Electrodiagnostic Assessment of Myopathy

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Myopathy: Issues for Electromyographers

Often perceived as challenging...
- Less common than neurogenic disorders
- NCS often unhelpful
- EMG abnormalities can be mild or patchy
- EMG abnormalities can be nonspecific

Important to let the clinical picture guide EDX assessment

EDX in Myopathies
Utility

1. Distinguish between muscle and non-muscle disease
2. Characterize (e.g., irritable vs. non-irritable) and focus differential diagnosis
3. Selection of muscle for biopsy

EDX in Myopathies
Limitations

1. Cannot determine specific cause
2. Relation between clinical and electrical changes may be poor (e.g., steroid myopathy)
3. Confounding by co-existing disease (e.g., neuropathy)
4. Anticoagulation

Causes of Myopathy

Inherited
- Muscular dystrophies
- Mitochondrial
- Metabolic
- Channelopathies
- Congenital

Acquired
- Inflammatory
- Endocrine
- Toxic
- Critical illness
- Infections

EDX findings depend on the degree to which disease affects muscle’s electrical (membrane) function

Nerve Conduction Studies in Myopathy

- Usually normal
- Routine motor NCS record from distal muscles
- If abnormal: Low CMAP amplitudes
- Sensory NCS normal unless coexisting neuropathy
- Utility is mainly in excluding alternative Dx
  - Poly(radiculo)neuropathy
  - Neuromuscular junction disorders
  - Motor neuron disease
What nerve conduction studies should be done when myopathy is suspected?

- ≥1 motor and 1 sensory in upper limb
- ≥1 motor and 1 sensory in lower limb
- Expand if abnormal
- Ulnar motor pre/post MVC if CMAP amplitudes low, i.e., possibility of LEMS
- 2-3 Hz repetitive stimulation if possibility of myasthenia

Needle EMG

- Muscle selection should be guided by weakness
- “Screening” needle exam
  - 1 distal & 2 proximal muscles in upper and lower limbs
  - Paraspinals (thoracic or upper lumbar)
- Inflammatory myopathy
- Pompe disease
- EMG dominant side & biopsy contralateral to EMG

Case 1

- 60 year old woman
- 3 month Hx myalgias
- Proximal limb, neck weakness
- Dysphagia requiring PEG tube
- Rash
- CK 1200

EMG: Muscle Irritability

- Normal muscle
  - No irritability
- Necrosis/Denervation
  - Fibrillations/PSW

Pathological Correlates of Irritable Myopathy

- Necrosis
- Fibre Splitting
- Inflammation

Irritable Myopathies

- Inflammatory myopathies
- Muscular dystrophies
- Rhabdomyolysis
- Toxic myopathies (some)
- Congenital myopathies (some)
- Trauma
- Infections
Case 2

- 45 year old Syrian man
- Weakness from infancy, non-progressive
- Able to walk with crutches and leg braces at age 10
- Knee and hip contractures; hip dysplasia; scoliosis
- No upper limb, bulbar or ocular Sx
- 3/8 siblings, 3/7 children affected
- Prox>distal weakness and atrophy, maximal in quads (grade 3)
- Normal CK

Genetic testing:
- Pathogenic variant (14582G>A) in RYR1
- AD central core disease

EDX findings depend on the degree to which disease affects muscle’s electrical (membrane) function

Case 2

- 60 year old woman
- 5 year Hx myalgias and progressive weakness, mainly lower limbs
- Brother with increased CK
- Large calves; mild weakness shoulders/quads; unable to stand on toes
- CK 857 U/L
- EMG:
  - Small motor unit potentials
  - Insertional activity increased in paraspinals but reduced in calf

EMG Reporting

**INTERPRETATION:** The findings are those of a proximal myopathy without distinctive electromyographic features. There is no evidence of a peripheral neuropathy or a defect in neuromuscular transmission.

"Bland" myopathy

**INTERPRETATION:** The EMG findings are those of a severe, active proximal myopathy. The presence of fibrillation potentials suggests underlying necrosis, fiber splitting and/or vascular change, and would be typical of an inflammatory myopathy.

- Decreased or absent insertional activity suggests fibrosis
- Implies chronic muscle damage
- Common in dystrophies but not unique to them
Myotonia

- Muscle fibre action potentials
- Classic *waxing and waning* in frequency +/- amplitude
- May present as waning-only discharges indistinguishable from a train of positive sharp waves
- Not found in every muscle (or patient) with myotonic disorders
  - Rarely absent in DM1
  - More often absent (16%) or minimal (16%) in DM2
  (Young 2010)

Myotonia DDx

Myotonic disorders
- Myotonic dystrophy
- Myotonia congenita
- Paramyotonia congenita
- Hyperkalemic PP

Other myopathies
- Pompe
- Myotubular
- Inflammatory
- Toxins
  - Chloroquine
  - Statins

EMG: Motor Unit Potentials

Myopathic Motor Unit Potentials

- Duration: short
- Amplitude: low
- Phases: may be increased

Motor unit duration: Normal vs. myositis
Motor Unit Size: What is “normal”?

- Quite variable
- Increasing size with age
- Sex?
- Specific muscles
  - Relatively small: biceps, ilopsoas
  - Relatively large: triceps, distal leg muscles
  - Vastus lateralis > vastus medialis
- Polyphasia (deltoid)

Quantitative EMG

- Occasionally helpful when MUP size is borderline
- Time consuming
- 20 MUPs: mean duration
- Compare to normative data

Case 4

- 67 year old man
- 2 year hx of progressive difficulty holding a golf club and opening jars
- Falls, especially coming down stairs
- Severe weakness in finger and wrist flexors & quads
- Mild weakness biceps & triceps
- CK 600 U/L

Mixed MUPs in Chronic Myopathy

Motor Unit Recruitment

- Neurogenic
- Myopathic

Number of Motor Units Firing
• Muscle involvement in myopathy is often patchy
• Needle EMG should be thorough and systematic

Case 5
• 74 year old woman
• Admitted 6 weeks ago for hip fracture treated with IM nail
• Hospital course complicated by CHF and delirium, now improving
• Slow to mobilize
• Proximal weakness noted by physiotherapist, maximal in hip flexors
• CK 60 U/L
• EMG requested: “Rule out myopathy”

Proximal Weakness with “Normal” EMG
• Not myopathy
  – Deconditioning
  – MSK disease, pain inhibition
• Myopathy
  – Steroid
  – Congenital
  – Mitochondrial
• NMJ disorders

Case 6
• 41 year old man
• Progressive proximal weakness over 2 years
• No sensory or craniobulbar Sx
• Weak neck flexion, proximal U/E & L/E muscles
• Mildly reduced reflexes
• Normal sensory exam
• CK 1600

Kennedy’s disease
(Bulbar and spinal muscular atrophy)
Exercise Testing

- Useful in assessment of channelopathies (myotonic disorders & PP)
- Short test: CMAP amplitude @ baseline/10s exercise/Q10s for 1 minute
- Long test: CMAP amplitude @ baseline/5m exercise/Q1-2 min for up to 1 hour

Short Exercise Test

Mainly useful in assessment of myotonic disorders

• Normal: -10% to +20% amplitude change
• Sensitivity 83%–100% in PC, 53%–100% in MC, 60% in SCM

Long Exercise Test

Mainly useful in assessment of periodic paralysis

• Specificity ~98%
• Sensitivity ~80% for Ca and Na channelopathies; low for Cl (Kuntzer 2000)

Selected References