What can neuromuscular ultrasound do for you?

2017 Gloor Lecture

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Radboud university medical center
Nijmegen, The Netherlands
Learning objectives

• How to recognize normal and pathologic nerves and muscle using ultrasound

• What the value of neuromuscular ultrasound is in screening for muscle or nerve disorders:
  • Adding to an ALS diagnosis
  • Helping out when EMG can’t (anymore)
  • Preventing needle sticks in unwanted places
  • As a biomarker for treatment trials in dystrophy
Disclosure Statement

Over the last 2 years I have been or am affiliated with:

☑ Ipsen Pharmaceuticals – Ultrasound trainer in botulinum toxin injection course for PM&R (payment goes to my employer)
Neuromuscular imaging

• Every neurologist uses cerebral and cervical imaging, but neuromuscular imaging is still relatively unknown and underused

• But imaging can be very helpful in NMD by providing information about:
  • lesion morphology, relation to surroundings
  • anatomical localisation of abnormalities (also for biopsy)
  • distribution of abnormalities
  • evaluating regions that can’t be accessed by EMG
Neuromuscular US
Muscle ultrasound: “256 shades of gray”
Muscle dystrophy: progressively more diffuse white

Biceps brachii

Rectus femoris

Tibialis anterior

48 y old male with FSHD1
Inflammatory myopathy: focal involvement
Neurogenic disorders: “moth eaten” pattern
Quantitative muscle US: grayscale analysis

- Mean echo intensity
- Compare to reference value
- Number of SD above normal (= z score)
Dynamic muscle ultrasound: fasciculations

Skin
Subcutis
Biceps brachii
Brachialis
Humerus
Dynamic muscle ultrasound: fasciculations
Nerve ultrasound

- Nerves are round, oval or triangular structures
- Internal hypechogenic, nodular aspect
- (For surgeons: they look just like a cut nerve ;-) )
Nerve ultrasound – median nerve example

Transverse view of the median nerve

Longitudinal view of the median nerve in the carpal tunnel
Dynamic nerve US: ulnar nerve trauma
Ulnar nerve trauma: stump neuroma
So, what can NM US do for you?

- Reliably screen for neuromuscular pathology
  - myopathies
  - neuropathies
- Provide more certainty about an ALS diagnosis at presentation
- Make a diagnosis when EMG can’t (anymore)
- Prevent you from sticking needles in unwanted places
- Be a non-invasive biomarker for treatment trials
- And maybe more?
Screening for neuromuscular disease
Muscle ultrasound - diagnostic values

- Visual muscle US screening: Sens ≈ 70%
- QMUS screening any NMD: Sens > 90%, PPV 90%, NPV 86%
- SMA type 2-4: Sens ≈ 100%
- Fasciculation screening vs EMG: ≈ 20% more fascics
- ALS versus mimics Sens 96%, Spec 84%
- QMUS added to EMG in ALS: 25% more certain diagnosis
- Diaphragm/phrenic neuropathy Sens 93%, Spec 100%
Recommended: US screening children for NMD
Nerve ultrasound screening for entrapment
CTS – nerve swelling and notch
Ulnar neuropathy at the elbow
Ulnar neuropathy at the elbow

CSA 8 mm$^2$ = normal

CSA 19 mm$^2$ = abnormal
US in inflammatory neuropathy - CIDP
Nerve US –
Screening for inflammatory neuropathies

*Diagnostic value of sonography in treatment-naive chronic inflammatory neuropathies.*


**Results:** Enlargement of median nerve at forearm >10 mm², upper arm >13 mm², and any trunk of brachial plexus >8 mm² was 99% specific for chronic inflammatory neuropathies. A shortened HRUS protocol for detecting this abnormal nerve enlargement showed high sensitivity (83%-95%), positive predictive value (100%), and negative predictive value (98%) in discriminating CIDP, LSS, and MMN from clinical mimics.
Providing a more certain ALS diagnosis
Typical US findings in ALS @diagnosis

- Lots of fasciculations
- Some muscles with increased echogenicity
- Atrophy in severely paretic muscles only
US in ALS and mimics

1. Cross sectional study
   - 48 ALS-patients and 27 ALS-mimics
   - 10 muscles measured for EI and fascics
   - 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)
   - Sensitivity 96%
   - Specificity 84%
Adding muscle ultrasound to EMG for ALS

• US can detect fasciculations in 10-30% of the muscles that are EMG negative

• US can increase diagnostic certainty by detecting subclinical involvement of EMG negative regions

• US added to EMG:
  • 5% of patients possible → probable/definite ALS
  • 20% of patients probable → definite ALS

  ➢ 25% gets a more certain diagnosis at presentation
Making a diagnosis when EMG can’t (anymore)
"Extreme carpal tunnel syndrome" (91 y.o. F)

### Motor NCS

<table>
<thead>
<tr>
<th>Nerve / Sites</th>
<th>Lat. ms</th>
<th>Ampl P-P mV</th>
<th>Dur. ms</th>
<th>Area mVms</th>
<th>Afstand cm</th>
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L MEDIANUS - APB: CMAP niet goed afgegrensbaar, proximaal niet opwekbaar

### Sensory NCS

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"Extreme carpal tunnel syndrome" (91 y.o. F)

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**Non localizing EMG**

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**Absent SNAPs**

**Very low - absent CMAPs**
US “Extreme carpal tunnel syndrome” (91 y.o. F)

Right median nerve

Wrist CSA 39 mm² (N < 11 mm²)

Left median nerve

Wrist CSA 46 mm²
Assessing denervation without needle EMG

Left calf: healthy

Right calf: S1 radiculopathy
Preventing needles in unwanted places
Manual needle placement
Manual needle placement?
Manual needle placement?
Ultrasound guided needle placement
Botulinum injection - “Blind” success rates

- Overall lower limb: 39% to 57%
- Cervical dystonia: 17% to 53%
- Upper limb dystonia: 37%
- Adult upper limb muscles in stroke: 39% to 63%
- Children limb muscles in CP: 13% to 78%
  - soleus / gastrocnemius 75%
  - hip adductors 67%
  - biceps brachii 62%
  - medial hamstrings 46%
  - forearm and hand 13%-35%
  - tibialis posterior 11%
Diaphragm EMG –
Why stick a needle in when you can have a look?
Diaphragm ultrasound

Courtesy dr. Andrea Boon, Mayo Clinic
Dynamic diaphragm ultrasound: see it move
Diaphragm US - normal versus slight atrophy

L inspiration: 3.5 mm
R inspiration: 1.9 mm
L expiration: 1.5 mm
R expiration: 1.2 mm
Would you stick a “blind” needle in this?

L diaphragm thickness: 0.8 mm

Courtesy dr. Andrea Boon, Mayo Clinic
Diaphragm US diagnostic values

Sensitivity and specificity of diagnostic ultrasound in the diagnosis of phrenic neuropathy

Andrea J. Boon, MD
Hiroshi Sekiguchi, MD
Caitlin J. Harper
Jeffrey S. Swanson, MD

ABSTRACT

Objectives: To determine the sensitivity and specificity of B-mode ultrasound in the diagnosis of neuromuscular diaphragmatic dysfunction, including phrenic neuropathy.

Conclusion: B-mode ultrasound imaging of the diaphragm is a highly sensitive and specific tool for diagnosis of neuromuscular diaphragm dysfunction.

Classification of evidence: This study provides Class II evidence that diaphragmatic ultrasound performed by well-trained individuals accurately identifies patients with neuromuscular diaphragmatic respiratory failure (sensitivity 93%; specificity 100%). Neurology® 2014;83:1-7
Sensitivity and specificity of diagnostic ultrasound in the diagnosis of phrenic neuropathy

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Neurology® 2014;83:1-7
A biomarker for treatment trials
Muscle ultrasound follow up in Duchenne

Healthy boy 10 years

Duchenne 8 years

Duchenne 16 years
Muscle ultrasound follow up in Duchenne

![Graph showing age, CMAS, Greyvalue, Echo intensity and Greyvalue CMAS relationship](image-url)
More things ultrasound could do for you

- Speed up the diagnostic process at the OPD for “Is this lump causing my hurting / tingling doctor?”
- Help with PNS injections (CTS, meralgia paresthetica, Morton’s neuroma, etc.
- Guide safe & accurate needle muscle biopsies
- Help you or your nurses with difficult lumbar punctures, IV and peripheral arterial lines
Take home: what NM US can do for you

• Reliably screen for neuromuscular pathology
  • myopathies
  • neuropathies
• Provide more certainty about an ALS diagnosis at presentation
• Make a diagnosis when EMG can’t (anymore)
• Prevent you from sticking needles in unwanted places
• Be a non-invasive biomarker for treatment trials in DMD
• And more
How to start using US tomorrow

• Come see us and learn hands-on @Radboudumc!
Neuromuscular ultrasound?