NERVE CONDUCTION STUDIES AND ELECTROMYOGRAPHY:

DIAGNOSTIC APPROACH WITHIN THE SCOPE OF CURRENT GUIDELINES 2020-2024

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IN ACCORDANCE WITH CURRENT GUIDELINES 2020-2024

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BİDGE Yayınları

Nerve Conduction Studies and Electromyography: Diagnostic Approach within the Scope of Current Guidelines 2020-2024

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EMG Pocket Companion

2026 EDITION

EMG
Pocket Companion

Featuring
EAN/PNS CIDP 2021 • EAN/PNS GBS 2023
Gold Coast Criteria 2020 • ENMC NMJ 2024
AANEM updates • ESTEEM consensus

Preface

This pocket companion distills the principles and clinical practice of electromyography (EMG) and nerve conduction studies (NCS) into a portable, evidence-based reference. It is written for neurology residents, fellows in clinical neurophysiology, and practicing electromyographers who need rapid access to current diagnostic criteria, normal values, and interpretive algorithms.

The field of electrodiagnostic medicine has evolved substantially in recent years. The 2020 Gold Coast criteria reshaped the diagnosis of amyotrophic lateral sclerosis. The 2021 European Academy of Neurology / Peripheral Nerve Society (EAN/PNS) guideline restructured chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) classification. In 2023, the same

societies issued an updated GRADE-based guideline for Guillain–Barré syndrome. The 2024 ENMC consensus refined the electrodiagnostic approach to seronegative myasthenia gravis. This volume integrates these contemporary frameworks while preserving the foundational concepts that remain essential.

The book emphasizes *pattern recognition*: each disease section pairs the clinical phenotype with characteristic electrophysiological signatures and the diagnostic criteria currently in use. Tables of normal values follow conventional reference ranges; readers should validate against their own laboratory norms. Quick-reference boxes flag pearls (💡), pitfalls (⚠), and recent updates (🔄).

Disclaimer: This book is intended as an educational reference. Clinical decisions must be individualized and made by qualified physicians integrating all relevant patient-specific data. Reference ranges and protocols vary between laboratories.

Foundations of Electrodiagnostic Medicine

1.1 What is the EMG study?

The electrodiagnostic (EDX) examination comprises two complementary techniques: **nerve conduction studies (NCS)**, which assess the integrity of peripheral nerves by stimulating and recording responses, and **needle electromyography**, which records the electrical activity of muscle fibers using an intramuscular needle electrode. Together they evaluate the entire lower motor unit — from the anterior horn cell, through the peripheral nerve and neuromuscular junction, to the muscle fiber.

Scope of EDX

The study localizes a lesion within the neuromuscular system, characterizes its pathophysiology (axonal vs demyelinating, focal vs diffuse), establishes severity and chronicity, and helps direct further investigation and management.

1.2 Common indications

Clinical question	Typical referrals
Focal weakness or numbness	Carpal tunnel syndrome, ulnar neuropathy, radiculopathy, plexopathy, peroneal palsy
Generalized weakness	Polyneuropathy, motor neuron disease, myopathy, NMJ disorder
Sensory symptoms	Distal neuropathy, small/large fiber assessment, ganglionopathy
Cramps / fasciculations	Motor neuron disease, cramp-fasciculation syndrome, neuromyotonia
Fatigability	Myasthenia gravis, Lambert-Eaton syndrome
Trauma / iatrogenic injury	Nerve transection, stretch injury, post-surgical palsy

1.3 Brief historical context

The clinical roots of EMG trace to Adrian and Bronk (1929), who first recorded single motor unit activity with a concentric needle electrode. Buchthal's work in Copenhagen during the 1950s-60s established quantitative MUAP analysis. Hodes, Larrabee and German (1948) measured motor nerve conduction velocity, while Dawson and Scott (1949) introduced sensory NCS. Modern protocols continue to refine these foundations with computerization, automated analysis, and ultrasound guidance.

1.4 Equipment essentials

- Differential amplifier with gain ~10^5–10^6, common-mode rejection > 100 dB
- Filters: NCS motor 2–10 kHz; NCS sensory 20 Hz–2 kHz; needle EMG 20 Hz–10 kHz
- Stimulator with constant-current output 0–100 mA, duration 0.05–1 ms

- Recording electrodes: surface (Ag/AgCl), monopolar needle, concentric needle, single-fiber needle
- Limb temperature monitoring — maintain ≥ 32 °C (upper) and ≥ 30 °C (lower)

Temperature is the most preventable source of error

A drop of 1 °C lowers conduction velocity by ~2 m/s and prolongs distal motor latency by ~0.2 ms while increasing amplitude. Always measure and record skin temperature; warm cool limbs before testing rather than relying on correction factors.

1.5 Safety

Electrical current delivered by stimulators is well below cardiac thresholds in healthy patients. However, certain situations require caution:

- **Implanted cardiac devices** — avoid stimulation directly over the device; transthoracic stimulation should not cross the heart axis. Modern pacemakers/ICDs are generally safe with peripheral stimulation but consult device specifications.
- **Anticoagulation** — needle EMG of deep muscles (e.g., paraspinals, iliopsoas, deep flexors) carries higher hematoma risk; consider alternatives or selective sampling.
- **Lymphedema, cellulitis, fresh wound** — avoid the affected segment.
- **Standard precautions** — single-use needles, sterile technique, sharps disposal.

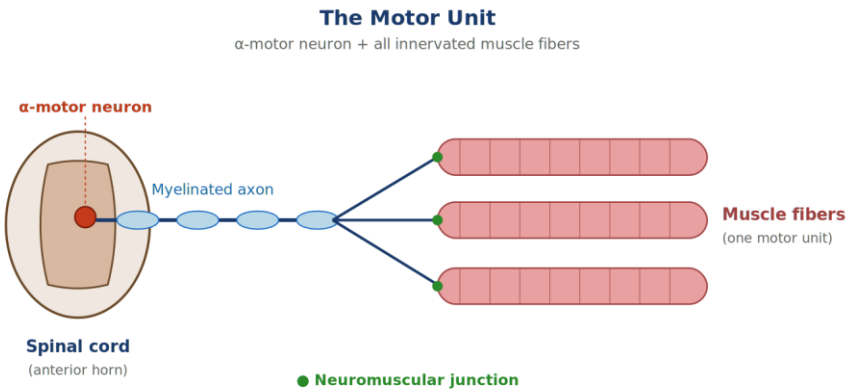
Documentation

A complete EDX report includes: indication, relevant history and exam, equipment and filter settings, side-by-side comparison with normal values (or laboratory norms), full needle EMG findings (insertional, spontaneous, MUAP analysis, recruitment), interpretation, and a clinical conclusion answering the referring question.

Neurophysiology & Anatomical Substrate

2.1 The motor unit

Sherrington's *motor unit* remains the elementary functional element of the motor system: a single α-motor neuron, its axon, and all the muscle fibers it innervates. The number of fibers per motor unit varies enormously, from ~10 in extraocular muscles to ~2000 in gastrocnemius, dictating fineness of control versus power.



Schematic of the motor unit: anterior horn cell, myelinated axon, terminal arborization, and innervated muscle fibers.

2.2 Peripheral nerve fiber classification

Class	Diameter (μm)	CV (m/s)	Function
Aα (Ia, Ib)	12-20	70-120	Muscle spindle primary, Golgi tendon, α-motor

Class	Diameter (μm)	CV (m/s)	Function
Aβ (II)	6-12	30-70	Touch, pressure, vibration, joint position
Aγ	2-8	15-30	γ-motor (intrafusal)
Aδ (III)	1-5	5-30	Cold, fast pain, crude touch
B	1-3	3-15	Preganglionic autonomic
C (IV)	0.2-1.5	0.5-2	Warmth, slow pain, postganglionic autonomic

What routine NCS captures

Standard NCS records only large myelinated fibers (Aα/Aβ). Small fiber neuropathy (Aδ, C) is not detected by routine NCS and requires skin biopsy, quantitative sensory testing, or autonomic studies.

2.3 Action potential generation & saltatory conduction

The resting nerve membrane potential (~−70 mV) is established by the Na^+/K^+ ATPase and selective ion permeability. Depolarization beyond threshold (~−55 mV) triggers voltage-gated Na^+ channels, producing the rising phase. Repolarization follows from Na^+ inactivation and delayed K^+ efflux. In myelinated fibers, action potentials propagate by jumping between nodes of Ranvier (saltatory conduction), which is dramatically faster than continuous conduction in unmyelinated fibers.

Implications for disease

- **Demyelination** → loss of saltatory conduction → marked slowing, conduction block, temporal dispersion

- **Axonal loss** → reduction in number of conducting fibers → decreased amplitude with relatively preserved velocity (until severe)
- **Channelopathy** → abnormal excitability (myotonia, neuromyotonia, periodic paralyses)

2.4 The neuromuscular junction

The chemical synapse between motor nerve terminal and muscle endplate uses acetylcholine as transmitter, packaged in vesicles of ~10,000 molecules each. A single nerve action potential normally releases ~100 quanta — far more than required to depolarize the postsynaptic membrane to threshold. This **safety factor** ensures reliable transmission and is reduced in NMJ disorders.

Failure points at the NMJ

Presynaptic	LEMS (VGCC antibody), botulism, hypocalcemia
Synaptic cleft	Acetylcholinesterase inhibitor toxicity
Postsynaptic	MG (AChR or MuSK antibody), congenital myasthenic syndromes

2.5 Muscle fiber excitation–contraction coupling

The endplate potential triggers a propagated action potential along the sarcolemma and into transverse tubules. Voltage sensors (DHP receptors) couple to ryanodine receptors on sarcoplasmic reticulum, releasing Ca^{2+} that initiates actin–myosin cross-bridging. Calcium reuptake by SERCA terminates contraction. Defects in this cascade underlie periodic paralyses, malignant hyperthermia, and central core disease, with characteristic EMG correlates.

2.6 The motor unit action potential (MUAP)

When a motor neuron fires, the summed extracellular potentials from all its muscle fibers — recorded by a needle electrode positioned among them — constitute the **motor unit action potential** (MUAP). MUAP characteristics reflect the structure and function of the motor unit:

Parameter	Normal	Reflects
Duration	5-15 ms (varies by muscle/age)	Number & spread of fibers
Amplitude	0.2-3 mV	Fibers near electrode tip
Phases	2-4 (≤4)	Synchrony of firing
Turns	Varies	Internal complexity
Stability	Stable from firing to firing	NMJ integrity

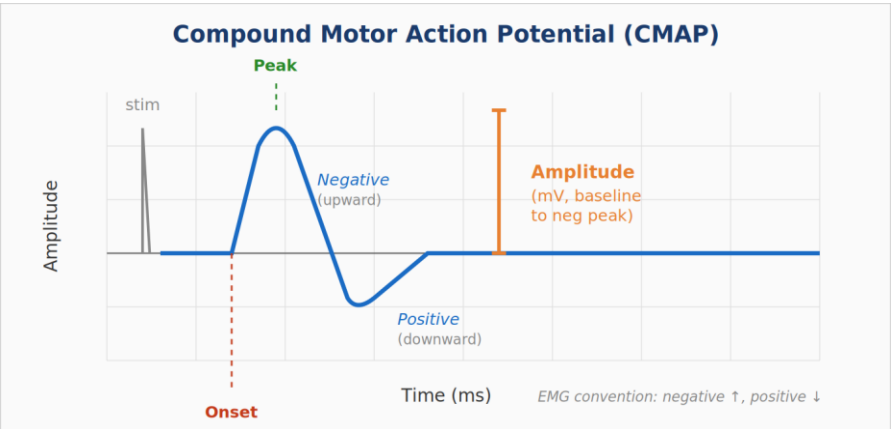
MUAP changes follow physiological logic

Reinnervation after axonal loss expands motor unit territory → **large, long-duration, polyphasic** MUAPs with reduced recruitment. Loss of muscle fibers (myopathy) produces **small, short-duration, polyphasic** MUAPs with normal or early recruitment for the force generated.

Nerve Conduction Studies

3.1 Motor NCS — principles & technique

Motor NCS supramaximally stimulates a peripheral nerve and records the compound muscle action potential (CMAP) from a distal target muscle. The recording belly–tendon montage uses an active electrode (G1) over the motor point and a reference (G2) on the tendon, with a ground between stimulator and recording sites.



Typical CMAP morphology — onset latency, peak, and baseline-to-negative-peak amplitude.

Key parameters

Parameter	Definition	Reflects
Distal motor latency (DML)	Stimulus onset to CMAP onset, distal site	Distal nerve + NMJ + muscle activation
CMAP amplitude	Baseline to negative peak (mV)	Number of activated muscle fibers
Conduction velocity (CV)	Distance between two stim sites / latency difference	Fastest myelinated fibers between sites
Duration	Onset to return to baseline	Synchrony of firing across fibers
Area	Negative-phase area under curve	More robust than amplitude alone

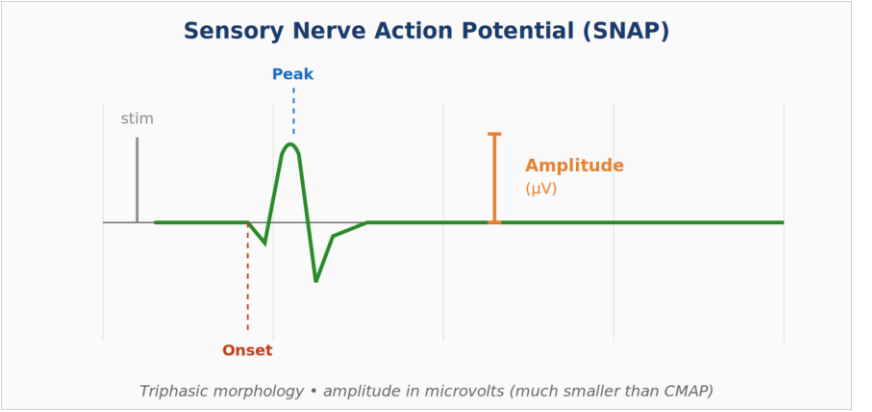
Calculation example — median motor

Wrist stim → DML 3.4 ms; Elbow stim → 8.2 ms; Distance wrist–elbow 24 cm.

CV (forearm) = 240 mm / (8.2 − 3.4) ms = 50 m/s

3.2 Sensory NCS

Sensory NCS records the sensory nerve action potential (SNAP) — the propagated action potential of sensory axons — typically antidromically (stimulating proximal, recording distal in a digit) or orthodromically. The signal is small (microvolts) and requires signal averaging in many cases.

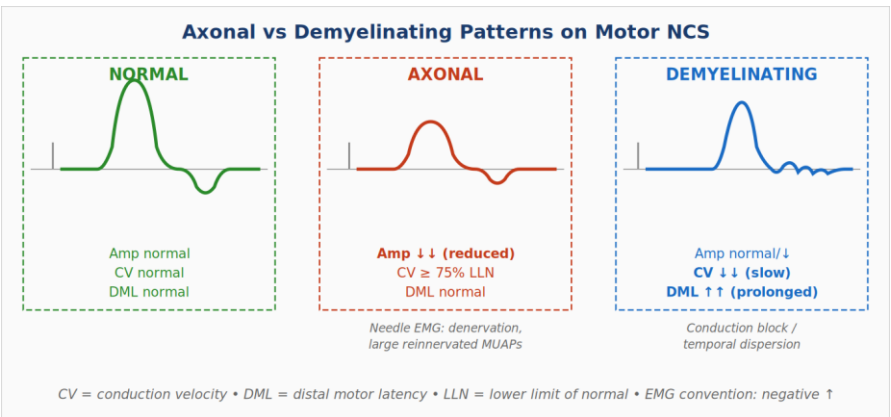


SNAP — characteristic triphasic morphology, amplitude in microvolts.

Why SNAP amplitude is the most sensitive marker

SNAPs reflect sensory ganglion neurons whose cell bodies sit in the dorsal root ganglion. Even mild axonal injury distal to the DRG reduces SNAP amplitude. In **radiculopathy** (lesion proximal to DRG), SNAP is preserved → useful for distinguishing root from plexus/nerve lesions.

3.3 Patterns of abnormality



Distinguishing axonal loss from demyelination on NCS.

Feature	Axonal	Demyelinating
CMAP amplitude	Reduced	Normal or mildly reduced
SNAP amplitude	Reduced	Normal/mild
Distal motor latency	Normal/mild prolongation	Markedly prolonged (>125-150% ULN)
Conduction velocity	≥75% LLN	<75% LLN (often <70%)
F-wave latency	Mildly prolonged	Markedly prolonged or absent
Conduction block	Absent	Present (acquired demyelination)
Temporal dispersion	Absent	Present (acquired demyelination)
Needle EMG denervation	Prominent	Absent / mild secondary

3.4 Conduction block & temporal dispersion

Both phenomena emerge when proximal stimulation produces a smaller or more prolonged CMAP than distal stimulation in the *same* nerve segment.

Conduction block (per EAN/PNS 2021)

- **Definite:** ≥50% drop in CMAP amplitude (or area) between distal and proximal stim, when distal CMAP ≥20% LLN
- **Probable:** 30–49% drop with distal CMAP ≥20% LLN
- CMAP duration increase <30%

Temporal dispersion

- Proximal CMAP duration >30% wider than distal CMAP duration
- Reflects desynchronized arrival of action potentials

- **Highly suggestive of acquired demyelination (CIDP, GBS, MMN)**

Pseudo-conduction-block

Inadvertent submaximal proximal stimulation, anomalous innervation (Martin-Gruber anastomosis), or technical issues can mimic conduction block. Always confirm supramaximal stimulation, verify by inching, and check for anastomoses.

3.5 Anomalous innervations

Variant	Pathway	Clue
Martin-Gruber anastomosis	Median → ulnar in forearm (~15-20% population)	Higher CMAP at elbow than wrist with median stim recording APB
Riche-Cannieu anastomosis	Recurrent median → deep ulnar in palm	"All-ulnar hand" appearance
Accessory deep peroneal	Superficial peroneal → EDB (~20%)	EDB CMAP smaller at fibular head than ankle

3.6 Practical NCS protocol

- Confirm temperature (≥32°C upper, ≥30°C lower); warm if cool.
- Place recording electrodes at standardized sites; measure distances precisely.
- Find supramaximal stimulus (increase ~20-30% above maximum CMAP).
- Record at multiple sites along the nerve; perform "inching" if focal lesion suspected.
- Always compare side-to-side and against laboratory normative data.
- Test sensory and motor for each nerve assessed.

- Examine ≥2 limbs (asymmetric findings suggest mononeuropathy or multiple mononeuropathies; symmetric distal pattern fits length-dependent polyneuropathy).

Late Responses & Reflex Studies

4.1 The F-wave

The F-wave is a small, late motor response evoked when supramaximal stimulation antidromically activates motor neurons; a fraction (1–5%) re-fire and produce an orthodromic descending action potential to the muscle. F-waves probe the entire motor neuron pathway — useful when proximal nerve segments cannot be stimulated directly.

Property	Notes
Latency	Variable trial-to-trial; report *minimum* of 10–20 trials
Amplitude	1–5% of M-response
Persistence	Number of F-waves observed / number of stimuli (normal ≥80% in upper limb, ≥50% in lower)
Chronodispersion	Range = max latency − min latency

Clinical interpretation

- Prolonged minimum latency → proximal demyelination (early GBS, CIDP)
- Absent or reduced persistence → loss of motor neurons (radiculopathy, ALS) or proximal block
- Increased chronodispersion → temporal dispersion / heterogeneous conduction
- F-wave normal when the motor pathway is intact — can normalize false positives

4.2 The H-reflex

The H-reflex is the electrical analogue of the monosynaptic stretch reflex. Submaximal tibial nerve stimulation activates Ia afferents from soleus muscle spindles, which monosynaptically excite α-motor neurons, producing the H-response.

Standard recording (S1 reflex)

- Stimulate tibial nerve in popliteal fossa with long-duration (1 ms) submaximal pulses.
- Record from soleus, midline of upper calf.
- Latency normal: ~28–32 ms (height-dependent).
- Side-to-side latency difference normal <1.5 ms.

When the H-reflex helps

- S1 radiculopathy: unilateral absent or prolonged H-reflex strongly supports the diagnosis.
- Length-dependent neuropathy: bilaterally absent H-reflexes are an early sign.
- Acute GBS: bilaterally absent H-reflexes can be the first electrophysiological abnormality.

4.3 The blink reflex

Stimulation of the supraorbital branch of the trigeminal nerve evokes two responses recorded over orbicularis oculi: an early **R1** (ipsilateral, oligosynaptic, brainstem reflex via main sensory trigeminal nucleus) and a later **R2** (bilateral, polysynaptic via spinal trigeminal nucleus, projecting to facial nuclei).

Response	Side	Normal latency	Pathway
R1	Ipsilateral	≤13 ms	V → pons → VII (oligosynaptic)

Response	Side	Normal latency	Pathway
R2 ipsi	Ipsilateral	≤41 ms	V → pons/medulla → VII (polysynaptic)
R2 contra	Contralateral	≤44 ms	V → bilateral → VII

Lesion patterns

Lesion site	Stim affected side	Stim healthy side
Trigeminal (afferent)	R1, R2 ipsi, R2 contra all delayed/absent	All normal
Facial (efferent)	Only ipsilateral R1, R2 absent	Only contralateral R2 absent
Pontine (R1)	R1 affected; R2 may be preserved	R1 normal
Lateral medullary	R2 ipsi delayed	R2 contra delayed

4.4 Other late responses

A-wave (axon reflex)

An intermediate response between M and F, fixed in latency, arising from collateral sprouting following axonal injury. Found in chronic neuropathies (e.g., recovered radiculopathies, demyelinating disease) and in normal individuals when stimulating tibial nerve.

Sympathetic skin response (SSR)

Records sweat-gland-mediated skin potentials following arousing stimuli (electrical, deep breath, startle). Tests postganglionic sympathetic sudomotor pathways. Useful but variable; absence (especially in young patients) suggests autonomic dysfunction (small fiber neuropathy, multiple system atrophy, amyloidosis).

Modern adjuncts

Quantitative sudomotor axon reflex testing (QSART), nerve excitability testing (axonal threshold tracking), nerve ultrasound, and MR neurography increasingly complement traditional EDX, especially for proximal lesions, small-fiber pathology, and inflammatory neuropathies.

Needle Electromyography

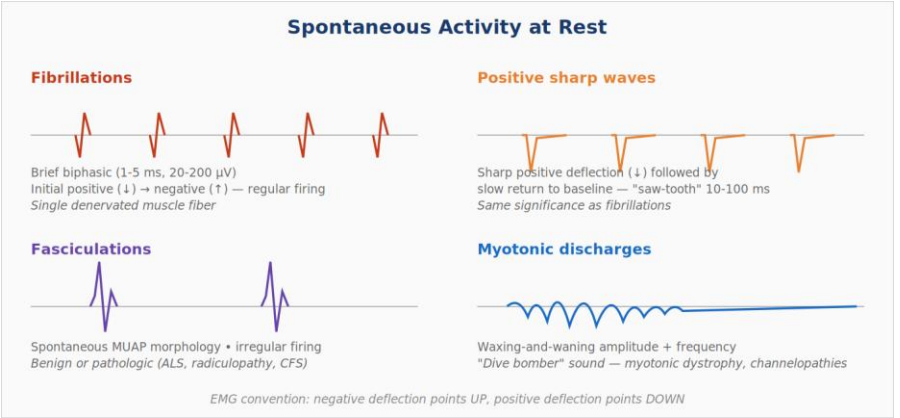
5.1 Technical aspects

Needle EMG records electrical activity of muscle fibers using either concentric needle (CNE) or monopolar needle (MNE) electrodes. CNE has a small recording area (~580 μm²), giving sharp localized recordings ideal for MUAP analysis. MNE has a larger pickup, useful for spontaneous activity. Filters are typically set at 20 Hz–10 kHz, with sweep speed 10 ms/division and gain 50–500 μV/division.

5.2 Phases of the needle examination

Phase	What is recorded	What is sought
Insertion	Brief burst as needle moves	Increased: denervation, myotonic disorders. Decreased: fibrosis, severe atrophy
At rest	Stationary needle, relaxed muscle	Spontaneous activity (fibs, PSWs, fascics, myotonic, complex repetitive)
Mild contraction	Single MUAP firing	Morphology: amplitude, duration, phases, stability
Increasing effort	Recruitment pattern	Order, rate, density of MUAPs
Maximal effort	Interference pattern	Full vs reduced; force–frequency match

5.3 Spontaneous activity at rest



Common spontaneous activity types at rest.

Potential	Origin	Significance
Fibrillation	Single denervated muscle fiber	Active denervation (begins 2-3 wk after axonal loss)
Positive sharp wave	Single denervated muscle fiber (initial positive deflection from injury)	Same as fibrillation
Fasciculation	Spontaneous discharge of single motor unit	Benign or pathologic; ALS, radiculopathy, cramp-fasciculation syndrome
Myotonic discharge	Sustained muscle fiber discharge	Myotonic dystrophy, channelopathies, hyperkalemic periodic paralysis
Complex repetitive discharge	Group of fibers firing as ephaptic chain	Chronic neurogenic or myopathic; non-specific
Myokymic discharge	Group of motor units in repetitive bursts	Radiation plexopathy, MS, facial myokymia
Neuromyotonic discharge	Very high frequency (150-300 Hz) burst	Isaacs syndrome, anti-VGKC complex
Endplate noise / spikes	Miniature/endplate potentials	Normal — needle in endplate zone

Grading spontaneous activity

1+ — persistent >1 sec in 1 area

2+ — persistent in >2 areas

3+ — persistent throughout muscle

4+ — fills baseline, contiguous in all areas

5.4 MUAP analysis

Pattern	Duration	Amplitude	Phases	Recruitment	Suggests
Normal	5-15 ms	0.2-3 mV	≤4	Full	—
Neurogenic chronic	↑ (long)	↑↑ (large)	↑ (polyphasic)	↓ (reduced)	Chronic axonal loss with reinnervation: ALS, chronic neuropathy, old radiculopathy
Neurogenic acute	Normal	Normal	Normal	↓↓	Acute axon loss before reinnervation
Myopathic	↓ (short)	↓ (small)	↑ (poly)	Early / increased	Myopathy
Unstable MUAP	Normal	Normal	↑	Variable	NMJ disorder, early reinnervation

5.5 Recruitment

Recruitment is the orderly addition of motor units with increasing voluntary effort. Two principles govern interpretation:

Recruitment frequency rule

First-recruited MUAP fires at ~5 Hz; second is recruited at ~10 Hz; third at ~15 Hz. The "5:1 rule" — if the firing rate of the fastest unit divided by the number of units exceeds 5, recruitment is reduced (neurogenic).

Pattern	Pattern characteristics	Etiology
Reduced	Few units firing at high rates → 5:1 ratio exceeded; "picket-fence" appearance	Neurogenic — axonal loss or conduction block
Early / increased	Many small units firing for minimal force; full pattern at low effort	Myopathy — fewer fibers per unit
Poor activation	Few units, slow firing rates	Central / suprasegmental cause; pain, lack of cooperation

5.6 Sequential changes after axonal injury

Time after injury	NCS	Needle EMG
0–3 days	Normal initially; CMAP drops as axons degenerate (3-10 d)	Reduced recruitment; no spontaneous activity yet
1–3 weeks	CMAP/SNAP minima reached	Fibrillations, positive sharp waves appear (~14-21 d)
3+ months	Stable amplitudes	Polyphasic, large MUAPs from reinnervation; reduced recruitment persists
6+ months	Recovery if reinnervation succeeds	Long-duration, very large MUAPs ("giant"), still reduced recruitment

Timing matters

Performing EMG too early misses denervation potentials. Wait ≥2-3 weeks after acute injury for the most informative needle exam, while NCS can be done immediately to confirm injury and estimate severity (degree of CMAP loss correlates with axonal loss).

Entrapment Neuropathies

6.1 Carpal tunnel syndrome (CTS)

CTS — compression of the median nerve at the wrist — is the most common entrapment neuropathy. EDX is the diagnostic gold standard, with sensitivity ~85% and specificity >95% when standardized criteria are applied.

Standard EDX evaluation

Test	Comment
Median motor (DML to APB)	Prolonged DML >4.4 ms; compare to ulnar
Median sensory (wrist to digit 2 or 3)	Prolonged peak latency; reduced amplitude
Ulnar sensory (digit 5)	Normal control on the same hand
Median–ulnar palmar comparison	Sensitive: latency difference >0.4 ms abnormal
Median–radial thumb comparison	Latency difference >0.5 ms abnormal
Median–ulnar lumbrical-interosseous	Useful when DML borderline

CTS severity scale (Bland 2000, AANEM)

Grade 1 (very mild)	Comparison studies abnormal only
Grade 2 (mild)	Sensory CV slow, normal motor
Grade 3 (moderate)	Sensory abn + DML prolonged
Grade 4 (severe)	Sensory absent + motor slowed
Grade 5 (very severe)	Motor & sensory absent
Grade 6 (extremely severe)	Thenar motor absent + needle EMG denervation

2024 nuance — adding ultrasound

Modern practice increasingly couples NCS with neuromuscular ultrasound. A median nerve cross-sectional area at the carpal inlet of >10–13 mm² (laboratory dependent) supports CTS, can identify anatomical variants (bifid median, persistent median artery), and is especially helpful in mild or NCS-negative cases.

6.2 Ulnar neuropathy at the elbow (UNE / cubital tunnel)

The second most common entrapment, typically at the cubital tunnel just distal to the medial epicondyle. Reliable diagnosis requires careful technique with elbow flexed 90–135°.

Finding	Threshold
Across-elbow segmental velocity	<50 m/s OR drop >10 m/s vs forearm
Conduction block across elbow	≥20% drop in amplitude (with proper technique)
"Inching" — focal slowing	>0.4 ms / 1 cm at compression site
Ulnar sensory amplitude (D5)	Reduced or absent

Distinguishing UNE from C8/T1 lesion

- **Medial antebrachial cutaneous (MAC) sensory** — preserved in UNE (its origin is from medial cord, not ulnar); reduced in lower trunk plexopathy
- **FDI vs APB needle EMG** — both abnormal in C8/T1; only ulnar-innervated muscles in UNE
- **Cervical paraspinals** — denervation favors radiculopathy

6.3 Other common entrapments

Site / Nerve	Compression point	Cardinal EDX finding
Radial — spiral groove	Humerus midshaft (Saturday night palsy)	CMAP loss recording EIP; sparing triceps
Posterior interosseous	Arcade of Frohse (supinator)	Pure motor weakness extensors; spares ECRL
Median — pronator teres	Forearm	Rare; preserved DML to APB; weakness FDS, FDP-I/II
Anterior interosseous	Forearm (often spontaneous)	Pure motor: FPL, FDP-I/II, PQ ("OK sign" loss)
Lateral femoral cutaneous	Inguinal (meralgia paresthetica)	Reduced/absent LFC sensory; pure sensory
Common peroneal — fibular head	Knee	Conduction block across fibular head; foot drop; spares short head biceps femoris
Tibial — tarsal tunnel	Medial malleolus	Prolonged medial/lateral plantar latencies
Sural	Posterior calf	Pure sensory; rare in isolation

6.4 Pearls for entrapment workup

Localize before labeling

A single sensory amplitude reduction without other findings is rarely a true entrapment. Confirm by:
1. Demonstrating focal slowing or block at the suspected site (inching)
2. Comparing to the contralateral side
3. Documenting needle EMG abnormalities in the appropriate distribution
4. Excluding more proximal lesions (radiculopathy, plexopathy)

Polyneuropathies — Diagnostic Approach

7.1 Classification framework

The first step in evaluating any polyneuropathy is to characterize it across four dimensions:

Pattern

- Length-dependent (distal-to-proximal)
- Non-length-dependent / multifocal
- Mononeuropathy multiplex
- Symmetric vs asymmetric

Modality

- Sensory predominant
- Motor predominant
- Sensorimotor
- Pure autonomic / autonomic-predominant

Pathology

- Axonal
- Demyelinating (acquired)
- Demyelinating (hereditary)
- Mixed / secondary axonal loss

Tempo

- Acute (<4 weeks)
- Subacute (4–8 weeks)
- Chronic (>8 weeks)
- Relapsing

7.2 Common etiologies by pattern

Pattern	Common causes
Distal symmetric axonal sensorimotor	Diabetes, alcohol, B12, uremia, HIV, chemotherapy (taxanes, platinum), idiopathic
Distal symmetric demyelinating	CMT (hereditary), POEMS, anti-MAG IgM
Acute symmetric demyelinating	GBS (AIDP), acute-onset CIDP
Chronic symmetric demyelinating	CIDP, anti-MAG, monoclonal gammopathy
Mononeuropathy multiplex	Vasculitis (AAV, PAN, RA, SLE), diabetes, leprosy, HNPP, lymphoma, Lyme, sarcoidosis
Pure motor	MMN with conduction block, MND, lead, porphyria, paraneoplastic
Pure sensory ataxic	Sjögren ganglionopathy, paraneoplastic anti-Hu, B6 toxicity, cisplatin, HIV
Small fiber	Diabetes, idiopathic, amyloid, sarcoid, SCN9A/SCN10A, Sjögren
Autonomic predominant	Diabetes, amyloid, paraneoplastic AAG, hereditary

7.3 EDX strategy in suspected polyneuropathy

- **Bilateral lower extremity NCS:** sural and superficial peroneal sensory; tibial and peroneal motor with F-waves and H-reflexes.

- **Upper extremity NCS:** at least one motor (median or ulnar) and one sensory (median or ulnar); add radial sensory if length-dependent suspected.
- **If patchy pattern or mononeuropathy multiplex:** sample additional asymmetric nerves, and consider proximal nerves (axillary, musculocutaneous).
- **Needle EMG:** sample at least one distal and one proximal muscle in each limb with abnormal NCS; in length-dependent cases, distal muscles (e.g., abductor hallucis, EHL, FDI, APB) typically show the earliest changes.

7.4 Charcot-Marie-Tooth disease (CMT)

The most common hereditary neuropathy. EDX subdivides CMT into three groups, guiding genetic testing strategy:

Type	Median motor CV	Inheritance	Most common gene
CMT1 (demyelinating)	<38 m/s	AD	PMP22 duplication (CMT1A) — ~70%
CMT2 (axonal)	>38 m/s	AD	MFN2 (CMT2A); MPZ also
Intermediate	25–45 m/s	AD/X-linked	GJB1 (CMTX1) — second most common overall

Clues to hereditary neuropathy

Slowly progressive, foot deformity (pes cavus), hammer toes, palpable nerves, family history, very young onset, uniform slowing across all motor nerves (unlike CIDP which is more multifocal).

7.5 Diabetic neuropathies

Diabetes can cause virtually any neuropathy phenotype. Recognize:

- **Distal symmetric polyneuropathy (DSPN)** — most common; mixed axonal sensorimotor
- **Diabetic radiculoplexus neuropathy** (lumbosacral or cervical, "diabetic amyotrophy") — painful asymmetric proximal weakness, often weight loss
- **Cranial mononeuropathies** — III, IV, VI, VII (vasculitic ischemia)
- **Treatment-induced neuropathy** ("insulin neuritis") — worsening pain after rapid glycemic control
- **Autonomic neuropathy** — orthostasis, gastroparesis, anhidrosis

7.6 Vasculitic neuropathy

Diagnostic clues

Acute or subacute, painful, asymmetric, stepwise progression. EDX shows multifocal axonal pattern (mononeuropathy multiplex). Sural or peroneal nerve biopsy may be diagnostic. Causes: ANCA-associated vasculitis (especially EGPA, GPA), PAN, RA, SLE, cryoglobulinemia, hepatitis B/C. Requires aggressive immunosuppression.

8 CIDP — EAN/PNS 2021 Guideline

What changed in 2021

The 2021 EAN/PNS revision **(1)** reduced electrodiagnostic certainty levels from three (definite/probable/possible) to two (CIDP and possible CIDP); **(2)** formally introduced "CIDP variants" (distal, multifocal, focal, motor, sensory) replacing the older "atypical CIDP" label; **(3)** excluded autoimmune nodopathies (anti-NF155, anti-CASPR1, anti-CNTN1) as a separate disease group; and **(4)** emphasized supportive criteria including ultrasound, MRI, and treatment response.

8.1 Clinical phenotypes

Phenotype	Clinical hallmarks
Typical CIDP	Symmetric proximal AND distal weakness, progressive ≥8 weeks, sensory involvement, areflexia/hyporeflexia
Distal CIDP	Distal predominant sensory or sensorimotor; "DADS" phenotype
Multifocal CIDP (Lewis-Sumner / MADSAM)	Asymmetric, multifocal, sensory + motor in distribution of individual nerves
Focal CIDP	Single limb or plexus involvement
Motor CIDP	Pure motor; no sensory symptoms or signs
Sensory CIDP	Pure sensory; chronic

8.2 Electrodiagnostic criteria (EAN/PNS 2021)

CIDP — needs ≥1 of the following

- **Motor distal latency prolongation** ≥50% above ULN in 2 nerves (excluding median DML at wrist if MUAP suggests CTS)
- **Reduction of motor conduction velocity** ≥30% below LLN in 2 nerves
- **Prolongation of F-wave latency** ≥20% above ULN (or ≥50% if motor amplitude is reduced) in 2 nerves; OR absence of F-waves in 2 nerves with normal CMAP and another demyelinating finding in 1 nerve
- **Motor conduction block**: ≥50% drop in amplitude on proximal vs distal stim in 2 nerves; OR in 1 nerve plus another demyelinating finding in 1 other nerve
- **Abnormal temporal dispersion** >30% increase in CMAP duration between proximal and distal in 2 nerves
- **Distal CMAP duration** increase in ≥1 nerve plus ≥1 other demyelinating finding

Possible CIDP

Demyelinating findings as above but in only 1 nerve.

8.3 Supportive criteria

- **CSF**: protein elevation (without pleocytosis >10/mm³)
- **MRI**: enhancement and/or hypertrophy of cervical/lumbosacral roots, brachial/lumbosacral plexus

- **Ultrasound**: nerve enlargement (cross-sectional area increase) — proximal segments and roots
- **Nerve biopsy**: rarely needed; macrophage-mediated demyelination, onion bulbs, inflammatory infiltrate
- **Objective response to immunotherapy** (IVIg, steroids, plasma exchange)

8.4 Differential to consider

Mimic	Distinguishing features
Charcot-Marie-Tooth (CMT1)	Family history, foot deformity, uniform slowing, no conduction block, no temporal dispersion
POEMS syndrome	Monoclonal gammopathy + endocrine + skin + organomegaly + edema; VEGF elevated
Anti-MAG neuropathy	Distal demyelinating; IgM monoclonal; anti-MAG antibody; very prolonged distal latencies (TLI <0.25)
Multifocal motor neuropathy (MMN)	Pure motor; conduction block; anti-GM1; responds to IVIg, NOT steroids
Autoimmune nodopathies	Sensory ataxia, tremor, severe disability, poor IVIg response, anti-NF155/CASPR1/CNTN1 antibodies — now classified separately

8.5 Treatment principles

First-line (per 2021 guideline)

- **IVIg** 2 g/kg over 2-5 days, then maintenance 1 g/kg every 3 weeks
- **Subcutaneous Ig (SCIg)** — strongly recommended for maintenance therapy (new in 2021)

- **Corticosteroids** — oral prednisolone or pulse IV methylprednisolone
- **Plasma exchange** — alternative or rescue

For refractory cases, second-line options include rituximab (especially with IgG4 anti-NF155 in nodopathies), cyclophosphamide, azathioprine, mycophenolate. Treatment response guides ongoing classification — true CIDP improves with first-line therapy; non-responders should prompt re-evaluation for nodopathies, MGUS-related neuropathy, or POEMS.

Guillain–Barré Syndrome — EAN/PNS 2023

2023 EAN/PNS guideline highlights

The first GRADE-based international guideline for GBS, developed jointly by 21 experts. Key recommendations: **(1)** use Brighton criteria for diagnostic certainty; **(2)** early electrodiagnosis aids subtype classification but is not required for treatment initiation; **(3)** IVIg 0.4 g/kg/day × 5 days OR plasma exchange in patients unable to walk unaided, ideally within 2 weeks of weakness onset (acceptable up to 4 weeks); **(4)** corticosteroids and other immunotherapies are not recommended.

9.1 Clinical phenotype

An acute, monophasic immune-mediated polyradiculoneuropathy with progressive weakness reaching nadir within 4 weeks. Cardinal features:

- Symmetric, ascending limb weakness
- Hyporeflexia or areflexia
- Variable sensory symptoms (often less prominent than motor)
- Cranial nerve involvement (especially VII bilaterally)
- Dysautonomia (BP fluctuations, arrhythmias, ileus)
- Respiratory failure in 20–30% (require monitoring of FVC, NIF)

- Recent antecedent infection in ~70% (Campylobacter, CMV, EBV, Mycoplasma, Zika; vaccinations rarely)

9.2 GBS subtypes

Subtype	Clinical / EDX features
AIDP (acute inflammatory demyelinating polyradiculoneuropathy)	Most common in West (~70-90%); demyelinating EDX (prolonged DML, slow CV, conduction block, prolonged or absent F-waves)
AMAN (acute motor axonal)	Common in Asia, summer; pure motor; reduced CMAP; preserved sensory; anti-GM1 / GD1a
AMSAN (acute motor and sensory axonal)	Severe; reduced CMAP and SNAP; slow recovery
Miller Fisher syndrome (MFS)	Ophthalmoplegia + ataxia + areflexia; anti-GQ1b in >90%
Bickerstaff brainstem encephalitis	MFS features + altered consciousness; anti-GQ1b
Pharyngeal-cervical-brachial	Bulbar + cervical + arm weakness; anti-GT1a
Paraparetic	Pure leg weakness

9.3 Brighton diagnostic certainty

Level	Required features
Level 1 (highest)	Bilateral flaccid limb weakness + decreased/absent reflexes + monophasic course nadir 12 h–28 d + CSF cytoalbuminologic dissociation + electrophysiology consistent with GBS subtype + no alternative diagnosis
Level 2	Above without one of CSF or EDX (other intact)
Level 3	Bilateral weakness + decreased reflexes + monophasic course + no alternative diagnosis (CSF and EDX may be missing/normal)

9.4 Electrodiagnostic features

Findings depend on timing and subtype. **Early** (within first week), the most sensitive findings are:

- Absent or markedly reduced H-reflexes (often bilaterally)
- Prolonged or absent F-waves
- Reduced sural-sparing pattern: sural SNAP preserved while upper limb sensory amplitudes reduced (~60% specific for GBS)
- Conduction block in non-entrapment sites
- Prolonged distal motor latencies

"Sural sparing" pattern

Reduced upper-limb sensory amplitudes (median, ulnar) with preserved sural amplitude is a useful early clue specific for AIDP, present in approximately 50–60% of cases. Likely reflects greater susceptibility of more proximal nerve segments to inflammatory injury.

EDX criteria for AIDP (Hadden, modified)

Demyelinating findings in ≥2 motor nerves: any combination of prolonged DML, reduced CV, prolonged F-wave latency or persistence, conduction block, or temporal dispersion.

9.5 Prognosis

Two scoring tools are widely used:

Tool	Use	Components
EGRIS (Erasmus GBS Respiratory Insufficiency Score)	Predicts mechanical ventilation	Days from weakness onset to admission, facial/bulbar weakness, MRC sum score
mEGOS (modified Erasmus GBS Outcome Score)	Predicts walking ability at 6 months	Age, preceding diarrhea, MRC sum score

9.6 Treatment summary (EAN/PNS 2023)

First-line therapy

For patients unable to walk 10 m unaided: **IVIg** 0.4 g/kg/day for 5 days OR **plasma exchange** 4–5 sessions over 1–2 weeks (200–250 mL/kg total) Both equally effective; choice based on local availability/cost; treatment ideally within 2 weeks of weakness onset (acceptable up to 4 weeks).

Not recommended

Corticosteroids (no benefit, possibly harmful in some studies). Sequential IVIg + PE (no added benefit). A second IVIg course in non-improving patients (no benefit demonstrated in I-SID trial). New agents under trial (eculizumab, imlifidase, FcRn inhibitors) — pending evidence.

Supportive care

- Respiratory monitoring: FVC <20 mL/kg, NIF less negative than −30 cmH_2O, or rapid decline → consider intubation
- Cardiac monitoring for autonomic dysfunction

- VTE prophylaxis
- Pain management (often neuropathic)
- Early rehabilitation

Motor Neuron Diseases — Gold Coast 2020

Why criteria changed

The El Escorial criteria (1994, revised 2000) and Awaji criteria (2008) had high specificity but limited sensitivity, particularly in early-stage and clinical-trial settings. The 2020 Gold Coast criteria, developed in Gold Coast, Australia, simplify to a **dichotomous decision**: ALS or not ALS. Recent meta-analyses confirm sensitivity ~95% (vs ~85% for older criteria) at the cost of slightly lower specificity (~66–88%).

10.1 Gold Coast diagnostic criteria for ALS (2020)

All three required

- **Progressive motor impairment** documented by history or repeated assessment, preceded by normal motor function
- **Upper motor neuron (UMN) and lower motor neuron (LMN) dysfunction** in at least one body region (bulbar, cervical, thoracic, lumbosacral); *OR* LMN dysfunction in at least 2 body regions
- **Investigations excluding other disease processes**

Notable update: Fasciculations are now treated as equivalent to chronic neurogenic EMG changes when assessing LMN involvement.

10.2 Body regions & LMN evidence

Region	Recommended muscles for needle EMG
Bulbar	Tongue (genioglossus), masseter; clinical fasciculations of tongue suffice
Cervical (C5–T1)	≥2 muscles from different roots and nerves: e.g., deltoid (C5), biceps (C5-6), pronator teres (C6-7), triceps (C7), FDI (C8-T1), APB (C8-T1)
Thoracic	Paraspinal muscles (T6–T12), high abdominal (rectus abdominis)
Lumbosacral (L2–S1)	Iliopsoas (L2-3), quadriceps (L3-4), tibialis anterior (L4-5), medial gastrocnemius (S1), abductor hallucis (S1-2)

LMN evidence (any of)

- Clinical: weakness with atrophy, fasciculations
- EMG: chronic neurogenic MUAPs (long duration, large amplitude, polyphasic), reduced recruitment, AND active denervation (fibrillations, positive sharp waves) OR fasciculation potentials

UMN evidence (any of)

- Increased reflexes (in a weak/wasted limb)
- Hoffmann sign, clonus
- Pseudobulbar features (emotional lability, brisk jaw jerk)
- Spasticity, extensor plantar response

10.3 ALS subtypes & mimics

Entity	Distinguishing features
Classic ALS (Charcot)	Mixed UMN + LMN, asymmetric onset, progressive, fasciculations
PMA (progressive muscular atrophy)	Pure LMN; ~10% of MND
PLS (primary lateral sclerosis)	Pure UMN ≥4 years; rare
Progressive bulbar palsy	Onset bulbar; later limb involvement
Flail arm / leg syndrome	LMN-predominant in single limb girdle
Familial ALS	~10%; SOD1, C9orf72 (most common, with FTD), TARDBP, FUS

Important mimics

- Multifocal motor neuropathy (MMN) — pure LMN; conduction block on NCS; responds to IVIg
- Cervical spondylotic myeloradiculopathy — UMN signs in legs from cord, LMN signs in arms from roots; MRI key
- Inclusion body myositis — slowly progressive, asymmetric finger flexor and quadriceps weakness; NCS normal
- Hexosaminidase A deficiency, Kennedy disease (X-linked spinobulbar muscular atrophy)
- Post-polio syndrome

10.4 Treatment landscape (2024–2026)

Disease-modifying therapy

- Riluzole — extends survival by ~3 months
- Edaravone — IV/oral; modest benefit in selected patients

- Tofersen (intrathecal antisense oligonucleotide) — for SOD1-mutated ALS, FDA approved 2023; reduces neurofilament light chain
- Multidisciplinary care including non-invasive ventilation, gastrostomy when appropriate, palliative input

Neuromuscular Junction Disorders

11.1 Repetitive nerve stimulation (RNS)

RNS exploits the fact that NMJ disorders impair the safety factor of neuromuscular transmission. Several closely-spaced supramaximal stimuli (typically 2–3 Hz, 9–10 stimuli) produce a CMAP train; the ratio of the 4th-or-5th to the 1st CMAP detects abnormalities.

Pattern	Finding	Suggests
Decrement >10% at 2-3 Hz	Progressive amplitude drop in train	Postsynaptic NMJ disorder (MG)
Increment >60-100% after exercise or 30-50 Hz	Marked CMAP enhancement	Presynaptic NMJ (LEMS, botulism)
Low baseline CMAP + increment	Small CMAP at rest, rises post-exercise	LEMS hallmark
No abnormality	Normal RNS	Doesn't exclude MG (sensitivity ~50-60%); proceed to SFEMG

Optimizing RNS technique

- Test clinically weak muscles (sensitivity rises with severity)
- Both proximal (deltoid, trapezius, anconeus, facial) and distal (ulnar to ADM/FDI) muscles
- Maintain temperature ≥35 °C

- Stop AChE inhibitors 12-24 hr before testing if possible
- Post-activation testing: brief maximum voluntary contraction (10 sec), then RNS at 1, 2, 3, 4 minutes — reveals post-tetanic exhaustion

11.2 Single-fiber EMG (SFEMG)

SFEMG is the most sensitive test for NMJ disorders (~95–99% in generalized MG, ~85–95% in ocular MG). It records action potentials from individual muscle fibers within a single motor unit.

Key parameters

Parameter	Definition
Jitter	Variability of inter-spike interval between two fibers of the same motor unit (mean consecutive difference, MCD)
Blocking	Failure of one fiber to fire on a given trigger
Fiber density	Average number of single fibers within recording radius (~270 μm for SFE)

Concentric needle (CN) jitter — current standard

Modern SFEMG often uses concentric needle electrodes rather than dedicated single-fiber needles (lower cost, infection risk). Reference values are slightly different and laboratory-specific. The 2024 ENMC consensus refines criteria for seronegative MG using jitter studies, emphasizing testing weak muscles and the importance of normal jitter excluding MG in that muscle.

Common test muscles

- Voluntary activation: extensor digitorum communis, frontalis, orbicularis oculi

- Stimulated SFEMG: useful in young, uncooperative, or deeply weak patients

11.3 Myasthenia gravis

Subtype	Antibody	Features
AChR-positive	Anti-AChR (~85% of generalized)	Variable phenotype; thymoma in 10-15%
MuSK-positive	Anti-MuSK (~5-8%)	Female predominance, bulbar/respiratory predominant, focal atrophy, less response to AChE inhibitors
LRP4-positive	Anti-LRP4 (1-3%)	Often milder; some overlap with AChR
Seronegative	None detected by standard assays	Cell-based assays may identify low-affinity AChR; SFEMG critical
Ocular MG	Variable	Confined to eyes ≥2 years; lower antibody yield; ~50% generalize

Diagnostic algorithm (per ENMC 2024 + MGFA 2020)

- Clinical features (fatigability, fluctuation, ocular onset)
- Antibody testing: AChR → MuSK → LRP4 → cell-based assays for seronegative
- RNS in weak muscles; SFEMG if RNS negative and clinical suspicion remains
- Ice pack test for ptosis (transient improvement)
- CT chest for thymoma (essential in all newly diagnosed MG)
- Edrophonium test rarely used today; oral pyridostigmine response can be assessed

11.4 Lambert-Eaton myasthenic syndrome (LEMS)

Presynaptic NMJ disorder caused by antibodies against P/Q-type voltage-gated calcium channels (VGCC). Reduces ACh release. Strongly associated with small cell lung carcinoma in 50-60% of cases (paraneoplastic).

LEMS triad

- Proximal muscle weakness (legs > arms) with characteristic improvement after brief exercise (post-exercise facilitation)
- Hyporeflexia / areflexia, often improving after exercise
- Dysautonomia: dry mouth, constipation, erectile dysfunction, orthostasis

Diagnostic EDX

- Low baseline CMAP (often <50% LLN) on routine NCS — major clue
- Decrement on slow RNS (2-3 Hz)
- Marked increment >60–100% (often >200%) after brief maximum voluntary contraction (10 sec) or high-frequency stimulation (30-50 Hz)
- Anti-VGCC antibody positive in >85%
- 3,4-diaminopyridine (amifampridine) is symptomatic treatment of choice; immunotherapy and oncological screening required

11.5 Botulism

Presynaptic NMJ block from botulinum neurotoxin (food-borne, wound, infant, iatrogenic). Toxin cleaves SNARE proteins → impaired ACh release.

- Descending flaccid paralysis: cranial nerves first (diplopia, ptosis, dysphagia, dysarthria), then limbs and respiratory muscles
- Pupillary dilation and dry mouth (autonomic)
- EDX: low CMAP, increment on RNS (50-100% at 50 Hz or post-exercise) — similar to LEMS but acute onset
- Equine antitoxin urgently; respiratory support

Myopathies

12.1 EMG features common to myopathies

The myopathic triad

- Short-duration, low-amplitude, polyphasic MUAPs
- Early recruitment — many small motor units recruited at minimal effort to generate force
- Full interference pattern at low force ("low-amplitude full interference")

NCS are typically normal in pure myopathies (unless very severe with marked muscle bulk loss reducing CMAP from the recording muscle). Spontaneous activity at rest may or may not be present, depending on the type:

- With spontaneous activity (fibs, PSWs): inflammatory myopathies, dystrophies, toxic, severe rhabdomyolysis
- Myotonic discharges: myotonic dystrophy, channelopathies, hyperKPP, statin myopathy (rare), acid maltase deficiency (Pompe)
- Without spontaneous activity: most metabolic, mitochondrial, and congenital myopathies

12.2 Inflammatory myopathies

Type	Clinical / Lab	EMG / Other
Dermatomyositis	Heliotrope rash, Gottron, proximal weakness; CK elevated; anti-Mi2, anti-MDA5, anti-TIF1γ	Myopathic + spontaneous activity; perifascicular atrophy on biopsy
Polymyositis	Proximal weakness; CK elevated	Now considered rare; many "PM" cases reclassified as IBM, IMNM, or overlap
IMNM (immune-mediated necrotizing myopathy)	Severe weakness; very high CK; anti-HMGCR (statin-related), anti-SRP	Myopathic with prominent spontaneous activity; necrotizing fibers without inflammation
Inclusion body myositis (IBM)	Asymmetric finger flexor + quadriceps weakness; older patients; modest CK elevation; refractory to therapy	Mixed myopathic + neurogenic-appearing MUAPs (long-duration); rimmed vacuoles on biopsy; anti-cN1A
Antisynthetase syndrome	Myositis + ILD + arthritis + Raynaud + mechanic's hands; anti-Jo-1, PL-7, PL-12, EJ, OJ	Myopathic; ILD often dominates prognosis

12.3 Muscular dystrophies

Disorder	Inheritance	Hallmarks
DMD / BMD	X-linked recessive	Calf pseudohypertrophy, Gower sign; very high CK; dystrophin gene
Limb-girdle MD	Multiple subtypes (AD/AR)	Proximal weakness; multiple genes; LGMD2A (calpainopathy) most common AR
FSHD	AD	Facial + scapular + humeroperoneal; D4Z4 contraction
Myotonic dystrophy 1 (DM1)	AD; CTG repeat in DMPK	Distal weakness, myotonia, cataracts, frontal balding, cardiac conduction block, insulin resistance
Myotonic dystrophy 2 (DM2)	AD; CCTG repeat in CNBP	Proximal predominant; milder myotonia; PROMM
OPMD	AD; PABPN1	Late-onset ptosis + dysphagia
EDMD	AD/X-linked	Joint contractures + cardiomyopathy + slowly progressive muscle weakness

12.4 Metabolic & mitochondrial myopathies

- **Glycogen storage** (McArdle V, Pompe II): exercise intolerance, cramps, rhabdomyolysis; ischemic forearm test
- **Lipid storage** (CPT II, MADD): exercise-induced rhabdomyolysis with prolonged exertion
- **Mitochondrial**: PEO, MELAS, MERRF; multisystem; muscle biopsy with ragged-red fibers

Pompe disease — don't miss

Late-onset Pompe (acid maltase deficiency) presents with proximal weakness mimicking LGMD, often with respiratory weakness and diaphragm involvement out of proportion. Needle EMG often shows myotonic discharges in paraspinal muscles. Enzyme replacement therapy is available — early diagnosis is critical.

12.5 Channelopathies (non-dystrophic myotonias & periodic paralyses)

Disorder	Channel / Gene	Features
Myotonia congenita (Thomsen, Becker)	CLCN1 (Cl^-)	Warm-up myotonia; "Hercules" phenotype
Paramyotonia congenita	SCN4A (Na^+)	Cold-induced; paradoxical (worsens with exercise)
Hyperkalemic periodic paralysis	SCN4A	Brief attacks; high K triggers
Hypokalemic periodic paralysis	CACNA1S, SCN4A	Long attacks; CHO/insulin trigger
Andersen-Tawil syndrome	KCNJ2	Periodic paralysis + dysmorphism + arrhythmia

EDX in channelopathies

Specialized exercise tests (long and short McManis protocols) measure CMAP changes after exercise; characteristic patterns help distinguish subtypes. Cooling protocols can unmask paramyotonia. Genetic testing increasingly replaces extensive electrodiagnostic protocols.

12.6 Toxic and endocrine myopathies

- **Statins** — myalgia/myopathy (~5%), rare immune-mediated necrotizing myopathy with anti-HMGCR

- **Steroids** — chronic high-dose prednisone, type II fiber atrophy; needle EMG often normal
- **Colchicine, chloroquine, amiodarone** — vacuolar myopathy
- **Critical illness myopathy** — ICU; often combined with critical illness polyneuropathy; preserved sensory NCS, low CMAP, myopathic MUAPs
- **Hypothyroidism, hyperthyroidism, Cushing** — proximal weakness; CK normal-elevated

ANormal Values Reference

Values shown are common reference ranges from major textbooks (Preston & Shapiro; Kimura; Dumitru). Always validate against laboratory-specific norms. Temperature: ≥32 °C upper limb, ≥30 °C lower limb.

A.1 Upper extremity motor NCS

Nerve	Recording	DML (ms)	Amp (mV)	CV (m/s)
Median	APB	≤4.4	≥4.0	≥49
Ulnar	ADM	≤3.3	≥6.0	≥49 (forearm); ≥50 (across-elbow)
Radial	EIP	≤2.9	≥2.0	≥50
Musculocutaneous	Biceps	≤4.0	≥4.0	≥55
Axillary	Deltoid	≤4.9	≥4.0	—
Spinal accessory	Trapezius	≤4.0	≥4.0	—
Phrenic	Diaphragm	≤8.0	≥0.4	—

A.2 Lower extremity motor NCS

Nerve	Recording	DML (ms)	Amp (mV)	CV (m/s)
Peroneal	EDB	≤6.5	≥2.0	≥41
Peroneal	Tibialis ant.	≤6.7	≥3.0	≥44
Tibial	AH	≤6.0	≥4.0	≥41
Tibial	ADQP	≤6.3	≥3.0	—
Femoral	Quadriceps	≤7.0	≥3.0	—

A.3 Sensory NCS

Nerve	Site	Peak latency (ms)	Amp (μV)	CV (m/s)
Median	Digit 2 / 3 (antidromic)	≤3.5	≥20	≥50
Ulnar	Digit 5	≤3.1	≥17	≥50
Radial	Snuffbox / dorsum hand	≤2.9	≥15	≥50
Median palmar	Mid-palm (8 cm)	≤2.2	≥40	—
Ulnar palmar	Mid-palm (8 cm)	≤2.2	≥20	—
LABC	Forearm	≤3.0	≥10	—
MABC	Forearm	≤3.2	≥5	—
Sural	Lateral malleolus	≤4.4	≥6	≥40
Superficial peroneal	Lateral leg	≤4.5	≥6	≥40
Saphenous	Medial leg	≤4.0	≥4	—
LFC	Lateral thigh	≤2.8	≥4	—

A.4 F-wave minimum latency (height-corrected approximations)

Nerve	Mean (ms)	Upper limit (ms)	Side-to-side diff
Median	26-30	32	≤2.0 ms
Ulnar	27-31	33	≤2.0 ms
Peroneal	48-55	58	≤4.0 ms
Tibial	48-56	58	≤4.0 ms

A.5 H-reflex (soleus, tibial nerve)

Height (cm)	Mean H latency (ms)	Upper limit (ms)
<160	27	31
160-170	29	33
170-180	31	35
>180	33	37

Side-to-side latency difference normally <1.5 ms.

A.6 Blink reflex

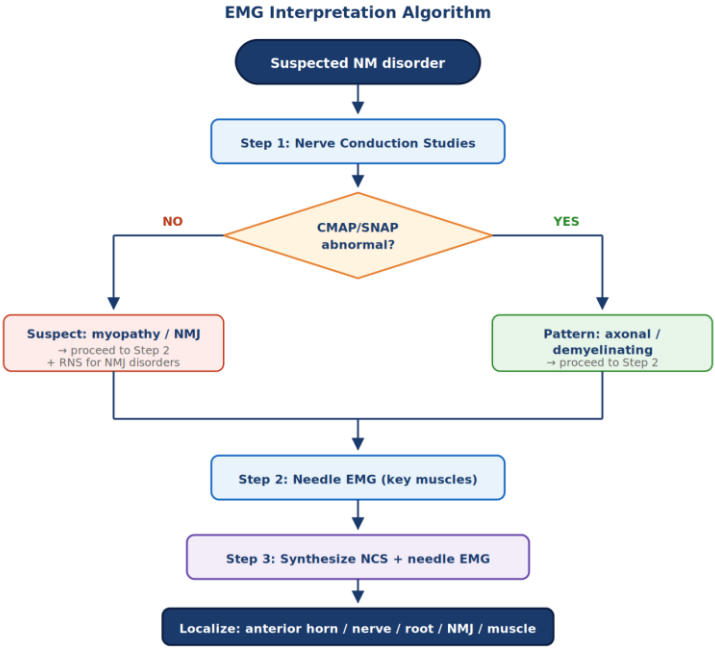
Component	Upper limit (ms)	Side-to-side diff (ms)
R1 (ipsilateral)	13	1.2
R2 ipsilateral	41	5
R2 contralateral	44	7

A.7 RNS & SFEMG criteria

Test	Abnormal
Slow RNS (2-3 Hz) decrement	>10% drop from 1st to 4th-5th CMAP
Brief exercise increment	>60% (highly suggestive of LEMS)
SFEMG jitter (single-fiber needle)	Mean MCD >34 µs (extensor digitorum communis)
SFEMG jitter (concentric needle)	Mean MCD >30-33 µs (muscle-specific; refer to lab norms)
Blocking	Any presence is abnormal

BClinical Algorithms

B.1 General EMG interpretation algorithm



Stepwise approach to localizing neuromuscular lesions.

B.2 Cervical & lumbosacral root reference

Cervical & Lumbosacral Roots — Key Levels

CERVICAL			LUMBOSACRAL		
C5	Deltoid, biceps	*Lateral arm, biceps reflex*	**L2-3**	Iliopsoas, quad	*Anterior thigh*
C6	Biceps, BR, ECRL	*Thumb, BR reflex*	**L4**	VM, TA	*Knee jerk, medial calf*
C7	Triceps, FCR, EDC	*Middle finger, triceps reflex*	**L5**	TA, EHL, peroneal	*Big toe dorsum, foot drop*
C8	FDP, FDI, APB	*5th digit, hand intrinsics*	**S1**	Gastroc, soleus	*Lateral foot, ankle jerk*
T1	Hand intrinsics	*Medial forearm*	**S2**	Glut max	*Posterior thigh*

⚡ **Key tips:**

- Each root supplies multiple muscles via DIFFERENT peripheral nerves; to confirm radiculopathy, sample ≥2 muscles from same root through different nerves.
- Paraspinal muscles separate pre- from post-ganglionic lesions: denervated in radiculopathy (pre-DRG); preserved in plexopathy (post-DRG).

BR = brachioradialis • ECRL = extensor carpi radialis longus • FDP = flexor digitorum profundus • DRG = dorsal root ganglion

Key roots, muscles, and tips for radiculopathy workup.

B.3 Acute weakness — focused electrodiagnostic strategy

Stepwise approach

Confirm pattern: ascending/symmetric → GBS likely; descending → botulism, brainstem; proximal fluctuating → MG crisis; pure motor → MMN, ALS bulbar

Rapid bedside tests: H-reflexes (absence supports GBS), F-waves (prolonged in early GBS), repetitive stim (MG, LEMS, botulism)

If GBS suspected: CSF, anti-ganglioside antibodies; treat empirically if Brighton level 2-3 with progressive disability

If MG crisis: AChR/MuSK antibodies, RNS, ice pack test; do not delay treatment for tests

If neuropathic mimic of myopathy: rapidly progressive symmetric weakness with retained reflexes → think IMNM, electrolyte, toxic

B.4 Suspected polyneuropathy — minimum dataset

Test	Why
Bilateral sural sensory	Most sensitive for length-dependent axonal
Bilateral superficial peroneal sensory	Confirms length-dependent pattern
Bilateral peroneal motor (EDB), F-waves	Distal axonal vs demyelinating
Bilateral tibial motor (AH), H-reflexes	Confirms; H-reflex sensitive early
Median & ulnar (motor + sensory) ≥1 side	Detects diffuse process; CTS controls
Needle EMG: AH or EHL, TA, vastus medialis, FDI, biceps	Distal-to-proximal gradient assessment

B.5 Distinguishing common entrapments from radiculopathy

Question	Median CTS	Ulnar UNE	C6/C7 radic	C8 radic
Median sensory	↓	—	↓ (if C6)	—
Ulnar sensory	—	↓	—	—
Ulnar D5 + MAC	—	D5 ↓; MAC normal	—	—
FDI / APB EMG	APB +; FDI normal	FDI +; APB normal	—	Both +
Cervical paraspinals	Normal	Normal	+ at corresponding level	+ at C8

B.6 Suspected NMJ disorder — workflow

When to test what

Antibody-positive with typical clinical picture → may not need EDX confirmation

Antibody-negative with high clinical suspicion → SFEMG of weak muscle is highly sensitive

RNS in clinically-affected muscles (proximal AND distal); always include facial muscles in suspected MG

For LEMS: low CMAP at baseline + post-exercise increment → highly specific

Always evaluate for thymoma (CT) in MG; for SCLC (CT chest, PET) in LEMS

B.7 Quick clinical pearls (15 high-yield points)

Temperature must be ≥32 °C (upper) or ≥30 °C (lower) before any NCS — single most preventable error.

SNAP preserved + CMAP reduced + motor weakness in same distribution = lesion is at or proximal to dorsal root ganglion (radiculopathy or anterior horn).

Sural sparing (preserved sural with reduced upper limb sensory) is ~60% specific for early AIDP/GBS.

Bilaterally absent H-reflexes can be the earliest electrophysiologic sign of GBS.

Conduction block at non-entrapment sites strongly suggests acquired demyelination.

"5:1 rule" for recruitment: if firing rate of fastest unit ÷ number of units exceeds 5, recruitment is reduced (neurogenic).

Fibrillation potentials take 2-3 weeks to develop after acute axonal injury — early needle EMG may be falsely negative for spontaneous activity.

Always confirm supramaximal stimulation by increasing 20-30% past the maximum CMAP.

Martin-Gruber anastomosis (~15-20%) can mimic ulnar conduction block recording APB with median stimulation.

In MG: clinically-weak muscles give the highest yield for both RNS and SFEMG.

Low baseline CMAP with normal motor exam = think LEMS; check post-exercise increment.

Gold Coast criteria (2020) for ALS: progressive impairment + UMN+LMN in 1 region OR LMN in ≥2 regions, plus exclusion of mimics.

EAN/PNS 2021 CIDP: simplified to "CIDP" or "possible CIDP"; variants now formally named (distal, multifocal, focal, motor, sensory).

EAN/PNS 2023 GBS: IVIg or PE in non-ambulatory patients, ideally within 2 weeks; do not give corticosteroids; do not give a second IVIg course based on poor response.

Always exclude technical artifact (temperature, electrode position, distance measurement) before declaring a finding pathological.

CUpdated References (2020–2026)

C.1 Foundational textbooks

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C.3 Selected technical & clinical reviews

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C.4 Disease-specific updates

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C.5 Useful online resources

AANEM (American Association of Neuromuscular & Electrodiagnostic Medicine): aanem.org — practice guidelines, position statements

EAN guidelines: ean.org/research/guidelines

Peripheral Nerve Society: pnsociety.com

WFN-ICNN (World Federation of Neurology, International Congress of Clinical Neurophysiology): wfneurology.org

Neuromuscular Disease Center (Washington University): neuromuscular.wustl.edu