Motor neuro- and neuronopathies

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Motor neuronopathies

- Amyotrophic lateral sclerosis (ALS)
- Progressive spinal muscular atrophy
- Spinal muscular atrophy
  - SMA1 (Werdnig-Hoffmann)
  - SMA2 (intermediate)
  - SMA3 (Kugelberg-Welander)
- Distal spinal muscular atrophy (heterogeneous)
- X linked spinal and bulbar muscular atrophy (Kennedy sdr)
- Polio (rarely other enteroviruses)
- Acute squeale following previous polio
- Monomelic spinal muscular atrophy
- Benign fasciculation

History of nomenclature

- Duchenne 1849 and Aran 1850
  - Progressive spinal muscular atrophy
- Charcot 1869
  - Amyotrophic lateral sclerosis
- Spiller 1904
  - Primary lateral sclerosis
- Brain 1962
  - Motor neuron disease (UK = ALS, PLS and PMA)
- Rowland 1982
  - Motor neuron diseases (USA = all motor neuronopathies)

Nomenclature

- Motor neuron disease = ALS
- Motor neuron diseases = all motor neuronopathies
- Progressive bulbar palsy (PBP)
- Progressive muscular atrophy (PMA)
- Primary lateral sclerosis (PLS)
- Hereditary spastic paresis (HSP)
- Spinal muscular atrophy (SMA)

Modified El Escorial criteria for ALS

- Lower motor neuron signs
  - Clinical, neurophysiological or pathological
- Upper motor neuron signs
  - Clinical
  - Neurophysiological
- Progressive spread of signs within a region or to other regions

Primary lateral sclerosis

- Only upper motor neuron signs
- Many of those with PLS onset develop classical ALS within 4 years from onset
- Pure PLS has a much better prognosis than ALS
- Rule out hereditary spastic paraparesis (HSP)
  - Family history
  - Onset usually < 40 years
  - Genetic testing
Progressive muscular atrophy (PMA)
- Only lower motor neuron signs
- Many ALS patients may present with pure lower motor neuron signs
- Within 1 year 20% develop upper motor signs
- At autopsy 50% of PMA patients have upper motor lesions
- Prognosis is similar to ALS, survival 12 months longer
- PMA is probably a form of ALS

Survival in PMA and ALS

Progressive bulbar atrophy
- Only bulbar symptoms
  - Dysarthria
  - Rarely dysphagia
- Upper and lower motor neuron involvement of cranial nerve innervated muscles (pseudobulbar palsy)
- Emotional lability
- Ocular muscle spared in the early stages

Hereditary spastic paraparesis
- Spasticity, mainly legs
- Mild weakness in distal part of legs
- Mild sensory symptoms
- Mode of inheritance
  - Autosomal dominant
  - Autosomal recessive
  - X-linked
- Onset 10-30 years or < 6 years
- > 45 genes
- HSP4 most common - spastin gene

ALS

Spectrum of motor neuron disorders

Primary lateral sclerosis | ALS | Progressive muscular atrophy
---|---|---
Sporadic | Hereditary | Lower motor neuron
Upper motor neuron | Hereditary spastic paraplegia | Spinal muscular atrophy
ALS aetiology

- Syndrome with heterogeneous aetiology
- Genetic
- Smoking increases risk x 3
- Diet, high fat, glutamate
- Autoimmune
- Oxidative stress
- Glutaminergic excitotoxicity
- Role of astrocytes
- Most cases are sporadic
- 5-10% hereditary, dominant or recessive
- Cu/Zn superoxide dismutase (SOD1) mutation on chromosome 21q22 (AD and AR)
- Chromosome 2q33-q35

ALS - Epidemiology

- Incidence 2-3/100,000/year
- Prevalence 4-6/100,000
- Usually onset 50-75 years
- Males:females below the age of 70
  - 1.3-1.6:1

Recent developments in epidemiology

Amyotrophic Lateral Sclerosis in Sweden, 1991-2005

Incidence of ALS in Sweden

ALS - Clinical features 1

- Painless, asymmetric weakness
- Upper limb muscles affected more frequently than lower limb muscles
- Bulbar muscles may be involved
- Dysarthria
ALS - Clinical features 2

- Rarely clinically affected muscles
- Extraocular muscles
- Anal or urethral sphincters
- Upper motor neuron signs
  - Increased tendon reflexes
  - Babinski sign
- Mild sensory symptoms may be present
  - Paresthesias
  - Tibial nerve SEP abnormal in 30%

Spreading of ALS

ALS motor phenotype heterogeneity, focality, and spread

Deconstructing motor neuron degeneration

Abstract

Heterogeneity of motor phenotypes is a clinically well-recognized fundamental aspect of amyotrophic lateral sclerosis (ALS) in determining variability of independent primary motor neuron body regions of arrest, relative rate of upper motor neuron (UMN) and lower motor neuron (LMN) deficits, and rate of progression. Motor phenotypes are determined by the anatomy of the underlying neuropathology and the various deficits reflected by these heterogeneities, are that motor neuron degeneration is a focal process and that it spreads progressively through the 3-dimensional anatomy of the UMN and LMN tissue, thus causing overlapping symptoms in different body regions. The clinical presentation, therefore, may be used to assess the underlying pathology for disease localization and to predict spread, disease progression, and survival. The objective of this study is to assess these clinical presentations and their relationship to pathology.

ALS and frontotemporal dementia (FTD)

- 5-15% have FTD
- Share similar inclusions
  - Immunopositive for TAR DNA binding protein TDP-43

Survival

- Average survival after diagnosis
  - 3 years in spinal form
  - 20% survive 5 years
  - 10% survive >10 years
  - If presenting with bulbar symptoms shorter

- Poor prognosis is related with
  - Onset > 60 years
  - Female > male
  - Bulbar onset

ALS and other neurodegenerative disorders

ALS + frontotemporal dementia (FTD)

5-15% have FTD

Share similar inclusions

Immunopositive for TAR DNA binding protein TDP-43

ALS and frontotemporal dementia (FTD)

Valdettaro and Rossi: Genetics of familial ALS. Neurology 2008
Familial ALS
- AD SOD1 mutation
  - 90 different mutations described
  - Does not differ from sporadic ALS
- AR SOD1 mutation - A90D
  - Tornio river valley in Sweden and Finland
  - 2.5% of population heterozygous for gene
  - LM > UM
  - Cramps, myalgias
  - Bladder dysfunction
  - Slow progression, survival 10-15 years

The ALSIN gene
- ALS2
  - Juvenile ALS starting in the second decade
  - Progression over 10-15 years
  - Recessive and dominant
  - Chromosome 2q33
  - Alsin also linked to HSP (hereditary spastic paraparesis) and PLS (primary lateral sclerosis)

The sentaxin gene
- ALS4
- Juvenile onset
- Chromosome 9

Twin studies
- In monozygotic twins 20% concordant for ALS
- In dizygotic twins very low concordance

Mutated single genes
- SOD1, chromosome 21q22.11
  - Dominant toxic gain of function most common
  - 110 different mutations described
  - 20% of familial ALS
  - Loss of normal SOD1 function is not the cause
  - Gene carriers have 80% probability of ALS
  - Recessive D90A mutation common in the northern part of Sweden and Finland
- VAPB (vesicle associated membrane protein)
  - Synaptobrevin

Definite familial ALS - laboratory supported
- Presence of defined pathogenic mutation
- Progressive UMN and/or LMN signs in a single anatomical region
- Absence of other causes
Electrodiagnostic adaptation

EMG features of chronic neurogenic change
- MUP amplitude and duration increased
- Decreased MU recruitment
- Unstable MUPS
- Fibrillation potentials
- Fasciculation potentials have equal significance as fibs
- Clinical and electrodiagnostic findings have equal significance

ALS requirements for diagnosis
- Presence of:
  - Evidence of lower motor neuron (LMN) degeneration by clinical, electrophysiological or neuropathological examination
  - Evidence of upper motor neuron (UMN) by clinical examination
  - Progressive spread of symptoms and signs within a region or to other regions, as determined by history or clinical examination

ALS requirements for diagnosis
- Absence of:
  - Electrophysiological and pathological evidence of other disease processes that might explain the LMN or UMN degeneration
  - Neuroimaging evidence of other disease that might explain the observed clinical and electrophysiological signs
    - MRI of the cervical spine will often show degenerative changes and spinal stenosis in this age group

The four regions of the body
- Bulbar
- Cervical
- Thoracic
- Lumbosacral

Clinically definite ALS
- UMN and LMN signs in bulbar and at least two spinal regions
- UMN signs in two spinal regions and LMN signs in three spinal regions
Clinically probable ALS
- UMN and LMN signs in at least two regions
- Some UMN signs above the LMN signs

Possible ALS
- UMN and LMN in one region
- UMN alone in two or more regions
- LMN signs rostral to UMN signs
- Absence of other disorders

Difficulty with El Escorial criteria
- Strict criteria developed for treatment trials
- Many ALS patients die before fulfilling criteria
- Usually it takes follow-up of months, sometimes years before the diagnosis is clear
  - 2-3 EMGs may be required

Strategy of EMG in ALS
- Confirm LMN lesion in clinically affected regions
- Detect electrophysiological evidence of LMN dysfunction in clinically uninvolved regions
- Exclude other pathophysiological processes

EMG features of LMN dysfunction
- Active denervation
  - fibrillation potentials
  - positive sharp waves
- Signs of chronic partial denervation
  - MUP duration and amplitude increased, polyphasic MUPs
  - Reduced interference pattern
  - Jiggle

Quantitative EMG techniques
- Motor unit number estimation (MUNE)
- SFEMG
- Macro EMG
- Turns/amplitude analysis
- MUP analysis
MUNE

- Abnormal threshold
- Normal conduction time or mild prolongation
- Cortical silent period is normal
- Abnormal peristimulus histograms

Decrement

- One bulbar muscle with denervation sufficient
- Thoracic region abdominal or paraspinal muscles at or below Th6
- In the cervical and lumbosacral region at least two muscles innervated by different roots and peripheral nerves must show EMG changes

MEP

- Abnormal threshold
- Normal conduction time or mild prolongation
- Cortical silent period is normal
- Abnormal peristimulus histograms

Topography of EMG findings

- One bulbar muscle with denervation sufficient
- Thoracic region abdominal or paraspinal muscles at or below Th6
- In the cervical and lumbosacral region at least two muscles innervated by different roots and peripheral nerves must show EMG changes

Fasciculation potentials in ALS

- Fasciculation potentials (FP) characteristic
- Absence of FPs does not rule out ALS, but raises diagnostic doubts
- FPs may be benign and occur in other neurogenic disorders as well
- Most ALS patients are not aware of the FPs
- Most patients that are referred for EMG because of FPs have benign fasciculation

Neurography in ALS

- Sensory neurography
  - Usually normal
  - Sometimes mild sensory abnormalities with reduced amplitudes
  - Slight reduction of MCV if amplitude is low
Conduction velocity in ALS

**ALS - expected abnormal findings**

- **EMG**
  - Subacute neurogenic muscle findings in at least 3 regions. Preferentially the findings should be asymmetric without any definite proximal or distal predominance.

- **Neurography**
  - MCS often show reduced amplitudes
  - If the AMPL is reduced significantly the conduction velocity may be reduced (loss of fastest conducting axons)
  - Central motor conduction time
    - sometimes abnormal

**ALS - expected normal findings**

- **Neurography**
  - MCS: if AMPL is normal or only moderately reduced, CV should be normal
  - no motor conduction block
  - normal number of F-waves in mildly affected muscles (indicating lack of proximal conduction block)

- **Evoked potentials**
  - VEP
  - BAEP
  - SEP (may be abnormal)

- **ALS - Procedure EMG**

  - **M. Orbicularis oris / m. masseter / m.genioglossus / m. trapezius (one abnormal sufficient)**
    - If dysarthria then chiroothyreoides
  - **M. Interosseus dorsalis I / m. biceps brachii / m. deltoideus .............**
  - Two abnormal, different nerves and myotomes
  - **M. Tibialis anterior / m. gastrocnemius caput mediale / m. vastus lateralis........**
    - Two abnormal, different nerves and roots
  - **M. Rectus abdominis**

**SEPs may be abnormal in ALS**

Abnormal sensory evoked potentials in amyotrophic lateral sclerosis
ALS - Procedure EMG

Cervical
- IOD I
- Delt.Bic
- Contralat
- 1 abnormal

Thoracic
- Rect. Abd
- Gastroc
- Vast.lat.
- 2 abnormal

Lumbosacral
- Tib.ant.
- M.orbic.ori
- Masseter
- Normal

Cranial
- Trapezius
- Geniohyoid
- Cricothy
- Normal

2 abnormal 1 abnormal 2 abnormal 1 abnormal

ALS - Procedure Neurography

- 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

Other diseases suggested if
- Conduction block is found
- MCV <70% and distal latencies >30%
- Decrement >20%
- SEP latencies >20%
- Full interference pattern in weak muscle
- Significant autonomic abnormalities

False positive diagnosis
- Cervical radiculopathies (spinal stenosis)
- Monomelic spinal muscular atrophy
- Multifocal motor neuropathy with conduction blocks
- CIDP
- Benign fasciculations
- Lumbar radiculopathies (spinal stenosis)
- Brachial plexus neuropathy
- Syringomyelia
- Spinal cord tumours
- Spinal cord AV malformations
- Foramen magnum tumour
- Previous polio
- IBM, polymyositis

10% of ALS patients have unnecessary surgery!!

Role of EMG
- Confirm suspected diagnosis
- Identify subclinical motor neuron loss
- Define severity of motor unit loss
- Define extent and distribution of disease
- Rule out other possible causes
- Provide tool for monitoring rate of progression and efficacy of treatment
Take home messages!

- All patients >50 years with muscle weakness without pain or sensory abnormalities have ALS until proven otherwise!!!
- Diagnostic errors are likely to happen
  - When the referral asks for a focal disorder
    - Radiculopathies
    - Traumatic neuropathy
    - Entrapment
  - Some diffuse disorders (IBM, polymyositis)
- A definite diagnosis may require several EMGs and a follow up of 0.3-3 years

Further reading

**Multifocal motor neuropathy with conduction block (MMN)**

**MMN - Clinical features 1**

- Slowly progressive weakness distributed over individual peripheral nerves
- Slow progression
- 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
- 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 different
- MMN is rare

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**ELECTRODIAGNOSTIC STUDIES IN AMYOTROPHIC LATERAL SCLEROSIS AND OTHER MOTOR NEURON DISORDERS**

JAPAN H. SAIDE, MD

Multifocal motor neuropathy with conduction block (MMN)

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MMN - Clinical features 3

- No signs of upper motor neuron lesion
- Rarely involvement of cranial nerves
- Diaphragm is rarely affected
- Clinically MMN and ALS different
- MMN is rare
MMN - Etiology

- Unknown, possibly an autoimmune reaction against gangliosides (GM₁)

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

MMN - Procedure: EMG

The muscles should be chosen based on the clinical muscle weakness; if weakness is widespread, the following muscles are recommended

- m.interosseus dorsalis I
- m.biceps brachii/m.deltodeus
- m.tibialis anterior/m.gastrocnemius caput mediale
- m.vastus lateralis
- m.trapezius/orbicularis oris/m.genioglossus

MMN - Procedure: neurography

- Motor nerves should be studied bilaterally
- N.Medianus
- N.ULNaris
- N.Peroneus
- N.Tibialis
- Sensory nerves
- N.Suralis
- N.Radialis
- It is recommendable to test the sensory nerve conduction in the segment with motor conduction block
MMN patient: history

- 52 year old engineer
- No significant disorders previously
- Since 10 years slowly increasing weakness of hand muscles
- During last year some weakness in the feet
- No obvious sensory symptoms

Clinical findings

- Slight atrophy of distal hand muscles
- Marked weakness of distal hand muscles, especially thenar muscles on the left side
- Slight weakness of ankle dorsiflexion
- Tendon reflexes symmetric
- No sensory abnormalities
- Plantar reflex normal

EMG findings

Clinical finding

Slight atrophy of distal hand muscles
Marked weakness of distal hand muscles, especially thenar muscles on the left side
Slight weakness of ankle dorsiflexion
Tendon reflexes symmetric
No sensory abnormalities
Plantar reflex normal

Patient: MCV findings

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<th>Sensor</th>
<th>Motor</th>
<th>Sensory</th>
<th>CV</th>
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<tr>
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<td>Ulnaris</td>
<td>5.9</td>
<td>18.1</td>
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Sensory neurography findings 1

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<th>MCV</th>
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SCV findings 2

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</tbody>
</table>
Left median nerve MCV

Median nerve sensory neurography

antidromic SCV: elbow to wrist

Right median nerve MCV

Spinal muscular atrophy (SMA)

Spinal muscular atrophy

- Hereditary motor neuronopathies
- Mostly proximal
- Some distal

SMA history

- 1891 Werdnig two brothers with proximal weakness
- 1893 Hoffman, two families with four children
- 1848 Aran distal SMA
- 1951 Sven Brandt systematic description
- Irina Hausmanowa.Petrusewicz
- John Pearn
SMA history
- 1990 International SMA Consortium
- SMA diagnostic criteria
- 1990 gene mutation detected

SMA – General aspects
- One of the most common recessive hereditary disorders
- 1: 10,000 newborn affected
- Gene carriers 1:50
- Homozygous mutation in the SMN1-gene

SMA genetics
- Chromosome 5q13
- SMN = survival motor neuron gene 1 & 2
- SMN1 in the telomeric part
- Homologous SMN2 in the centromeric part
- SMN includes 8 exons (1, 2a, 2b, 3-8), stop codon at the end of exon 7
- SMN1 and 2 differ from each other only in exons 7 & 8 (one base pair in each)

SMN gene on chromosome 5

SMA gene
- SMN1 and SMN2 code the survival motor neuron – protein
- SMN1 gene produces 90% of the SMN protein
- SMN2 alone is not capable of producing enough SMN protein
- 94% of SMA patients lack both SMN1 genes

SMA I (Werdnig-Hoffman)
- Onset usually < 3 months of age, before 6 months
- Often intrauterine onset
- Reduced movements of the fetus
- Symmetric weakness of arms and legs
  - Diffuse or proximal > distal
  - Hypotonia, swallowing difficulties, unable to sit
  - Fasciculations may be seen
  - Lack tendon reflexes
  - Weakness of respiratory muscles
  - Normal cognitive function
  - 50% die before 7 months
SMA1

Neurophysiology

- EMG
  - Abundant fibrillations in all muscles
  - Often fasciculations
  - MUPs difficult to evaluate
- Neurography
  - Sensory normal (superficial peroneal, radial)
  - Motor: low amplitudes

SMA II (Intermediate)

- Onset around 6 months, before 18 months
- Learn to sit
- All muscles weak
- Normal cognitive function

SMA II

SMA III (Kugelberg-Welander)

- Onset 2-17 years
- Muscle weakness, proximal > distal
SMA diagnosis
- Clinical findings
- SMN-gene test abnormal in 95 % a deletion
- ENMG
- Neurography
- Muscle biopsy
- Fiber type grouping and group atrophy
- SMA I ja II: type 1 hypertrophy
- SMA III (ja IV): reinnervation

SMA I differential diagnosis
- Floppy infant syndrome
- CNS malformation
- Perinatal asphyxia
- Metabolic disorders
- Prader-Willi, Zellweger sdr
- Congenital myopathies

SMA III differential diagnosis
- May clinically present like a proximal limb-girdle dystrophy

Kennedy syndrome
- Onset 15-60 years, mean 27
- Muscle weakness
  - Legs > arms
  - Proximal > distal
- Bulbar symptoms
  - Dysphagia
  - Dysarthria
  - Gynecomastia

EMG findings
- EMG
- Neurogenic findings
- Bulbar muscles affected
  - Usually not much fasciculations
- Neurography
  - Sensory amplitudes reduced or absent
Kennedy syndrome
- X-chromosomal (Xq12 recessive)
- Androgen receptor
- CAG repeat
  - Normal 9-39
  - MSMA 40-65
- Toxic gain of function
- Frequency 1:50,000
- In Scandinavia common founder haplotype
  - Common ancestor

Postpolio syndrome

Acute polio
- Poliovirus types 1, 2 & 3
- Infection rate 95%
- Most infections very mild
- 1-2% develop poliomyelitis
  - 85% paralysis caused by type 1
  - High fever, myalgia, nausea, headache
  - Flaccid paralysis maximum within 48 hours
  - Some recovery
  - 1/1000 in children, 1/75 in adults

Polio
- Vaccination started 1956
  - Salk trivalent inactivated virus
  - Sabin attenuated live virus
- Dramatic reduction in poliomyelitis
- Still endemic
  - India, Pakistan, Nigeria, Niger, Afghanistan, Egypt
- Patients with perivious polio
  - European patients born before 1956
  - Immigrants may be born later

Postpolio syndrome

Postpolio syndrome (PPS)

PPS
- Past history of polio
  - In Northern Europe vaccination started 1956
  - In developing countries not all vaccinated
  - In Eskilstuna I have seen 5 immigrants from the near east and Africa < 40 years with postpolio muscular atrophy in 2009
- Stable period after poliomyelitis
- Development of new impairment
  - Generalized fatigue
  - Weakness
  - Joint and muscle pain
Epidemiology in Sweden

PPS is multifactorial
- Severe primary involvement of muscles
- Aging
- Arthrosis
- Depression
- Concurrent other diseases
  - Perceived functional deficit

PPS
- PPS – macro EMG & MRI
- No objectively measurable parameter discriminated between stable and unstable
  - EMG
  - Histology
  - Imaging
  - Muscle strength
  - Pain correlated with loss of function
Muscle strength

Macro EMG m.tibialis anterior

Macro EMG m.biceps brachii

Macro vs concentric EMG

Role of EMG in PPS

- Ascertain that the patient really had polio
- Cerebral palsy
- GBS
- Detect other concurrent disorders
- CTS
- Radiculopathies
- Polyneuropathy
- Conventional EMG does not discriminate between stable and unstable patients with previous polio
- Fibrillation potentials do not indicate PPS
- Severe involvement may be suggestive
- Macro EMG or MU counting may be helpful in evaluating the degree of MU loss
- Research tool

Normal findings with history of previous polio

- Primary diagnosis erroneous
- CP
- GBS
- Meningitis
- Other CNS disorders
- Functional
- Paralytic polio
- Motor neuron loss minimal
Hirayama's disease

**Monomelic amyotrophy**

- Male : female  10:1
- 15-25 years
- Usually sporadic, also hereditary
- C7-Th1 innervated muscles
- May be bilateral
- Progressive weakness over 1-4 weakness

**Benign fasciculations**

- Only fasciculations without other abnormalities
- Common problem
- No epidemiological studies
- Usually young subjects with no other symptoms
- Medical students or health care personnel
- Duration of fasciculations variable
  - Sometimes lifelong
- Not a prelude to motor neuron disease
- Fasciculations are very rarely the first symptom of ALS
- Most ALS patients are not aware of their fasciculations

**EMG in benign fasciculations**

- 4-6 muscles
- Demonstrate fasciculations
- No fibrillations
- MUPs normal
- Normal neurography

**Game over**