



CLINICAL GUIDELINES

Peripheral Nerve Disorders (PND) Imaging Guidelines

Version 2.0

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eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies: This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

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Peripheral Nerve Disorders (PND) Imaging Guidelines

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Abbreviations for Peripheral Nerve Disorders Imaging Guidelines

AIDS	Acquired Immunodeficiency Syndrome
ALS	Amyotrophic Lateral Sclerosis
CIDP	Chronic Inflammatory Demyelinating Polyneuropathy
CNS	central nervous system
CPK	creatinine phosphokinase
CT	computed tomography
EMG	electromyogram
LEMS	Lambert-Eaton Myasthenic Syndrome
MG	myasthenia gravis
MRI	magnetic resonance imaging
MRN	magnetic resonance neurography
MRS	magnetic resonance spectroscopy
NCV	nerve conduction velocity
PET	positron emission tomography
PNS	peripheral nervous system
PNST	Peripheral Nerve Sheath Tumor
POEMS	Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes
TOS	Thoracic Outlet Syndrome

PN-1: General Guidelines

PN-1.0: General Guidelines

A pertinent clinical evaluation is required before advanced imaging can be considered. The clinical evaluation should include a pertinent history and physical examination, including a neurological examination, appropriate laboratory studies, non-advanced imaging modalities, electromyography and nerve conduction (EMG/NCV) studies. Other meaningful technological contact (telehealth visit, telephone call or video call, electronic mail or messaging) by an established patient can serve as a pertinent clinical evaluation.

Nerve conduction studies are often normal early in the disease course with changes peaking at 2 weeks after symptom onset in the majority of patients. This will be taken into consideration on a case by case basis in regards to the NCV/EMG requirement in each section requirement of Peripheral Nerve Disorders Imaging Guidelines.

During the current COVID-19 pandemic, with limited face-to-face visits, the electrodiagnostic (EMG/NCV) study requirements may be waived with necessity to be determined by the treating neurologist or team coordinating the patient's care.

If imaging of peripheral nerves is indicated, ultrasound is the preferred modality for superficial peripheral nerves. MRI may be used for imaging deep nerves such as the lumbosacral plexus or nerves obscured by overlying bone such as the brachial plexus or for surgical planning. CT is limited to cases in which MRI is contraindicated.

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PN-2: Focal Neuropathy		
Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Carpal Tunnel Syndrome	YES	➤ Ultrasound Wrist or MRI Wrist without contrast (CPT® 73221) to estimate size of the carpal tunnel and diameter of the median nerve may be helpful in the evaluation and confirmation of carpal tunnel syndrome pre-operatively when EMG findings are equivocal and clinical findings are uncertain. ➤ See MS-21: Wrist in the Musculoskeletal Imaging Guidelines and SP-3: Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma in the Spine Imaging Guidelines.
Ulnar Neuropathy	YES	Ultrasound for evaluation when clinical findings and EMG/NCV findings are uncertain. MRI Elbow without contrast (CPT® 73221) or MRI Upper Arm or Forearm without contrast (CPT® 73218) for complex cases when diagnosis remains uncertain after EMG and US or for pre-op planning.
Radial Neuropathy	YES	➤ MRI Upper Arm or Forearm without contrast (CPT® 73218) in severe cases when surgery is being considered. ➤ MRI Upper Arm or Forearm without and with contrast (CPT® 73220) if there is a suspicion of a nerve tumor such as a neuroma.
Radial Neuropathy Notes: Leads to wrist drop with common sites of entrapment the inferior aspect of the humerus (Saturday night palsy) or the forearm (Posterior Interosseus Syndrome). Trauma or fractures of the humerus, radius, or ulna can damage the radial nerve.		
Sciatic Neuropathy	YES	MRI Pelvis without contrast (CPT® 72195) may be performed in the evaluation of these entities. CT Pelvis without contrast is not indicated due to lack of soft tissue contrast. It should only be performed in the rare circumstance of contrast allergy and contraindication to MRI such as pacemaking device.
Sciatic Neuropathy Notes: Trauma to the gluteal area with hematoma, injection palsy, hip or pelvic fractures, or hip replacement (arthroplasty) and rarely Piriformis Syndrome involves entrapment of the sciatic nerve at the sciatic notch in the pelvis by a tight piriformis muscle band.		
Femoral Neuropathy	NO	MRI Pelvis without contrast (CPT® 72195) may be performed in the evaluation of these entities.

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Femoral Neuropathy Notes: May occur as a complication of pelvic surgery in women or those on anticoagulants with retroperitoneal bleeding, or as a mononeuropathy in diabetics		
Meralgia Paresthetica	NO	MRI Pelvis without contrast (CPT® 72195) may be performed in cases of diagnostic uncertainty or for pre-op planning. CT Pelvis without contrast is not indicated due to lack of soft tissue contrast. It should only be performed in the rare circumstance of contrast allergy and contraindication to MRI such as pacemaking device.
Meralgia Paresthetica Notes: Sensory loss in the lateral femoral cutaneous nerve as it exits the pelvis under the inguinal ligament (lateral thigh without extension into lower leg), and is usually easily diagnosed based on a careful history and physical exam. EMG/NCV testing is often technically difficult and not required.		
Peroneal Neuropathy	YES	MRI Knee without contrast (CPT® 73721) or MRI Lower Extremity other than joint without contrast (CPT® 73718) in severe cases when surgery is considered.
Tarsal Tunnel Syndrome	N/A	See MS-27: Foot (Tarsal Tunnel Syndrome) in the Musculoskeletal Imaging Guidelines.

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PN-3: Polyneuropathy			
Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
PNS/CNS Crossover Syndromes	YES	MRI Brain and/or Spinal Cord without and with contrast if clinical findings point to abnormalities in those areas.	Examples: Guillain-Barré syndrome and Lyme disease
AIDS Related Cytomegaloviral Neuropathy/ Radiculopathy	YES	MRI Lumbar Spine without and with contrast (CPT® 72158) if suspected.	Urinary retention and a clinically confusing picture in the legs.
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	YES	MRI Lumbar Spine without and with contrast (CPT® 72158) if uncertain following EMG. See <u>PN-4: Brachial Plexus</u> , <u>PN-5: Lumbar and Lumbosacral Plexus</u> , and <u>PN-6.2: Muscle Diseases</u>	
Multifocal Motor Neuropathy	YES	MRI Brachial Plexus without and with contrast (CPT® 71552 or CPT® 73220) if uncertain following EMG.	
POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes)	YES	Advanced imaging is for the non-neurological entities of this rare osteosclerotic plasmacytoma syndrome.	See <u>ONC-25: Multiple Myeloma and Plasmacytomas</u> in the Oncology Imaging Guidelines.
Subacute Sensory Neuronopathy & Other Paraneoplastic Demyelinating Neuropathies	YES	Advanced imaging should be guided by specific clinical concern (See relevant guideline). For evaluation of suspected paraneoplastic syndromes: See <u>ONC-30.3: Paraneoplastic Syndromes</u> in the Oncology Imaging Guidelines	

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PN-4: Brachial Plexus

- Brachial plexus studies can be coded either as MRI Upper Extremity other than joint without or without and with contrast (CPT® 73218 or CPT® 73220), MRI Chest without or without and with contrast (CPT® 71550 or CPT® 71552) or MRI Neck without or without and with contrast (CPT® 70540 or CPT® 70543) (if upper trunk) after EMG/NCV examination for:
 - ◆ Malignant infiltration (EMG not required)
 - ◆ Radiation plexitis to rule out malignant infiltration
 - ◆ Brachial plexitis (Parsonage-Turner Syndrome or painful brachial amyotrophy).
 - Self-limited syndrome characterized by initial shoulder region pain followed by weakness of specific muscles in a pattern which does not conform to involvement of a single root or distal peripheral nerve
 - Consider MRI Cervical Spine if radiculopathy.
 - See **SP-3: Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma** in the Spine Imaging Guidelines
 - ◆ Traumatic injury (MRI Cervical Spine CPT® 72141 may be approved)
 - ◆ Neurogenic Thoracic Outlet Syndrome (TOS) failed a 2 to 3 month trial of conservative management and are being considered for surgical treatment.
 - ◆ See **CH-31: Thoracic Outlet Syndrome (TOS)** in the Chest Imaging Guidelines
 - ◆ Preoperative study which requires evaluation of the brachial plexus
 - ◆ MRI Chest and Neck are inherently bilateral; whereas MRI Upper Extremity is unilateral.
 - ◆ MRI should be performed prior to consideration of PET imaging.
 - ◆ If unable to have a MRI (e.g. implanted device), CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy and injury
 - ◆ For PET imaging requests, See **ONC-1.4: PET Imaging in Oncology** in the Oncology Imaging Guidelines.

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PN-5: Lumbar and Lumbosacral Plexus

- The following studies can be considered: MRI Pelvis without and with contrast with fat suppression imaging (CPT® 72197) **OR** MRI Abdomen and Pelvis without and with contrast with fat suppression imaging (CPT® 74183 and CPT® 72197) **OR** if MRI is not available, CT Pelvis with contrast (CPT® 72193) **OR** CT Abdomen and Pelvis with contrast (CPT® 74177) can be considered after EMG/NCV based on whether the upper lumbar plexus (abdominal retroperitoneal space) or the lumbosacral plexus (pelvis), respectively, is involved based on:
- ◆ Malignant infiltration (EMG not required)
 - ◆ Radiation plexopathy to rule out malignant infiltration
 - ◆ Traumatic injury (MRI Lumbar Spine without contrast CPT® 72148 including post-surgical cases may be approved)
 - ◆ Inflammation including sarcoidosis and infection
 - ◆ Toxic including iatrogenic during delivery (obstetric) or related to nerve blocks (ex. Botox®)
 - ◆ Metabolic including etiologies including diabetes
 - ◆ MRI should be performed prior to consideration of PET imaging.
 - ◆ If unable to have a MRI (e.g. implanted device), CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy and injury.
 - ◆ For PET imaging requests, See **ONC-1.4: PET Imaging** in the Oncology Imaging Guidelines

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PN-6: Muscle Disorders

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PN-6.1: Neuromuscular Junction Disorders

- Myasthenia Gravis (MG) is associated with thymic disease and can undergo:
 - ◆ CT Chest with contrast (CPT® 71260) after an established diagnosis of MG.
 - Can be repeated if initial CT previously negative and now symptoms of chest mass, rising anti-striated muscle antibody titers, or need for preoperative evaluation (clinical presentation, electro-diagnostic studies, and antibody titers).
 - ◆ CT Chest without contrast (CPT® 71250) may be used if there is concern regarding adverse effects of contrast in patients with MG.
- Lambert–Eaton myasthenic syndrome (LEMS) is associated with small cell lung cancer and can undergo:
 - ◆ CT Chest with contrast (CPT® 71260) with a suspected diagnosis (Chest x-ray, symptoms of lung mass, clinical presentation, electro-diagnostic studies, and antibody titers).
 - Can be repeated if initial CT previously negative after 3 months with persistent suspicion.
- Stiff-person syndrome is associated with small cell lung cancer and breast cancer
 - ◆ CT Chest with contrast (CPT® 71260) if Stiff-person syndrome is suspected based on clinical findings.

Practice Notes

- Myasthenia gravis is an autoimmune disease of the neuromuscular junctions, manifested by fatiguable weakness of the cranial nerves (examples - ocular: ptosis, diplopia, bulbar: dysphagia, dysarthria, dysphonia), as well as generalized limb weakness, depending on the severity of the disease. Associated antibodies: acetylcholine receptor (AChR), muscle specific kinase (MuSK).
- Lamber Eaton Myasthenic Syndrome is also an autoimmune disease affecting the neuromuscular junction presenting with ocular and bulbar symptoms and proximal limb weakness. Associated antibodies: P/Q voltage-gated calcium channel (VGCC).
- Stiff-person syndrome is an autoimmune disease associated with muscle spasm and muscle rigidity affecting the trunk and limb muscles. Associated antibodies: Glutamic acid decarboxylase (GAD)

PN-6.2: Muscle Diseases

- MRI and ultrasound are increasingly being used in the evaluation of muscle disease. MRI may be helpful in demonstrating abnormalities in muscles that are difficult to examine or not clinically weak, and MRI can also help distinguish between different types of muscle disease. MRI is also useful in determining sites for muscle biopsy.
- MRI Lower Extremity other than joint without contrast (CPT® 73718) or MRI Lower Extremity other than joint without and with contrast (CPT® 73720) and/or MRI Upper Extremity other than joint without contrast (CPT® 73218) or MRI Upper Extremity

other than joint without and with contrast (CPT® 73220), usually the most affected muscle is imaged (when criteria is met imaging can be approved for bilateral studies) for:

- ◆ Additional evaluation of myopathy or myositis (based on clinical exam and adjunct testing with EMG/NCV and labs)
 - ◆ To plan muscle biopsy
 - ◆ See **PEDMS-10.3: Inflammatory Muscle Diseases** in the Pediatric Musculoskeletal Imaging Guidelines
 - ◆ See **CH-11.1: Interstitial Disease** in the Chest Imaging Guidelines for interstitial lung disease associated with inflammatory myopathies
- All cases with dermatomyositis and polymyositis can undergo search for occult neoplasm See **ONC-30.3: Paraneoplastic Syndromes** in the Oncology Imaging Guidelines

PN-6.3: Gaucher Disease (Storage Disorders)

- See **AB-11: Gaucher Disease and Hemochromatosis** in the Abdomen Imaging Guidelines.
- Gaucher disease is group of autosomal recessive inborn errors of metabolism characterized by lack of the enzyme acid β -glucuronidase with destructive ceramide storage in various tissues. Gaucher disease is a treatable disorder (enzyme replacement) in which the liver, spleen, and bone marrow/bones are the most affected organs.
- Three types of Gaucher disease are recognized:
- ◆ **Type I** (non-neuropathic form or adult form): progressive hepatomegaly, splenomegaly, anemia and thrombocytopenia, and marked skeletal involvement; lungs and kidneys may also be involved, but central nervous system is spared
 - ◆ **Type II** (acute neuropathic form or infantile form): severe progressive neurological involvement with death by 1 to 2 years of age; hepatomegaly, splenomegaly, is also present (usually evident by 6 months of age)
 - ◆ **Type III**: type I with neurological involvement
- MRI Lumbar Spine without contrast (CPT® 72148) and Bilateral Femurs (CPT® 73718) is indicated to evaluate bone marrow involvement at initial diagnosis.
- ◆ Repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- MRI Abdomen without contrast (CPT® 74181) is indicated to assess liver and spleen involvement at initial diagnosis.
- ◆ Repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- Pulmonary involvement is less common, but CT Chest without contrast (CPT® 71250) is indicated for patients with new or worsening pulmonary symptoms.

- ◆ For patients with documented pulmonary involvement, repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- DXA may be approved every 2 years beginning at 6 years of age if requested by an institution with technical and interpretive expertise with DXA in pediatrics.
- Patients with Gaucher disease are at risk for osteonecrosis, osteomyelitis, and bony tumors. Acute bone pain should be imaged with plain xray.
 - ◆ MRI of affected areas with and without contrast may be approved if xray is non-diagnostic or indicates the need for further imaging, such as equivocal for osteonecrosis, infection or malignancy.
- PET/CT imaging is considered investigational in the evaluation of Gaucher disease. ¹⁸F-FDG does not reliably detect Gaucher disease in the marrow, and other isotopes are not yet FDA-approved for clinical use.

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PN-7: Magnetic Resonance Neurography (MRN)

- MRN is currently considered investigational by most payers.
- Use limited to evaluation of complicated cases and diagnostic uncertainty when other studies (EMG/NCV, ultrasound) are equivocal or non-diagnostic and results will determine intervention and/or surgical planning for peripheral nerve surgery and repair

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PN-8: Neuromuscular Disorders

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PN-8.1: Amyotrophic Lateral Sclerosis (ALS)

- For an established patient with ALS, an neurological examination is not required.
- MRI Brain, Cervical, Thoracic, and Lumbar Spine without contrast or without and with contrast are approvable.
 - ◆ Can be considered when ALS is suspected (combination of upper and lower motor neuron findings) to establish a diagnosis.
 - ◆ Repeat imaging can be evaluated based on the appropriate **Spine Imaging Guidelines**.

PN-8.2: Spinal Muscular Atrophy

- Molecular genetic testing is the standard tool for diagnosis for the early consideration in any infant with weakness or hypotonia.
 - ◆ MRI is usually unnecessary for the diagnosis.
- In patients with adult onset disease, the differential includes later onset disorders such as ALS with approvable MRI Brain and Spinal Cord imaging as per **PN-8.1: Amyotrophic Lateral Sclerosis (ALS)** when upper and lower motor neuron findings are present.

Practice Notes

- Evidence of lower motor neuron dysfunction in a muscle may include clinical examination of muscle weakness/wasting or EMG abnormalities to meet the criteria for the diagnosis of ALS.

PN-8.3: Fasciculations

- Fasciculations are spontaneous, erratic movements of muscle that may be secondary to benign and nonbenign etiologies.
- Prior to advanced imaging the following is required:
 - ◆ Clinical history should include the time course of symptoms, any associated weakness, areas of involvement, as well as the presence or absence of pain, sensory loss, or sphincter dysfunction.
 - ◆ Laboratory investigation consists of complete blood count, comprehensive metabolic panel, serum calcium, thyroid function testing, vitamin B12 level, sed rate, ANA, rheumatoid factor, and serum protein electrophoresis with immunofixation.
 - ◆ Certain clinical scenarios may require specialized lab testing (e.g. Lyme testing, HIV testing, heavy metals, etc.)
- The presence of upper motor neuron signs (e.g. increased tone, hyperreflexia, presence of Babinski or Hoffman signs) necessitates central nervous system imaging.

- ◆ MRI Brain (CPT® 70551 or CPT® 70553), MRI Cervical Spine (CPT® 72141 or CPT® 72156) and/or MRI Thoracic Spine (CPT® 72146 or CPT® 72157) are necessary to exclude mimics of non-benign etiologies of muscle fasciculations (i.e. motor neuron disease).
 - Typically, lumbar spine imaging is not indicated unless there is sphincter involvement, or there is a need to rule out lower motor etiologies in the lower extremities (e.g. lumbar radiculopathy, See **SP-6.1: Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain**).
- ◆ However, electrophysiologic studies (including but not limited to nerve conduction studies (NCS) AND needle EMG testing should be completed prior to CNS imaging during the evaluation of muscle fasciculations.
- ◆ Fasciculations may be present on electrodiagnostic testing (EMG/NCV). Spine imaging requests that do not meet under neuromuscular or muscle disorders should follow Spine Imaging Guidelines including for Radiculopathies.

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PN-9: Peripheral Nerve Sheath Tumors (PNST)

- PNST such as (Schwannomas or Neurofibromas) arise from Schwann cells or other connective tissue of the nerve. They can be located anywhere in the body. When suspected, advanced imaging may include:
 - ◆ MRI Brain without and with contrast (CPT® 70553) for a Vestibular Schwannoma. See **HD-33.1: Acoustic Neuroma and Other Cerebellopontine Angle Tumors** in the Head Imaging Guidelines
 - ◆ MRI Cervical, Thoracic, and Lumbar Spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158) for suspected paraspinal neurofibroma
 - ◆ Follow-up imaging is not needed unless:
 - New symptoms or neurological findings develop
 - Post operatively, at the discretion of or in consultation with the surgeon or to reestablish baseline if the tumor was not completely removed
 - Malignant transformation (5%) is known or suspected. This may include a metastatic work-up with CT Chest and Abdomen with contrast (CPT® 71260 and CPT® 74160).
- See **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines

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PN-10: Nuclear Imaging

- Nuclear Medicine
 - ◆ Nuclear medicine studies are not generally indicated in the evaluation of peripheral nerve disorders. See **PEDPN-2: Neurofibromatosis** in the Pediatric Peripheral Nerve Disorders Imaging Guidelines for specific imaging guidelines regarding PET/CT in evaluation of peripheral nerve tumors.