CIGNA MEDICAL COVERAGE POLICIES - RADIOLOGY

Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines

Effective Date: February 1, 2025

Instructions for use

The following coverage policy applies to health benefit plans administered by Cigna. Coverage policies are intended to provide guidance in interpreting certain standard Cigna benefit plans and are used by medical directors and other health care professionals in making medical necessity and other coverage determinations. Please note the terms of a customer's particular benefit plan document may differ significantly from the standard benefit plans upon which these coverage policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a coverage policy.

In the event of a conflict, a customer's benefit plan document always supersedes the information in the coverage policy. In the absence of federal or state coverage mandates, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of:

1. The terms of the applicable benefit plan document in effect on the date of service
2. Any applicable laws and regulations
3. Any relevant collateral source materials including coverage policies
4. The specific facts of the particular situation

Coverage policies relate exclusively to the administration of health benefit plans. Coverage policies are not recommendations for treatment and should never be used as treatment guidelines.

This evidence-based medical coverage policy has been developed by EviCore, Inc. Some information in this coverage policy may not apply to all benefit plans administered by Cigna.

These guidelines include procedures EviCore does not review for Cigna. Please refer to the **Cigna CPT code list** for the current list of high-tech imaging procedures that EviCore reviews for Cigna.

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Abbreviations for Peripheral Nerve and Neuromuscular Disorders 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

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Abbreviations for Peripheral Nerve Disorders Imaging Guidelines	
PNST	Peripheral Nerve Sheath Tumor
POEMS	Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes
TOS	Thoracic Outlet Syndrome

General Guidelines (PN-1.0)

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- 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, (since the onset or change in symptoms), appropriate laboratory studies, non-advanced imaging modalities, and electromyography/nerve conduction (EMG/NCV) studies. Other meaningful technological contact (telehealth visit, telephone call or video call, electronic mail or messaging) since the onset or change in symptoms, by an established individual can serve as a pertinent clinical evaluation.
- Nerve conduction studies are often normal early in the disease course with changes occurring from one to four weeks after symptom onset in the majority of individuals. This will be taken into consideration on a case-by-case basis in regards to the EMG/NCV requirement in each section requirement of the Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines.
- Due to the termination of the federal public health emergency declaration, the COVID-19 pandemic is no longer considered an indication to waive electrodiagnostic (EMG/NCV) study requirements within the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines.[7]
- 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.

Evidence Discussion (PN-1.0)

- Electromyography (EMG) and nerve conduction velocity (NCV) studies are useful in establishing the origin of peripheral nerve pathology and in guiding further diagnostic evaluation. Needle EMG following traumatic nerve injury may detect denervation of muscles that do not seem clinically affected. The optimal time to search for denervation changes is 10 to 14 days after the injury. Needle EMG may show residual innervation to paralyzed muscles. Follow-up EMG and NCV studies may demonstrate early evidence of re-innervation or evolving abnormalities that objectively demonstrate the temporal course of peripheral nerve pathology.
- Deferring EMG due to COVID-19 is less relevant at this time.
- For superficial peripheral nerves, ultrasound has significantly higher resolution than MRI. In terms of expense, safety, and noninvasiveness, ultrasound has clear advantages over MRI and the few comparative reports available confirm the value of ultrasound as an initial imaging choice.

- Advantages of ultrasound over MRI for detecting peripheral nerve pathology include lower cost, rapidity of examination, higher spatial resolution, imaging of the nerve in continuity, and ease of side-to-side comparisons. Ultrasound may better detect subtle changes in nerve caliber. This is important because peripheral nerve pathology is often fusiform in shape and can extend along the length of the nerve without greatly altering its cross-section area. MRI frequently misses multifocal (71%) and occasionally single pathologies.
- Advantages of MRI over ultrasound include superior contrast between tissues, imaging of structures that are deep or surrounded by bone, and tissue characterization using multi-sequence analysis and IV contrast.
- There is greater accuracy (96%) of diagnoses in cases of peripheral nerve sheath tumor, traumatic neuroma or neuropathy, idiopathic mono-neuropathy or plexopathy, fibrosis of nerves, nerve compression caused by ganglion or synovial cysts or any other soft tissue structures, non-neural soft tissue tumors, intra-neural granulomas, and vasculitis with ultrasound than MRI.

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Focal Neuropathy (PN-2)

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Focal Neuropathy (PN-2.1)

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Carpal Tunnel Syndrome	YES	• When EMG/NCV and clinical findings are equivocal **AND** only when requested for pre-operative planning, MRI Upper Extremity Joint (Wrist) without contrast (CPT® 73221) is indicated. • For radiculopathy, see **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)** in the Spine Imaging Guidelines.
Ulnar Neuropathy	YES	After EMG/NCV, only **ONE** of the following is indicated if requested for surgical consideration: • MRI Upper Extremity Joint (Elbow or Wrist) without contrast (CPT® 73221), **OR** • MRI Upper Extremity Other Than Joint (Forearm or Hand) without contrast (CPT® 73218)
Radial Neuropathy	YES	• MRI Upper Extremity Other Than Joint (Arm or Forearm) without contrast (CPT® 73218) when surgery is being considered. • MRI Upper Extremity Other Than Joint (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 at the inferior aspect of the humerus (Saturday night palsy) or the forearm (Posterior Interosseous Syndrome). Entrapment of the nerve at the wrist (Wartenberg syndrome or handcuff palsy) typically spares motor involvement and results only in sensory changes.

Trauma or fractures of the humerus, radius, or ulna can damage the radial nerve.

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Pudendal Neuropathy[7,8]	NO	• **Documented concern specifically for pudendal neuropathy, pudendal neuralgia, or pudendal entrapment** : MRI Pelvis without contrast (CPT® 72195) **OR** MRI Pelvis without and with contrast (CPT® 72197) ◦ If there is a contraindication to MRI **and** the above documented concern is present, then **ONE** of the following is indicated: ▪ CT Pelvis without contrast (CPT® 72192) ▪ CT Pelvis with contrast (CPT® 72193) ▪ CT Pelvis without and with contrast (CPT® 72194) • For all other pelvic concerns, see the following Pelvic Imaging Guidelines (as indicated): ◦ **Pelvic Pain/Dyspareunia Female (PV-11.1)** ◦ **Impotence/Erectile Dysfunction (PV-17.1)** ◦ **Male Pelvic Disorders (PV-19.1)** ◦ **Scrotal Pathology (PV-20.1)**
Pudendal Neuropathy Notes : Causes pain, sexual dysfunction, or sensory change in the genitals, perineum, and perianal region. May be caused by trauma, recurrent injury from exercise such as cycling, pelvic mass, or after viral infection (e.g., post-herpetic neuralgia).		
Sciatic Neuropathy	YES	• MRI Pelvis without contrast (CPT® 72195) • CT Pelvis without contrast (CPT® 72192) is **NOT** routinely indicated due to lack of soft tissue contrast. ◦ It should only be performed in the rare circumstance of contrast allergy and/ or contraindication to MRI such as pacemaking device.

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Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Sciatic Neuropathy Notes : May be caused by trauma to the gluteal area with hematoma, injection palsy, hip or pelvic fractures, or hip replacement (arthroplasty). **Piriformis Syndrome** involves entrapment of the sciatic nerve at the sciatic notch in the pelvis by a tight piriformis muscle band. Concerns for piriformis syndrome should be imaged according to the sciatic neuropathy criteria.		
Femoral Neuropathy	NO	• MRI Pelvis without contrast (CPT® 72195)
Femoral Neuropathy Notes : May occur as a complication of pelvic surgery in females or those on anticoagulants with retroperitoneal bleeding, or as a mononeuropathy in diabetics		
Meralgia Paresthetica	NO	• MRI Pelvis without contrast (CPT® 72195) is indicated for **ANY** of the following scenarios: ◦ Cases of diagnostic uncertainty ◦ Pre-operative planning • CT Pelvis without contrast (CPT® 72192) is **NOT** routinely indicated due to lack of soft tissue contrast. ◦ It should only be performed in the rare circumstance of contrast allergy and/ or 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 diagnosed based on a careful history and physical exam. EMG/NCV testing is often technically difficult and not required.		
Peroneal Neuropathy	YES	• MRI Lower Extremity Joint (Knee) without contrast (CPT® 73721) **OR** MRI Lower Extremity Other Than Joint without contrast (CPT® 73718) when surgery is considered or when ordered by or in consultation with a surgeon.

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Tarsal Tunnel Syndrome	N/A	• See **Foot (Tarsal Tunnel Syndrome) (MS-27)** in the Musculoskeletal Imaging Guidelines.

Evidence Discussion (PN-2.1)

- Focal neuropathies are typically diagnosed by a combination of clinical history, thorough neurological examination, and electrodiagnostic testing with electromyography (EMG) and nerve conduction studies (NCS).
- When clinical evaluation and electrodiagnostic testing are inconclusive, MRI may allow for better identification and anatomic localization of lesions and is considered the gold standard for imaging of the peripheral nerve.
- The sensitivity and specificity of MRI findings for carpal tunnel syndrome are low (sensitivity, 23%–96%; specificity, 39%– 87%), and for this reason MRI imaging does not play a role in the routine clinical assessment of carpal tunnel syndrome. However, MRI of the wrist can help identify surgical candidates when clinical and electrodiagnostic findings are inconclusive.
- When caused by nerve entrapment or compression, focal neuropathies may benefit from surgical release or decompression. MRI can provide visualization of the cause of compression, rule out other causes of nerve injury, and allow for a more focused operative approach, particularly when surgery is considered to decompress common entrapment neuropathies of the ulnar, radial, and peroneal nerves.
- Sciatic, femoral, and pudendal neuropathies often occur secondary to trauma, compression, or entrapment of the affected nerve. These are often diagnosed clinically or localized with electrodiagnostic testing. MRI imaging of the pelvis may be indicated to assess for sources of compression, including occult malignancy.
- Meralgia paresthetica is the common term describing pathology of the lateral femoral cutaneous nerve of the thigh. The nerve is prone to injury and compression but may have a variable anatomic course. Meralgia paresthetica is primarily diagnosed by clinical history and exam, as neuroimaging and electrodiagnostic testing results may be difficult to interpret. Neuroimaging is most useful in cases of diagnostic uncertainty, particularly when surgical exploration and treatment are considered.

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Polyneuropathy (PN-3)

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Polyneuropathy (PN-3.1)

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Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
Polyneuropathies with Central Nervous System (CNS) Involvement	YES	If clinical findings point to abnormalities in those areas, then **ANY** of the following are indicated: • MRI Brain without and with contrast (CPT® 70553), **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)[6,7]	Examples: Guillain-Barré syndrome, inflammatory polyneuropathies unspecified, and Lyme disease
AIDS-Related Cytomegaloviral Neuropathy/ Radiculopathy[1]	YES	• MRI Lumbar Spine without and with contrast (CPT® 72158) • If concern for myelopathy, **ANY** of the following imaging are **ALSO** indicated: ◦ MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR** ◦ MRI Thoracic Spine without and with contrast (CPT® 72157)	• Often presents with urinary retention and a clinically confusing picture in the legs. • For myelopathic signs and symptoms, see **Myelopathy (SP-7.1)** .

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Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	YES	MRI Lumbar Spine without and with contrast (CPT® 72158) **AND/OR** MRI Cervical Spine without and with contrast (CPT® 72156) if diagnosis uncertain following EMG/NCV.[7] • For imaging requests of the brachial or lumbosacral plexus or muscle: See **Brachial Plexus (PN-4.1)**, **Lumbar and Lumbosacral Plexus (PN-5.1)**, and **Muscle Diseases (PN-8.5)**	
Multifocal Motor Neuropathy	YES	If diagnosis is uncertain following EMG/NCV, MRI of the Brachial Plexus is supported with **ONE** of the following: • MRI Upper Extremity other than joint without and with contrast (CPT® 73220) • MRI Chest without and with contrast (CPT® 71552) • MRI Neck without and with contrast (CPT® 70543)	
POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin changes)	YES	Advanced imaging is for the non-neurological etiologies of this rare osteosclerotic plasmacytoma syndrome.	See **Multiple Myeloma and Plasmacytomas (ONC-25)** 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 **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.	

Background and Supporting Information

- Central Nervous System (CNS) Imaging (Brain and Spinal Cord) is not required for Polyneuropathy without CNS signs/symptoms.[6]
- Distal symmetric polyneuropathy is the most common pattern of generalized peripheral neuropathy. It is typically sensory predominant and may demonstrate neurological abnormalities including reduced or absent deep tendon reflexes (DTRs), reduced sensation to multiple testing modalities (vibration, proprioception, etc). In more advanced staging, mild motor weakness may be present. It is most often associated with diabetes and metabolic abnormalities. In the absence of atypical findings (such as asymmetrical presentation, significant weakness, or upper motor neuron exam findings such as hyperreflexia or spasticity), distal symmetric polyneuropathy does not require central nervous system (CNS) imaging.[6]

Evidence Discussion (PN-3.1)

- Polyneuropathies are typically diagnosed by a combination of clinical history, thorough neurological examination, lab work up, and electrodiagnostic testing with electromyography (EMG) and nerve conduction studies (NCS).
- For systemic polyneuropathies with potential for central nervous system (CNS) involvement, such as Lyme disease-related polyneuropathy and some inflammatory polyneuropathies, MRI imaging of the brain and/or spinal cord may be helpful identify typical patterns of involvement or to rule out other pathologies when clinical findings suggest CNS involvement.
- Neuropathy is the most common neurological complication of human immunodeficiency virus (HIV) infection and, in its most common form, is treated with symptom management and anti-viral therapy. However, other acquired immunodeficiency syndrome (AIDS)-related neurological disorders may be difficult to clinically differentiate from common HIV polyneuropathy and may require more aggressive treatment. Accurate diagnosis of AIDS-related cytomegalovirus (CMV) polyradiculopathy, HIV vasculitis, or AIDS-related motor neuron disease is required for appropriate treatment; MRI imaging of the spinal cord or nerve roots may assist with diagnosis when clinically indicated.
- Chronic acquired demyelinating polyneuropathies, including chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN), are diagnosed by clinical history and results of electromyography and nerve conduction studies. If the diagnosis remains uncertain after these studies, neuroimaging may help establish the diagnosis. Evidence of lumbar nerve root involvement on MRI lumbar spine is supportive of a CIDP diagnosis. T2-weighted signal change on MRI of the brachial plexus is often present in MMN patients.
- POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein, Skin Changes) syndrome is a disorder affecting multiple organ systems which occurs in the setting of a plasma cell disorder. Diagnosis is based on electrodiagnostic confirmation of polyneuropathy and work up of the underlying oncologic condition.

- Electrodiagnostic testing can provide valuable findings in the investigation of some paraneoplastic polyneuropathies; however, identification of the underlying malignancy and appropriate oncological management are key to management.

References (PN-3)

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Brachial Plexus (PN-4)

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Brachial Plexus (PN-4.1)

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- EMG/NCV examination is required prior to advanced imaging except in cases of malignant infiltration or radiation plexitis as detailed below.[8-12]

Brachial Plexus Imaging		
Indication	**Imaging**	**Notes**
Malignant infiltration (EMG not required)	Any **ONE** of the following: • MRI Upper Extremity other than joint without contrast (CPT® 73218) • MRI Upper Extremity other than joint without and with contrast (CPT® 73220) • MRI Chest without contrast (CPT® 71550) • MRI Chest without and with contrast (CPT® 71552) • MRI Neck without contrast (CPT® 70540) • MRI Neck without and with contrast (CPT® 70543)	
Radiation plexitis to rule out malignant infiltration (EMG not required)		
Neurogenic Thoracic Outlet Syndrome (TOS)[10]		
Preoperative work up requiring evaluation of the brachial plexus		
Brachial plexitis (Parsonage-Turner Syndrome or painful brachial amyotrophy)	• Any **ONE** of the above studies **AND** • If there is concern for radiculopathy in addition to plexopathy, MRI Cervical Spine without contrast (CPT® 72141)	• For concern for cervical radiculopathy, see **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)** • For details of brachial plexitis (Parsonage-Turner Syndrome), see **Background and Supporting Information**.
Traumatic injury[13]		

- MRI Chest and Neck are inherently bilateral, whereas MRI Upper Extremity is unilateral.
- If MRI is not available or is contraindicated, CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy

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and injury. In this circumstance, when the above criteria are met, only **ONE** of the following studies is indicated:
  - **CT Neck Soft Tissue** : CT Neck without contrast (CPT® 70490); **or** , CT Neck with contrast (CPT® 70491); **or** , CT Neck without and with contrast (CPT® 70492)
  - **CT Upper Extremity** : CT Upper Extremity without contrast (CPT® 73200); **or** , CT Upper Extremity with contrast (CPT® 73201); **or** , CT Upper Extremity without and with contrast (CPT® 73202)
  - **CT Chest** : CT Chest without contrast (CPT® 71250); **or** , CT Chest with contrast (CPT® 71260); **or** , CT Chest without and with contrast (CPT® 71270)
- MRI should be performed prior to consideration of PET imaging.
  - For PET imaging, see **PET Imaging in Oncology (ONC-1.4)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Brachial plexitis (Parsonage-Turner syndrome or painful brachial amyotrophy) is a 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.

Evidence Discussion (PN-4.1)

- MRI is the imaging study of choice to evaluate the brachial plexus due to superior soft-tissue contrast and good spatial resolution, providing detailed definition of intraneural anatomy as well as localizing pathologic lesions in conditions in which electrodiagnostic and physical findings are nonspecific. A variety of findings may be seen within the brachial plexus on MRI, including increased T2 signal intensity, focal or diffuse enhancement, or enlargement or edema of nerve segments. Furthermore, signal abnormalities or atrophy in muscles supplied by the brachial plexus can help support a plexopathy. MRI is more sensitive than CT at identifying subtle infiltrative lesions regions or areas of enhancement.
- Regarding fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT, there is no relevant literature to support the use of FDG-PET/CT in the evaluation of traumatic or nontraumatic brachial plexopathy in the absence of a known malignancy.

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Lumbar and Lumbosacral Plexus (PN-5)

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Lumbar and Lumbosacral Plexus (PN-5.1)

PN.LP.0005.1.A
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- EMG/NCV examination is required prior to advanced imaging.
  - EMG/NCV is **NOT** required if there is concern for malignant infiltration.
- For suspected lumbar and/or lumbosacral plexopathy, **ONE** of the following is indicated
  - MRI Pelvis without contrast (CPT® 72195) with fat suppression imaging, **OR**
  - MRI Pelvis without and with contrast (CPT® 72197) with fat suppression imaging, **OR**
  - MRI Abdomen without contrast (CPT® 74181) and MRI Pelvis without contrast (CPT® 72195) with fat suppression imaging, **OR**
  - MRI Abdomen without and with contrast (CPT® 74183) and MRI Pelvis without and with contrast (CPT® 72197) with fat suppression imaging
- If suspected lumbar and/or lumbosacral plexopathy is due to a traumatic injury, then MRI Lumbar Spine without contrast (CPT® 72148) is **ALSO** indicated.
  - See **Low Back (Lumbar Spine) Trauma (SP-6.2)**
- If MRI is not available or is contraindicated, CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy and injury. In this circumstance, when requested for suspected lumbar and/or lumbosacral plexopathy, **EITHER** of the following is indicated:
  - CT Pelvis with contrast (CPT® 72193), **OR**
  - CT Abdomen and Pelvis with contrast (CPT® 74177)
- For PET imaging, see **PET Imaging in Oncology (ONC-1.4)** in the Oncology Imaging Guidelines.

Background and Supporting Information

- Lumbar and lumbosacral plexopathy may be caused by any of the following:
  - Malignant infiltration
  - Radiation
  - Traumatic injury
  - Inflammation including sarcoidosis and infection
  - Toxic including iatrogenic during delivery (obstetric) or related to nerve blocks (ex. Botox®)
  - Metabolic including etiologies including diabetes

Evidence Discussion (PN-5.1)

- MRI is the imaging study of choice to evaluate the lumbosacral plexus due to superior soft-tissue contrast and good spatial resolution, providing detailed definition of intraneural anatomy as well as localizing pathologic lesions in conditions in which electrodiagnostic and physical findings are nonspecific. Abnormal MRI findings in lumbosacral plexopathies include increased T2 signal intensity, focal or diffuse enhancement, or enlargement or edema of nerve segments. MRI is more sensitive than CT at identifying subtle infiltrative lesions, although CT may be useful to assess for psoas hematoma.
- Regarding fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT, there is no relevant literature to support the use of FDG-PET/CT in the evaluation of traumatic or nontraumatic lumbosacral plexopathy in the absence of a known malignancy.

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Muscle Disorders (PN-6)

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Muscle Disorders (PN-6)

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- See **Neuromuscular Junction Disorders (PN-8.4)**
- See **Muscle Disease (PN-8.5)**
- See **Gaucher Disease (Storage Disorders) (PN-8.6)**

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Magnetic Resonance Neurography (MRN) (PN-7)

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Magnetic Resonance Neurography (MRN) (PN-7.1)

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- MRN is supported when **ALL** of the following criteria are met:
  - The study is to evaluate a traumatic or compressive focal neuropathy **or** a brachial plexus injury.
  - The study is requested by a neurosurgeon, orthopedic surgeon, neurologist, or podiatrist after an in-person clinical evaluation **AND** when surgery is being considered.
  - EMG/NCV has been performed and results provided.
  - The diagnosis remains unclear following prior imaging of the region with x-ray, ultrasound, or conventional imaging (CT or MRI).
    - For conventional imaging criteria, see **Focal Neuropathy (PN-2.1)** and **Brachial Plexus (PN-4.1)**.
- MRN is reported as **ONE** of the following:
  - Unlisted MRI procedure code (CPT® 76498), **OR**
  - MRI extremity with **ONE** of the following codes:
    - MRI Upper Extremity, other than joint, without contrast (CPT® 73218)
    - MRI Upper Extremity, other than joint, without and with contrast (CPT® 73220)
    - MRI Lower Extremity, other than joint, without contrast (CPT® 73718)
    - MRI Lower Extremity, other than joint, without and with contrast (CPT® 73720)
- MRN for **ANY** other indication is considered **NOT medically necessary** at this time, including for assessment of lumbosacral plexopathy, neuromuscular disease, and polyneuropathy.

Background and Supporting Information

Magnetic resonance neurography utilizes standard MRI equipment with sequences and technology that allow for optimized viewing of the peripheral nerve. MRN creates greater contrast between the nerve and other surrounding soft tissue to allow a detailed view of the nerve tissue and layers. This allows for more accurate diagnosis of the location and degree of nerve injury.

Evidence Discussion (PN-7.1)

- Magnetic resonance neurography (MRN) offers advantages over standard MRI imaging by utilizing sequences and technology that optimize viewing of the peripheral nerve. MRN presents no increased risk to safety over standard MRI.1 MRN is a non-

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invasive, accurate, reliable method of demonstrating normal and abnormal nerve and assessing regional muscle denervation with good surgical correlation to findings.

- Efficacy and reliability of MRN have been clinically validated in the diagnosis and localization of traumatic and compressive focal neuropathies and brachial plexus injuries for the purpose of surgical consideration. A clinical study assessing the impact of MRN data on surgical planning noted that review of MRN altered the suspected nerve involvement in 23% and changed the nerve injury grade in 27% of patients studied. Surgeons reported MRN altered their determination of the need for surgery in 63%, timing of surgery in 41%, length of skin incision in 27%, and time in the operating room in 30% of cases reviewed. This data suggests that MRN may improve the selection of candidates for surgical repair of these lesions and may narrow the focus of surgery.
- There is insufficient literature to support the role of MRN for evaluation of other pathologies, including but not limited to, lumbosacral plexopathy, neuromuscular disease, and polyneuropathy. Thus, MRN is considered not medically necessary at this time for indications other than traumatic and compressive focal neuropathies and brachial plexus injuries.

References (PN-7)

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Neuromuscular Disorders (PN-8)

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Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)

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- A neurological examination is **NOT** required for an individual with established diagnosis of motor neuron disease/ALS **or** when diagnosis is suspected by a neurologist, geneticist, or a physical medicine and rehabilitation (PM&R) specialist.
- For initial evaluation of suspected motor neuron disease/ALS, **ANY** of the following are indicated
  - **Brain** : MRI Brain without contrast (CPT® 70551) **or** MRI Brain without and with contrast (CPT® 70553), **AND/OR**
  - **Cervical Spine** : MRI Cervical Spine without contrast (CPT® 72141) **or** MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR**
  - Thoracic Spine: MRI Thoracic Spine without contrast (CPT® 72146) **or** MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR**
  - **Lumbar Spine** : MRI Lumbar Spine without contrast (CPT® 72148) **or** MRI Lumbar Spine without and with contrast (CPT® 72158)
- Repeat imaging can be evaluated based on the appropriate **Spine Imaging Guidelines.**

Background and Supporting Information

- 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.
- Motor Neuron Diseases (also known as Anterior Horn Cell Diseases) are heterogeneous and encompass either upper motor neurons, or lower motor neurons, or both. Upper motor neurons begin in the cerebral cortex and descend into the brainstem (corticobulbar), or spinal cord, where there is a connection to the lower motor neuron that exits the central nervous system and reaches out to the muscle.
  - The various types can be divided into the areas so affected:
    - Amyotrophic Lateral Sclerosis (Lou Gehrig's disease) – both Upper and Lower Motor Neurons
    - Primary Lateral Sclerosis – Upper Motor Neurons
    - Progressive Muscular Atrophy – Lower Motor Neurons
    - Progressive Bulbar Palsy – Rare and limited to bulbar muscles (muscles innervated by the Cranial Nerves – dysarthria and dysphagia)
  - Other rare conditions:

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  - Monomelic Amyotrophy (Hirayama disease)
  - Spinal Bulbar Muscular Atrophy (Kennedy Disease)
- Signs of lower motor neuron pathology include weakness, fasciculations, atrophy, decreased muscle tone, decreased reflexes, and a plantar extensor response (Babinski sign).
- Signs of upper motor neuron pathology include weakness, increased muscle tone, increased reflexes, and a plantar flexor response.[11]

Evidence Discussion (PN-8.1)

MRI of the Brain and/or Spine is indicated to evaluate for amyotrophic lateral sclerosis (ALS)-associated changes as well as evaluation for disorders that may mimic ALS.

Spinal Muscular Atrophy (PN-8.2)

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- Molecular genetic testing is the standard tool for diagnosis for the early consideration in any infant with weakness or hypotonia.
  - MRI is **NOT** supported for diagnosis in children, unless other diseases are being considered. See **Spinal Muscular Atrophy (PEDPN-5.1)** .
- In individuals with adult-onset disease, the differential includes later-onset motor neuron disorders, such as ALS
  - For these conditions, advanced imaging is indicated when upper and lower motor neuron findings are present. For imaging, see **Motor Neuron Disease/ Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**.

Evidence Discussion (PN-8.2)

- Spinal Muscular Atrophy (SMA) is a genetic/hereditary disorder. Molecular genetic testing is the standard tool for diagnosis of Spinal Muscular Atrophy (SMA). MRI is NOT supported for diagnosis of SMA unless other diseases are being considered.

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Fasciculations (PN-8.3)

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Fasciculations are involuntary, irregular movements of muscle caused by activation of a single motor unit that may be secondary to benign or non-benign etiologies.[12]

- **ALL** of the following evaluations are required prior to advanced imaging:
  - **History and physical exam** should include documentation of the following: time course of symptoms, areas of involvement, weakness, and any associated symptoms such as pain, sensory loss, or bowel or bladder dysfunction.
  - **EMG/NCV evaluation**
  - **Laboratory evaluation** (e.g., complete blood count; comprehensive metabolic panel; serum calcium; thyroid function testing; vitamin B12 level; sed rate; ANA; rheumatoid factor; serum protein electrophoresis with immunofixation; Lyme testing; HIV testing; testing for heavy metals; etc.)

  In the setting of clinical concern for radiculopathy, neuromuscular disorders, or muscle disorders, see the following imaging guidelines:
  - **Neuromuscular Junction Disorders (PN-8.4)**
  - **Muscle Diseases (PN-8.5)**
  - **Neck (Cervical Spine) Pain without and with Neurological Features (Including Stenosis) (SP-3.1)**
  - **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)**
- In the presence of upper motor neuron signs (e.g. increased tone; hyperreflexia; presence of Babinski or Hoffman signs) when there is concern for motor neuron disease, including amyotrophic lateral sclerosis (ALS), **ANY** of the following CNS studies are indicated:
  - **Brain** : MRI Brain without contrast (CPT® 70551) **or** MRI Brain without and with contrast (CPT® 70553), **AND/OR**
  - **Cervical Spine** : MRI Cervical Spine without contrast (CPT® 72141) **or** MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR**
  - **Thoracic Spine** : MRI Thoracic Spine without contrast (CPT® 72146) **or** MRI Thoracic Spine without and with contrast (CPT® 72157)
  - See **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**
- **Lumbar Spine** : Lumbar spine imaging is **NOT** indicated unless there is sphincter involvement **or** there is a need to rule out lower motor neuron etiologies in the lower extremities (e.g., lumbar radiculopathy). See the following Spine Imaging Guidelines:
  - **Red Flag Indications (SP-1.2)**

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- **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)**

Evidence Discussion (PN-8.3)

- Fasciculations in isolation are usually benign, especially when they occur repetitively for seconds at a single site and in a single muscle. Fasciculations are more likely to be pathologic if they occur simultaneously in multiple muscles or if they are associated with objective weakness, atrophy, or hyperreflexia.
- Although fasciculations are characteristic of Motor Neuron Disease/Amyotrophic Lateral Sclerosis (MND/ALS) and may occur in other neurological conditions, they are also a very common occurrence in the general population, being noticed by about 70% of normal healthy individuals.
- EMG/NCV evaluation may help differentiate patients with benign fasciculations from those who warrant further investigation.
- Appropriate laboratory evaluation and imaging would depend on suspected etiology.

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Neuromuscular Junction Disorders (PN-8.4)

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Myasthenia Gravis (MG)

- For imaging requests related to ptosis and ocular movements associated with MG, see **Eye Disorders and Visual Loss (HD-32.1)**
- After an established diagnosis of MG or when MG is suspected by a neurologist, rheumatologist, or ophthalmologist, ONE of the following is indicated to assess for MG related thymic disease:[13,16]
  - CT Chest with contrast (CPT® 71260), **OR**
  - CT Chest without contrast (CPT® 71250), **OR**
  - MRI Chest without and with contrast (CPT® 71552), **OR**
  - MRI Chest without contrast (CPT® 71550)
- Repeat of **ANY ONE** of the above imaging studies is indicated if the initial imaging study was negative for **ANY** of the following scenarios:
  - Symptoms of chest mass
  - Rising anti-striated muscle antibody titers
  - Need for pre-operative evaluation (clinical presentation, electro-diagnostic studies, and antibody titers)

Lambert–Eaton Myasthenic Syndrome (LEMS)

Lambert–Eaton Myasthenic Syndrome (LEMS) is associated with malignancies, especially small cell lung cancer.

- For a suspected diagnosis, **ANY** of the following are indicated: CT Chest with contrast (CPT® 71260) **AND/OR** CT Abdomen and Pelvis with contrast (CPT® 74177)[17,18]
  - See **Paraneoplastic Syndromes (ONC-30.3)**
- If initial CT was negative and there is persistent suspicion, **ANY** of the above imaging studies are indicated every 6 months for 2 years from date of initial negative imaging.[17,18]
  - See **Paraneoplastic Syndromes (ONC-30.3)**

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Stiff-Person Syndrome

Stiff-person syndrome is associated with cancers such as, but not limited to, small cell lung cancer, pancreatic neuroendocrine cancer, and breast cancer.[19,20]

- If Stiff-person syndrome is suspected based on clinical findings, **ANY** of the following are indicated:
  - **Abdomen/Pelvis** : CT Abdomen and Pelvis with contrast (CPT® 74177) **or** CT Abdomen and Pelvis without and with contrast (CPT® 74178); **OR** , MRI Abdomen without and with contrast (CPT® 74183) **and** MRI Pelvis without and with contrast (CPT® 72197)
  - **Chest** : CT Chest with contrast (CPT® 71260) **or** CT Chest without contrast (CPT® 71250)
  - **Symptomatic Body Areas** : CT with contrast **or** MRI without and with contrast of any other symptomatic body areas
  - See **Paraneoplastic Syndromes (ONC-30.3)**

Background and Supporting Information

- Myasthenia gravis is an autoimmune disease of the neuromuscular junctions, manifested by fatigable 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).
- Lambert Eaton Myasthenic Syndrome (LEMS) 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).
- LEMS can occur as a paraneoplastic syndrome associated with malignancy (cancer-associated LEMS) or as an autoimmune phenomenon in the absence of malignancy (non-tumor LEMS). Between 50% and 60% of all LEMS cases are associated with malignancy, particularly small cell lung carcinoma (SCLC), although LEMS has been described in individuals with non–small cell and mixed-cell lung carcinomas, neuroendocrine tumors such as prostate cancer, thymoma, and lymphoproliferative disorders.[17]
- 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).

Evidence Discussion (PN-8.4)

- In patients with Myasthenia Gravis, advanced chest imaging with CT or MRI is preferred over X-Ray for the evaluation of thymic disease and for planned thymectomy.

- Lambert-Eaton Myasthenic Syndrome has been associated with malignancies. Initial and repeat imaging with CT of Chest and/or Abdomen and Pelvis are supported to evaluate for associated cancers, especially small cell lung cancer and neuroendocrine tumors.
- Stiff-Person Syndrome has been associated with malignancies. Initial and repeat imaging of the Chest and/or Abdomen and/or Pelvis and/or any symptomatic body area with CT Chest and/or CT Abdomen and Pelvis or MRI Abdomen and/or Pelvis and/or CT or MRI of any symptomatic body area are supported to evaluate for associated cancers, such as small cell lung cancer, pancreatic neuroendocrine cancer, and breast cancer.

Muscle Diseases (PN-8.5)

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- MRI may be helpful in demonstrating abnormalities in muscles that are difficult to examine or not clinically weak and can help distinguish between different types of muscle disease. MRI is also useful in determining sites for muscle biopsy.

Imaging for Muscle Disease		
Disease	**Indication**	**Imaging**
Any Known or Suspected Muscle Disease	To plan muscle biopsy	Typically an affected muscle is imaged. ◦ Upper Extremity: MRI Upper Extremity other than joint without contrast (CPT® 73218); **OR** , MRI Upper Extremity other than joint without and with contrast (CPT® 73220)* **AND/OR** ◦ Lower Extremity: MRI Lower Extremity other than joint without contrast (CPT® 73718); **OR** , MRI Lower Extremity other than joint without and with contrast (CPT® 73720)* * When indication column criteria are met, bilateral studies are supported if requested
Myopathy or Myositis	Additional evaluation after clinical exam, EMG/NCV, **OR** labs	
Inflammatory Muscle Diseases ◦ Dermatomyositis ◦ Polymyositis ◦ Inclusion body myositis	◦ Evaluation of differential diagnosis ◦ Selection of biopsy site ◦ Clinical concern for progression ◦ Treatment monitoring ◦ Detection of occult malignancy	

- For interstitial lung disease associated with inflammatory myopathies, see **Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)** in the Chest Imaging Guidelines.
- For dermatomyositis and polymyositis with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.

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Evidence Discussion (PN-8.5)

- MRI is supported in known or suspected muscle disease to identify involved muscle(s). MRI may highlight muscle edema and pathology at the potential biopsy site.2 MRI is helpful to avoid a false-negative biopsy.
- The ordering of tests should be based on the differential diagnosis arrived at by the history and examination. Laboratory evaluation is often a critical initial step to guide further investigations. Nerve conduction studies and EMG aid in making the diagnosis of neuromuscular disorders and are best conceptualized as extensions of the history and neurologic examination.
- MRI of the affected muscle is supported in the evaluation of patients with suspected Inflammatory Myopathy to help identify a reversible etiology such as Immune-Mediated Necrotizing Myopathy.
- MRI of affected muscle is supported in the diagnosis and follow-up of patients with Inflammatory Myopathies, such as Dermatomyositis, Polymyositis and Inclusion Body Myositis to identify disease specific patterns and evaluate response to treatment.
- Inflammatory Muscle Diseases, including Dermatomyositis and Polymyositis, have been associated with malignancy. Initial and repeat imaging with CT Chest and/or Abdomen and Pelvis are supported to evaluate for associated cancers, such as adenocarcinomas.

Gaucher Disease (Storage Disorders) (PN-8.6)

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Imaging for Gaucher Disease
Initial Imaging
• MRI Lumbar Spine without contrast (CPT® 72148) • Bilateral femurs with MRI Lower Extremity, other than joint, without contrast (CPT® 73718) • MRI Abdomen without contrast (CPT® 74181) • DEXA scan • CT Chest without contrast (CPT® 71250) for individuals with new or worsening pulmonary symptoms
Every 12 months
• To assess treatment response for individuals on enzyme replacement therapy or assess disease progression for individuals in surveillance ◦ MRI Lumbar Spine without contrast (CPT® 72148) ◦ Bilateral femurs with MRI Lower Extremity, other than joint, without contrast (CPT® 73718) ◦ MRI Abdomen without contrast (CPT® 74181) ◦ CT Chest without contrast (CPT® 71250) for individuals with documented pulmonary involvement
New or worsening pulmonary symptoms
• CT Chest without contrast (CPT® 71250)
DEXA scans
• Every 12-24 months until it is normal • Enzyme replacement therapy dose change • Every 3 years
Acute bone pain

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Imaging for Gaucher Disease
• X-ray ◦ MRI of affected areas with and without contrast if x-ray is non-diagnostic or indicates the need for further imaging, such as equivocal for osteonecrosis, infection, or malignancy

- PET/CT imaging is considered not medically necessary in the evaluation of Gaucher disease. ^{18}F-FDG does not reliably detect Gaucher disease in the marrow, and other isotopes are not yet FDA-approved for clinical use.

Background and Supporting Information

- Gaucher disease is group of autosomal recessive inborn errors of metabolism characterized by lack of the enzyme acid ß-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. Diagnosis is established by decreased enzyme activity or genetic testing.
- Three major 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 and death by 2 to 4 years of age; hepatomegaly, splenomegaly, is also present (usually evident by 6 months of age)
  - **Type III** : type I with neurological involvement and slowly progressive disease. Onset may be present before two years of age with survival to the third or fourth decade of life.
- Additionally, there is a perinatal-lethal and a cardiovascular form. The cardiovascular form involves the heart, spleen and eyes. Note that cardiopulmonary complications may be present, with varying frequency and severity, in all subtypes.
- Individuals with Gaucher disease are at risk for osteonecrosis, osteomyelitis, and bony tumors

Evidence Discussion (PN-8.6)

- Initial imaging and lifelong re-imaging is supported due to Gaucher Disease progressive, multisystem involvement.
- Due to bone involvement, including increased risk for Multiple Myeloma, Skeletal X-rays, MRI of Lumbar Spine and MRI Bilateral Femurs are supported. Delineating extent of disease can have positive impact on developing appropriate treatment strategies.

- Dual-energy x-ray absorptiometry (DXA) Scan is supported to evaluate for bone disease including increased risk of osteoporosis. This study helps to predict and avoid pathologic fractures.
- CT is the preferred study for the evaluation of lung parenchyma and is supported to evaluate for pulmonary involvement.
- MRI Abdomen is supported to evaluate for associated visceral disease, such as hepatic, splenic and biliary disease. This Modality has better signal to noise ration and soft tissue contrast helping to make more precise diagnosis.
- The role of PET/CT imaging in Gaucher Disease is yet to be established. In the absence of malignancy, PET/CT is not considered medically necessary in the evaluation of Gaucher disease. Unnecessary use of this study would expose the patient to excess radiation and noncontributory imaging.

References (PN-8)

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Peripheral Nerve Sheath Tumors (PNST) (PN-9)

Guideline Page

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Peripheral Nerve Sheath Tumors (PNST) (PN-9.1)

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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 Peripheral Nerve Sheath Tumors (PNST) is suspected, the following advanced imaging is indicated:	
Suspected Lesion/Indication	**Imaging**
Vestibular Schwannoma	• MRI Brain without and with contrast (CPT® 70553) See **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)** in the Head Imaging Guidelines
Paraspinal Neurofibroma	**ANY** of the following imaging: • MRI Cervical Spine without and with contrast (CPT® 72156), **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157), **AND/OR** • MRI Lumbar Spine without and with contrast (CPT® 72158)
Neurofibroma of the Limb or Torso (other than Paraspinal)	• MRI without and with contrast or without contrast of the area of interest after plain x-ray* ◦ See **Soft Tissue Mass (MS-10.1)** in the Musculoskeletal Imaging Guidelines *Plain x-ray is not required in an individual with a cancer predisposition syndrome. • See **Screening Imaging in Cancer Predisposition Syndromes (PEDONC-2)** in the Pediatric and Special Populations Oncology Imaging Guidelines

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Routine follow-up imaging is NOT indicated *except in the following scenarios:*	
Suspected Lesion/Indication	**Imaging**
New symptoms or neurological findings	• MRI without and with contrast of the known body area containing PNST
Post-operatively for ANY of the following scenarios: • At the discretion of or in consultation with the surgeon; • If the tumor was not completely removed and the imaging is requested to re-establish baseline	• MRI without and with contrast of the known body area containing PNST or from which PNST was removed
Request for metastatic work-up when malignant transformation is known or suspected	**ANY** of the following imaging: • CT Chest with contrast (CPT® 71260) **AND/OR** • CT Abdomen with contrast (CPT® 74160)

- For guidelines related to known malignancies in individuals with Neurofibromatosis 1 (NF1), see the appropriate imaging guideline for the specific cancer type.

Background and Supporting Information

- The role of PET imaging in Peripheral Nerve Sheath Tumors is not yet well established.[8]
- Malignant transformation may be present in approximately 5% of Peripheral Nerve Sheath Tumors.

Evidence Discussion (PN-9.1)

- Peripheral Nerve Sheath Tumors (PNSTs) may arise from any body region. PNSTs are susceptible to malignant transformation. Therefore, MRI of the known or suspected body region is supported for evaluation.
- MRI is the preferred imaging modality for soft tissue tumors, such as PNSTs, and is a relatively safe imaging modality since radiation exposure is not involved. The role of PET imaging in Peripheral Nerve Sheath Tumors is not yet well established. Otherwise, PET imaging in this clinical scenario would not add any clinical value and would unnecessarily expose patients to radiation.

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