve → optic atrophy.
- Compression of the olfactory tract → anosmia.

2. Posteriorly:

- Compression of the upper brain stem → bilateral pyramidal signs and ophthalmoplegia.

3. Laterally:

- Compression of the optic tract → homonymous hemianopia.
- Compression of the cavernous sinus → 3rd ,4th & 6th cranial nerve paralysis and loss of sensations over the area of the face supplied by the ophthalmic division of the 5th nerve.

4. Superiorly:

- Compression of the hypothalamus → hypothalamic manifestation e.g. Diabetes insipidus, hypersomnia, adiposity or autonomic epilepsy.

C. Headache:

Typically it passes through 3 phases:

- It starts bitemporal due to increased intrasellar pressure.
- Then it disappears due to rupture of the sella turcica.
- Lately it reappears & is generalised with the increased I.C.T.

VI) Cerebello-pontine angle tumours

(C.P.A. tumour): They include:

1. Neurofibroma (acoustic neuroma) of the 8th cranial nerve.
2. Less commonly: • Meningioma. • Cholesteatoma. • Arachnoid cyst.

Clinical Picture of C.P.A. tumour:

1. Ipsilateral cerebellar ataxia.
2. Ipsilateral affection of the 8th, 7^{th} & 5^{th} nerves running in the CPA

→ progressive deafness, facial palsy, facial pain & weakness of the muscles of mastication.

3. Contralateral hemiparesis (pyramidal compression in pons).

N.B.: The neurofibroma of the 8th nerve may be:

- Solitary. associated with subcutaneous fibromas along peripheral nerves, cutaneous fibroma (mollusca fibrosa) & café- au- lait patches in,

Von Recklinghausen's disease (neurofibromatosis type 1).

Investigation of a case with brain tumour:

1) Proper history taking and clinical examination.

2) Examination of visual field.

3) Fundus examination for papilloedema.

4) Local examination of the skull for ▸:

a) Enlargement.

b) Dilated tortuous veins.

c) Bony bosses.

d) Angiomatous malformations.

e) McEwen's sign: cracked pot note on percussion.

5) Plain x-ray of the skull

which, might show:

▸ a) Features related to ↑ ICT.

1- Separation of the cranial sutures.

2- Beaten-silver appearance or finger prints.

3- Sellar changes:

- Enlargement of the sella turcica.
- Rarifaction and destruction of the dorsum sellae and the posterior clinoids.
- Encroachment on the sphenoid air-sinus

- b) Lateralising signs denoting the side of the tumour:

Shift of calcified pineal body or falx cerebri.

- c) Signs denoting the site of the tumour:

1.Localised calcification.

2.Localised erosion and destruction of the skull bones.

6) C.T. scan: Fig;(93 a,b)

(computerised tomography of the brain):

It shows:

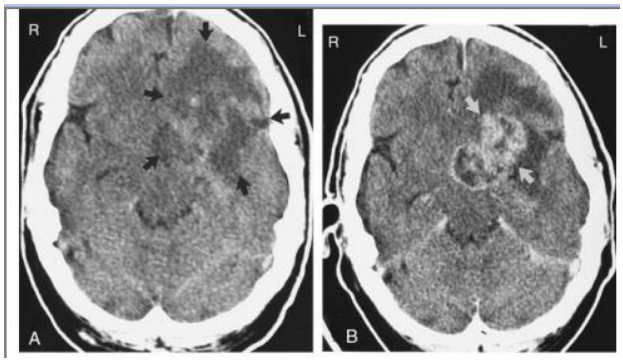
A. The site and size of the tumour, its density and any cystic degeneration or calcification.

B. The size of the cerebral ventricles.

C. Any midline shift.

a-shows only a large area of low density that represents the tumor and edema (arrows).

b-A contrasted CT scan (B) shows enhancement of the tumor (arrows) surrounded by the dark or low-density area of edema.



a ← Fig;(93) → b

11) M.R.I.: (Magnetic resonance imaging):

This is one of the most recent methods of investigation. Tumours can be diagnosed not only anatomically but also pathologically.

Treatment:

1.Symptomatic For increased I.C.T., fits.

2.Surgical if accessible.

3.Palliative Radiotherapy, chemotherapy if not accessible.

Dementia

Definition:

Cognitive impairment of severity sufficient to interfere with work , social & behavioural activities of daily living with normal arousal.

Cognitive impairment include memory loss, plus one or more of the following cognitive deficits:

1.Apraxia
2.Agnosia
3.Aphasia
4.Impairment of judgment
5.Impairment of abstract thinking

Some important considerations:

■ Dementia is characterized by progressive decline of all cognitive functions including memory resulting in decline in work performance and daily activities over the course of many years.

■ Dementia is summation of many cognitive deficits resulting from progressive cerebral disease.

■ It is called → **presenile dementia** if occured in people under age of 65 years & → **senile dementia** in older people.

■ Approximately 70% of demented people have Alzheimer disease which is considerd the major clinical consideration.

■ There are different causes in which the brain is affected in different ways but the final expression is widespread neuronal failure and dementia.

■ The nature & severity of deficits in cognition and memory differ markedly according to the site of pathology e.g profound amnesia & disorientation may result from circumscribed lesions in the thalamus,

■ language disturbance has been recognized following discrete dominant hemispheric lesions, visual agnosia may result from lesions in visual association cortex.

■ Since dementia is at least the sum of a number of specific cognitive deficits, the features of dementia may differ qualitatively and quantitatively according to the site and severity of pathology. Furthermore, a diffuse pathology is not essential for the production of widespread cognitive impairment, where small lesions in selective critical brain regions may produce cognitive deficits similar to that of extensive pathology. Consequantly, the features of dementia will differ according to the aetiology& rate of advance of the responsible cause.

Causes of dementia:

1) Degenerative diseases:

-Alzheimer disease -Progressive supranuclear palsy

-Pick's disease -Parkinson's disease

-Huntington's disease -Multisystem atrophy

-Progressive myoclonic epilepsy -Leucodystrophy

2) Trauma:

-Open & closed head injuries -Boxer's dementia

-Subdural heamatoma

3) Metabolic & Endocrinal disorders:

-Hypothyroidism -Wilson's disease

-Hypopituitarism -Renal failure, Dialysis dementia

-Crushing disease -Hepatic failure

4) Difficiency disorders :

-Thiamine difficiency (korsakoff syndrome)

-B 12 & Folic acid deficiency

-Pellagra

5) Anoxia :

-Hypoperfusion state

6) Vascular disease:

-Multi-infarct dementia -Vasculitis

-Bins wanger's encephalopathy

7) Infection :

-Cerebral abscess -Encephalitis

-Creutzfield jakob encephalopathy -AIDS encephalopathy

8) Neoplasm :

Brain tumours (frontal, temporal, thalamic, corpus callosum)

9) Toxins :

-Alchol -Heavy metals

-CO -Organic solvent

-Drugs(barbiturates, amphetamines, bromides)

10) Others :

-Communicating or obstructive hydrocephalus

-SLE, Behcet's disease,Sarcoidosis

-D.S

Clinical features:

Major features of dementia of any cause:

1. Memory impairment
2. Deterioration of attention, language & abstract thinking.
3. Change in personality, behaviour & mood.
4. Focal neurological symptoms & signs.

Features of subcortical dementia :e.g (Parkinson's disease,Huntington's disease.).

Mainly affects some specific patterns of cognitive impairment:

a) Poor recall of memories (retrograde).
b) Slowness of cognition
c) Prominent personality disturbances
d) Depression
e) Motor deficits

Features of cortical dementia: e,g Alzheimer disease

a) Prominent dysphasic errors.
b) Apraxia, agnosia, acalculia.
c) Poor registration of memories (immediate).
d) Less disturbances of mood, personality & motor control

Alzheimer disease:

*Pathology:

- Diffuse cortical atrophy with neuronal loss.
- Enlargement of ventricles and sulci.
- Large numbers of senile plaques, granulovascular degeneration.
- These changes are maximal at frontal, temporal and hippocampal cortex.
- The number of characteristic plaques per unit volume of cerebral cortex correlates with severity of dementia.

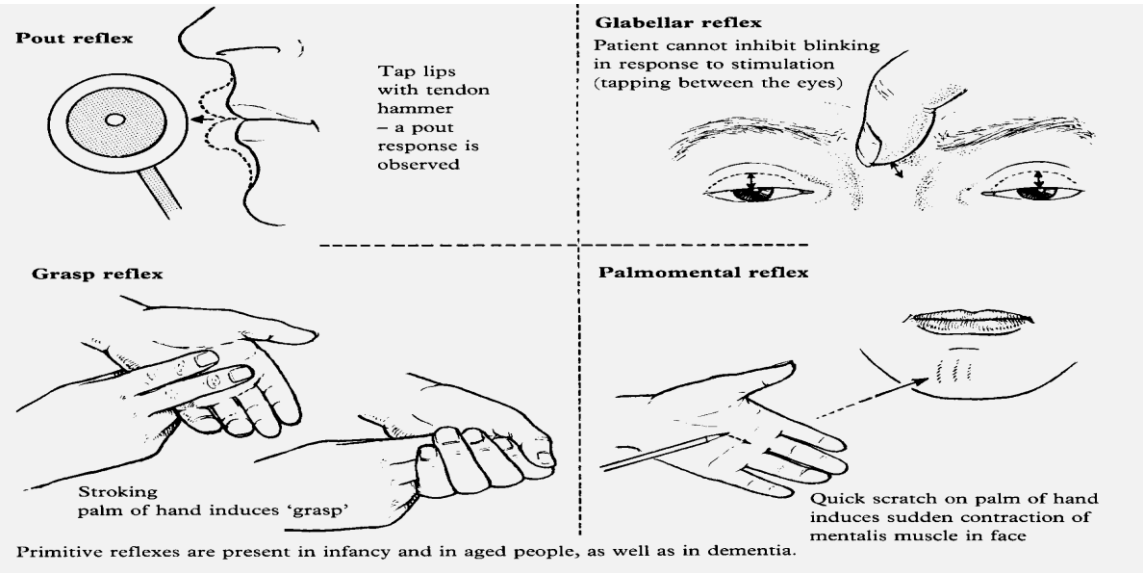
Clinical picture:

1) Immediate memory impairment with ability to recall remote events.
2) Anomia (language impairment affect the use of meaning or concept).
3) Impairment of comprehension of speech & writing.
4) Visuospatial deficits.
5) Aphasia (receptive, expressive or transcortical).
6) Apraxia of face, tongue, limbs and that of dressing.
7) Rigidity, akinesia in 60-80% of patients.
8) Gait disturbances & postural instability.
9) Myoclonus & dyskinesia in 87% of patients.

10) GTC seizures in 10-20% of patients.

11) Oculomotor dysfunction in the form of delayed saccadic initiation.

12) Presence of primitive reflexes e.g pulmomental reflex, glapellar tap reflex, suckling reflex and Tom's grasp reflex Fig(94)



Fig(94)

N.B: in some cases, the disease may presents with focal onset(e.g aphasia) which may preceeds intellectual deterioration by 2 years.

Multi-infarct dementia:

-It's considered the second most common cause of dementia in elderly after Alzheimer disease.

- Affect males more than females.

***Pathology:**

-There are multiple infarctions in large vessels territories.

-The extent of cognitive dysfunctions depends upon site, size & severity of lesions.

-There is a loss of 50-100 ml of cerebral volume , this is considered certain to produce dementia.

***Clinical Picture :**

1) Recurrent defined acute strokes or TIAS.

2) Predominance of focal neurological signs.

3) Dysphagia, dysphonia & emotional incontinence.

4) Bilateral pyramidal signs.

5) Stepwise progression of dementia.

N.B: control of cerebrovascular risk factors may improve cognitive performance, decrease progression & severity of dementia.

Investigation of dementia:

(A) Laboratory:

1) Blood count & films : for anaemia, macrocytosis of vitamin B12 or folic acid defiency.
2) ESR : for connective tissue disorders.
3) Chest radiography: for cardiac lesions, bronchial neoplasm.
4) Biochemical tests: for liver & renal failure.
5) Serology for $ (neurosyphilis)
6) Thyroid function tests: for hypothyroidism.

(B) Imaging:

1) Skull x-ray: -Pineal body shift
   -Calcified mass
   - Erosion of bone in space occupying lesions .
2) C.T & MRI brain: -Cortical atrophy
   -Focal lesions
   -Infarctions
   -Tumours
3) PET (positron emission tomography): areas of hypo metabolism

(C) EEG:

- Generalized slowing in primary cerebral atrophy & metabolic causes.
- Focal changes in S.O.L.

Treatement

Control of vascular risk factors as DM.IHD,HTN
Healthy life style
Healhy balanced diet
Neuroprotectives
Drug therapy eg Aricept,Ibexa,

Demyelinating diseases

Myelination in CNS is mediated by oligodendrocytes which give multiple axons ,so damage to these cells is not followed by regeneration in contrast to peripheral nervous system in which each axon is covered by its own Schwann cell so ,it's liable for regeneration

Demylinating diseases are diseases in which the structure and function of myelin axons are distorted mostly due to inflammatory and autoimmune disorder. In Multiple Sclerosis (MS) ; demyelination is the usual explanation of these disorders with episodic neurological symptoms and signs

Demyelinating diseases ;either inflammatory {usually episodic }
or noninflammatory {usually progressive}.

Demyelination occurs throughout the CNS {especially white matter },**optic nerves** ,**brain stem** ,**spinal cord** and **cerebellum**

***Classification of demyelinating diseases*:**

A-Isolated Demyelinating syndrome:

1. Optic neuritis
2. Acute dissiminated encephalomyelitis
3. Transverse myelitis
4. Chronic progressive myelopathy

B-Multiple Sclerosis(MS) :

1. Relapsing remitting
2. 1ry Progressive
3. 2ry Progressive
4. Devic's disease (variant of MS)

C-Leucodystrophies (non inflammatory demyelination):

Characterized by progressive neurological disorders usually in childhood or young patient, arising from genetically determined disorders of myelin formation {abnormal myelin} , the most common leucodystrophies are ;

-Metachromatic leucodystrophies

-Adrenoleucodystrophies (Krabb's disease.)

Multiple sclerosis:

Multiple sclerosis is the commonest demyelinating disease

Aetiology ;

A- Genetic factors;

The most accepted theory is genetic one which explain why MS is common in some families than others

B- **Environmental factors;where M**S occur in certain areas,more than others,

-Areas of high risk.

Northen Europe ,northen USA ,Canada ,southern Australia and Newzland

-Areas of medium risk.

Southern Europe southern US nourthen Australia

-Areas of low risk .

Asia ,Africa

Also reported that MS is common in white persons more than blacks

-Male :female ratio the same

Pathology.

Demyelinating diseases are characterized by inflammatory cell infiltration with focal and multifocal lesions .

The hallmark of demyelinating diseases in case of multiple sclerosis is the formation of **sclerotic plaques** these represent the end stage of a process which includes :

1. Demyelination
2. Remyelination
3. oligodentrocyte depletion, astrocytosis and
4. Axon degeneration

Sites of plaques.

.These sclerotic plaques are clustered around the lateral ventricles , corpus callosum , 4th ventricle , cortical and subcortical white matter, optic nerves, brain stem and cervical portion of spinal cord

Clinical features;

The C/P usually reflects the term of multiple sclerosis or disseminated sclerosis which mean that, multiple areas through the CNS are affected that anatomically discrete, on the other hands these lesions in different areas of the CNS ,in about 80% of MS ,these lesions differ in time {episodic } and differ in site 'anatomically not related e.g optic nerve ,spinal cord ,brain stem.

The first attack comes without warning and may be mono-or poly-symptomatic .in 1/5 of cases the onset is acute i.e the deficit attain its maximum severity in minutes or hours

i) Motor; weaknes, spasticity, Seizures

ii) Sensory; Numbness,pain

iii) Cerebellar; Ataxia ,nystagmus

iii)Sphincteric ; Urgency hesitancy incontinence

iv)Brain stem &cranial nerves; optic nerve affection → blurring of vision ,diminution of vision up to monocular blindness(optic atrophy)

-Facial weakness - Internuclear ophthalmoplegia - Dysartheria ,deafness,vertigo diplopia

- Trigeminal neuralgia

v) Mental changes; Learning disabilities , -Dementia

vi) Others; Depression , -Fatigue .

Course;

■ 80% of patients with MS are presentrd with episodic neurological symptoms {relapsing remitting}either multifocal or anatomically discrete which reflect the CNS areas mentioned before {optic nerve, brain stem ,spinal cord }and the symptoms recover completely.

■ Further episodes occur at random frequency and on unpredictable period seldom exceeding one year

■ With time, recovery from each episode is incomplete, and persisting symptoms begin to occur, finally the patient become completely handicapped

■ In 20% of patients the disease is progressive from the start

Diagnosis and differential diagnosis :

Once theres is evidence of multiple CNS lesions with remitting & relapsing symptoms over a period of time ,with exclusion of other causes (DD);

- Metastatic tumours
- Cerebral arteritis (Behcet's disease ,SLE)
- CNS infection

A single lesion causing recurrent symptoms must be regarded with suspicion (may be due to MS or other causes);

-Vascular malformations of brain stem

- Chiari malformation

- Tumours of the foramen magnum ,clivus,or cerebellopontine angle

***Investigations*:**

1-CSF

-Abnormal in 80 % of cases

-Mild mononuclear pleocytosis

-Modest increase in total protein (great increase in gamma globulin fraction)

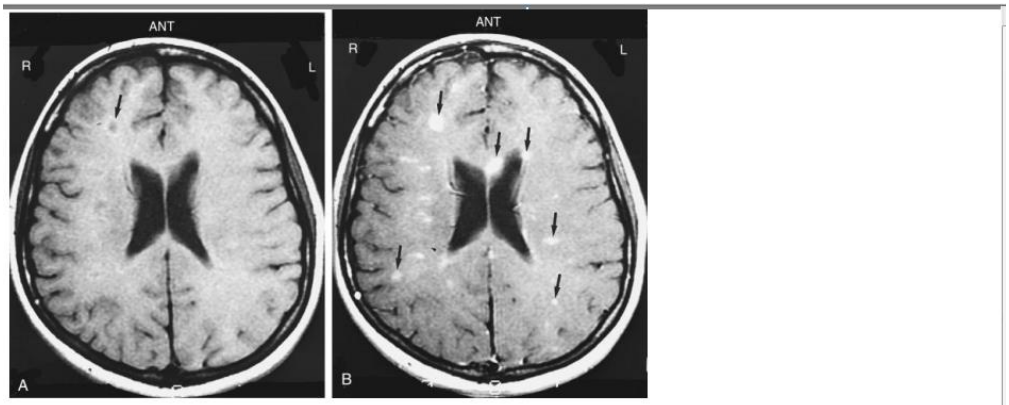
-More specifically the presence of oligoclonal band (IgG band) by electrophoresis

2-Evoked potentials.

Visual, Auditory and Somatosensory evoked potentials for not clinically manifest lesions

3-MRI(T2 weighted) Fig(95)

Multiple ,periventricular lesions

Fig(95 

Multiple sclerosis. The noncontrasted T1–weighted magnetic resonance imaging (MRI) scan (A) is generally unremarkable, with the exception of one the right frontal lobe (arrow). A gadolinium-enhanced scan (B) is much better and shows many enhancing lesions, only some of which are indicated by the arrows.)

4-CT; Hypodense lesions

***Treatment*:**

A) Treatment of acute episode &relapse:

- **Corticosteroids,** Over a period of weeks

IV methylprednisolone 500 mg daily for 5 days,or 1gm for 3 days in patient with acute symptomatic deterioration,to be followed by IV pulse dose once/month,or for 3 days/2 month

- **Immunmodulating therapy: interferon**

Interferon beta-1a 30 micro gm IM/week

Interferon beta-1a 22-44 micro gm SC every other day

Interferon beta 1-a 8 million Iu SC every other day

-Lessen the frequency of attacks in relapsing remitting cases

-Side effects flue like symptoms,leucopenia

- **Immunosuppressive therapy:** Azathioprine, cyclophosphamide given for a period of years
- **Combined therapy** both interferon & immunosuppressive therapies.
- **Other therapies;IV IG.Plasma exchange.Monoclonal antibodies**

B) Symptomatic treatment :

Spasticity; Baclofen dantrolene ,diazepam

Dizziness & nystagmus; Cinnarizine

Fatigue; Amantadine,

Sexual; Yohimbine

Pain; Anticonvulsant(carbmazepine, gapabentin)

Neuromyelitis optica (Devic's disease)

Attacks of , optic neuritis , progressive paraparesis of pyramidal tract features.

-May be a form of MS,

-Treatment

- steroids (poor response)

COMA

Definition:

Coma is a state of loss of consciousness from which the patient can not be aroused by any stimulation whatever vigorous & painful.

Pathophysiology:

■Consciousness consists of two components-

(1)-Awareness (content of consciousness).

Depend on the activities of the cerebral cortex & thalamus, permitting higher level of integration of sensory inputs reaching the cortex leading to reasonable understanding of self & environment.

(2)-Arousal (state of consciousness).

Which is a primitive one depending on the activities of the central reticular formation of the brain stem.

■Thus lesion of the cortex alone affect the content of consciousness (awareness) e.g {emotions, sensations, memories, ideas, experience},without changing the state of consciousness.

■Therefore coma can be produced by :-

1-bilateral or diffuse brain damage.

2-brain stem failure or damage

3-combined cortical & brain stem failure.

Aetiology:

Either → **organic** or **hysterical**

■Organic causes include→ **cranial** and **extra cranial** causes

(1)-Cranial causes:

*** Trauma:** leading to intracranial hge .

***Cerebrovascular disease:** can produce coma directly or indirectly by interfering with the functions of the reticular activating system .

either through -Increase ICP

-Direct pressure of haematoma or edema

-Impairment of blood supply , e.g

- Massive subarachnoid hge.
- Large intracerebral or intracerebellar hge
- Large cerebral infarction
- Brain stem hge or infarction
- Hypertensive encephalopathy

*Space occupying lesions: as tumors & abscess

NB. Haemorrhge into the tumor , sudden development of edema or cerebellar herniation →coma can occur

*Inflammatory causes:

Meningitis or encephalitis

*Other causes:

-Post icteal Todd's paralysis.

-Leucodystrophy , M.S

-Central pontine myelinolysis

(2)-Extra cranial causes:

■ -A-Metabolic causes:

*Uraemic coma:

.In acute or Ch.R.F

.due to -Metabolic acidosis

-Raised blood urea

-Electrolyte disturbance

-water intoxication

-Raised blood pyruvate : → decrease cerebral oxygen consumption

.Cerebral disequilibrium syndrome:

Commoner in children & young adults, characterized by temporary impairment of consc.

Due to rapid change of electrolyte

*Hepatic coma:

- Either spontaneously in -Acute viral hepatitis

-Eclampsia

-Acute chemical poisoning

- Chronic hepatic Patienst with - Cirrhosis

-Hemochromatosis

-Wilson's disease

-.*Hepatic coma* occurs usually due to:

-Increase in blood ammonia→ toxic to the brain

-Electrolyte disturbances

-Increase of circulatory Amino.Acids

- Cerebral depletion of dopamine

-Increase of false neurotransmitters,as octopamine

*Diabetic comas:

Either –Diabetic ketotic coma

- Lactic acidotic coma

- Hyperosmolar non ketotic hyperglyceamic coma

*Hypoglycemic coma:

-Diabetic patients

-Hypopituitarism

-Alcoholism

-Hepatic patients

-Addison's disease

*Hyperthermia (Heat stroke):

-After prolonged exertion in hot surroundings

-Tetanus , -Pontine hge, -Intraventricular.hge

-Malignant hyperpyrexia as a complication of general anaesthesia.

-Malignant neuroleptic syndrome as a complication of anti psychotics

* Hypothermia:

- Prolonged exposure in cold surroundings

- Hypopituitarism. - Older patients with arthritis and parkinsonism.

- Drugs as chlorpromazine

- Shapiro's syndrome.(Recurrent hypothermia with polyuria, polydepsia, hyponatremia, and agenesis of corpus callosum).

*Hypo and hypernatremia *Hypo and hyperkalaemia:

*Hypo and hypercalcemia *Acidosis or alkalosis:

*Porphyrias *Pancreatic encephalopathy→ as in chronic relapsing or acute pancreatitis.

■-B- Respiratory causes:

*Hypoventillation in COPD leading to anoxia.

*Diffusion or perfusion defects.

■-C- Cardiovascular causes leading to cerebral ischemia:

* Cardiac arrest. * Severe anaemia.

* Severe hypotension * Myocardial infarction. * Valvular heart diseases.

* Complications of open heart surgery.

■-D- Endocrinal causes:

* Hypopituitarism. * Adrenal crisis.

* Hypo and hyper parathyroidism. * Hypothyroidism.

■-E- Toxins:

* CO_2 poisoning. * CO poisoning. * Heavy metals. *Alcohol.

■-F- Drugs:

* Sedatives and hypnotics. * Anticonvulsants.

* Antidepressants. * Anaesthetic agents.

▸Grades of impairment of consciousness:-

1) **Coma:** previously defined.
2) **Semi coma:** complete loss of consciousness with response only at reflex level.
3) **Stupor:** state of impairment of consciousness from which the patient can be aroused by vigorous stimuli.
4) **Lethargy:** state of drowsiness and indifference in which increased stimulation may be needed to arouse the patient.
5) **Confusion:** impairment of consciousness with poverty of mental process.
6) **Delerium:** confusion with excitement.

▸Now the **Glasgow Coma Scale** replaced the old classification and has great immediate prognostic value and assessed as following :Table (5)

Eye-opening	*Score*	*Verbal response*	*score*	*Motor response*	*score*
Spontaneously (with blinking)	4	Orientated	5	Voluntary	6
To voice	3	Disorientated	4	Localizing	5
To pain	2	Words	3	Withdrawal	4
Nil	1	Sounds	2	Flexion	3
		Nil	1	Extension	2
				Nil	1

Diagnostic approach for a case of coma:

A- **History: for trauma, medications, previous illness.**

B- **General physical examination:**

1) Vital signs
2) Skin: signs of trauma, needle marks in addicts, jaundice, cyanosis, signs of liver disease, severe pallor, myxodema, cherry red color in CO poisoning.
3) Head: signs of trauma, enlarged head with tense anterior fontanelle in children means increased ICP.

4) Battle sign: hematoma and odema overlying mastoid bone indicating fracture of the petrous part of temporal bone.
5) Raccon's eyes: bilateral orbital hematoma and tenderness indicating bilateral orbital fracture.
6) Hemorrhage from nose or ear means fracture base of skull.
7) Neck stiffness: meningitis, SAH, tonsillar herniation
8) Tongue bitting: epileptic, Todd's paralysis.

C- Chest and abdomen:

1) Odour of breath: fetour hepaticus in colemia, acetone odour in diabetics, uriniferous odour in uremia.
2) Chyne-stock breathing
3) Ascites, hepatomegaly, shrunken liver, splenomegaly

D- Heart:

1) Myocardial infarction
2) Subacute bacterial endocarditis. 3)Valvular heart diseases

E- Extremities, pelvic and rectal examination:

F- Neurological examination:

1) **Signs of increased ICP as papillodema and tense anterior fontanelle**.
2) **Visual field and pupils:**
   - We can examine visual field in unresponsive patients by threatening movements which should evoke a blink, asymmetry of the blink response indicate hemianopia.
   - Mid position and unreactive pupils mostly evidence of midbrain affection.
   - Unilateral dilated fixed pupil means third nerve compression.
   - Pin point pupil means opiate, organophosphorous poisoning, pontine heamorrhage, or parasympathomimitics toxicity.
3) **Occular movements:**
   - Doll's head maneuver, means lesion of the brainstem or barbiturate poisoning.
   - Conjugate deviation of the eyes opposite to the paralyzed side indicate cerebral damage.
   - Conjugate deviation towards the paralysis means brainstem damage.
   - Unequal pupil with divergent squint and irregular jerky movemnts suggests heamorrhage into corpus striatum or internal capsule.
   - Paralysis of vertical movement with downward displacement means thalamic or subthalamic heamorrhage.

4) Motor system:

- Seizures ---- focal or generalized.
- Myoclonic jerks, asterexis mostly metabolic causes.
- Asymmetry of movements, asymmetry of tone and asymmetry of Babineski means lateralizing signs with cerebral lesions.
- Reflex movements, decerebrate, decorticate, abduction of a limb, usually indicate good prognostic value if the cause of coma is released.

Investigations:

* Trauma →signs of ↑ ICT → plain X ray ,CT,MRI brain
* Focal neurological signs→ CT,MRI brain
* Meningitis , encephalitis→ Lumbar puncture

- No signs of ↑ ICT→ metabolic screen
- No signs of meningitis→ metabolic screen
- No focal neurological signs→ metabolic screen
- Urea & creatinine - Electrolytes
- Blood gases ,ph - Blood sugar
- Liver function tests - Blood culture - Drug screen

→→ →→ **if not diagnostic**

- Serum ca & phosphate & Mg - Folic acid &VIT B12
- Serum amylase & cortisol - Porphyrins & thyroid function tests

Key points with some types of coma:

1)-Hysterical coma. -Hysterical personality -Rare to get deep coma
-Resistance to passive movements -Preceeded by psychic trauma
-Absence of signs of lateralization
-Any attempt to elicit corneal reflex →vigorous contraction of orbicularis occuli
While in organic coma →corneal reflex is lost
-Vigorous ocular movements on raising the eye lid ,

2)- Uremic coma . -Early headache ,vomiting, insomnia,dypsnea,restlessness,confusion
-Patient with history of renal troubles
-Earthy look face -Oedema with puffy eyelids -Uremic frost, uriniferous odour -Flapping tremors -Uremic lungs, uremic pericarditis -Coma preceeded by muscle twitches, convulsions -Absence of focal neurological deficits,or cranial nerve affection ,(**rare to get focal deficit)** -lab. investigation→ ↑ blood urea & creatinine

3)-Cerebral coma.

▸ ***In hypertensive patients***;

-With → negligect of ttt, mental stressors, physical stress

- Acute onset of severe headache ,vomiting, blurring of vision ,convulsion

→Coma with focal neurological deficits ,cranial nerve affection,±neck stiffness

→Diagnostic CT ,MRI

▸***Also in cardiac patient***s ;

→Sudden onset of impairement of consciousness ,with focal neurological deficit,cranial nerve palsies

▸***In atherosclerotic or diabetic old patient;***

→Gradual development of focal neurological deficit over hours , in a stepwise manner (stuttering stroke)with sometimes past history of TIA

→diagnostic CT,MRI

4)- Hepatic coma; → Hepatic pts , either acute or chronic

→ In chronic patient →there is ascites, shrunken liver, splenomegally, spider naevi, divercation of recti, oedema of LL, feminine distribution of hair, hematemesis, melena, jaundice

→ Coma preceeded by personality changes, change in mood & behavior, flapping tremors presence of ppt factor, and absence of focal neurological signs

5)- Drug induced coma; →History of medications →Anti depressants

→Anti convulsants

→Drug abuse,sedatives ,hypnotics

→lost eye movements with preservation of pupillary reflex,where most of these drugs act by depressing the vestibuloccular reflex

→Alcoholic coma →→flushed face ,rapid pulse ,low BP,congested conjuctiva, dilated pupil,odour of alcohol

6)- Hypoglycemic coma; Occur in diabetic pts with over dose of insulin or oral hypoglycemics, or other ppt factors ***clinically*** → muscle twitches ,sweating,dilated pupil ,? convulsion,hypothermia,confusion,disturbed behaviour→ if prolonged → irreversible brain damage

7)-Diabetic coma ;→ In diabetic patients → with negligence of ttt, & other factors

-Pale skin , dehydrated tongue & axilla, rapid weak pulse, decreased BP, ocular tension is low, odeur of acetone , reflexes may be lost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