Ultrasound of muscle disorders

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Disclosures

None

# "Muscle ultrasound disclaimer"

Muscle ultrasound

Ø Introduction

Ø Specific NMD

Ø Quantification

Ø Dynamic imaging

Ø Does US help diagnose NMD patients?

Muscle ultrasound

Muscle ultrasound: introduction

THE LANCET, JUNE 28, 1980

Preliminary Communication

DETECTION OF PATHOLOGICAL CHANGE IN DYSTROPHIC MUSCLE WITH B-SCAN ULTRASOUND IMAGING

JOHN Z. HECKMATT   VICTOR DUBOWITZ
SIDNEY LEEMAN

Department of Pædiatrics and Medical Physics and Jerry Lewis Muscle Research Centre, Hammersmith Hospital, Du Cane Road, London W12 0HS
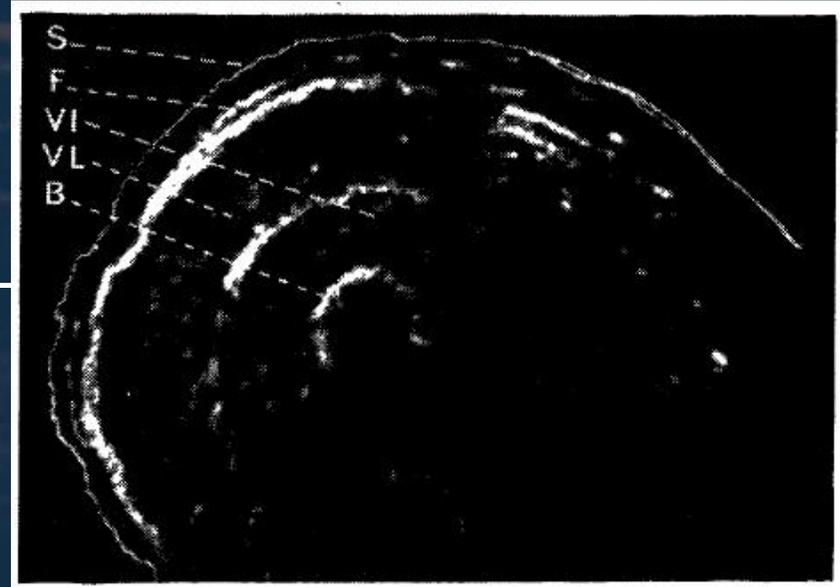
Introduction: muscle ultrasound

Rectus femoris

3.12 cm

vastus

femur
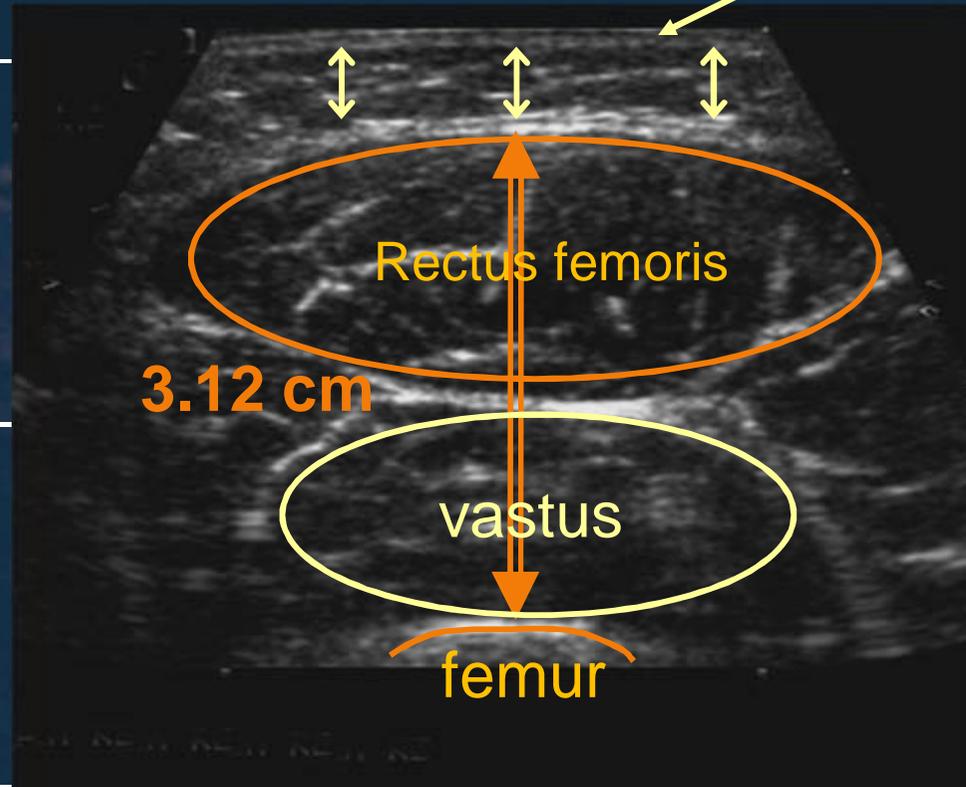
Normal quadriceps muscle

Muscle type and build


biceps brachii

Tibialis anterior

Latissimus dorsi

Muscle trauma

Normal gastrocnemius

Rupture with hematoma

Muscle identification & anatomy teaching

Flexor carpi radialis
and pronator teres
muscles in forearm

Forearm extensors
with panoramic view
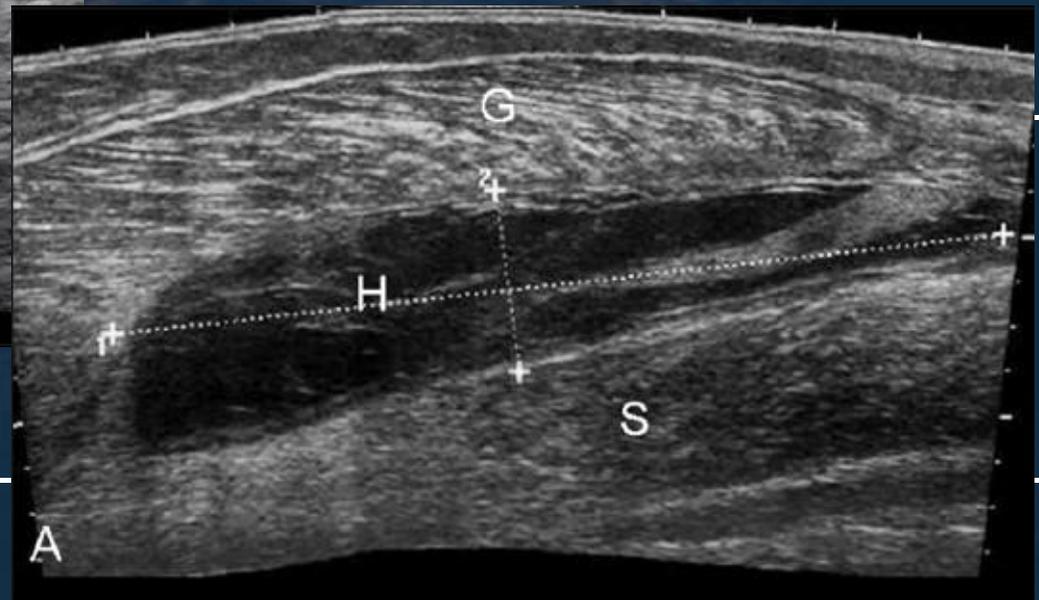
Needle guidance for EMG / injection therapy

Muscle ultrasound in neuromuscular disorders:

"256 shades of grey"
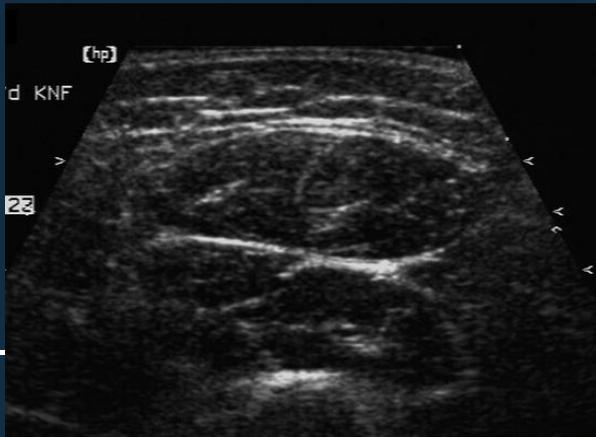

Healthy
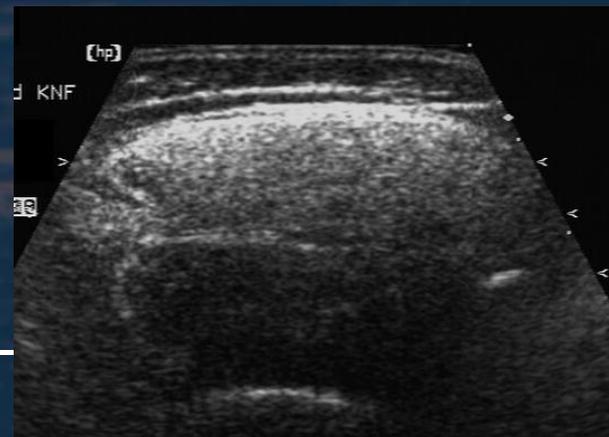
Duchenne muscular dystrophy

Muscle ultrasound

**Every NMD has is own appearance:**
DMD: homogeneous affected and strong attenuation

Healthy   Ambulant DMD   Advanced stage

Typical picture of SMA:
severe atrophy and moth eaten pattern
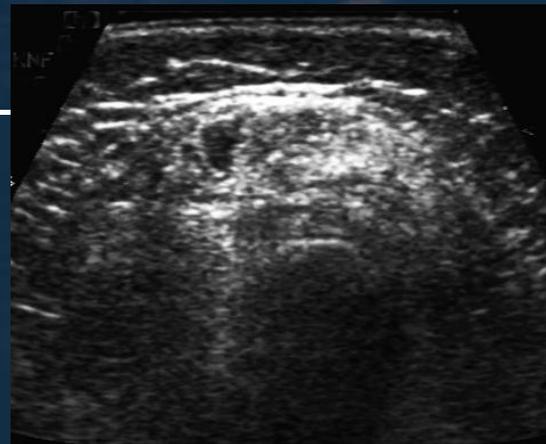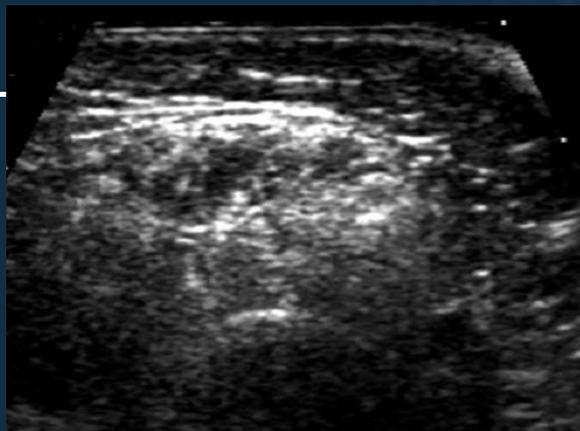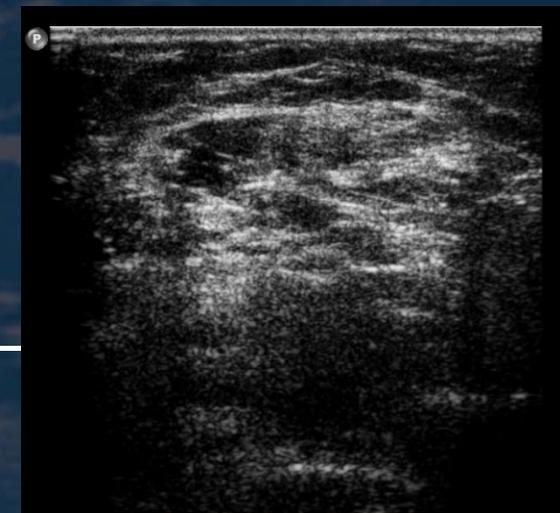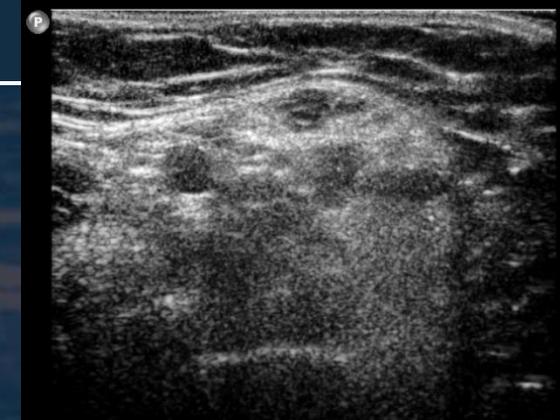
Inflammatory myopathies:

Focal changes

Increased perfusion

Fasciitis
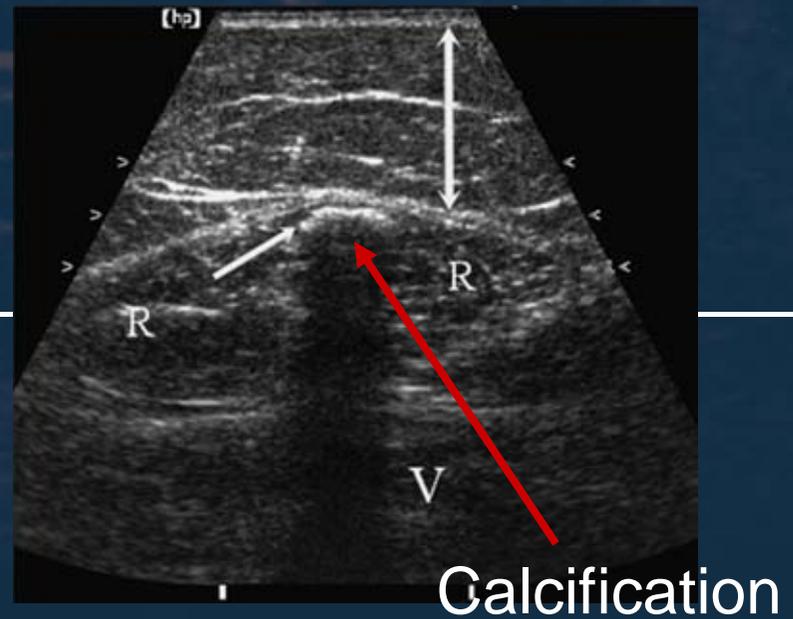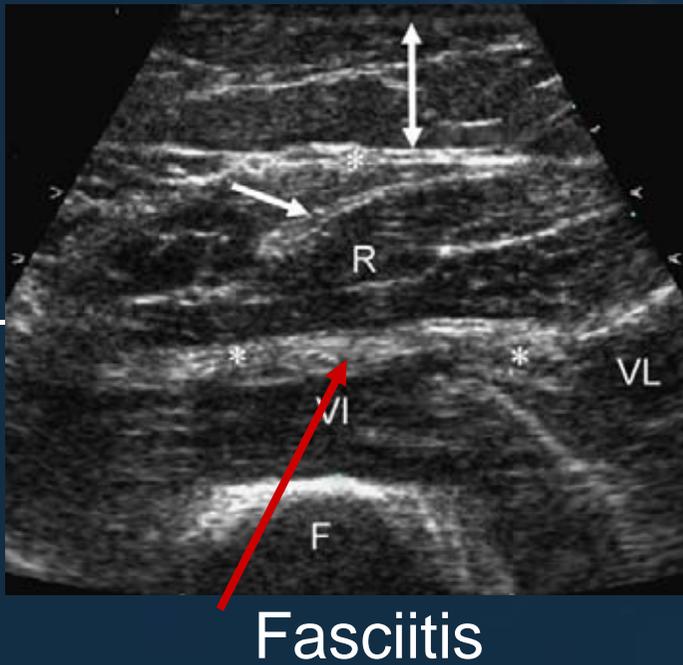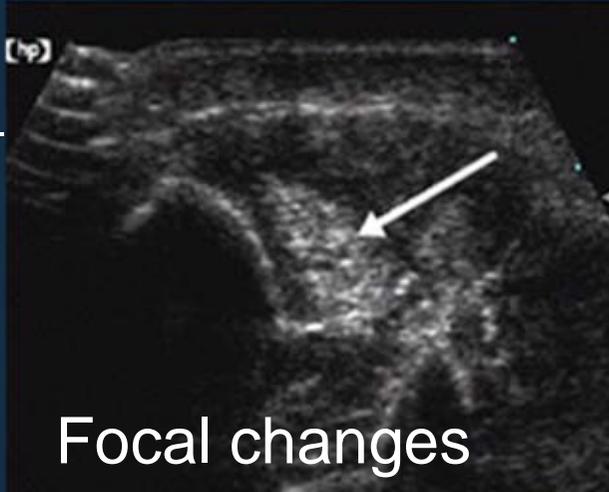
Calcification

Ultrasound in Bethlem myopathy
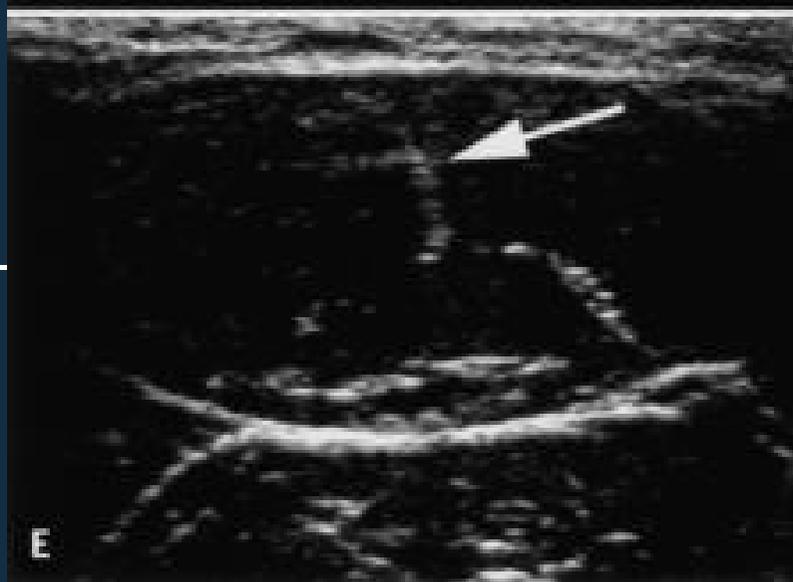
Healthy: central aponeurosis visible
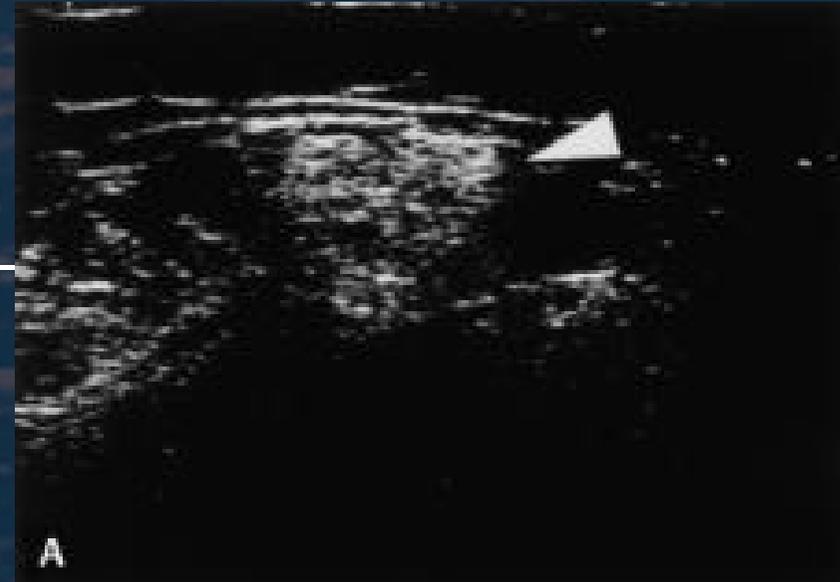
Bethlem myopathy: central shadow sign

Bonnemann Neuropediatrics 2003

Muscle ultrasound

Subjectivity of our visual system

How grey is that image: age dependency

Brother — Normal?

Dad

Grandpa — NMD?
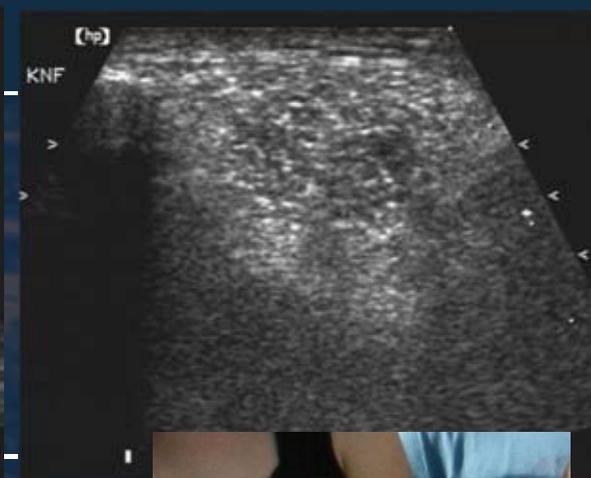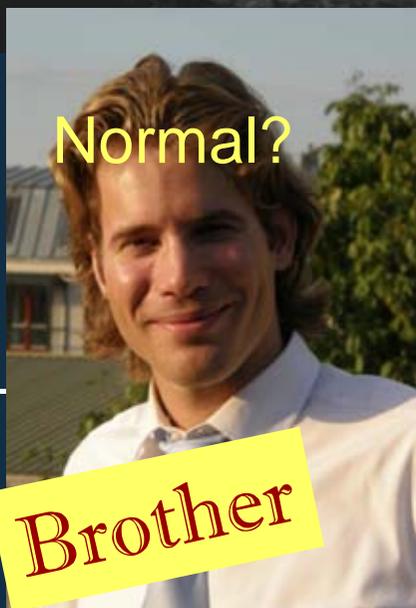
Normal aging!

Echo intensity: age dependent
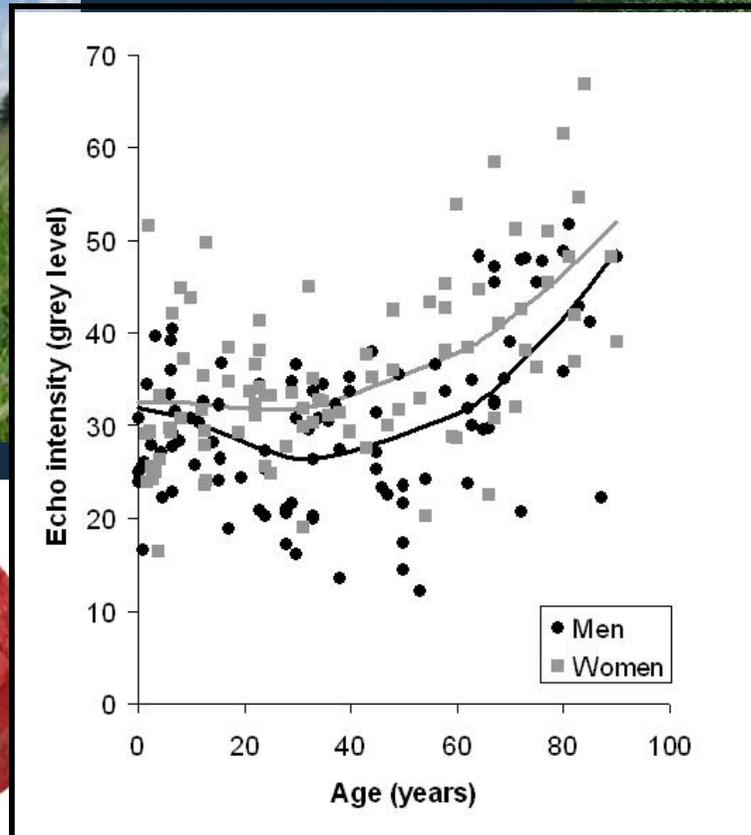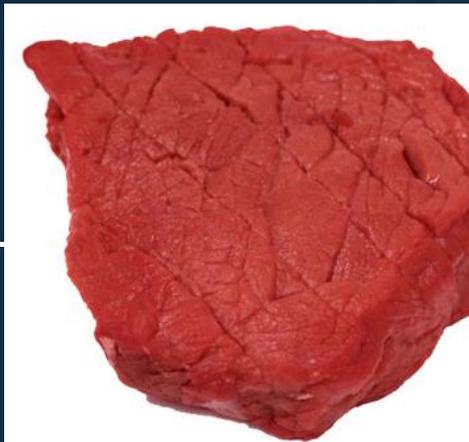
Quantitative echo intensity analysis
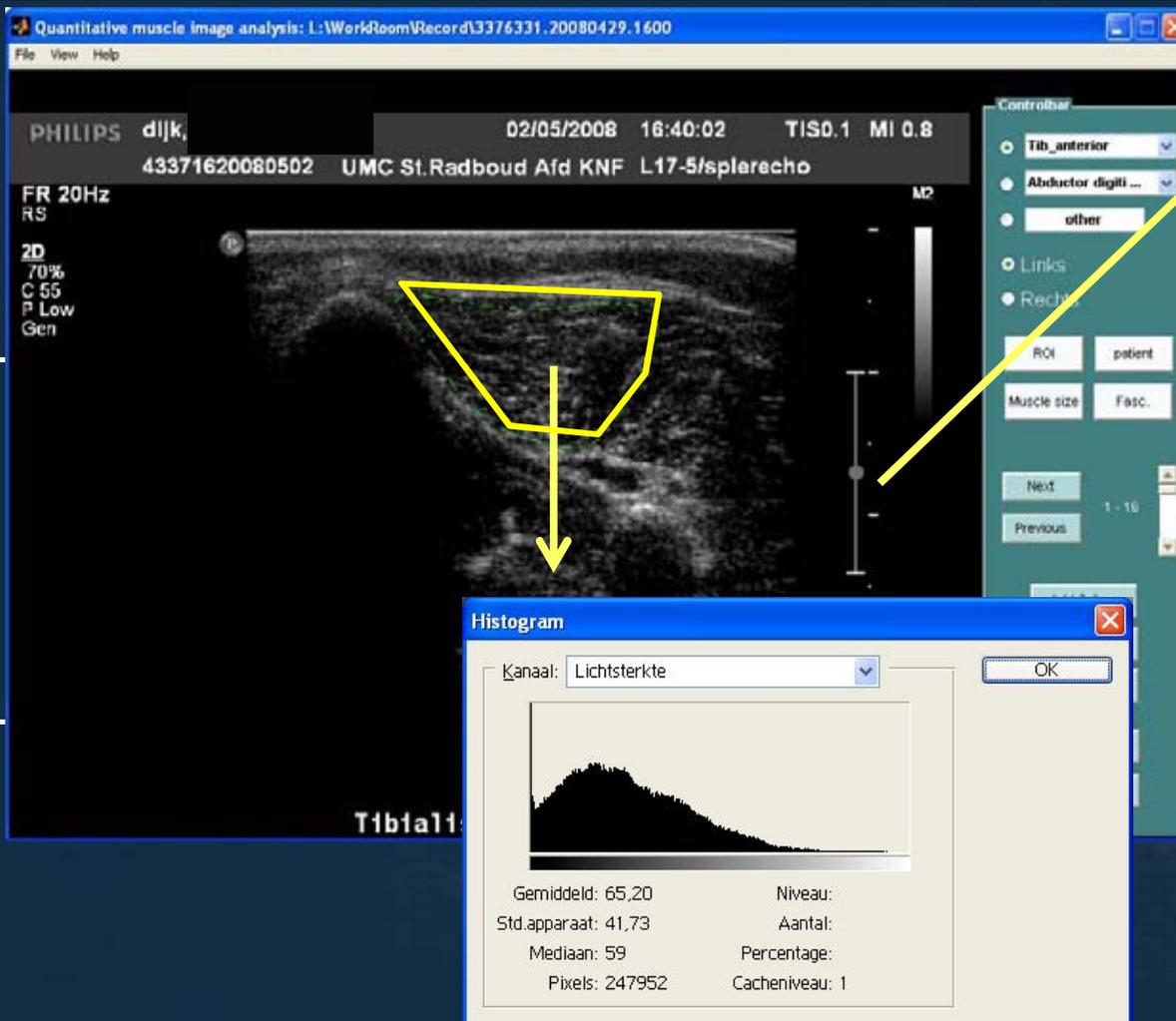
Mean echo intensity

Compare with normal value

Number of SD above normal (= **z score**)

Muscle echo intensity quantified
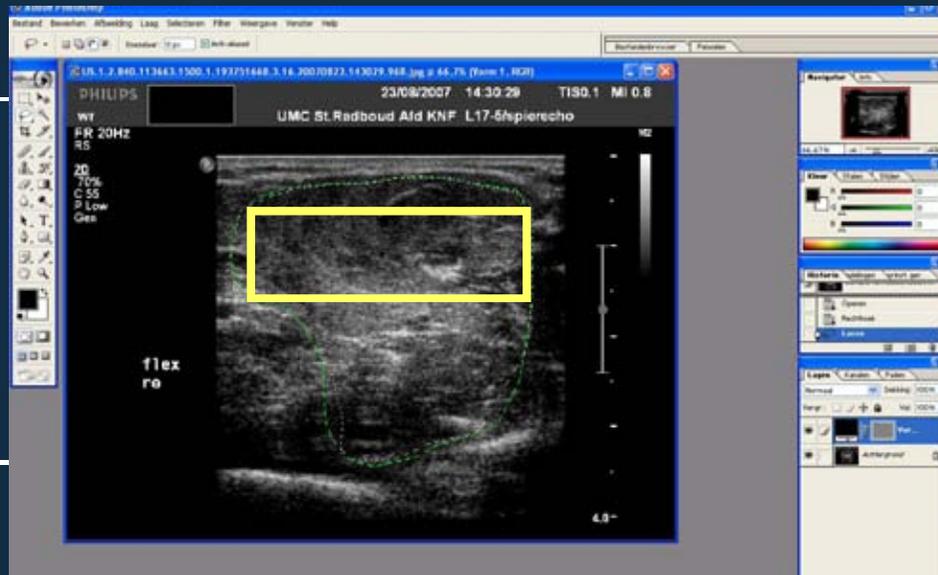
Mean grey value

Pro:

• Well investigated, high diagnostic value

Contra:

• Software and hardware dependent

• New reference values for each US device

Alternative quantification methods
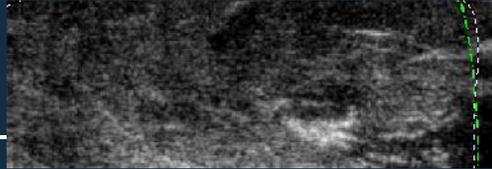
ROI: Mean grey value

Compare with:

Phantom

Subcutaneous tissue
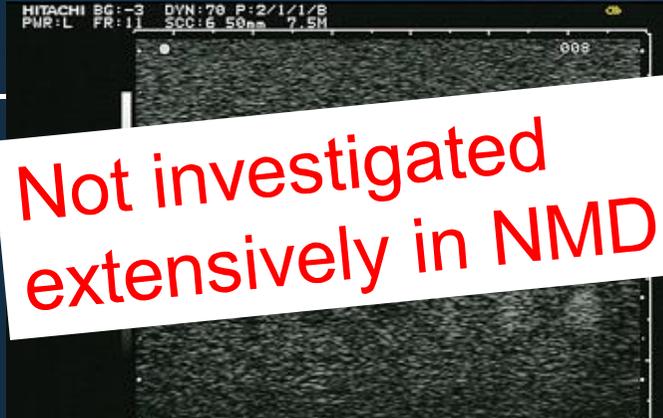
Not investigated extensively in NMD

Not reliable
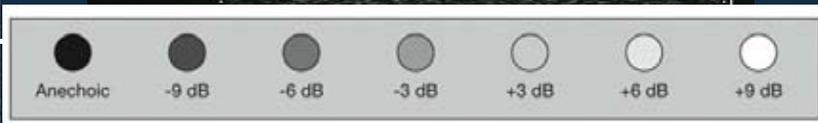
Backscatter analysis
- Not software dependent
- Less but still hardware dependent

Luminosity ratio
- Less but still software *and* hardware dependent
- Difficult in very thin patients, edema..

QMUS reporting

Ø Neuromuscular disorder yes/no?

Ø Distribution of abnormalities: differential diagnosis

Ø Muscle biopsy location
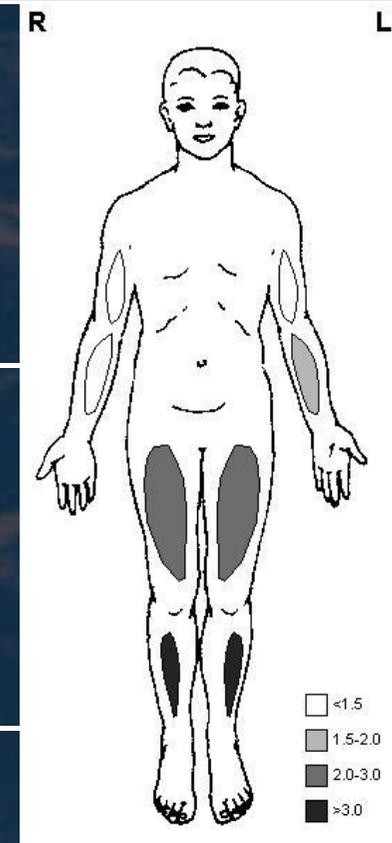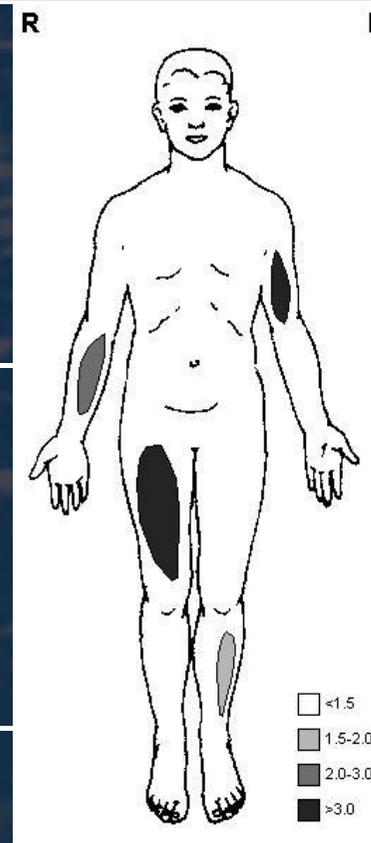
Dermatomyositis Polyneuropathy

Optimal muscle biopsy site identification
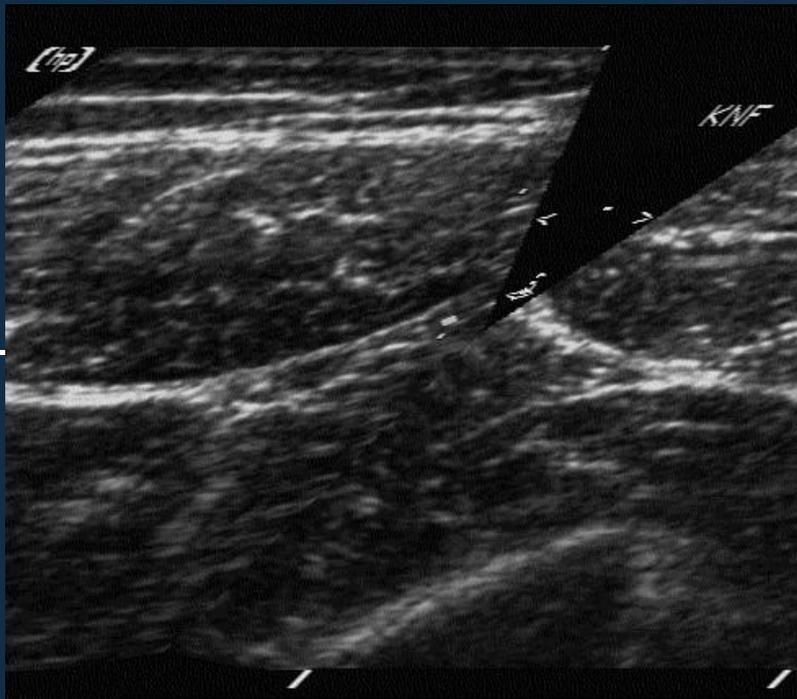
Rectus femoris:
+ 1 SD
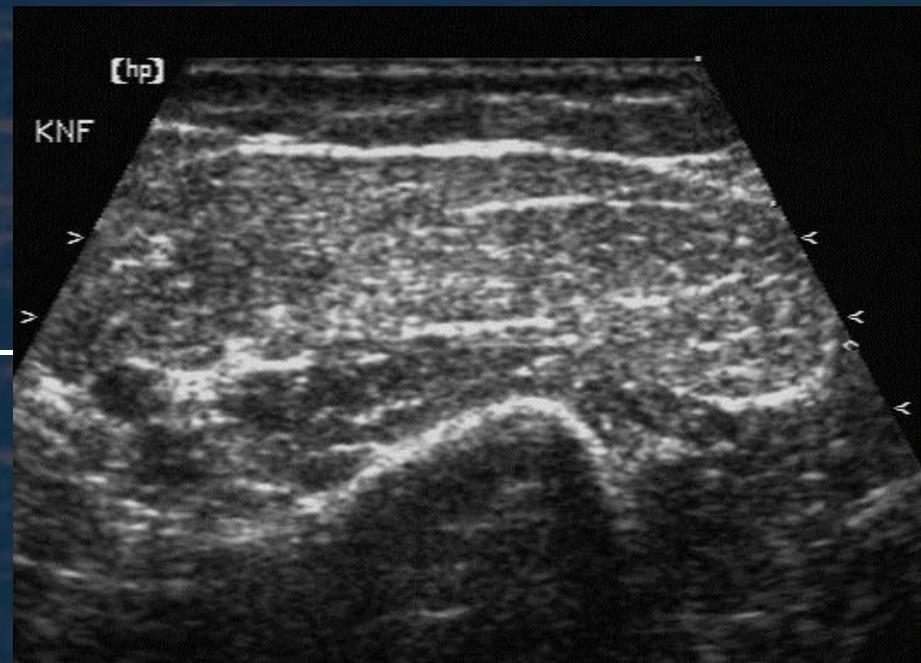
Biceps brachii:
+ 3 SD

Optimal biopsy site: avoid "too abnormal"
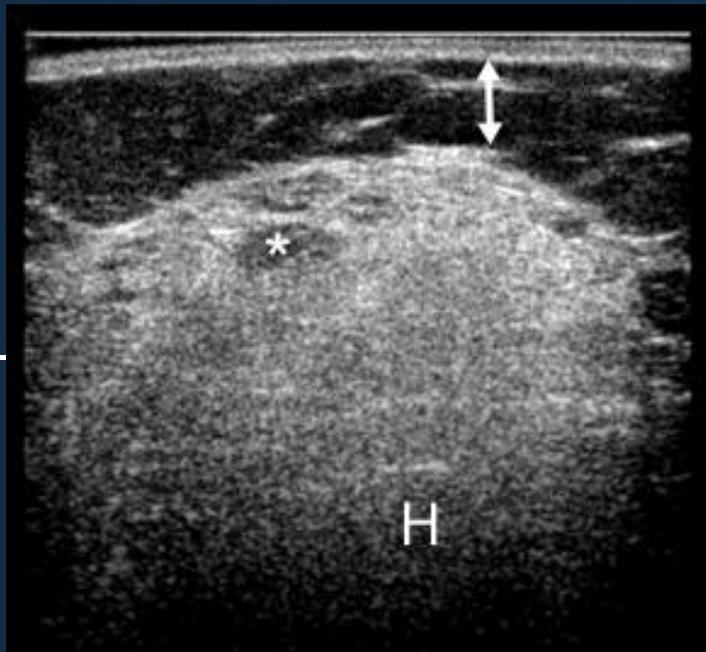
Biceps brachii:
only fat/fibrosis

Search for muscle with
remaining muscle tissue

Muscle ultrasound

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Ø Dynamic imaging

Ø Does US help diagnose NMD patients?

You see more…

… when it's moving
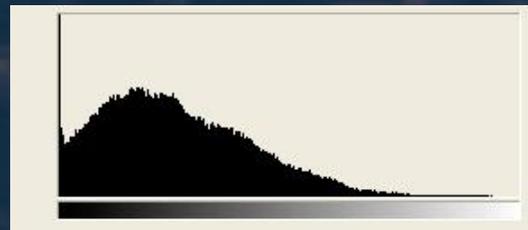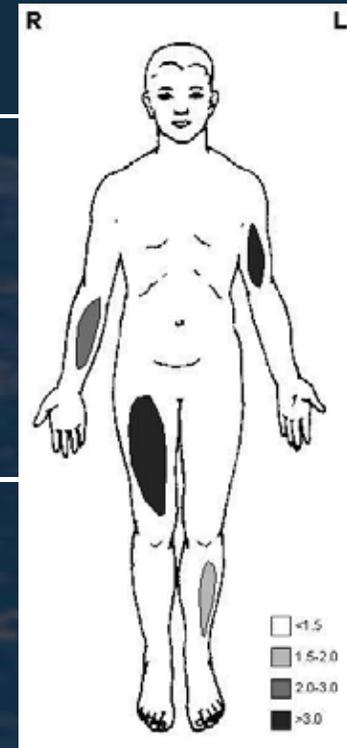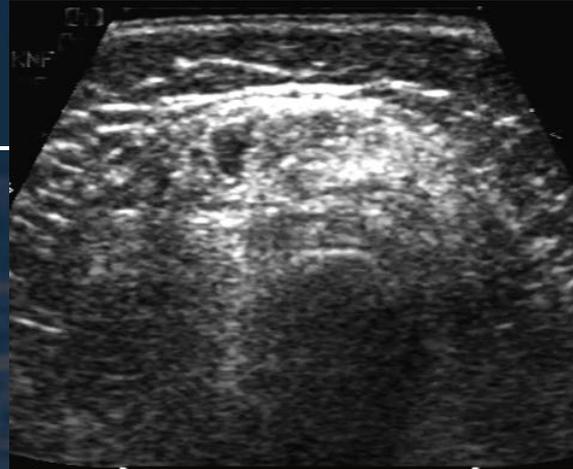
Muscles: you see more when they´re moving!

Healthy biceps brachii muscle

US is more sensitive for detecting fasciculations than needle EMG
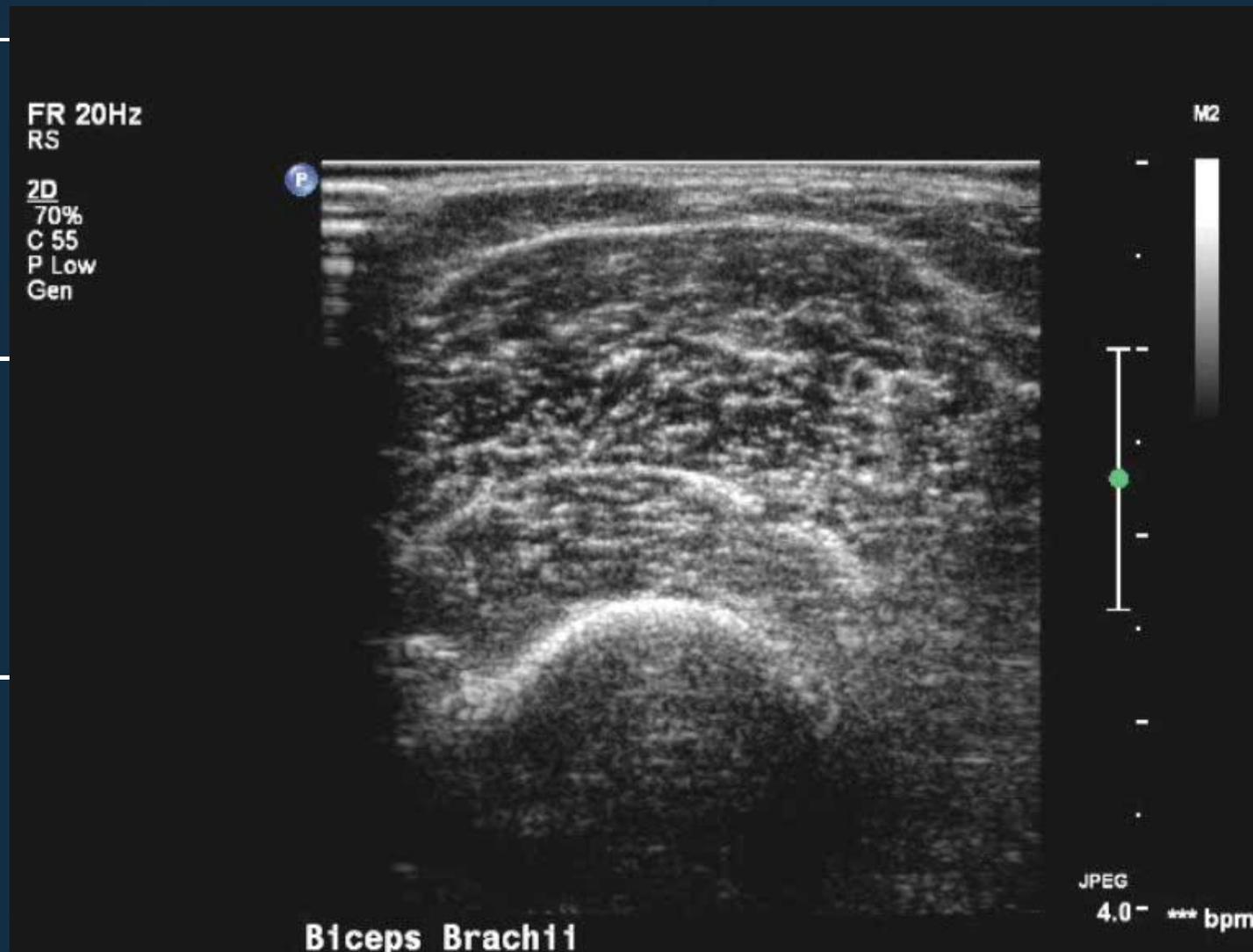
Fibrillations on ultrasound

Proof: combined ultrasound – EMG

EMG needle

Great! But...

Ø Overall sensitivity 45%, specificity 85%

Ø Proximal muscles sensitivity 63% but distally 33%

Why?

Ø Detection only possible when at least 5 fibs/second

Ø Temperature dependent: 100% at 40?C vs 63% at 30?C

And:

Ø Only with high-end ("expensive") US-machines: high frequency, high frame rate, little noise

Fasciculations and fibrillations can easily be differentiated
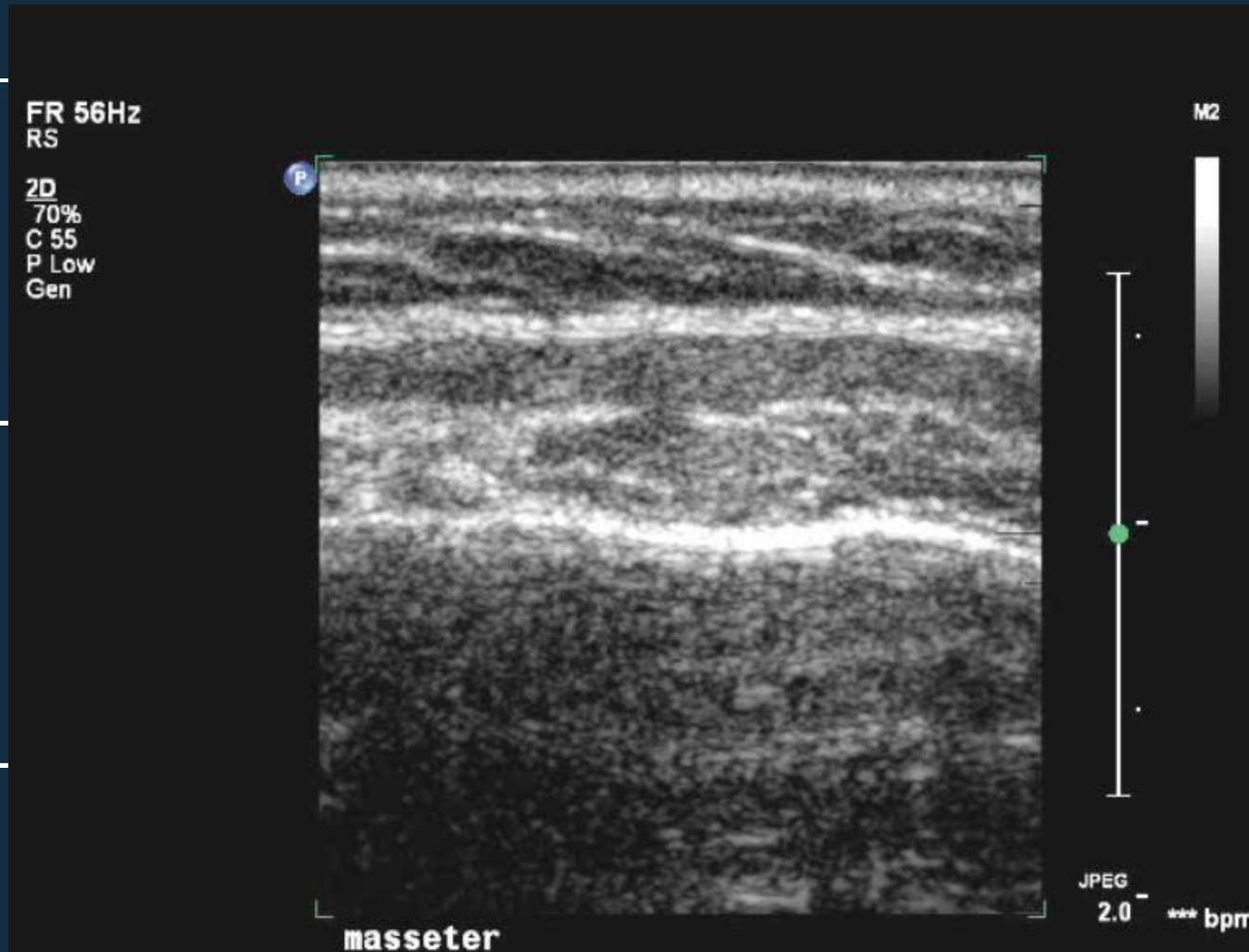
Pseudotremor: contraction of large neurogenic units
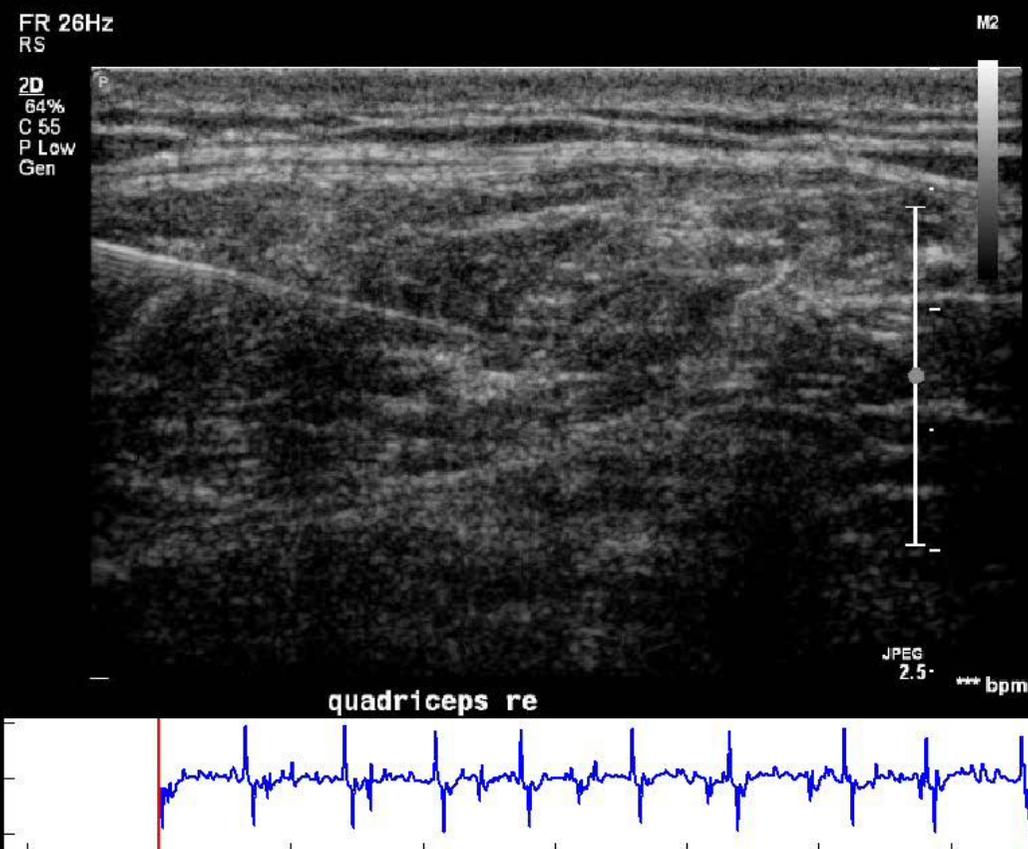
Muscle ultrasound

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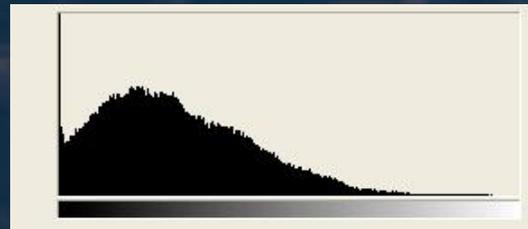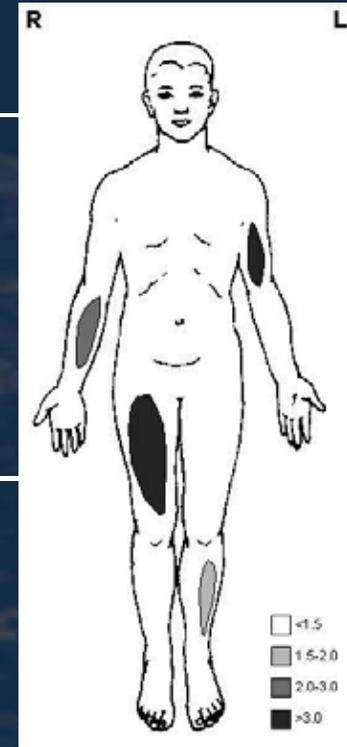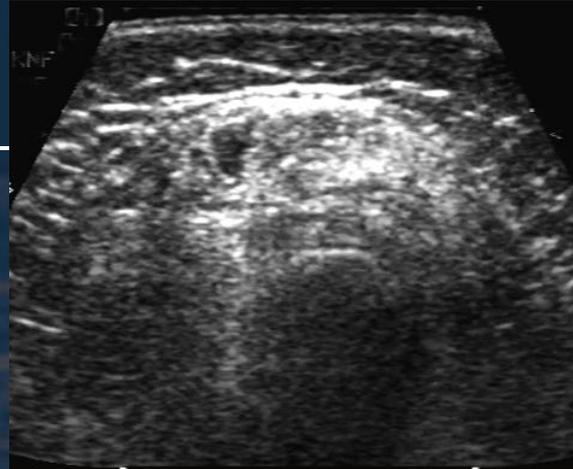
Diagnostic value in children with suspected NMD

Visual evaluation: sensitivity at best +/- 70%

Quantitative evaluation: sensitivity >90%

- ø Positive predictive value: 90%

- ø Negative predictive value: 86%

Lower sensitivities in:
- ø Children < 3 years: sensitivity 75%
- ø Mitochondrial myopathies: sensitivity 45 – 70%

Pillen et al NMD 2007
Pillen et al Neuroped 2006
Pillen et al US Med Biol 2006

Can US aid in diagnosing ALS? - revised El Escorial criteria
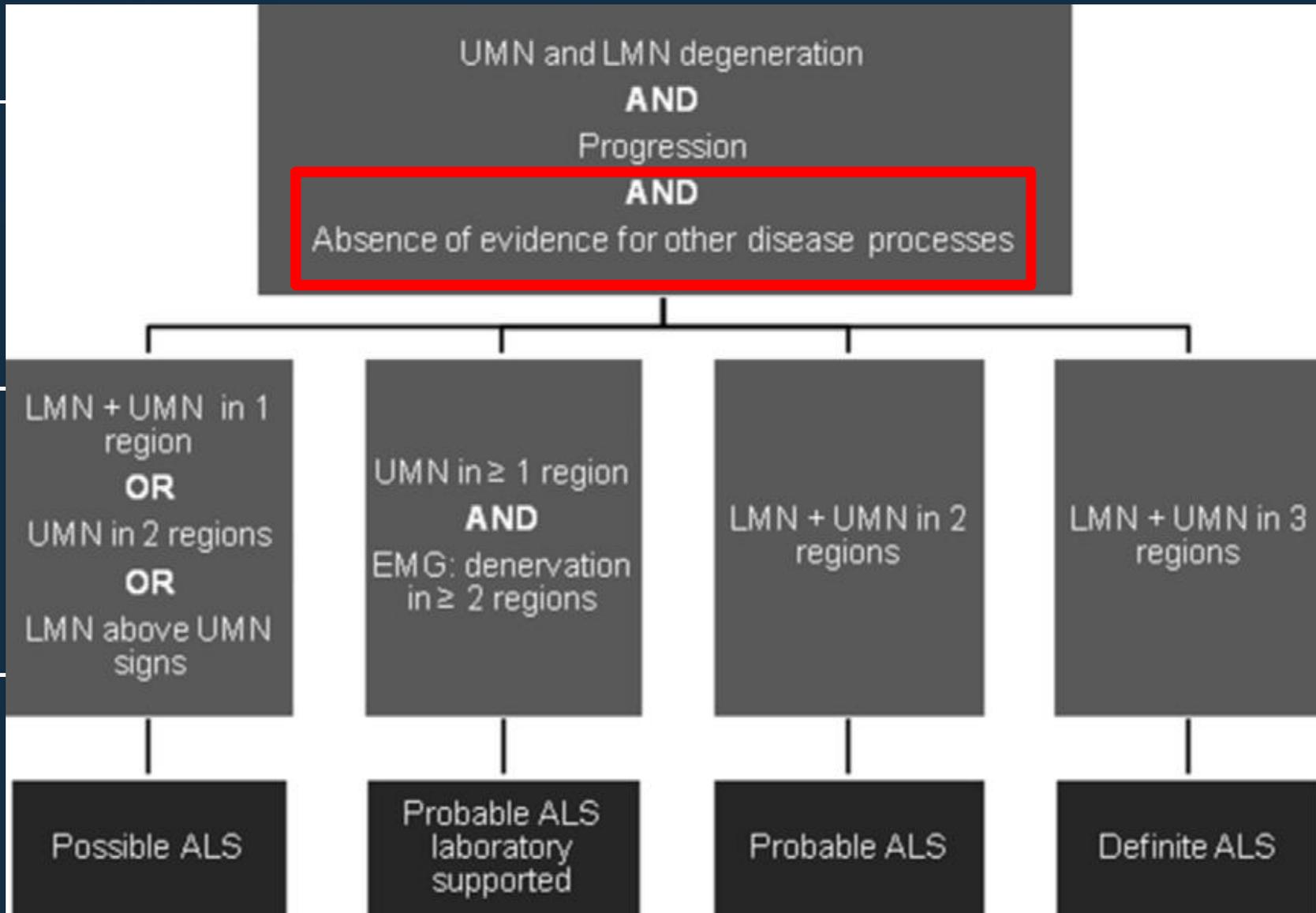
Typical US findings in ALS at diagnosis

Ø Lots of fasciculations

Ø Mild to moderately increased EI

Ø Atrophy only in paretic muscles
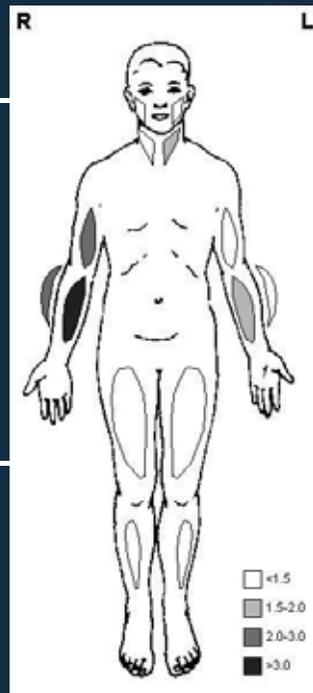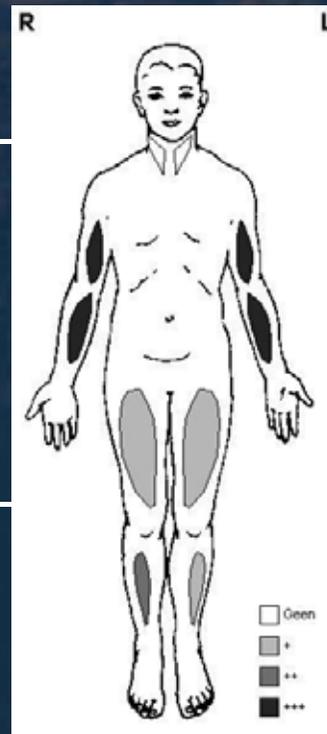
EI   Fasciculations   Arts US med biol 2008

60-year old patient with symptoms suggestive of ALS

biceps

rectus femoris

2cm: atrophy

EI
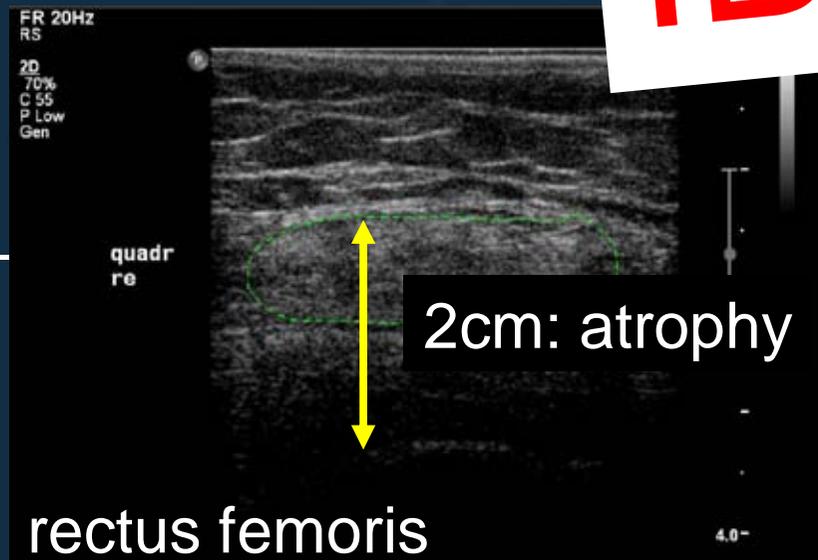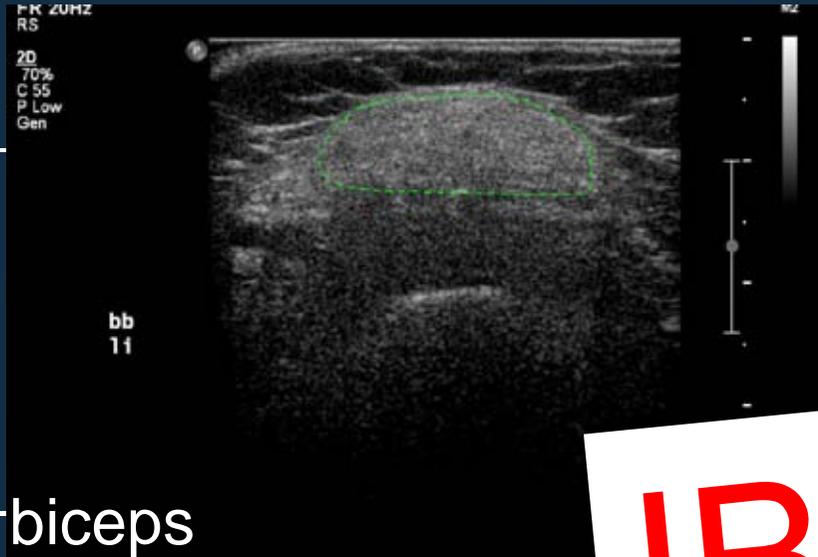
Fasciculations

IBM
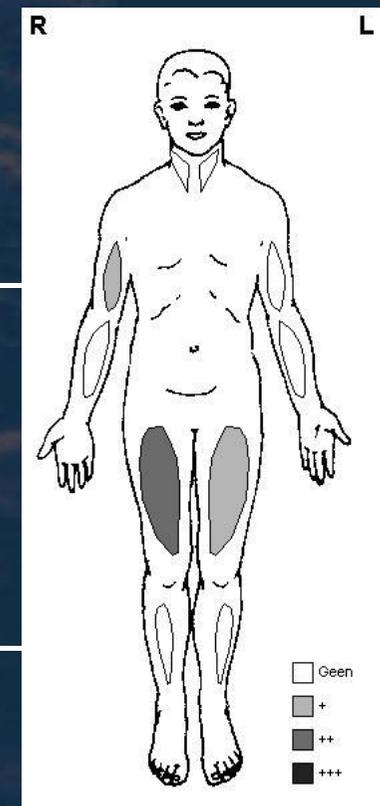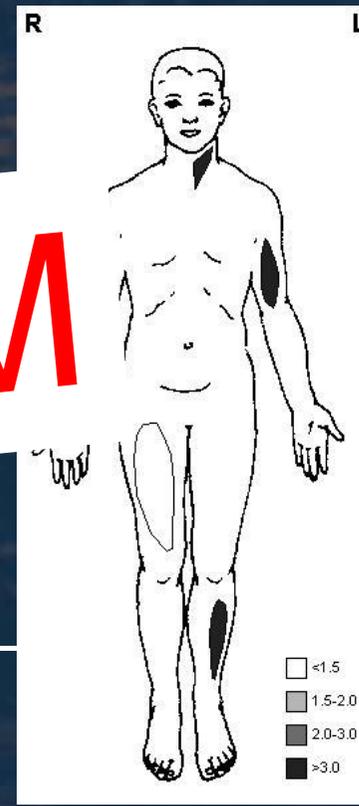
Typical US findings in IBM

Ø Severely increased EI

Ø Pronounced atrophy

Ø No/some fasciculations
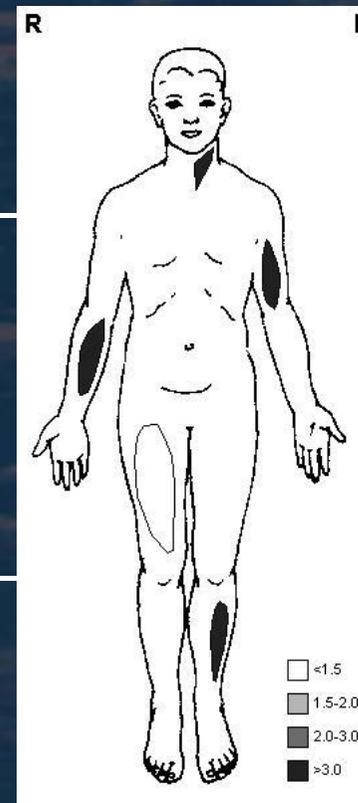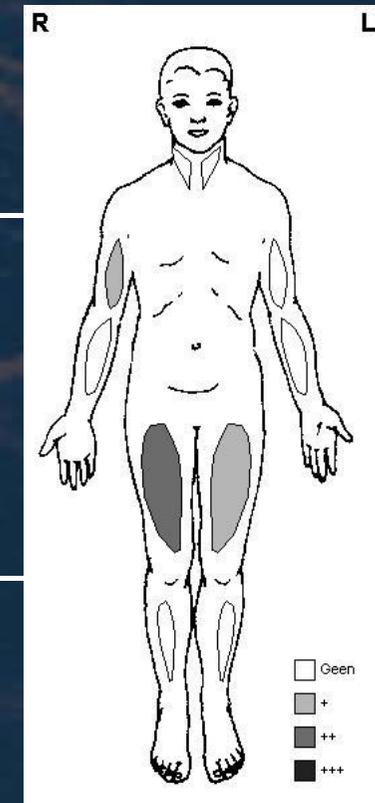
EI   Fasciculations

How to differentiate ALS mimics

- Ø Cervical and lumbar spinal stenosis with myelopathy
  - Ø NO BULBAR OR THORACIC ABNORMALITIES
- Ø Benign cramp fasciculation syndrome
  - Ø NORMAL EI
- Ø Inclusion body myositis
  - Ø EI MORE INCREASED, LESS FASCICS
- Ø Multifocal motor neuropathy (with silent cerebral lesion)
  - Ø FOCAL NERVE ABNORMALITIES?

US in ALS and mimics

1. Cross sectional study

    Ø 48 ALS-patients and 27 ALS-mimics

    Ø 10 muscles measured for EI and fascics

    Ø Optimal cut-off point defined:

        • ? 2 muscles with EI > 1.5

        • ?≥4 muscles with fasciculations

2. Prospective study 59 patients with suspected ALS (27 ALS, 32 mimics)

    Ø Sensitivity 96%

    Ø Specificity 84%

Arts US med biol 2008
Arts Clin Neurophys 2012

Diagnosing ALS - revised El Escorial criteria
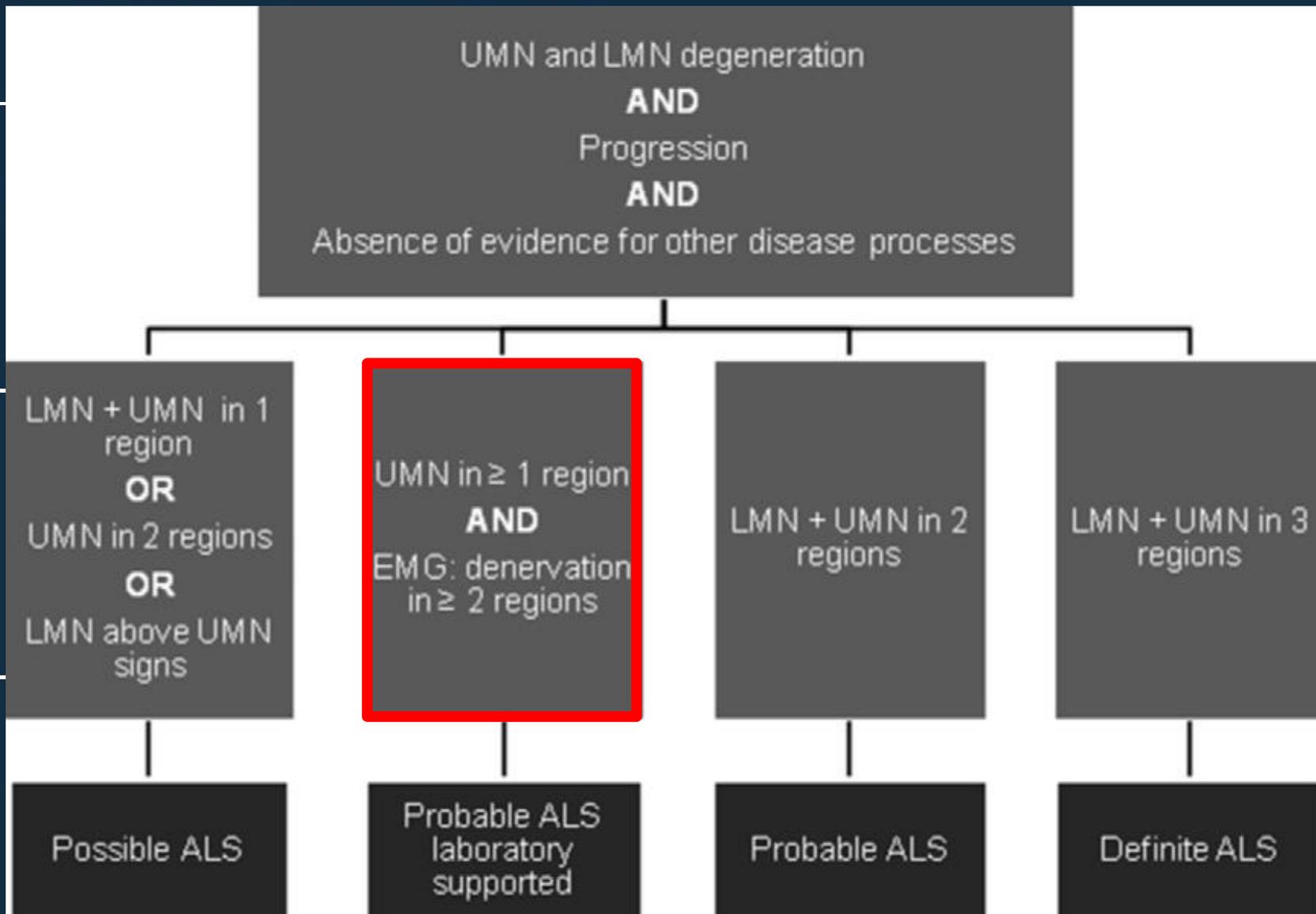
Diagnosing ALS - revised El Escorial criteria
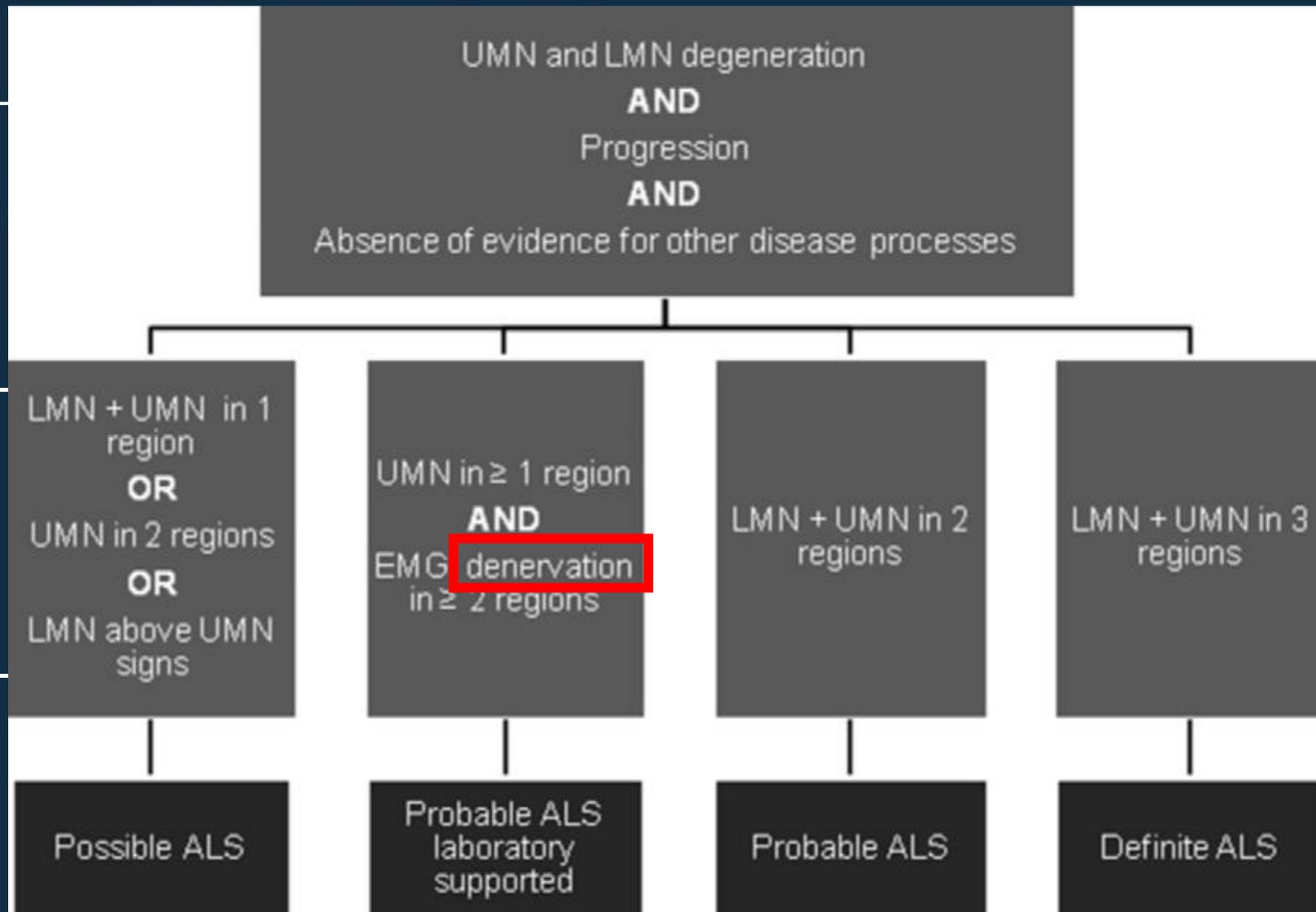
"Fasciculations = denervation" ?Œwaji criteria

Ø US can detect fasciculations in 10-30% of the *muscles* that are EMG negative [1,2,3]

Ø US can increase diagnostic certainty by detecting subclinical involvement of EMG negative *regions* [4]

Ø US added to EMG:[5]

 Ø 5% of patients possible ?^ probable/definite ALS

 Ø 20% of patients probable ? definite ALS

[1] Walker Muscle Nerve 1990
[2] Wenzel J Neuroimaging 1998
[3] Reimers J Neurol 1996
[4] Arts Clin Neurophys 2012
[5] Misawa Neurology 2011

Fasciculations in ALS: screening of "unusual muscles"
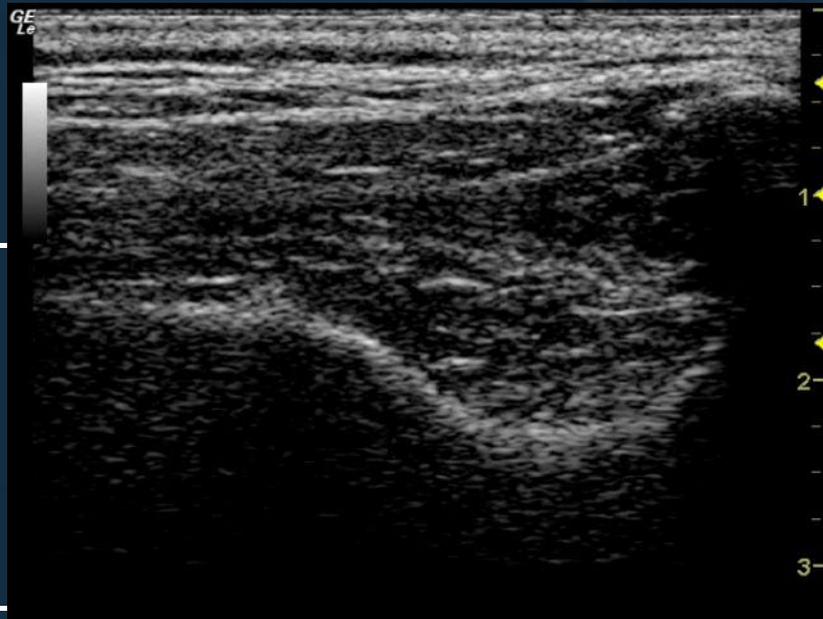
Temporalis muscle
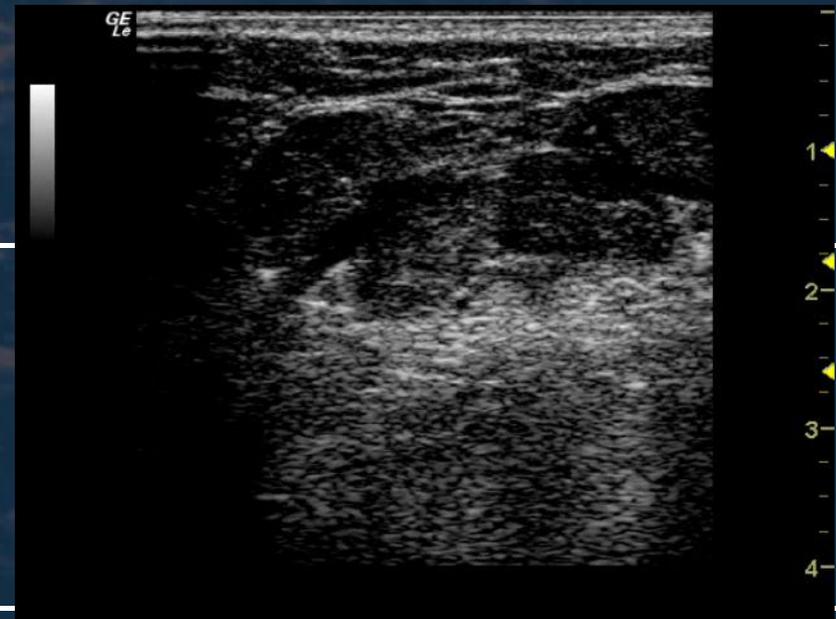
Tongue (submental muscles)

To conclude

Muscle ultrasound

Ø is a good screening tool for NMD in children

Ø can be used to determine the optimal muscle biopsy site

Ø is better in detecting fasciculations than EMG

Ø (as an add-on tool to EMG): increases *clinical* diagnostic certainty in ALS

Thanks for your attention!