
بسم الله الرحمن الرحيم
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صدق الله العظيم

Motor Neuron Disease

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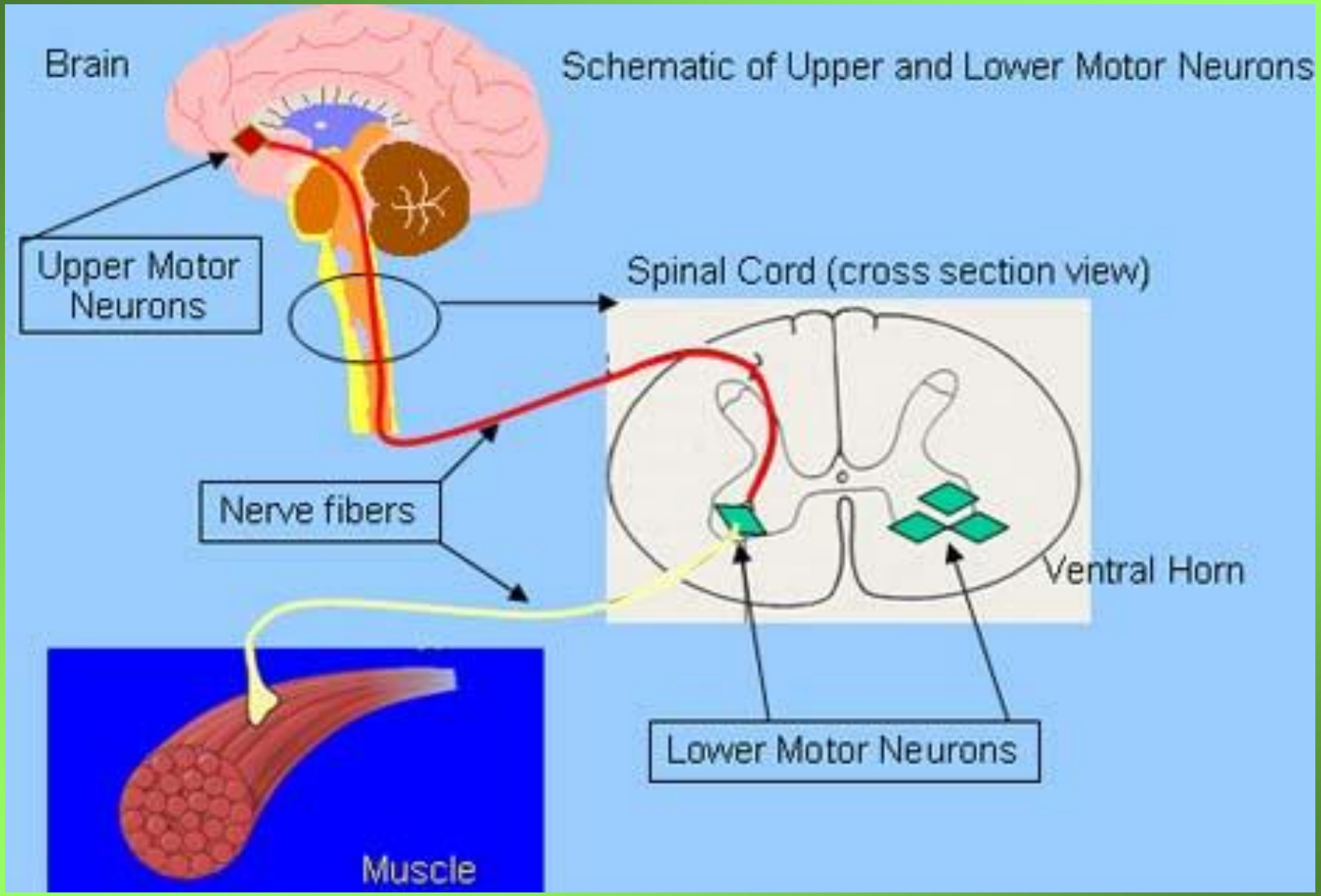
Definition:

- Disorders which are characterized by selective loss of function of the lower and/ or upper motor neurons
- duo 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.


Brain
Schematic of Upper and Lower Motor Neurons
Upper Motor Neurons
Spinal Cord (cross section view)
Nerve fibers
Ventral Horn
Lower Motor Neurons
Muscle

Classification

1. ***Pure upper motor neuron diseases***
    1. Hereditary spastic paraplegia
    2. 1ry lateral sclerosis
    3. Pseudobulbar palsy
2. ***Pure lower motor neuron diseases***
    1. Progressive muscular atrophy
    2. Progressive bulbar palsy (hereditary bulbar palsy)
    3. Spinal muscular atrophies.
    4. Post-polio syndrome.
    5. Post-irradiation syndrome.
3. ***Combined upper and lower motor neuron diseases***
    1. Amyotrophic lateral sclerosis; ALS.

(I)

Hereditary spastic paraplegia

- *Characters*

* AD or AR inheritance
* Age of onset; may be as early as before 20ys., but not later than 40ys.
* Gradual onset progressive course.

- ***Pathology***

* There is degeneration of the crossed pyramidal tract.
* The dorsal root ganglia, posterior root & the peripheral nerve are spared.

Clinically:

- Weakness- both LL is more severe with UMN feature
- Gait :difficult spastic gait
- Reflexes: hyperreflexia, clonus in both LL
- Stiffness and Spastisity:both legs
- Planter reflex: bilateral extensor planter reflex
- Deep Sensory impairment: vibration, position
- Urgency +/- in 10% only.

(II)
Primary lateral sclerosis

- **Cardinal clinical features**:

  Spasticity, clonus, hyperreflexia, extensor planter and pyramidal patterns of weakness in both LL. mainly and UL.
- Bladder, bowel, sensory and sexual functions are usually normal.

(III)
Progressive muscular atrophy

- 1st involve the hands >>> forearm muscle
- Usually asymmetrical, flexors before extensors
- Early proximal presentation is rare

Cardinal features:

- Weakness, hypotonia, lost reflexes, wasting and fasciculation of progressive course with involvement of LL. Muscles and respiratory muscles.
- Bladder, bowel, sensory and sexual functions are usually normal.

(IV)
Progressive bulbar palsy

- With involvement of the motor cranial nerve nuclei in the brainstem
- Tongue >>> 1st to be affected >>> wasting, fibrillation, dysartheria.
- Next with involvement of bulbar nerves >>> manifestations of true bulbar palsy ??!
- Weakness of neck extensors……
- Weakness of orbicularis oculi and jaw muscles.

(V)
Amyotrophic lateral sclerosis (ALS)

Epidemiology:

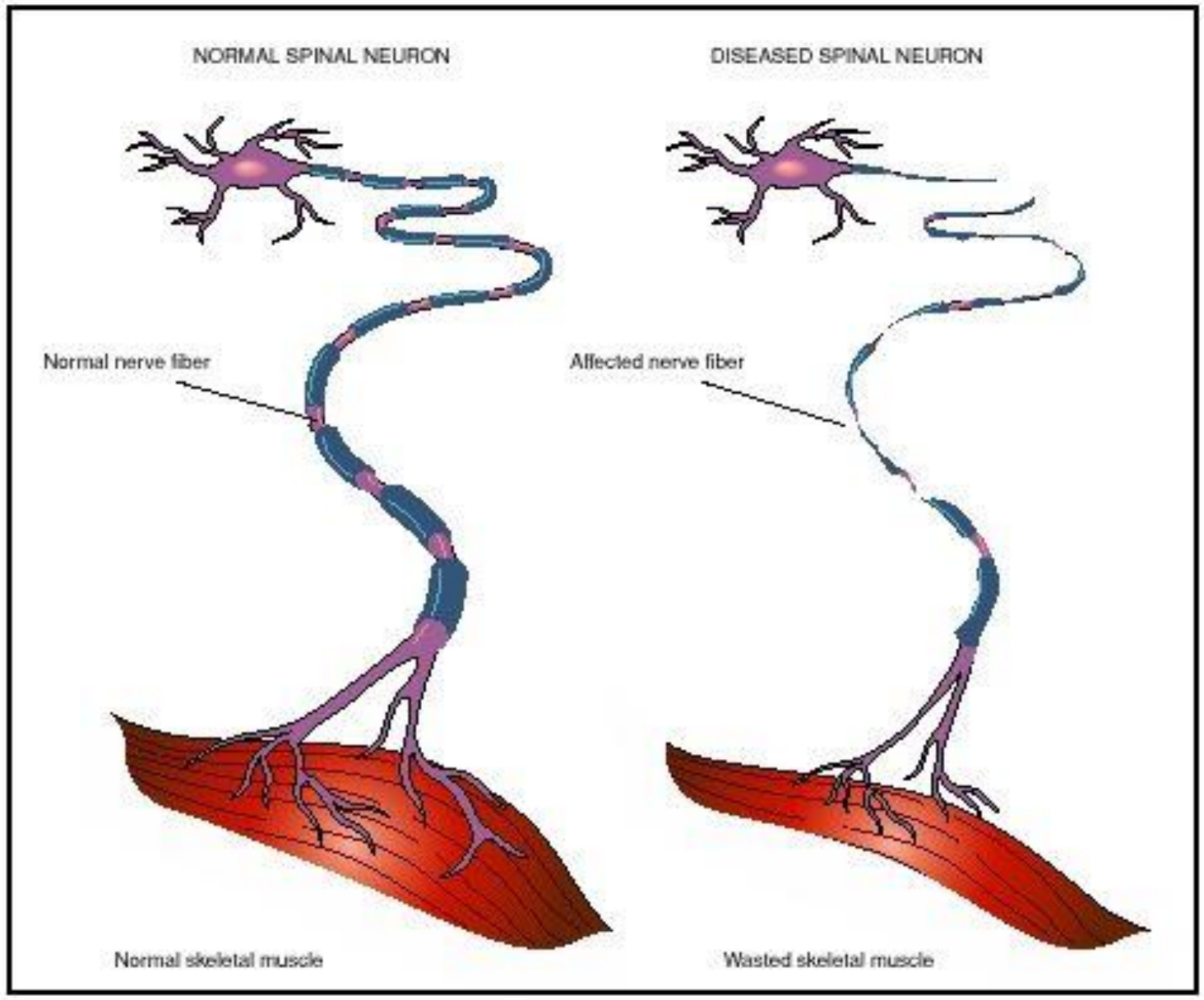
- Age (47 to 63)
- Inheritance (sporadic 90%, familial 10%)

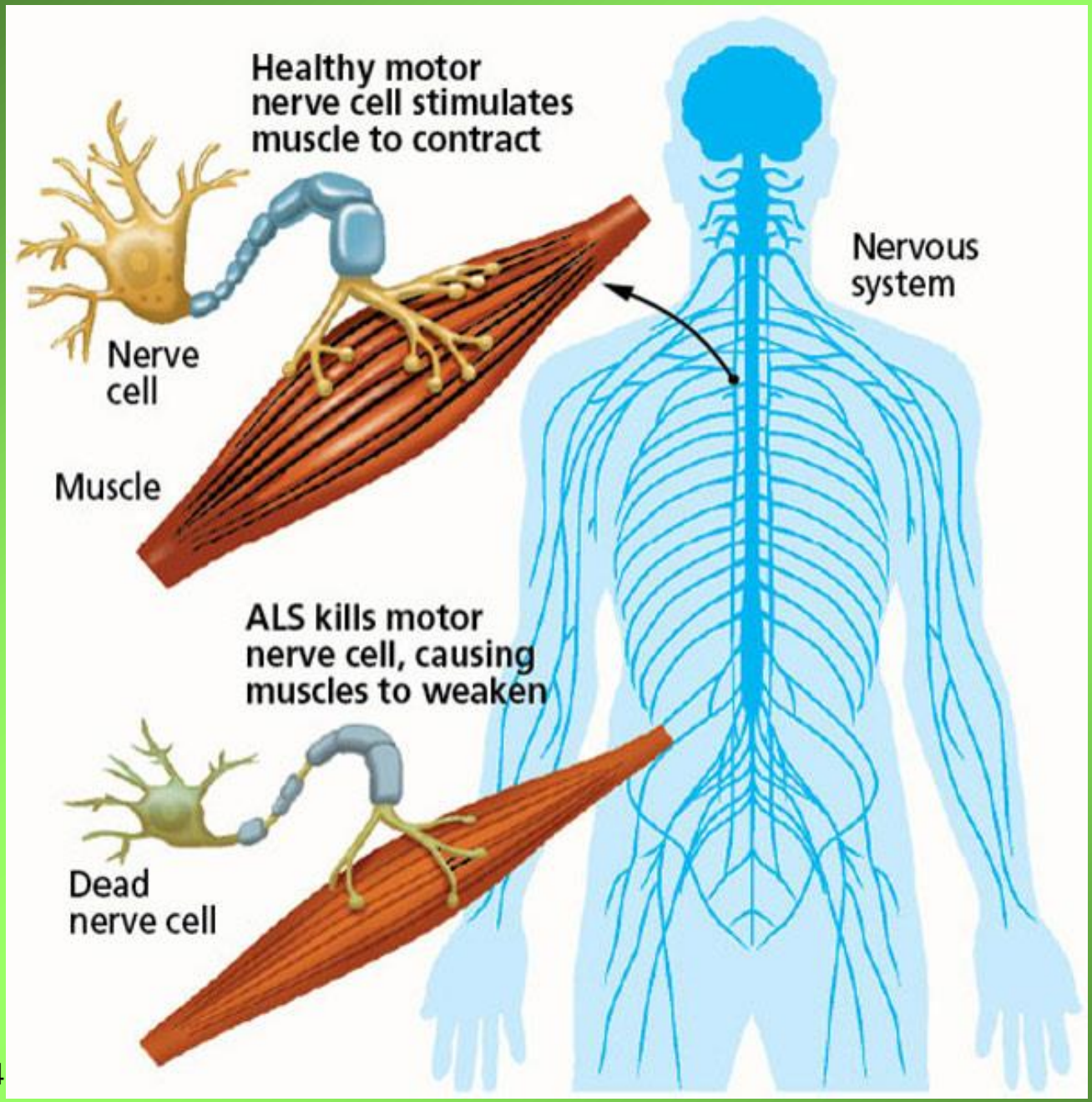
Aetiology:

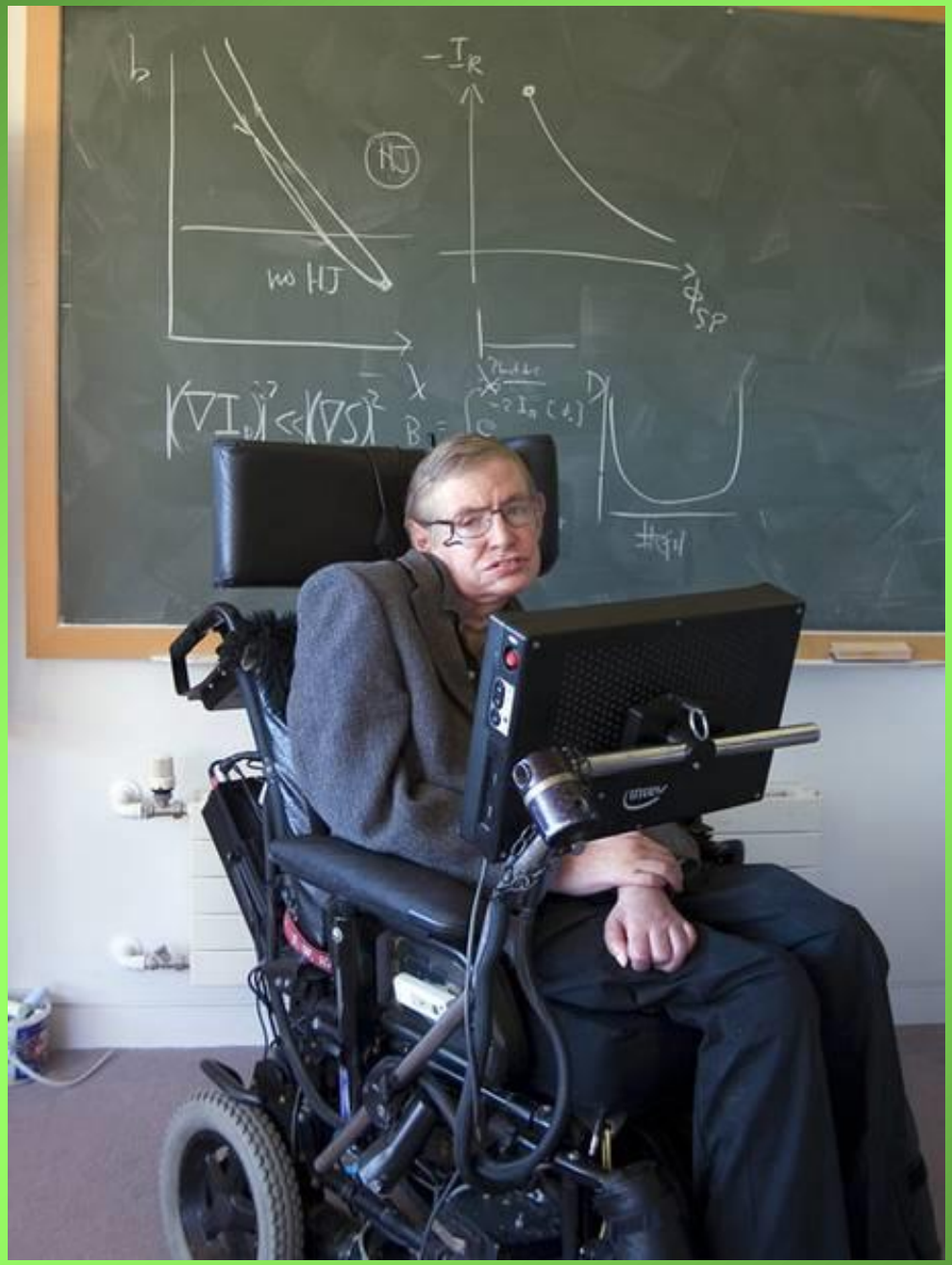
- Genetic inheritance
- Other aetiological factors
  - Post-viral infection
  - Premature aging of motor neurons
  - Immunological role
  - toxins

Clinical features

- Presence of Mixed UMN & LMN signs within the muscle supplied by the same spinal segment is an essential feature for diagnosis of ALS.
- UL. Wasting and weakness
- Fasciculations
- Spastic paraparesis
- Abd. Reflexes-normal
- Pseudobulbar palsy
- Frontal dementia
- Bladder, bowel, sensory and sexual functions are usually normal.


NORMAL SPINAL NEURON
DISEASED SPINAL NEURON
Normal nerve fiber
Affected nerve fiber
Normal skeletal muscle
Wasted skeletal muscle


Healthy motor
nerve cell stimulates
muscle to contract
Nervous
system
Nerve
cell
Muscle
ALS kills motor
nerve cell, causing
muscles to weaken
Dead
nerve cell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$-I_R$
HJ
no HJ
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λ
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Investigations

- **NCS:** usually normal
- **EMG :**fasciculation and fibrillation potentials,Giant motor unit potentials
- **CPK:** mild to moderate increase
- **Muscle biopsy:** features of denervation
- **CSF:** increased CSF protien
- MRI brain ;spine (DD myelopathy)

Differential diagnosis

1) Cord lesions:
    1) Cervical spondylosis
    2) Syringomyelia
    3) Cord tumers
2) 1ry muscle disease
3) CVS
4) Lower motor causes of bulbar palsy; eg; mythenia gravis, basal skull disorders….

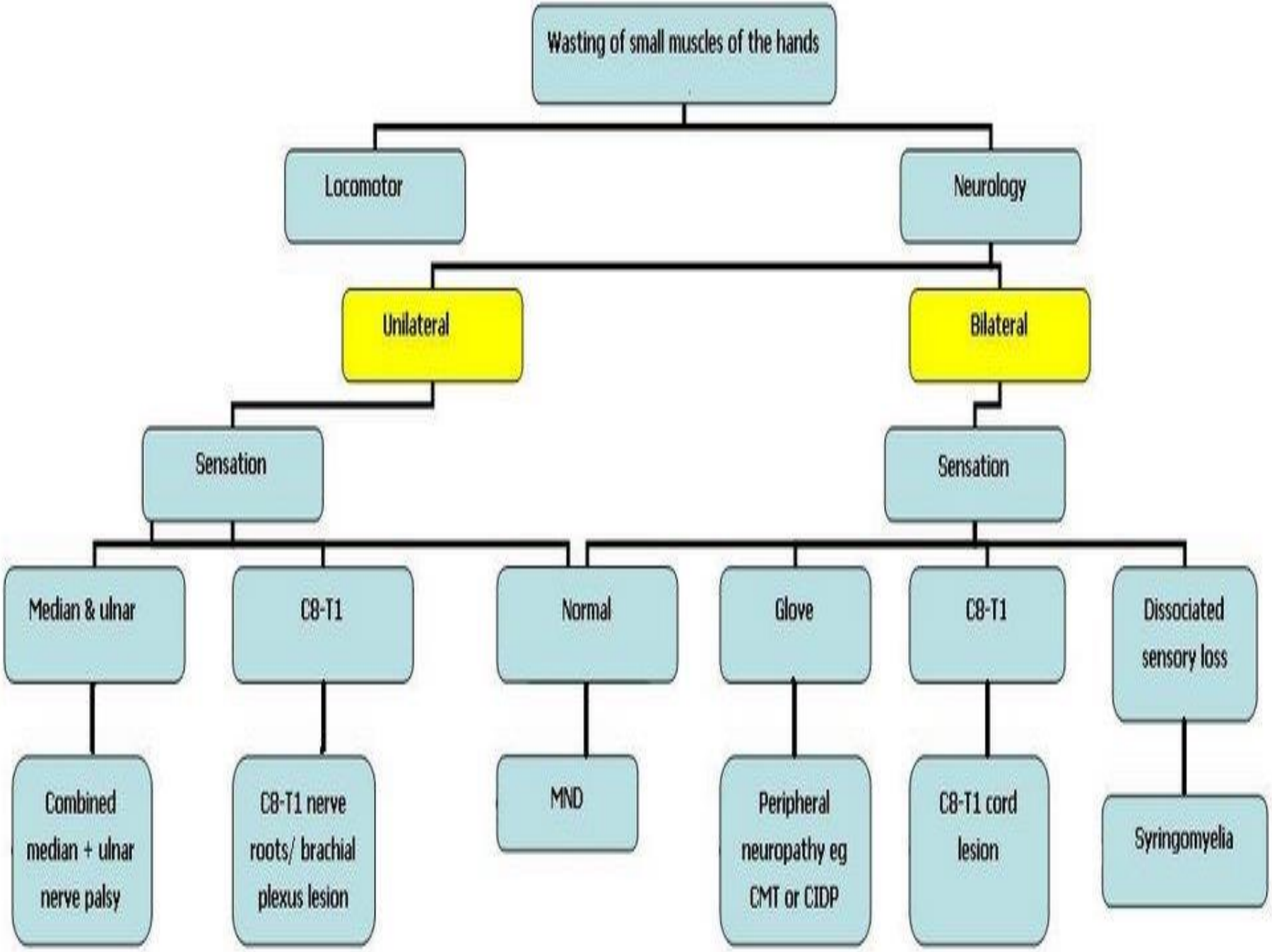
- * Prognosis

* patients with later onset or those with bulbar palsy have the worst prognosis;

* inhalation pneumonia, respiratory failure and malnutrition are common sequales

Management

- baclofen pump can be used for severe spasticity
- Riluzole prolong survival by about 2–3 months
- Breathing support
- Physical therapy
- Occupational therapy (assisted technology)
- Speech therapy (voice amplifiers)
- Nutritional support (feeding tube, percutaneous endoscopic gastrostomy)
- Stem-cell therapy


Wasting of small muscles of the hands
Locomotor
Neurology
Unilateral
Bilateral
Sensation
Sensation
Median & ulnar
C8-T1
Normal
Glove
C8-T1
Dissociated sensory loss
Combined median + ulnar nerve palsy
C8-T1 nerve roots/ brachial plexus lesion
MND
Peripheral neuropathy eg CMT or CIDP
C8-T1 cord lesion
Syringomyelia

Inflammatory muscles diseases (myositis)

Acquired or idiopathic.

Clinically→ muscle pain, tenderness, weakness, wasting.

Histopathologically→ muscles fiber necrosis, regeneration and interstitial inflammatory cell infiltration.

Types & causes:

1) **Infective agents :**

a. **Viral myositis: influenza virus, HIV, HSV.**

b. **Bacterial: acute suppurative myositis.**

c. **Parasitic myositis: toxoplasmosis, cysticercosis.**

2) **Connective tissues disease:" collagen diseases " as in :**

a. **Rheumatoid arthritis, sarcoidoisis, SLE.**

b. **Polyarteritis nodosa, progressive systemic sclerosis.**

Inflammatory muscles diseases (myositis)

3) Idiopathic polymyosites and dermatomyositis :

a. May be autoimmune.

b. Common in females, adult life.

d. clinically :

I. Onset is subacute, rarely to be acute.

II. Muscles pain & tenderness in > 90 % of cases.

III. Subacute proximal weakness.

IV. Skin manifestations in dermatomyositis: Butterfly erythema.

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. Respiratory disorders and pericarditis may occur.

XI. Associated malignancies in >20% of of dermatomyositis.

Diagnosis of Inflammatory muscle diseases

- **Clinical diagnosis.**
- **EMG study:**

Decreased duration and amplitude of action potential.

Increased incidence of polyphasic potential.

Detection of fibrillation potentials.

- **Histopathologically :**

Muscle fiber necrosis, phagocytosis and regeneration.

Interstitial, perivascular and perifasicular inflammatory cell infiltration.

- **Laboratory: increased levels of:**

CPK.

Transaminase.

Treatment of Inflammatory muscles diseases

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

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

- Immune suppression (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 plasmapharesis


Thank You