

(area 4, pyramidal tract) : is the upper motor neuron system * ∴
(ant. horn cell, Nerve, muscle) : " lower " " " * ∴

Paralysis to Muscles Below level of lesion? $\Rightarrow$ paralysis deno LMN $\Rightarrow$ لو خويبت
" at " " ? " " LMN " "

* اي شئام ماساعة دة لا مبيعات لوظائفه disuse atrophy ؟

ازلاء جسمي جازلا مع عضلاته وانما ناسية ؟

← Muscle Tone (يقي' العضلات Contract)

نة الـ هو حفظ عليها ، ازلاء - ريو ؟

هنا لو اخذنا biceps وقسمنا الطول بطول [20 cm] ، الـ نسبة بين الـ origin

بنا والـ insertion بنا [25 cm] ، يعني هـ يتكون Stretched

فنجين الـ stretched fibers هتجـ دائرة الـ Dorsal root ganglion ← نقوم

باسة impulses الـ post. horn cells ودي هتجـ الـ ant. horn cell

دي يعني هتجـ الـ muscle fibers هـ impulse عتـ Contract

يعني حق وانما مش مستقيمة العضلة ، الـ muscle Contracting

هـ خذ باله ! يعني الـ fibers لو كلها كـ زيالك تتنقب وانما تـ

المكونة الـ m. tone

* لو استقامت الخلية الانما ممتاحة فـ m. tone في حالة

يقي طـ الـ Pyramidal tract هـ activation للعضلة ، هـ

در Tone مع جنب ملوش دقوة فـ فيقوم عامله : inhibition

يقي لو قـ Pyramidal T. هـ الـ ؟ (hypertonia)

pyramidal Tract → UMN → hypertonia below the level → m. wasting

LMN → hypotonia at the level → m. wasting

* لانجـ تقوى بالهـ الـ hne مش ريو ؟ stretch all fiber فـ الـ fibers

تجـ الـ dorsal ganglion ؟ Post. horn ؟ ant. horn فـ فـ الـ الـ

(Deep reflex الـ الـ) Contract

* الـ deep relax سـ الـ m. Tone الـ الـ

Some fibers

spontaneous

all fibers

induced

• UMN → hyperreflexia

• LMN → hyporeflexia

• Sp (pathological reflex) : Sp St St ✓

Tow reflex, Patellar reflex, adductor nl

حیبت = adductor tendon، ایسی = ischio

Clinical Significance of Contraction of adduc-m. of stretch test ← (in Normal)

Trifurcation $\Rightarrow$ (in LMNL)

Contraction of the Thigh muscle (in UNNL) ←

✓ یقیناً ۱) Pathology = So = ۲) UMNL

(abdominal reflex, cremasteric reflex) ← (superficial reflex) ✓

دی۔ بقی تختی مع UMANL, LMNL, طبع UMANL وعودہ کل شیہ hyper

للإفصاح مفسد لغفوه ...

extrapyramidal system : نظام خارجي motor system (pyramidal) *

وہمیشہ تفسیر کا حوالہ عطا کرے اور Touch تابع اور emotion، عین :

Contract - α & β cells $\leftarrow$ Pyramidal Tract

extrapyramidal ← "ما خارج" "تبعي اذا انا متعلقة / مبرطة"

* الـ pyramidel يعبر Voluntary

involuntary? - extrapyramidal: i.e.

عنه وجهه بابه اغنى منكم ب انا بالحق مباله

Pyramidal : لقولنا بكه باقية رحمة الله " جامعة " دة لله , قول

extrapyram : lip oli Ssp Evlp // // iS

5. extra pyram , pyramidal cell is not x

(Cerebellum) وظيفته حفظ البناء وتنسيق الحركات وتنظيمها

المغني لويسين ولسي الكونابة 20 cm المخفي هسقول للمخ لـ

فتيوم الخ محرك ايدى 20 cm

"Motor neuron disease"

- ✓ it's a degenerative systemic dz of gradual onset and slowly progressive nature, which affects the motor system bilat. and symmetrical

Spinal Cord H. 19 and 6 upper motor neuron (pyramidal tracts) *

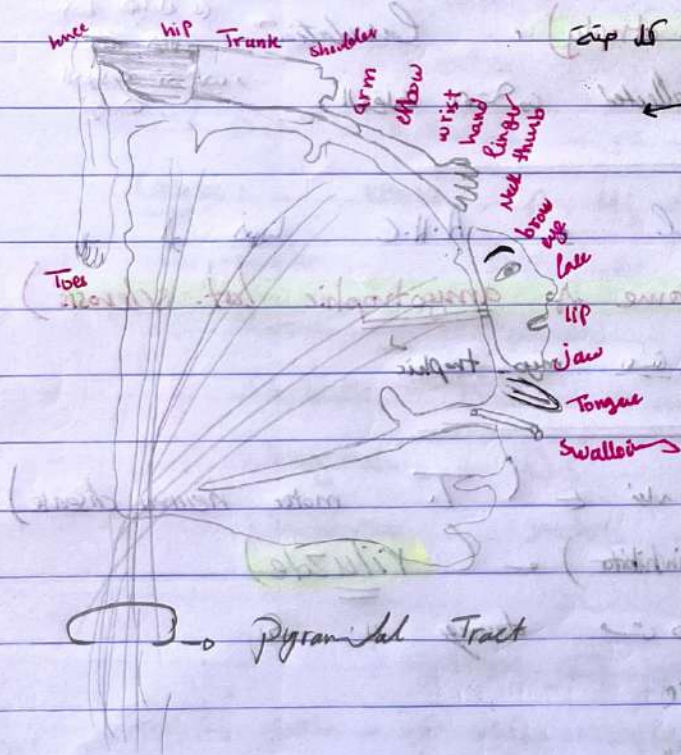
Cortico spinal : ant. horn cell

Cortico bulbar : Cranial N. nuclei

Lower motor N. : motor nuclei of C.N., A.H.C *

motor nuclei : sensory CN 8 = 6 motor, sensory CN 12 = 1 motor

medulla



area 4, area 4

Quadriplegia (both LL Paralyzed) Paraplegia

area 4 in medial of LL

Paraplegia

Paraplegia - below C5 lesion

Quadriplegia - above C5

Iron disease

dual onset and slo

System

mmetrical

motor neuron

horn cell

N. nuclei

motor N

sory via

excessive glutamate
release

apoptosis

motor neuron

vidal tracts)

A.H.C

UMNL

? brainstem & Pyramidal

(Syndrome of lateral sclerosis)

fibrosis في الجذع، spinal cord & lateral part & Pyramidal

Pseudobulbar Palsy, hypertonia, hyperreflexia, clonus, Babinski
 Quadriplegia (إذا شلت جميع الأطراف) Para (إذا شلت 3 أطراف)

LMNL

? motor nuclei

True Bulbar Palsy, hypotonia, hyporeflexia, no Quadriplegia/Para

extrapyramidal & لا يوجد في LMNL، emotion expression lost في LMNL

نوى الدماغ motor nuclei أصلاً وبقوة لها علاقة بالوضعية / النعش / التوتر
 LMNL & LMNL

LMNL

? ant. horn cells (A.H.C)

(Syndrome of progressive m. atrophy), **Emaciation**

wasting - affected muscles (العضلات تتقلص)
 LMNL & LMNL

LMNL + UMNL

? Pyramidal & A.H.C

(syndrome of amyotrophic lat. sclerosis)

atrophy in muscle

Progression (T. motor neuron disease)

Prognosis (glutamate inhibitor) - **riluzole**

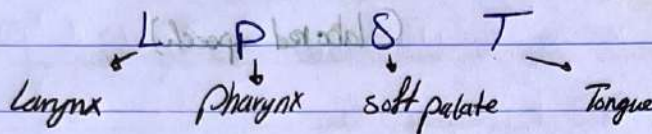
damage

Caudate nucleus, lentiform nucleus, Cortex, Corticobulbar tract

brainstem, Thalamus

nucleus 9, 10, 11, 12, medulla

supply



Pseudobulbar palsy

UMNL

bilat. lesion of corticobulbar tract

Central Palsy

Causes: stroke, CP, MS, atherosclerosis

vascular

bilat. stroke

inflammatory

encephalitis

Neoplastic

brainstem tumor, syringobulbia

Demyelinating

D.S.

Degenerative

motor neuron disease

True bulbar palsy

LMNL

bilat. lesion of nuclei in medulla

Peripheral Palsy

Gullian Barré, tumor, meningo-encephalitis

Vertebrobasilar insufficiency

diphtheria, bulbar poliomyelitis

brain stem tumor

Motor neuron dz

bulbar paralysis

Palatal pharyngeal reflexes

UMNL

hyper

exaggerated reflexes

Brainstem reflex

suppression

Pyramidal Tract

(Pyramidal Tract)

Corticospinal

Corticobulbar

lat. C.S.T

ventral

direct C.S.T

lat.

medial

↓
Cerebr

↓
N. Cerebr

Motor
nuclei
of

Motor
nuclei
of

• Trigeminal

• Oculomotor N.

• dorsal part

• Trochlear N.

• facial motor

• abducens

• facial N.

• ambiguous Nucleus

Vagus, glossopharynx, accessory N.

• ventral nucleus of facial N.

• hypoglossal nucleus

مسؤول عن
upper
quadrant
of face

lower quadrant
of face

مسؤول عن

الوجه من الجانبين

opposite cerebellar
hemisphere

lat. Column
أي هيمس لار ✓

Ventral Column
بندوا ✓

Crossing
لا يتقاطعا

• Resp. m.
نفسا ✓

m. ذ
تفصية ✓
LL, UL

bilat. hemi.

= Cross C.S.T
indirect C.S.T

10-15%

Lecture: 5

Motor and Sensory System Lesions

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The Pyramidal Tract

- The pyramidal tract is a bundle of motor nerve fibers.
- It begins in the cerebral cortex and ends in:
 - a. Anterior horn cells of the spinal cord (corticospinal tract).
 - b. Motor nuclei of the cranial nerves in the brain stem (corticonuclear or corticobulbar tract).
- The pyramidal tract consists of:
 1. Corticospinal tracts (lateral & ventral).
 2. Corticonuclear or corticobulbar tracts (Lateral & Medial).



The Upper Motor Neuron System (Corticospinal Tract)

Function:

1. Initiation and control of voluntary motor activity.
2. Inhibition of muscles tone.
3. Inhibition of deep reflexes.

عظام أعلى لو حمار UMN ← hyperreflexia
hypertonia

Pathway of the Upper Motor Neuron (Pyramidal or Corticospinal tract)

- From Betz cells in motor area 4 (in the upper 2/3 precentral gyrus of frontal lobe) → descends in the corona radiata → pass in the posterior limb of the internal capsule → descends in midbrain, pons and medulla oblongata.

- In the medulla the pyramidal fibers collect to form the pyramid → pyramidal decussation.
 في ههنا نازل في lat. column دلت ، ينتهي عند

Cross d20
A.H.C من fibers level بيدي S3-S4
ذلك لما يوصل level بيدي

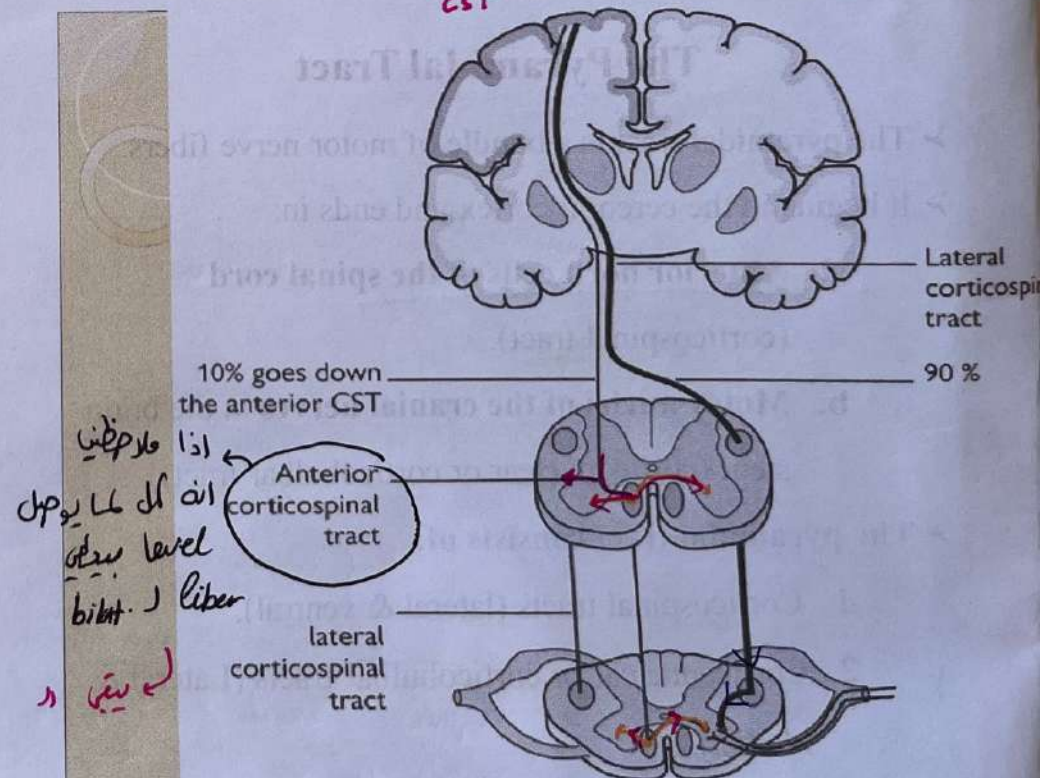
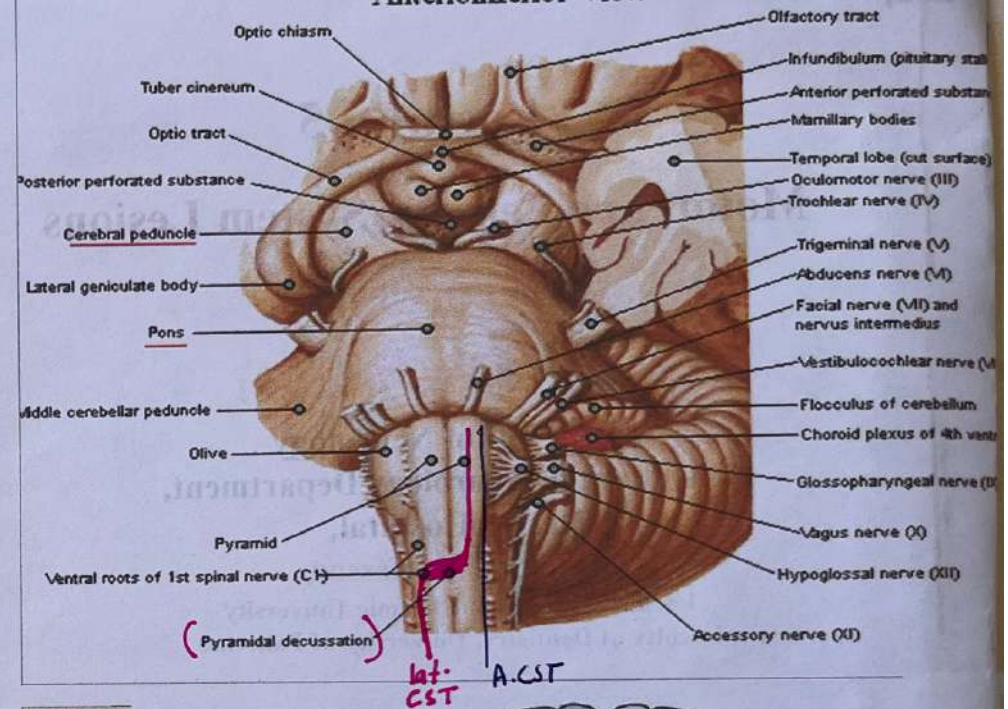
➤ In the lower medulla:

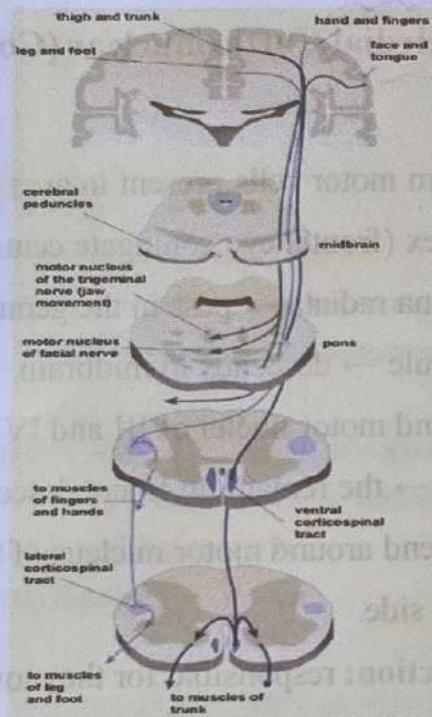
- 85 - 90%** of fibers decussate to descends → in the lateral column of the opposite side of the spinal cord (**indirect or lateral crossed corticospinal tract**) and end in the anterior horns.

- 10 - 15%** of fibers descend directly → in the anterior column of the same side (**direct or anterior corticospinal tract**) then gives fibers for AHCs at the segmental level in **both side**. They descend in the spinal cord until they disappear in the **midthoracic region**.

both side chest m. innervation A.H.C من المسئولة عن

Brainstem Anterioinferior View





C.S.T
 upper M.N.
 ↳ above the cross
 hemiplegia in opposite side
 ↳ below the cross
 Paralysis in same side.

The Corticospinal Tract

- The motor area of one side of cerebral hemisphere is responsible for movement of the muscles of the opposite $\frac{1}{2}$ of the body (mainly the limbs).
- The muscles of respiration are received fibers from both cerebral hemispheres. This explain the absence of respiratory paralysis in hemiplegic person.

لِإِنَّ الِ لِمَبْدُ الرَّئِيسِ فِي هَذِهِ النِّصْفِ
 وَفِي هَذِهِ النِّصْفِ فِي هَذِهِ النِّصْفِ
 لِكِنَّ الرِّسْمِ فِي هَذِهِ النِّصْفِ

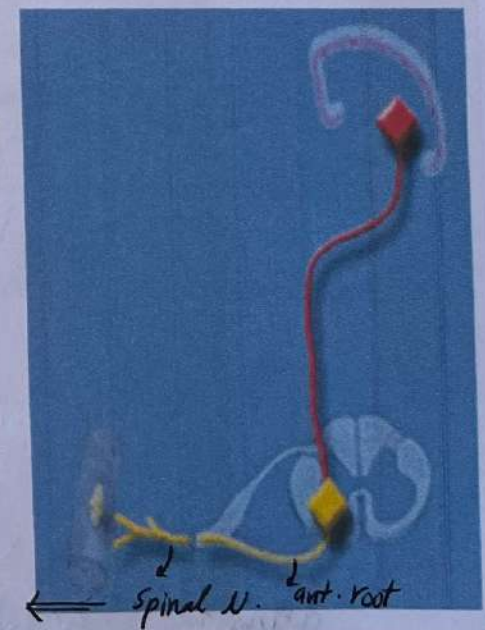
The Lower Motor Neuron System (Spinomuscular Fibers)

Function:

1. Stimulation of voluntary motor at the level of the same side.
2. Stimulation of muscles tone at the level of the same side.
3. Stimulation of deep reflexes at the level of the same side.

Pathway of the Lower Motor Neuron System (Spinomuscular fibers)

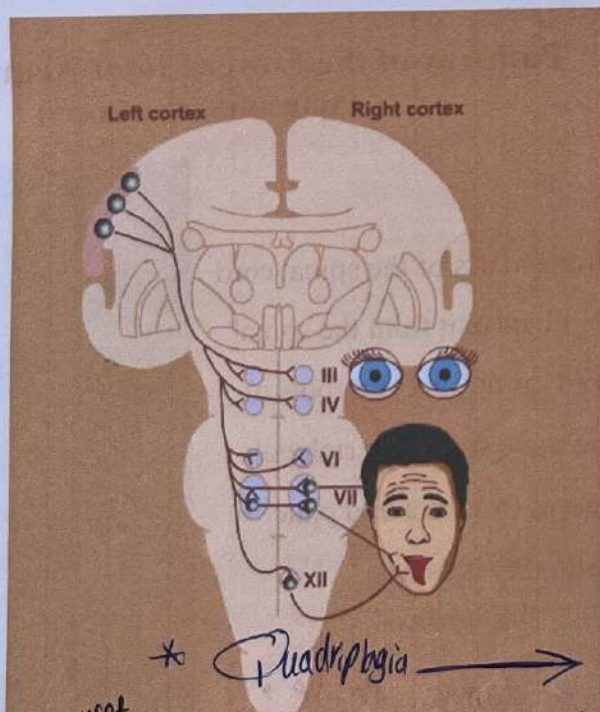
From AHCs of the spinal cord
 → fibers exit from the spinal cord as the anterior roots of the same side → peripheral nerves → motor end plate → voluntary muscles.



لَوْ هُوَ فِيهِ دَمَاجِ مَبْدُ

The Lateral Corticonuclear (Corticobulbar) Tract

- ❖ From Betz cells in motor area 4 (in the lower 1/3 of precentral gyrus of frontal lobe – the face region) → descends in the corona radiata → pass in the genu (apex) of the internal capsule → descends in midbrain, pons → some fibers end around the motor nuclei of V and VII cranial nerves → give fibers for motor nuclei of IX, X, XI and XII cranial nerves in the medulla oblongata.
- ❖ **Termination:** the fibers of corticonuclear tract end in the motor nuclei of V, dorsal VII, IX, X and XI cranial nerves of both side except the motor nuclei of ventral VII and XII cranial nerves are receive fibers only from the opposite side.
- ❖ **Function:** Motor control of the head and neck.



different onset

* bilat. hemip. → Rt. hemip. stroke

m. , nerve , function

The Medial Corticonuclear (Corticobulbar) Tract

- ❖ From motor cells present in area 8 of the cerebral cortex (frontal eye conjugate center) → descends in the corona radiata → pass in the genu (apex) of the internal capsule → descends in midbrain → some fibers end around motor nuclei of III and IV cranial nerves of both side → the remaining fibers descend to reach the pons and end around motor nucleus of the VI cranial nerve of both side.
- ❖ **Function:** responsible for the movement of both eyes.

Classification of the Motor Neuron Lesion

I. According to the location of the lesion:

1. Upper motor neuron lesion (spastic or central paralysis).
2. Lower motor neuron lesion (flaccid or peripheral paralysis).

II. According to the affected limbs:

1. **Monoplegia** (Rt. UL or LL, Lt. UL or LL): paralysis of one limb.
2. **Hemiplegia** (Rt. or Lt. sided): paralysis of one side of the body.
3. **Paraplegia**: paralysis of both limbs.
4. **Triplesia**: paralysis of three limbs (one upper and two lower limbs).
5. **Quadriplegia** (tetraplegia): paralysis of four limbs.

Same onset of disease, Complete spinal cord injury

stroke only affect one side of the body

Causes of the Upper Motor Neuron Lesion

- ✓ 1. CVA.
- ✓ 2. Multiple sclerosis.
- ✓ 3. Brain and spinal cord tumors.
- ✓ 4. Inflammatory: encephalitis, transverse myelitis.
- ✓ 5. Traumatic: head trauma, supdural hematoma, spinal cord trauma.
- ✓ 6. Congenital: cerebral palsy, hereditary spastic paraplegia.
- ✓ 7. Degenerative: vertebral spondylosis and myelopathy.
- ✓ 8. Subacute combined degeneration.
- ✓ 9. Motor neuron disease.

Causes of the Lower Motor Neuron Lesion

- ✓ 1. Anterior horn cells lesion: epiconus lesion, spinal muscular atrophy, poliomyelitis.
- ✓ 2. Peripheral nerve lesion: mononeuropathy, polyneuropathy.
- ✓ 3. Motor end plate lesion: myasthenia.
- ✓ 4. Muscle disease: myopathy.
- ✓ 5. Lumbosacral disc protrusion.
- ✓ 6. Cauda equina syndrome.

Differential between the UMNL & LMNL

	U.M.N. LESION	L.M.N. LESION
1. Location of lesion	From area 4 until AHCs	From AHCs until muscles <i>or motor nuclei in brainstem</i>
2. Location of paralysis	Below the level of lesion: <u>Contralateral</u> (above decussation) and <u>ipsilateral</u> (below decussation)	<u>Ipsilateral</u> at the level of lesion
3. Paralysis	Spastic or central	Flaccid or peripheral
I. MUSCLES		
1. State of the muscles	Wasting occurs <u>very late</u> due to disuse	Wasting occurs <u>early</u> (hypotrophy)
2. Muscle tone	Hypertonia (<u>spasticity</u>)	Hypotonia (<u>flaccidity</u>)
3. Muscle fasciculation (spontaneous contraction of muscles group).	<u>Absent</u>	May be present in the irritative lesions of AHCs
II. REFLEXES		
1. Deep reflexes	<u>Hyperreflexia</u>	<u>Hyporeflexia</u> or areflexia
2. Clonus (Rhythmical series of contraction in response to sudden stretch of muscle tendon).	May be <u>present</u>	<u>Absent</u>
3. Pathological reflexes (Babinski reflex)	+ve (dorsiflexion of big toe + fanning of other toes)	<u>Absent</u>
4. Superficial reflexes	Lost if the lesion is <u>above</u> segmental supply of the reflex	Lost if lesion <u>involve the</u> segmental supply of the reflex

Causes of Cerebellar dysfunction

- **Developmental:** agenesis, Dandy-walker syndrome, Arnold-chiari malformation,
- **Demyelinative disease:** MS, acute disseminated encephalomyelitis (ADEM).
- **Degenerative "Hereditary":** cerebellar degeneration, multi-system atrophy, spinocerebellar ataxia.
- **Neoplastic:** astrocytoma, medulloblastoma, hemangioblastoma, metastasis.
- **Paraneoplastic:** subacute cerebellar degeneration
- **Infectious:** cerebellitis (viral), abscess.
- **Metabolic:** myxedema, hypoglycemia, vitamin B1 deficiency,
- **Vascular:** cerebellar hemorrhage, cerebellar infarction
- **Drug/Toxins:** alcohol, phenytoin, *phenobarbital*.

عامل LMN
زمن سلة زني
لشقة لوفيلته
الشقة به ساني
صناعة إلهو
اي لنب
level

Lesion of the Cerebellum

1. **Ataxia**: discoordination.
2. **Muscle hypotonia**.
3. **Intention tremor**: a tremor which increases as the limb approaches its target (tremor during and maximal at the end of movement). Absent at rest.
4. **Dysmetria**: (error in termination of movement).
5. **Dysdiadochokinesia**: a failure to perform a rapid alternating movement.
6. **Dysarthria**: a type of slurred speech.
7. **Nystagmus**.
8. **Rebound phenomenon**: ask the patient to flex elbow against resistance. Sudden release may cause the hand strike the face due delay in triceps contraction.

Handwritten notes in Arabic:
 Flexion elbow: فلكي إبقاء اليد
 Delay contraction: تأخر انقباض العضلة
 Extension: امتداد
 Rebound phenomenon: ظاهرة الارتداد
 Delay: تأخر

Classification of the Somatic Sensation

The somatic sensations can be classified in two ways:

A. According to the Site of the Sensation:

1. **Superficial (exteroceptive) sensations**: the sensations from the skin (pain, touch and temperature).
2. **Deep sensations**: the sensations from skeletal muscles, tendons, joints, bones and ligaments:
 - a. **Proprioceptive sensations**: sense of position and the sense of movement (kinesthetic sensation).
 - b. **Pressure sense**.
 - c. **Muscle tension sense**.
 - d. **Muscle sense**.

Sometimes the vibration sense is included in this group.

3. **Combined or synthetic senses**: stereognosis, tactile discrimination and vibration sense.

Handwritten note: Superficial + Deep Sensation

The Sensory System

The various sensations in the body include:

1. **Somatic sensation**: from the skin and deep tissues e.g. muscles, joints and bones, conducted to CNS through the somatic nerves.
2. **Special senses**: reaching the CNS through specialized cranial nerves:
 - a. Sense of the vision. optic
 - b. Sense of the hearing and equilibrium. Vestibulocochlear
 - c. Sense of the smell. Olfactory
 - d. Sense of the taste. Lingual, glossopharyngeal
3. **Visceral sensation**: including all sensations from the internal viscera and reaching the CNS through the autonomic nerves.
4. **Organic sensations**: e.g. hunger, thirst and sexual sensations

B. According to the Modality (type) of the Sensation: (More common classification)

I. Mechanoreceptive sensations: these include the:

1. Touch sensations.
2. Pressure sensations.
3. Muscle tension sense.
4. Vibration sensations.
5. Tickle and itch sensations.
6. Proprioceptive sensations.

II. Thermoreceptive sensations: heat and cold sensations.

III. Pain sensation.

The Sensory Pathway or Axis

❖ A sensory pathway includes:

- Receptors.
- Afferent (sensory) nerve that transmits the signals to the nervous system.
- Transmitting tract to the higher centers.
- Cortical sensory areas.

❖ All somatic sensation (superficial and deep), pass through **three order neurons**, from receptors in the skin and tendons, to reach the cortical sensory area.

Pathway of the Superficial Sensation

2. The second neuron:

Is the cell of **Substantia Gelatinosa of Rolandi** and its axon. This axon crosses to the opposite side and ascends in the lateral column of the SC as the **lateral spinothalamic tract** → medulla oblongata → the pons → the mid brain → to relay the sensory impulse at the thalamus.

3. The third order neuron:

Starts in the cell of **the posterolateral ventral nucleus (PLVN) of the thalamus**, its axon ascends to pass through the posterior limb of the internal capsule conducting the impulse to the cortical somatic sensory area (1, 2, 3) in the postcentral gyrus of the parietal lobe.

Pathway of the Superficial Sensation

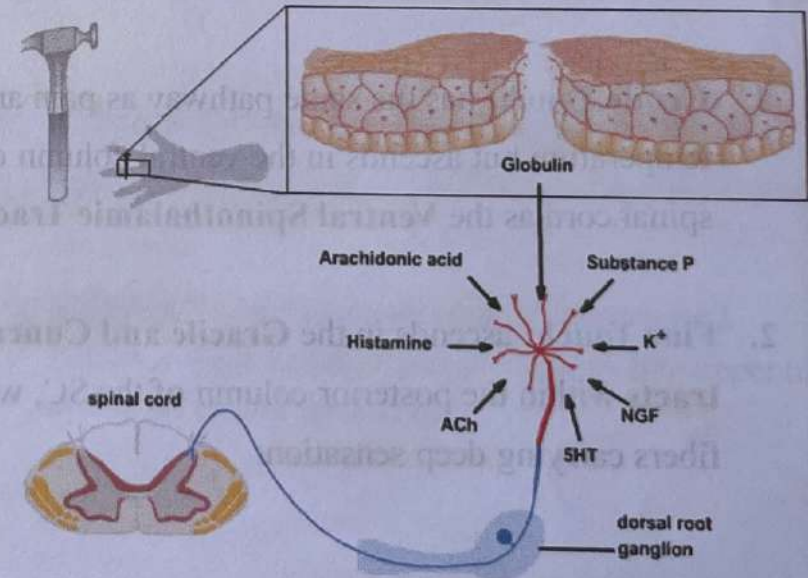
Pathway of Pain and Temperature sensations

1. The first order neuron:

Is the cell of **the posterior root ganglion** and its axon. This axon divided into lateral and medial branch.

- The lateral branch** goes peripherally forming the afferent sensory nerve (the ends of this nerve are receptors of the skin).
- The medial branch** enters the spinal cord through the **posterior root** and relays in the cells of the **Substantia Gelatinosa of Rolandi (S.G.R.)** in the posterior horn of the gray matter.

Handwritten notes:
 ipsi. loss sensation $\rightarrow$ damage $\rightarrow$ ganglion, root, post. horn $\rightarrow$ sensory nerve, up $\checkmark$
 loss Pain, Temp $\rightarrow$ spinothalamic $\checkmark$
 Below the level



Ach- Acetylcholine; NGF- Nerve Growth Factor; 5HT- 5-hydroxytryptamine

Crude touch 1st 2nd order neuron
Let a Post. horn cell $\equiv$ 100%

(main sensory nucleus)

Cross labeled fibers up

~~cross~~

100% 2 opposite side

as ventral column

ant. spinothalamic
Tract

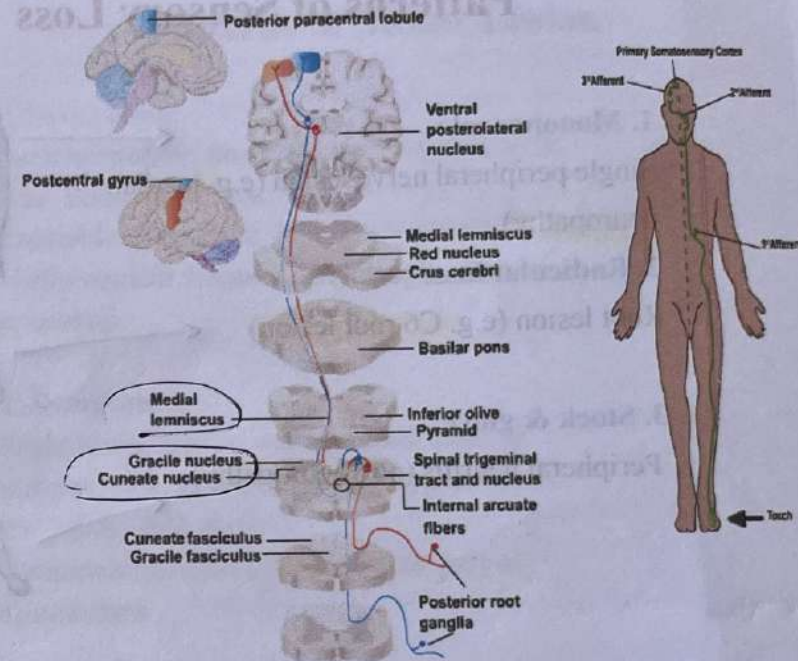
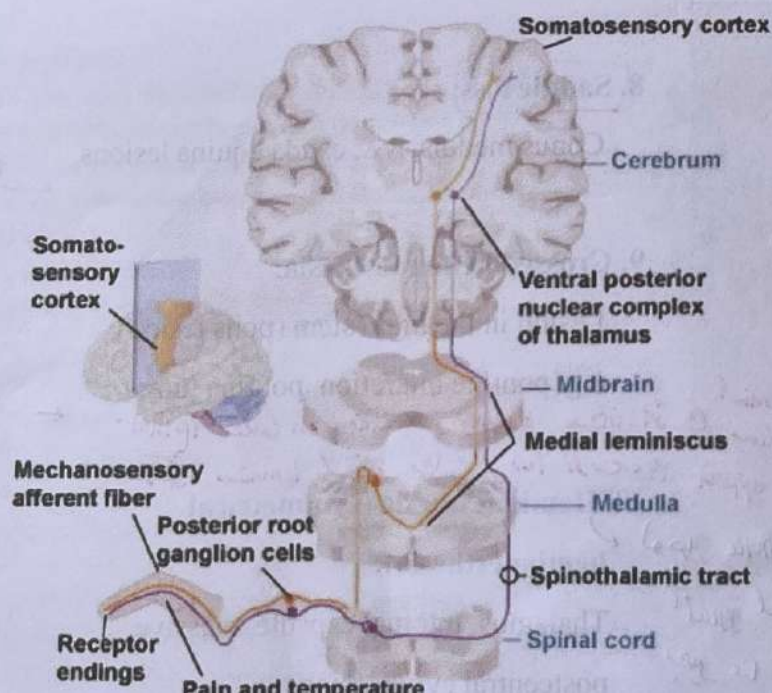
Pathway of Deep Sensation

2. The second neuron:

Is the cell of **the Gracile and Cuneate nuclei** in the medulla → decussating (crossing) to the opposite side of the medulla oblongata ascends with spinothalamic Tract as **Medial Lemniscus** through → the pons → the mid brain → to relay in the **thalamus**.

3. The third order neuron:

Starts in the cell of **the posterolateral ventral nucleus (PLVN) of the thalamus**, its axon ascends to pass through the posterior limb of the internal capsule conducting the impulse to the cortical somatic sensory area (1, 2, 3) in the **postcentral gyrus** of the **parietal lobe**.



Lamination of the Gracile and Cuneate Tracts

- ❖ **The Gracile Tract:** lies more **medially** and received from the **lower part** of the body (the lower limbs and lower half of the trunk).
- ❖ **Cuneate Tract:** lies more **laterally** and received impulses from the **upper part** of the body (the upper limbs and upper half of the trunk).

Patterns of Sensory Loss

1. Mononeural :

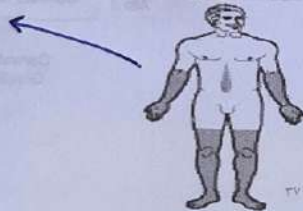
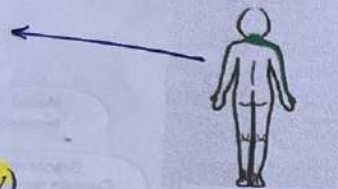
Single peripheral nerve lesion (e.g. median neuropathy).

2. Radicular loss:

Root lesion (e.g. C6 root lesion)

3. Stock & glove:

Peripheral neuritis (polyneuropathy).



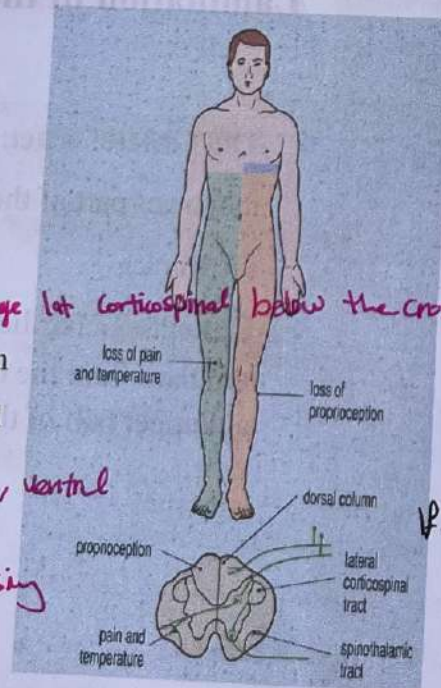
6. Dissociative sensory loss:

Brown-Sequard syndrome

(hemisection or hemicord lesion):

- Ipsilateral paralysis due to damage lat corticospinal below the cross.
- Ipsilateral loss of deep sensation
- Contralateral loss of Superficial sensation due to damage lat, ventral

Spinothalamic
after crossing



4. Sensory level:

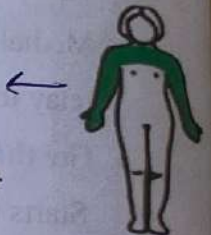
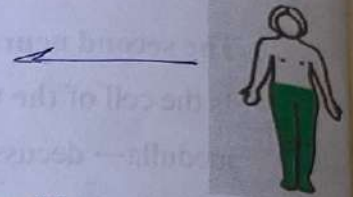
Spinal cord lesion.

5. Jacket sensory (superficial) loss with preserved deep sensation:

Intramedullary lesion in the gray matter (posterior horns) → syringomyelia (C5-T2).

6. Deep sensory loss:

Lesion in the posterior columns: tabes dorsalis, SCD, post-cord \$



8. Saddle loss:

Conus medularis & cauda equina lesions



9. Crossed hemianesthesia:

Lesion in the brain stem (pons lesion):

e.g. pontine infarction, pontine tumor.

Trigeminal descend nucleus damage sensation on face ipsilat. opposite spinal lemniscus

10. Hemianesthesia (symmetrical hemianesthesia):

Thalamus, Internal capsule, massive postcentral gyrus lesions.

stroke, post central gyrus, contra lat. hemianest.



ant. Cerebral art.
 area 11 upper part
 LL dominant CS Contralateral hemihypoesthesia

11. Cortical sensory loss:

Lesion in the postcentral gyrus (areas 1, 2,

3) of parietal lobe

➤ Lower or upper mononesthesia, asymmetrical hemianesthesia, astereognosis, discrimination of single or multiple stimulation in the contralateral limb.

➤ Symmetrical hemihypoesthesia in massive postcentral gyrus lesion



3. Symptoms:

Adult low back pain radiating to right posterolateral surface of left lower limb, pain radiating into foot, hypoesthesia, right ankle reflex decreased, right foot paralysis

Probable diagnosis:

Herniation of disc L5-S1 (Radiculopathy).

4. Symptoms:

Spastic paraplegia, hyperreflexia; Muscle hypertonia, loss deep and superficial sensation, double incontinence.

Probable diagnosis:

- Total transection of spinal cord at T11 segment.
- Midline tumor at the level of the central sulcus.



Patterns of Motor Lesion

1. Symptoms:

Muscle atrophy, fasciculation, hyperreflexia, + ve Babinski reflex.

Probable diagnosis:

Motor neuron disease (Amyotrophic lateral sclerosis).



2. Symptoms:

Right lower flaccid monoparesis, muscle atrophy, muscle fasciculation, hyporeflexia.

Probable diagnosis:

Poliomyelitis infection of lumbar part of spinal cord.



5. Symptoms:

Right: peripheral facial palsy.

Left: Spastic hemiparesis.

Probable diagnosis:

Right sided pontine lesion- Millard-Gubler Syndrome, Raymond-Foville syndrome (Pontine tumor or pontine CVA).



6. Symptoms:

Left sided hemiparesis, lower limb more than upper limb, hyperreflexia.

Probable diagnosis:

Stroke of the right anterior cerebral artery.



middle Cerebral art.

faciobrachial and lower limb type

7. Symptoms:

Ascending flaccid symmetrical paraparesis, decrease superficial and deep sensation.

Probable diagnosis:

Guillain-Barre syndrome. Commonly ascends to upper limbs and face.



8. Symptoms:

Right: central facial palsy; spastic hemiparesis. *due to damage Cortico nuclear tract*

Left: ptosis, dilated pupil, difficulty in adduction eye.

Probable diagnosis:

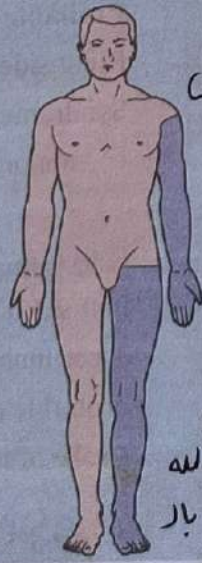
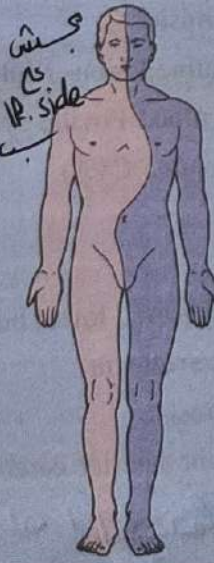
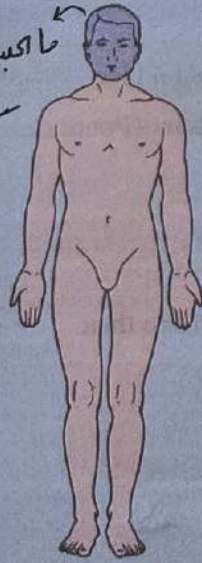
a. Lesion of left midbrain (Weber Syndrome).

b. Stroke in left hemisphere (without oculomotor nerve paralysis).



damage to motor nucleus of oculomotor ipsilat. + Cortico spm - globe cross -> Contralat hemiplegia

Non-Organic Sense Loss (Functional or Hysterical Sensory Loss)



*جانب
بای
و
ل
ب
Trunk*

*ل
کذا له
ما
Organic*

10. Symptoms:

Right: hemihypoesthesia and sensitive hemiataxia.

Left: ptosis, dilated pupil, difficulty in adduction eye.

Probable diagnosis:

Lesion of left midbrain (tumors or CVA).



♥ (Symptoms of sensory system lesion)

* Pain :
- peripheral : Trigeminal neuralgia
- radicular Pain
- Central : Thalamic Pain
- Phantom Pain : due to amputation of limb.
- allodynia : Pain provoked by a nonPainful stimulus.

↳ Causalgia: Severe Sympathetic Pain

2* Numbness, Paresthesia

Positive 2, 11

(Positive Phenomena)

3* hyperesthesia

↑ sensitivity to various stimuli

4* sensory ataxia: loss deep sensation

5* allerent (sensory) paralysis

6* astereognosis

ما يقدر على الاستيعاب بدون النظر

7* hyperpathia, hyperalgesia:

exaggerated response to painful stimulus

(severe pain)

8* hypoesthesia (partial loss of)

all forms of sensation

Lecture: 7

Motor Neuron Disease

Dr. Eyad M. Hussein

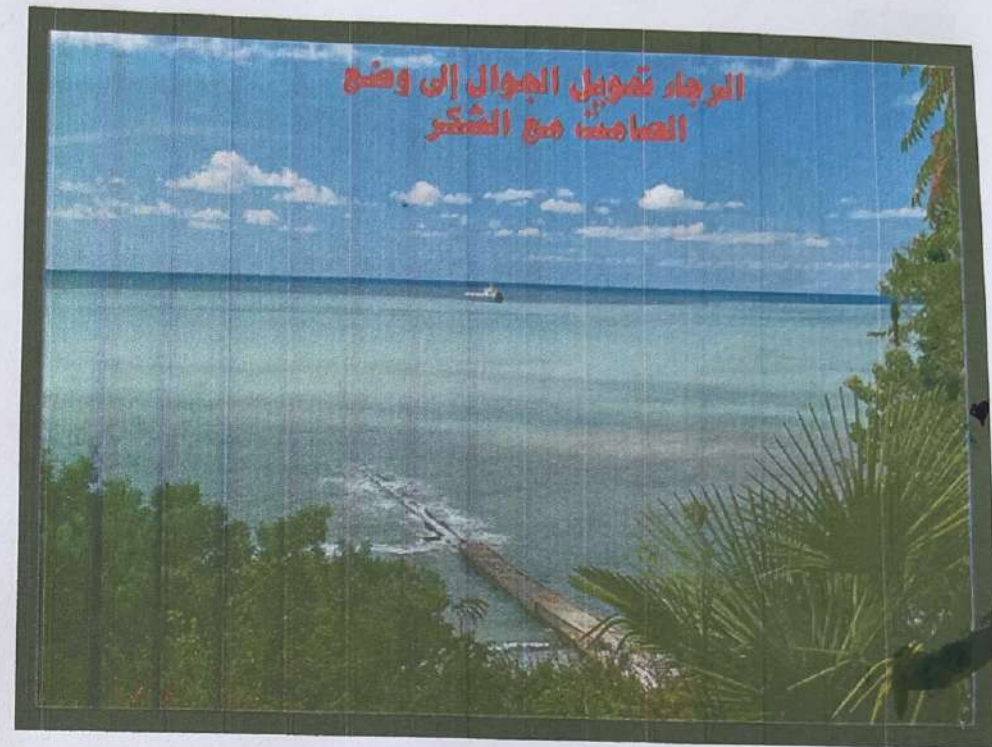
MD, Ph.D of Neurology
Consultant & Head of Neurology Department,
Nasser Hospital,
Assistant Professor,
Faculty of Medicine, Islamic University
Faculty of Dentistry, University of Palestine



قبل صيد الحمازة بالحرمة Case :-

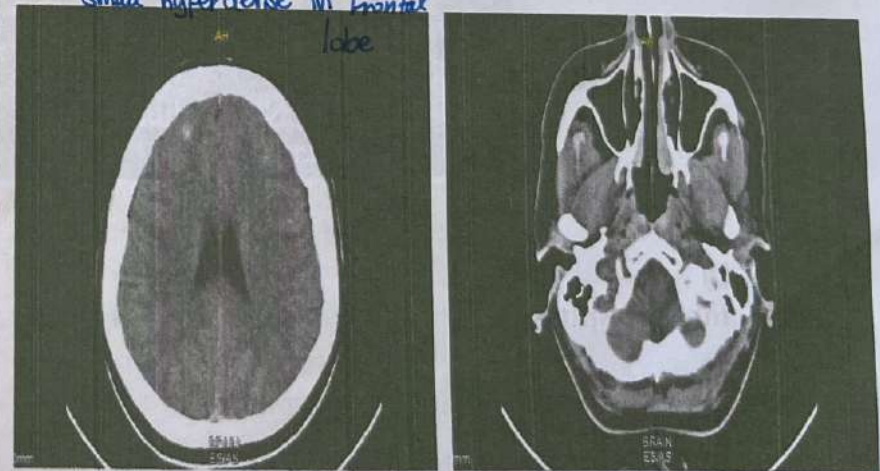
Case- M. In Gaza

- A 47-year old lady, well known case to have HTN, complaining of progressive since two years:
 - Speech difficulty "dysarthria, dysphonia with nasal tone of speech"
- She was seen by Neurologist, initial diagnosis was CVA.
- **Brain CT was done twice:** normal study
- CBC, Full blood chemistry, ANA, TFT were normal
- **Medication was given:** Aspirin, Lozartan, HCT, atorvastatine, Vit. B-complex without improvement

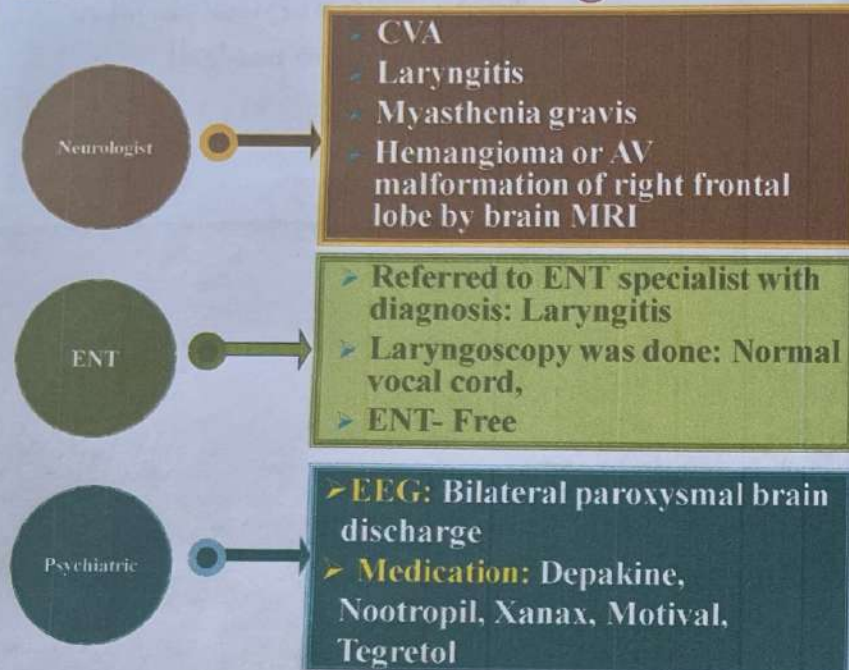


Case- M. Brain CT

تقريرا لورمال لكن لوركت :
small hyperdense in frontal lobe



Case- M. In Gaza During 2009-2011



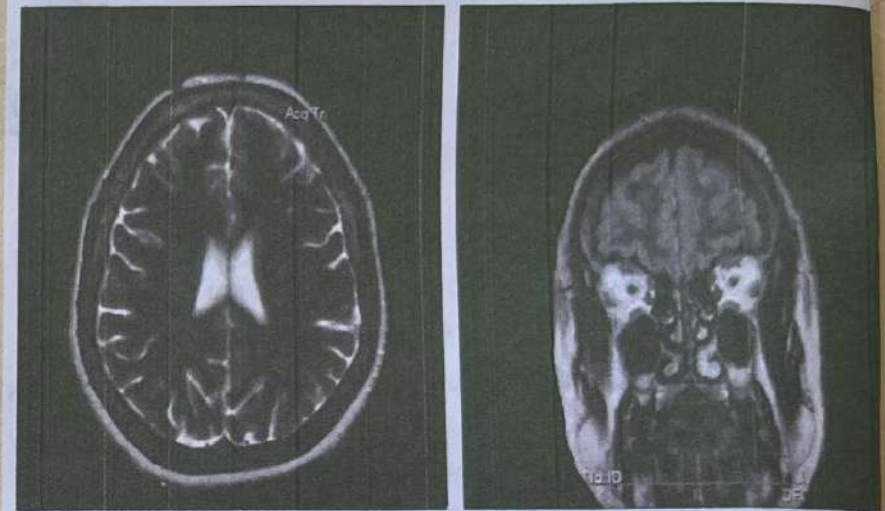
In Makassed Hospital

- > Head CT and MRI were done
- > Final Diagnosis:
 - Sphenoid tumor or infection
 - Very small right frontal angioma
- > Recommendation: *طعامات عذبة بآرام*
 - Neurosurgical management not indicated
 - Needs ENT evaluation and management.

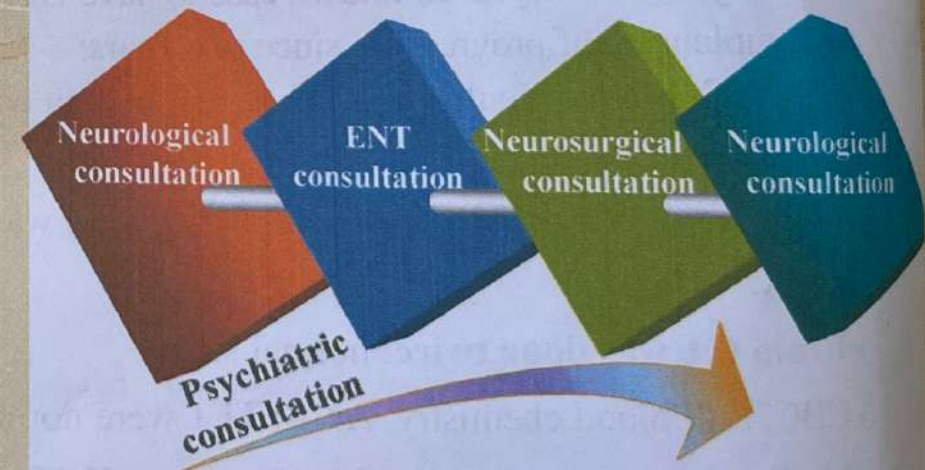
In Augusta Victoria Hospital

- > Laryngoscopy: no any pathological finding
- > Diagnosis: sphenoiditis, treated by antibacterial and anti-inflammatory drugs
- > Follow up with neck, brain and sinuses CT after one month.

Case- M. Brain MRI



Case- M. In Gaza

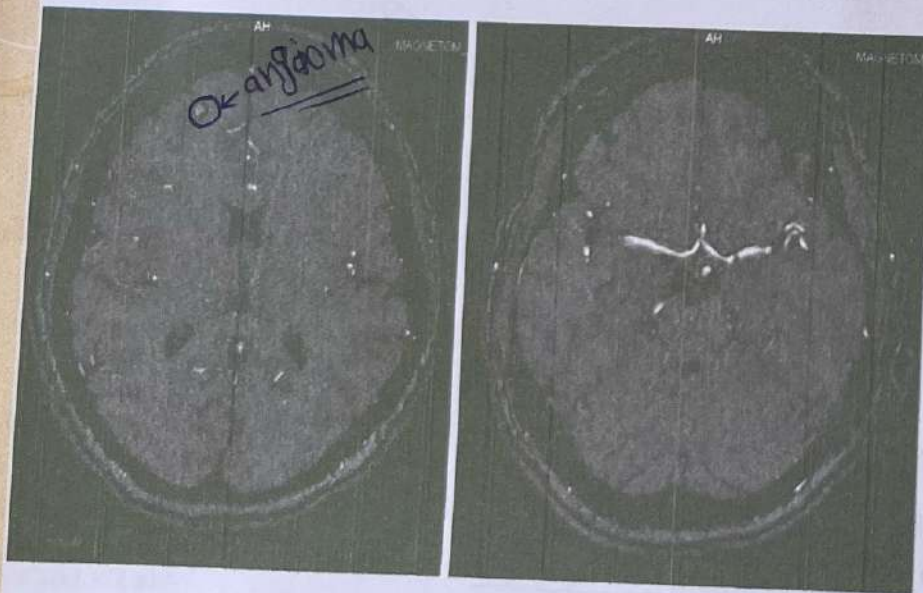


Case- M. In Nasser Hospital

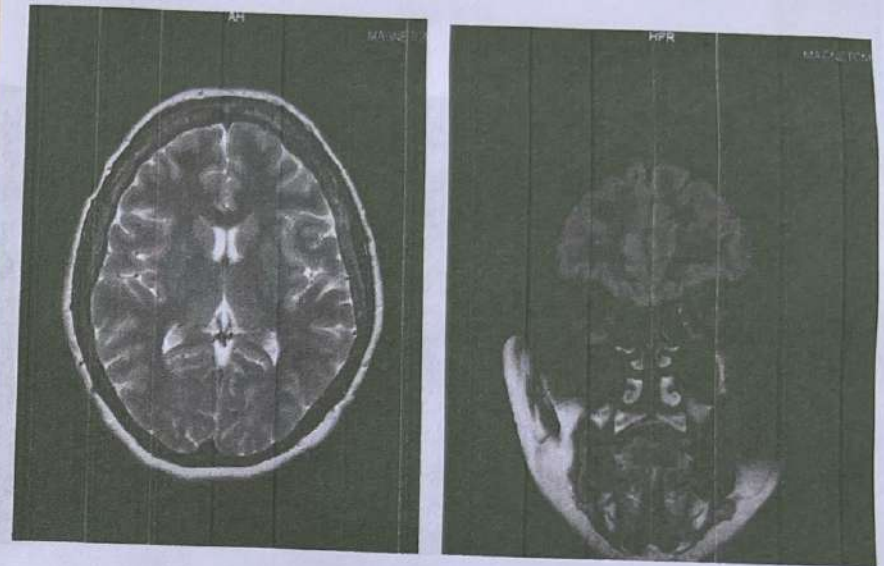
- The patient was referred to our hospital for consultation.
- She suffering from a gradually progressive dysarthria, dysphonia with nasal speech.
- **Family history:** no familial history of neural or muscular disease
- **On Examination:**
 - ❖ Normal cognitive function
 - ❖ Cranial nerves I-VIII intact
 - ❖ Nasal tone of speech, dysarthria
 - ❖ Bilateral wasting and fasciculation of tongue muscles
 - ❖ Decrease of palatal and gag reflexes
 - ❖ **Absolutely there were no motor, coordination or sensory disturbances in upper and lower extremities**
 - ❖ **Autonomic sphincters were intact**

له تعبيل اليد يسطور الرضن ومها عندها
UL, LL weakness

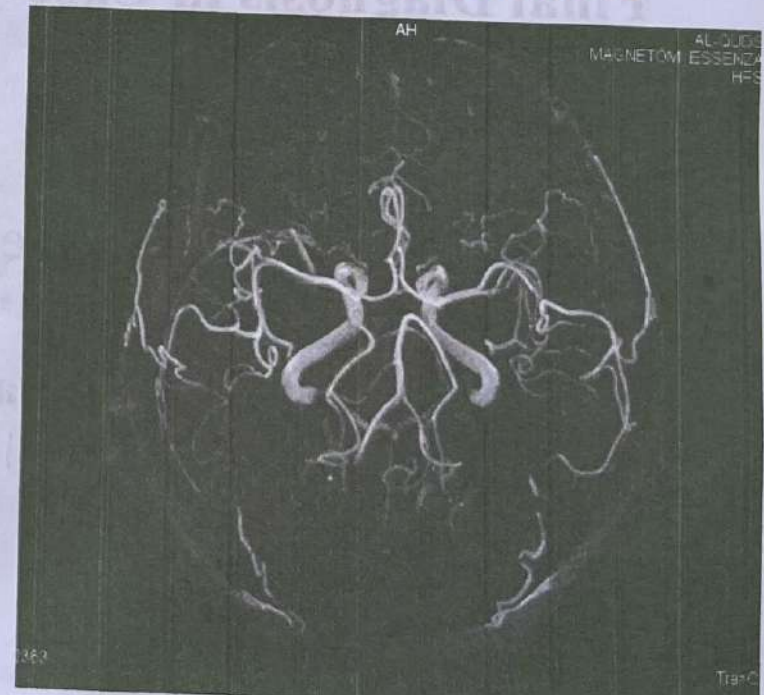
Case- M. Brain MRA



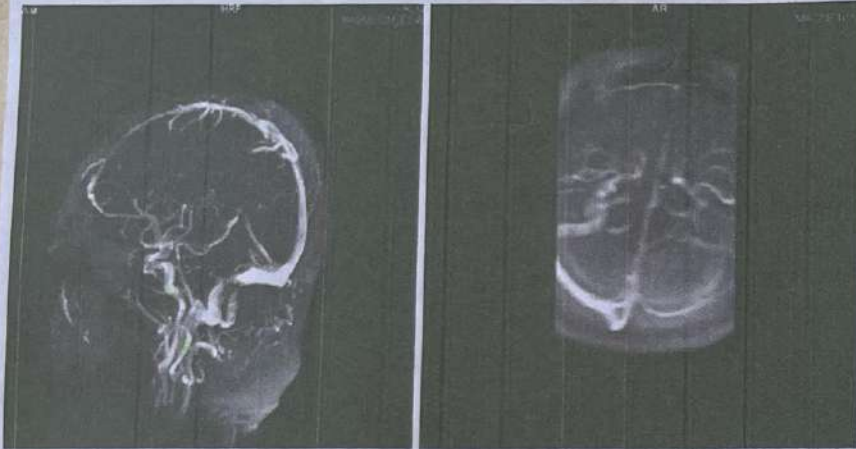
Case- M. Brain MRI



Case- M. Brain MRA



Case- M. Brain MRV



Final Diagnosis in Case- M

Motor Neuron Disease
"Lou Gehrig's Disease"
Isolated Progressive Bulbar Palsy

لأنه بدأ المرض بـ أعراض الـ bulbar

لكن بـ بطانة
 نقول amyotrophy
 نوع من الـ MND
 وفي المص #

Case- M. EMG & NCS

EMG & NCS of upper and lower extremities:

Normal study

EMG & NCS of tongue: Signs of fibrillation, fasciculation and decreased amplitudes of compound muscle action potentials with normal conduction velocities

الـ nerve الـ لـ

Motor Neuron Disease (MND)

- MND is a progressive neurodegenerative disorders characterized by progressive degeneration of the motor neuron in the brain and spinal cord.
- It is characterized by the presence of **both upper and lower motor neuron lesions**.
- What is the Motor Neuron Disease?**
 - ❖ Amyotrophic Lateral Sclerosis (ALS) is the term usually given to MND in the US.
 - ❖ Lou Gehrig's disease. = MND عند الأمريكيين

What is the ALS?

A-myo-trophic =
no-muscle-nourishment

A = absence of

myo = muscle

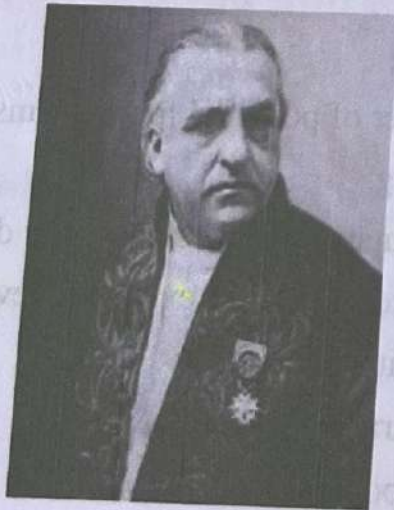
trophic = nourishment

lateral = side (of spine)

**sclerosis = hardening or
scarring**

Lateral Sclerosis refers to
involvement of the lateral
corticospinal tracts.

History of ALS



1869 - First described in publication by Dr. Jean-Martin Charcot, in Paris.

1881 - Lectures translated into English.

Lou Gehrig

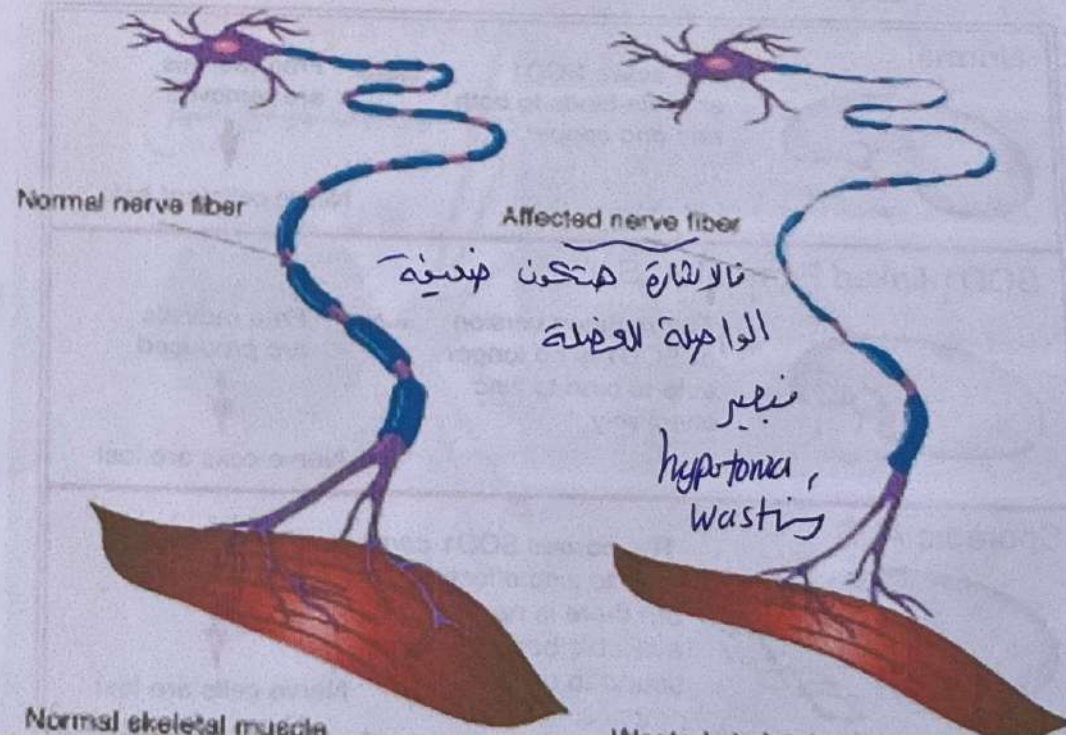
- The famous baseball player who was affected with the ALS.
- Poor 1938 season:
 - Leg > arm weakness/
- 1938-1939, many falls when skating.
- Played 2130 games:
 - Played 8 games in 1939.
 - Stopped May 2, 1939.
- Diagnosed June 19, 1939:
 - His 36th birthday.
- 1940, developed dysphonia.
- Died - June 2, 1941, aged 38 years.



مشاكل
بالصوت
speech

NORMAL SPINAL NEURON

DISEASED SPINAL NEURON

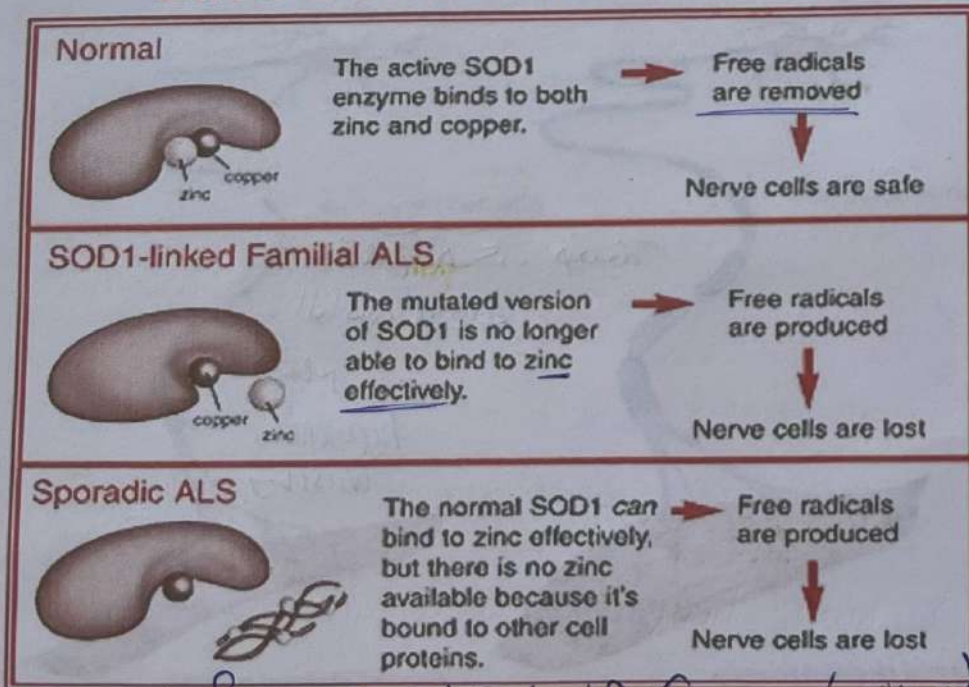


ALS: Epidemiology

- ❖ Incidence rates for ALS: 2 per 100,000 per year.
- ❖ The male to female ratio is about 3:2. ♂ ♀
- ❖ Main age onset - 55 years
- ❖ The incidence of ALS increases with each decade, especially after age 40 years. يبدأ الارتفاع مع كل عقد
- ❖ 50-60% died within 3-5 years of diagnosis.
- ❖ 10-20% survive for more than 10 years.

← تحدث عن كيفية إصابة (motor) و (sensory) في حالة

ZINC-SOD1 INTERACTION



← الإنزيم لو سبب ذلك لما يربطوا به، يربطوا

ALS: Etiology

ALS is classified as either sporadic or familial:

- **Sporadic forms** account for about 90% of ALS cases.
- **Familial forms** make up approximately 10%. Mutations in the **superoxide dismutase 1 (SOD1) gene** are found in 20% of familial cases of ALS. SOD1 is important in removing toxic superoxide radicals.

هذا الإنزيم مرتبط مع free radicals وينزل في Urine ...
 mutation من جين مرتبط مع zinc
 Zinc مرتبط مع جين آخر، accumulation over motor neuron

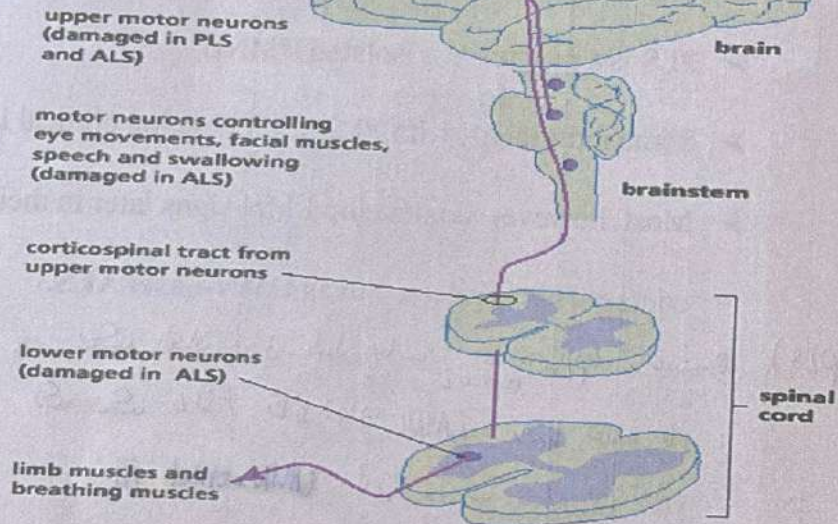
ALS: Etiology

A number of potential mechanisms have been proposed including:

- Mutation in the superoxide dismutase 1 gene.
- Excitotoxicity: excessive level of glutamate.
- Mitochondrial dysfunction.
- Viral infections.
- Apoptosis.
- Growth factor abnormalities, and others.

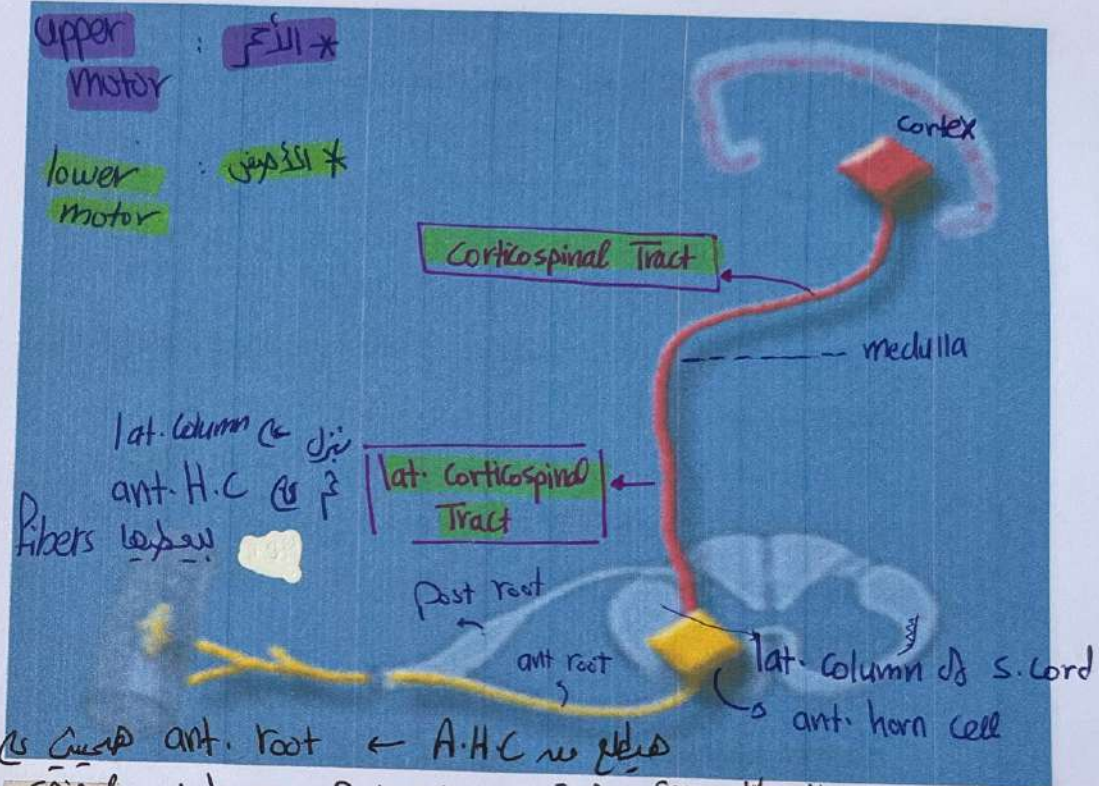
?????

The human motor system



upper motor : * الأخرى

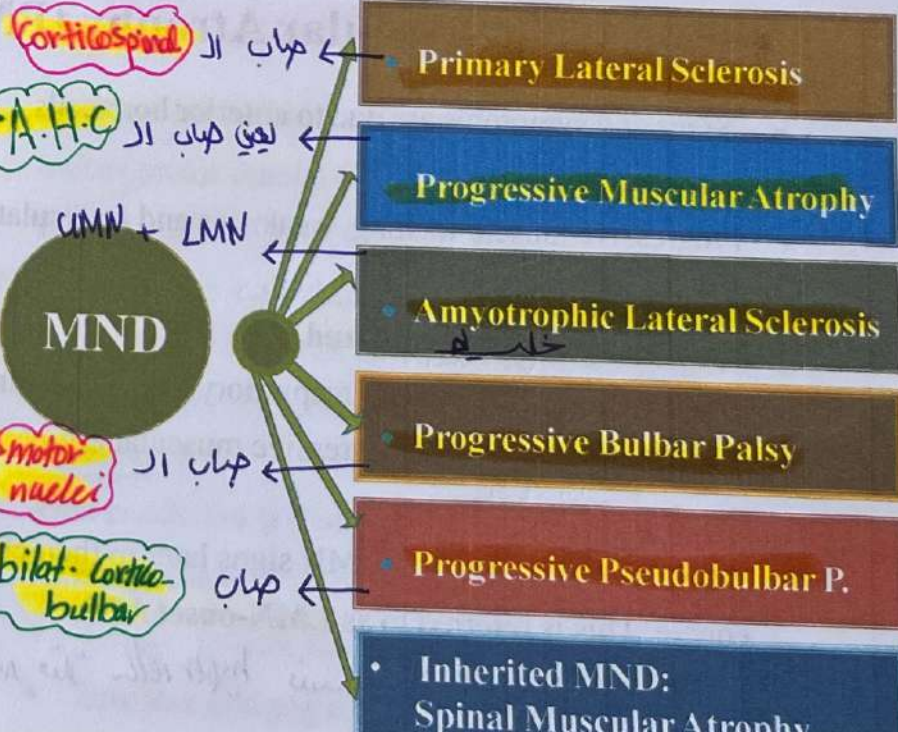
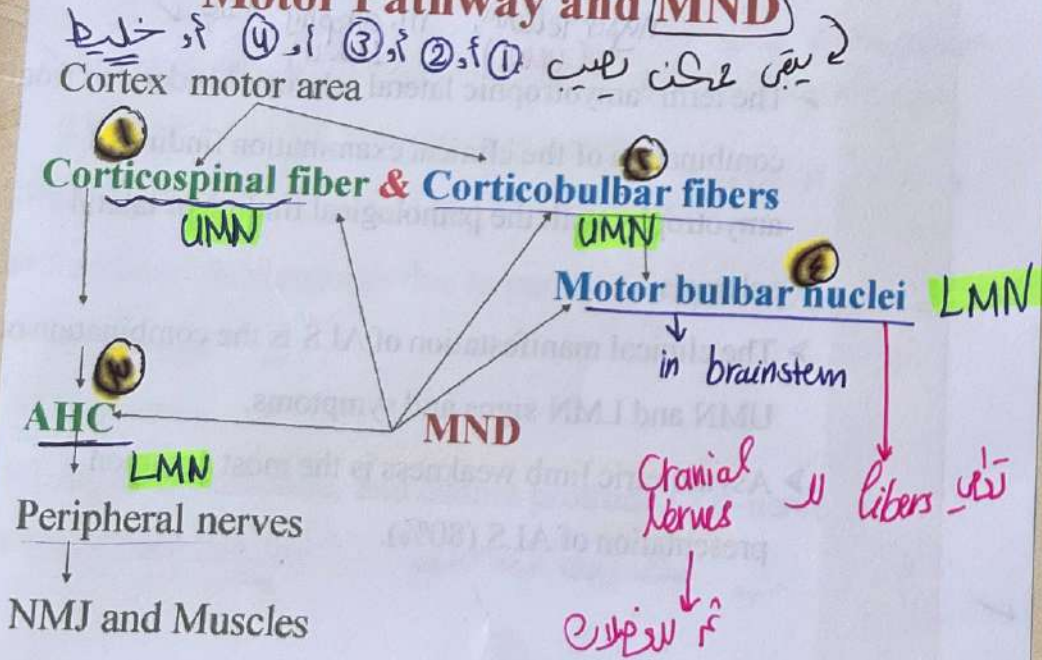
lower motor : * الأخرى



ant. root ← A.H.C

Spinal nerve

Motor Pathway and MND



Motor Neuron Disease: Classification

- **Amyotrophic lateral sclerosis (UMNL+LMNL)** is used synonymous with motor neuron disease: the corticospinal tract and anterior horn cells are affected.
- **Primary Lateral sclerosis (PLS)** when the corticospinal tract alone affected (UMNL).
- **Progressive muscular atrophy (PMA)** when the anterior horn cells alone affected (LMNL).
- **Progressive bulbar palsy:** damage of the bulbar cranial nerve nuclei.
- **Progressive pseudobulbar palsy:** damage of the corticobulbar tract.

Progressive Muscular Atrophy (PMA)

- Signs and symptoms are due to anterior horn cells involvement.
- Progressive muscle wasting, weakness and fasciculation are the cardinal features.
- **Hand wasting- Skeleton Hand.**
- The usual cause of death is respiratory muscle weakness.
- Some individuals with progressive muscular atrophy never develop clinical UMN signs.
- Most, however, develop UMN signs later in their clinical course. This is referred to as **LMN-onset ALS**.

✓ بالآخر تظهر قلة hyperreflexia و LMN onset ALS

Primary Lateral Sclerosis (PLS): Clinical Features

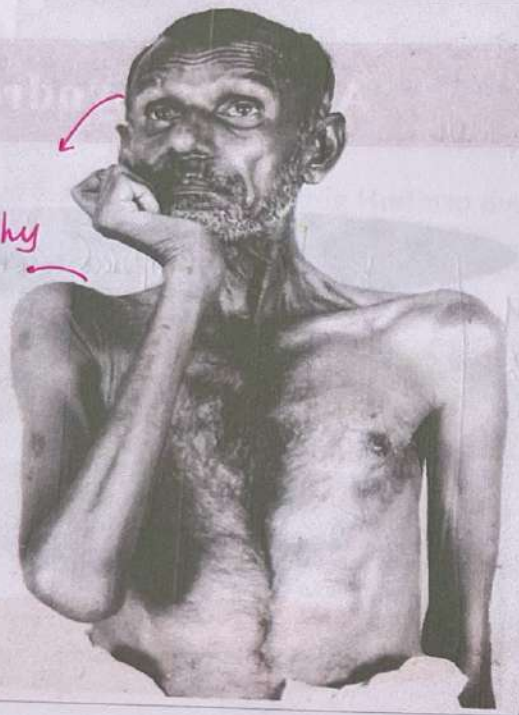
- PLS is a progressive isolated UMNL.
- Some individuals with PLS never develop clinical LMN.
- Most, however, do develop LMN signs later in their clinical course. This is referred to as **UMN-onset ALS**.

(PLS) * من العيان يخل طول حياتهم upper motor neuron L.
 لكن بعد تظهر أعراض LMNL وقتها هيمنة :
 UMN-onset ALS لأنها بدأت بـ UMN

Amyotrophic Lateral Sclerosis (ALS): Clinical Features

- hyperreflexia, m. atrophy (UMNL) (LMNL) ✓ عنده
- The term "amyotrophic lateral sclerosis" is derived from the combination of the clinical examination finding of amyotrophy with the pathological finding of lateral sclerosis.
- The clinical manifestation of ALS is the combination of UMN and LMN signs and symptoms.
- **Asymmetric limb weakness is the most common presentation of ALS (80%).**

atrophy

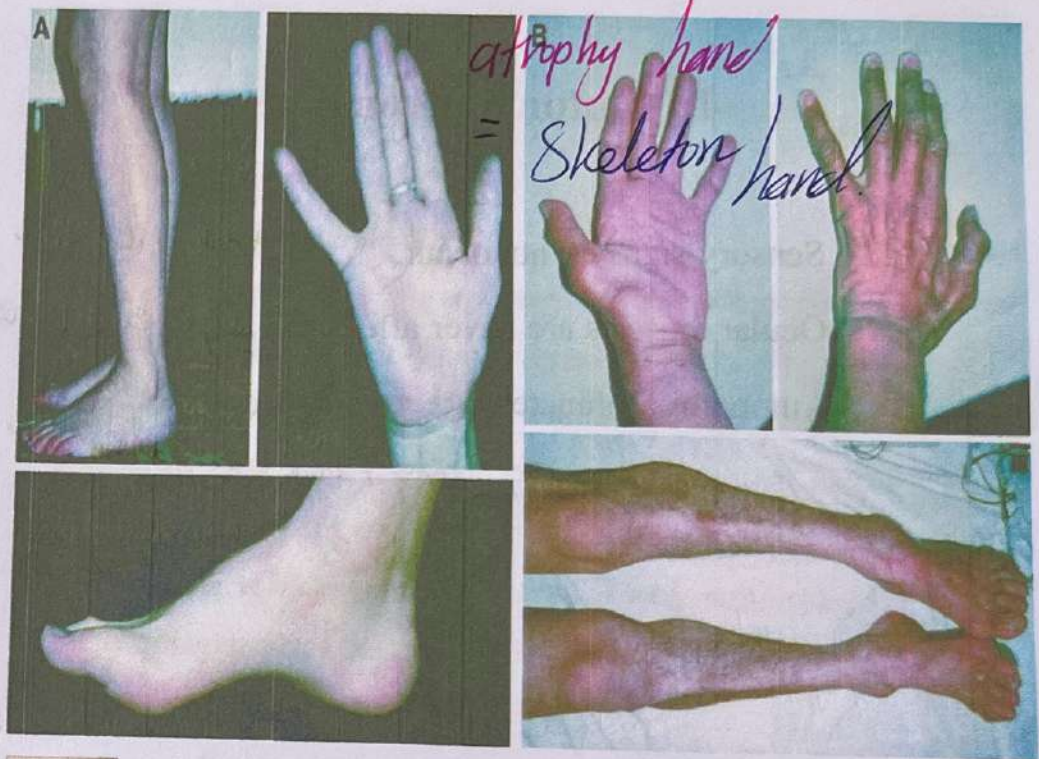


Pseudobulbar Palsy

= corticobulbar.

- Features are due to degeneration of corticobulbar fibers to IX, X, XI and XII nerve motor nuclei.
- Exaggeration of jaw and gag reflexes. * bulbar ۱۱ زراف
- Food and fluid regurgitate due to paralysis of palate (X-nerve).
- Dysphagia.
- Dysphonia.
- Tongue is immobile, and cannot protrude (XII-nerve).
- Emotional lability. عین بیضه درده

أو ليد



Progressive Bulbar Palsy

- Progressive bulbar palsy is a progressive disturbance of the bulbar motor cranial nuclei.
- Bulbar onset in 20-30% of all ALS cases.
- Some individuals with progressive bulbar palsy stay isolated to the bulbar segment. But more commonly, spread to involve other segments. This is then referred to as **bulbar-onset ALS**. عید آ - bulbar symptoms بالآخر تظهر أعراض أخرى
- This condition is distinguished from pseudobulbar palsy by the presence of LMN (nuclear) signs:
 - Tongue muscle fasciculations and atrophy.
 - Jaw jerk and gag reflex are absent.

In Motor Neuron disease

- Sensory signs do not occur. منه مشاكل بالحواس
- Ocular muscles are never affected. منه مشاكل بالعضلات البعيدة
- Autonomic sphincters are never affected. منه مشاكل بال incontinence / urine

Inherited Motor Neuron Disease

Spinal Muscular Atrophies (SMAs)

- SMA is inherited MND (autosomal recessive disease).
- The disorder characterized by progressive degeneration of the anterior horn cells and symmetrical muscle weakness and wasting.

يعتقوا على عمر 18 شهر من resp, aspiration failure

Types of SMA:

- Type I – **Werdnig Hoffman disease** (Acute Infantile SMA)
- Type II – **Kugelberg Welander disease** (Late Infantile or juvenile SMA)
- Type III – (Adult onset SMA)

ALS Plus Syndrome



SMA Type I – Werdnig Hoffman Disease

- ❖ Characterized by reduction fetal movement in late pregnancy with weakness at birth.
- ❖ Swallowing and sucking are impaired.
- ❖ Generalized muscle hypotonia, wasting and fasciculation.
- ❖ The most severe type, 95% of all patients are dead by 18 months.

يبتلع الأم وهي حامل تنحني مع قلة حركة الطفل
هذا ناتج عن LL, LL weak بالجانبين

ALS: Neuroimaging

خاصة Cervical MRI
تأكد من وجود تumor

- ❖ Neuroimaging has traditionally been used to exclude other possible diagnoses in the evaluation of suspected ALS.

لأنه عنده صعوبة البلع و حماري عنده تumor
aspiration Pneumonia
عنه نكاح Saliva Production
بكون Botulinum toxin refractory
تكون NG هذا قد حار لاسوأ
العيان مش حالة لكأ، طابع

Riluzole (Rilutek)

- **Riluzole (Rilutek)** is the **first drug** treatment for the disease in a dose of **100 mg daily** 50 mg مرتين باليوم
- The goal of the treatment is to control the symptoms and maintaining an optimal quality of life.
- Riluzole is anti-glutamate properties.
- Glutamic acid is major excitatory neurotransmitter in CNS

MND: Treatment

- There is no radical therapy for MND
- **Riluzole (Rilutek)** is the only approved medication for treatment of MND
- Antioxidant therapy
- Symptomatic therapy
- Injections of the **Botulinum toxin type B** into the salivary glands in patients with medically refractory sialorrhea
- **Percutaneous endoscopic gastrostomy (PEG)**
- Noninvasive ventilation (NIV)
- Tracheostomy and permanent ventilation
- Under research: Ceftriaxone, Memantine HCL, stem cell and gene therapy

ALS: Symptomatic Treatment

- **Cramps:** Quinine sulfate, Tizanidine, Carbamazepine, Phenytoin, Magnesium
- **Spasticity:** Baclofen, Tizanidine, diazepam.
- **Pseudobulbar affect:** Amitriptyline
- **Dysphagia and aspiration:** Percutaneous endoscopic gastrostomy (PEG).
- **Respiratory care**
- **Psychosocial Care**

SMA Type II – Kugelberg Welanders Disease

- ❖ Pathological features similar Werdnig Hoffman disease.
- ❖ It is slowly progressive disease. Median age at death 12 years.

SMA Type III – (Adult onset SMA)

- ❖ Onset between 2nd and 5th decade with progressive limb girdle weakness.
- ❖ Differential between type – III SMA and progressive muscular atrophy is difficult → genetic test (هنا)

ALS: Laboratory Testing

- Routine lab work usually includes CBC, ESR, electrolytes including calcium and phosphate, LFT, CK, TFT, PTH (ALS is rarely associated with primary hyperparathyroidism), ANA, rheumatoid factor, vitamin B12,
- CSF analysis for cells, protein, oligoclonal bands and cytology for systemic malignancy. → MS (لوشكل ب MS)
- Screening for heavy metals in the blood and urine (lead intoxication).
- Testing for the acetylcholine receptor antibodies (found in myasthenia gravis)

ALS: Diagnosis

- ✓ History
- ✓ Physical examination
- ✓ Electromyography and nerve conduction studies
- ✓ Neuroimaging: brain and spinal cord MRI to exclude other pathology
- ✓ Laboratory testing
- ✓ Genetic testing: genetic testing is not a routine part of the diagnostic evaluation in ALS.
- ✓ Muscle biopsy: muscle biopsy is not a routine part of the diagnostic evaluation of ALS but may be performed if myopathy is suspected.

ALS: Electrodiagnostic studies

- ❖ Electromyography (EMG)- To confirm the Dx. The EMG findings in ALS include fibrillations and denervation signs. Fasciculation potentials may also appear in denervated muscle.
- ❖ Nerve conduction studies (NCS)- Sensory and motor NCS are most often normal in ALS, may be reduced in severely atrophic and denervated muscles.

بالبراق نورمال يكون لكن لو
slightly ↓ conduction severe atrophy

ALS: Exercise

- Prolongs survival
- Improves quality of life
- Reduces spasticity ✓
- Strengthens stronger muscles ✓ *↑ power*
- Elevates mood ✓ *↓ depression*



MND: Life Expectancy

Depends on stage of disease at diagnosis

Over 50% live 3-5 years

20% live 5+ years

10% live 10+ years

5% live 20+ years

لغبي 50٪ بالمقابل غلوتوا
خلال 3-5 و

20٪ لغبيثوا 5 وائلز

10٪ لغبيثوا 10 سنوات
والكن

ALS: Prognosis

- ALS is a fatal disease. *مرض قاتل*
- Younger patients may have a slower rate of progression.
- Patients with bulbar onset, particularly the lower motor neuron type, have a poorer prognosis.

55 سنة عند مرضه بـ ALS



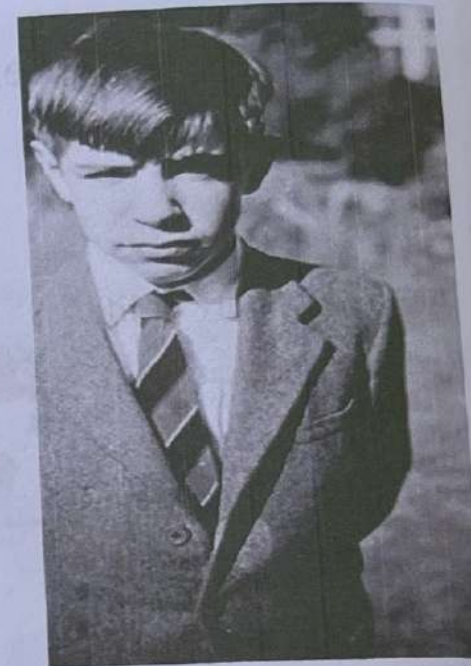
January 8, 1942 – March 14, 2018 Stephen Hawking, and Jane Hawking - 1965

Stephen Hawking, a famous physicist and mathematics who was diagnosed at the age of 21.

Stephen William Hawking

- **Date of Birth:** January, 8, 1942, in Oxford, England,
- Received an **honorary BA degree** from Oxford University, and a PhD from Cambridge,
- In 1959 he won a scholarship to Oxford University, and in 1962 he got his degree with honors and went to Cambridge University to pursue a **PhD degree in cosmology**.
- Diagnosed ALS when he was **21 years old**.
- Hawking has an amyotrophic lateral sclerosis, a condition that has progressed over the years and has left him almost completely paralyzed.
- His father was a physician and **specialist in tropical diseases**

سنة
الطول؟
يعيش مع
ALS



Stephen Hawking at St. Albans School – 1951

Stephen Hawking - Awards and honours

- ❖ 1975 Eddington meda
- ❖ 1976 Hughes Medal of the Royal Society
- ❖ 1979 Albert Einstein Medal
- ❖ 1981 Franklin Medal
- ❖ 1982 Order of the British Empire (Commander)
- ❖ 1985 Gold Medal of the Royal Astronomical Society
- ❖ 1986 Member of the Pontifical Academy of Sciences
- ❖ 1988 Wolf Prize in Physics
- ❖ 1989 Prince of Asturias Awards in Concord
- ❖ 1989 Companion of Honour
- ❖ 1999 Julius Edgar Lilienfeld Prize of the American Physical Society
- ❖ 2003 Michelson Morley Award of Case Western Reserve University
- ❖ 2006 Copley Medal of the Royal Society
- ❖ 2008 Fonseca Prize of the University of Santiago de Compostela
- ❖ 2009 Presidential Medal of Freedom the highest civilian honour in the United States



Stephen Hawking – 1963



