Cigna Medical Coverage Policies – Radiology Peripheral Nerve Disorders Imaging Guidelines

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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 <u>Cigna CPT</u> <u>code list</u> for the current list of high-tech imaging procedures that eviCore reviews for Cigna.

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Abbrevia	tions for Peripheral Nerve Disorders Imaging Guidelines
AIDS	Acquired Immunodeficiency Syndrome
ALS	Amyotrophic Lateral Sclerosis
CIDP	Chronic Inflammatory Demyelinating Polyneuropathy
CNS	central nervous system
СРК	creatinine phosphokinase
СТ	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
тоѕ	Thoracic Outlet Syndrome

General Guidelines (PN-1)

General Guidelines (PN-1.0)

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, electromyography and 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 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 individual'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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Focal Neuropathy (PN-2)

Focal Neuropathy (PN-2.1)

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Carpal Tunnel Syndrome	YES	Ultrasound Wrist and/or MRI Wrist without contrast (CPT® 73221) to estimate size of the carpal tunnel and diameter of the median nerve in the evaluation and confirmation of carpal tunnel syndrome pre-operatively when EMG findings are equivocal and clinical findings are uncertain.
		 SeeNeck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3) in the Spine Imaging Guidelines.
Ulnar Neuropathy	Ultrasound may be used for evaluation is not required prior to MRI. MRI Upper Extremity Joint (Elbow or Wrist) without contrast (CPT® 73221) or MRI Upper Extremity Non Joint (Forearm or Hand) without contrast (CPT® 73218) after EN surgical consideration	
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.

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
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.
palsy, hip or pelvic	fractures, or hip repla entrapment of the sci	ne gluteal area with hematoma, injection cement (arthroplasty) and rarely Piriformis iatic nerve at the sciatic notch in the pelvis by
Femoral Neuropathy	NO	MRI Pelvis without contrast (CPT® 72195) performed in the evaluation of these entities.
-	-	as a complication of pelvic surgery in women toneal bleeding, or as a mononeuropathy in
Meralgia Paresthetica	NO	 MRI Pelvis without contrast (CPT® 72195) may be performed for ANY of the following: Pre-operative Cases of diagnostic uncertainty
		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.
it exits the pelvis u leg), and is usually	nder the inguinal ligan	loss in the lateral femoral cutaneous nerve as nent (lateral thigh without extension into lower ed on a careful history and physical exam. icult 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.

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

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Polyneuropathy (PN-3)

Polyneuropathy (PN-3.1)

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>Brachial Plexus (PN-4.1)</u> , <u>Lumbar and Lumbosacral Plexus (PN-5.1)</u> , and <u>Muscle Diseases (PN-6.2)</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 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	

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Brachial Plexus (PN-4)

Brachial Plexus (PN-4.1)

- 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:8-12
 - Malignant infiltration (EMG not required)
 - Radiation plexitis to rule out malignant infiltration (EMG not required)
 - 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
 - MRI Cervical Spine if radiculopathy.
 - See <u>Neck (Cervical Spine) Pain Without/With Neurological Features</u>
 (Including Stenosis) and Trauma (SP-3) in the Spine Imaging Guidelines
 - Traumatic injury (MRI Cervical Spine CPT® 72141)¹³
 - Neurogenic Thoracic Outlet Syndrome (TOS)¹⁰
 - 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 <u>PET Imaging in Oncology (ONC-1.4)</u> in the Oncology Imaging Guidelines

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Lumbar and Lumbosacral Plexus (PN-5)

<u>Lumbar and Lumbosacral Plexus (PN-5.1)</u>

- 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) 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 postsurgical)
 - 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 <u>PET Imaging in Oncology (ONC-1.4)</u> in the Oncology Imaging Guidelines

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Neuromuscular Junction Disorders (PN-6.1)

- 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) if there is concern regarding adverse effects of contrast in individuals 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.

Background and Supporting Information

- 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)

Muscle Diseases (PN-6.2)

- 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 affected muscles is imaged (when criteria are met imaging for bilateral studies is appropriate) for:
 - Additional evaluation of myopathy or myositis (based on clinical exam and adjunct testing with EMG/NCV and labs)
 - To plan muscle biopsy

- See <u>Interstitial Lung Disease (ILD)/Diffuse Lung Disease (DLD) (CH-11.1)</u> for interstitial lung disease associated with inflammatory myopathies
- Inflammatory Muscle Diseases:
 - These include dermatomyositis, polymyositis, and sporadic inclusion body myositis. MRI of a single site is indicated in these disorders for the following purposes:
 - Selection of biopsy site
 - Clinical concern for progression
 - Treatment monitoring
 - Detection of occult malignancy
- All cases with dermatomyositis and polymyositis can undergo search for occult neoplasm See <u>Paraneoplastic Syndromes (ONC–30.3)</u> in the Oncology Imaging Guidelines

Background and Supporting Information

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.

Gaucher Disease (Storage Disorders) (PN-6.3)

- 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. 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.

Imaging for Gaucher Disease

Initial Imaging

- ➤ MRI Lumbar Spine without contrast (CPT® 72148)
- ➤ MRI Bilateral Femurs without contrast (CPT® 73718)
- ➤ MRI Abdomen without contrast (CPT® 74181)
- DXA 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)
 - MRI Bilateral Femurs 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)

DXA scans

- Every 12-24 months until it is normal
- Enzyme replacement therapy dose change
- Every 3 years

Acute bone pain

- X-ray
 - MRI of affected areas with and without contrast 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.

Background and Supporting Inforrmation

Individuals with Gaucher disease are at risk for osteonecrosis, osteomyelitis, and bony tumors

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Magnetic Resonance Neurography (MRN) (PN-7)

Magnetic Resonance Neurography (MRN) (PN-7.1)

➤ MRN (Unlisted MRI procedure code (CPT® 76498)) - 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

Reference

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Neuromuscular Disorders (PN-8)	
Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-	
8.1)	20
Spinal Muscular Atrophy (PN-8.2)	20
Fasciculations (PN-8.3)	20

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

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

Spinal Muscular Atrophy (PN-8.2)

- 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 diagnosis in children, unless other diseases are being considered.
 - In individuals with adult onset disease, the differential includes later-onset motor neuron disorders such as ALS. For these conditions, advanced imaging may be approved, MRI Brain and Spinal Cord imaging, per <u>Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)</u> when upper and lower motor neuron findings are present.

Fasciculations (PN-8.3)

- 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 <u>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 Spine Imaging Guidelines).</u>
 - 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 guideline requirements under neuromuscular or muscle disorders, Neuromuscular Junction Disorders (PN-6.1) and Muscle Diseases (PN-6.2) respectively, should follow requirements under Spine Imaging Guidelines, and this includes Lumbar Radiculopathies, See 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 Spine Imaging Guidelines.

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

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

- 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 Schwannomas See <u>Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)</u> 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 is known or suspected. (Malignant transformation may be present in approximately 5% of Peripheral Nerve Sheath Tumors.)
 This can include a metastatic work-up with CT Chest and Abdomen with contrast (CPT® 71260 and CPT® 74160)
- For guidelines related to known malignancies in individuals with NF1, see the appropriate imaging guideline for the specific cancer type.

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