Neuromuscular Ultrasound in Action

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# Neuromuscular Ultrasound in Action

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*No one involved in the planning of this CME activity had any relevant financial relationships to disclose.*

**Chair:** Francis Walker, MD

The ideas and opinions expressed in this publication are solely those of the specific authors and do not necessarily represent those of the AANEM.
Objectives - Participants will acquire skills to (1) Evaluate the diagnostic yield of combining US of nerve and muscle with electrodiagnosis, (2) recognize common MSK disorders of the wrist and elbow that may be present when EDX studies for extremity symptoms are negative, (3) identify patients referred for EDX studies who would likely have informative US studies and list key elements that would show up in an US report on such individuals, and (4) discuss potential benefits and risks of incorporating NM US into the practice of EDX medicine.

Target Audience:
- Neurologists, physical medicine and rehabilitation and other physicians interested in neuromuscular and electrodiagnostic medicine
- Health care professionals involved in the diagnosis and management of patients with neuromuscular diseases
- Researchers who are actively involved in the neuromuscular and/or electrodiagnostic research

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INTRODUCTION

Ultrasound (US) is a painless, noninvasive technique to identify skeletal muscle and nerve pathology. Its use was pioneered by Heckmatt and Dubowitz in the early 1980s in the evaluation of Duchenne muscular dystrophy (DMD). Since this time, studies of US imaging of skeletal muscle and nerve have expanded to include neuromuscular disorders of various etiologies. US provides a practical and effective supplement to the physical examination in the evaluation of neuromuscular pathology. It can be performed at the bedside and allows for examination of multiple muscles quickly and without reliance on patient participation. This allows for a directed electrophysiologic examination and can be of assistance when selecting sites for biopsy. These qualities make US a practical and informative tool in the evaluation of the infant or child with a suspected neuromuscular disorder.

NORMAL AND ABNORMAL NERVE

Normal nerves appear on US as round or ellipsoid structures with a hypechoic (bright) rim representing the epineurium and multiple, round structures internally that are the nerve fascicles/perineurium. Nerves can be traced along their course and do not show abrupt changes in direction (with the exception of the peroneal nerve at the fibular head) or size. Both proximal nerves, such as the brachial plexus, and distal small nerves, such as the dorsal ulnar cutaneous nerve, can be visualized. In the leg, the sciatic and femoral nerves typically can be seen proximally, and distally the tibial, peroneal, and sural typically are readily identified.

Abnormal nerves show a variety of overlapping pathologies on US. Typically, a nerve will be enlarged. This can be focal, as seen in an entrapment neuropathy. In this case, the nerve will typically enlarge just proximal to the site of entrapment and then can show a “neck” or constriction at the site of entrapment. Focal nerve enlargement also can be seen in peripheral nerve tumors, often with a “tail sign,” or from focal demyelination associated with conduction block in demyelinating neuropathies. More extensive nerve enlargement can be seen from either inherited or acquired demyelinating neuropathies, or more rarely from nerve infiltration or infection (e.g., leprosy, lymphoma). Nerve enlargement from compression or edema often is associated with a hypechoic appearance with loss of the normal fascicular architecture. Nerve trauma also causes nerve enlargement, and it can demonstrate neuroma in continuity or in a partial or complete laceration.

US demonstrates a variety of pathologies associated with nerve disorders that have direct clinical implications. For instance, US of the ulnar nerve at the cubital tunnel not only can localize the nerve lesion but can add anatomic information to guide surgical decisionmaking. In traumatic neuropathies, US can be used to determine the presence, location, and extent of nerve trauma. Focal compressive neuropathies resulting from peripheral nerve tumors or ganglion cysts can be identified and differentiated from idiopathic entrapment neuropathies. Peripheral neuropathies also can result in changes in nerve morphology that can be readily detected using US. The identification of the type, pattern, and location of nerve pathology using US in these instances can guide management and treatment decisions.
NERVE ULTRASOUND IN CARPAL AND CUBITAL TUNNELS

One of the more common utilizations of nerve US is to assist in the localization and diagnosis of entrapment neuropathies. In carpal tunnel syndrome (CTS), the median nerve most commonly becomes enlarged just proximal to the tunnel at the level of the distal wrist crease. This can best be appreciated by scanning the nerve in cross section from the forearm through the carpal tunnel. Cutoff values for normal nerve size vary by laboratory and study, and they are best determined for each individual practitioner.5,6 Other studies have reported an increase in the intraneural blood flow, detected using power Doppler, within the median nerve in CTS.7 Currently, the size of the median nerve at the wrist is the most widely reported measure for identifying CTS and has been most commonly shown to have the greatest sensitivity and specificity for CTS.

Ulnar neuropathy at the elbow (UNE) can be identified on US as an enlargement in the nerve cross sectional area, often with a hypechoic appearance of the nerve, typically just proximal to the site of compression.5,9 In UNE, nerve enlargement can be appreciated most commonly at the level of the medial epicondyle of the humerus but can occur anywhere throughout the ulnar groove or cubital tunnel. Additionally, dynamic visualization with US of the cubital tunnel during elbow flexion and extension readily allows for identification of ulnar nerve subluxation or dislocation, or other conditions such as “snapping” of the triceps tendon over the medial epicondyle of the humerus. Importantly, the presence of ulnar nerve subluxation or dislocation is not in itself diagnostic of a cubital tunnel syndrome, as this condition is similarly present in normal subjects as it is in patients with UNE.10

NERVE ULTRASOUND IN TRAUMATIC AND OTHER FOCAL NEUROPATHIES

US of nerves is a powerful technique to use in addition to the clinical and electrophysiologic examination to aid in localization and diagnosis. Nerve US confirmed or augmented the electrodiagnostic (EDX) examination in half of patients on whom both were performed.11 For traumatic lesions, US can be of great practical value to diagnosis nerve transection. In one study, it modified the approach in 58% of patients.12 Specifically, US can identify the presence or absence of nerve continuity, measure the distance between proximal and distal nerve stumps for surgical planning in cases of discontinuity, and identify mechanical entrapments following fractures.13 The ability to visualize the nerve in high spatial resolution, and in continuity, distinguishes US from magnetic resonance imaging (MRI) and may account for the superior performance of US in a recent retrospective evaluation of mononeuropathies.14

Nerve tumors and cysts also can be readily identified on US. Nerve tumors appear as fusiform (spindle) shaped enlargements that result in focal enlargement of the nerve.15 Neurofibromas typically are intra-axial; they grow within the fascicles within the nerve. Schwannomas typically are extra-axial; they originate from a portion or select fascicle of the nerve and tend to displace the unaffected fascicles. Nerve tumors can have varied echogenicity, including simple, homogenous, isoechoic, or hyperechoic signal, or they may have varying cystic and non-cystic components. Blood flow can be increased within the tumor, but is not a universal finding. Ganglion cysts have a characteristic appearance, with a hyperechoic border and anechoic center, often with posterior acoustic enhancement. Extraneural cysts can directly compress the nerve, while intraneural ganglion cysts infiltrate the nerve and extend along its axis. This is most commonly encountered in the peroneal nerve at the fibular head and should be evaluated in patients with foot drop.16

NERVE ULTRASOUND IN POLYNEUROPATHIES

US frequently can detect abnormalities in nerves of patients with demyelinating polyneuropathies.17 In the most common form of Charcot–Marie–Tooth (CMT) disease (CMT1A), nerves are commonly and often massively enlarged. In other forms of CMT, such as CMTX or CMT2, nerves may be larger than in control subjects but are not as enlarged as in CMT1A.18,19 In acquired demyelinating polyneuropathies, nerves also are often enlarged, although not to the same extent or frequency as in CMT1A. This is most common in chronic inflammatory demyelinating polyneuropathy but also occurs in Guillain-Barré syndrome and multifocal motor neuropathy.17,20 Focal enlargement of nerves at sites of conduction block has also been described.20,22 In diffuse peripheral axonal neuropathies, nerves typically are normal sized.

ULTRASOUND OF NORMAL AND ABNORMAL MUSCLE

US also can be used to detect the presence and pattern of muscle abnormalities in both myopathic and neuropathic disorders. Patterns of muscle involvement in myopathies can be used to guide biopsy site selection and to narrow the differential diagnosis, and it has potential use for monitoring disease progression.

Normal muscle shows low echogenicity (mostly dark) on US. Interspersed within this low-signal are multiple, homogeneously distributed, well-defined brighter punctate or curvilinear bright areas. These represent the fibroadipose septa and tendinous fibrils interspersed among the muscle fibers. The myofascial fibrils coalesce near the myotendinous junction. At these areas, the echo intensity is increased and there is higher anisotropy. For diagnostic purposes, it is best to avoid these areas and focus on the bulk of the muscle belly. The fascia around the muscle belly is brighter and thicker than the fibrous tissue in the muscle belly. At high magnification, multiple low-signal, dark honeycombed structures surrounded by a thin, medium intensity ribbon can be seen. These structures appear like individual muscle fibers; however, US does not have sufficient resolution to visualize individual muscle fibers. Rather, these areas likely are bundles of muscle fibers surrounded by brighter fibroadipose tissue. Bone is very bright (highly echogenic) with a well-defined, crisp edge and casts a shadow on US deep the this boney reflection. Subcutaneous fat has similar echo intensity to muscle and is interspersed with poorly organized threads of brighter connective tissue.

The appearance of skeletal muscle on US changes with age. In infants, muscle is more echo-dark than in older children. At this age, there are few myofascial planes in the muscle parenchyma. By...
age 2-3 years, there are more myofascial planes than in an infant. By age 5, myofascial planes are seen in a homogenous pattern typical of the adult. After age 5, muscle echogenicity increases more gradually, if at all, through most of early adulthood. In later life, at about age 60 years, muscle echogenicity increases more rapidly with advancing age. These changes vary with muscle group and are most pronounced in the biceps brachii and quadriceps muscles, particularly in males. There is no difference in echogenicity between males and females until the teenage years. From this age forward, muscle in males is slightly darker on US than in females.

Muscle bulk also changes throughout the lifespan and varies with age, gender, and muscle group. In the first 20 years of life, muscle thickness increases. As with muscle signal intensity, differences in gender begin in the early teenage years, with males having larger muscles than females. Until age 40 years, muscle thickness remains relatively stable in both genders. In older adults, muscle thickness in some muscles declines considerably. Muscle thickness decreases in the quadriceps by 30% in women and 50% in men between ages 40 to 90. In the biceps brachii, similar but less severe declines (20-30%) are seen. In contrast, muscle thickness in adults in the sternocleidomastoid, tibialis anterior, and, in women, the forearm flexors remains more stable through the lifespan.

Muscle atrophy also can be assessed qualitatively by comparison of the thickness of muscle to subcutaneous fat. The typical ratio of muscle to subcutaneous fat is approximately 2:1. While using this ratio is helpful as a quick screening tool, the ratio can be misleading in obese patients and in infants, as the thickness of the subcutaneous fat changes rapidly through the first year of life. Thus, assessment of muscle thickness must be interpreted using norms adjusted for patient characteristics, including age and body habitus.

MUSCLE ULTRASOUND AS A DIAGNOSTIC TOOL

Myogenic weakness typically appears as a homogenous increase in echo signal intensity with relatively preserved muscle bulk. In contrast, US in a patient with neurogenic weakness shows reduced muscle size with an increase in the subcutaneous fat to muscle ratio and a pattern of streaky, increased echoes scattered heterogeneously within the muscle. Central hypotonia does not cause substantial alterations in the muscle signal intensity on US, although in the author’s experience disuse can reduce muscle size. Using these criteria, US can play a role as a well-tolerated screening test and can be performed at the bedside without need for sedation or patient discomfort.

The sensitivity and specificity of diagnostic US for detecting neuromuscular disorders has been best studied in children and depends on the type and severity of disorder (reviewed by Pillen and colleagues). In the evaluation of infants, in whom electrophysiologic evaluation can be technically challenging, US has similar sensitivities and specificities to needle EMG for identifying and characterizing neuromuscular pathologies. In a study of 41 hypotonic infants aged 2-24 months, qualitative US was highly concordant with needle EMG.31 US and needle EMG both showed abnormalities consistent with myopathy in 6 of 6 infants with myogenic hypotonia and both showed neuropathic abnormalities in 16 of 16 infants with neurogenic hypotonia. In 17 infants with central hypotonia, both US and needle EMG were normal. Needle EMG and US results conflicted in only two infants. For these patients, the muscle and nerve biopsy ultimately did not reveal a diagnosis.

In older children, qualitative US also is sensitive and specific for detecting neuromuscular disease. In a study of 134 patients with suspected neuromuscular disorders, qualitative US showed a sensitivity of 81% and a specificity of 96% in the assessment of any neuromuscular disorder. US was less sensitive (71%) in identifying abnormalities in children with neuromuscular disorders under age 3.

In a study of 100 children with suspected neuromuscular disease, US was 78% sensitive and 91% specific for identifying any neuromuscular disease, was more reliable in children over age 3 years, and was least reliable in those under 1 year of age. Sensitivity and specificity varied with the degree of US abnormality. A mildly abnormal US is neither sensitive nor specific for a neuromuscular disorder. Only 7 of 13 children with a mildly abnormal US scan had a neuromuscular disorder. In contrast, all of the children with moderate or severely abnormal USs (Heckmatt Grade III or IV) had a neuromuscular disorder. Similarly, nearly all (62 of 69) children with a normal US (Heckmatt Grade I) did not have a neuromuscular disorder. Quantitative US results in similar sensitivities for detecting neuromuscular disorders. A prospective study of quantitative grey-scale US analysis of 150 children referred for evaluation for neuromuscular disorders was 71% sensitive and 91% specific for identifying neuromuscular disorders. Again, the sensitivity of US in children younger than 3 years old was lower than in older children; however, specificity was 100%, with no false-positives in the younger age group.

US has higher specificity than sensitivity in differentiating myopathic and neuropathic changes. Thus, abnormalities that distinguish the two pathologies such as muscle size and patterns of homogeneity are helpful when present but are less useful as a screening tool. US was more specific than sensitive for detecting myogenic (92 versus 67%) and neurogenic (98 versus 77%) changes in 134 children studied by Brockmann and colleagues. The pattern of muscle involvement also can help identify neuropathies, which affect the distal more than proximal muscles of the legs. A quantitative US study of 31 children with myopathic and 27 with neuropathic disorders, brighter echoes and more atrophy in the legs than arms was 67% sensitive and 94% specific for identifying neurogenic disease. This type of analysis did not distinguish myopathic disease from non-neuromuscular conditions. In adults, a study comparing 145 healthy control subjects with 17 myopathic and 15 neuropathic patients, brighter signal in the biceps brachii (increased grey-scale values) was 94% sensitive and 93% specific for myopathy while increased signal inhomogeneity was 100% sensitive and 93% specific for neuropathy. However, in children, this same quantitative approach did not distinguish between myopathic and neuropathic disease.

The relationship between strength, function, and the degree of image abnormality may vary with differences in the underlying pathologies. In patients with muscular dystrophies, subclinical abnormalities and changes with disease progression have been detected on MRI
and US, suggesting that imaging can be used to evaluate disease severity and progression. In contrast, the severity of imaging findings in children with mitochondrial myopathies or congenital muscular dystrophies may not correlate with disease severity or function. Additional studies of the neuromuscular pathologies are needed comparing patient strength, function, and prognosis with the degree of image abnormality.

**ULTRASOUND ABNORMALITIES IN INHERITED MYOPATHIES**

Hereditary myopathic disorders are a diverse group of pathologies that include the muscular dystrophies and the metabolic, mitochondrial, and congenital myopathies. They generally present with progressive, symmetric weakness more than atrophy of the proximal arms and legs and are classified by the pattern of inheritance, clinical findings, histopathology, and genetic abnormality. Muscular dystrophies are the most common type of myopathy and are characterized by findings on muscle biopsy of early and extensive muscle fiber degeneration and regeneration. Later in the disease, prominent increased connective tissue and fatty replacement of muscle fibers are seen. The muscular dystrophies include DMD and Becker muscular dystrophy (BMD), as well as fascioscapulohumeral, Emery-Dreifuss (EDMD), limb-girdle (LGMD), and congenital muscular dystrophies.

**ULTRASOUND OF MUSCULAR DYSTROPHIES**

Abnormal ultrasonography in neuromuscular disease was first described in males with DMD, an X-linked muscular dystrophy caused by mutations of the dystrophin gene. Much of the ongoing work in US of myopathies continues to be performed in patients with D/BMD, a less severe form of the disease. US in D/BMD, as well as other muscular dystrophies characterized by increased connective tissue and fatty replacement of muscle, shows a diffuse and often marked increase in muscle echogenicity. The muscle shows a granary, ground-glass like appearance, typically with preserved muscle bulk. In more advanced pathology, the muscle echoes are very bright and the attenuation greatly increased, resulting in a relatively darker appearance in the deep rather than superficial portion of the muscle and reduced or absent bone echoes.

The sensitivity of US to pathology associated with muscular dystrophy increases with age and disease severity. In DMD, US often is abnormal in children by the time they are toddlers. In one study, qualitative US was abnormal in nearly all (21 of 22) boys with DMD aged 3-7 years but in none of the 7 boys aged 2-30 months. In a study of quantitative US, abnormalities were detected in 32 of 38 boys aged 1 to 11 years; all six normal USs were in children under age 6 years. In another quantitative US study, grey-scale levels were abnormally high in 10 of 11 boys with DMD ages 3-9 years but in only 1 of 2 boys aged 3 weeks and 7 months.

In D/BMD, US measurement of muscle pathology varies with the severity of muscle pathology. Both US and strength abnormalities are more severe in the quadriceps than the biceps brachii. US backscatter is higher and increases twice as much with age in patients with DMD than those less severely affected with BMD. US signal abnormalities also increase with worsening strength and function in DMD and are more or similarly sensitive to changes in pathology over time as compared to functional measures of disease progression. Additional studies in the dystrophinopathies are needed to determine the sensitivity of US to detect effects of treatment.

Calf enlargement is a common clinical finding in patients with dystrophinopathies and other neuromuscular disorders. Calf enlargement can be either associated with normal or increased echogenicity and is a common and nonspecific finding in neuromuscular disorders. Interestingly, when there is severe fatty infiltration in the calf muscle, the US actually appears dark, similar to subcutaneous fat. As muscle pathology from increased fat typically results in brighter US echoes, the US appearance of severe pathology in the calf may be misleading.

**ULTRASOUND OF MITOCHONDRIAL MYOPATHIES**

Mitochondrial myopathies are a heterogeneous group of neuromuscular disorders that can affect multiple organ systems. Symptoms are related to dysfunction of energy metabolism and include weakness and exercise intolerance. Skeletal muscle US in mitochondrial myopathies can be abnormal but is less sensitive than in other myopathies. In 14 children with mitochondrial myopathies, US assessment was concordant with histlogic findings in only 8 patients, including 1 myogenic, 2 neurogenic, 4 nonspecific, and 1 normal pattern on both US and histology. In a prospective study of quantitative US in 53 children with suspected mitochondrial disorders, only 7 of 28 children with definite or probable mitochondrial disorders had abnormal US echogenicity. An additional 6 children had only borderline US abnormalities. Echo intensity in the 28 subjects with mitochondrial disorders did not correlate with strength or the percentage of intramuscular fat or connective tissue, but it did increase with age. Six of the 8 children with abnormal USs were over 5 years old. A normal US thus cannot exclude a mitochondrial disorder but does detect pathology that, when found, is independent, and complimentary, to functional and histologic results.

**OTHER CONGENITAL AND HEREDITARY MYOPATHIES**

The congenital and hereditary myopathies are comprised of a large group of heterogeneous disorders and phenotypes. Radiologic studies of specific congenital or hereditary myopathies often are limited to small case series. Although relatively few radiologic studies of specific congenital or hereditary myopathies have been reported, US and MRI can detect the presence and pattern of skeletal muscle pathology in patients with these disorders (reviewed by Pillen and colleagues and Mercuri and colleagues). Certain patterns of muscle involvement can direct focused genetic testing or guide selection of a muscle for biopsy. However, as with many neuromuscular disorders, the patterns and degree of muscle involvement on US varies within a genotype and with disease severity. The heterogeneity in phenotype and the small number of reported cases makes it difficult to determine the specificity of a pattern of muscle involvement on US or MRI. For instance, selective involvement of the medial gastrocnemius, with sparing of the lateral gastrocnemius, has been described in several inherited myopathies including EDMD/LGMD1B (Lamin A/C), LGMD1C.
Nonetheless, a few case reports in hereditary myopathy describe unique, specific radiologic patterns. Imaging of Bethlem and the more severe Ullrich congenital muscular dystrophies, both collagen type VI disorders characterized by proximal weakness and contractures, show a unique “outside-in” pattern of muscle involvement. These studies show relative sparing of the central portion of the muscle belly with involvement of the outer rim of the muscle in a concentric pattern. This pattern is best described in the rectus femoris of patients with Bethlem myopathy, and it is termed the “central shadow” sign. This “shadow” seen on US refers not to darkening but rather increased echogenicity and thickening along the central fascia—a normally thin, bright, band that vertically divides the rectus femoris from the superficial fascia to the middle of the muscle belly. Similar but more severe findings to are seen in Ullrich myopathy. In a study of nine patients with Ullrich myopathy, the involved concentric rim with central sparing was best seen in the vastus lateralis and was present but less distinct in the rectus femoris. Increased signal also is seen in the connective tissues between the soleus and gastrocnemius in both Bethlem and Ullrich myopathies. This finding was more distinct in Bethlem myopathy. In contrast, Emery-Dreifuss muscular dystrophy, which shares some clinical features with collagen VI disorders, shows more diffuse thigh and selective medial gastrocnemius involvement and does not show the “outside-in” pattern or central shadow sign. Another unique pattern of pathology on US has been reported in six patients with hereditary inclusion body myositis with homozygous GNE mutations. These patients showed selective involvement of the rectus femoris with relatively spared vastus medialis, lateralis, and intermedius muscles. Additionally, these patients showed areas of increased echogenicity within the central portion of the hamstring muscles, producing a “target” like brightness inside a rim of relatively spared, hypoechoic muscle. This study also showed more severe atrophy of the anterior foreleg compared to the calf and more severe involvement of the hamstring muscles compared to the anterior thigh.

Patients with acid maltase deficiency (Pompe disease) have a specific pattern of muscle involvement that can be detected using US. This pattern includes sparing of the superficial but not deep portion of the biceps brachii, differential involvement of the biceps brachii but sparing of the triceps brachii, and differential involvement of the vastus intermedius more than the rectus femoris. The degree of increased echogenicity in the muscle of patient with acid maltase deficiency correlated with strength. One distinguishing feature of this study in patients with acid maltase deficiency was that findings were compared to patients with other myopathies. This demonstrates the potential for imaging studies in neuromuscular disease to identify patterns of muscle involvement that are specific and could direct further testing. Additional radiologic imaging studies are needed to describe more myopathies with specific patterns of muscle involvement.

US is particularly useful in the identification of fasciculations, which are more frequent and widespread in patients with neuromuscular disorders. US has been shown to detect fasciculations with higher sensitivity than the clinical examination or electromyography. Fasciculations are rapid (0.2-0.5 s) contractions of focal areas of muscle that deform the surrounding areas and occur at random intervals and areas. Occasional fasciculations (≥2/10 s observation) are detectable on US in as many as 8-43% of healthy subjects, are more common in older patients, but rarely occur in muscles proximal to the knee or in the arm. In contrast, fasciculations were very common (85 of 92 [92%]) in subjects with spinal muscular atrophy (SMA), CMT, or motor lumbar radiculopathies and in 24 of 25 (96%) subjects with early amyotrophic lateral sclerosis (ALS). Most patients with ALS in this study had on average more than 4 fasciculations/10 s. The presence of fasciculations can facilitate the diagnosis of ALS and is augmented by incorporating US into the evaluation. In the author’s practice, he screens for fasciculations for 10 s in each imaged muscle and comments on their presence if they are frequent (>2/10 s), in multiple muscles (especially in muscles above the knee as this is uncommon in normal subjects), or if they occur in the presence of other abnormalities.

Two studies report the detection of fibrillations using US as small amplitude muscle movements. Fibrillations appear as small amplitude, irregularly recurring movements within the muscle without other movement of the surrounding tissues or muscle. Detection of fibrillations requires a high frame-rate. This can be easily achieved by using the zoom function. Detection of fibrillations using US did not reach high levels of sensitivity or specificity. This is due likely to several pitfalls that can mimic the small amplitude movements seen from fibrillations. These include movement of tissue due to nearby arterial blood or artifact usually encountered along the lateral edge of the image or at a tissue-to-bone interface. Thus, the appearance of small amplitude movements on US should be interpreted in the appropriate clinical and EDX context.

Other abnormalities that can be detected in motor neuron disease using US include abnormalities in muscle echogenicity and size. In motor neuron disease, muscle bulk is decreased and muscle echogenicity increases in some areas of the muscle while other areas remain normal appearing. In ALS, the rate of increase in muscle echogenicity over time is an independent predictor of survival. In children with SMA, muscle echogenicity is highest in children with the most severe weakness. These studies demonstrate the potential for quantitative muscle measurements using US to inform prognosis and assess in disease progression and severity.
REFERENCES


What to Do When the Needle EMG is Negative?  
An Argument for the Reasonable Use of Musculoskeletal Ultrasonography in the Neurodiagnostic Laboratory

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HISTORY

Ultrasound (US) of peripheral muscle and nerve was first described in the 1980s\(^1,2\) and now is being promoted as a valid and reliable tool to be employed in the neuromuscular laboratory.\(^3\) The first reported use of musculoskeletal US was by the Austrian neurologist Karl Dussik (1958) and his colleagues\(^4\) who measured the acoustic attenuation of articular and periarticular tissues including skin, adipose tissue, muscle, tendon, articular capsule, cartilage, and bone. Subsequently, musculoskeletal US was used to differentiate a Baker’s cyst from deep vein thrombosis,\(^5\) and since that time it has been used to investigate a wide range of soft tissue pathologies.\(^6\)

The first demonstration of synovitis in rheumatoid arthritis was performed in 1978 by the Canadian radiologist Peter Cooperberg\(^7\) using a 5 MHz probe, and he correlated grey-scale images of synovial thickening and joint effusion in the knee with clinical and arthrographic findings before and after treatment with an yttrium-90 injection.

In 1994, the first paper demonstrating the ability of power Doppler to visualize abnormal joint vascularity\(^8\) led the way for rheumatologists to confirm a primary diagnosis and assess responses to often expensive new therapies. With improved technology there is now the ability to scan in grey-scale but also use color/power Doppler settings\(^9\) to illustrate low pressure blood flow in soft tissues that serves as a marker of local inflammation, disease activity, and probable disease progression. The validity of Doppler US has been demonstrated\(^10-12\) and now it is increasingly used as an essential tool in the assessment of synovial perfusion in rheumatoid arthritis\(^13\) and other inflammatory conditions. The appropriate tools, settings, image acquisition and grading scales, and artifacts will be demonstrated and discussed.

The American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) believes that those proficient in neuromuscular US should also be familiar with musculoskeletal US of common disorders of joints and tendons, as they may frequently be an associated or primary disorder that triggers the electrodiagnostic (EDX) study.\(^14\)

The American College of Rheumatology (ACR) Report on the Reasonable Use of Musculoskeletal Ultrasonography in Rheumatology Clinical Practice highlights 14 validated indications to employ US for the diagnosis and management of rheumatic and musculoskeletal disorders.\(^15\) Point #11 states:

“For a patient with regional neuropathic pain without a definite diagnosis on examination, it is reasonable to use [musculoskeletal US] to diagnose entrapment of the median nerve in the carpal tunnel, the ulnar nerve at the cubital tunnel, and the posterior tibial nerve in the tarsal tunnel.”

Rheumatologists are now encouraged—in the absence of rheumatic conditions—to search for and diagnose entrapment syndromes using US.

The subject of this discussion is to explore—in the absence of a defined neurologic diagnosis by needle electromyography (EMG)/nerve conduction study—what added value can the electrophysiologist/sonographer expect from the utilization of musculoskeletal US, with specific attention to the wrist and elbow sites.
PRINCIPLES

The same principles for image acquisition in standard US apply when studying musculoskeletal structures:

- Ensure appropriate and comfortable positioning of the patient.
- Employ high frequency linear transducers (which are best suited for the majority of applications in musculoskeletal US) that are adjusted for gain, depth, and focal point(s).
- Scan in two planes, using the contralateral side for comparison, and employ dynamic scanning.
- Review the gross anatomy and become familiar with the sonomorphology to differentiate soft tissue structures through their shape, echogenicity, texture, and location. This can be of enormous value.

The common pathologies detected by musculoskeletal US in regional pain syndromes include: effusions, synovitis, tenosynovitis, enthesitis, erosions, and miscellaneous lesions (e.g., ganglia). In addition (not discussed here), rheumatologists are afforded the ability to: detect early arthritis (preclinical), engage in sonographic-guided procedures, and monitor the response to disease-modifying antirheumatic drugs (DMARDs) and biologic therapy.

STANDARDIZATION, CONSENSUS, AND DEFINITIONS

As US was being increasingly employed, several problems were identified:

- The lack of standardization for indications.
- The lack of consensus on scanning methodology.
- The lack of clear definitions of pathology.

This lead to an international collaboration resulting in the formation of an Outcome Measures in Rheumatology (OMERACT) US special interest group. The European League Against Rheumatism (EULAR) Working Group for Musculoskeletal Ultrasound published guidelines for musculoskeletal US in rheumatology, which include descriptions of standardized image acquisitions and illustrations of standard scans at multiple sites.


The recognition of common pathological entities require a clear definition of both normal and sonographic features of pathology:

- Synovial fluid is an abnormal echo (i.e., hypo-, an-, or isoechoic) of intra-articular material that is displaceable and compressible, but does not exhibit a Doppler signal.
- Synovial hypertrophy is an abnormal echo (maybe iso- or hyperechoic, relative to subdermal fat) of intra-articular tissue that is nondisplaceable, poorly compressible, and may exhibit a Doppler signal.
- Bony erosion is an intra-articular discontinuity of the bone surface that is visible in two perpendicular planes.
- Tenosynovitis is a hypo- or anechoic thickened tissue with or without fluid within the tendon sheath. This is seen in two perpendicular planes and may exhibit a Doppler signal.
- Enthesopathy is an abnormally hypoechoic or thickened tendon or ligament (loss of fibrillar architecture at its bony attachment) seen in two perpendicular planes, may exhibit calcification, enthesophytes, erosions, and calcifications.

Once recognized how can these pathologies be scored for severity rather than just presence or absence and does it make a difference?

The OMERACT Ultrasound Task Force is working towards a standardized US scoring system for synovitis and other features. Currently, a popular scoring method is semiquantitative, with scores from ranging from 0 to 3, representing mild, moderate, and severe pathology. This method can be applied to effusions, synovial thickening, and power Doppler scoring. These scores are easy to apply in practice, but it must be remembered that differences between grades are not equal. The feasibility of multiple joint assessments has lead to the exploration of composite indices using a reduced number of sites such as a 726- or 1227-joint scoring system.

Power Doppler has been preferred over color Doppler for the assessment of disease activity because of certain technical advantages: less angle dependency, greater sensitivity for low flow states, and lack of aliasing. But, it remains very dependent on the use of appropriate settings and, in particular, probe pressure as applied by the examiner, as excessive pressure will compress low pressure and low velocity blood flow resulting in false-negative results.

Tendon sheath inflammation is common in inflammatory diseases like rheumatoid arthritis and systemic lupus erythematosus. US is uniquely sensitive in detecting these conditions, and it can be useful for diagnosis and prognosis (e.g., extensor carpi ulnaris tendinopathy predicts erosive progression in rheumatoid arthritis).

A more common condition seen in the EMG suite is first extensor compartment tenosynovitis (i.e., involving the abductor pollicis longus and extensor pollicis brevis tendons), the second most common entrapment being tendinopathy (also known at De Quervain’s tenosynovitis). US features include tendon thickening and peritendinous edema appearing as a halo. Detection of an intratendinous septum may be an indication for surgical release if unresponsive to local therapy.

Flexor tendon tendinopathy—the most common disorder affecting tendons and responsible for so-called trigger finger, snapping finger, or stenosing flexor tenosynovitis—is commonly associated with diabetes and carpal tunnel syndrome. The
pathology is more often found in the A1 pulley that undergoes thickening, hypervascularity, and fibrocartilagenous changes.31,32

Lateral epicondylitis of the elbow, the most common cause of lateral elbow pain, can be due to repetitive traction on the osteotendinous attachment of the common forearm extensors. There are characteristic but varied US changes that assist in validating the diagnosis and therapeutic decisions. These can include:

- Tendon changes, such as a loss of fibrillar pattern, focal thickening or swelling, and tears or vertical splits
- Spurring and cortical irregularities, calcification, and erosions
- Increased vascularity

However, the differential diagnosis may include enthesis, an inflammatory lesion at tendon/bone junction, which is a common manifestation of spondyloarthropathy.33 The diagnosis of this condition often is delayed when back mobility is good and radiographic changes are absent. Other areas of inflammation could include the triceps insertion, patellar tendon, achilles tendon, and plantar fascia. US detection usually is more sensitive than the clinical examination.

Posterior interosseous nerve entrapment, an uncommon focal neuropathy, is thought to play a role in resistant tennis elbow. Focal swelling at the Arcade of Frohse or through the osteotendinous attachment of the common forearm extensors. These include the American College of Rheumatology, the Canadian Rheumatology Ultrasound Society, and EULAR. A superb sonoanatomy and cadaveric workshop is run yearly in Barcelona by the University of Barcelona and the Spanish School of Ultrasound and an interventional cadaveric workshop is planned for Chicago in 2014 by the ACR.

Clinicians interested in gaining more experience in musculoskeletal US can receive didactic workshops and hands-on training in several American, Canadian, and European centers. These include the American College of Rheumatology, the Canadian Rheumatology Ultrasound Society, and EULAR. A superb sonoanatomy and cadaveric workshop is run yearly in Barcelona by the University of Barcelona and the Spanish School of Ultrasound and an interventional cadaveric workshop is planned for Chicago in 2014 by the ACR.

Point-of care US for neuromuscular and musculoskeletal disease is a powerful tool for evaluating a wide spectrum of disorders and provides a opportunity for improved diagnosis and therapeutic decision making.34

34. Palau J. Complex anatomic areas and difficult diagnosis in the elbow. Presentation at Advanced European League Against Rheumatism Sonography Course in Madrid, June 2013. (jpalau@centredelama.com)

WHAT TO DO WHEN THE NEEDLE EMG IS NEGATIVE? AN ARGUMENT FOR THE REASONABLE USE OF MUSCULOSKELETAL ULTRASONOGRAPHY IN THE NEURODIAGNOSTIC LABORATORY
INTEGRATION OF NEUROMUSCULAR ULTRASOUND WITH ELECTRODIAGNOSIS

One of the most obvious uses for neuromuscular ultrasound (US) in the electromyography (EMG) laboratory is to facilitate safe and accurate needle placement during needle EMG and, in some cases, nerve conduction studies (NCSs). In addition to facilitating NCSs and needle EMG, in the hands of a skilled operator US provides detailed anatomical and pathophysiological information about nerve and muscle disease and real-time information regarding muscle activation and movement patterns. US should be considered complementary to electrodiagnostic (EDX) medicine; with EDX medicine providing physiologic and prognostic information, while US provides additional structural information. Just as the EDX examination must be individualized for each patient and modified based on ongoing findings, the US examination is also dynamic; the sonographer can rule in or out various diagnoses based on the information acquired while performing the study.

Most nerves amenable to electrophysiologic testing also can be visualized with US. Larger, superficial nerves are easier to identify, but smaller or deeper nerves also can be quickly identified with current US technology. Although US is not needed in the vast majority of NCSs, there are certain settings in which use of US may allow accurate localization of a nerve at the stimulation or recording site. For example, when anatomy is altered or the usual anatomical landmarks are not palpable due to body habitus, it may not be clear whether an absent response is due to true pathology or technical factors. One such example would be an ulnar motor NCS after ulnar nerve transposition surgery where the position of the transposed nerve can be mapped out in the antecubital fossa and segmental studies performed along the relocated course of the nerve. US-guided NCSs also have allowed more accurate and reliable evaluation of nerves that are typically challenging for technical or anatomical reasons. Examples include the lateral femoral cutaneous in the proximal thigh (Fig. 1) and the saphenous in the calf. Near-nerve needle stimulation or recording can be performed more quickly, accurately, safely, with lower current, and less patient discomfort when needles are placed under direct US guidance. This can be important when confirming true conduction block.

**Figure 1.** An example of near nerve needle placement for stimulating or recording. The image on the left shows the setup for the near nerve needle recording technique for the lateral femoral cutaneous sensory nerve conduction study. The image on the right shows a monopolar recording needle electrode adjacent to the lateral femoral cutaneous nerve (LFCN) in the thigh.
Similar to NCSs, US guidance has a role in needle EMG in certain clinical scenarios, the most apparent of which is facilitating accurate muscle localization. Although EDX physicians are trained to use various techniques for accurate localization during needle EMG, there are clinical scenarios in which it may be difficult to confidently isolate the targeted muscle. Inadvertent needle placement into an adjacent muscle (with a different peripheral nerve or nerve root supply) can lead to an erroneous conclusion. Most experienced EDX physicians are comfortable localizing muscles for needle EMG without using image guidance; however, muscles rarely examined can be more challenging as can deeper muscles, particularly in obese patients, or in cases where the anatomy is altered after surgery or trauma. US-guided needle placement also may be indicated in patients with severe denervation or spasticity preventing voluntary and/or isolated muscle activation. US-guided needle placement is a clear advantage in anticoagulated patients, or when examining muscles in close proximity to vascular structures (Fig. 2), as power Doppler can be used to quickly identify blood vessels. If there is clinical concern for bleeding, US can be used at the bedside to evaluate for that.

US imaging of nerve and muscle can provide additional diagnostic information when performed in conjunction with NCSs and needle EMG in the setting of nerve entrapment, nerve inflammation, and muscle disease. Its unique features include the ability to image structures dynamically in real time and the ability to perform sonopalpation. The dynamic aspect of US allows one to identify spontaneous activity such as fasciculations and, with increasing levels of resolution, even fibrillation potentials. The role of sonographic assessment of spontaneous activity is unclear at this time and may only be clinically applicable in the evaluation of neuromuscular disease in pediatric patients, where needle EMG is challenging. However, given that a larger area of muscle can be examined for fasciculation potentials using US compared with needle EMG, this has potential application when testing for certain diseases such as amyotrophic lateral sclerosis (ALS). The dynamic aspect of US also can be used to advantage in certain settings, such as diagnosing a subluxing ulnar nerve, and simultaneously evaluating for a snapping head of the medial triceps, an entity that commonly goes unrecognized, and it has implications in the surgical treatment of ulnar neuropathy. Dynamic entrapment related to anomalous muscle or compressive structures such as fascial bands can be visualized using real-time US, allowing the clinician to more confidently implicate those as a cause of entrapment. Sonopalpation (direct pressure from the transducer applied over the area of interest) is a unique feature that can help determine if a neuroma is symptomatic.

Grey scale imaging of muscle has been used for many years to diagnose muscle disease, but it may become more widely applicable in the future with further development of quantitative grey scale imaging. Muscle size, presence of atrophy or hypertrophy, changes in echotexture, and pattern of muscle involvement can be evaluated. The echo intensity of a muscle changes as it develops myopathic or neurogenic changes; such changes may manifest on US as increasing homogeneity and/or an overall increase in signal intensity, accompanied by muscle atrophy in some cases (Fig. 3). Quantitative imaging of muscle is a relatively simple technique to learn, and once normal values and technology to apply those values to different machines and transducers become more available, it has the potential for widespread use in screening for, diagnosing, and following nerve and muscle disease as well as in targeting biopsies.

![Figure 2](image2.png)

**Figure 2.** An example of the iliopsoas muscle (IP) with important neurovascular structures nearby: the femoral artery (FA) and the femoral nerve (FN). Ultrasound can increase the safety and accuracy of needle placement in anticoagulated or obese patients.

![Figure 3](image3.png)

**Figure 3.** An example of (A) a normal biceps brachii muscle and (B) a denervated biceps brachii muscle which shows markedly increased echogenicity and severe muscle atrophy.
EDX medicine provides diagnostic information regarding the presence of neuropathy and useful prognostic information based on severity of axon loss and demyelination. In contrast to EDX studies, high-resolution US in nerve disease provides anatomic information about the nerve itself and surrounding structures. It can be used to identify many types of nerve pathology including focal entrapment, nerve transection, neroma, nerve tumor, and intraneural ganglion (Fig. 4), and more diffuse involvement of the nerve as seen in multifocal motor neuropathy with conduction block, hereditary motor sensory neuropathy (Charcot–Marie–Tooth [CMT] disease), and acute and chronic inflammatory demyelinating polyradiculoneuropathy (AIDP/CIDP).

Structural lesions that can result in focal neuropathies can also be identified, such as fascial bands, anomalous muscles, hematomas, pseudoaneurysms (Fig. 5), lipomas, fibromas, and hemangiomas. US is useful in identifying anatomic variants such as a bifid median nerve or persistent median artery, which may have implications for surgical treatment.

The potential role of US in EDX studies is highlighted in a study in which diagnostic US was performed in a series of 77 patients after various mononeuropathies were diagnosed on NCSs and needle EMG. In 26% of cases, an underlying cause for the neuropathy was identified with US, resulting in modification of the management of those cases. In another study, intraneural ganglion cysts, which should be surgically resected, were seen on US in 18% of patients with EDX evidence of common peroneal neuropathy at the fibular head. In one series of 77 patients (96 wrists) with clinical and electrophysiologic evidence of carpal tunnel syndrome (CTS), 17% had underlying flexor tenosynovitis evident on US, and in another study where US was performed in patients presenting with electrophysiologic evidence of unilateral CTS, 35% had evidence of an underlying structural abnormality.

**PATIENT SELECTION**

As outlined above, there are many different scenarios in which neuromuscular US is likely to be informative, either as part of, or in addition to, an EDX study. Suggested uses are outlined below, for both US-guided needle placement and diagnostic US. As this field evolves it is likely that the role of neuromuscular US in the diagnosis of peripheral nerve and muscle disease will continue to expand.

Suggested uses of US-guided needle placement in the EMG laboratory setting include:

- When the accuracy of needle EMG is critical (neighboring muscles with differing nerve supply)
- In obese patients, where bony landmarks cannot be palpatated
- In anticoagulated patients (particularly for high risk muscles such as iliopsoas, flexor pollicis longus, and posterior tibialis)
- When spasticity or severe weakness of some muscles prevents isolated activation (e.g., rhomboid [Fig. 6], middle and lower trapezius, flexor digitorum profundus to the index and long finger versus flexor digitorum profundus to the ring and little finger, iliopsoas, and supinator)
- When needling high risk muscles such as the diaphragm (Fig. 7) or serratus anterior
- To localize nerves or muscles when the anatomy is altered due to surgery or trauma
- When placing monopolar needles to perform near nerve stimulation to rule out conduction block
- Suggested indications for diagnostic US after an EDX examination:
  - Any EDX identified mononeuropathy that may be due to entrapment (evaluate for mass lesions, anatomic variants, amyloidosis, and synovitis); consider routinely for:
    - Peroneal neuropathy (high rate of intraneural ganglion cyst, requiring surgical management)
• Unilateral median neuropathy at the wrist (high association with compressive lesions and flexor tenosynovitis)

• After fracture, with associated nerve injury

• A nonlocalizable ulnar neuropathy (may be able to identify focal swelling)

• Any patient in whom there is a high index of clinical suspicion for nerve entrapment, but the EDX study is negative or inconclusive

• When evaluating for CTS in patients with coexisting peripheral neuropathy or radiculopathy (with use of patient-derived control measurements)

• A suspected nerve transection or neuroma

• When evaluating for hypertrophic hereditary neuropathy (CMT or hereditary neuropathy with liability to pressure palsies), AIDP, CIDP, multifocal motor neuropathy with conduction block, or suspected Lepromatous neuropathy

• A brachial plexopathy (evaluate for root avulsion and neuroma)

• An evaluation for increased echo intensity (denervation or myopathy) when needle EMG is contraindicated or refused

• A dynamic evaluation for subluxation of the ulnar nerve or medial head of the triceps when ulnar neuropathy at the elbow evident on NCS

• An evaluation for fasciculation potentials in cranial muscles when ALS is suspected

DOCUMENTATION: KEY ELEMENTS OF A NEUROMUSCULAR ULTRASOUND REPORT

As with NCS/needle EMG reports, adequate documentation of peripheral nerve and muscle US is essential for high-quality patient care. The report must include basic elements and present data in a structured and succinct format that is easily interpreted. Below are specific recommendations and options for information to include in a neuromuscular US report.\textsuperscript{33} These recommendations do not preclude other reasonable methods of presenting EDX data. An example of a report may be found on the American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) website at http://www.aanem.org/Practice/Position-Statements.

When neuromuscular US is performed in conjunction with and on the same day as an EDX examination, a separately identifiable written report, or at least separate paragraphs within the same report for each test performed, should be generated. When billing for diagnostic US, images of any findings of note should be saved and available for review, with measurements included where appropriate. When billing for needle guidance, at least one image of the targeted structure should be saved, ideally with the needle in the image.

Figure 6. An ultrasound image of the chest wall muscles: rhomboid (R), scapula (S), and trapezius (T). Underlying the rhomboid is the intercostal muscle and deep to that the lung. The use of ultrasound when performing needle electromyography of these muscles can be very helpful in situations where obesity, altered anatomy, or severe denervation is present, making accurate and safe muscle localization more challenging.

Figure 7. Ultrasound can significantly enhance the accuracy and safety of needle electromyography of the diaphragm.

D=diaphragm, IC=intercostal muscle, L=liver, P=lung, SC=subcutaneous tissue
Important Elements to Include in the Report

Description of the Clinical Problem and Patient Data Section
A description of the clinical problem and patient data selection should include:

- Study indication and medical necessity; choose the appropriate ICD-9 code (may include brief pertinent history and physical examination findings that support the performance of neuromuscular US)
- Referring physician
- Patient demographic data
- Examining physician (and technician/sonographer, if appropriate)
- Type of US machine
- Type of transducer (linear versus curvilinear)
- Transducer frequency and gain setting

Results Section
For the results section of the report:

- List the side examined
- Document the patient positioning
- List the nerves, muscles, and other structures visualized (e.g., “the ulnar nerve was imaged from the axilla to the wrist”)
- Document the orientation of the US transducer to the nerve or muscle (ideally image the structure of interest in at least two planes).
- Provide numerical values for the results of nerve and muscle imaging; cross-sectional areas (measured just inside the hyperechoic epineurium) are the most reliable measures for nerve pathology and should be provided in standard units (i.e., mm² or cm²; measurements of nerve or muscle diameter, if included, should be in mm or cm)
- Include the location of each measurement.
- Include normal values for nerve measurements (ideally note the source of the normal values)
- Describe any abnormalities in nerve appearance, vascularity, or motion, and the site at which these were observed
- Note any abnormalities extrinsic to the nerve, such as ganglion cysts, with appropriate measurements, location, and presence or absence of vascularity; include a differential diagnosis for those findings
- Include the transducer frequency, depth of imaging, and gain settings (If reporting grey-scale imaging for muscle, US machine system settings strongly affect the appearance of muscle signal, thus these data must be included with each report. Also include a notation as to whether any post-processing, such as compound imaging or time-gain compensation, was used for the study. Gray-scale values may be provided as a means of quantifying muscle signal but the report should describe how these were calculated and should reference the source for the normal values used.

- Describe the muscle or nerve targeted, and the reason for the use of US guidance (e.g., decreasing risk of inadvertent puncture of vital structures or improved accuracy after failing to locate the target with a nonguided attempt)
- Describe length and gauge of needle used
- Note type of transducer probe used (curvilinear or linear, MHz range)
- Note that an aseptic technique was used
- Note any complications that occurred
- Include a paragraph documenting informed consent and observation of procedural pause

Interpretation Section
For the interpretation section:

- Provide an interpretation of the findings
- Provide a clinically meaningful conclusion with a definitive diagnosis when possible, or a list of relevant possibilities (e.g., “This is an abnormal sonographic study revealing a cystic mass compressing the right fibular nerve at the fibular head. This is most likely a synovial ganglion cyst, although a peripheral nerve sheath tumor cannot be excluded.”)
- Provide a clear statement of whether findings were normal or abnormal
- Provide any recommendations for further testing to clarify diagnosis
**Image Documentation**

Regarding image documentation:

- Images do not need to be included in the report
- Images should be saved and available for review; document the location of stored images (digital storage recommended)
- When billing for US-guided needle placement, it is recommended that at least one image of the needle once it has reached the target be saved

**Billing**

When billing for the US:

- Choose the appropriate evaluation and management (E/M) code, if indicated
- Choose the appropriate Current Procedural Terminology (CPT®) code
  - 76942: US guidance for needle placement
  - 76881: US, extremity, nonvascular, real-time with image documentation; complete
    - Code each limb as a separate unit
    - Add a 26 modifier if you do not own the US machine
  - 76882 (US, extremity, nonvascular, real-time with image documentation; limited anatomic specific)
    - Code each limb as a separate unit
    - Add a 26 modifier if you do not own the US machine

The CPT® code book states: “A complete ultrasound examination of an extremity (76881) consists of a detailed evaluation and real time scans of a specific joint that includes assessment of the muscles, tendons, joint, other soft tissue structures, and any identifiable abnormalities.”

- 76882 (US, extremity, nonvascular, real-time with image documentation; limited anatomic specific)
  - Code each limb as a separate unit
  - Add a 26 modifier if you do not own the US machine

The CPT® code book states: “76882 refers to an examination of an extremity that would be performed primarily for evaluation of muscles, tendons, joints, and/or soft tissues. This is a limited examination of the extremity where a specific anatomic structure such as a tendon or muscle is assessed. In addition, the code would be used to evaluate a soft tissue mass that may be present in an extremity where knowledge of its cystic or solid characteristics is needed.”

- Both 76881 and 76882 have medically unlikely edits to allow no more than two units billed per day

**Medicare allowables in 2013 include:**

- 76881: 2013 base Medicare payment is
  - $124.52 for global billing
  - $30.96 for professional component (use 26 modifier)
  - $93.56 for technical component (TC modifier)

- 76882: 2013 base Medicare payment is
  - $35.38 for global billing
  - $23.82 for professional component (26 modifier)
  - $11.57 for technical component (TC modifier)
REFERENCES


Neuromuscular ultrasound (US) involves the use of high-
frequency US to image peripheral nerves and muscles and assist
in the diagnosis of a variety of neuromuscular conditions. US
of peripheral muscle was first described in the early 1980s and
US of nerve in 1988.1,2 In 1991, Buchberger and colleagues
described the first use of neuromuscular US in the evaluation of
focal nerve disease, as they demonstrated nerve enlargement in
carpal tunnel syndrome (CTS).3 Over the past 20 years, the field
of neuromuscular US has evolved and matured, with publications
detailing its use in a large number of focal neuropathies, acquired
and hereditary polyneuropathies, motor neuron diseases, muscular
dystrophies, inflammatory myopathies, and other conditions.
Over the past 10 years, some electrodiagnostic (EDX) laboratories
have incorporated neuromuscular US into their routine clinical
practice. As this has occurred, some have questioned whether it is
a useful or necessary diagnostic test.

The systematic and regimented evidence-based medicine
approach for the evaluation of the efficacy of a specific diagnostic
imaging test is well described by Fryback and Thornbury.4 They
outline the following progression of six levels that are necessary
to confirm that a diagnostic imaging test is effective: (1) validity
and reliability, (2) accuracy, (3) change of diagnosis, (4) change
of treatment plan, (5) improved patient outcomes, and (6) cost-
effectiveness (Fig. 1). Therefore, an appropriate approach to
determining if neuromuscular US is an effective diagnostic
imaging modality is to determine how it performs when assessed
at each of these six levels.

The Argument for Using Neuromuscular Ultrasound in Combination With Electrodiagnosis

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Figure 1. The six levels used to assess an effective diagnostic imaging
test, as described by Fryback and Thornbury, are detailed, along with the
specific answers in regards to neuromuscular ultrasound.
LEVEL 1: IS NEUROMUSCULAR ULTRASOUND VALID AND RELIABLE?

Yes, neuromuscular US is valid and reliable. A diagnostic test has high validity if the measures obtained with the test correlate with the actual structure being measured. Nerve and muscle validity has been assessed by measuring nerve cross-sectional area and muscle thickness as determined by US, and then comparing these measures to actual measures in a cadaver model. The cross-sectional area of 96 nerves (12 unique sites, bilaterally, in four cadavers) correlated very closely with actual measures of nerve area (Pearson product-moment correlation coefficient of 0.97, p<0.001). Similarly, the thickness of 16 muscles (2 unique sites in the arm and leg, bilaterally, in four cadavers) correlated very closely with actual measures of muscle thickness (Pearson product-moment correlation coefficient of 0.99, p<0.001).

Assessment of reliability is not quite as direct as it is for validity. First, there is both intra-rater and inter-rater reliability, and then there is the issue of whether to measure the exact same image or to have each observer obtain a new image each time a nerve or muscle is assessed. No matter the measurement technique (i.e., intra or inter-rater, still image or real time), reliability for muscle measurements is very high, with the lowest intra-class correlation coefficient being 0.82 and all others being higher than 0.90. This included sonographers of varying skill level, from less than 3 months to over 10 years of experience. Similarly, intra-rater reliability for nerve cross-sectional area was extremely high, with the correlation coefficients being 0.98 or higher. Finally, correlation coefficients for inter-rater reliability were more varied, but for still images they were all significant at the p<0.001 level. Intra-class reliability for muscle measurements is very high, with the lowest intra-class correlation coefficient being 0.82 and all others being higher than 0.90.

LEVEL 2: IS NEUROMUSCULAR ULTRASOUND ACCURATE?

Yes, neuromuscular US is accurate. An accurate diagnostic test is one that has both high sensitivity and specificity. Highly sensitive tests have a low number of false-negative results (i.e., very few cases are missed) and highly specific tests have a low number of false-positive results (i.e., very few cases are incorrectly diagnosed with the suspected condition). Over the past 20 years, more than 45 articles have dealt specifically with the accuracy of neuromuscular US for the diagnosis of CTS alone. Similar accuracy studies have also been published regarding ulnar neuropathy at the elbow and traumatic focal neuropathies. A 2012 American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) clinical practice guideline evaluated the accuracy of neuromuscular US measurement of median nerve cross-sectional area for the diagnosis of CTS, and it identified four Class 1 and two Class 2 articles. This provided evidence to make the Level A recommendation that: neuromuscular US measurement of median nerve cross-sectional area at the wrist is accurate and may be offered as a diagnostic test for CTS. Similar evidence has not been published for the use of nerve conduction studies or needle electromyography (EMG) to diagnose CTS.

LEVEL 3: DOES NEUROMUSCULAR ULTRASOUND CHANGE THE DIAGNOSIS?

Yes, neuromuscular US changes the diagnosis. There are over 100 cases in the literature, from case reports and case series, in which US refined or changed the diagnosis of focal mononeuropathy. Examples include identification of extrinsic compressive structures (e.g., cysts, tumors, vessels, hematomas, fibrous bands, muscles, etc.), extrinsic irritating or inflammatory causes (e.g., synovitis, adjacent myositis), and intrinsic compressive or destructive structures (e.g., cysts, tumors, neuromas, foreign bodies, etc.) (Fig. 2).

Several studies have also examined this question on a more systematic level. In 2003, Kele and colleagues used US to assess 110 wrists in 77 individuals with CTS. They found that in 35% of the cases, US added information to the diagnosis of CTS. This included identification of compressive muscles within the tunnel, abnormal branching patterns of the median nerve, and compressive vascular structures. Padua and colleagues have published three important studies addressing this question. In 2007, they reported that US modified the diagnosis in 26% of 77 cases in which a single nerve was found to be abnormal clinically and with EDX studies. Then, in 2012, they evaluated 130 consecutive patients with nerve disease and found US impacted the diagnosis in 42.3% of cases. Finally, in 2013, they examined 112 nerves in 98 consecutive patients with traumatic nerve lesions and found that US strongly modified the diagnosis in 58% of cases. In this study of traumatic nerve injury the authors were able to identify the presence or lack of nerve continuity, the etiology of nerve trauma, and the location of trauma in those with multiple injuries to a single nerve.

The 2012 AANEM clinical practice guideline on the use of neuromuscular US for the diagnosis of CTS also examined the question of “added value” obtained when using US in combination with EDX studies. Twenty-three articles were identified that addressed this question, and four were graded as class II studies. This led to the Level B recommendation that: if available, neuromuscular US should be considered to screen for structural abnormalities at the wrist in those with CTS.
LEVEL 4: DOES NEUROMUSCULAR ULTRASOUND CHANGE TREATMENT PLAN?

Yes, neuromuscular US changes the treatment plan. In many of the case reports and series describing the utility of neuromuscular US, the treatment plan was changed based upon the US findings. This included resection of cysts, tumors, and aberrant muscles as well as repair of vascular structures, such as compressive aneurysms and pseudoaneurysms. Another example of an alteration in therapeutic strategy based on US occurs when a persistent median artery is detected, which can occur in up to one-quarter of patients. The presence of a persistent median artery makes a blind injection, or endoscopic carpal tunnel release, more challenging. On a more systematic basis, the prospective studies from Padua and colleagues in 2012 and 2013 indicated that between 42.3% and 58% of cases had an alteration in the therapeutic plan based upon the US findings.

LEVEL 5: DOES NEUROMUSCULAR ULTRASOUND IMPROVE PATIENT OUTCOMES?

It is unknown whether neuromuscular US changes outcomes. This is the first level of the six outlined by Fryback and Thornbury in which there is not published evidence to address the question. While it can be implied from case reports, case series, and prospective cohort studies that a more refined or accurate diagnosis, as obtained when US is used in combination with the EDX examination, leads to improved outcomes, this can only be proven with a randomized trial. Such a trial would compare the clinical outcomes in those who had diagnostic US to those who did not, to determine if the US group had better outcomes. Although such a study does not exist for neuromuscular US, it is also important to note that one also does not exist for EDX studies.

LEVEL 6: DOES NEUROMUSCULAR ULTRASOUND HAVE A GOOD COST-BENEFIT PROFILE?

It is unknown whether neuromuscular US has a good cost-benefit profile. Similarly to Level 5, this question has not been directly studied. However, it is highly likely that if Level 5 is satisfied and neuromuscular US is shown to change outcomes, then it will also have a good cost-benefit profile, since US is a relatively inexpensive diagnostic modality. The Current Procedural Terminology (CPT®) codes used most often for neuromuscular US are 76881 (complete) and 76882 (limited), and, as of 2013, the North Carolina Medicare allowable reimbursement rates are $116.60 for 76881 and $32.62 for 76882.

CONCLUSION

Four of the six levels outlined by Fryback and Thornbury for an effective diagnostic imaging test are met when assessing the evidence supporting neuromuscular US, and a clinical practice guideline endorsed by the AANEM and National Guideline Clearinghouse (which can be found at Guidelines.gov) provides Level A and Level B recommendations for the use of neuromuscular US in the assessment of CTS. Although two of the six levels for an effective diagnostic test have not been met (because the proper studies have not been performed), this is unfortunately common for many diagnostic tests used in routine clinical practice. In fact, the evidence for the use of neuromuscular US for the diagnosis of CTS is greater than for EDX studies, which have also only satisfied the first four levels and do not have a clinical practice guideline to support their use. (Of note, it is possible that the evidence for EDX studies is lacking because the techniques were developed before the emphasis on evidence-based medicine, or because the comprehensive review needed for a clinical practice guideline has just not been conducted.) If future studies (currently underway) show neuromuscular US to improve outcomes in a cost-effective manner, then it will be one of the few diagnostic modalities to meet all six levels of an effective diagnostic imaging test.

Clinicians interested in obtaining experience with neuromuscular US have several options to receive training in the technique. The AANEM offers workshops, courses, and symposia for a variety of experience levels at the annual meetings, and the International Society for Peripheral Neurophysiological Imaging (ISPNI) has annual meetings with a large number of hands-on workshops as well. In addition, some academic medical centers now offer courses in neuromuscular US, and the technique is being introduced into an increasing number of clinical neurophysiology, neuromuscular medicine, neurology, and physical medicine and rehabilitation residencies and fellowships. Hands-on experience with neuromuscular US, when combined with the growing body of evidence supporting the technique, is the best way to fully appreciate the valuable information gained when imaging nerve and muscle and is perhaps the greatest argument for using neuromuscular US in combination with EDX studies.

REFERENCES


BACKGROUND

Neuromuscular ultrasound (US) has been in use since the early 1980s in pediatric patients, then in the late 1980s it was utilized for the assessment of peripheral nerves. Neuromuscular US’s multiple benefits include its noninvasiveness, painlessness, and portability, which make it an attractive clinical and research tool for establishing diagnosis and treatment of neuromuscular disorders. Electrodiagnostic (EDX) testing, including nerve conduction studies (NCSs) and needle electromyography (EMG), has been in use since the 1940s and has played an increasingly important role in the clinical evaluation of patients suspected to have neuromuscular disorders. However, the sensitivity and specificity of neuromuscular US compared to needle EMG is variable based upon the neuroanatomic location in question. Neuromuscular US complements EDX studies well by providing anatomic information regarding nerves, muscles, and other non-neurological structures that cannot be recognized with NCSs and needle EMG. Neuromuscular US has established its role in detecting focal compressive neuropathies and currently is used for determining atrophy in muscle disorders such as myotonic dystrophy, a motor neuron disorder. However, its usefulness in other areas of the peripheral nervous system (PNS) is limited at this time. The combined use of neuromuscular US along with EDX studies (NCSs and needle EMG) currently is not a common practice.

THE CONTRIBUTION OF ULTRASOUND IN DOUBTFUL CASES

Neuromuscular US has only been in the market for less than half as long as needle EMG/NCSs. The inclusion of somatosensory evoked potentials, quantitative EMG, single fiber EMG, and autonomic testing in the EDX examination has further improved its sensitivity and specificity. This, in part, is because of a greater understanding of the pathophysiology of these disorders. Also of importance is the development of the techniques themselves, beginning with the basic needle EMG and NCS. Moreover, the improvement in computer technology has added speed and accuracy to the testing. US is off to a great start, continues to expand its role in various aspects of neuromuscular disorders. However, the sensitivity and specificity of neuromuscular US compared to EDX studies is variable based upon the neuroanatomic location in question. Neuromuscular US complements EDX studies well by providing anatomic information. US is more sensitive than MRI (93% versus 67%), has equivalent specificity (86%), and better identifies multifocal lesions than MRI. Neuromuscular US adds value to EDX studies when diagnosing carpal tunnel syndrome (CTS) and should be considered in screening for structural abnormalities at the wrist in those individuals with CTS.

Despite its limitations, neuromuscular US can be utilized when the clinical or neurophysiological picture is unclear. The simultaneous study of the PNS through both neuromuscular US and EDX studies may provide additional information. In a study conducted by Padua and colleagues, for about 25% of the patients neuromuscular US results confirmed the clinical needle EMG diagnosis. In most of these cases, the contribution of US was important for the detection of tumors or cysts, thus showing the cause of nerve involvement. The combination of needle EMG and US performed in the same session (or in collaboration with an US examiner) may be useful for diagnosis and determination of appropriate therapy. The area which has seen the most effort with neuromuscular US is in the evaluation of focal neuropathies.
caused by entrapment, and hundreds of articles have been written about neuromuscular US for CTS alone. Of the several changes which occur with entrapment, nerve enlargement is likely the most accurate for diagnosis, so efforts have been made to establish reference values for nerve cross-sectional area using US.\textsuperscript{9,10} However, the extent of the demyelinating injury or the axonal loss is not completely defined.

Neuromuscular US has emerged as a noninvasive technique that can be used in the structural and functional assessment of patients in intensive care units (ICUs). Neuromuscular clinicians often are asked to evaluate the diaphragm for diagnostic and prognostic purposes. Traditionally, this evaluation is accomplished through a history, physical examination, the fluoroscopic sniff test, and EDX studies.\textsuperscript{25} NCSs and needle EMG in this setting are challenging, uncomfortable, and can cause serious complications, such as pneumothorax. In addition, US is an informative technique for assessing muscles of patients with critical illness neuromyopathy. This technique can be examined further for diagnosing and tracking those with acquired weakness in the ICU stay.\textsuperscript{4} EDX studies are able to identify the chronicity of the problem based upon changes in recruitment pattern, fibrillations and positive waves, morphology, amplitude and duration of the motor unit action potentials. It remains to be seen whether US can differentiate between acute and chronic problems in patients with acquired weakness.

Dermatomyositis and polymyositis traditionally have been diagnosed with needle EMG as well as muscle biopsies. In one of the studies comparing magnetic resonance imaging (MRI) with US, it was found that MRI is more sensitive in detecting edema-like muscular changes and thereby acute myositis. Contrast-enhanced US with its capability of measuring perfusion has become a useful diagnostic tool in diagnosing acute inflammation in polymyositis and dermatomyositis.\textsuperscript{31} As far as the neuroimaging is concerned, T2-weighted MRI with fat suppression or short tau inversion recovery sequences are the most sensitive and specific routine method of imaging to detect polymyositis and dermatomyositis. US may be a cost-effective alternative to MRI, with contrast-enhanced US also permitting the assessment of muscle vascularization.\textsuperscript{30} It remains unclear whether US is better than needle EMG in diagnosing patients with inflammatory myopathies. Regarding hereditary muscle disease, in a study of US comparing types of myotonic dystrophy (i.e., myotonic dystrophy type 1 [DM1] and myotonic dystrophy type 2 [DM2]), all muscles tested were affected and structurally abnormal in DM2 patients. Proximal arm muscles were more affected in DM2 as compared to DM1, which corresponds to clinical findings.\textsuperscript{24} Unfortunately, neuromuscular US is unable to recognize myotonia in these patients, which is a characteristic identifying feature on the needle examination. The myotonia equivalent for the neuromuscular US examination has not yet been explored.

US is considered a useful method for evaluating cervical nerve root hypertrophy, which is frequently seen in patients with chronic inflammatory demyelinating polyneuropathy, particularly in patients with an elevated level of cerebrospinal fluid protein.\textsuperscript{30} Clinical evaluation, neurophysiology, and MRI are common tools in diagnosing lumbosacral radiculopathy. US can occasionally be helpful to determine the etiology of radiculopathy including tumor.

The utilization of both the EDX evaluation and US, possibly in the same session, may avoid misdiagnosis.\textsuperscript{21} Neuromuscular US has limited utility in diagnosing nerve root injuries. However, both studies may be complimentary in some cases.

Neuromuscular US can differentiate between amyotrophic lateral sclerosis (ALS) and mimics with high sensitivity and specificity, and it is a sensitive tool to screen for regional lower motor neuron involvement. Neuromuscular US detection of fasciculations and muscular change in motor neuron disease has been reviewed.\textsuperscript{28} Neuromuscular US demonstrates nerve and muscle atrophy in ALS and should be further explored as a disease biomarker.\textsuperscript{2,23} So, how this does provide additional information clinically? Currently, criteria for neuromuscular US clinical diagnosis of ALS are not yet developed.

THE ROLE OF ULTRASOUND ALONGSIDE NEEDLE ELECTROMYOGRAPHY

While US is clearly useful, there are several arguments to suggest that its use is not mandatory that it should be used with every needle EMG study. The answers to the following questions need to be established:

- Can US help physicians with every neuromuscular disorder?
- Can US provide additional information when used with needle EMG?
- Should US and needle EMG be used for just select or all conditions?
- Has the correct code for billing the joint use of US and needle EMG been determined?
- Has US reached the level of importance that it should be considered a mandatory piece of equipment in the EMG laboratory and should it be performed routinely?

US currently has some limitations in the following areas:

- Scope of practice/design of the EMG laboratory.
- Training for EDX physicians in the use of US.
- Interpretation of US images.
- Lack of current individual certification in US.
- Cost effectiveness of performing dual studies of US and needle EMG.
- Medicare needle EMG reimbursement cuts (effective January 2013).
Scope of Practice/Design of the Electromyography Laboratory

EMG laboratories are designed to perform NCSs and needle EMG studies. Patients are referred to EMG laboratories to get a needle EMG, not an US. EMG laboratories can perform all of the necessary EDX evaluations without having an US machine. This historical model has served well in most laboratories. Most EMG laboratories in United States and elsewhere do not utilize needle EMG and US in the same setting.

Training for Electrodiagnostic Physicians in the Use of Ultrasound

The development of professional parameters defining scope of practice, training requirements, and training methods to enable physicians in practice or during residency to obtain the skills needed to make this technology available to patients falls under the umbrella of future directions. The fundamental questions relating to US will be who is qualified to perform neuromuscular US, what training is needed, how competence is to be maintained and improved with continuing medical education, and how neuromuscular US should be classified. At present, there is not sufficient training within neurology training programs or EMG fellowship programs for an individual to acquire the skills needed to interpret an US.

Current neuromuscular US indications are not limited to but include needle guidance for NCSs and needle EMG, diagnosis of nerve entrapment, diagnostic muscle imaging via gray-scale analysis, and dynamic real-time imaging, including sonopalpation, to provide additional diagnostic information. Training and credentialing are reviewed, specifically noting the challenge of the lack of formal training programs and the relatively long, flat learning curve of diagnostic US. EDX physicians are not trained to perform US. Even if there is an US machine in the laboratory, one needs to develop special qualifications to make it useful. As the former residency program director, the author can attest that there is not sufficient training within neurology training programs or EMG fellowship programs for an individual to acquire the skills needed to interpret an US.

Interpretation of Ultrasound Images

US has limitations whether used independently or in addition to needle EMG, including limited penetration (e.g., obese patients), lower resolution at greater depths, and a high degree of reliance on operator skill (i.e., education/knowledge of anatomy, scanning skills, and interpretation). In addition, US images are often of poor quality and difficult to interpret. Even trained sonographers face challenges. There is a need to standardize US scanning techniques and definitions of various pathological US lesions as well as improve inter-observer agreement and reproducibility.

With improvements in resolution, the introduction of US contrast agents, and objective measures of nerve echogenicity, there is promise for further expanding its role in the diagnosis of all peripheral neuropathies. There are no formal guidelines on how to appropriately report neuromuscular US results. The American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) convened an expert panel to develop recommendations for reporting neuromuscular US findings. Neuromuscular US reports should provide comprehensive information along with a succinct conclusion, mirroring guidelines for EDX reports.

No data exists at present to outline comfort levels of laboratories utilizing the technique.

Lack of Current Individual Certification in Ultrasound

Currently, there is no individual certification in neuromuscular US. The American Board of Psychiatry and Neurology (ABPN) offers a subspecialty certification in neuromuscular medicine and the American Board of Electrodiagnostic Medicine (ABEM) provides an EDX medicine certificate. No medical board has begun to offer individual certification in neuromuscular US. Requirements for the eligibility, certification, and recertification need to be determined.

Cost Effectiveness of Performing Dual Studies of Ultrasound and Needle Electromyography

US-guided needle placement can be used to increase the accuracy and safety of NCSs and needle EMG. However, there remains an ongoing need for high-quality studies that evaluate the role and cost-effectiveness of neuromuscular US, both when used alone and in combination with EDX studies. In patients with CTS, the sensitivity of the combination of US and needle EMG is higher than the sensitivity of needle EMG or US alone. Boutte and colleagues revealed that the diagnostic value of US is not as good as that of the EDX examination. This means that in clinical practice, US is a complementary tool in cases of equivocal needle EMG.

The results of imaging examinations from US, computed tomography, and low-field MRI were compared with needle EMG and muscle biopsy findings. None of the imaging methods revealed specific diagnostic details, but they provided valuable information on the extent and distribution of muscle involvement. In myopathies, all imaging modalities corresponded well with needle EMG and histopathology findings, but in the neuropathies with minimal tissue destruction needle EMG was, understandably, more sensitive. In a study by Schwennicke and colleagues to evaluate the diagnostic value of muscle US in the diagnosis of focal neuropathy, US was less reliable than needle EMG.

Medicare Needle Electromyography Cuts (Effective January 2013)

In November 2012, the Centers for Medicare & Medicaid Services (CMS) issued the Medicare Physician Fee Schedule (MPFS) for calendar year 2013. Under this schedule, physicians who provide NCS and needle EMG services face cuts of more than 50% for some services starting January 1, 2013. According to the CMS, the reduction is a result of the “potentially misvalued code” initiative, which impacted EMG and nerve conduction tests. According to the American Academy of Neurology (AAN), the needle EMG and NCS test codes were not part of the proposed rule released in August 2012. The Neuropathy Association confirms the same, that reimbursement for needle EMG and NCSs will be severely...
NEUROMUSCULAR ULTRASOUND IN ACTION: A WAYS TO GO

reduced by 30-70%. Therefore, based upon this recent experience with reimbursement, an appropriate code needs to be established.

Physicians will need to know whether the performance of both studies in the same setting will be reimbursed. Criteria needs to be set for utilizing dual studies, including parameters such as:

- Both needle EMG and US, or
- Use of US more than needle EMG, or
- Use of needle EMG more than US.

Other Limitations

Audio and visual components, which are integral to the needle EMG examination, are missing with neuromuscular US. Will US be able to help replace fibrillations and positive waves from cranial and paraspinal muscles?

Myotonia, neuromyotonia, and complex repetitive discharges observed during needle EMGs help narrow down the differential diagnosis for a variety of neuromuscular disorders. However, their equivalent presence in neuromuscular US is lacking. High-resolution US has been used to evaluate several neuromuscular conditions, but it has only been used on a limited basis in ALS patients. Neuromuscular US demonstrates nerve and muscle atrophy in ALS and should be further explored as a disease biomarker. Is there a single fiber EMG and repetitive nerve stimulation US equivalent for neuromuscular junction (NMJ) deficits?

COMPARISON OF NEEDLE ELECTROMYOGRAPHY WITH ULTRASOUND

The Table summarizes the current utility of needle EMG and US for various disorders of the neuromuscular system. It appears that EDX studies are helpful with all the lower motor neuron localization. However, US is helpful for predominantly nerve and to some extent muscle disorders. The utility of US with nerve root as well as anterior horn cell disorders is very limited. There is no literature on the use of US with disorders of the NMJ.

<table>
<thead>
<tr>
<th>Neuroanatomic localization</th>
<th>Needle EMG</th>
<th>US</th>
<th>Needle EMG and US results</th>
<th>Needle EMG and US cost</th>
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</thead>
<tbody>
<tr>
<td>Nerve</td>
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<td>Helpful</td>
<td>Complimentary</td>
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</tr>
<tr>
<td>Muscle</td>
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<tr>
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<tr>
<td>AHC</td>
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<td>Limited</td>
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</tbody>
</table>

AHC=anterior horn cell, EMG=electromyography, NMJ=neuromuscular junction, US=ultrasonography

SUMMARY

Needle EMG has been used for almost 70 years and it has served well in the diagnosis of all lower motor neuron disorders of the nervous system. US has now been utilized for the last 20 years and it has shown great benefits in helping to diagnose most nerve and some muscle disorders. Neuromuscular US is an excellent complement to EDX studies for the evaluation of suspected neuropathies and a variety of other disorders. However, currently—based upon the current organization of EMG laboratories, the lack of appropriate training for EDX physicians, the limitations in interpretation of US images, the cost-benefit analysis of dual studies, and other limitations of US in comparison to needle EMG—EMG laboratories can function without it. The combined benefit at present is mainly in the areas of nerve and muscle in select cases. The cost of billing both the procedures together needs to be determined. EDX studies are predominantly the diagnostic studies of choice for evaluating disorders of the NMJ, nerve roots, and anterior horn cells. Neuromuscular US is an emerging diagnostic subspecialty field and it has become an important extension of the EDX examination. However, it has not replaced EDX studies, nor has it found a role in every aspect of PNS disorders.
REFERENCES


Neuromuscular Therapeutics
CME Questions:

1. Which of the following regarding ultrasound of nerves is true?
   A. Dislocation/subluxation of the ulnar nerve over the medial epicondyle of the humerus is sufficient to diagnosis ulnar neuropathy at the elbow.
   B. For evaluation of carpal tunnel syndrome, the best place to measure the median nerve cross sectional area is at the distal margin (outlet) of the carpal tunnel.
   C. Ultrasound may be more sensitive than magnetic resonance imaging for identifying nerve pathology.
   D. All of the above.

2. Ultrasound of nerves in polyneuropathies is often normal:
   A. In inherited, demyelinating neuropathies.
   B. In acquired, demyelinating neuropathies.
   C. In diffuse, axonal peripheral neuropathy.
   D. In all of the above.

3. Ultrasound echogenicity in normal skeletal muscle:
   A. Increases linearly throughout the lifespan.
   B. Increases most rapidly in both men and women after age 60.
   C. Is different in prepubertal males and females.
   D. Changes with age at the same rate in all muscles.

4. Skeletal muscle ultrasound in children:
   A. Has higher sensitivity than specificity in differentiating myopathic and neuropathic changes.
   B. Is more sensitive in children under age three than older children for detecting neuromuscular disorders.
   C. Is more sensitive for detecting mitochondrial disorders than muscular dystrophies.
   D. Yields similar results to EMG in evaluating hypotonic children ages 2-24 months.

5. Ultrasound of skeletal muscle in boys with Duchenne muscular dystrophy:
   A. Can be normal in children under age 3.
   B. Shows more severe pathologies with increasing age.
   C. Shows brighter signal in children with weaker strength and function.
   D. All of the above.

6. Fasciculations are:
   A. Detected with high sensitivity using ultrasound than electromyography.
   B. Often seen in patients with motor neuron disease.
   C. Present in normal people but are infrequent and often limited to muscles distal to the knee.
   D. All of the above.

7. The best strategy to differentiate a joint effusion from synovial thickening is to:
   A. Compress the region of interest with the transducer.
   B. Reduce the pulse repetition frequency to 1.0.
   C. Scan area of interest in 2 planes.
   D. Scan with power Doppler.

8. Power Doppler is preferred over Color Doppler for assessing inflammation for all the reasons listed except one?
   A. Not angle dependent to detect flow.
   B. Improved sensitivity for low flow rates.
   C. Avoids concerns for aliasing artifact.
   D. Is sensitive to probe pressure.

9. Enthesitis is a common inflammatory lesion in SpA and can be found at all listed sites except for one?
   A. Extensor carpi ulnaris insertion at wrist.
   B. Achilles tendon insertion at heel.
   C. Common extensor tendon at lateral epicondyle of humerus.
   D. Distal patellar tendon insertion at tibia.
10. Flexor tendon tenosynovitis in the hand is commonly associated with only one of the following features:
   A. Loss of fascicular pattern.
   B. Reduced cross sectional diameter.
   C. A1 pulley hypervascularity.
   D. Fibronodular thickening.

11. In which situation could ultrasound increase the safety or accuracy of needle EMG?
   A. Obese patients.
   B. Patients with altered anatomy.
   C. When examining chest wall muscles.
   D. When examining the iliopsoas in anticoagulated patients.
   E. All of the above.

12. Which pair of adjacent muscles has different peripheral nerve and/or nerve root innervation?
   A. Biceps and brachialis.
   B. Supraspinatus and infraspinatus.
   C. Tensor fascia lata and rectus femoris.
   D. Posterior tibialis and flexor digitorum longus.
   E. Extensor carpi radialis and supinator.

13. Advantages of diagnostic ultrasound over MRI include which of the following?
   A. Cost.
   B. Portability.
   C. Lack of exposure to radiation.
   D. Ability to dynamically image nerves and muscles.
   E. Speed of the examination.

14. In which situation would ultrasound be expected to add information to the EDX test?
   A. Common peroneal neuropathy at the knee.
   B. Fractured humerus with wrist drop.
   C. Median neuropathy that occurred after IV line placement.
   D. Patient presenting with sudden onset of shortness of breath and orthopnea.
   E. All of the above.

15. What is it not necessary to include in an ultrasound report?
   A. Description of the clinical problem.
   B. Results section.
   C. Interpretation.
   D. Reference to where images are stored.
   E. Ultrasound images.

16. What cannot be identified on ultrasound?
   A. Ganglion cyst causing compression of the tibial nerve in the tarsal tunnel.
   B. Flexor tenosynovitis causing median neuropathy at the wrist.
   C. Severe atrophy and lack of motion of the diaphragm.
   D. Conduction block in the ulnar nerve at the elbow.
   E. Swelling of the ulnar nerve at the elbow.

17. All of the following are questions proposed by Fryback and Thorbury to address the effectiveness of a diagnostic imaging test EXCEPT:
   A. Is the test valid?
   B. Is the test accurate?
   C. Is the test expensive?
   D. Does the test change treatment plans?

18. Which of the following methods could be used to assess the validity of neuromuscular ultrasound?
   A. Compare ultrasonographic nerve area measurements with a direct measurement of the same nerve.
   B. Compare ultrasonographic nerve area measurements between two different ultrasonographers.
   C. Compare ultrasonographic nerve area measurements obtained by the same sonographer at different time points.
   D. Compare ultrasonographic nerve area measurements between those with carpal tunnel syndrome and healthy controls.

19. A 35-year-old woman presents with paresthesias in the first three fingers of her right hand. Evidence based medicine supports which of the following statements:
   A. Muscle echogenicity is an accurate test to diagnose carpal tunnel syndrome.
   B. Ultrasonographic measurement of the median nerve area at the wrist is an accurate test for the diagnosis of carpal tunnel syndrome.
   C. Ultrasonographic measurement of median nerve flattening within the carpal tunnel is an accurate test for the diagnosis of carpal tunnel syndrome.
   D. Ultrasound guidance is required for the injection of corticosteroids into the carpal tunnel.

20. A 45-year-old man presents after a traumatic injury to the left forearm and subsequent weakness in the ulnar innervated intrinsic hand muscles. Ultrasound of the left ulnar nerve in this setting should be performed to assess for all of the following EXCEPT:
   A. Nerve continuity.
   B. Traumatic neuroma.
   C. Hematoma compressing the nerve.
   D. Fibrohamartoma.

21. Neuromuscular ultrasound (NM US) is most complimentary to electrodiagnostic studies for which of the following disorder?
   A. Focal compressive neuropathies.
   B. Myotonic dystrophy.
   C. Myasthenia gravis.
   D. Motor neuron disease.
22. Which of the following is true regarding NM-US training in the US?
   A. It has been ACGME approved part of the residency program training.
   B. Mandatory three months training is part of the ABPN certificate requirement.
   C. Neuroimaging certification requires at least six months of time spent in NM-US.
   D. No formal guidelines are published for NM-US training during the residency or fellowship programs.

23. NM-US is helpful in ALS patients by identifying which of the following features?
   A. Denervation in the tongue muscles.
   B. Denervation in thoracic muscles.
   C. Atrophy of the muscles.
   D. Loss of peripheral nerve myelin.

24. Which of the following can be detected with NM-US?
   A. Fasciculations.
   B. Myotonia.
   C. Neuromyotonia.
   D. Complex repetitive discharges.

25. Which of the following may represent NM-US equivalent of conduction block seen on NCS?
   A. Variation in the size of the nerve proximal to the lesion.
   B. Variation in the size of the nerve distal to the lesion.
   C. Variation in the size of the nerve both proximal and distal to the lesion.
   D. Rarely there are seen any changes in the nerve on US.