Ultrasound in Neuromuscular Disorders

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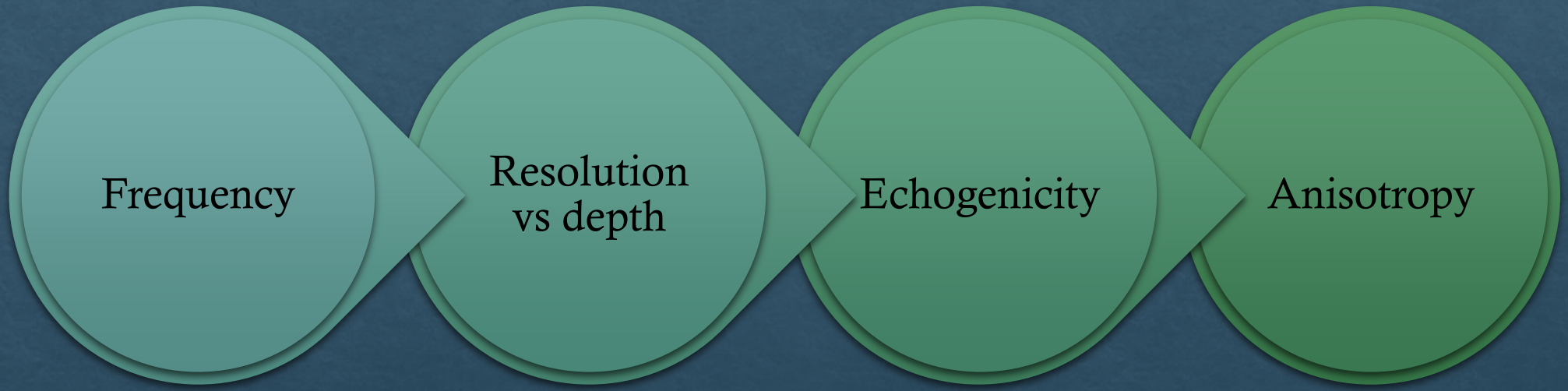
Disclosure

- I have nothing to disclose

Basic principles

- Insonation of tissue
- Analysis of Echoes
- Tissues with different acoustic impedance
- Impedance affects speed of travel of sound waves
- Bone conducts faster than soft tissue faster than water faster than air
- Use of gel minimizes reflection between skin and air

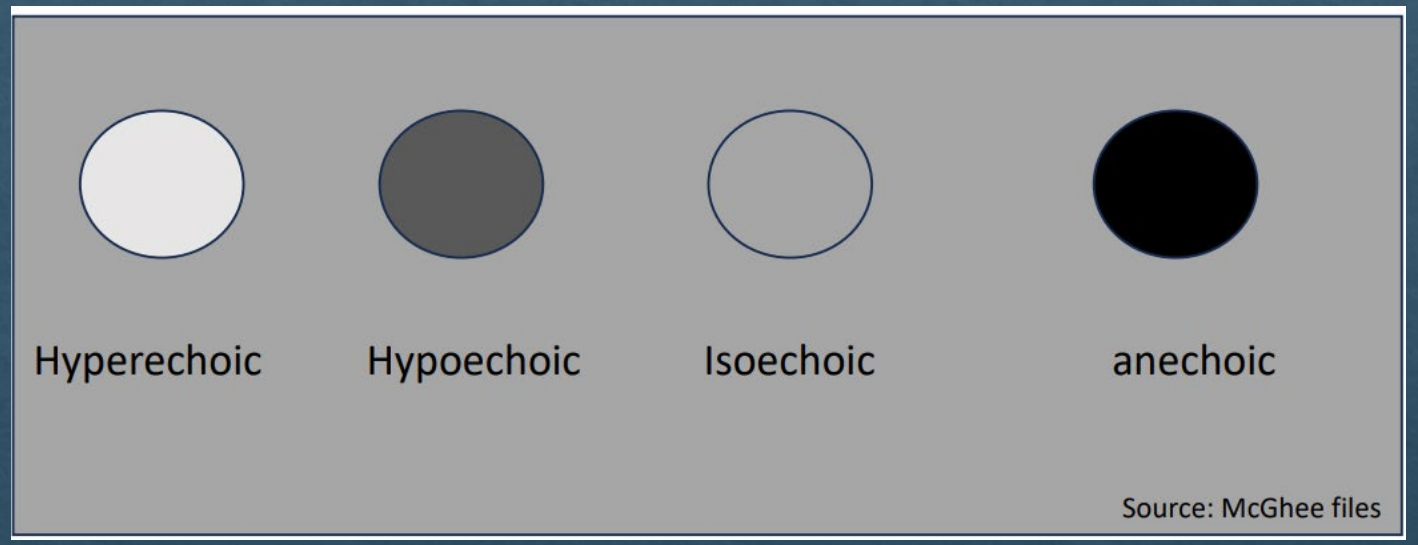

Frequency
Resolution vs depth
Echogenicity
Anisotropy

Frequency

- Frequency determines resolution and depth
- Higher frequencies for superficial structures
- Lower frequencies for deeper structures

Echogenicity

- Image brightness is referred to as echogenicity. Objects that appear bright, like bone, are hyperechoic and darker structures, like fluid, are hypoechoic.



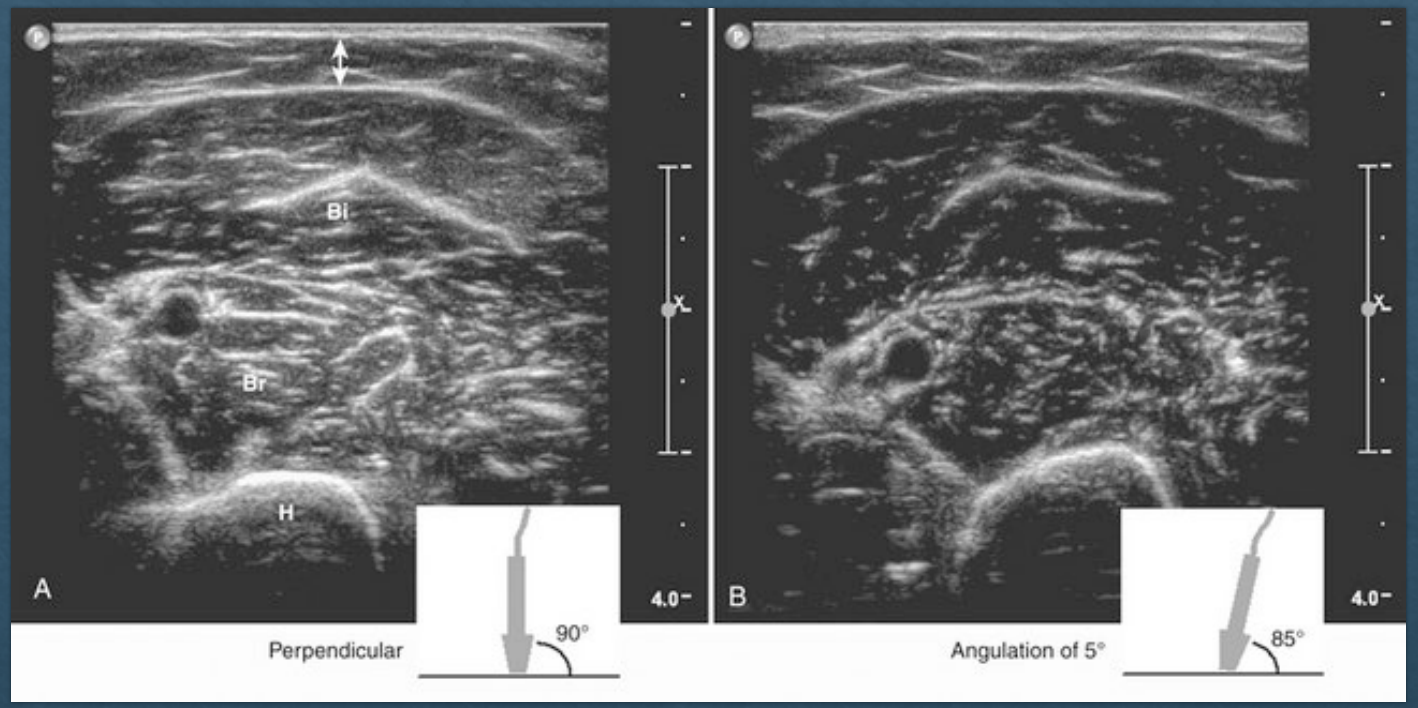
Courtesy of Dr. Barclay McGhee

Anisotropy

- Tendency of tissues to reflect sound in a directionally dependent manner
- Probe perpendicular to structure minimizes anisotropy
- Some tissues exhibit more anisotropy compared to others.

Probe angle

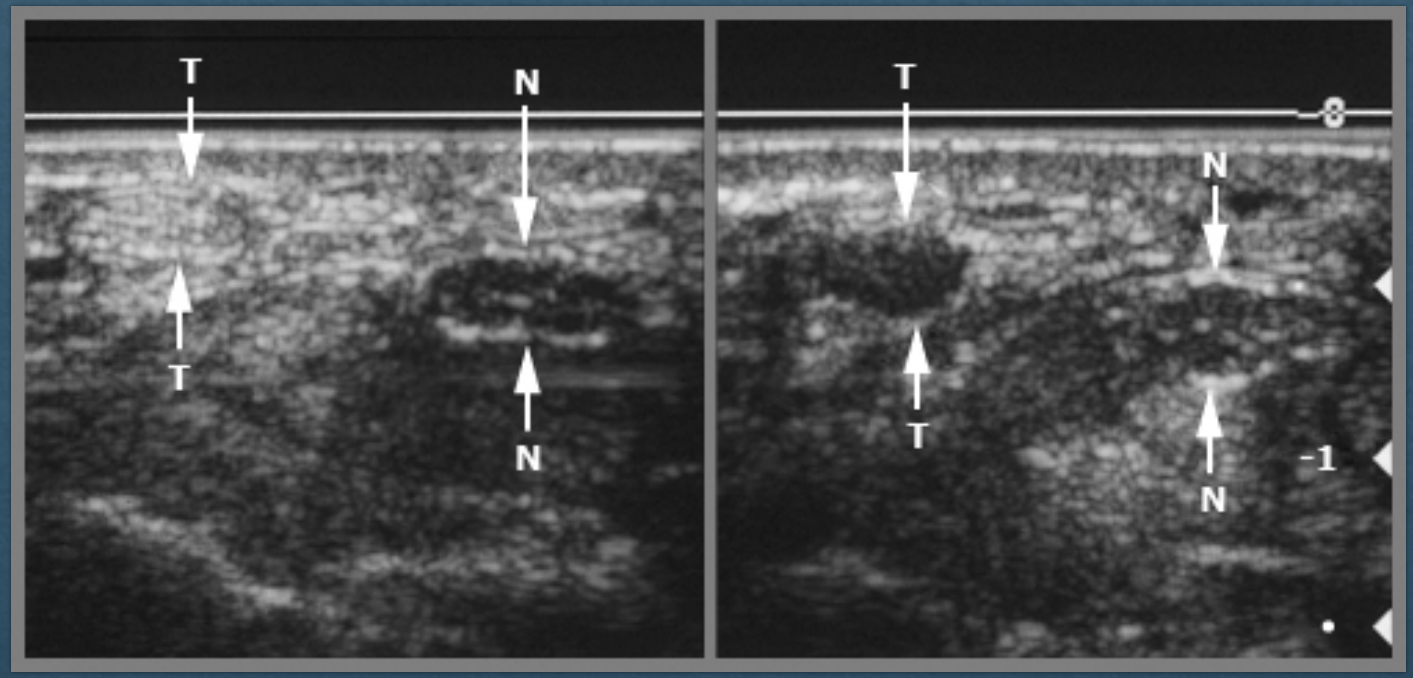
Ultrasound image of the upper arm perpendicular **(A)** and after angulating the transducer 5 degrees **(B)**. After angulation of the transducer, the sound reaching the fascia within and surrounding the muscle as well as the bone gets partially deflected. Because this sound does not return to the transducer, the image is relatively black compared with the image with a perpendicular position of the transducer



Tendon vs. muscle

This is an ultrasound axial image of the wrist at the distal wrist crease. The image on the left is obtained with the probe perpendicular to the flexor carpi radialis tendon (T); note that it is hyperechoic in this image. In the right image, at the same level, the probe is tilted just off the perpendicular axis by aiming the probe slightly proximal. Note that this shift of just a few degrees causes the tendon to become hypoechoic. The median nerve (N) varies less in echogenicity because the tendon exhibits a higher degree of anisotropy. Note that some of the flexor tendons below alis tendon, also show marked variations in echogenicity with the slight shift of probe angle. (The numeral "-1" to the right indicates a depth of 1 cm, and the dot below it represents an additional 0.5 cm.)

Courtesy of Francis O Walker, MD



Common indications

- Entrapment neuropathies
- Nerve injuries
- Demyelinating disorders
- Myopathies
- Motor neuron disease
- Mass lesions
- Guided procedures
- Diaphragm

Advantages

- Complements EMG/NCS
- Non invasive and pain free
- Bedside accessible
- Cost-effective

Normal nerve

- A typical nerve has a honeycomb appearance with a somewhat echogenic external perineurium punctuated by hypoechoic rounded fascicles
- Not freely movable
- Less anisotropy than tendons

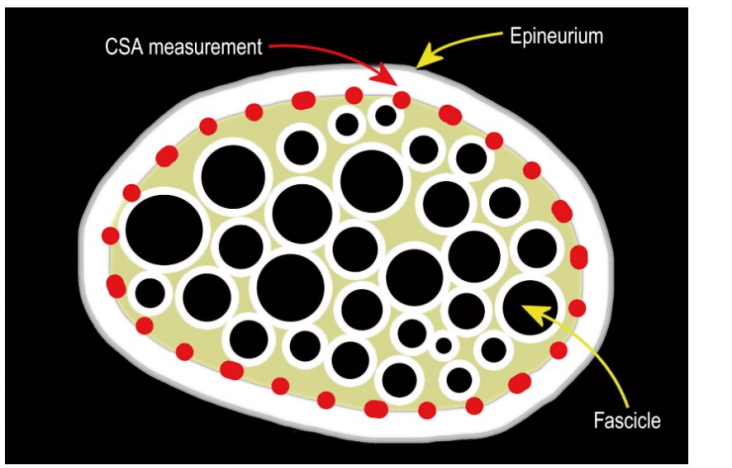
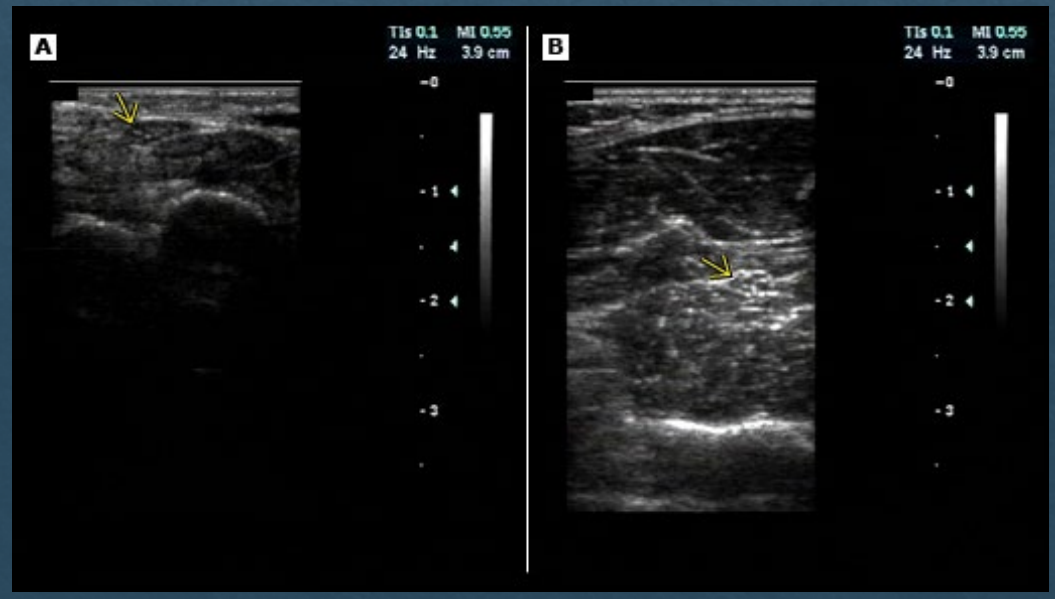


FIG. 18.7

Manual tracing of the cross-sectional area (CSA).

Preston et al.



Entrapment mononeuropathies

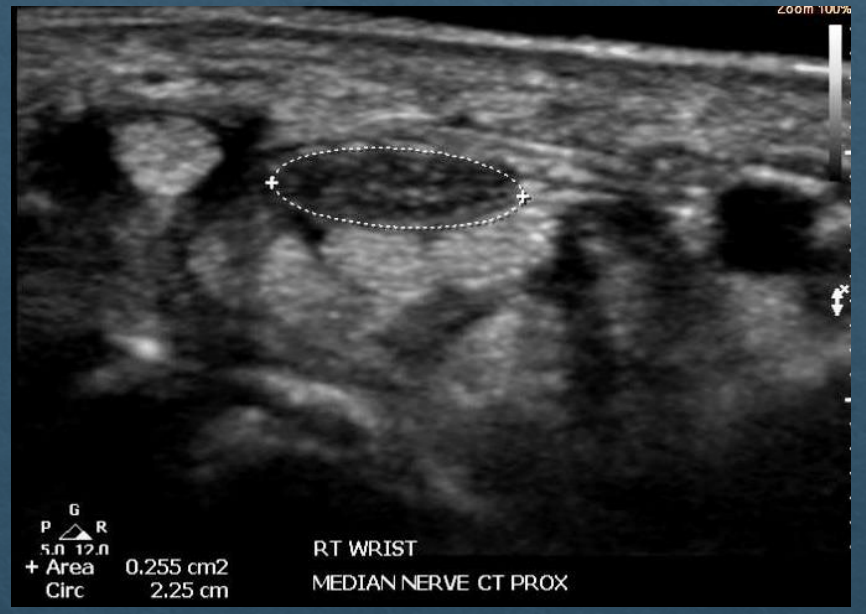
Nerve enlargement proximal to area of compression

Hypoechoic enlarged fascicles.

Structural lesions/anatomical variations compressing the nerve

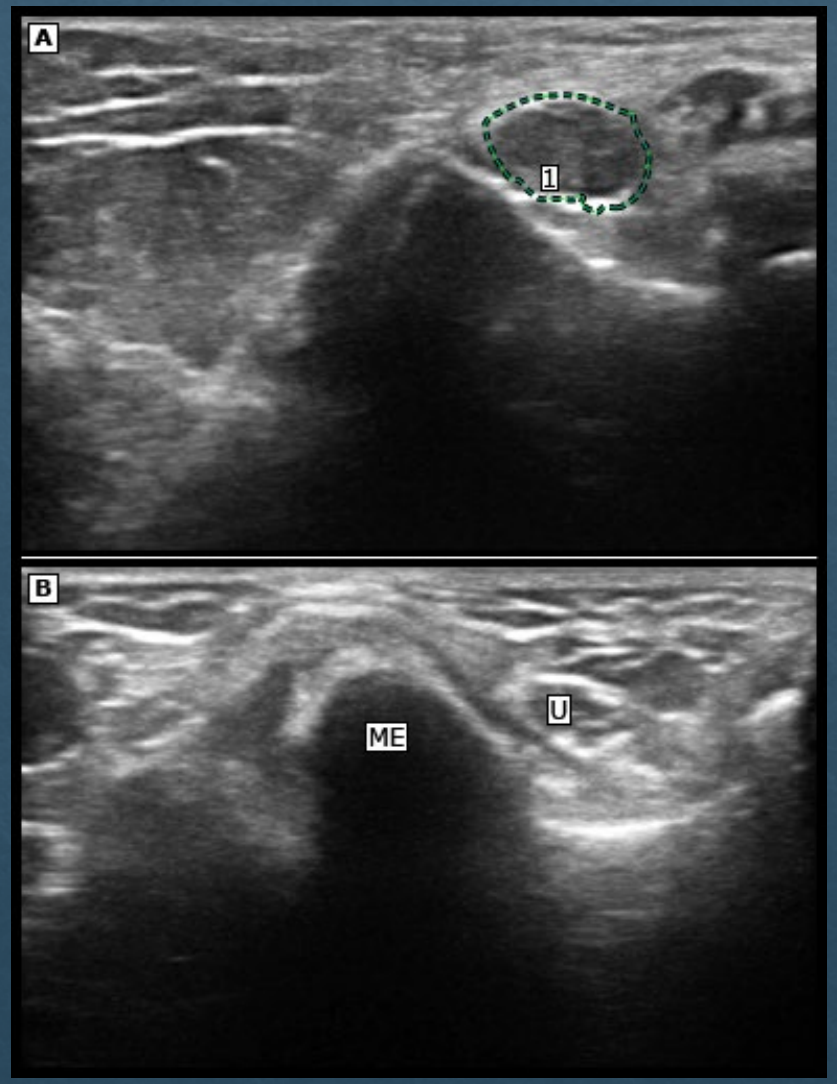
Carpal tunnel syndrome

- Wrist CSA 10-12 mm 2 borderline, >12mm2 abnormal
- WFR (CSA wrist to forearm ratio)
  - Ratio of >1.4 is 100% sensitive
- Flattening ratio >3:1
- Anatomical variation
  - Bifid median nerve, Persistent median artery
- Structural lesions
  - Ganglion cysts, neuromas
- Mobility, bowing of flexor retinaculum



Ulnar neuropathy

- Swelling ratio (CSA ratio at elbow compared to forearm/upper arm) >1.5
- Subluxation out of retrocondylar groove with elbow flexion
- Anatomical variants (accessory epitrochlearis muscle)
- Especially helpful when ulnar neuropathy is not localizable on EMG



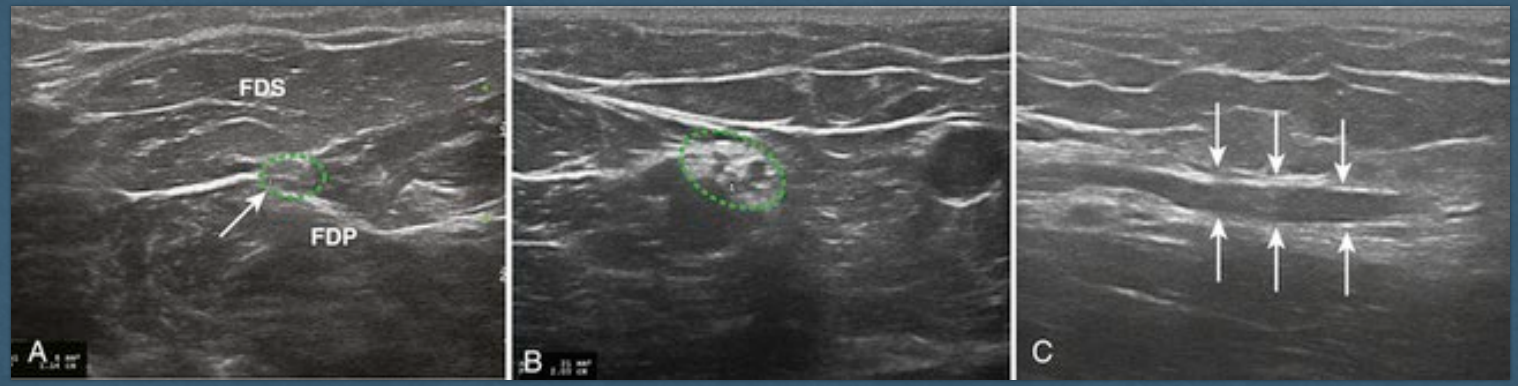
Demyelinating Polyneuropathy

Acquired demyelinating polyneuropathies: multifocal enlargement of nerves

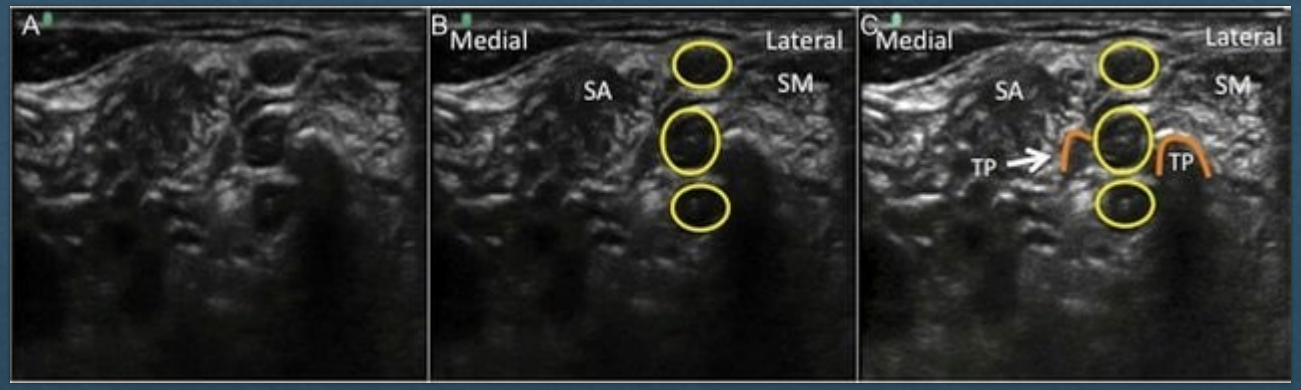
Hereditary disorders show uniform enlargement

Standard protocol involves scanning the median nerves in the forearm and upper arm, and trunks of the brachial plexus

Goedee et al. found that 2 or more enlarged areas is 100% specific for acquired demyelination (CIDP, MMN, LSS)



Median nerve in a patient with chronic inflammatory demyelinating polyneuropathy (CIDP). **A,** Cross-sectional area of the median nerve (*arrow*) 7.5 cm proximal to the distal wrist crease measuring 8 mm^2*FDP,* Flexor digitorum profundus; *FDS,* flexor digitorum superficialis. **B,** Cross-sectional image at 7 cm proximal to the antecubital fossa, measuring 21 mm^2 (*nerve outlined in green*). **C,** Longitudinal image at the same site demonstrating focal enlargement and hypoechogenicity of the nerve (*arrows*).



Interscalene brachial plexus. Ultrasound image illustrating nerve roots [yellow rings in (b)] between scalenus anterior (SA) and scalenus medius (SM). (c) The proximity of these nerve roots to the transverse process (TP)

Situations where US is especially useful

- Most or all of the responses are absent
- The conduction velocity is in the borderline demyelinating range
- The demyelination may be limited to the proximal nerve segments
- CSA of nerves can be used to monitor treatment of CIDP

Other disorders with nerve enlargement

- Diabetes
- Acromegaly
- Leprosy
- Vasculitis
- CMT
- AIDP

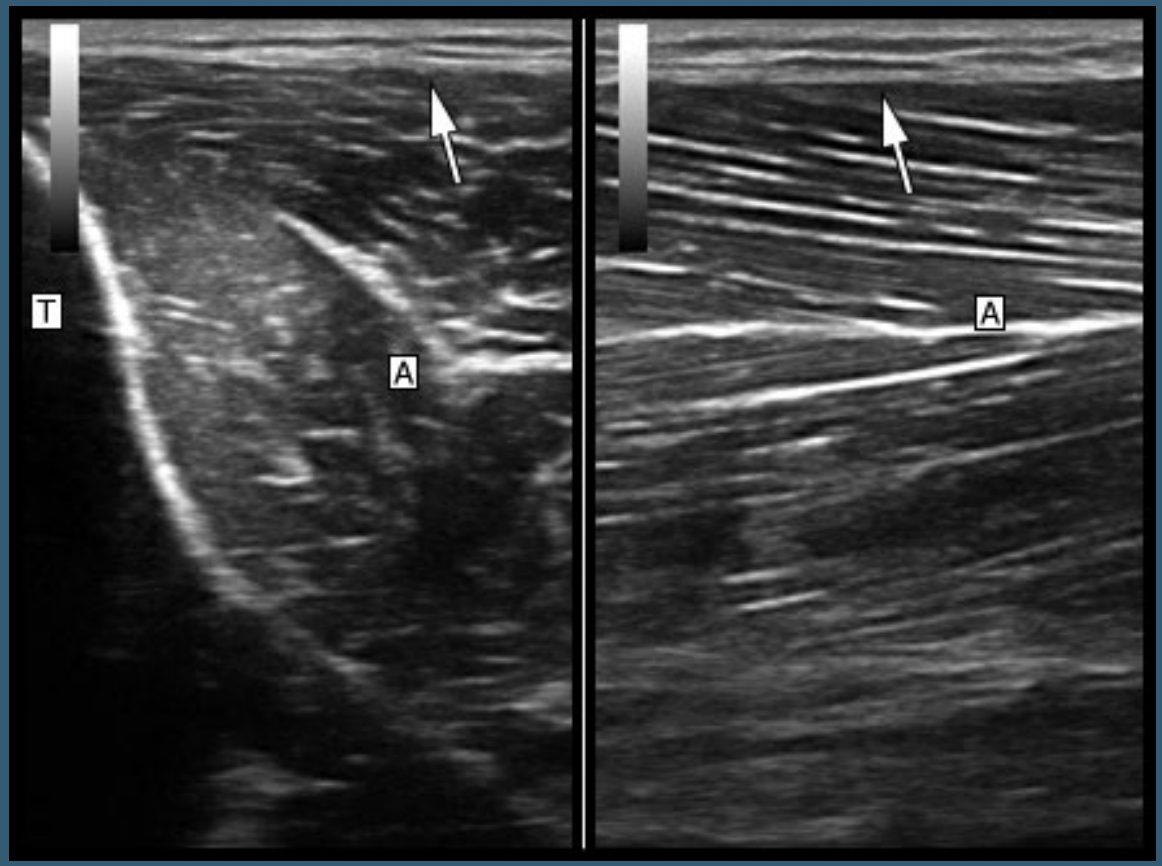
Motor neuron disease

- Fasciculations
  - More sensitive than EMG
  - Can be detected in muscles that are difficulty to relax (genioglossus, rectus abdominis)
  - Fasciculations can be used of evidence of denervation

Normal muscle

In the axial image, muscle consists of primarily echolucent (dark) areas interspersed with small bright curved echoes of seemingly random orientations.

In the sagittal plane, however, these bright echoes are seen to be the fibrous tissue that surrounds muscle fibers and fascicles and which organizes into recognizable striations



These are axial (left) and sagittal (right) images of the tibialis anterior muscle. Note the distinction of the muscle from the thin layer of subcutaneous tissue marked by arrows. The aponeurosis (A) stands out from the rest of the muscle in both images. The sagittal image (right) shows the bipennate attachments of the muscle fibrous tissue, which joins the aponeurosis at a distinct angle of pennation. The bone edge above the tibia (T) is bright and crisply defined

Diseased muscle

- 3 patterns occur with muscle disease
  - Diffuse homogenous hyperechogenicity with attenuation of the ultrasound bearm
  - Diffuse homogenous hyperechogenicity without attenuation of ultrasound beam
  - Patchy hyperechogenic pattern with a “moth-eaten” pattern

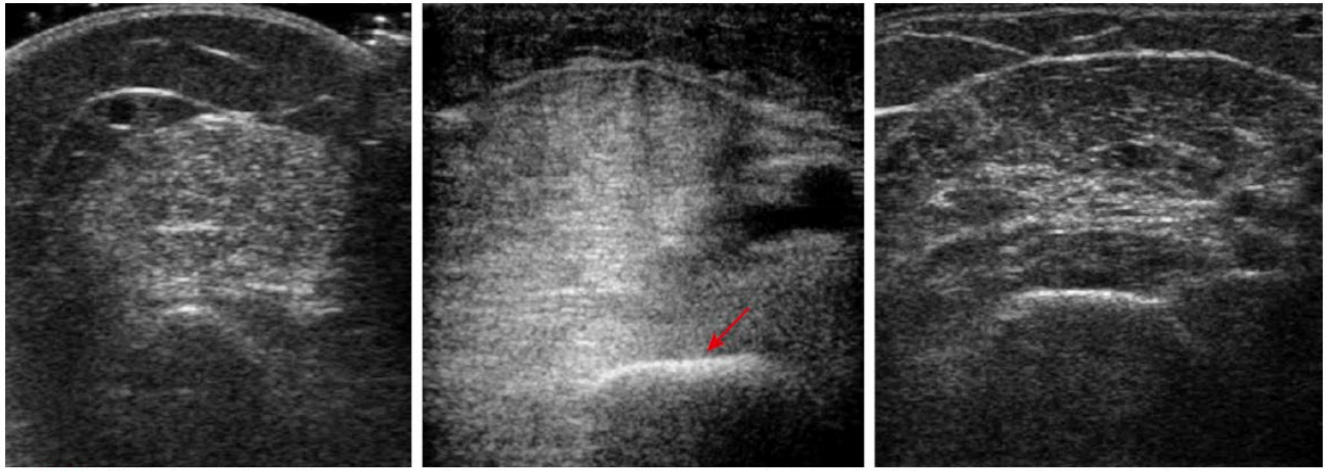
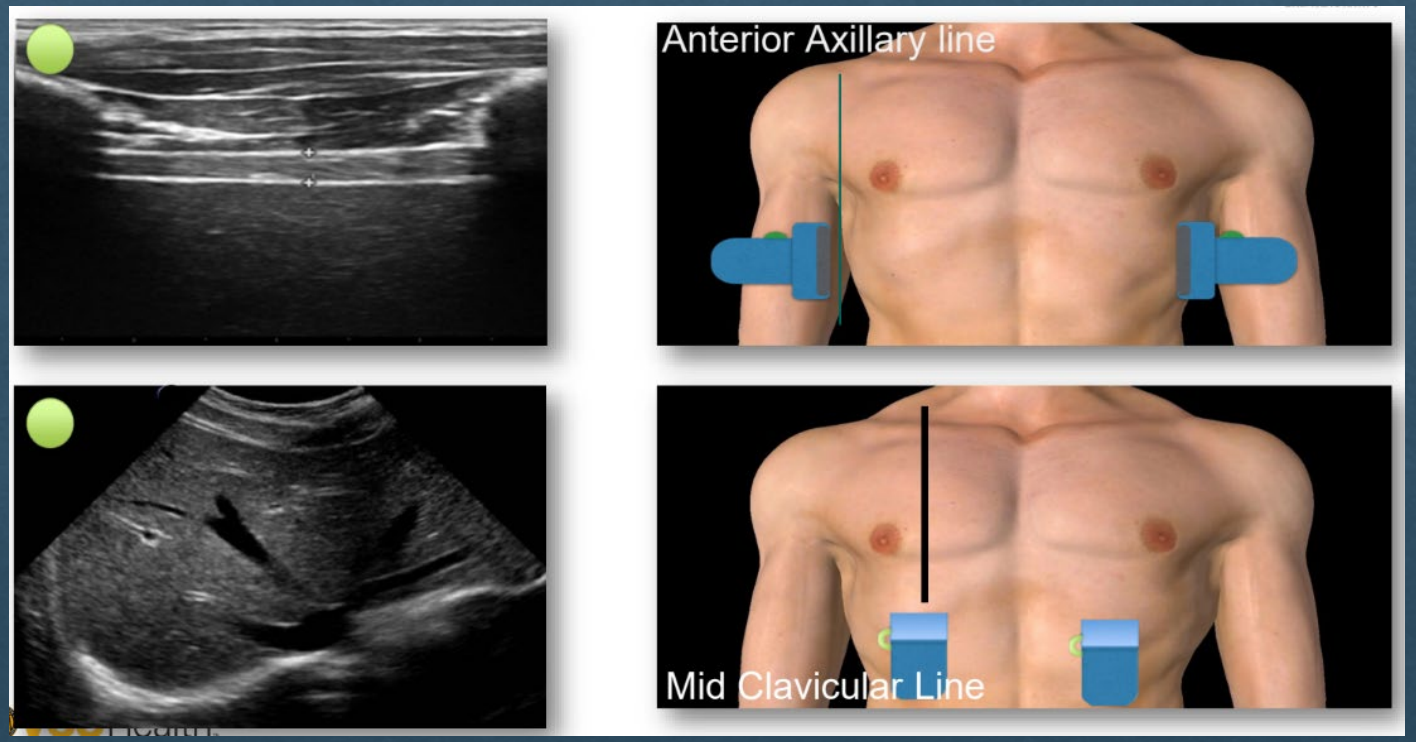


FIG. 38.9 Ultrasound images of myogenic and neurogenic pathology.Transverse views. *Left*, Duchenne muscular dystrophy. *Middle*, Immune myositis. *Right*, Denervation atrophy in amyotrophic lateral sclerosis. In the muscular dystrophy and myositis patients, the muscle has homogenously increased echogenicity. However, the bone shadow is well maintained in myositis patients *(red arrow)*. Note the moth-eaten appearance in denervation atrophy.
From Zaidman CM, vanAlfen N. Ultrasound in the assessment of myopathic disorders. *J Clin Neurophysiol*. 2016;33:103–111.

Heckmatt Scale

- The Heckmatt scale evaluates the severity of neuromuscular disease based on echo intensity and underlying bone definition:
- **Grade 1 (Normal):** Muscle is dark (hypoechoic) with distinct, white (hyperechoic) bone echo.
- **Grade 2 (Mild):** Increased muscle echogenicity (whiter), but bone echo is still distinct.
- **Grade 3 (Moderate):** Strong increase in muscle echo, reducing the definition of the bone echo.
- **Grade 4 (Severe):** Muscle is very white, with complete loss of bone echo and internal structure.

Diaphragm



Essential Anatomy 5

Measurements

- Thickness at the end of inspiration

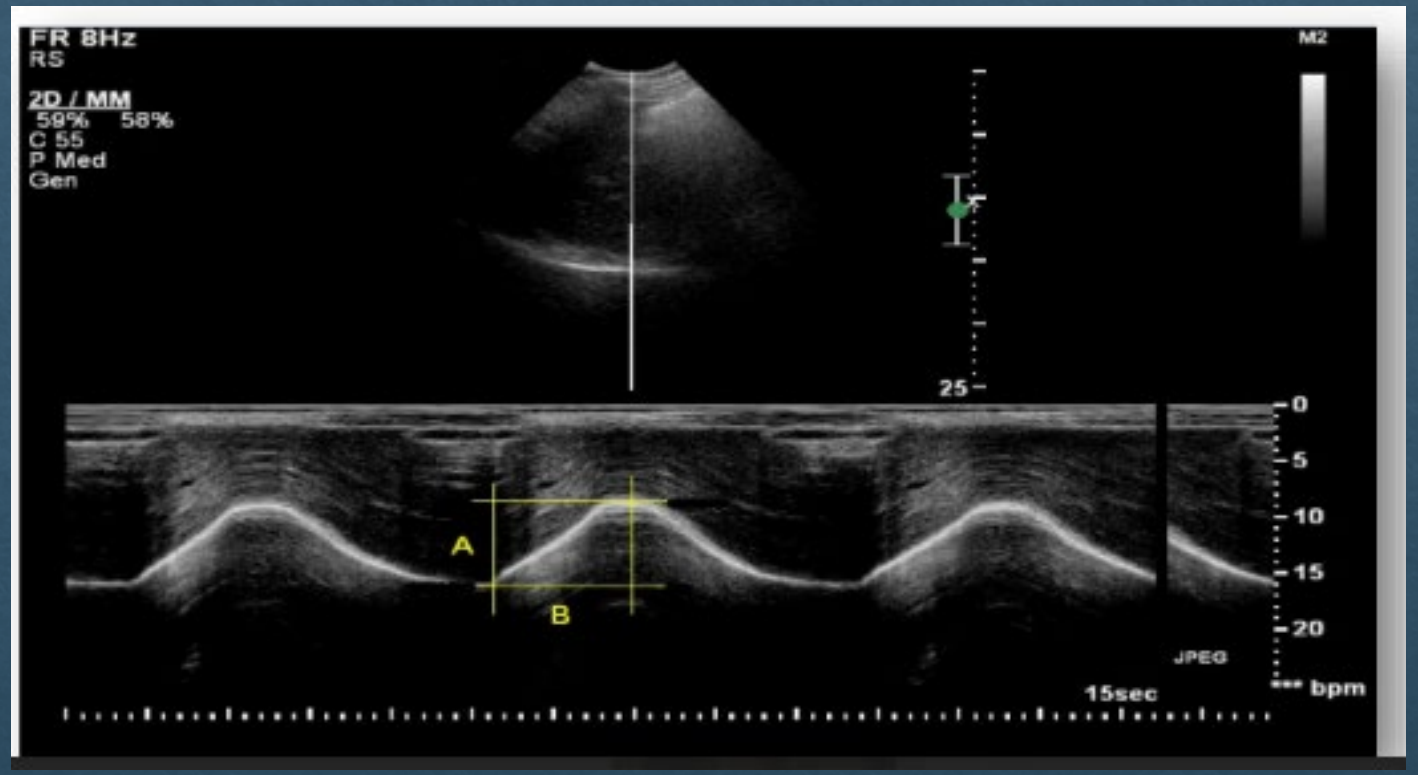
- Thickness at the end of expiration

- Excursion of the dome of diaphragm

Courtesy of Dr. Aarthi Sarwal

M mode excursion

- Excursion >2.5 cm excludes severe diaphragmatic dysfunction
- Normal side to side difference is <50%



Courtesy of Dr. Aarthi Sarwal

- Thickness at end of expiration <20mm is indicative of diaphragm atrophy
- Thickness in normal individuals should increase by 20% during end inspiration
- Any side to side difference in end expiratory thickness >3.3mm is abnormal.
- Measurement of diaphragmatic excursion and end inspiratory thickness requires adequate patient effort.

References

- All ultrasound images taken from Walker et al. unless otherwise specified.
- Walker, Francis, and Michael S Cartwright. *Neuromuscular Ultrasound E-Book*. Elsevier Health Sciences, 6 May 2011.
- Preston, David C. *ELECTROMYOGRAPHY and NEUROMUSCULAR DISORDERS : Clinical-Electrophysiologic-Ultrasound... Correlations.* S.L., Elsevier, 2020.