# Neurophysiological tests in MND

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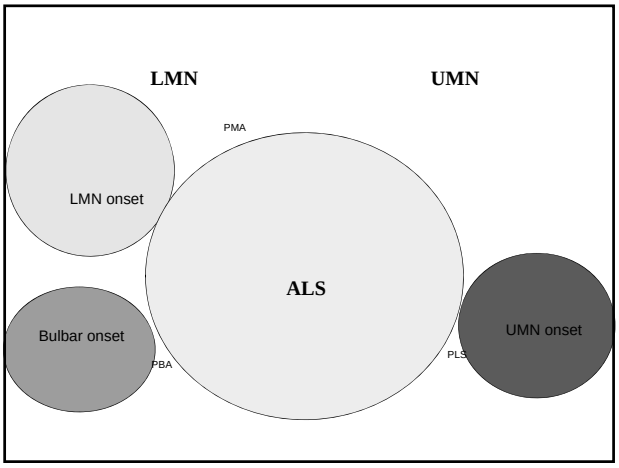
## Clinical types of ALS

- **Sporadic ALS**
- **Genetically determined ALS**
  - **SOD-1 mutations**
- **Two clinical forms**
  - **Spinal ALS**
  - **Bulbar ALS**






## Clinical hallmarks of ALS

- Painless weakness and atrophy in several regions
- Often focal onset
- Increased reflexes
- Progressive history
- Imaging and other tests exclude other diagnoses

## EMG findings

## Neurogenic EMG

(has the same significance as clinical LMN signs)

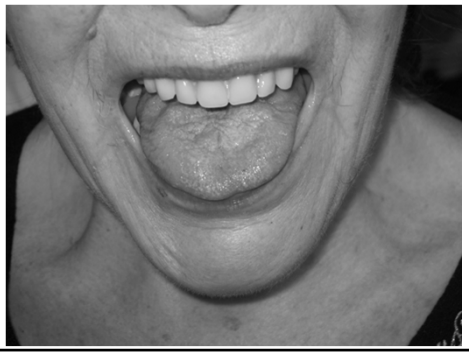
*EMG features

- MUP: Increased ampl, dur, phases
- MUP complex, with jiggle
- Decreased recruitment=rapid firing in red # of MU. (UMN may give low firing)

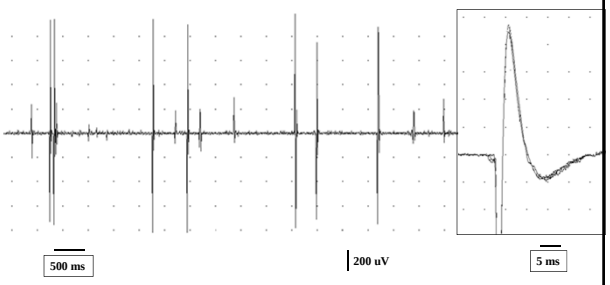
*Fibs/-psw usually in strong non-wasted muscles

*In neurogenic EMG, fasc, preferably complex have the same significance as fib-psw

## Generalized tounge atrophy in ALS



## Fasciculation potentials

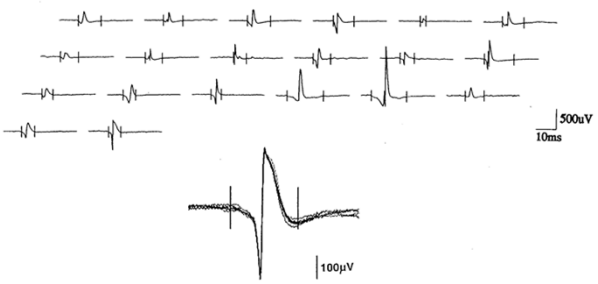



## Fasciculations

- FP in ALS
  Complex, jiggle (instability)
- Benign
  - Simple, stable
- Benign FP normal
- Complex FP
  - can alone not make ALS diagnosis
  - not always present in ALS
  - can be seen in other neurogenic disorders

Awaji, 2008

## MUPs in a normal subject

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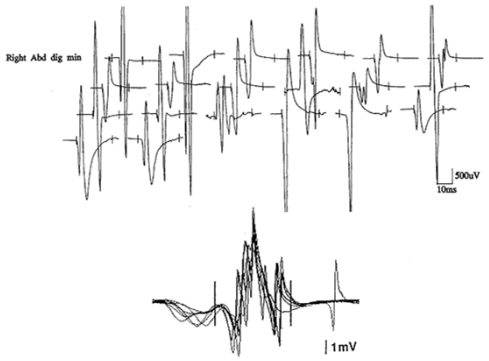
## MUPs in a patient with ALS

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## Peripheral nerve in ALS

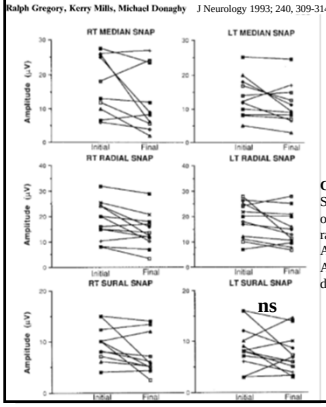
## Neurography, to exclude other disorders

- ”Normal SNS”
- MCS, CV >75% of normal, F <130% of normal
- Distal latency and dur < 150% of normal
- Absence of CB and dispersion
- CMAP ampl often low
- If CMAP ampl < 2 mV, CV value low

## “Sensory normal”

**Progressive sensory nerve dysfunction in amyotrophic lateral sclerosis: a prospective clinical and neurophysiological study**

Ralph Gregory, Kerry Mills, Michael Donaghy J Neurology 1993; 240, 309-314)




**Conclusion**
Subclinical sensory deterioration (6-17 months) but only occasionally outside normal range.
At one test, normal sensory function is the rule.
ALS may be a generalized neurodegenerative disorder, not restricted to motor neurons.

## Upper motor neurone testing

## EDX signs of UMN

- Low irregular firing rate of MUPs
- Increased F-persistence (Stålberg unpubl)
- TMS
  - Decreased / Increased threshold
  - Increased CMCT (30%)
  - Increased absolute latency
  - Absent limb responses in pat with bulbar symptoms supports UMN
- Triple stim technique sensitive, needs confirmation

## ALS criteria

- El Escorial WFN criteria (Brooks), 1994
- Revised El Escorial criteria, Airlie House 2000
- Awaji Island criteria, 2008

**http://www.wfnals.org**

Modified criteria, Awaji Island, Japan, 2006, publ 2008

Electrodiagnostic criteria for diagnosis of ALS
J Clin Neurophysiol, 119, 2008

De Carvalho, Dengler, Eisen, England, Kaji, Kimura, Mills, Mitsumoto, Nodera, Shefner, Swash

*Awaji 2008*

## Some news compared to El Escorial

- EDX equally important as clinical signs
- Fasciculation potentials (FP) indicate ongoing denervation, equally important as fibs/psw
- FP are complex and unstable
- FP in ALS usually start distally
- Jiggle of MUPs is useful information

*Awaji 2008*

Diagnostic categories,

## Clinically definite ALS

Clinial or EDX evidence of

LMN + UMN in bulbar and at least 2 spinal regions

or

LMN + UMN in 3 spinal regions

*Awaji 2008*

Diagnostic categories

## Clinical probable ALS

Clinical or EDX evidence of

LMN + UMN in at least two regions with some UMN rostral to the LMN signs

*Awaji 2008*

Diagnostic categories

## Clinically possible ALS

Clinical or EDX signs of

UMN + LMN dysfunction in only one region

or

UMN in two or more regions

or

LMN rostral to UMN signs

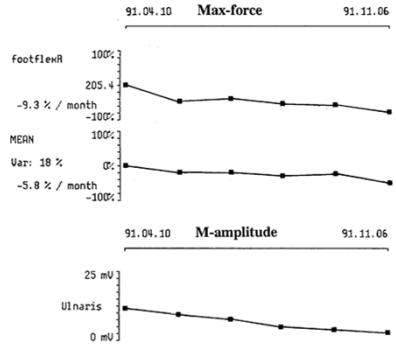
and

Other diagnoses excluded (imaging + lab tests)

*Awaji 2008*

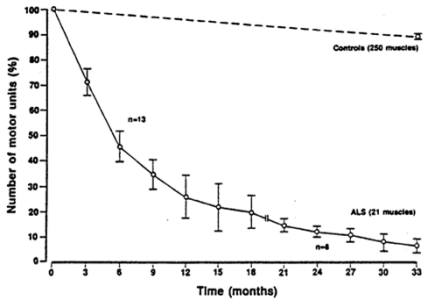
## Monitoring changes over time

Change in force and CMAP-amplitude in a patient with ALS



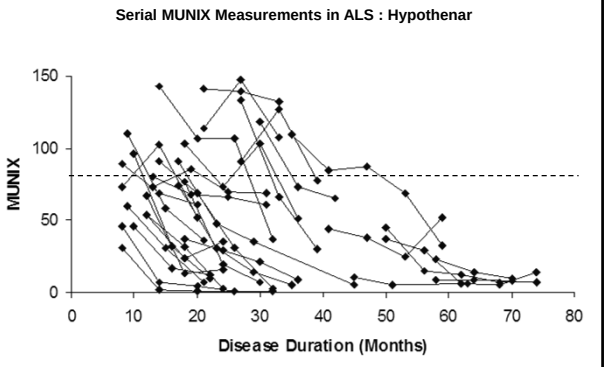

Motor unit number estimation (MUNE)

Rate of loss of MU-function in patients with ALS




EDB, thenar and hypothenar m

Dantes, McComas -91




Barkhaus,Nandedkar

EDX strategies

## Principle strategy of EDX in ALS

- **Confirm LMN dysfunction in clinically affected regions**
- **Detect electrophysiological evidence of LMN dysfunction in clinically uninvolved regions**
- **Exclude other pathophysiological processes**

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## Practical strategy of EMG in ALS

- Chose some weak/atrophic muscle and some clinically normal
- Muscles should repr different nerves and segments
- Assess fib-psw, fasc pot (frequency, shape), MUP parameters incl jiggle, IP

## EMG in ALS, suggested muscles

- Spinal:
  - IOD
  - Biceps
  - Paraspinal Th10
  - Rect abd
  - Tib ant
  - Vast lat
- Upper cervical and bulbar
  - Trapezius
  - Sternocleid
  - Masseter
  - Genioglossus

## Practical strategy of Neurography in ALS

- ***Neurography MCS (bilaterally)***
  - n.medianus
  - n.ulnaris (also including supraclavicular stimulation)
  - n.peroneus
  - n.tibialis
- ***Neurography SCS (bilaterally)***
  - n.suralis
  - n.radialis
- ***MEP***
  - upper and lower extremity

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## Multifocal motor neuropathy with conduction block (MMN)

## MMN - Clinical features1

- Slowly progressive weakness distributed over individual peripheral nerves rather than myotomes (in ALS the distribution follows spinal myotomes)
- Progression usually slow over years
- Weakness is often distal, rarely proximal

## MMN - Clinical features 2

- Muscle atrophy of weak muscles is less pronounced than would be expected (weakness is partly due to conduction block)
- Fasciculations, cramps and myokymia
- Although MMN is predominantly a motor neuropathy, there may be mild sensory symptoms and findings

## MMN - Clinical features 3

- *No signs of upper motor neuron lesion*
- *Rarely involvement of cranial nerves*
- Diaphragm is rarely affected
- Clinically MMN and ALS present usually differently
- Sometimes be difficult to distinguish MMN from ALS clinically

## MMN - Etiology

- Unknown, possibly an autoimmune reaction against gangliosides ($GM_1$)

MMN - Expected abnormal EMG findings

- Subacute or chronic neurogenic EMG findings in muscles innervated by different nerves
- Weakness and EMG findings are distributed according to peripheral nerves rather than myotomes

MMN - Expected abnormal neurography

- Motor nerves show conduction blocks (amplitude and area decay, reduced number of F-waves)
- Often reduced M wave amplitudes
- Motor conduction velocity may be reduced

## MMN - Expected normal findings

- Sensory nerve conduction studies
- Central motor conduction time normal

## Post-polio conditions

- Post-polio syndrome -PPS
- Post-polio muscular atrophy - PPMA

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## Post-polio muscle dysfunction
### clinical criteria

- history of paralytic polio
- functional stability for 15 years
- new (and/or)
  - weakness
  - atrophy
  - pain
  - fatigue
- neurological exam = lower motor neurone
- no other disorders to explain symptoms

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## Post-polio syndrome - etiology for deterioration in n-m function

- disuse
- overuse
- weight gain
- chronic weakness

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## Post-polio syndrome - causes for deterioration in n-m function

| | |
|---|---|
| • reactivated virus | - |
| • loss of motor units | + |
| • excessive metabolic demand | + |
| • muscle fibre defect | + |

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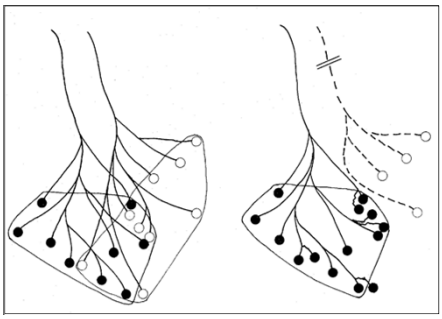
## Schematic fig of reinnervation



2 normal motor units

grouping but no extension outside borders

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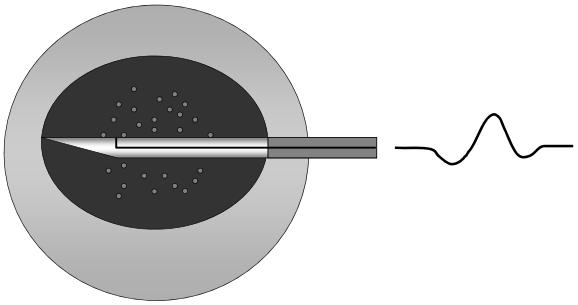
## Macro EMG signal from the entire motor unit



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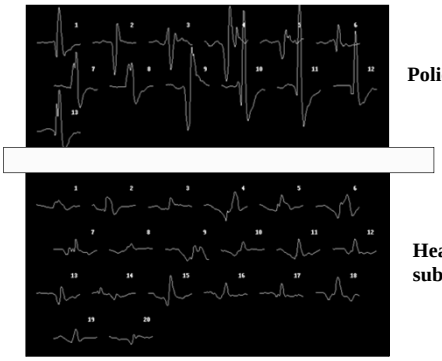
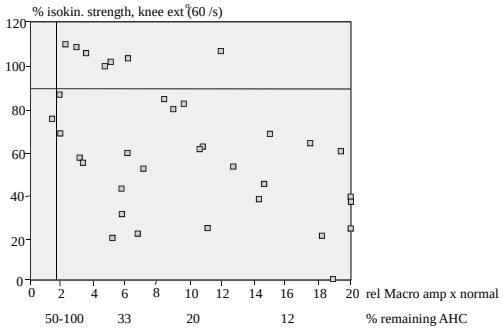
## Macro MUPs in Tibial anterior muscle



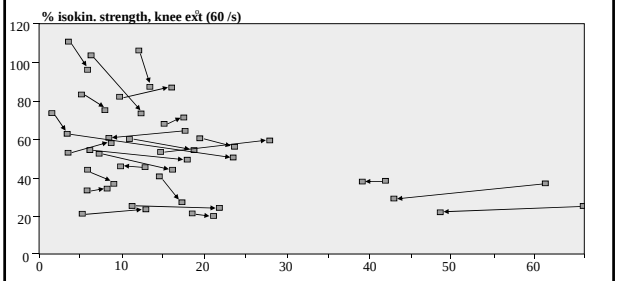
**Polio**

**Healthy subject**

## Macro MUPs in patients with acute polio >30 years ago

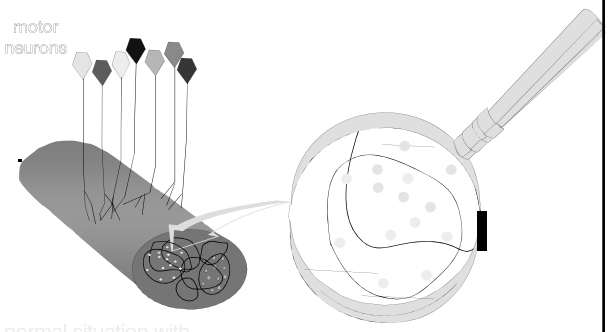


## Change in Macro EMG in patients with polio more than 30 years ago 4 year follow up




Stålberg, Grimby

## Normal

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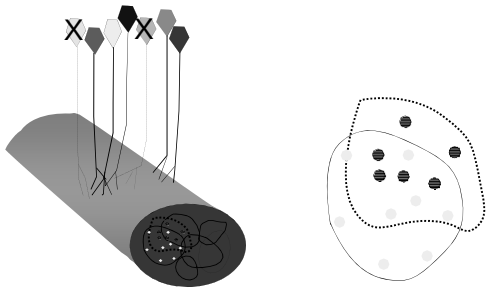
## Polio - after 1 week

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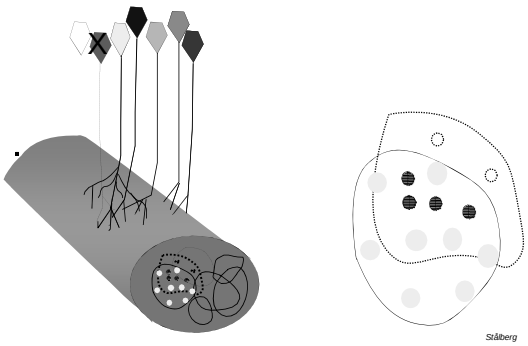
## Polio - after 3 months

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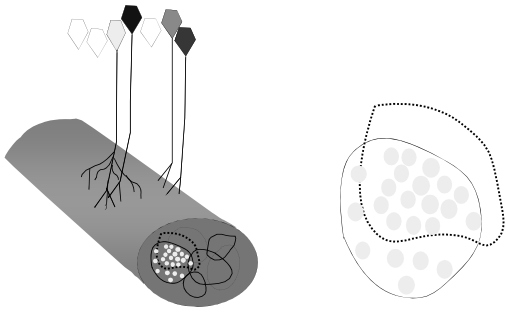
## Polio - after "10 years"



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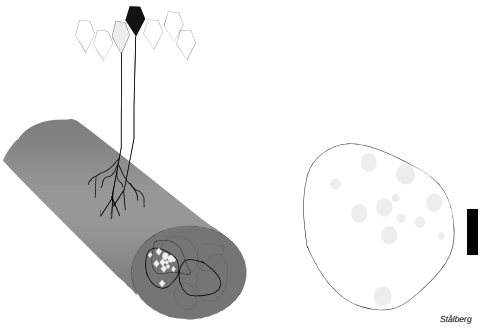
## Polio - after "20 years" - PPS



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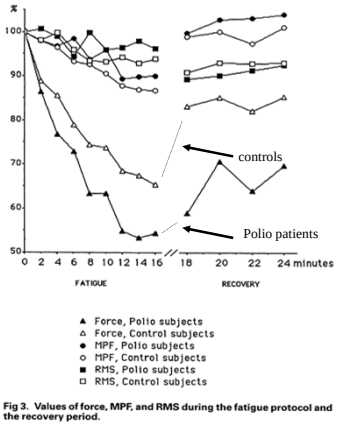


**Fig 3. Values of force, MPF, and RMS during the fatigue protocol and the recovery period.**

More fatigue during exercise

and

Poor recovery
in post polio patients
after fatiguing exercise

Stibrant-Sunnerhagen et al.
Arch Phys Med Rehab
81:2000

# Conclusion

- Nearly all post polio subjects have EMG changes
- If EMG is normal, reconsider the diagnosis
- A ”normal EMG” does not completely exclude a previous polio

Sandberg,Stålberg