

Electrodiagnostic Studies

What?

When?

Why?

Why not?

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Europe's last case of wild poliomyelitis



AĞRI-TURKEY

26 NOV 1998

Now, he is 13 years old.

All our efforts are to keep him as the last child suffering from polio in the Region.



Electrodiagnostic Studies

What?

When?

Why?

Why not?



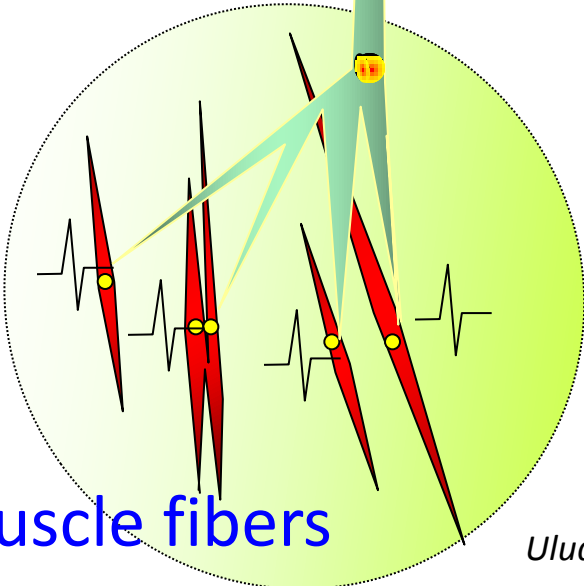
WHAT??

diagnostic techniques used to evaluate
structure of a motor unit electrophysiologically

Motor Neuron

Motor Unit

Axon



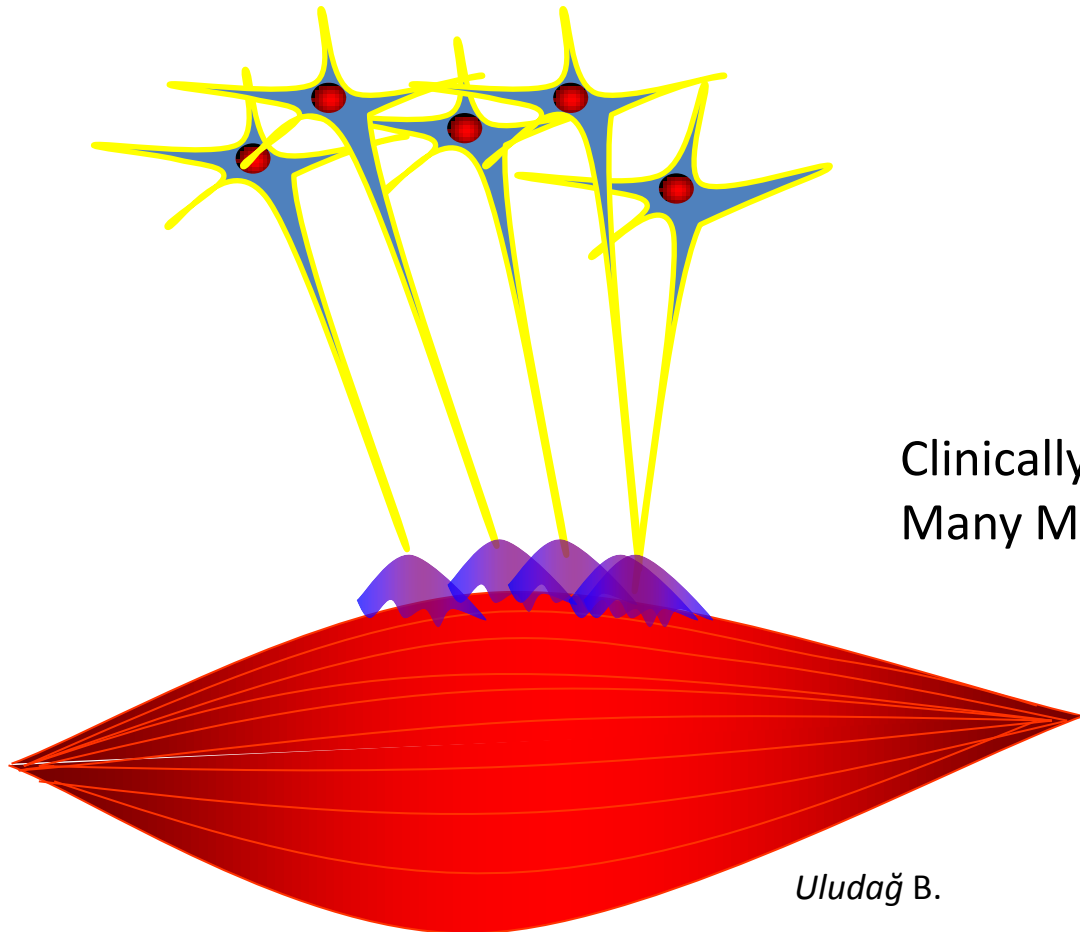
Muscle fibers



SMUP

Uludağ B.

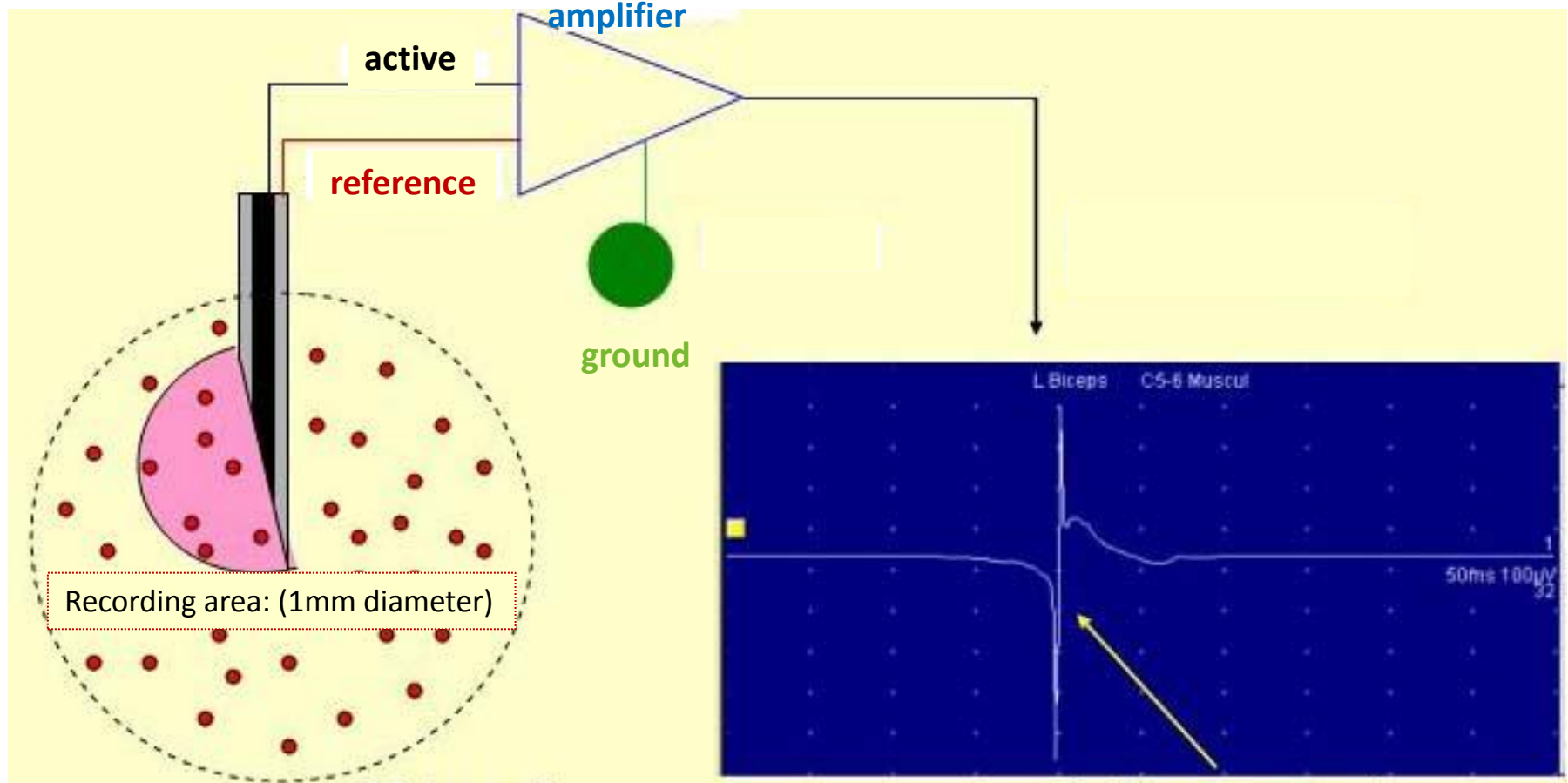
Motor Neuron Pool



Clinically apparent muscle contraction
Many MUPs

Uludağ B.

Conventional Needle EMG



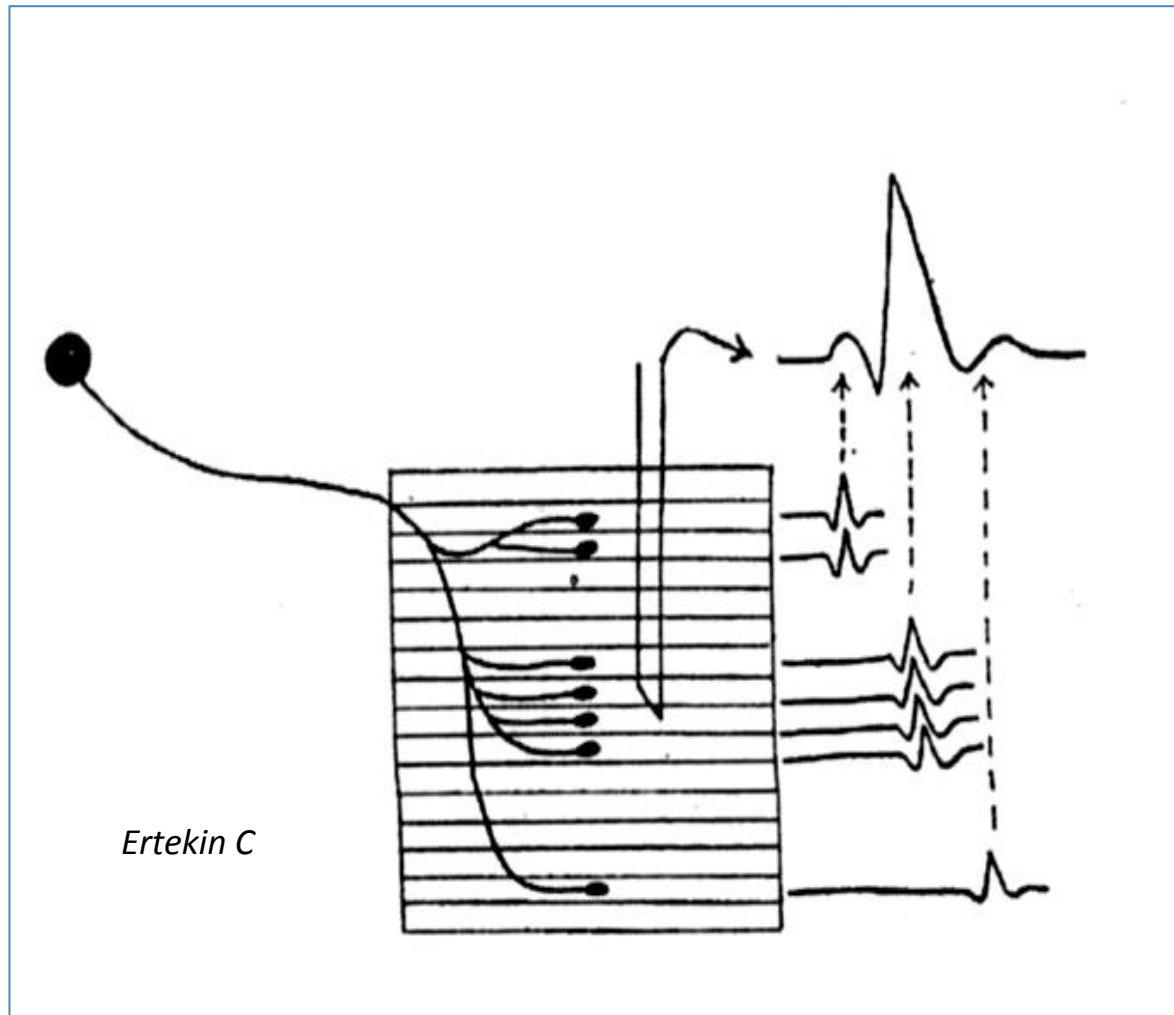
motor unit (2 to 10mm diameter)

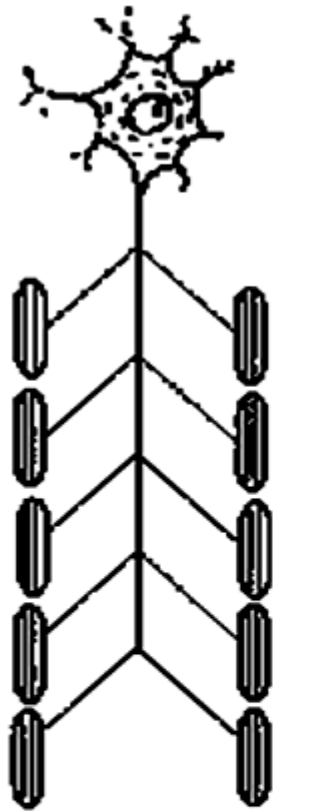
motor unit action potential(MUP)

Signals from 2-15 muscle fibers

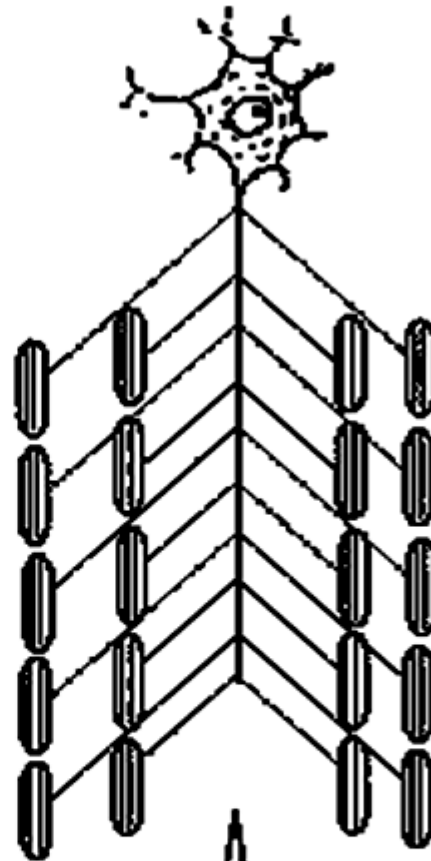
Amplitude, duration of a MUP:

number and size of muscle fibers in the motor unit,
synchronicity of their action potentials





normal



neurogenic

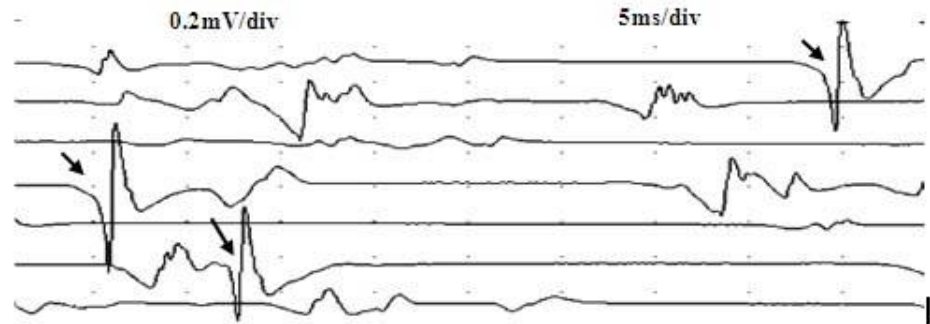
Observations in an EMG study

1. at rest

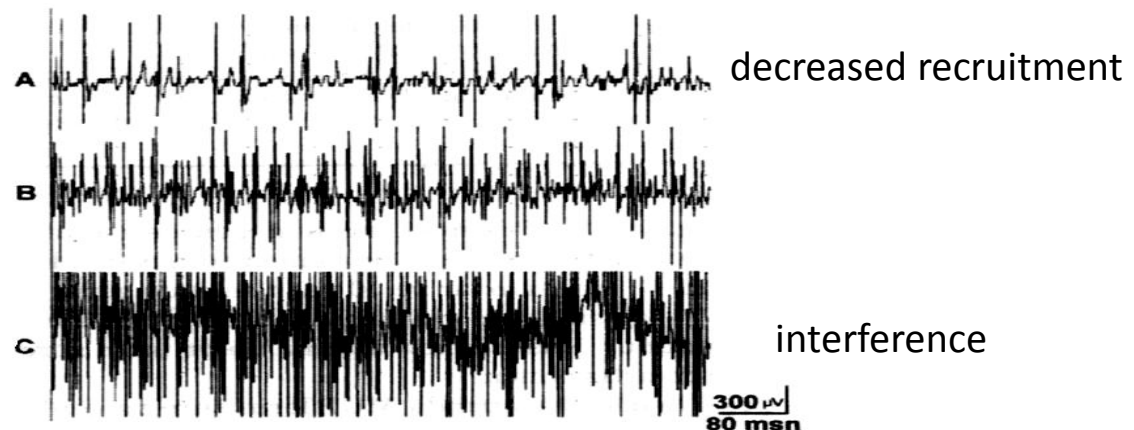


Normally silent
Active denervation

2. with slight contraction

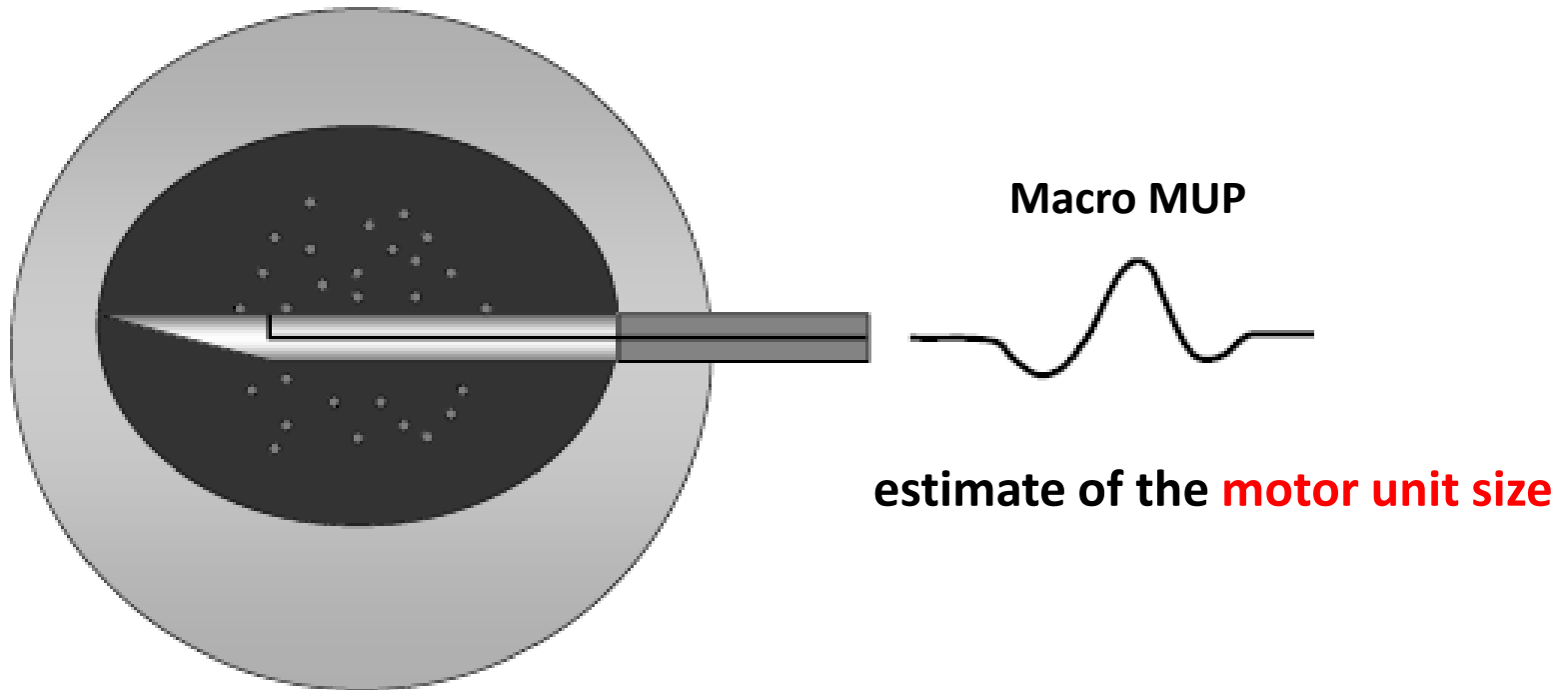


3. at full effort



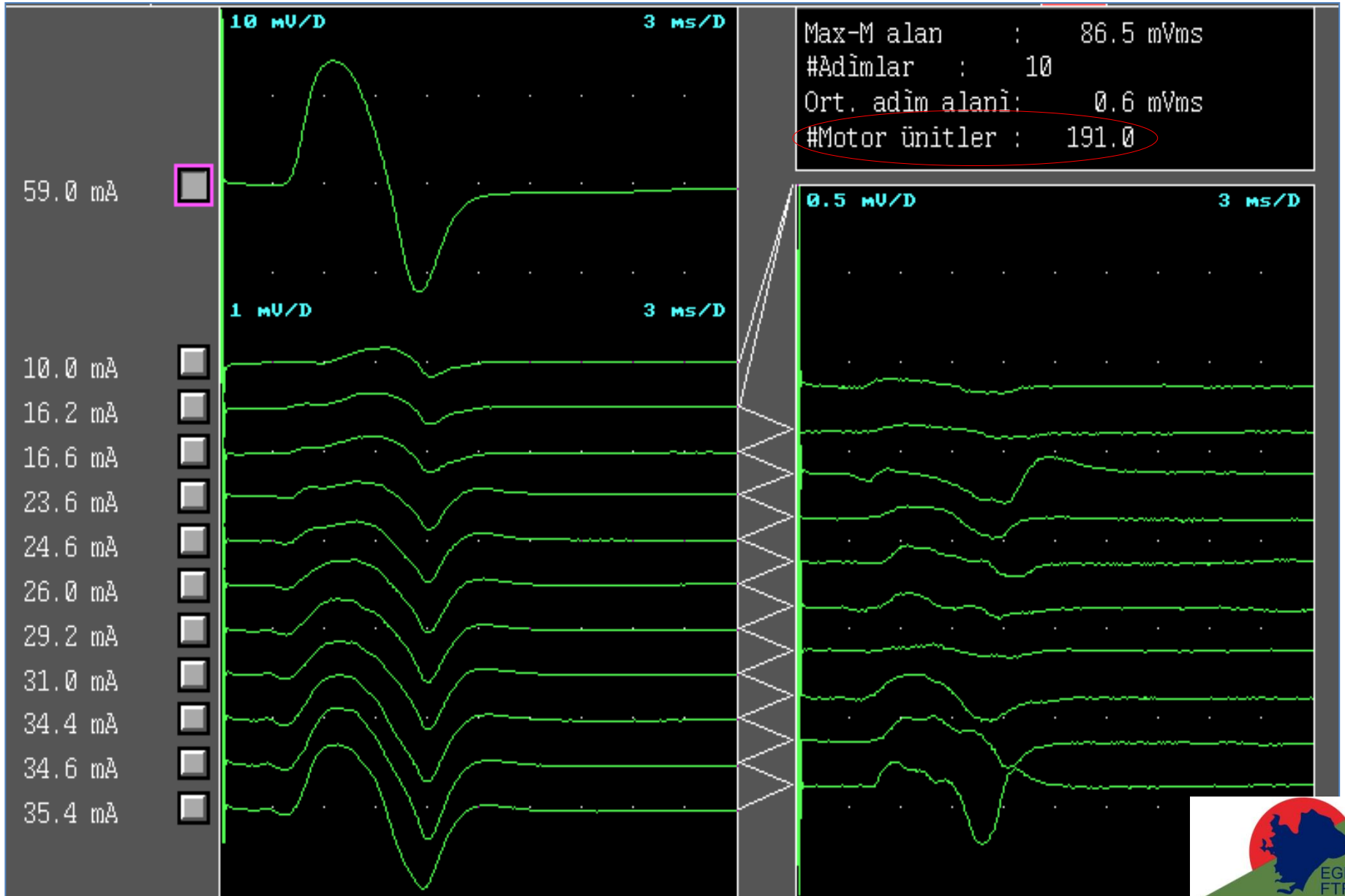
Macroelectromyography (macro-EMG)

Signal from the **entire** motor unit



Stålberg

Motor Unit Number Estimation (MUNE)

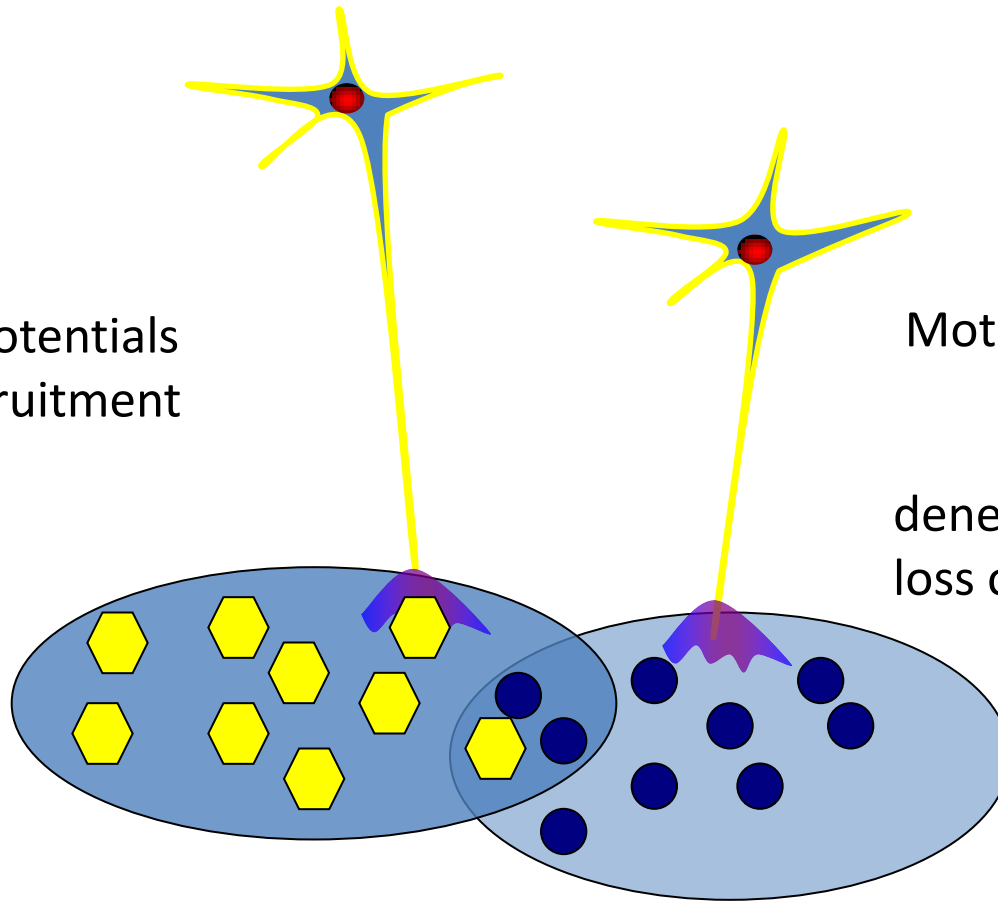


Electrophysiological findings in the muscles affected by poliomyelitis

acute poliomyelitis

EMG findings

Denervation potentials
Decreased recruitment



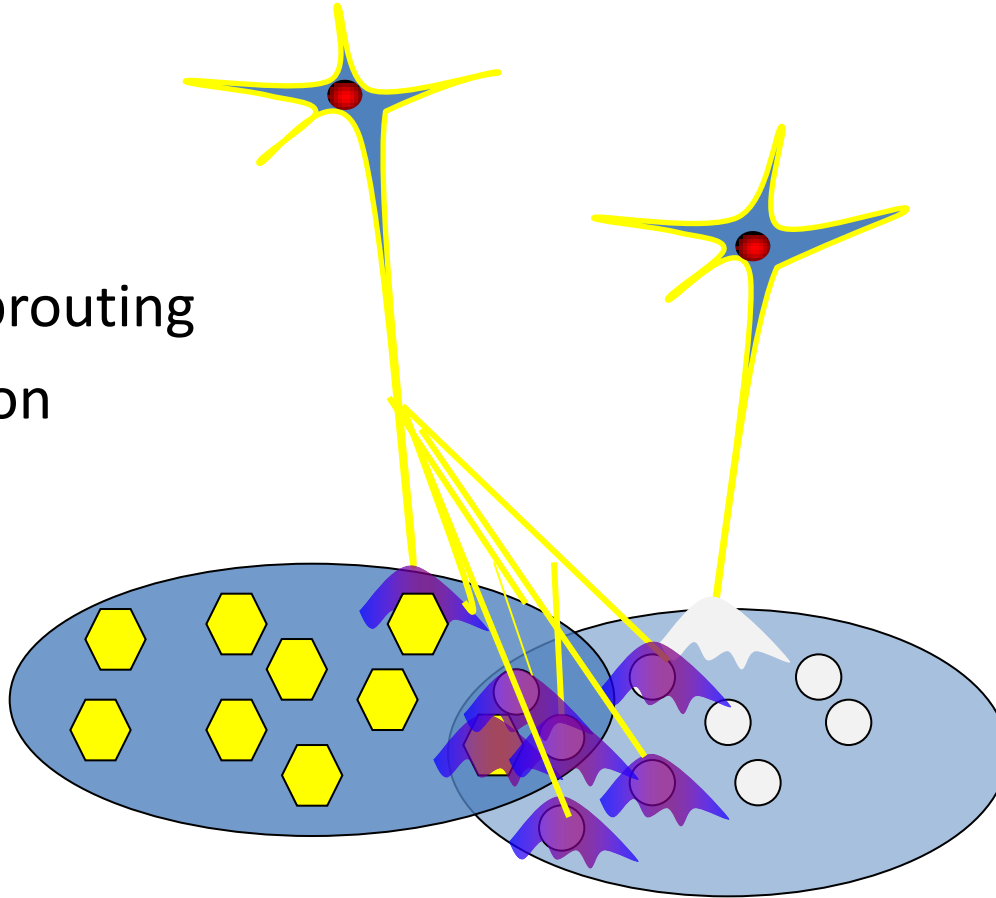
Motor neuron death



denervation
loss of voluntary contraction

recovery process

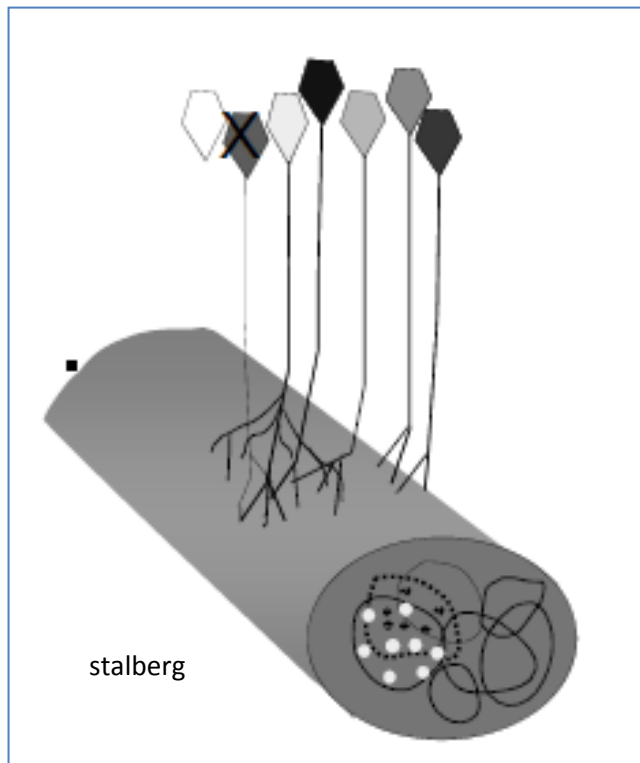
Collateral sprouting
Reinnervation



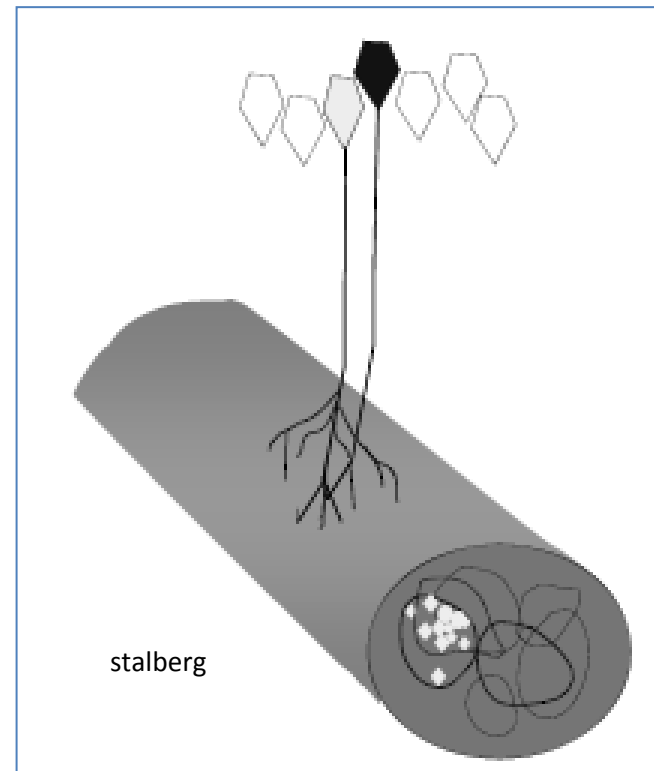
recovery process

Number of surviving MNs

Ability to form sprouts



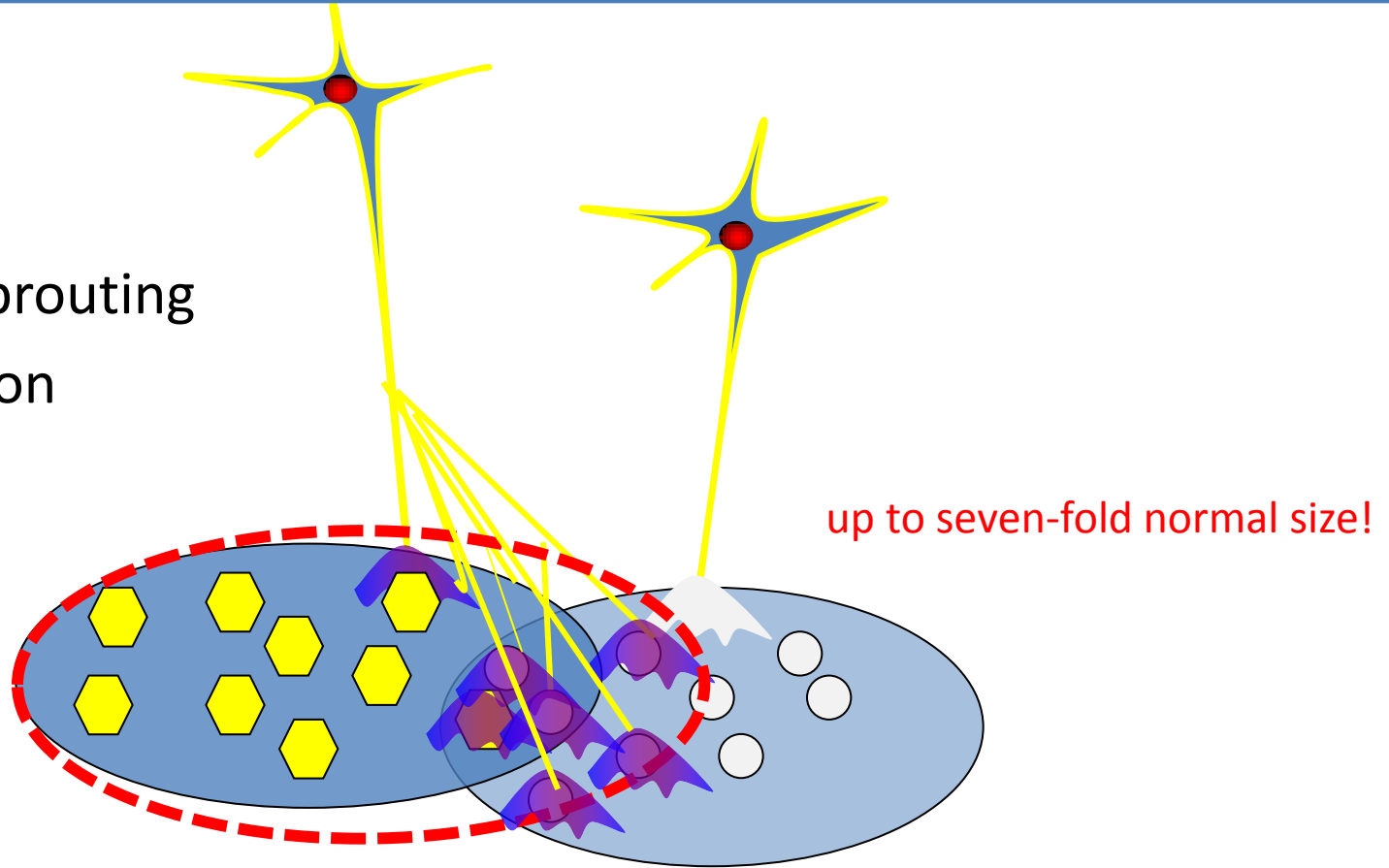
Loss of up to 50% of motor neurons
Complete recovery in muscle strength



weakness
progressive atrophy

After recovery process

Collateral sprouting
Reinnervation



Loss of motor units

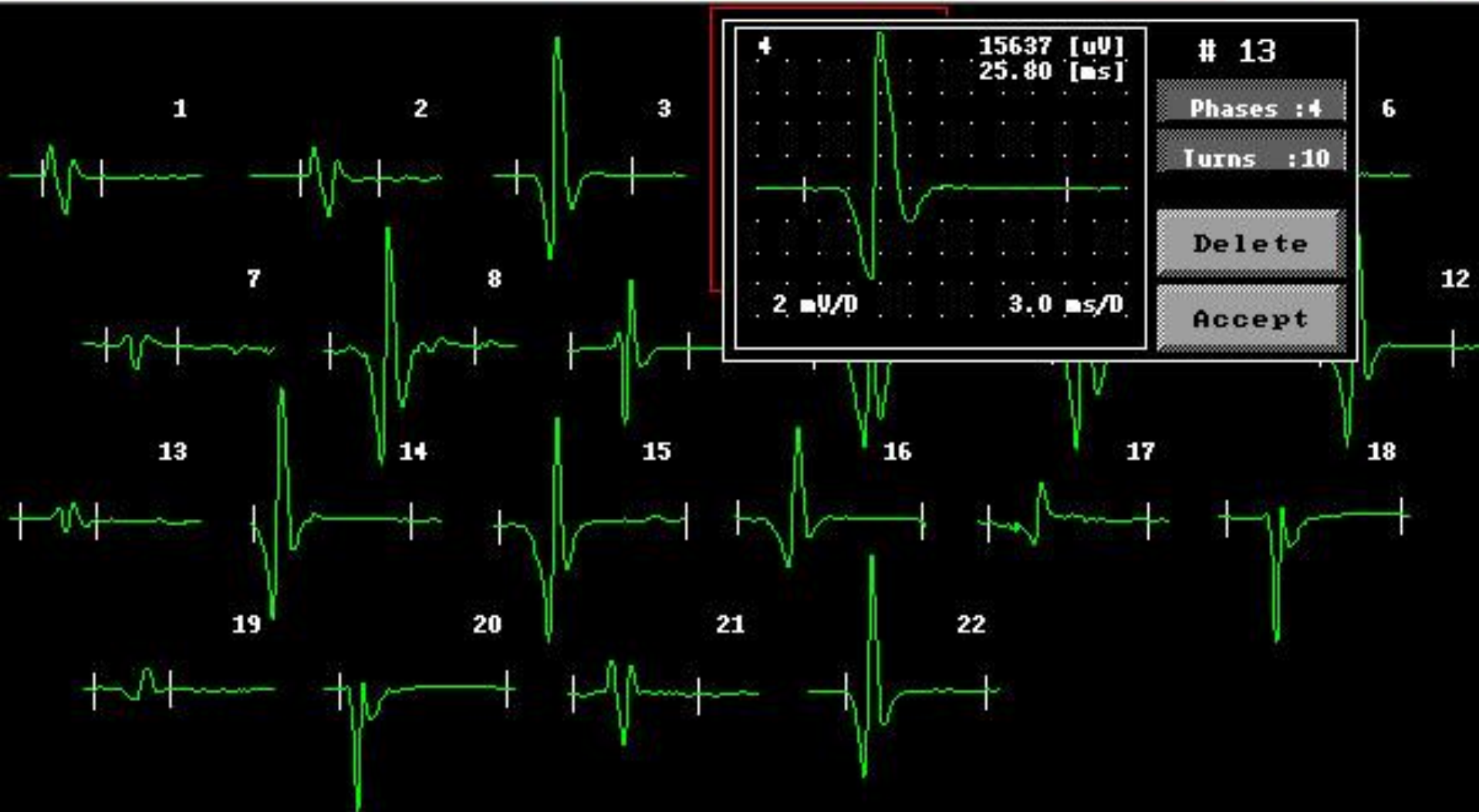
Decreased MUNE
Decreased recruitment
Denervation potentials

Increased motor unit size

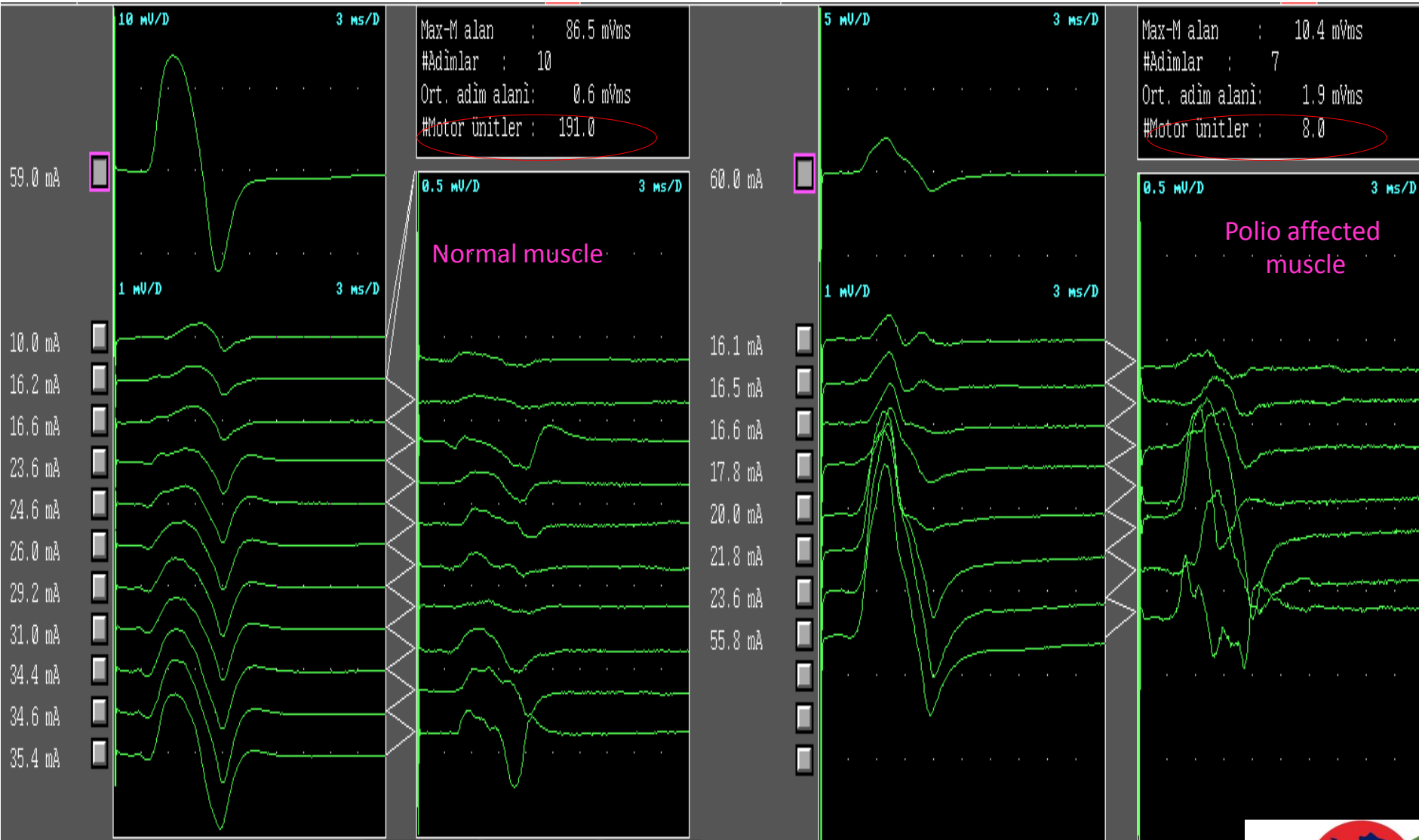
Increased amplitude and duration of the MUPs
Increased macro MUP amplitude

long after recovery

- Continue to have electrophysiological abnormalities....
- **always leaves clear abnormalities on ED studies.**
 - Persistence of denervation potentials
 - Decreased recruitment
 - Enlarged MUPs in needle EMG and macro EMG
 - Decreased MUNE

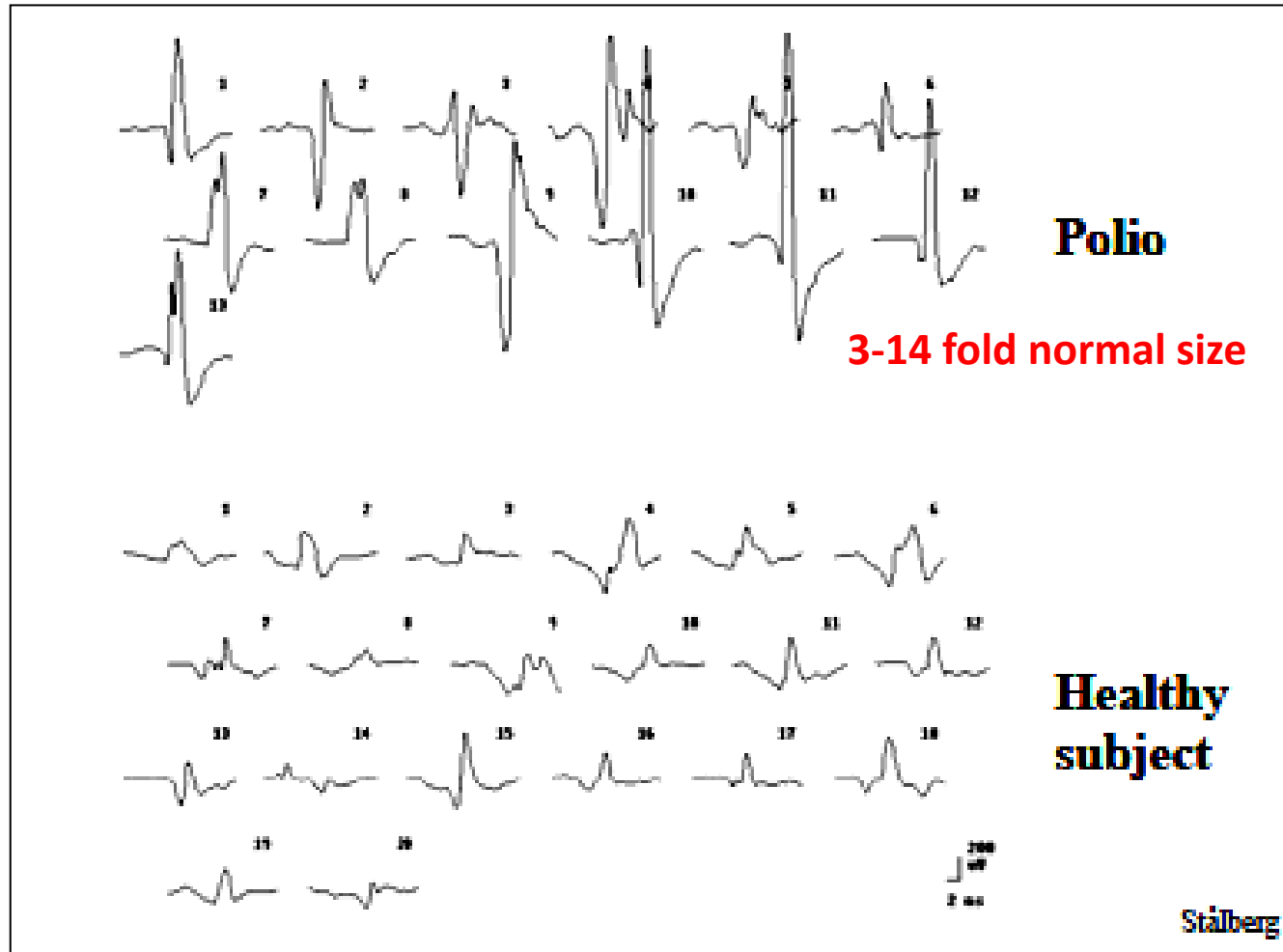


Motor Unit Number Estimation(MUNE)



Reduced MUNE in the 87% of the affected muscles (McComas 1997)

Macro-MUP amplitude



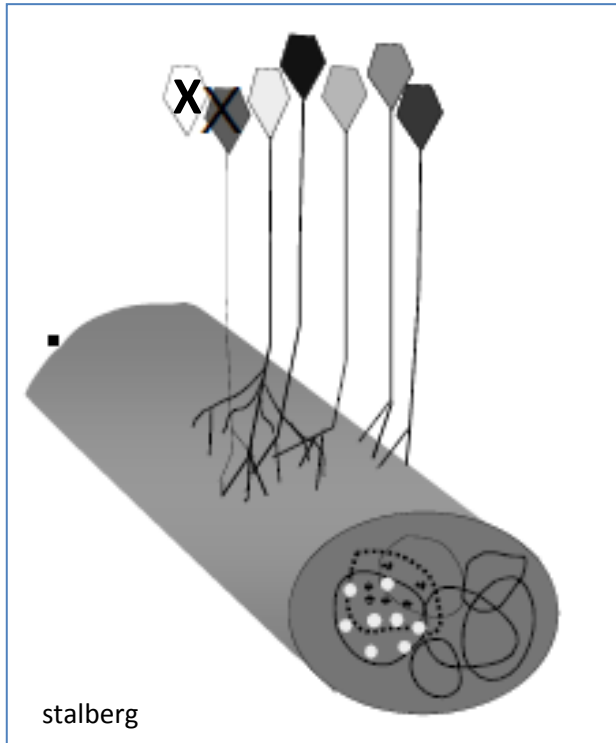
Why???

1. for confirmation of past poliomyelitis involving motor neurons

if neurophysiological findings are normal,
the **original diagnosis of polio must be revised.**

Why??

2. To detect motor neuron involvement in the muscles considered **unaffected** (subclinical polio)



Loss of up to 50% of motor neurons:
complete recovery
normal muscle strength
abnormalities in ED studies

NEW MUSCLE WEAKNESS IN THE SUBCLINICALLY INVOLVED LIMBS IN PATIENTS WITH POLIOMYELITIS



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Ege University Medical Faculty, Department of Physical and Rehabilitation Medicine, İzmir, Turkey

Methods and material:

- We investigated systematically 448 limbs of 112 patients (72 women, 40 men, age between 22 to 65 years) who had admitted to our post-polio clinic.
- Detailed clinical histories, including the affection of the limbs by polio and presence of new muscle weakness were recorded.
- Muscle strengths were measured in all limbs of the patients.
- Needle EMG investigations were performed in selected proximal and distal muscles in all of the limbs.
- An un-affected limb** was defined as one that the survivor had identified as not weakened or paralyzed during the acute phase of polio
- A subclinically involved limb** was defined as one that polio survivors had regarded as unaffected but needle EMG had revealed motor neuron involvement.

All limbs (n=448)		
Limbs considered unaffected by the survivor(n)	Limbs identified as affected by needle EMG (n)	rate of subclinical polio
311	112	36%

limbs of the survivors with post-polio syndrome (n=296)		
Limbs considered unaffected by the survivor(n)	Limbs identified as affected by needle EMG (n)	rate of subclinical polio
184	86	47%

Widespread involvement is a risk factor for PPS???

new muscle weakness in 49 out of 112 limbs with subclinical polio (23%)

the muscles affected by polio virus is vulnerable for later functional impairment, even if they achieved complete recovery after the acute stage of poliomyelitis.



Why???

- for exclusion of other conditions that may explain the new symptoms of post polio syndrome (peripheral neuropathy, radiculopathy, myelopathy)
- to find concomitant nerve or muscle disorders (entrapment, radiculopathy)



Why not??

can ED studies detect the presence of post-polio syndrome??

NO

**symptomatic and asymptomatic muscles have the same findings
no specific ED test for the syndrome is currently available**



Polio survivor presented with new symptoms

If clinical and EMG investigations indicate polio-induced lower motor neurone involvement,
the patient is said to have developed post polio syndrome.

the lack of clear evidence for previous denervation after extensive electrodiagnostic testing is a valid means for
excluding the diagnosis of postpolio syndrome

- (Bromberg and Waring. Arch Phys Med Rehabil 1991)



Clinical utility of ED studies in polio survivors

a patient
with a history of polio
with obvious atrophic paralysis

EMG
unnecessary

To confirm past
poliomyelitis

EMG
helpful

To detect the
presence of
subclinically involved
muscles

To identify or rule out
other conditions

Should be performed at least once

a patient
with a vague history of polio
misdiagnosed with polio

EMG
helpful

To rule out poliomyelitis

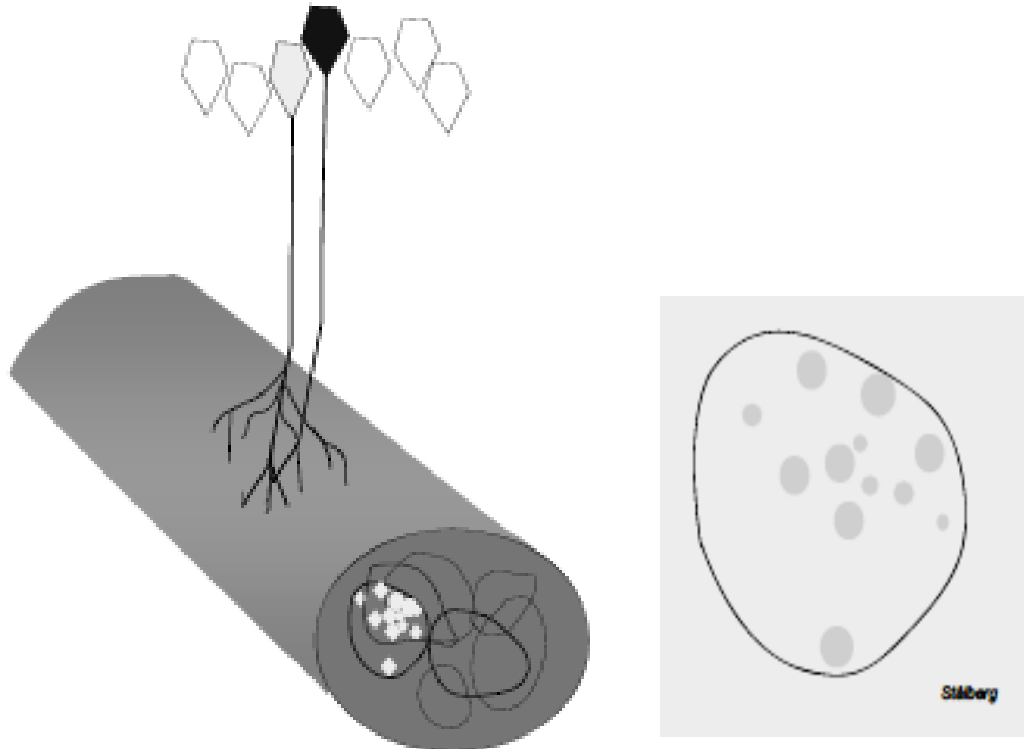
To identify the presence of
another disorder



Research purposes

to provide information on the possible etiology of some PPS symptoms.

**New weakness appears
when the compensatory mechanisms are no longer sufficient”**



Grimby G , Stalberg E , Sandberg A , Sunnerhagen KS . An 8 - year longitudinal study of muscle strength, muscle fiber size, and dynamic electromyogram in individuals with late polio . *Muscle Nerve* 1998 ; 21 (11) : 1428 – 37



ELSEVIER

Clinical Neurophysiology 113 (2002) 478–484



www.elsevier.com/locate/clinph

Motor evoked responses from the thigh muscles to the stimulation of the upper limb nerves in patients with late poliomyelitis

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^aDepartment of Clinical Neurophysiology, Ege University Medical School, 35100 Bornova, İzmir, Turkey

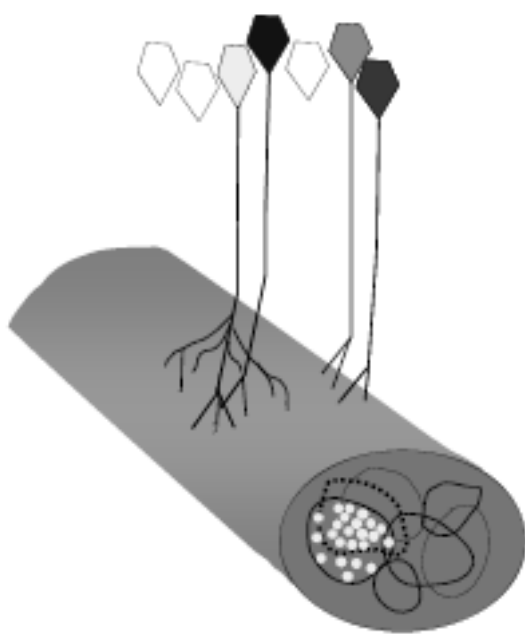
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the proximal muscles of the lower limbs (Fig. 5). Such an approach concerning the double source of continuous excitation on the motoneurons (descending and segmental) may also contribute to the weakness and fatigue encountered in post-polio syndrome.

Future research

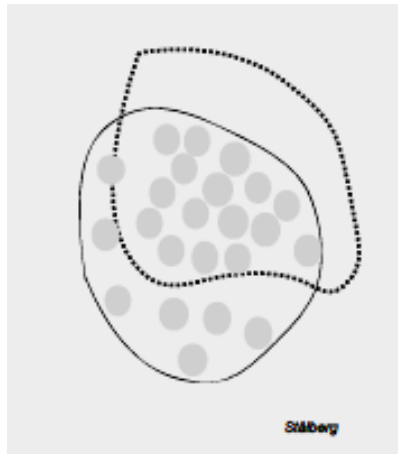
- Relations with the symptoms of PPS.
- Diagnostic value of macro EMG
- Changes in motor units over time or in response to treatment.

Thank you very much

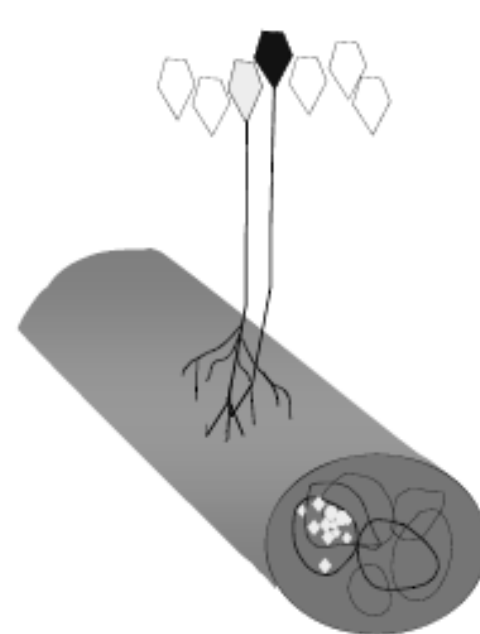


Great metabolic demand

Distal degeneration
reinnervation of
enlarged MUs



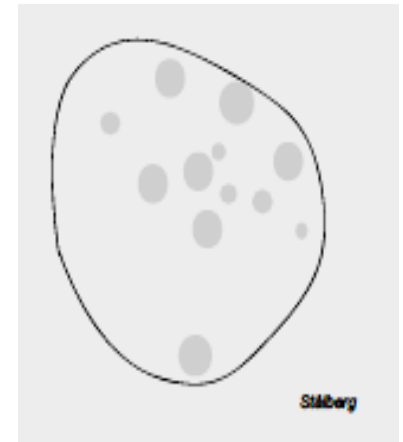
Continuous denervation – reinnervation
Continuous increase in macro-MUP amp



Upper capacity reached
(macro MUP exceeds 20
times the normal size)
permanent denervation



new weakness
reduction in size of MU



Persistence of fibrillation potentials long after polio attack

- Disappear within 6 to 12 months
 - maturation of the new terminal sprouts.
- Never disappear
 - Ineffective reinnervation of the enlarged motor units
 - Ongoing denervation / reinnervation of the enlarged motor units
 - Metabolic abnormality of the motor neurons affected by the polio virus

Normal electromyographic findings in patients with a history of polio

1. Vague history of polio
2. Non-paralytic polio

Normal electromyographic findings in patients with a history of **paralytic** polio

1. Incomplete neurophysiological investigation
2. Insensitivity to detect MUP changes after degeneration of a small number of neurons
3. Transient functional loss without degeneration of anterior horn cells
4. Additional causes for muscle weakness (central involvement, pain inhibition)

SFEMG can provide information about neuromuscular transmission with determination of jitter and blocking, and about motor unit reorganization with fiber density.

During the reinnervation process after acute polio, axonal sprouts are initially unmyelinated and conduction is slow and variable. The newly formed neuromuscular junctions may also be a site of transmission slowing or failure. These abnormalities will present as increased jitter and blocking on SFEMG ([Fig 1A](#)). In addition, fiber type grouping as a result of motor unit remodeling during reinnervation will result in increased fiber density on SFEMG in the involved muscle. Increased jitter and blocking persist in the polio survivor.[\[29,36\]](#) Thus, abnormalities observed on SFEMG, as on EMG, are not helpful in distinguishing symptomatic from asymptomatic individuals.[\[29\]](#) However, a significant correlation between the percentage of fibers exhibiting jitter and fiber density has been found, suggesting that muscles with the most enlarged motor units as a result of sprouting are more likely to exhibit instability later in life.

Evidence for neuromuscular junction deficits are frequently observed in polio survivors, and may be a cause of muscular fatigue. Earlier studies demonstrated a reduction of compound motor action potentials on supramaximal repetitive stimulation,[\[55\]](#) and **numerous recent studies demonstrate increased jitter and blocking on SFEMG.**[\[28,29,35-37\]](#) There appears to be a relationship between the degree of neuromuscular junction deficits and the degree of motor unit enlargement after polio. Jitter has been shown to be increased in patients with a higher fiber density (indicating greater motor unit reorganization),[\[29\]](#) increased macro-MUP amplitudes (suggesting larger motor units), and fiber-type grouping on muscle biopsy.[\[66\]](#) Therefore, the extent of initial recovery may predict the degree of electrophysiological deficits later in life.