Motor Neuron Disease: Diagnostic Pitfalls

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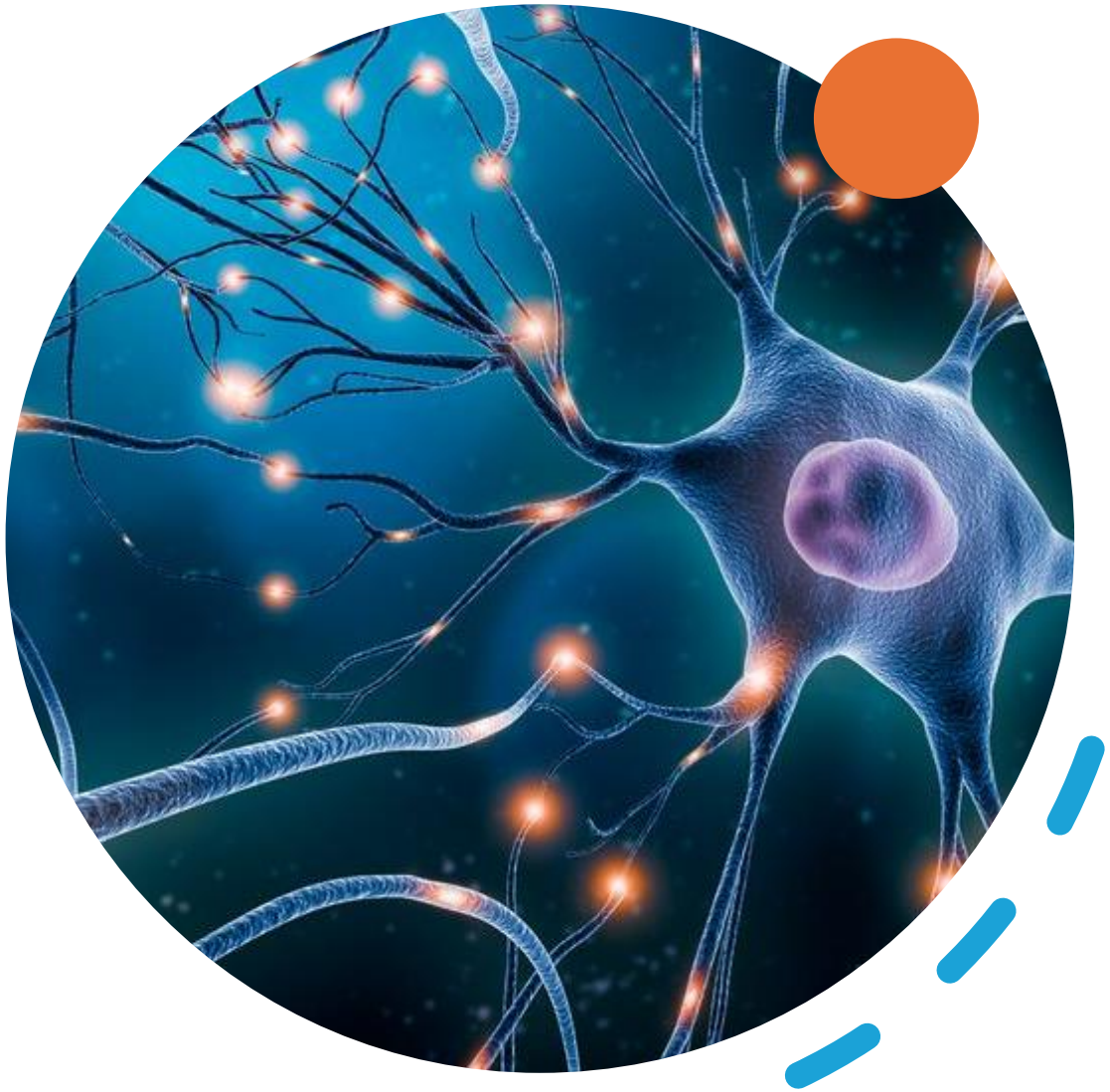
Outline

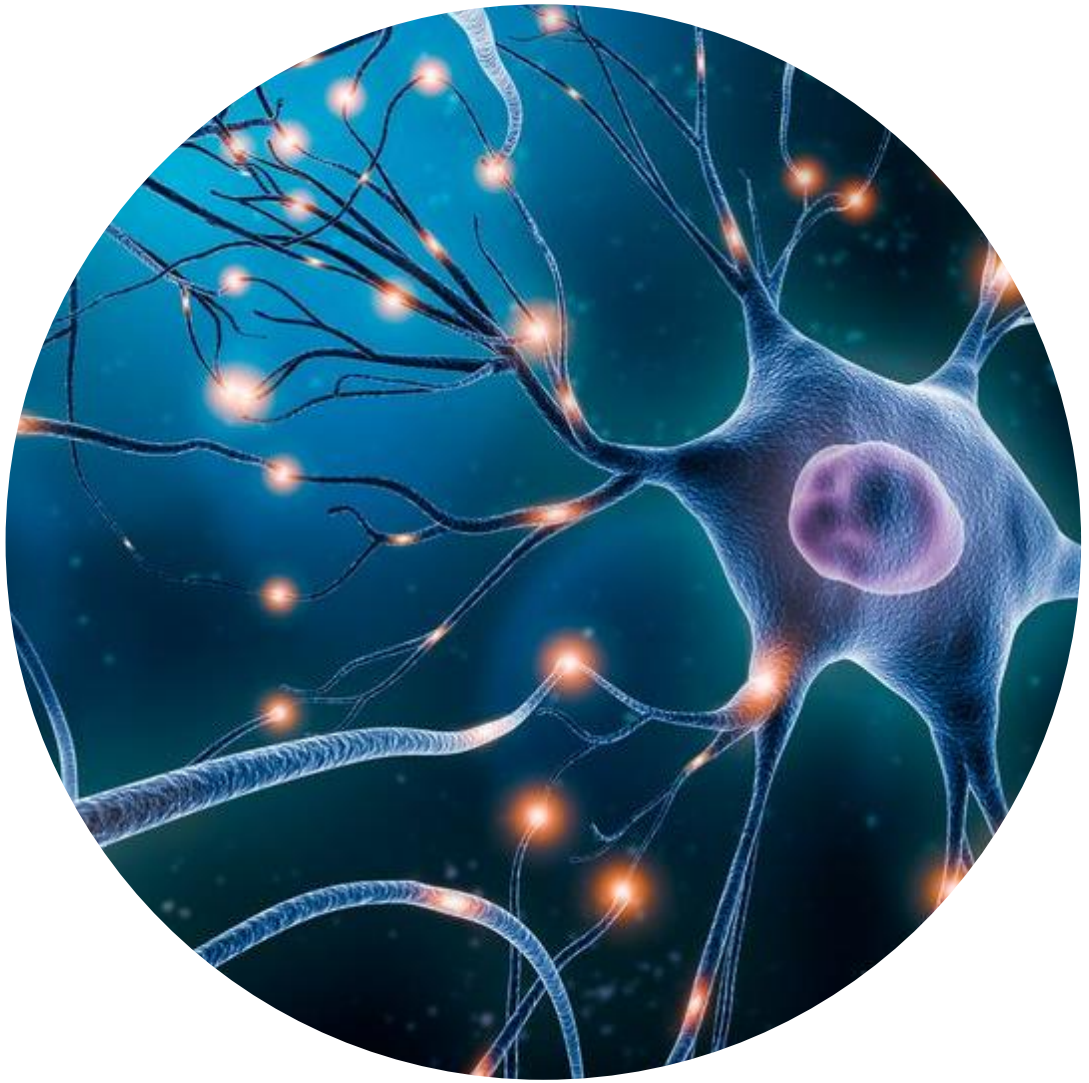
I. Overview of MND
   - Introduction
   - Types
   - Phenotypic presentations
   - MND Mimic

II. Diagnosis of MND

III. Case study



Overview of MND



Motor Neuron Disease (MND)

Amyotrophic lateral sclerosis (ALS) or Lou Gehrig's disease (LGD)

- Heterogeneous neurodegenerative disease that is characterized by the degeneration of both ***upper motor neurons*** (that is, neurons that project from the cortex to the brainstem and the spinal cord) and ***lower motor neurons*** (that is, neurons that project from the brainstem or spinal cord to muscle), leading to ***motor and extra-motor symptoms***.
- Diagnosis is mainly clinical, but treatable mimics must be excluded before the diagnosis is ascribed.
- It has an invariably fatal outcome, usually from respiratory failure, with 50% of patients dying within 30 months of symptom onset.
- Management therefore properly focuses on symptom relief and the preservation of independence and quality of life.

Epidemiology

- MND is relatively uncommon with an annual incidence of 2 in 100,000 and prevalence of 5-7 per 100,000.
- Incidence is lower in the South and East Asian community. (less than 1 per 100,000)
- Most MND is sporadic but approximately 10% is inherited.
- Incidence peaks in the age group 50 to 70 years.
- Young onset MND (age <45 years) – slower progression
- Juvenile onset MND (age <25 years) – very rare, family history+
- Male to female ratio is three to two.

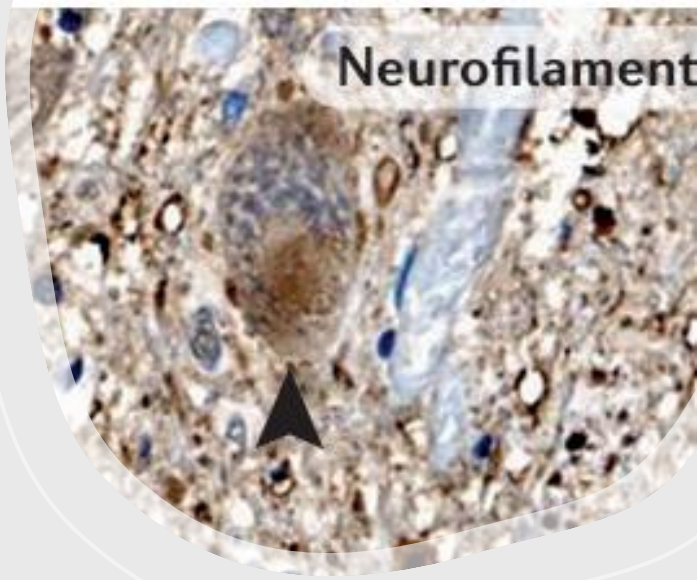
Types of MND

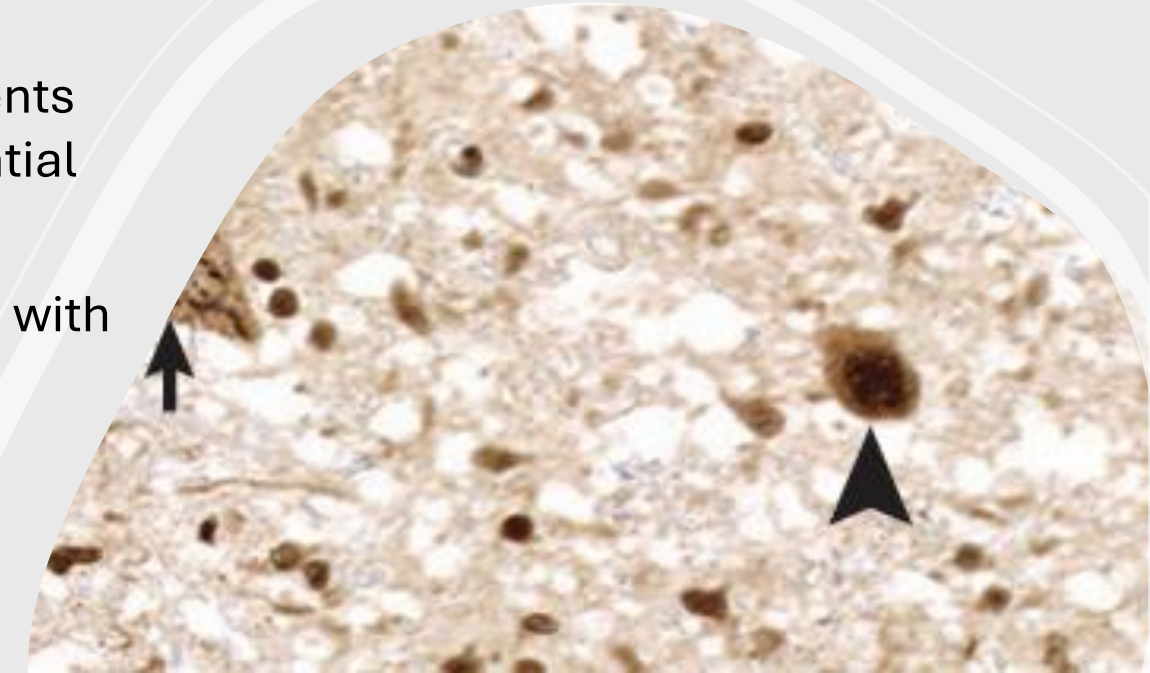
❖Classically, three distinct MND phenotypes

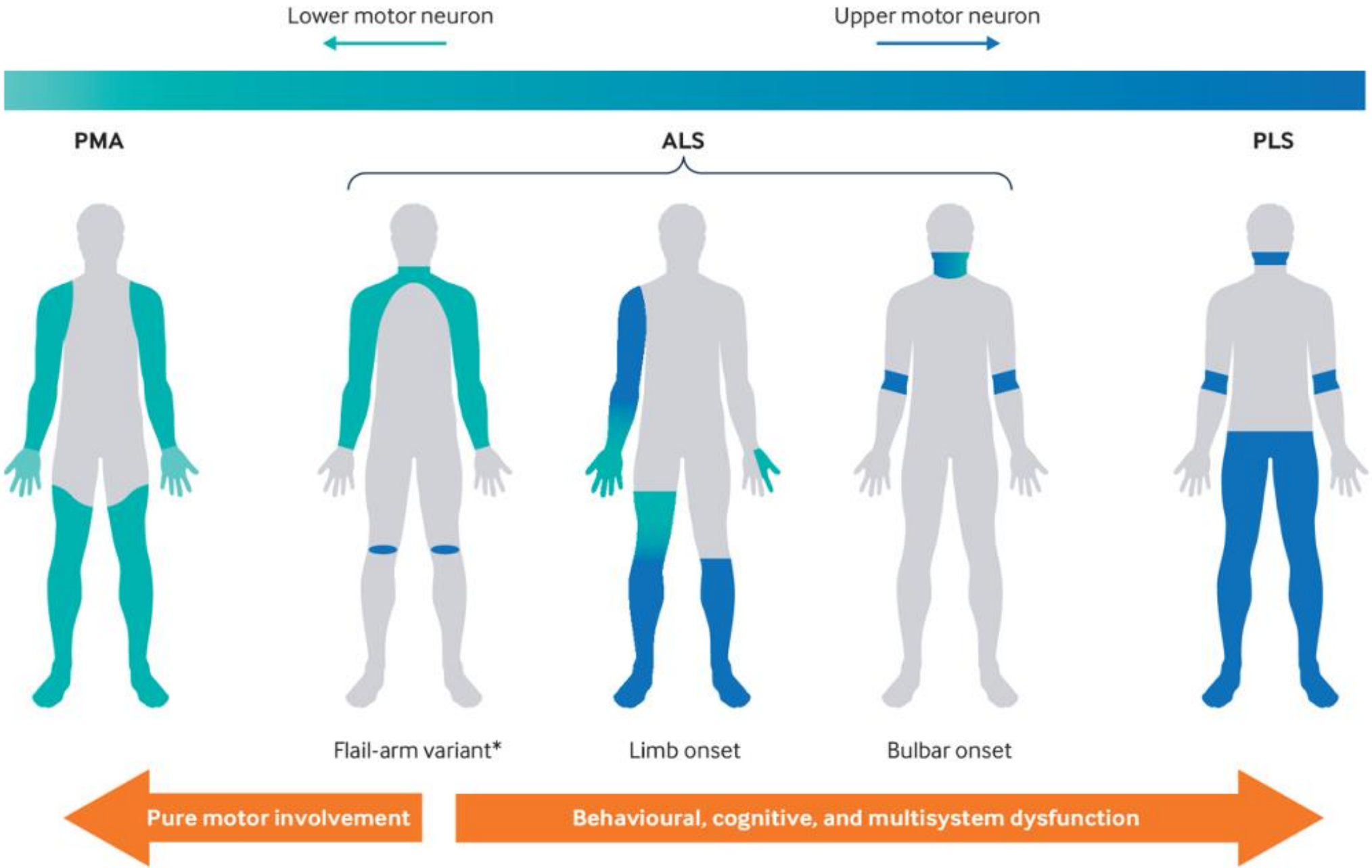
1. Amyotrophic lateral sclerosis (ALS) represents 85% of all MND cases and features a combination of *upper and lower motor neuron dysfunction*.
2. Progressive muscular atrophy(PMA) (5% of MND) presents with *lower motor neuron dysfunction*, with flaccid weakness and muscle atrophy.
3. Primary lateral sclerosis (PLS) (2-3% of MND) presents with *upper motor neuron dysfunction*, with substantial muscle spasticity.

❖ALS is typically associated with rapid clinical decline, with survival typically 3-4 years from symptom onset

❖but PMA and more so PLS are associated with longer survival.







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Phenotypic presentations of ALS

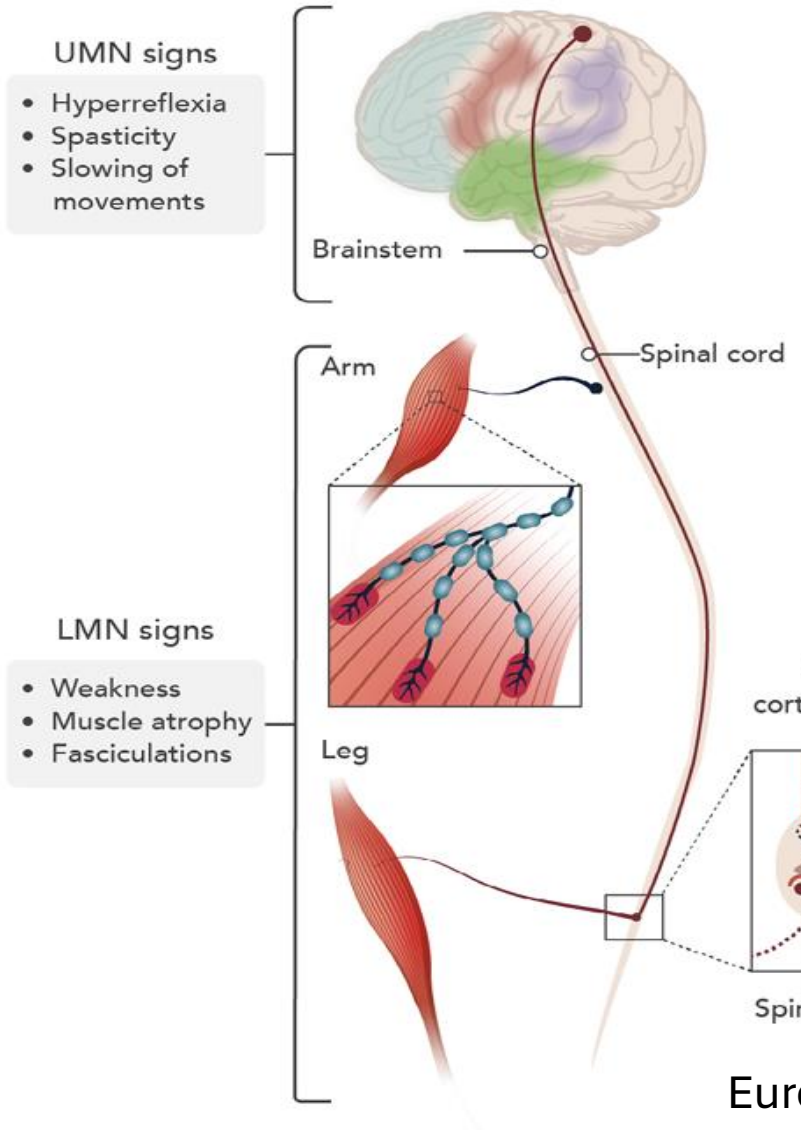
Motor features

Subtypes by regional onset

Classic ALS
- Bulbar ALS
- Spinal ALS

Specific subtypes
- Pseudobulbar palsy
- Progressive bulbar palsy
- Mill's syndrome (hemiplegic)
- Respiratory ALS
- Axial ALS
- Flail arm syndrome
- Flail leg syndrome
- Pseudopolyneuritic ALS

Subtypes by UMN vs LMN involvement
- Primary lateral sclerosis (PLS)
- UMN predominant ALS
- ALS
- LMN predominant ALS
- Progressive muscular atrophy (PMA)



Cognitive features

- Behavioral variant (bvFTD)
- Primary progressive aphasia (PPA)
  - Progressive nonfluent aphasia (naPPA)
  - Sematic dementia (svPPA)
  - Logopenic progressive aphasia (lvPPA)

EuropeanJournalof Neurology2020,27:1918 1929

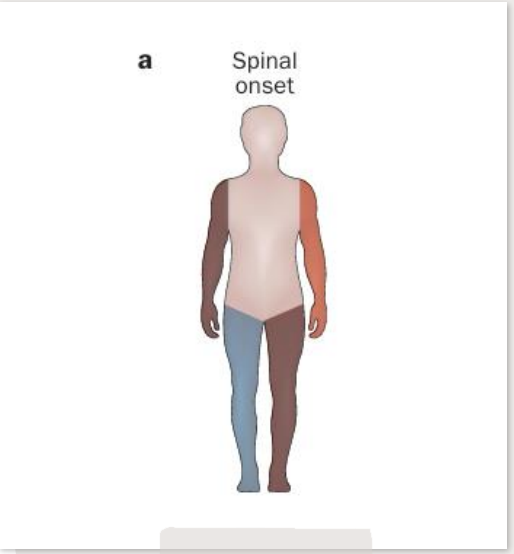
Presentation of ALS (Variability in location of onset)

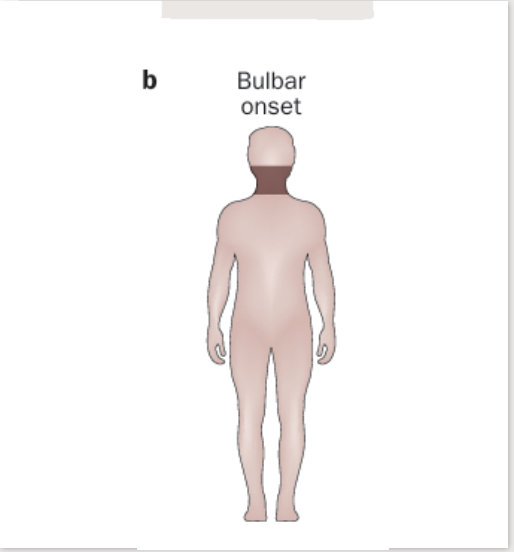
Spinal onset ALS (65%)

- Asymmetric, painless weakness and atrophy occurring, often in the dominant limb, before spreading to the contralateral limb.
- Medium survival ~ 3-5 years.

Bulbar onset ALS(25%) ~ progressive bulbar palsy

- Dysarthria, dysphagia and tongue fasciculations
- Pseudobulbar affect (an inaccurate term mainly referring to uncontrolled crying or laughing)
- More common in women and elderly
- Worse prognosis than patients with spinal onset, with a mean survival of 2 years.





Presentation of ALS (Variability in location of onset)

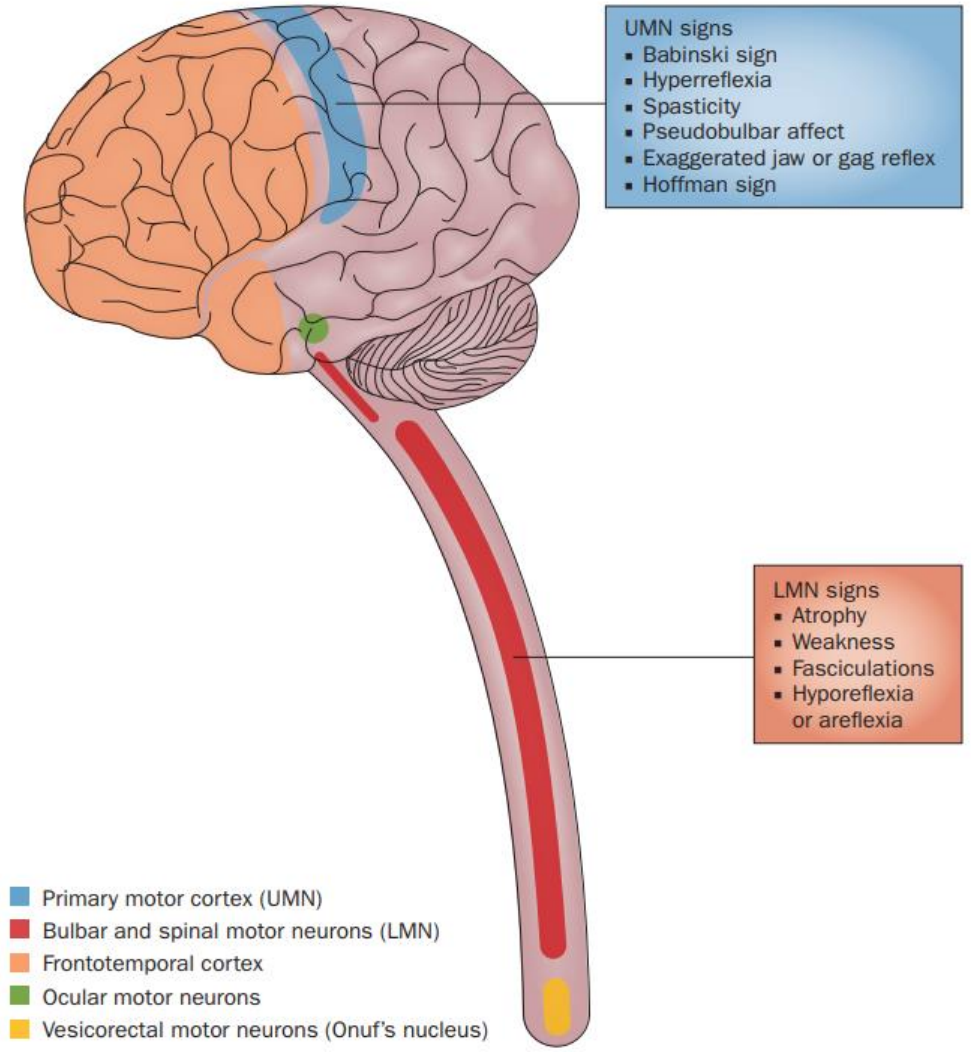
Respiratory onset(<5%)

- characterized by orthopnea or dyspnea(due to diaphragm weakness) , and mild or even absent spinal or bulbar signs.
- The patients with respiratory onset have a notoriously poor prognosis. with a mean survival of 1.4 years.

Axial variant ALS

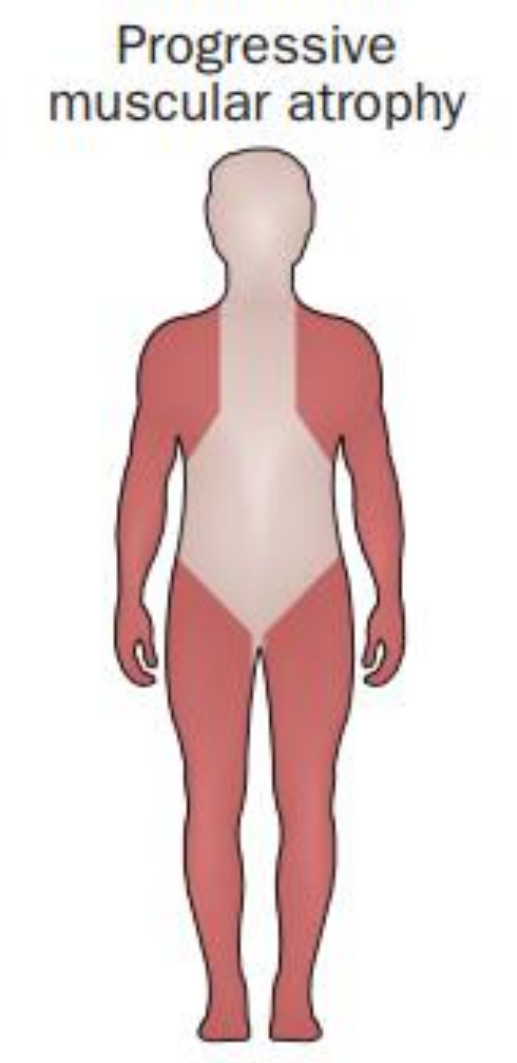
- The disease starts in paravertebral muscles, with stooped posture as a presenting symptom.
- Male predominance

Presentation of ALS (based on relative UMN versus LMN)



Swinnen, B. & Robberecht, W. Nat. Rev. Neurol. 10, 661–670 (2014)

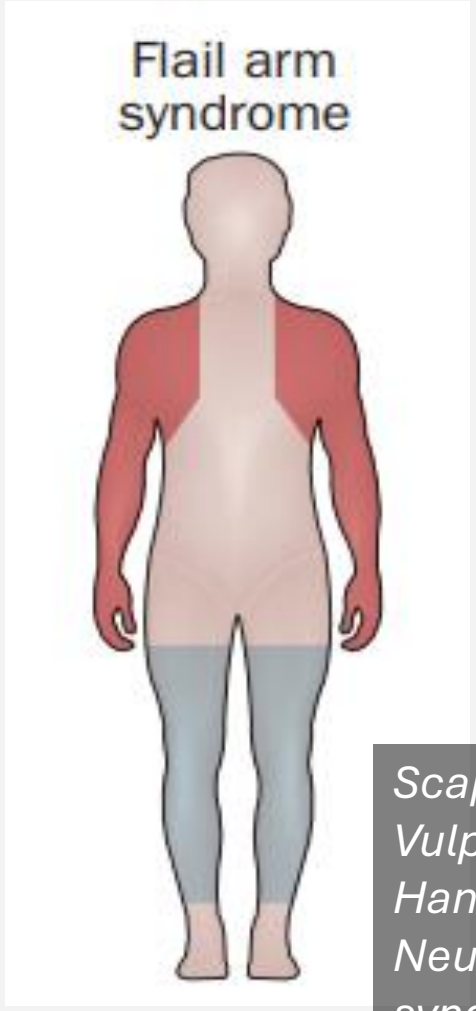
Presentation of ALS (LMN predominance)



➢PMA(5%)

- Typical LMN symptoms of progressive flaccid paralysis, muscle atrophy, hyporeflexia/ areflexia, and fasciculations.
- Asymmetric weakness and atrophy in the distal limbs at presentation.
- Progressive worsening LMN signs and appearance of UMN signs in 20-30 percent of the cases, usually within 5 to 10 years of the onset of the disease.
- Relatively better prognosis and slower rate of progression.

Presentation of ALS (LMN predominance)

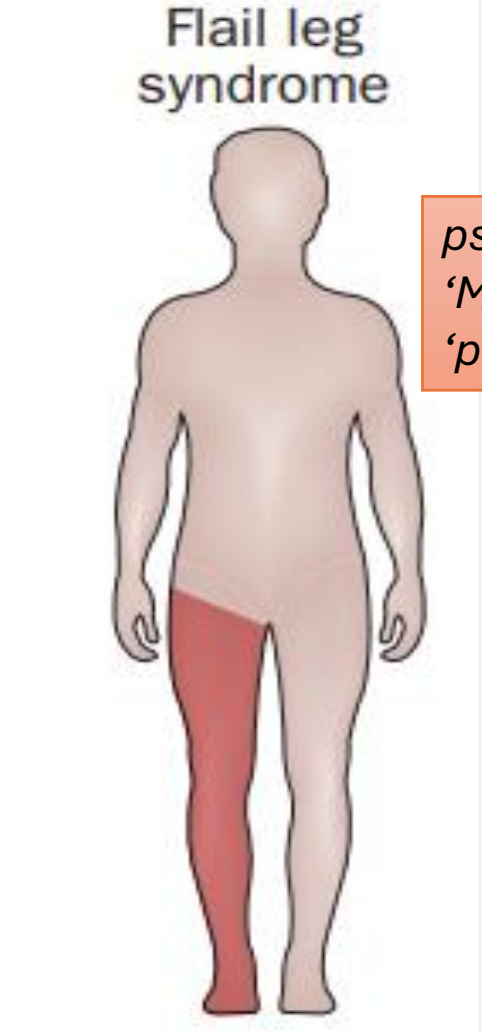


Scapulo-humeral form of ALS
Vulpian–Bernart syndrome
Hanging arm syndrome
Neurogenic man-in-a-barrel syndrome
Brachial amyotrophic diplegia

➢Flail arm syndrome

- characterized by is a progressive predominantly LMN pattern of weakness in the upper limbs, a mostly symmetrical pattern of weakness that typically begins in proximal muscles with progression to distal involvement.
- Bulbar symptoms develop in up to 77%.
- There is a high male preponderance (male to female ratio 3:1)
- Prognosis is some what better than that of classic ALS, with mean survival of 4 years, long term survival 17%.

Presentation of ALS (LMN predominance)



pseudo polyneuritic', 'Marie–Patrikios' or 'peroneal' form of ALS

➢ Flail leg syndrome

- characterized by progressive, asymmetrical, predominantly LMN pattern of weakness with distal-onset weakness and wasting of the lower limbs.
- Progression is slightly slower compared to classic ALS.

➢ Dropped head syndrome

- onset in the cervical region, limited to the extensors of the neck
- needs to be differentiated from myasthenia gravis or a (typically inflammatory) myopathy

Presentation of ALS (UMN predominance)

➢PLS (3-5%)

- Primary lateral sclerosis (PLS) is characterized by progressive spasticity and slowing of movements with isolated UMN signs on clinical examination.
- Slower progression, median survival of PLS patients is more than 20 years.
- But can evolve into ALS, typically within 3–4 years after disease onset.

Presentation of ALS (UMN predominance)

Mills syndrome or progressive hemiplegia

➢ Hemiplegic variant

- Extreme rare
- usually begins with unilateral upper motor neuron involvement in the lower limb, followed by slowly progressive ipsilateral involvement of the arm, with relative sparing of the face.
- After a variable time period, the disease spreads to the initially unaffected side.

Non-motor involvement in ALS/MND

Cognitive impairment

- Up to half of those with ALS develop some cognitive impairment.
- Onset of cognitive problems usually precedes that of motor dysfunction.
- The commonest cognitive and behavioural abnormalities are executive dysfunction and apathy, respectively, both being associated with poorer survival.
- 25% of patients with ALS meet all criteria for the clinical manifestation of FTLD, frontotemporal dementia (FTD), mostly of the behavioural variant.

MND mimics

LMN

- Benign fasciculations
- Multifocal motor neuropathy with conduction block
- Motor-predominant CIDP
- Neuralgic amyotrophy
- Kennedy's syndrome (spinobulbar muscular atrophy)
- Hirayama disease
- Inclusion body myositis

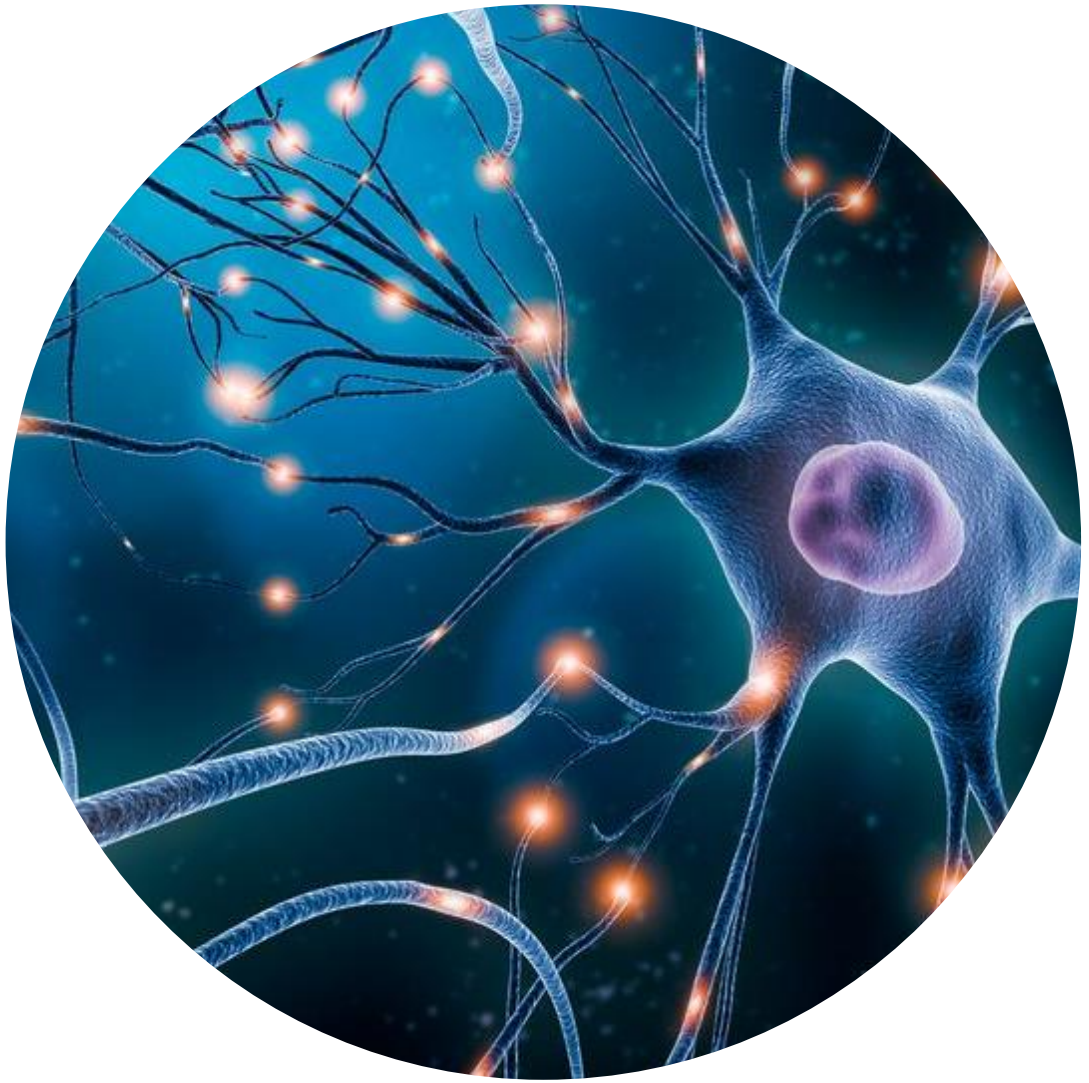
UMN

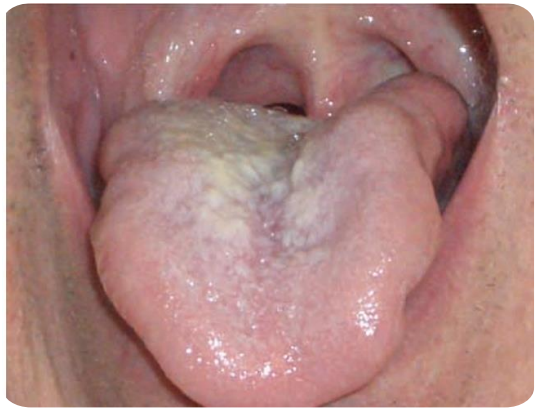
- Hereditary spastic paraparesis
- Primary progressive multiple sclerosis

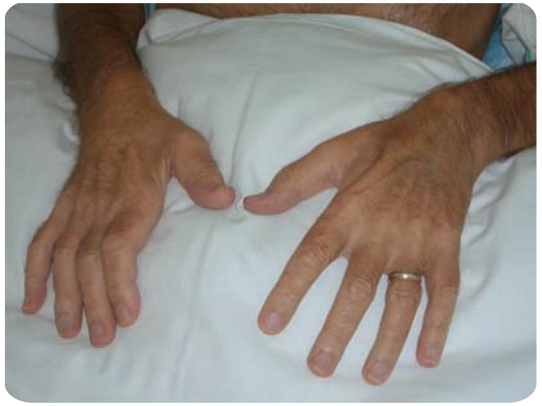
Mixed

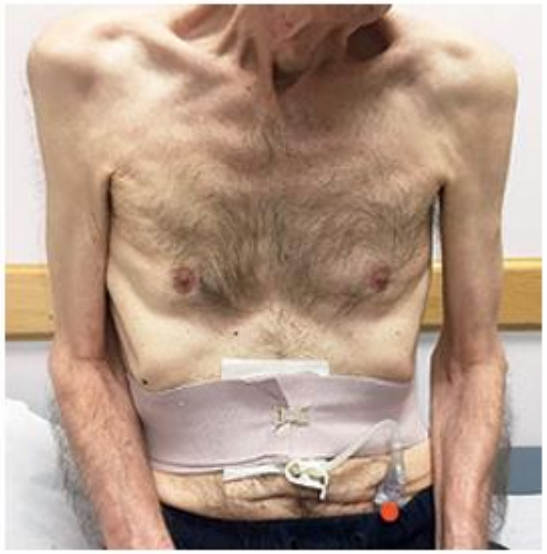
- Cervical myeloradiculopathy
- Syringomyelia/bulbia

Diagnosis of MND









N Engl J Med 2017;377:162-72

Diagnosis

- Diagnosis of MND was mainly based on clinical findings with support of electrophysiological, imaging and laboratory techniques to exclude other diseases.

mnda motor neurone disease association

Royal College of General Practitioners

Painless, progressive weakness – Could this be Motor Neurone Disease?

1. Does the patient have one or more of these symptoms?

Bulbar features

- Dysarthria
  - Slurred or quiet speech often when tired
- Swallowing difficulties
  - Liquids and/or solids
  - Excessive saliva
  - Choking sensation especially when lying flat
- Tongue fasciculations

Limb features

- Focal weakness
- Falls/trips – from foot drop
- Loss of dexterity
- Muscle wasting
- Muscle twitching/ fasciculations
- Cramps
- No sensory features

Respiratory features

- Hard to explain respiratory symptoms
- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache
- Orthopnoea

Cognitive features (rare)

- Behavioural change
- Emotional lability (not related to dementia)
- Fronto-temporal dementia

2. Is there progression?

Supporting factors

- Asymmetrical features
- Age – MND can present at any age
- Positive family history of MND or other neurodegenerative disease

Factors NOT supportive of MND diagnosis

- Bladder / bowel involvement
- Prominent sensory symptoms
- Double vision / Ptosis
- Improving symptoms

If yes to 1 and 2 query MND and refer to Neurology

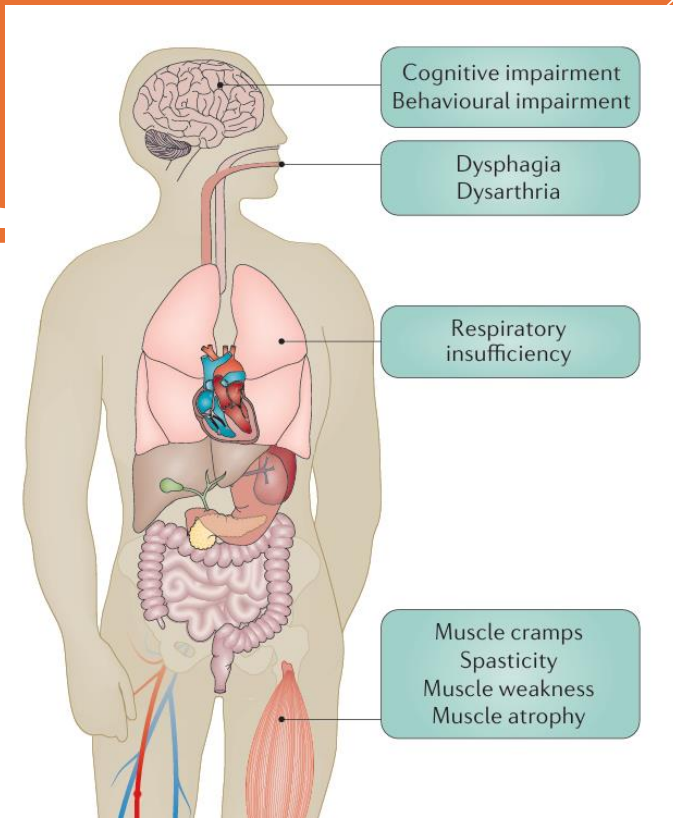
If you think it might be MND please state explicitly in the referral letter.
Common causes of delay are initial referral to ENT or Orthopaedic services.

When will you suspect MND?

Diagnostic pointers in primary care

- Asymmetrical distal weakness
- Brisk reflexes in a wasted limb
- Absence of major sensory symptoms, pain, and bladder dysfunction
- Slurring of the speech, caused by impaired tongue movement, which may be accompanied by obvious wasting and fasciculation of the tongue
- Relentless progression of symptoms and signs during follow-up period

How to diagnose MND?



History

- Painless progressive asymmetric weakness
- Initially localized- Limb(60%, typically asymmetric), Bulbar(30%, voice changes, swallowing), early behavior changes(10%)
- Absence of sensory symptoms

Examination

- Presence of UMN and LMN signs- wasted tongue and brisk jaw jerk, muscle atrophy with hyperreflexia
- Muscle wasting- focal, but beyond one nerve root
- Split hand sign (95% specificity)
- Widespread fasciculation- more obvious proximally
- Non-motor findings- apathy, disinhibition, emotional lability

Investigations

- Laboratory investigations
- Neuroimaging
- Neurophysiology (NCS/EMG)

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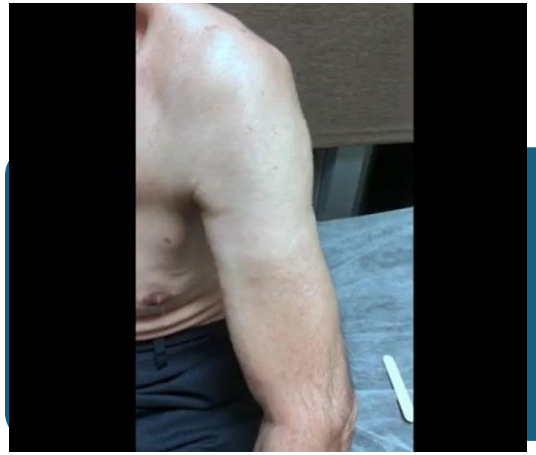
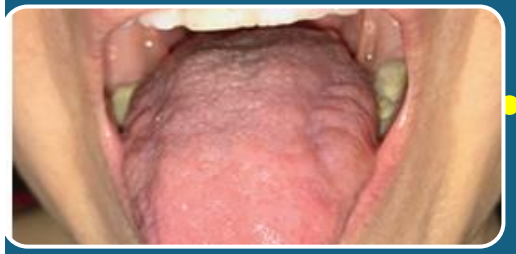
Signs with a high positive predictive value* for motor neurone disease where there is a history of progressive motor-only weakness



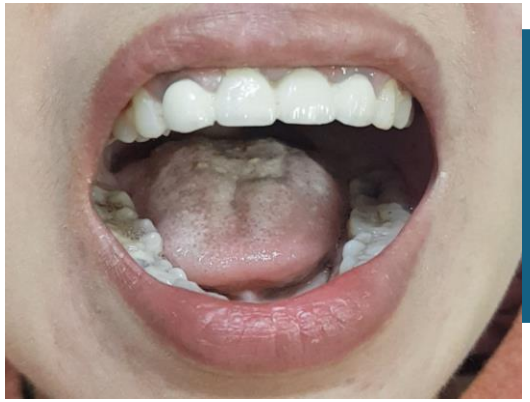
Generalized Fasciculations

easily missed over the anterior shoulders



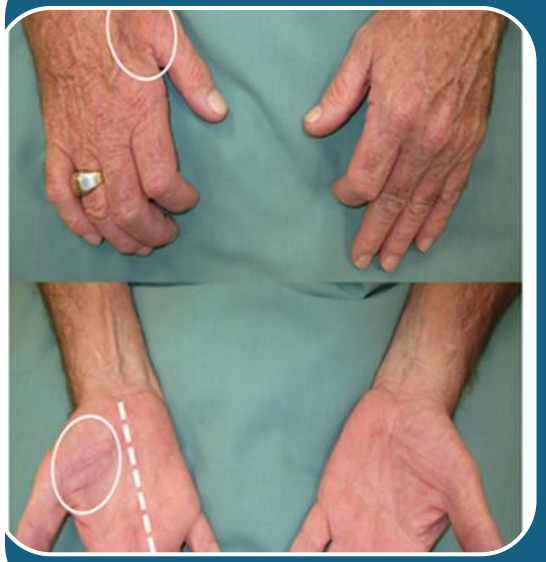
Bilateral wasting of the tongue

- Lateral borders, particularly where there are also fasciculations (best observed without protrusion) and a brisk jaw jerk or orbicularis oris reflexes



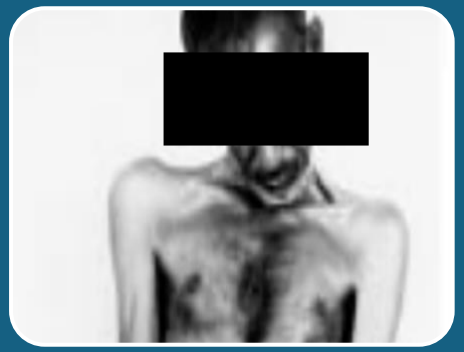
Tongue wasting and fasciculation

Signs with a high positive predictive value* for motor neurone disease where there is a history of progressive motor-only weakness



The ‘split hand’

Preferential wasting of the lateral border of the hand, that is, first dorsal interosseous and abductor pollicis brevis. This is thought possibly to reflect cortical organization.



Head drop

- Weakness of neck extensors.
- Myasthenia gravis is a consideration, but this sign should not be attributed to cervical spondylosis

Signs with a high positive predictive value* for motor neurone disease where there is a history of progressive motor-only weakness

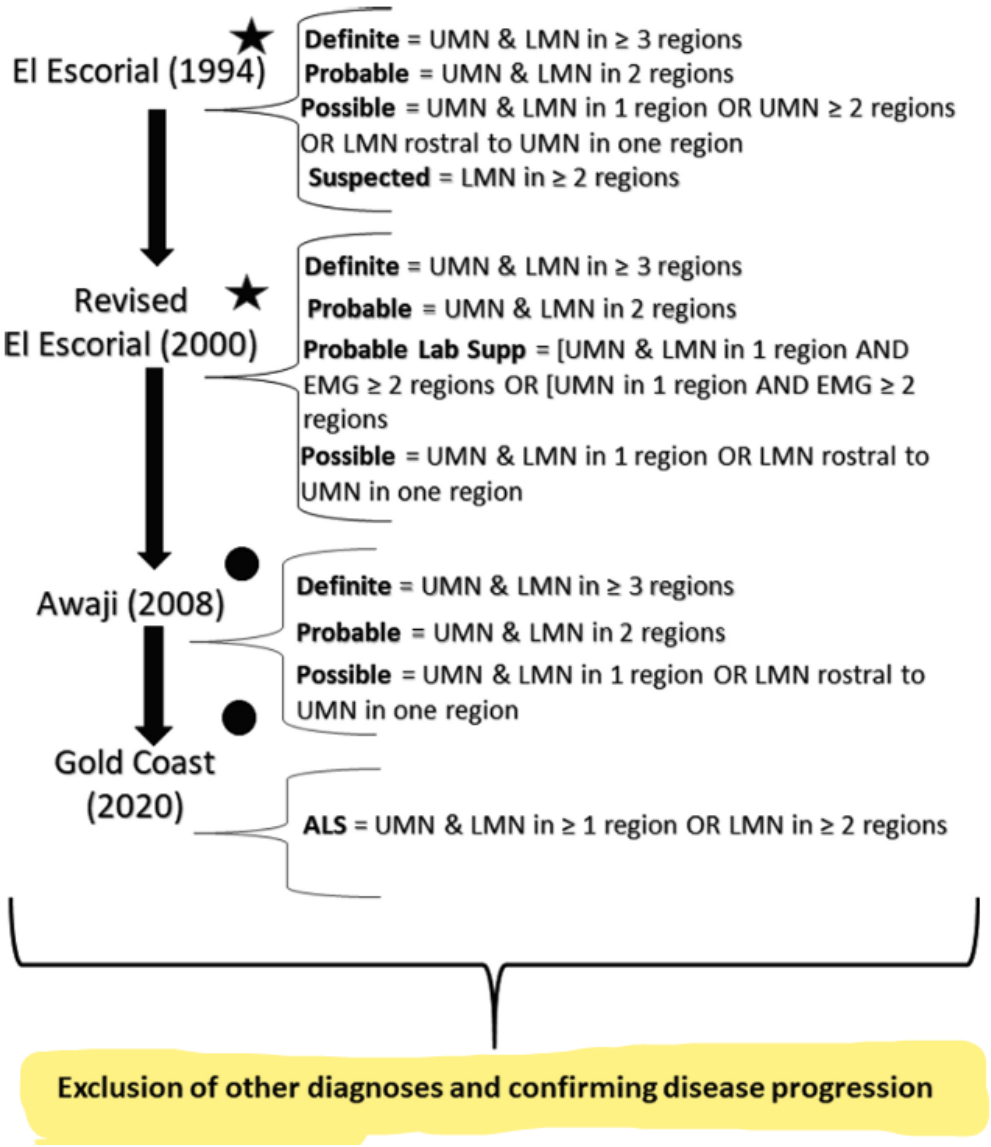
Emotionality	• Exaggerated response to emotional stimuli, usually crying, typically with bulbar weakness and often with an abnormal response to glabellar tap
Cognitive or behaviour impairment	• Frontotemporal dementia overlap features

Gold Coast criteria

1) a documented history or repeated clinical assessments that demonstrate progressive motor impairment after a period of normal motor function;
2) the presence of both upper and lower (clinical or EMG) motor neuron signs in at least one body region (or UMN and LMN dysfunction in the same body region if only one region is affected), or LMN dysfunction in at least two body regions;
3) **thorough investigations must be conducted to rule out any other potential disease process**

❖Sensitivity greater than 90% for diagnosing ALS

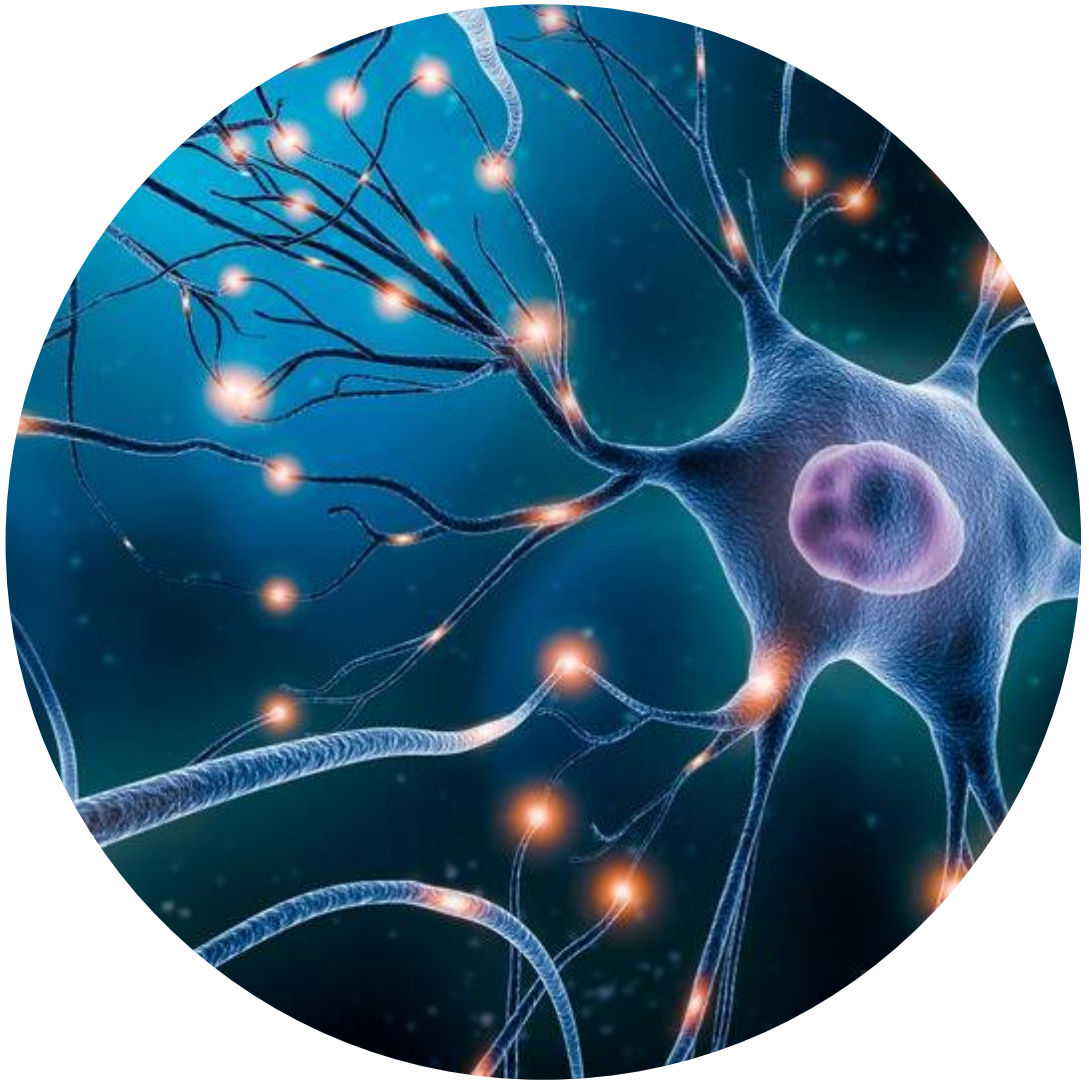
DIAGNOSTIC CRITERIA FOR ALS/MND



EMG changes in MND

- Provide the evidence of LMN dysfunction necessary to support the diagnosis of ALS and should identify at least two of the four CNS regions: brain (bulbar), neck, thoracic, or lumbosacral spinal cord (anterior horn motor neuron).
- Signs of active denervation : Fibrillation and positive sharp waves.
- Signs of chronic denervation : large motor unit action potential (MUAP) with increasing duration, polyphasic, often increasing amplitude.
- The fasciculation potential is very important as a characteristic feature of ALS especially when long duration and polyphasic features are obtained; the absence of fasciculations raises doubts but does not rule out the diagnosis, but these features in EMG are considered helpful in the diagnosis of ALS .

Case study



Case 1

- A 58-year-old male presented with 2 months history of left lower limb limping, difficulty climbing stairs, and left foot weakness. After 1 month, he noticed the weakness spreading to right leg. He was unable to stand from sitting without using his arms for support, nor able to walk unassisted.
- After 3 months, he noticed difficulty using his left hand, loss of strength and began troubling his day-to-day activities. He also noted 'muscle twitching' occasionally in thigh and arms.
- He subsequently noticed slurring of speech, but there was no swallowing difficulty nor shortness in breath.
- He has no sensory symptoms nor sphincter dysfunction. He has no known past medical history.
- He was apparently normal before the onset of his symptoms.

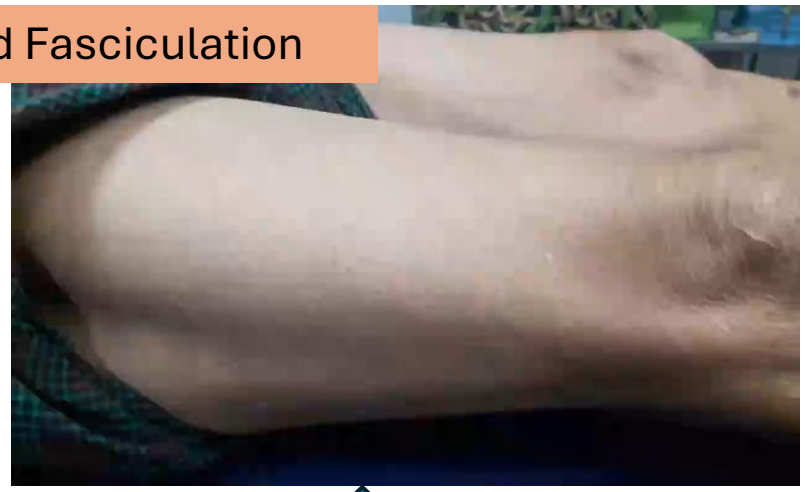
Case 1

Neurological examination

- Higher mental function- Intact
- Sensory, cranial nerves and cerebellar examination – Normal

A

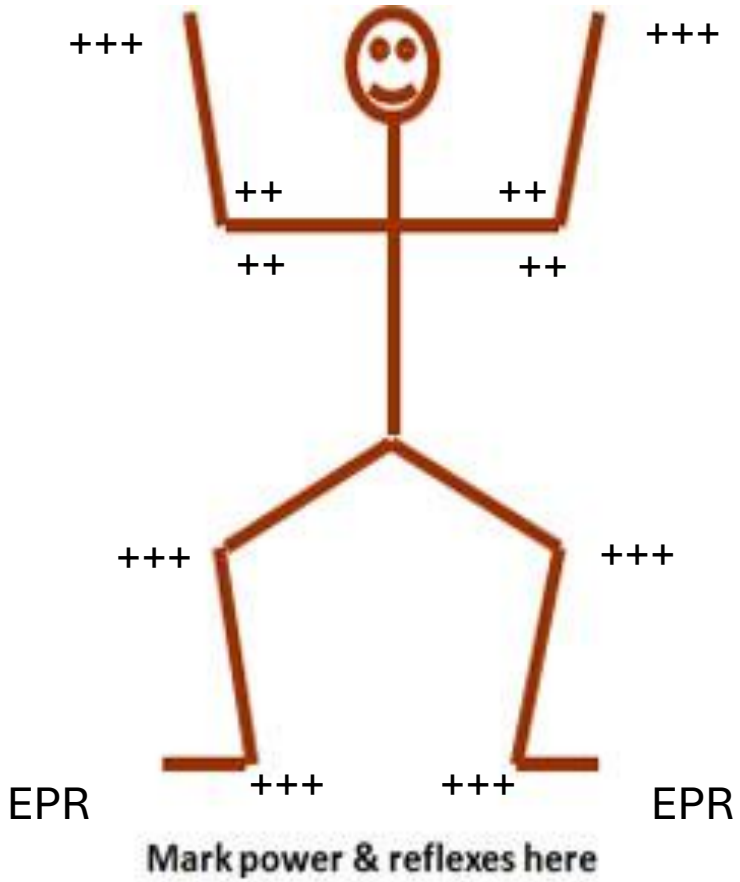
B

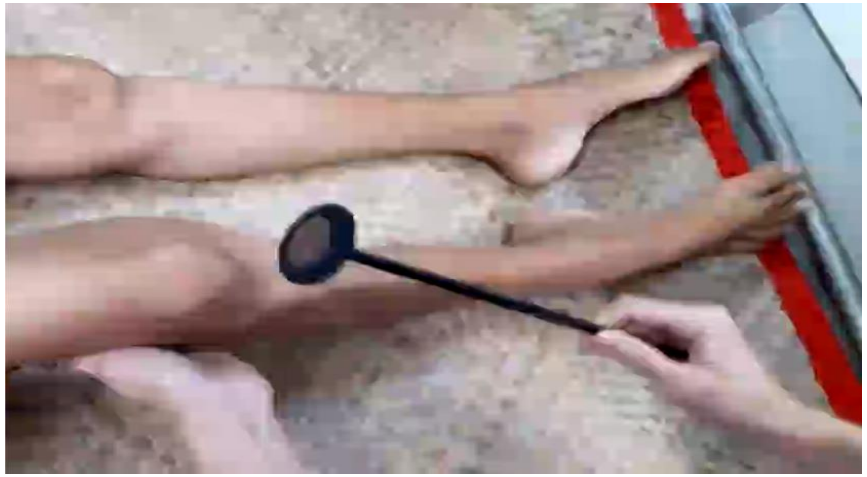


C

UL	Rt	Lt	**LL**	Rt	Lt
Shoulder	5	4	Hip	4	4
Elbow	5	3	Knee	4	4
Wrist/fingers	4	2	Ankle	3	3

Case 1





MND mimics

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- Motor-predominant CIDP
- Neuralgic amyotrophy
- Kennedy's syndrome (spinobulbar muscular atrophy)
- Hirayama disease
- Inclusion body myositis

UMN

- Hereditary spastic paraparesis
- Primary progressive multiple sclerosis

Mixed

- Cervical myeloradiculopathy
- Syringomyelia/bulbia

Case 1 Investigations

- MRI(brain and cervical spine)- unremarkable
- NCS is normal.
- EMG shows fasciculations and chronic reinnervation in bulbar, cervical, thoracic and lumbosacral segments.

EMG

Side	Muscle	Nerve	Root	Ins Act	Fibs	Psw	Amp	Dur	Poly	Recrt	Int Pat	Comment
Right	Rectus Abdom	Intercostals	T6-12	Nml	Nml	Nml	**3+**	**3+**	**3+**	**Reduced**	Nml	
Right	1stDorInt	Ulnar	C8-T1	**Incr**	Nml	Nml	**3+**	**3+**	**3+**	**Reduced**	Nml	fasciculation
Right	Biceps	Musculocut	C5-6	Nml	Nml	Nml	**1+**	**1+**	**1+**	**Reduced**	Nml	
Right	SternoMast	SpinAcc	CN XI, C2-3	Nml	Nml	Nml	**1+**	**1+**	**1+**	**Reduced**	Nml	
Right	AntTibialis	Dp Br Fibular	L4-5	Nml	Nml	Nml	**3+**	**3+**	**3+**	**Reduced**	Nml	
Right	VastusMed	Femoral	L2-4	**Incr**	Nml	Nml	**2+**	**3+**	**3+**	**Reduced**	Nml	fasciculation

Pure motor distal asymmetric weakness and wasting of hands and feet, rapidly progressive in 6 months

Presence of both upper and lower motor neuron signs (clinical + EMG) in 4 regions

Exclusion of other diagnosis

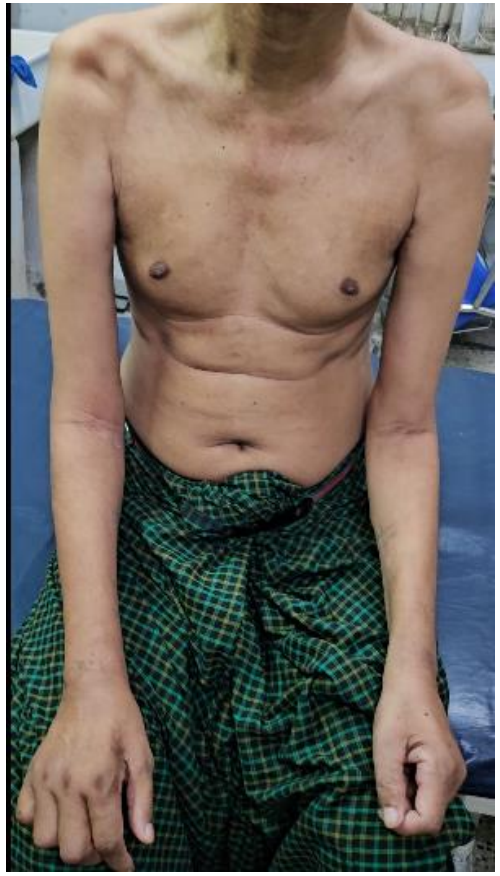
Classical spinal onset ALS

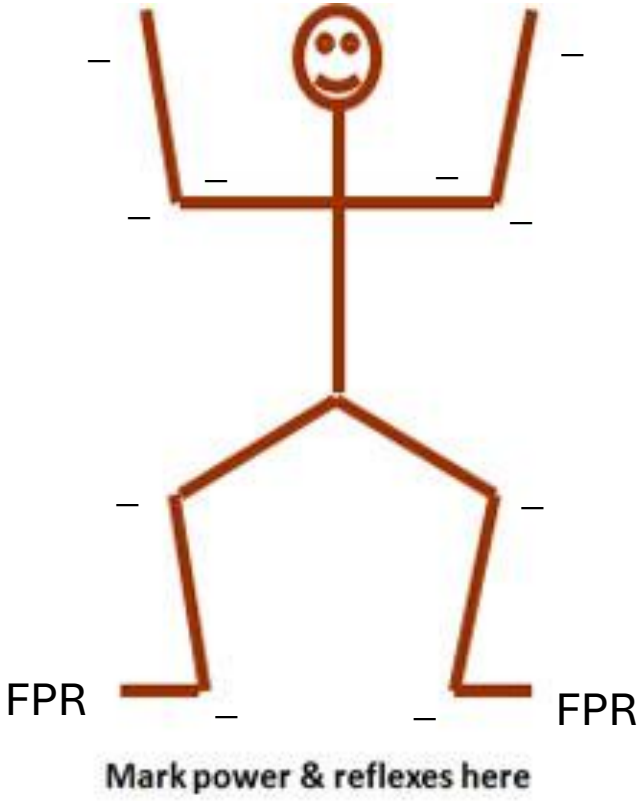
Case 2

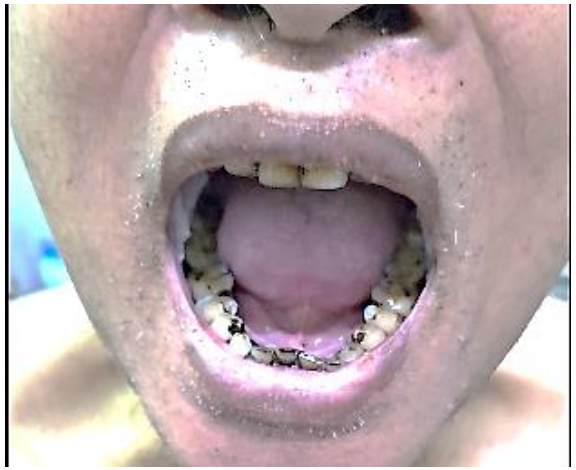
- 50-year-old man presented with difficult in lifting arms for 3 months followed by difficult standing from squatting.
- There was no symptoms of tingling and numbness apart from aching pain in thigh.
- Weakness progressed over 1 year and he encountered difficultly in using his hands and problem doing daily activities.
- Then he needs assistant in walking for long distance.
- No swallowing difficulty and slurring of speech
- No problem with bladder and bowel dysfunction.
- Apart from mild hypertension, there was no previous medical illnesses.
- No chronic exposure to fertilizers nor insecticide.

Case 2

Neurological examination



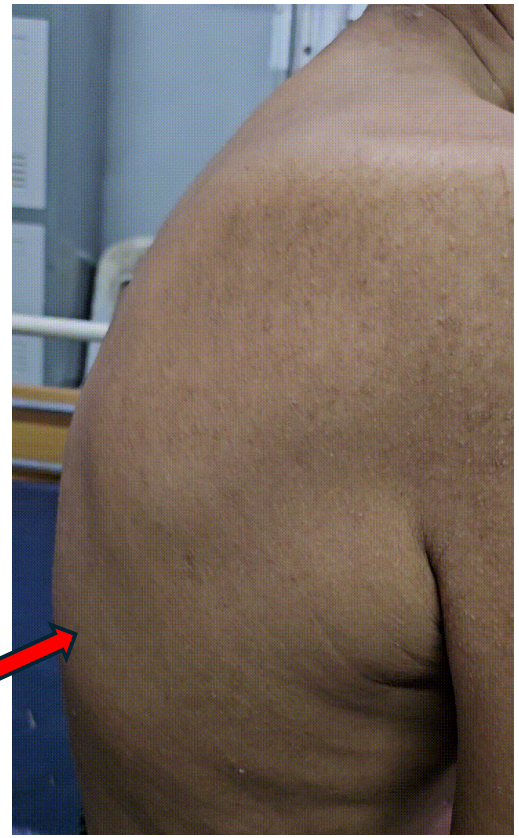




UL	Rt	Lt	LL	Rt	Lt
Shoulder	1	1	Hip	3	3
Elbow	2	2	Knee	4	4
Wrist/fingers	3	3	Ankle	2	2

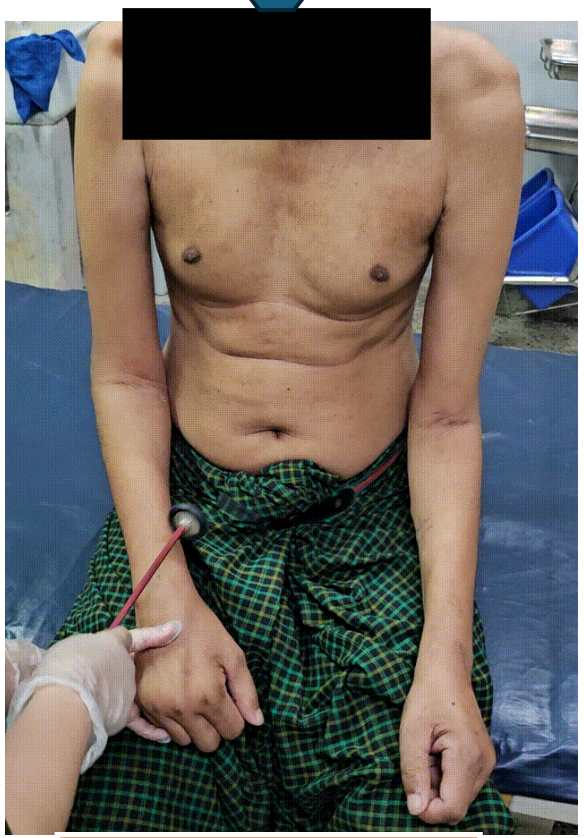
Case 2

A



Fasciculation

B



Areflexia

C

Hanging arms

MND mimics

LMN

- Benign fasciculations
- Multifocal motor neuropathy with conduction block
- Motor-predominant CIDP
- Neuralgic amyotrophy
- Kennedy's syndrome (spinobulbar muscular atrophy)
- Hirayama disease
- Inclusion body myositis

UMN

- Hereditary spastic paraparesis
- Primary progressive multiple sclerosis

Mixed

- Cervical myeloradiculopathy
- Syringomyelia/bulbia

Case 2

Investigations

Laboratory

- Creatine kinase- not raised

MRI (Cervical spine)

- OA changes of upper cervical vertebra, no cord compression nor spinal stenosis

NCS

- Normal sensory potentials, low normal CMAP, no conduction block

EMG

- Widespread active denervation and chronic reinnervation changes in cervical and lumbar region

Side	Muscle	Nerve	Root	Ins Act	Fibs	Psw	Amp	Dur	Poly	Recrt	Int Pat	Comment
Right	1stDorInt	Ulnar	C8-T1	Incr	3+	3+	1+	Nml	0	Reduce	Nml	
Right	Biceps	Musculocut	C5-6	Nml	1+	1+						no effort
Right	AntTibialis	Dp Br Fibular	L4-5	Nml	2+	3+	1+	1+	1+	Reduce	Nml	

Slowly progressive symmetrical motor weakness in proximal muscles of UL followed by distal weakness

Normal sensory NCS
Presence of lower motor neuron signs (clinical + EMG) in 2 regions

Exclusion of other diagnosis

Flail arm syndrome
(LMN variant of ALS)

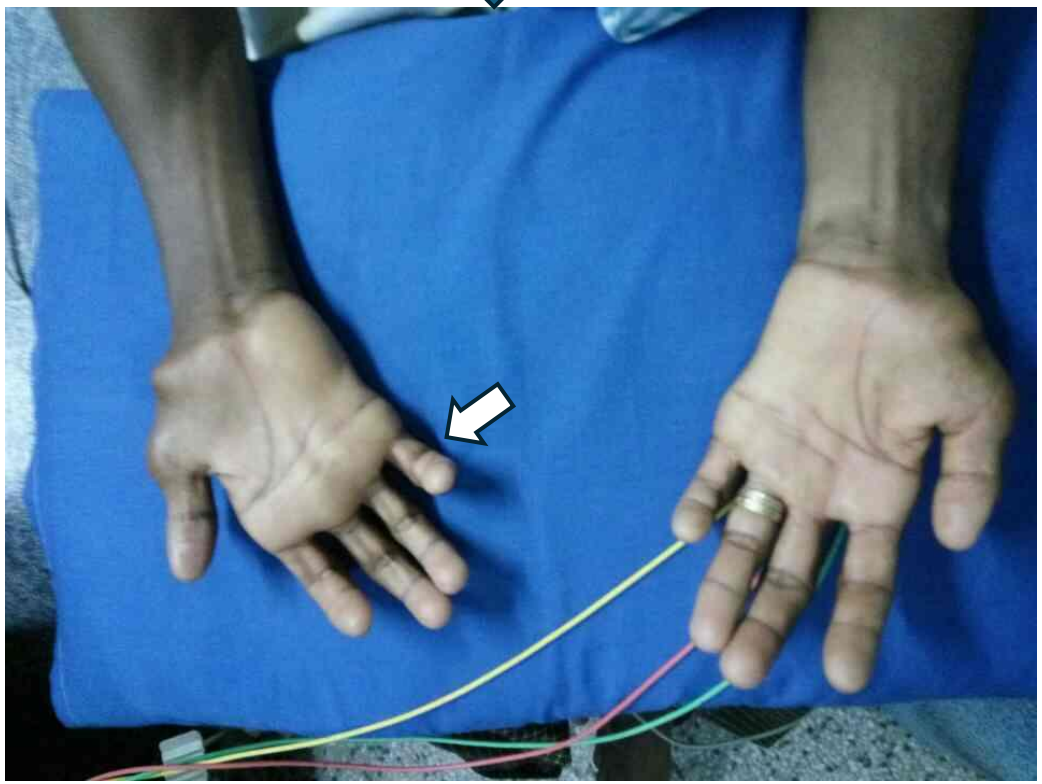
Case 3

- A 22-year-old man gentleman came with history of insidious onset of weakness in both the hands for 4 years duration.
- Weakness started in the right-hand muscles which was gradually progressed to the forearm within 6 months.
- He did not have any pain, loss of sensation, diplopia, dysphagia, ptosis, muscle cramps, fasciculations, headache or neck pain.
- There was no history of trauma, febrile illness, poliomyelitis or exposure to toxins or heavy metals in the past.
- There was no family history of similar complaints or neuromuscular disease

Case 3 Neurological examination

-Clawing of 5th finger on right (white arrow)

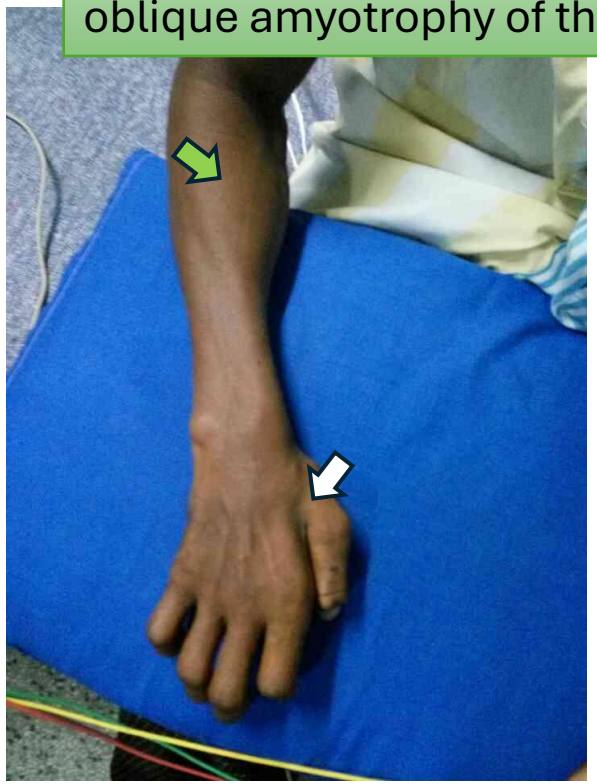
-Thenar & hypothenar muscle wasting and atrophy , no fasciculation

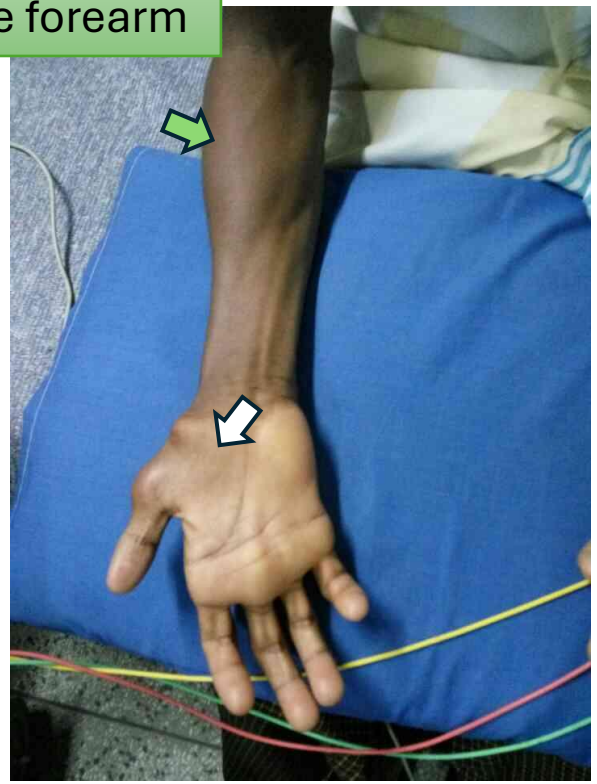


-Wasting of first dorsal interosseous and other small muscles of the hand (white arrow)

-Wasting of forearm with relative preservation of brachioradialis muscles (green arrow)

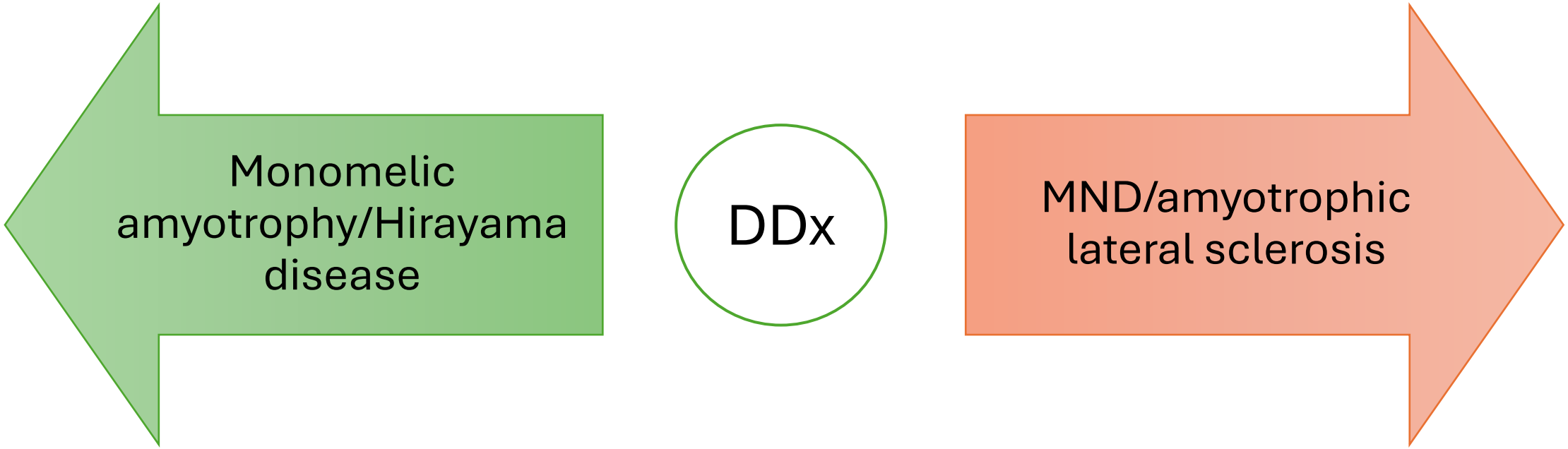
oblique amyotrophy of the forearm





Case 3

Slowly progressive weakness and atrophy of right hand and forearm without sensory symptoms

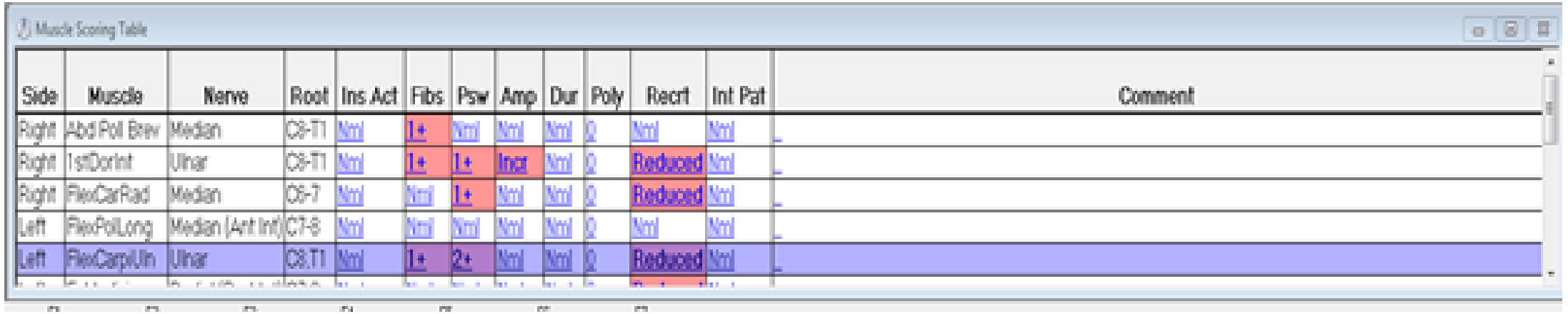


Case 3 Neurophysiology

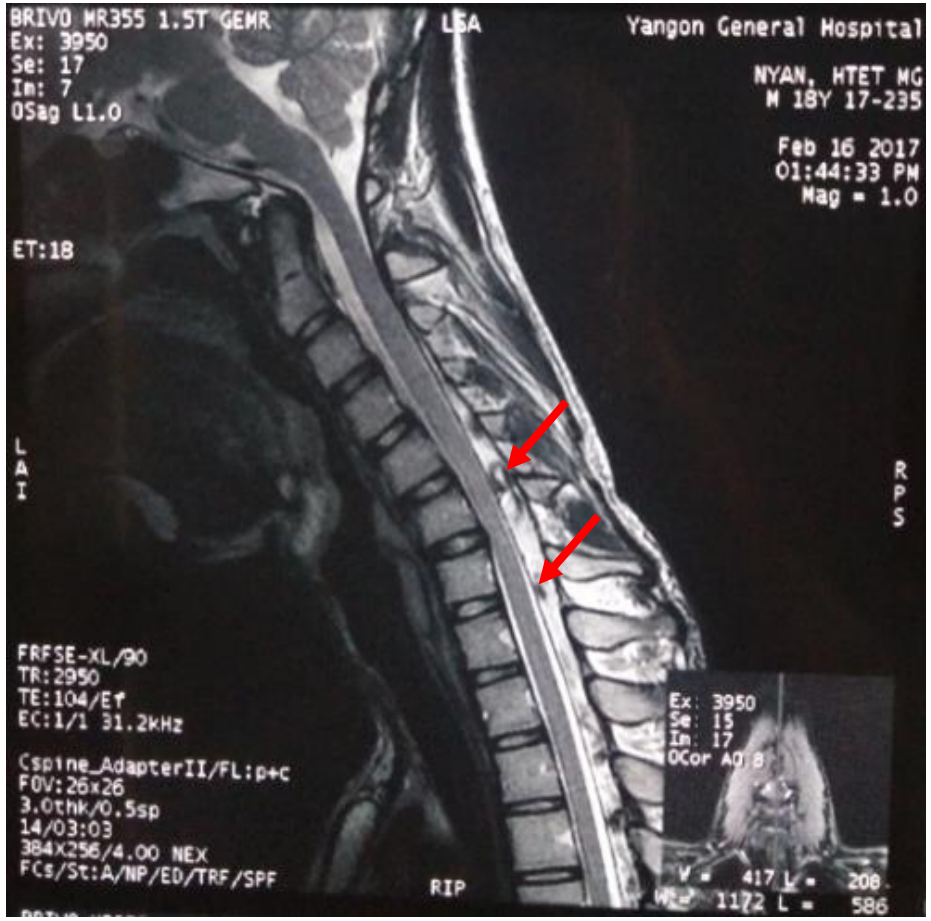
NCS – normal

EMG

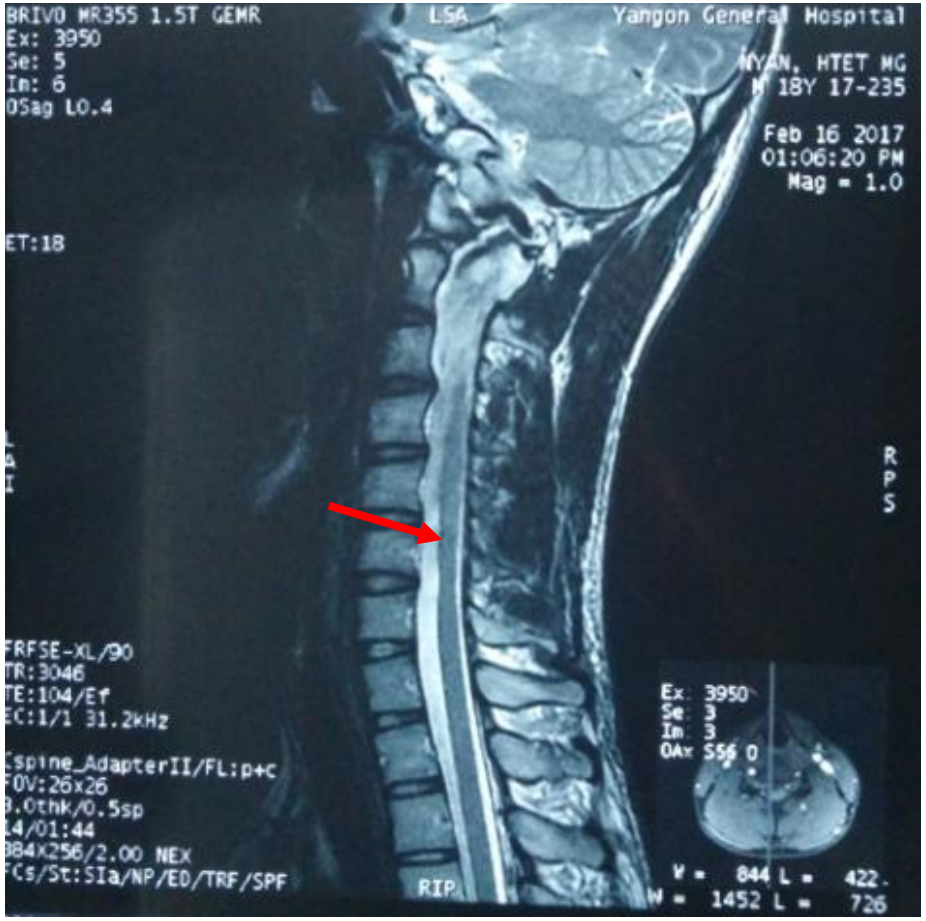
- Active denervative changes(fibrillations and positive sharp waves, ↑amplitude in motor unit potentials (MUPs)).
- Chronic renervative changes(reduced recruitment & and polyphasia) in the right upper limb.



Case 3 MRI (Flexion –Extension cervical spine)



Posterior epidural space is widened in flexion sequences. Epidural flow voids.



Thinning of cervical cord(cord atrophy)
Anterior displacement of the posterior dura from C3 to T1 levels

Hirayama's Disease

Monomelic amyotrophy (MMA), Juvenile non progressive amyotrophy, Sobue disease

- Hirayama's disease is a rare benign disorder.
- It is a focal, lower motor neuron type of disease.
- Mainly young males in their second and third decades of age are most affected.
- It is seen mostly in Asian countries like India and Japan.
- In majority of people cause of this disease is unknown.
- MRI of cervical spine in flexion will reveal the cardinal features of Hirayama disease.
- Early diagnosis is necessary as the use of a simple cervical collar which will prevent neck flexion, has been shown to stop the progression.

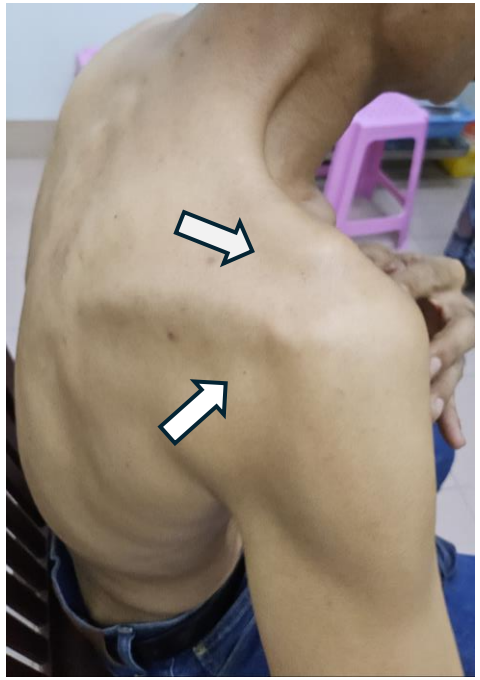
	Hirayama's disease	MND
Age	Young	Older
Onset	Wasting start in one hand Oblique amyotrophy	Wasting start in one hand
Disease progression	Slow	Rapid
Course	Arrest	Progressively worsen
EMG	Active denervation & chronic renervation	Active denervation & chronic renervation
MRI flexion sequence	Posterior epidural space widen	
Prognosis	Good	Poor

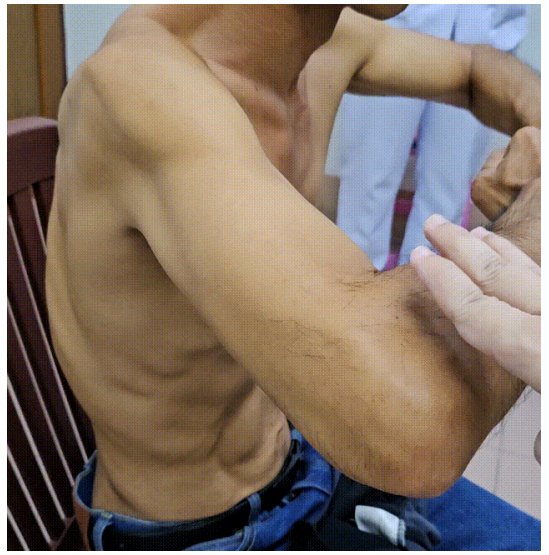
Case 4

- 37-year-old man presented with acute onset of throbbing pain in right scapular and lateral shoulder areas, extending to right side of neck.
- Pain was very severe and intolerable, worsened in the supine position, preventing sleep, but were slightly relieved when sitting or standing.
- Pain was not relieved by NSAIDs and significantly impact daily life and work.
- No fever, headache, tingling and numbness in the limbs
- No problem with walking, speech and swallowing.
- Three weeks after the onset, the patient experienced a decrease in strength in right shoulder joint abduction and external rotation.
- At 8 weeks post-onset, pain significantly reduced, not affecting sleep or work, but the patient still experienced right upper limb weakness.

Case 4 Neurological examination

- Marked atrophy of Rt scapular region
- Right shoulder abduction and external rotation strength rated at 4.
- No abnormalities observed in the strength of elbow flexion, extension, or grip.
- No abnormalities in the sensation of the rt scapular area and upper limb.
- Bilateral biceps tendon reflex (++) bilateral triceps tendon reflex (++)
- Negative Hoffmann's sign.

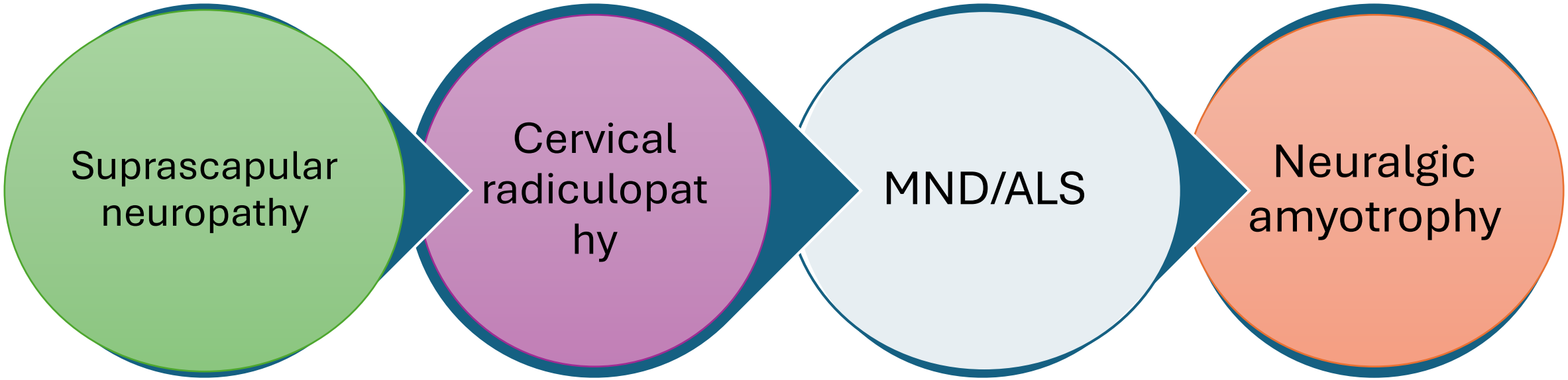




- No tongue wasting, fasciculation
- Other neurological examination was normal.

Case 4

Acute onset severe pain of right scapula followed by wasting and weakness of right upper arm



Case 4 Investigations

MRI(C spine and brachial plexus)

- protrusion of the C4/5 and C5/6 intervertebral discs.

NCS

- normal

EMG

- chronic moderate Rt C5 radiculopathy and mild S1 radiculopathy

Motor Summary Table

Stim Site	NR	Onset (ms)	Norm Onset (ms)	O-P Amp (mV)	Norm O-P Amp	Neg Dur (ms)	Full Dur (ms)	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Left Median Motor (Abd Poll Brev)													
Wrist		3.0	<4.2	9.7	>5	5.70	8.05	Elbow	Wrist	3.9	23.0	59	>50
Elbow		6.9		9.3		5.55	35.08						
Right Median Motor (Abd Poll Brev)													
Wrist		2.9	<4.2	9.7	>5	6.02	26.64	Elbow	Wrist	4.1	24.0	59	>50
Elbow		7.0		9.2		6.02	15.78						
Left Ulnar Motor (Abd Dig Minimi)													
Wrist		2.3	<4.2	7.1	>3	6.09	26.25	B Elbow	Wrist	3.9	23.0	59	>50
B Elbow		6.2		6.9		6.02	18.28	A Elbow	B Elbow	1.8	10.0	56	>53
A Elbow		8.0		6.8		6.02	18.36						
Right Ulnar Motor (Abd Dig Minimi)													
Wrist		2.1	<4.2	7.6	>3	7.11	21.80	B Elbow	Wrist	3.8	23.0	61	>50
B Elbow		5.9		7.0		6.48	22.42	A Elbow	B Elbow	1.6	10.0	63	>53
A Elbow		7.5		6.3		6.88	27.27						
Left Proneal (Fibular) Motor (Ext Dig Brev)													
Ankle		4.6	<6.1	3.3	>2.5	7.97	12.50	B Fib	Ankle	5.9	31.0	53	>38
B Fib		10.5		3.0		8.44	31.72	Poplt	B Fib	1.7	10.0	59	>40
Poplt		12.2		2.9		8.20	29.14						
Right Proneal (Fibular) Motor (Ext Dig Brev)													
Ankle		4.5	<6.1	3.9	>2.5	7.89	38.05	B Fib	Ankle	6.0	31.0	52	>38
B Fib		10.5		3.7		8.44	33.05	Poplt	B Fib	2.0	10.0	50	>40
Poplt		12.5		3.6		8.05	31.88						
Left Radial Motor (Ext Ind Prop)													
8cm		1.6	<2.5	3.5	>1.7	7.66	32.50	Up Arm	8cm	3.1	20.0	65	>60
Up Arm		4.7		3.3		7.97	34.69	Axilla	Up Arm	1.5	10.0	67	
Axilla		6.2		2.9		7.11	34.61						
Right Radial Motor (Ext Ind Prop)													
8cm		1.7	<2.5	3.2	>1.7	8.36	22.19	Up Arm	8cm	3.3	20.0	61	>60
Up Arm		5.0		3.2		8.75	19.92	Axilla	Up Arm	1.5	10.0	67	
Axilla		6.5		2.9		8.75	37.19						
Left Tibial Motor (Abd Hall Brev)													
Ankle		4.4	<6.1	16.3	>3.0	6.02	39.92	Knee	Ankle	8.1	39.0	48	>35
Knee		12.5		15.4		6.88	13.67						
Right Tibial Motor (Abd Hall Brev)													
Ankle		4.6	<6.1	16.4	>3.0	5.86	37.27	Knee	Ankle	8.1	39.0	48	>35
Knee		12.7		15.3		6.64	28.52						

Side	Muscle	Nerve	Root	Ins Act	Fibs	Psw	Amp	Dur	Poly	Recrt	Int Pat
Right	1stDorInt	Ulnar	C8-T1	Nml	Nml	Nml	Nml	Nml	0	Complete	Nml
Right	PronatorTeres	Median	C6-7	Nml	Nml	Nml	Nml	Nml	0	Complete	Nml
Right	BrachioRad	Radial	C5-6	Nml	Nml	Nml	**1+**	**1+**	**1+**	**Incomplete**	Nml
Right	Biceps	Musculocut	C5-6	Nml	Nml	Nml	**2+**	**2+**	**2+**	**Reduce**	Nml
Right	Deltoid	Axillary	C5-6	Nml	Nml	Nml	**2+**	**2+**	**2+**	**Reduce**	Nml
Right	AntTibialis	Dp Br Fibular	L4-5	Nml	Nml	Nml	Nml	Nml	0	Complete	Nml
Right	Gastroc	Tibial	S1-2	Nml	Nml	Nml	**1+**	**1+**	**1+**	**Reduce**	Nml
Right	Rectus Abdom	Intercostals	T6-12	Nml	Nml	Nml	Nml	Nml	0	Complete	Nml
Right	C5 Parasp	Rami	C5	Nml	Nml	Nml					
Right	SternoMast	SpinAcc	CN XI, C2-3	Nml	Nml	Nml	Nml	Nml	0	Complete	Nml

Neuralgic amyotrophy

Idiopathic brachial plexitis/ Parsonage Turner syndrome

- Idiopathic inflammatory condition affecting the brachial plexus, characterized by the sudden onset of acute severe pain in one or both shoulders and the rapid onset of weakness in the muscles of the shoulder girdle and upper arm.
- NA is a diagnosis of exclusion.
- Usually monophasic and self-limiting with good (albeit often incomplete) recovery.

Key points

- Motor neuron disease is a devastating progressive neurodegenerative disease, which is irreversible.
- Consider the diagnosis when faced with progressive painless weakness in patients over the age of 50.
- Weak and wasted muscles with retained reflexes is highly suggestive of MND until proven otherwise.
- Cognitive impairment is a common feature of ALS.
- There is no definitive test to confirm MND, but investigations may aide in diagnosis and assist in the exclusion of other conditions.
- Lower motor neurone-predominant monomelic clinical sub-types of MND present the greatest diagnostic challenge but are frequently more slowly progressive.
- The prompt referral of such patients to specialist neurological services for assessment is thus important.

Thank you for attention