ROLE OF EMG IN DIAGNOSIS OF MYOPATHY

Associate Professor
Steve Vucic

Western Clinical School, University of Sydney

Department of Neurology

Westmead Hospital

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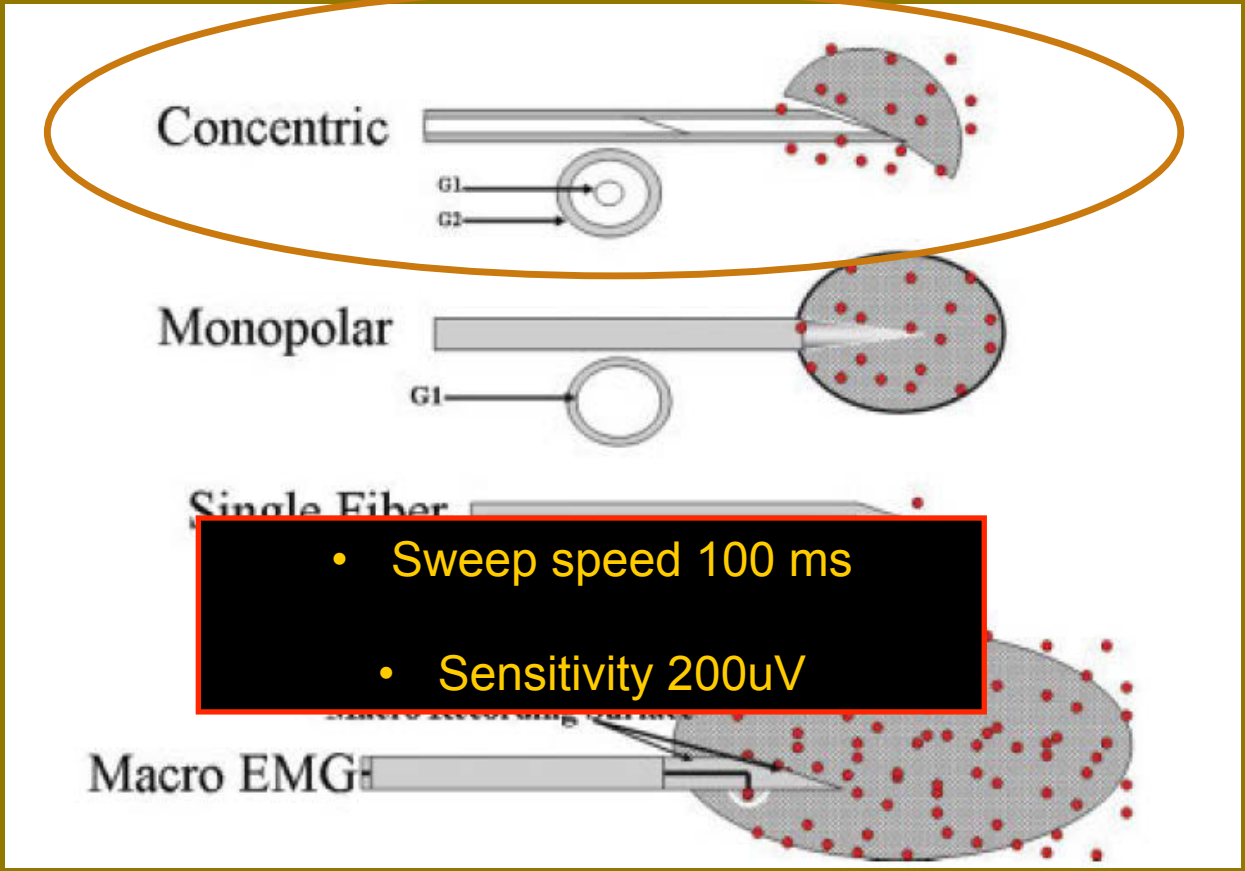
EMG and myopathy

- Infer the presence of myopathy
  - "Myopathic –like"
- Process may be patchy
- Qualitative
- Quantitative techniques
  - Mean vs outlier method
- **DIAGNOSTIC UTILITY**
  - Sensitivity variable
    - 33-92%
  - Specificity
    - 49-84%

EMG needles



- Sweep speed 100 ms
- Sensitivity 200uV

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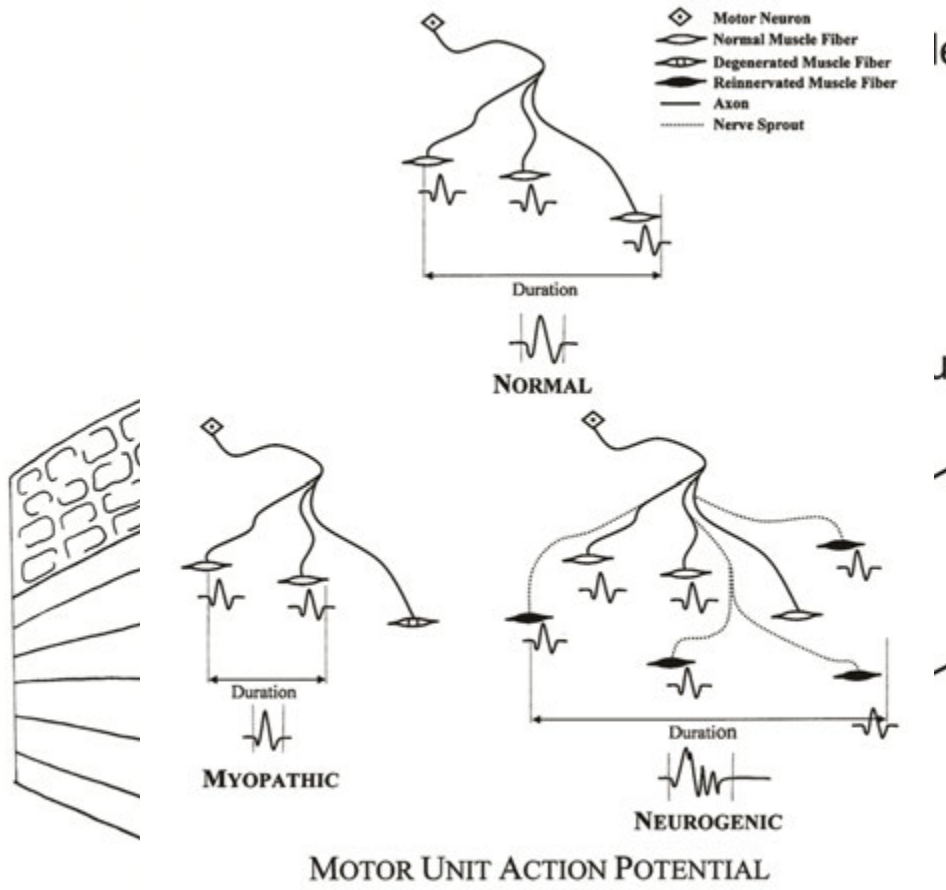
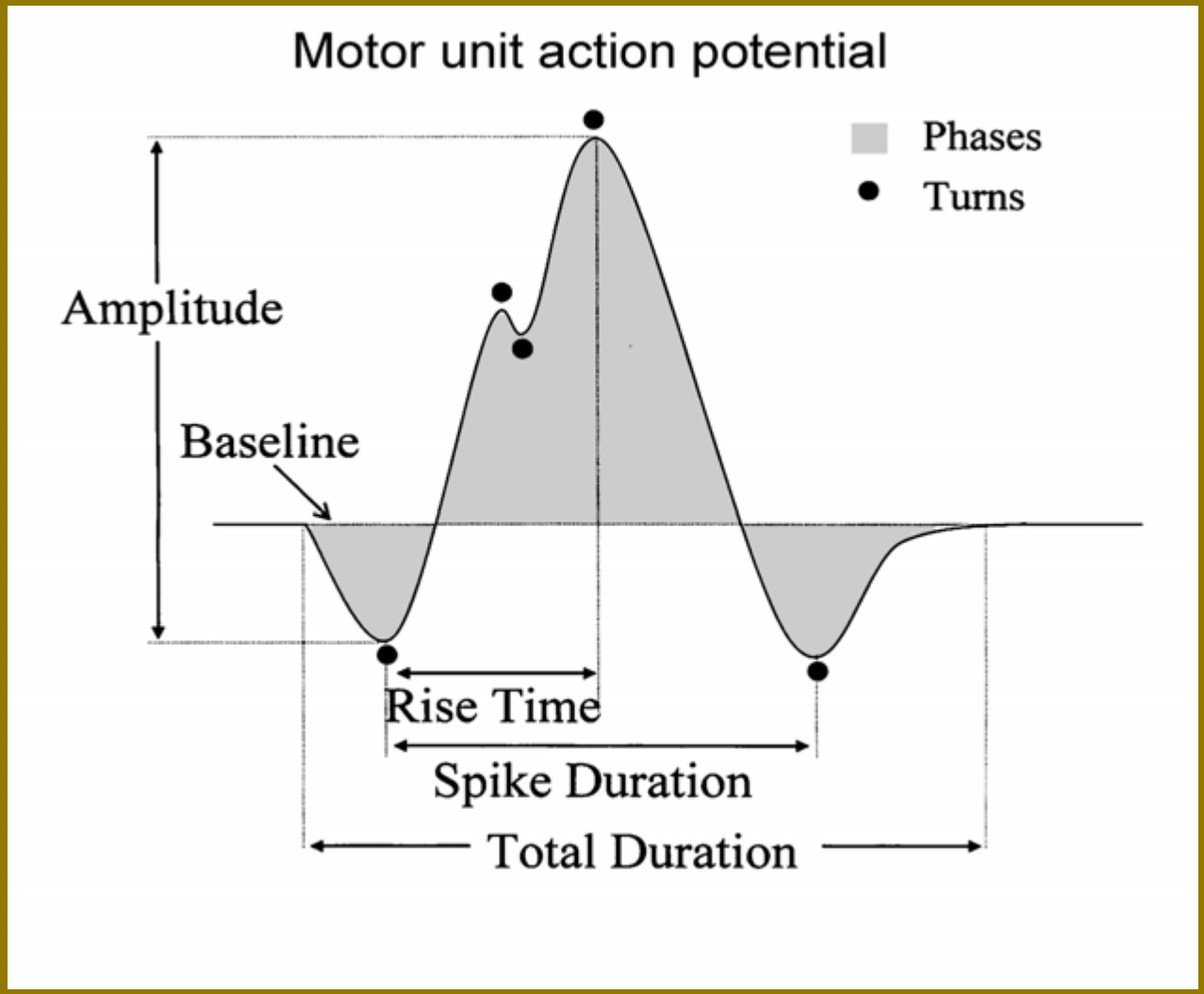
MOTOR UNIT ACTION POTENTIAL

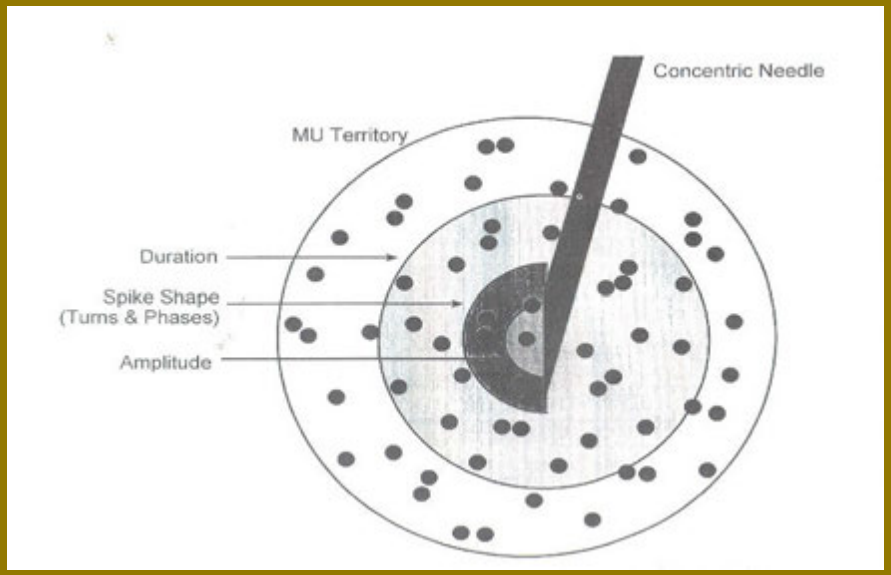


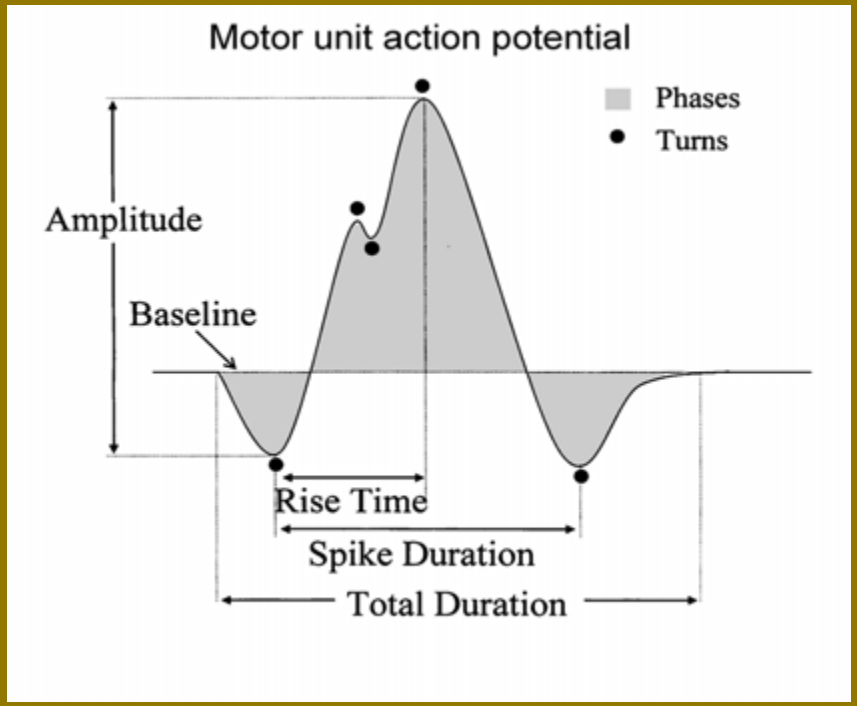
MOTOR UNIT ACTION POTENTIAL



MUAP DURATION

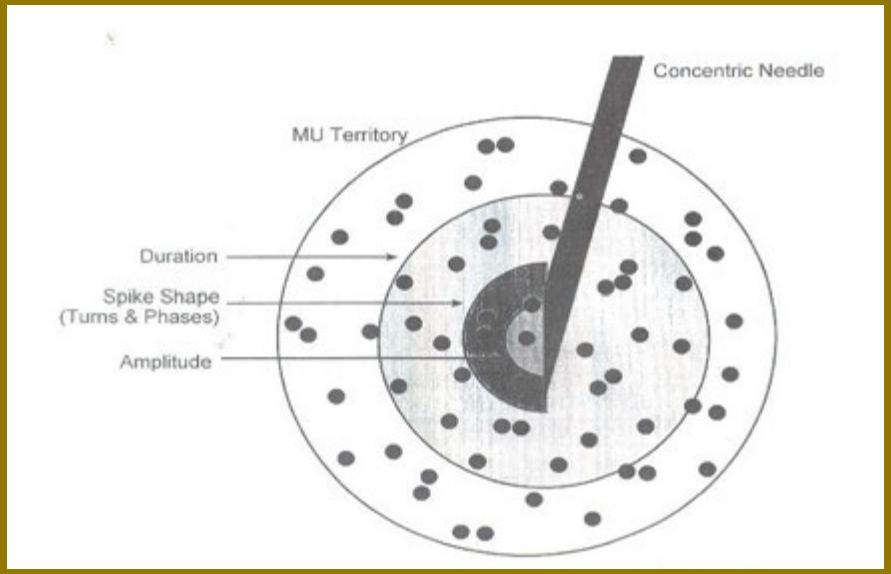
- Depends on electrical activity 2.5 mm
- Measured at high display gain
- Robust

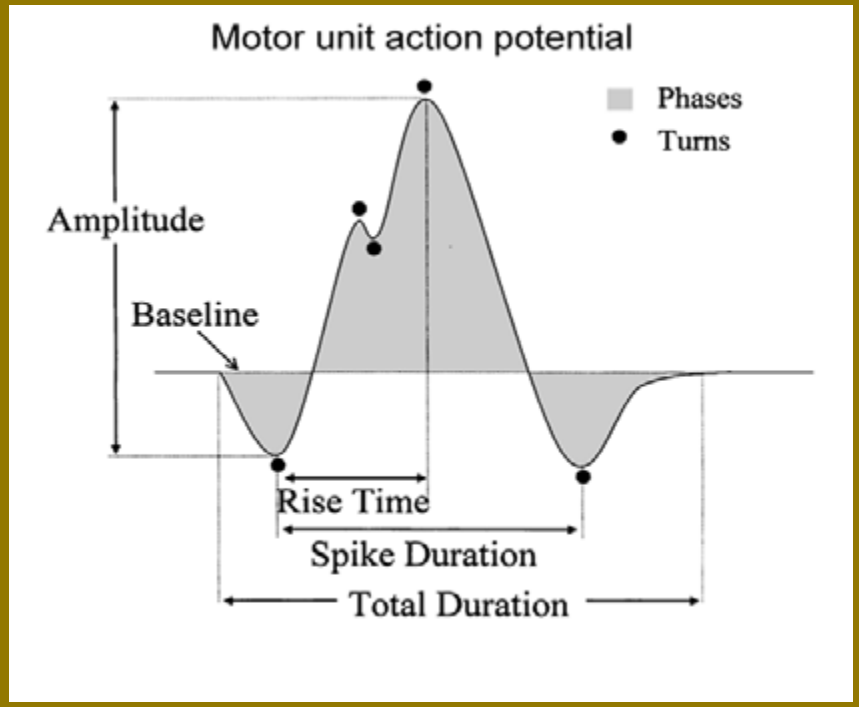




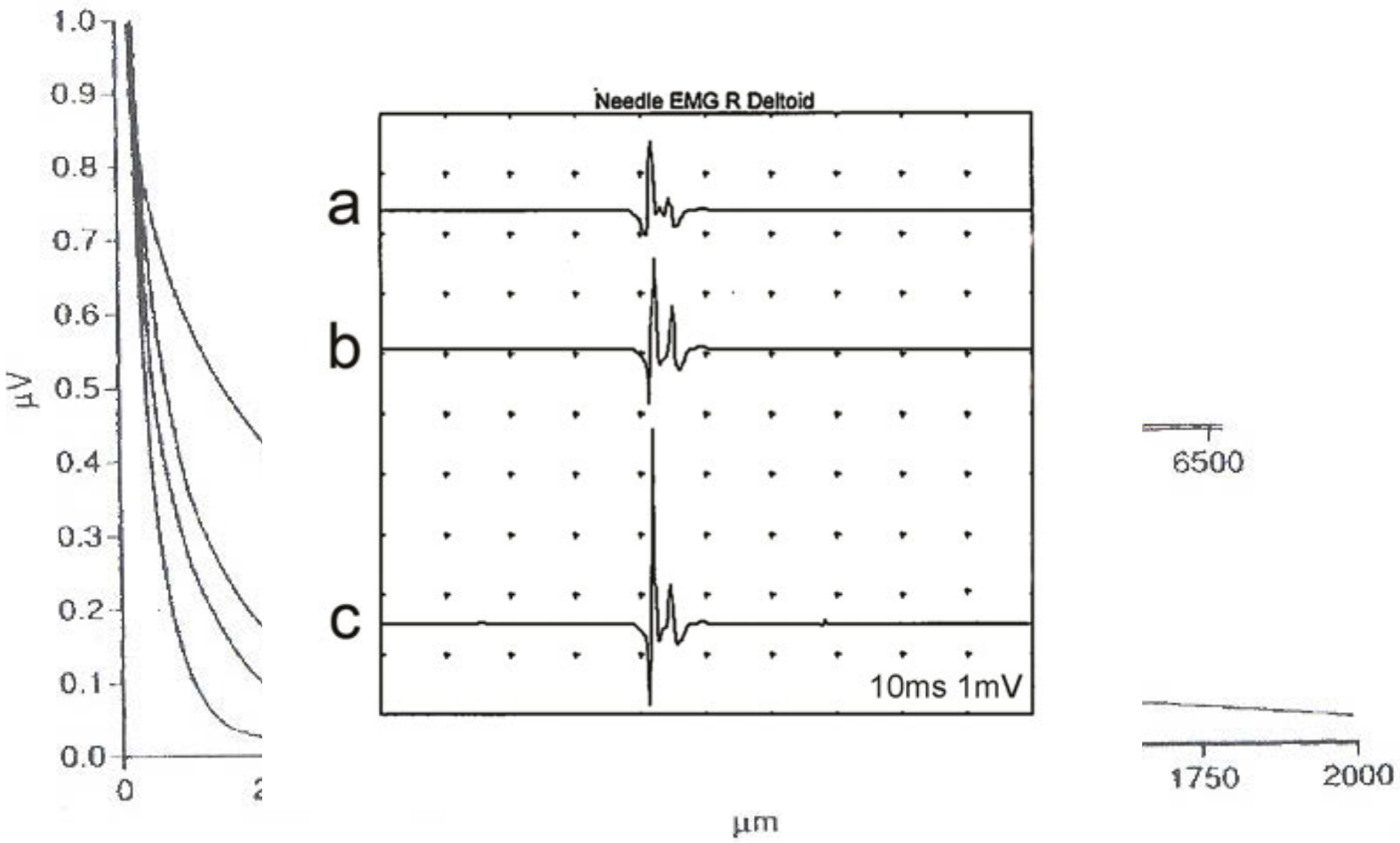
MUAP AMPLITUDE/RISE TIME

- Muscle fibers 0.5 mm
- Muscle fiber closest to the recording tip



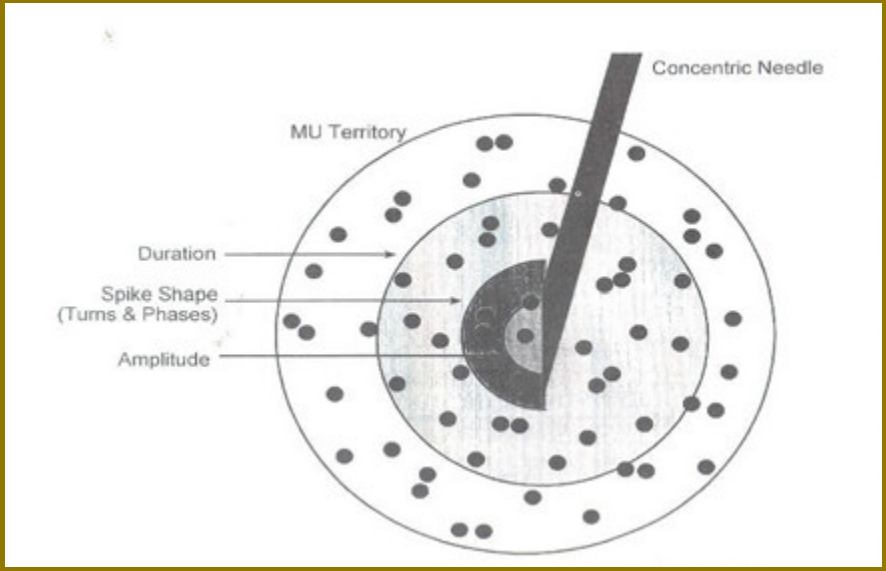


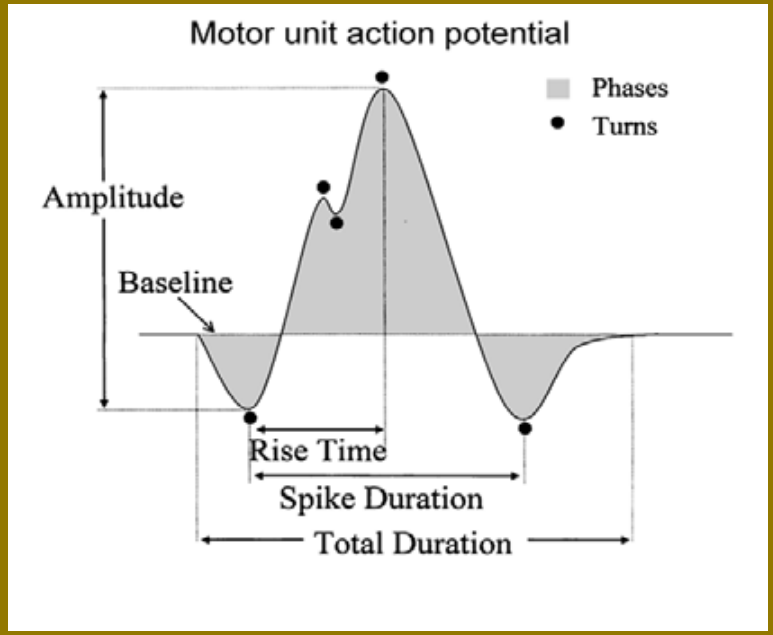
AMPLITUDE CHANGE WITH DISTANCE



MUAP PHASE

- Muscle fibers 1 mm
- Synchrony of muscle fiber APs
- Normal ≤ 4 phases

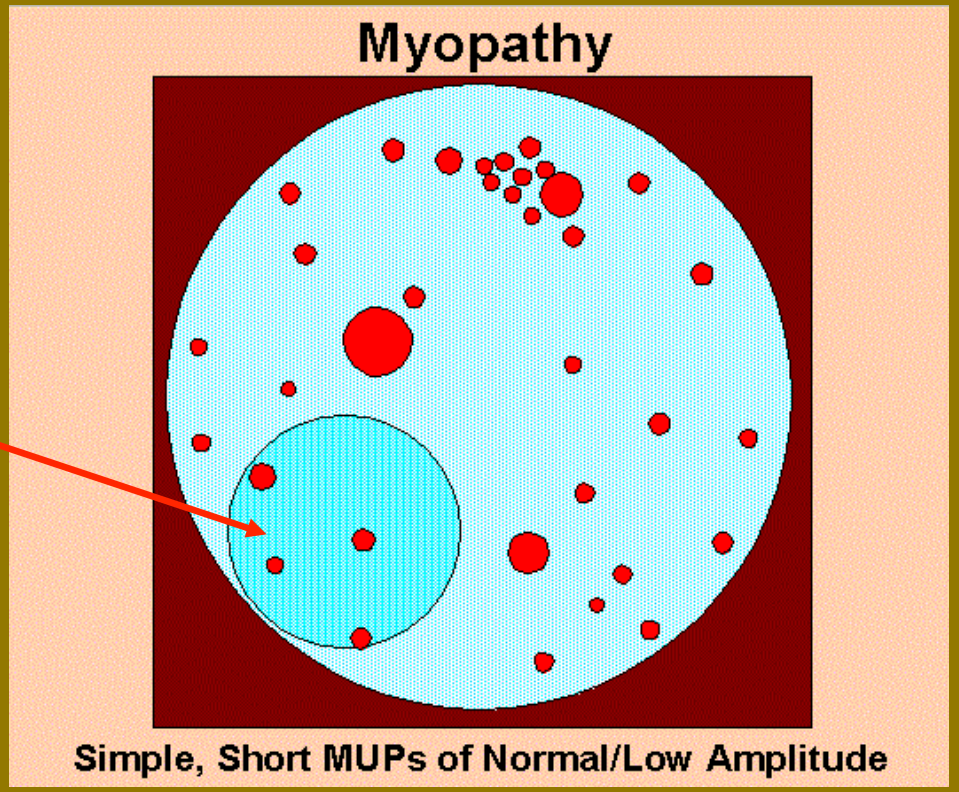




Pathological changes in myopathy

- **Loss of muscle fibers & necrosis**
  - **segmental**
- **Change in fibre size (atrophy and hypertrophy)**
- **Regeneration fibers**
- **Fibre splitting**

"MYOPATHIC-LIKE UNITS"

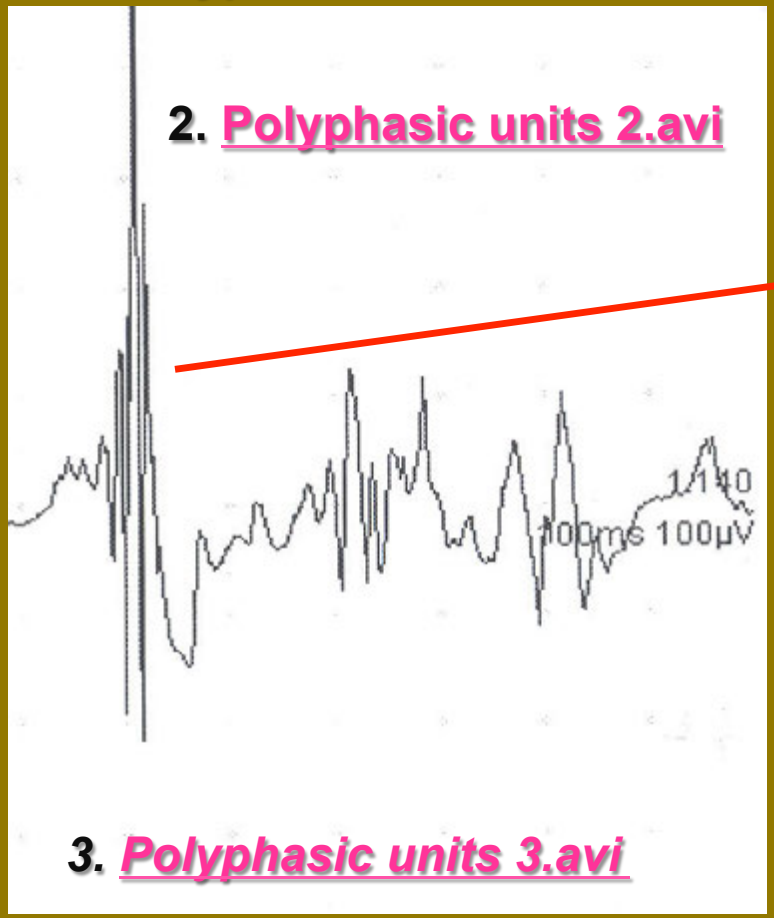


1. Small simple units 1.avi

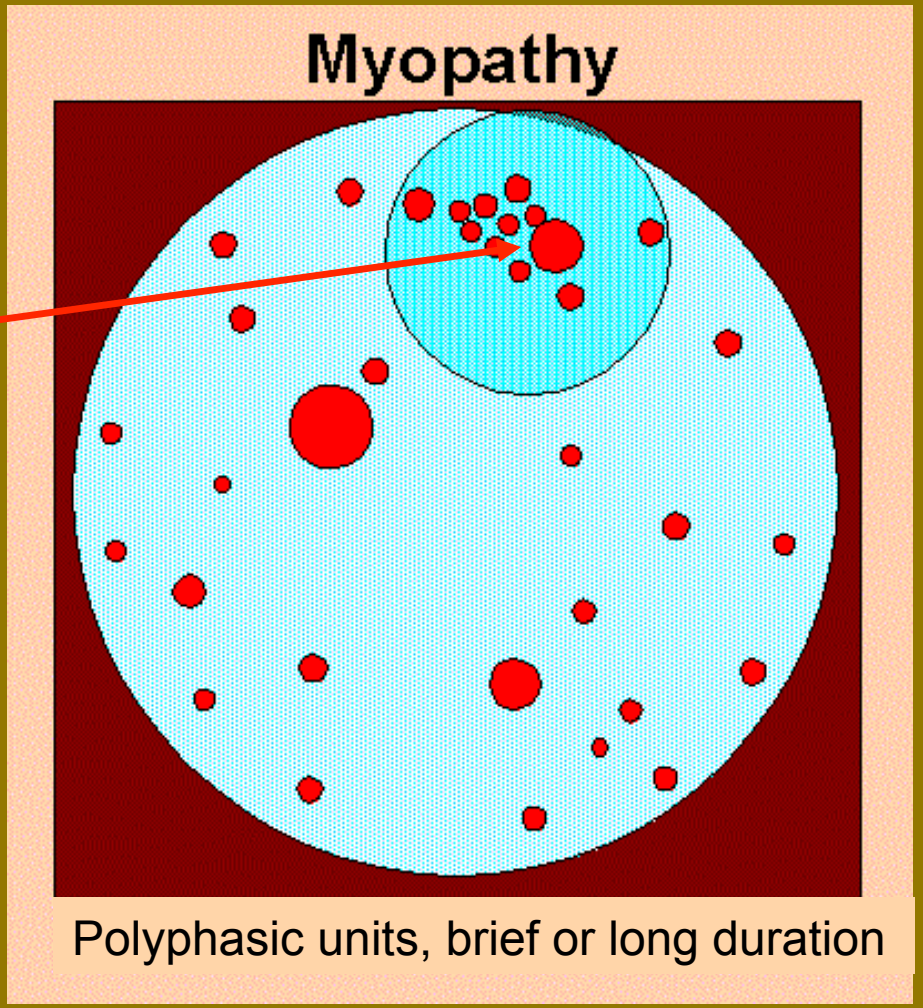
2. Small simple units 2.avi

"MYOPATHIC-LIKE UNIT"

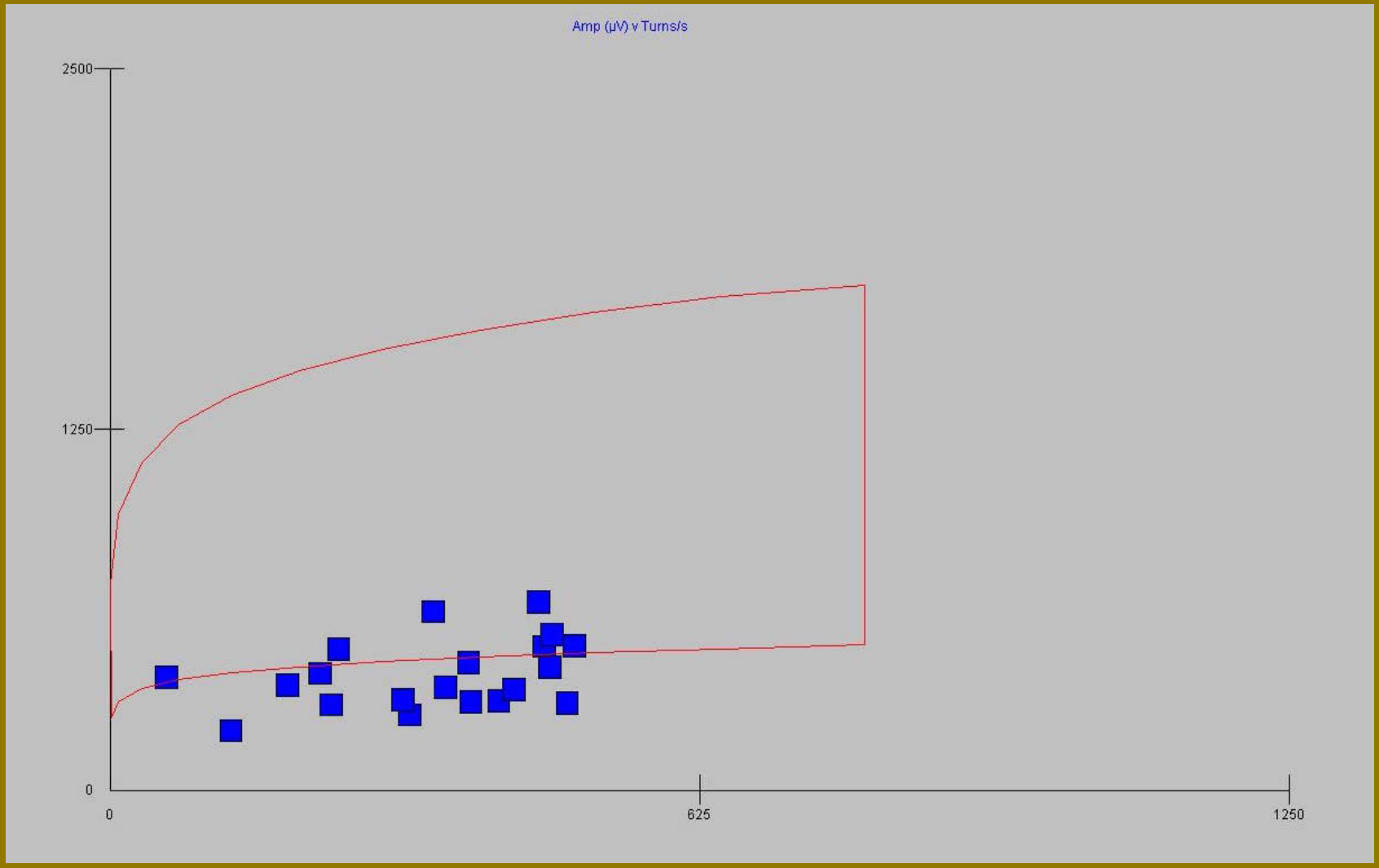
1. Polyphasic units 1.avi



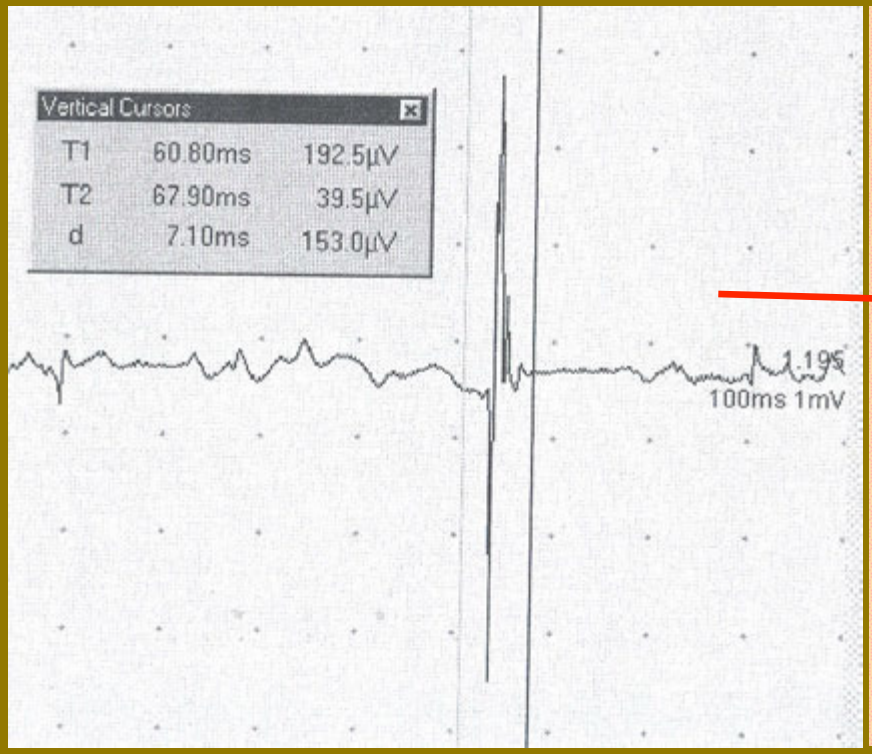
4. Polyphasic units 4.avi

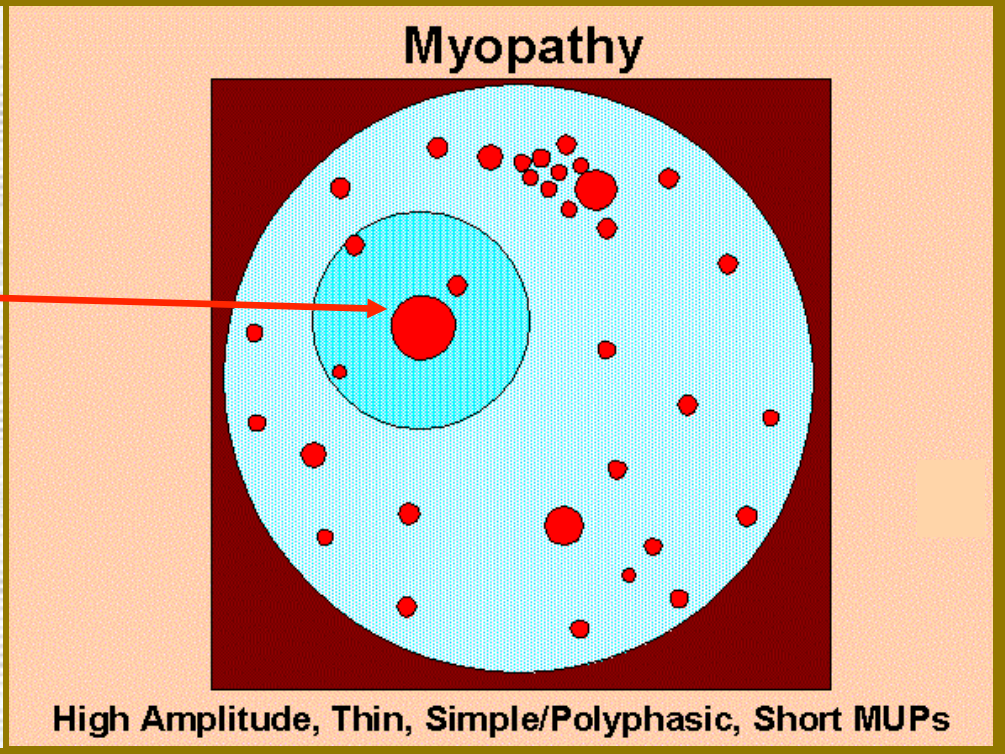


TURNS AMPLITUDE ANALYSIS



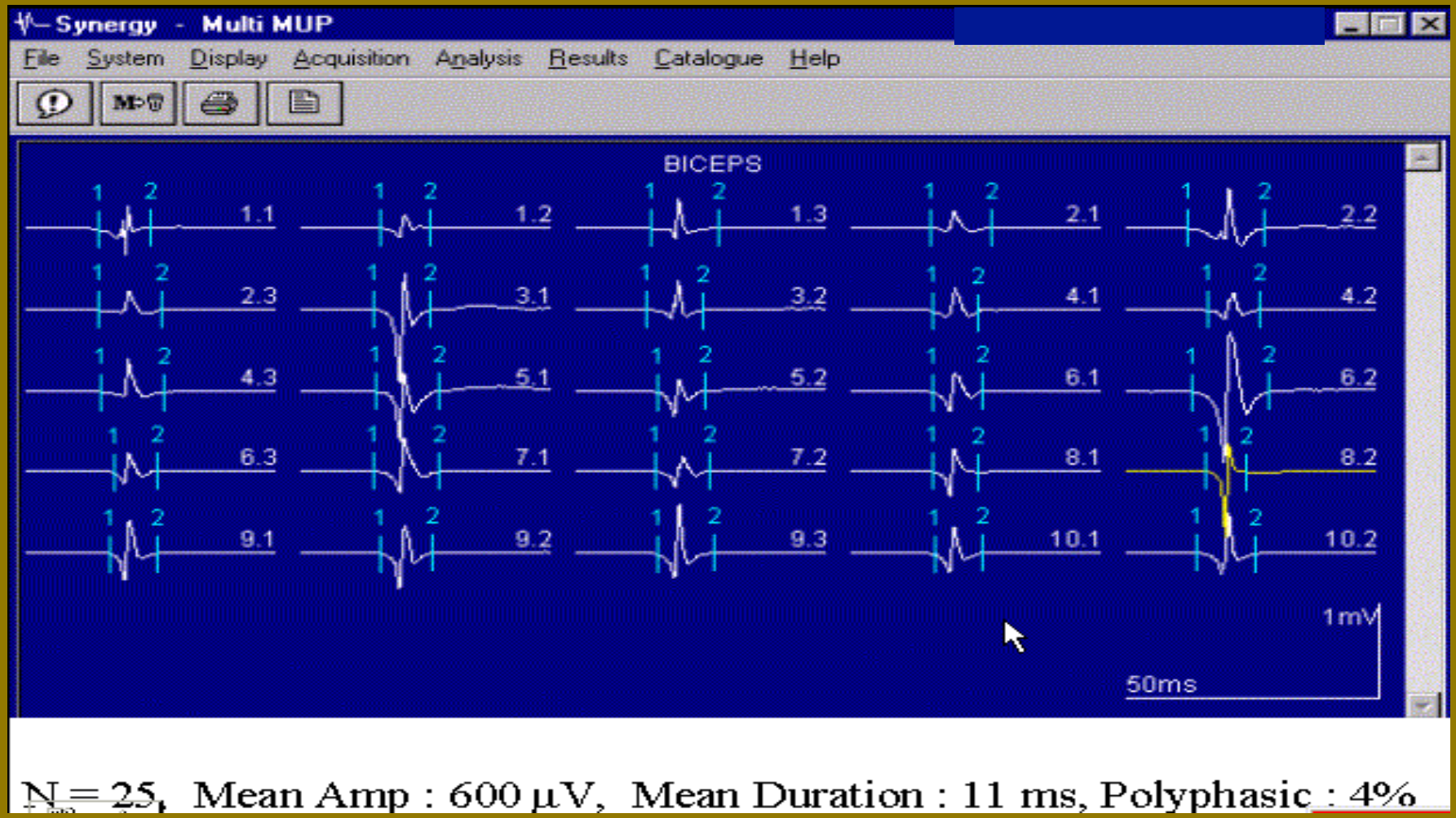
"MYOPATHIC-LIKE UNITS"





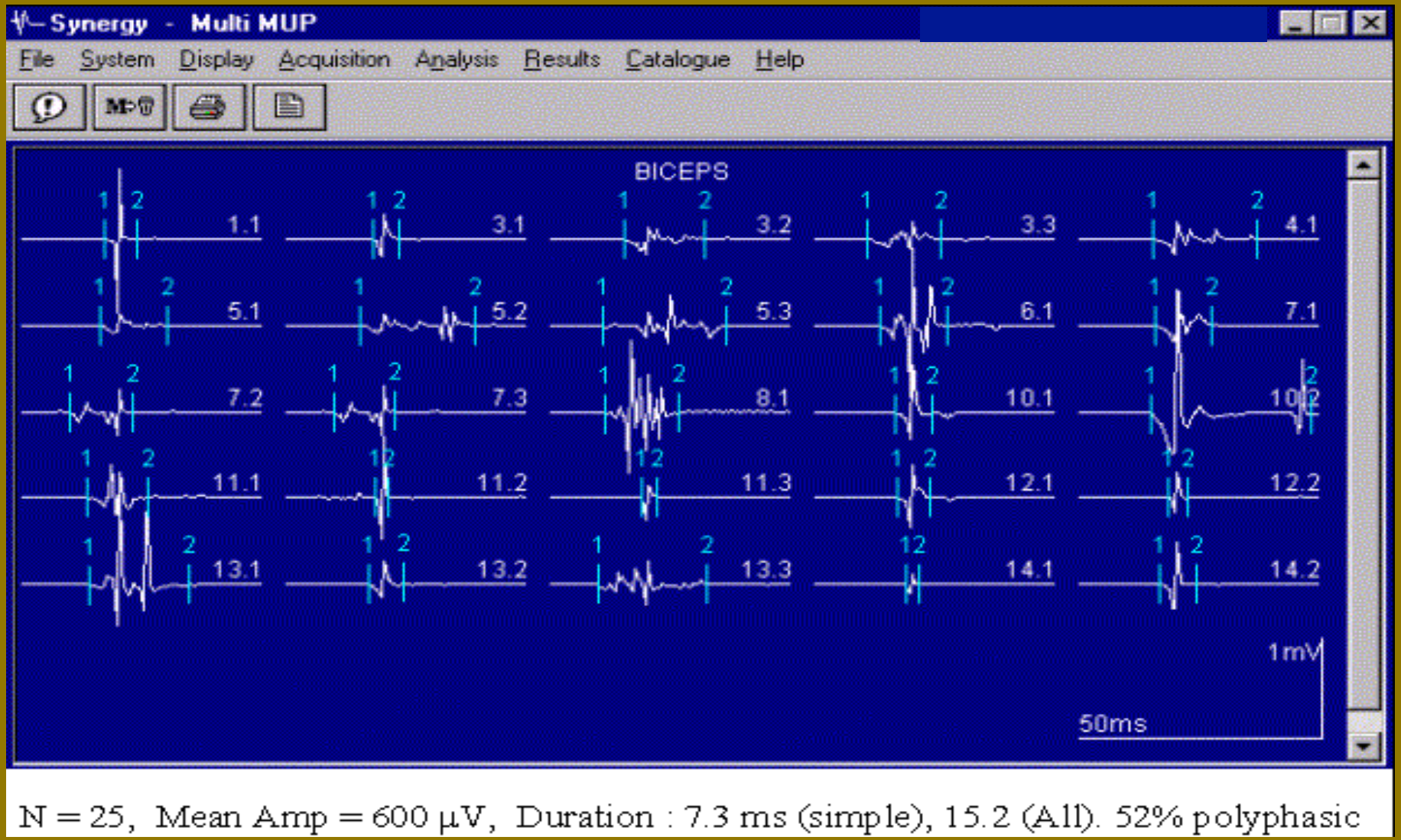
1. Large amplitude.avi

Quantitative EMG
MULTI-MUP ANALYSIS



N = 25, Mean Amp : 600 μV, Mean Duration : 11 ms, Polyphasic : 4%

QEMG IN MYOPATHY



Acta Myologica • 2011; XXX: p. 37-41

ORIGINAL ARTICLES

A correlative study of Quantitative EMG and biopsy findings in 31 patients with myopathies

E. Dardiotis[1,2], E. Papathanasiou[1], I. Vonta[3], G. Hadjigeorgiou[2], E. Zamba-Papanicolaou[1], T. Kyriakides[1]

	Myopathic findings in muscle biopsy (n = 29)	Sensitivity
Classical Q-EMG	All (n = 29)	31,0%
	M1 (n = 23)	39,1%
	M2 (n = 7)	28,6%
	M3 (n = 6)	33,3%
	M4 (n = 9)	22,2%
	M4 without M1 (n = 5)	0%
Amplitude outliers	All (n = 29)	68,9%
	M1 (n = 23)	69,5%
	M2 (n = 7)	71,4%
	M3 (n = 6)	50%
	M4 (n = 9)	77,7%
	M4 without M1(n = 5)	80%
Duration outliers	All (n = 29)	24,1%
	M1 (n = 23)	30,4%
	M2 (n = 7)	14,3%
	M3 (n = 6)	33,3%
	M4 (n = 9)	11,1%
	M4 without M1 (n = 5)	0%

- **M1= Increase fibre size variability**
- **M2 = Necrosis and/or regeneration**
- **M3 = Endomysial fibrosis and fibre loss**
- **M4 = Alteration in fibre architecture**

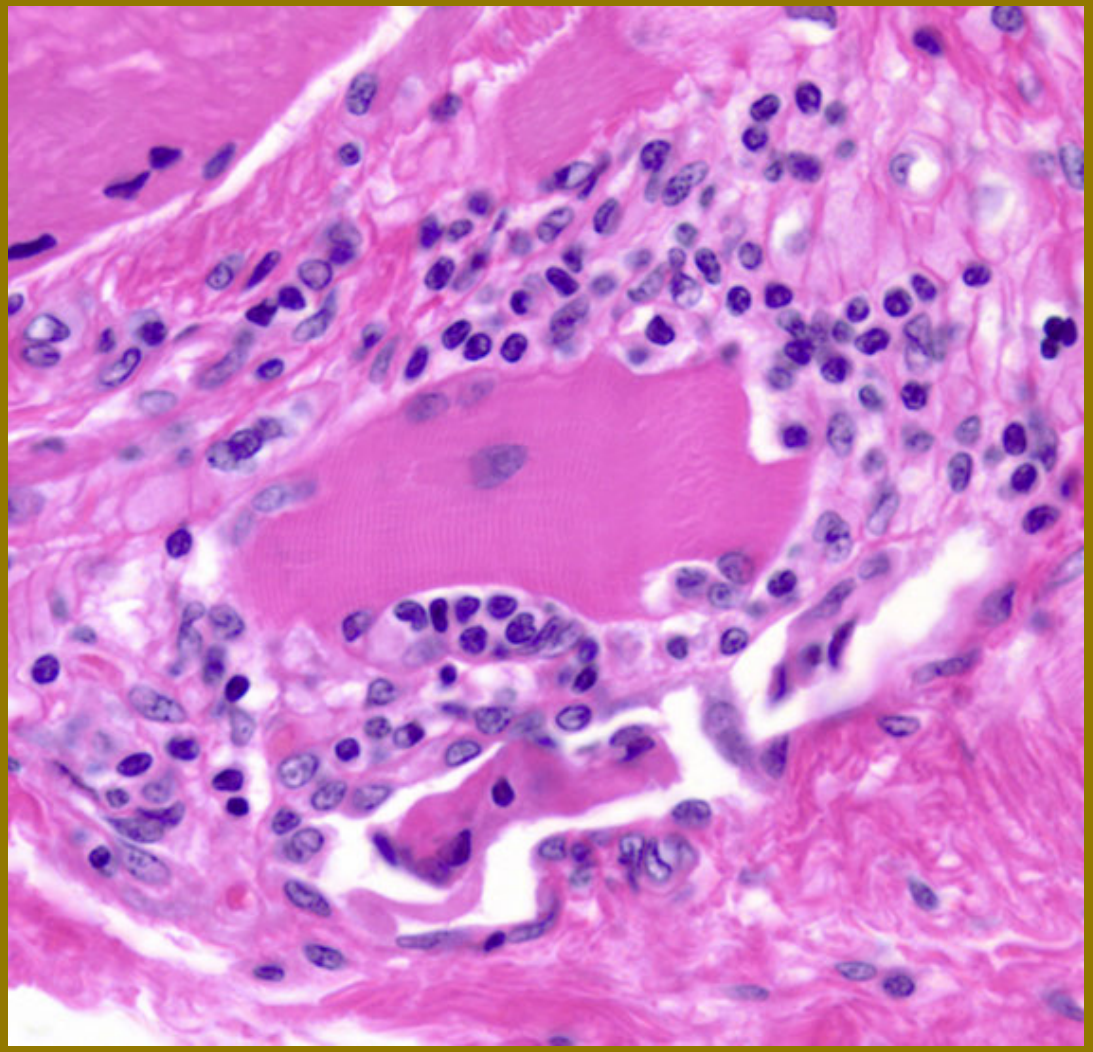
CAN EMG HELP WITH THE DIAGNOSIS OF THE UNDERLYING CAUSE OF MYOPATHY?

Accompaniments

- Spontaneous activity
- Pattern of motor unit recruitment
  - “neurogenic”= chronic myopathies
- Pattern of muscle disease

(1) Fibrillation potentials & PSW

1. F_P 1 (1).avi
2. F_P 1 (2).avi
3. F_P 3.avi



Fibs/PW

- Inflammatory myopathies
- Infiltrative myopathies
- Inclusion body myositis
- Muscle trauma
- Muscular dystrophies/Congenital myopathies
- Rhambdomyolysis
- Muscle membrane disorders (hyperkalaemia periodic paralysis)
- Toxic myopathies
- Metabolic myopathies
- Infectious myopathies

Complex repetitive discharges

1. CRD 1.avi

2. CRD 2.avi

3. CRD 3.avi

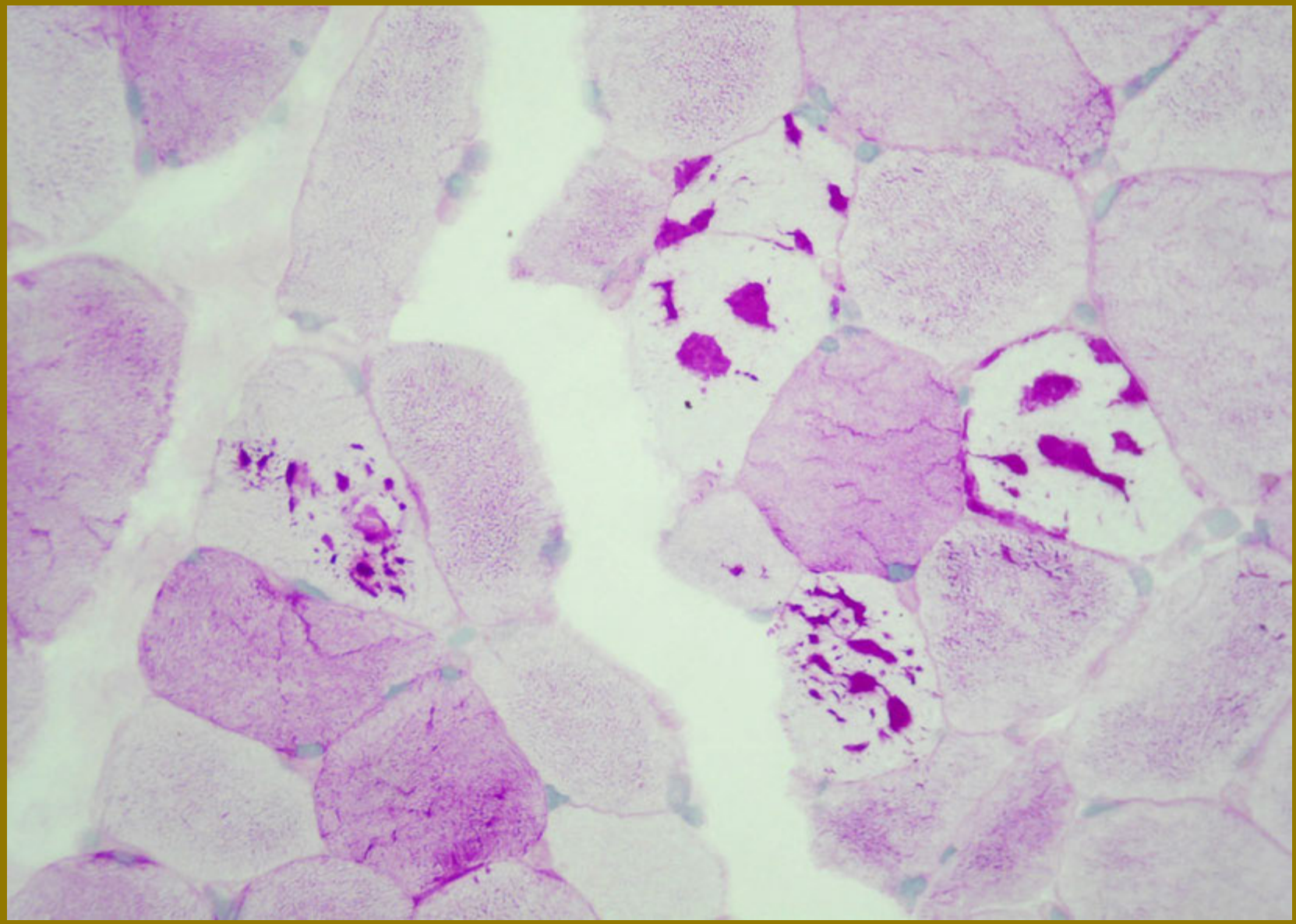
Case of myopathy with CRDs

- 55-year-old female
  - Right foot drop
    - 18 months before presentation
  - Right hand weakness
    - 3-4 months before presentation
  - Left foot drop, left hand & bilateral shoulder girdle muscle weakness
    - At presentation

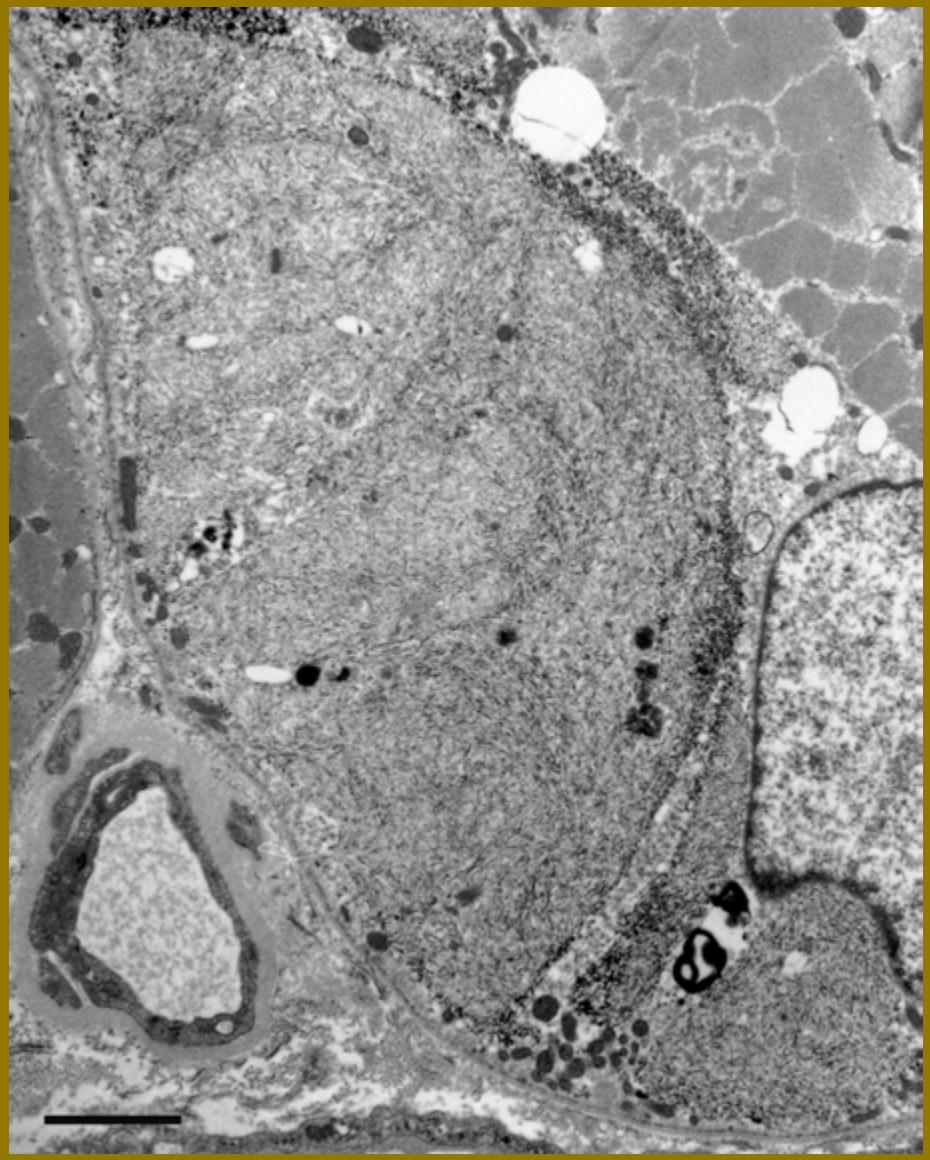
Physical examination

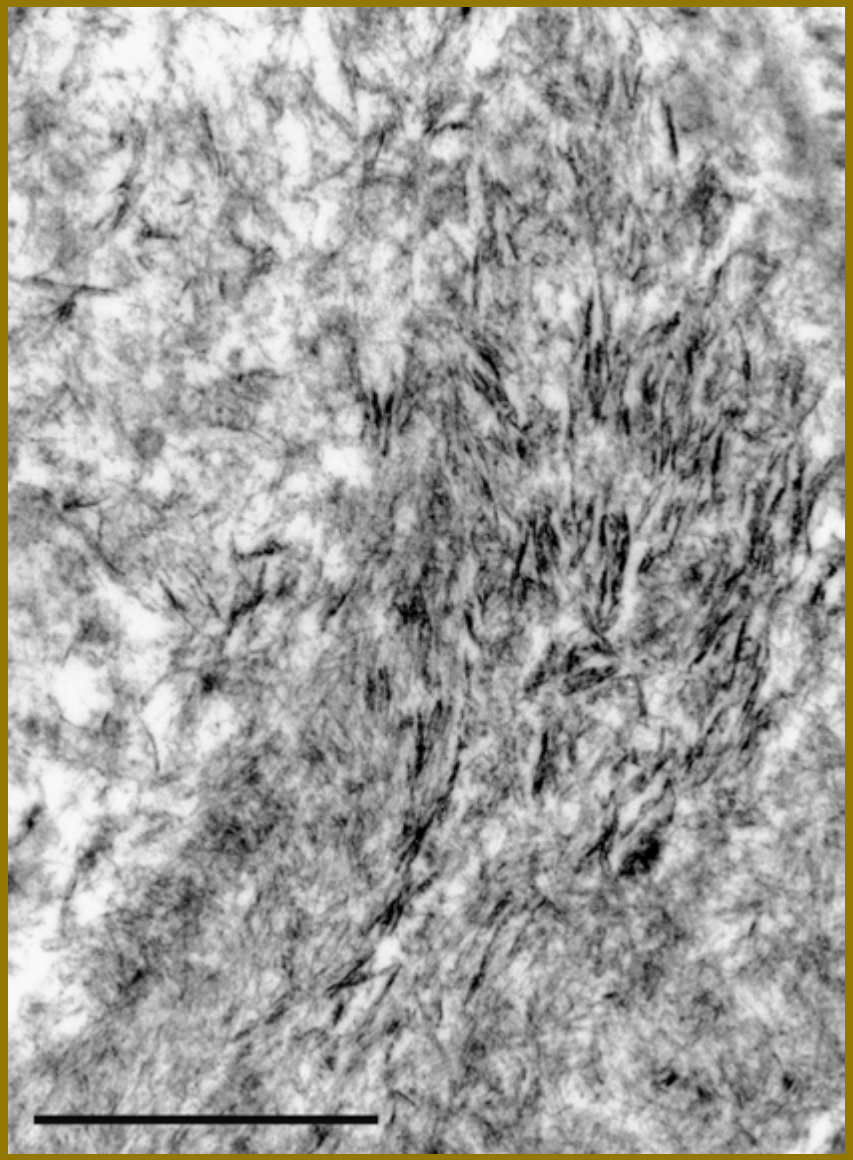
- Wasting
  - Intrinsic hand muscles
  - Right wrist & finger extensor muscles
  - Right tibialis anterior muscle
- Weakness
  - Global weakness in upper limb
    - MRC grade 3-4, Distal>Proximal
  - Distal lower limb; right>left
    - MRC grade 1-4
  - Hyperreflexia/ flexor plantar responses

Periodic acid Schiff



Polyglucosan Bodies

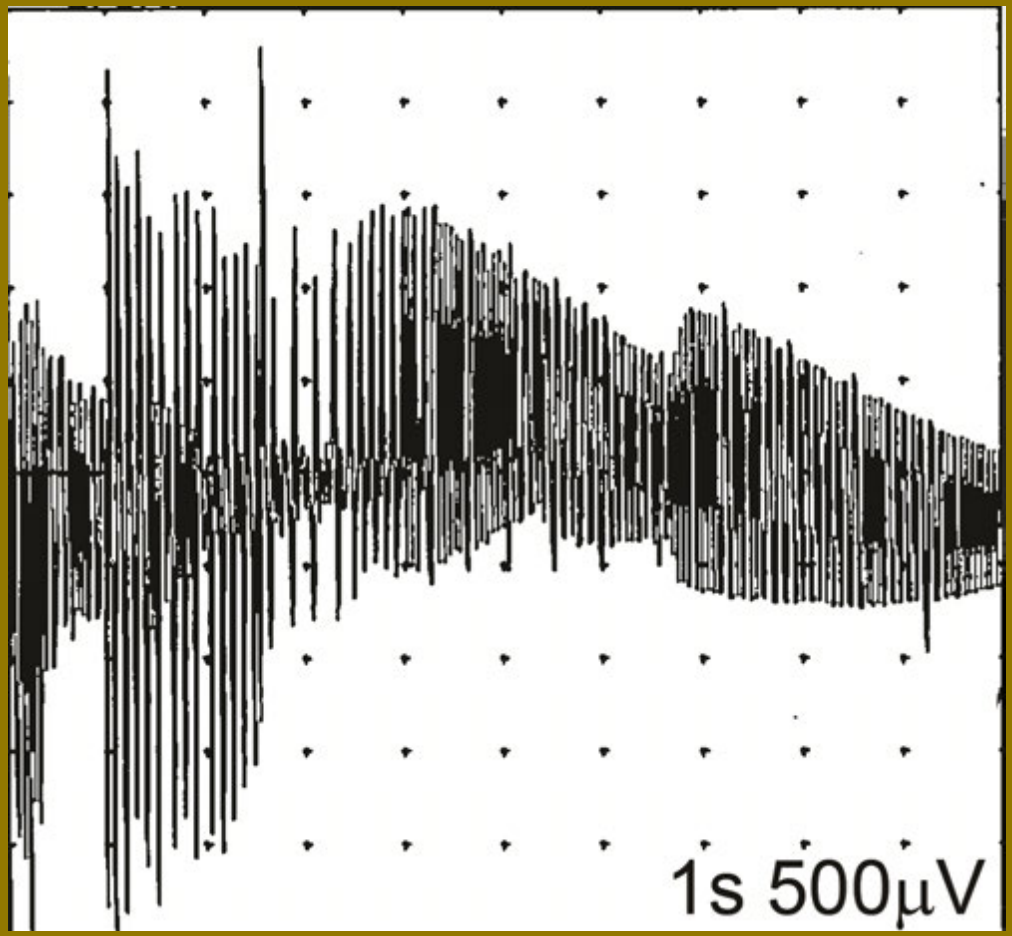




Complex repetitive discharges (CRD)

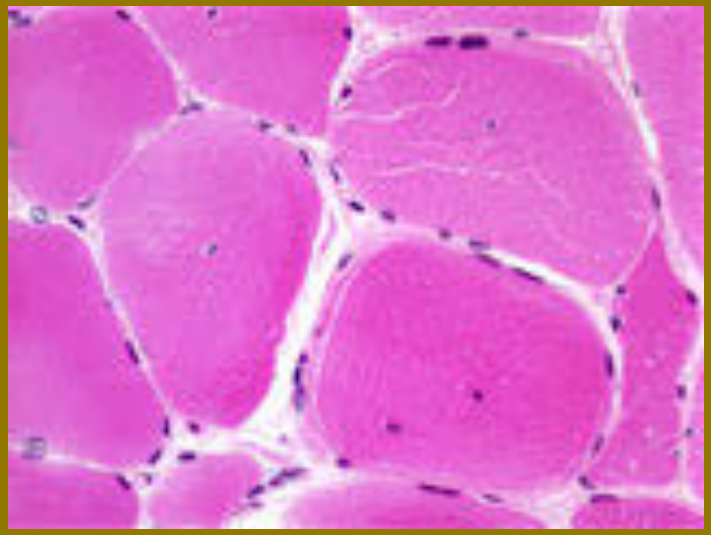
- Inflammatory myopathy
- Muscular dystrophies
- Channelopathies
- Glycogen storage disorders
  - Acid maltase deficiency
  - Debrancher deficiency
  - Polyglucosan body disease
- Myxedema
- Schwartz-Jampel syndrome
    - Rare genetic disorder
    - Mutation in HSPG2 gene (perlecan)
    - Bone dysplasia and joint contractures
    - Dwarfism

Myotonic discharges



1. Myotonic dc.avi

Case with myotonic discharges

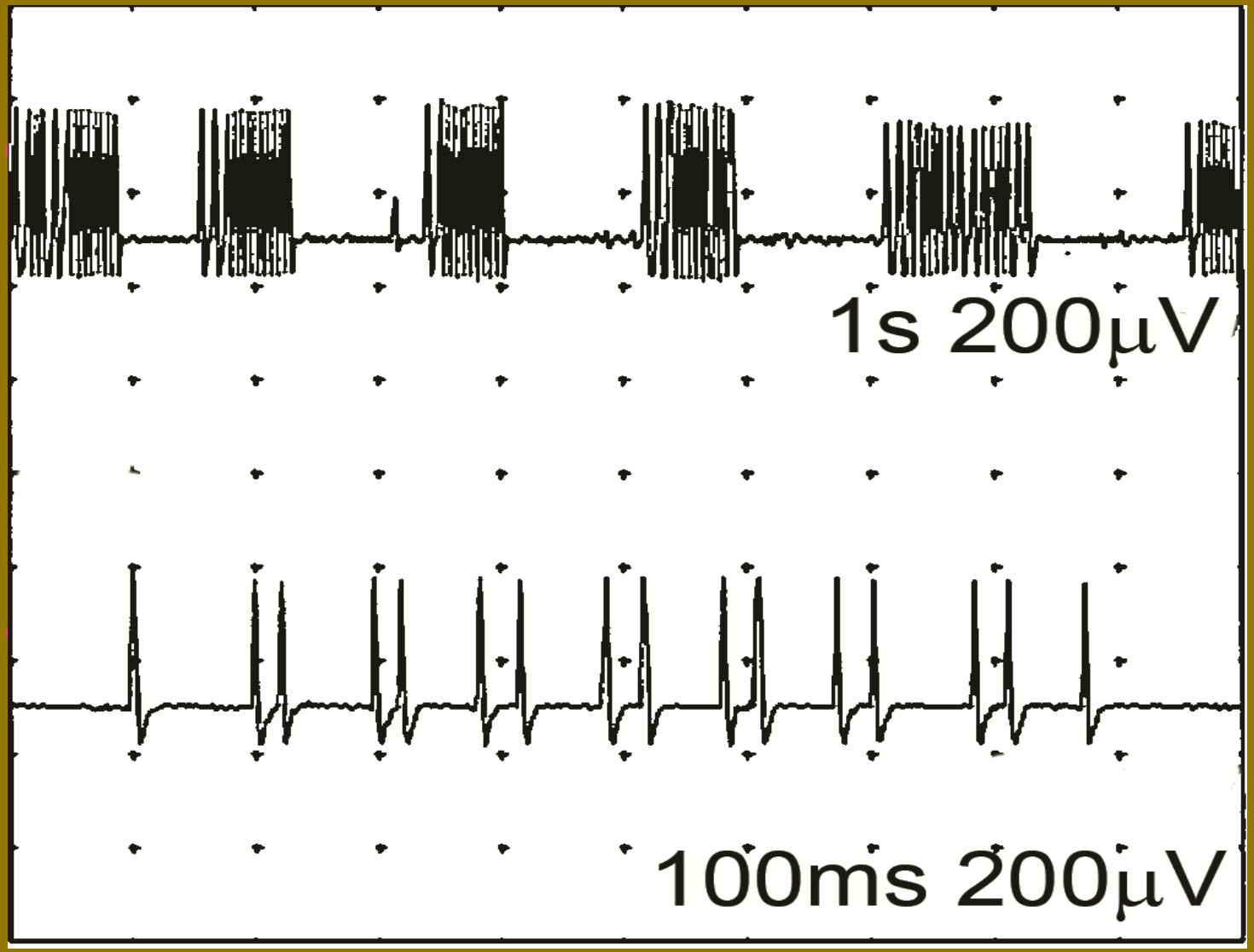


Myotonic discharges

- **Myotonic dystrophy type II (PROMM)**
- **Channelopathies**
  - **Myotonia & paramyotonia congenita**
  - **Hyperkalemic PP**
- **Glycogen storage disorders**
- **Inflammatory disease**
- **Toxic myopathies**
  - **Chloroquine, statins, cyclosporine**
- **Hypothyroid myopathy**

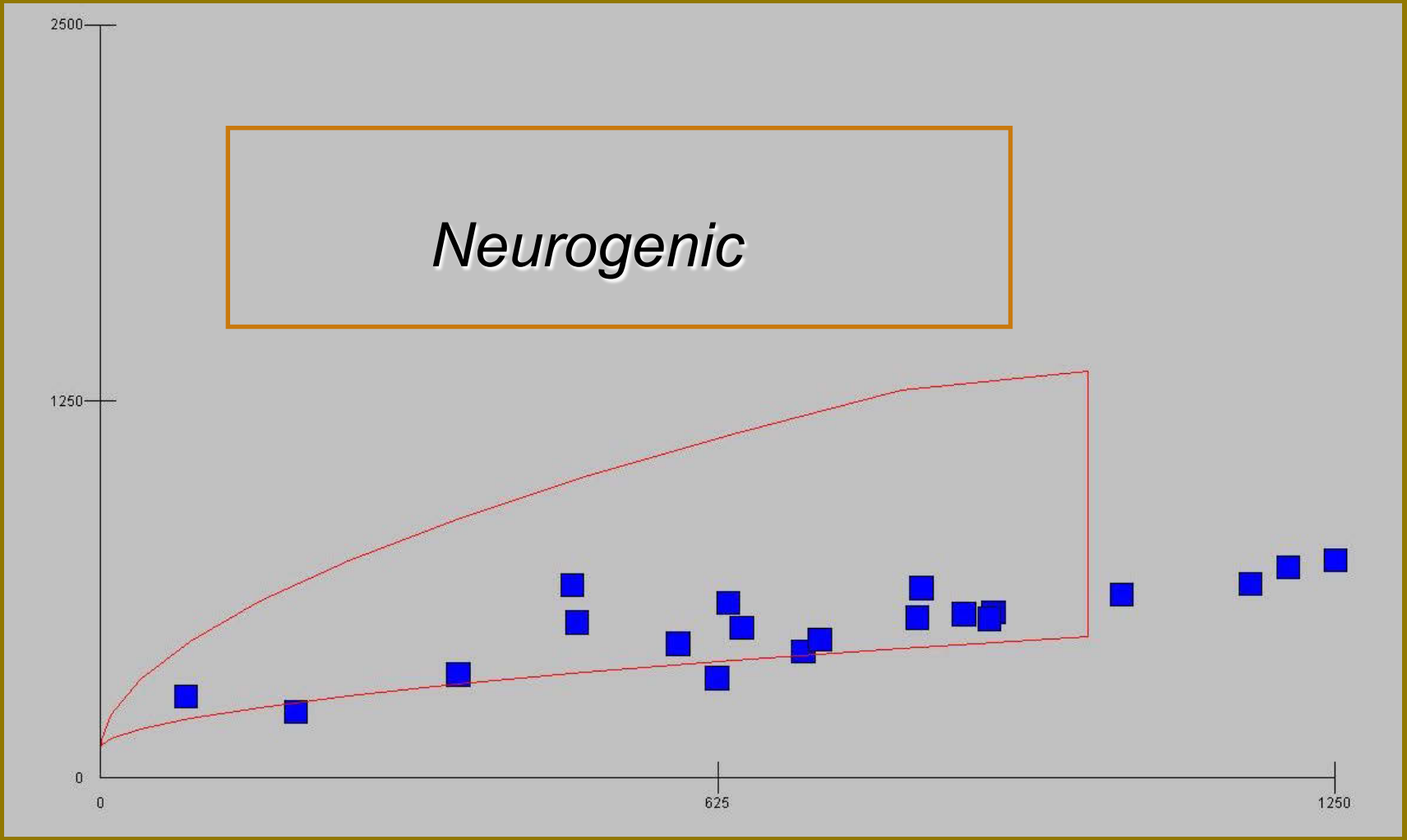
1.Myokymia.avi

MYOKYMIA



Pattern of motor unit recruitment

Inclusion body myositis



Often performed to diagnose a myopathy

1. CK levels
2. Thyrotropin, electrolytes, renal and liver function tests, complete blood count, erythrocyte sedimentation rate, and serum protein electrophoresis/immunofixation
3. Genetic testing (hypothesis-driven, step-wise; avoid panels)
4. NCS/EMG
5. Muscle biopsy (if above are nondiagnostic)

Box 4
Myopathies that may have a normal EMG

1. Type II muscle atrophy
   a. Steroid myopathy
   b. Disuse myopathy
2. Some mitochondrial myopathies
3. Some congenital myopathies
4. Metabolic myopathies with dynamic[a] phenotype if not performed during acute exacerbations
   a. GSD V (McArdle disease or myophosphorylase deficiency)
   b. GDS VII (phosphofructokinase deficiency)
   c. Carnitine palmityltransferase II deficiency

[a] Here, metabolic myopathies are referred to as having a dynamic phenotype when they are associated with exercise-induced symptoms (exertional myalgias, cramps, and myoglobinuria) as the dominant clinical features.

Conclusion

- Assess motor units
  - Quantitative EMG
- Accompaniments
  - Spontaneous activity
  - Pattern of recruitment

- MUSCLE BIOPSY

Pattern of muscle disease

Distal weakness

Weakness.mpg

Weakness.mpg

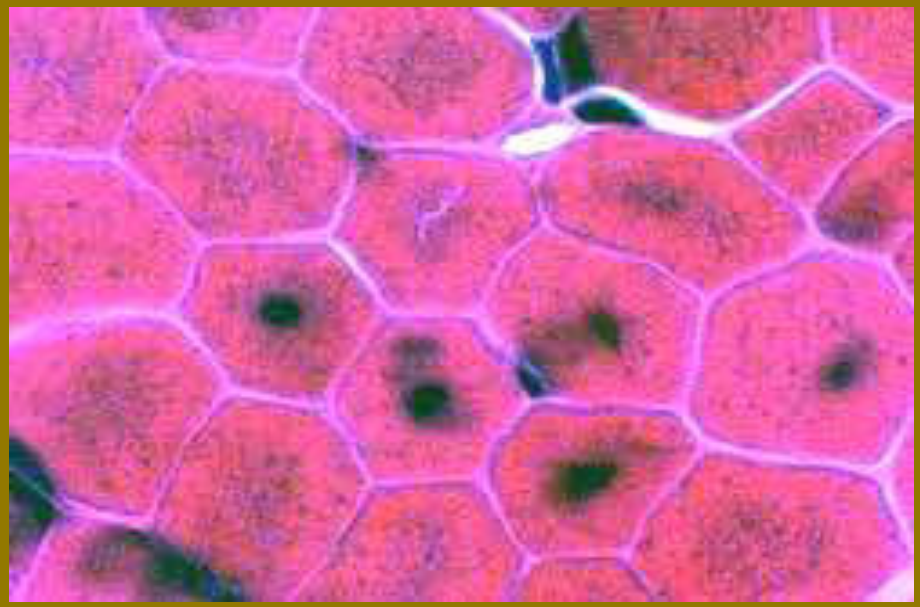
(1) Distal myopathy

- **Early onset < 40**
  - **Miyoshi myopathy**
    - **Deficiency Dysferlin**
    - **Calf muscle weakness**
  - **Laing myopathy**
    - **Skeletal/beta cardiac myosin gene (MyH7)**
    - **Chromosome 14**
    - **Weakness of toe & ankle dorsiflexor & neck flexors**
  - **Nonaka myopathy***
    - **Bilateral foot drop**
    - **N-acetylglucosamine 2 epimerase/N-acetylmannosamine kinase gene (GNE)**
- **Late onset > 40 years**
  - **Welander**
    - **2p13**
    - **Weakness of finger & wrist extensors**
  - **Markesbery-Griggs/Udd**
    - **Titin gene**
    - **Chromosome 2**
    - **Ankle dorsiflexor**

Other myopathies that present with distal weakness

- Myotonic dystrophy
- Inflammatory myopathies
  - IBM
- Metabolic myopathies

- Hereditary myopathies
  - Emery-Dreifuss
  - OPD
- FSH
- Congenital myopathies
  - Nemaline
  - Centronuclear
  - Central core



Other patterns

- Quadriceps and wrist & finger extensors
  - IBM
- FSH (FSH FACE.mpg, FSH anterior.mpg, FSH POSTERIOR VIEW.mpg)
- Scapuloperoneal pattern (Prox. arm & distal leg)
  - Scapuloperoneal dystrophy
  - E-D
  - LGMD1B (laminopathies)
  - LGMD2A (calpain)
  - LGMD 2C-F (sarcoglycan)
  - Congenital myopathies
  - Acid maltase

Other patterns

- Dropped head syndrome
  - Focal myositis
    - Inflammatory myopathy
  - Isolated neck extensor myopathy
  - Congenital myopathy
  - Mitochondrial myopathy
  - Carnitine deficiency
  - MD
  - Hypothyroidism/hyper PTH
  - Myasthenic gravis

- EMG of limb muscles may be normal

Other patterns

- Ptosis with ophthalmoplegia
  - Oculopharyngeal MD
  - Oculopharyngealdistal MD
  - CPEO (mitochondrial myopathy)
  - NMS (MG)
- Ptosis without ophthalmoplegia
  - Myotonic dystrophy
  - Congenital myopathies
  - Desmin (myofibrillar) myopathy