

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 [Cigna CPT code list](#) for the current list of high-tech imaging procedures that eviCore reviews for Cigna.

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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

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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 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 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.

References

1. Bowen BC, Maravilla KR, Saraf-Lavi. Magnetic Resonance Imaging of the Peripheral Nervous System. In Latchaw RE, Kucharczyk J, Moseley ME. Imaging of the Nervous System. Diagnostic and Therapeutic Applications. Vol 2, Mosby, Philadelphia, 2005, pp.1479-1497.
2. Walker WO. Ultrasonography in peripheral nervous system diagnosis. Continuum. 2017 Oct; 23 (5, Peripheral Nerve and Motor Neuron Disorders):1276-1294.
3. Ohana M, Moser T, Moussaoui A, et al. Current and future imaging of the peripheral nervous system. Diagnostic and Interventional Imaging. 2014;95(1):17-26.
4. Stoll G, Bendszus M, Perez J, et al. Magnetic resonance imaging of the peripheral nervous system. J Neurol. 2009 Jul;256(7):1043-51.
5. Stoll G, Wilder-Smith E, and Bendszus M. Imaging of the peripheral nervous system. Handb Clin Neurol. 2013;115:137-153.
6. Kim S, Choi J-Y, Huh Y-M, et al. Role of magnetic resonance imaging in entrapment and compressive neuropathy—what, where, and how to see the peripheral nerves on the musculoskeletal magnetic resonance image: part 1. Overview and lower extremity. Eur Radiol. 2007 Jan;17(1):139-149.
7. Russell JA. General Approach to Peripheral Nerve Disorders. CONTINUUM: Lifelong Learning in Neurology. 2017;23(5):1241-1262. doi:10.1212/con.0000000000000519.
8. Sheikh, KA. Guillain-Barré Syndrome. CONTINUUM: Lifelong Learning in Neurology. 2020;26(5):1184-1204. doi:10.1212/con.0000000000000929.
9. Kassardjian CD, Desai U, Narayanaswami P. Practical Guidance for Managing EMG Requests and Testing during the COVID-19 Pandemic. Muscle & Nerve. Published online April 11, 2020. doi:10.1002/mus.26891
10. London ZN. A Structured Approach to the Diagnosis of Peripheral Nervous System Disorders. CONTINUUM: Lifelong Learning in Neurology. 2020;26(5):1130-1160. doi:10.1212/con.0000000000000922.

PN-2: Focal Neuropathy

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Carpal Tunnel Syndrome	YES	<ul style="list-style-type: none"> ➤ Ultrasound Wrist and/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 <u>MS-21: Wrist in the Musculoskeletal Imaging Guidelines</u> and <u>SP-3: Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma in the Spine Imaging Guidelines.</u>
Ulnar Neuropathy	YES	<ul style="list-style-type: none"> ➤ Ultrasound may be used for evaluation, but 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 EMG for surgical consideration
Radial Neuropathy	YES	<ul style="list-style-type: none"> ➤ 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.
<p>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.</p>		

Focal Disorder	EMG/NCV Initially?	Advanced Imaging
Sciatic Neuropathy	YES	<ul style="list-style-type: none"> ➤ 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.
<p>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.</p>		
Femoral Neuropathy	NO	<ul style="list-style-type: none"> ➤ MRI Pelvis without contrast (CPT® 72195) may be performed in the evaluation of these entities.
<p>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.</p>		
Meralgia Paresthetica	NO	<ul style="list-style-type: none"> ➤ MRI Pelvis without contrast (CPT® 72195) may be performed for ANY of the following: <ul style="list-style-type: none"> ◆ 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.
<p>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.</p>		
Peroneal Neuropathy	YES	<ul style="list-style-type: none"> ➤ 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>MS-27: Foot (Tarsal Tunnel Syndrome)</u> in the Musculoskeletal Imaging Guidelines.

References

1. Andreisek G, Crook DW, Burg D, et al. Peripheral neuropathies of the median, radial, and ulnar nerves: MR imaging features. *RadioGraphics*. 2006 Sep-Oct;26(5):1267-1287.
2. Iverson DJ. MRI detection of cysts of the knee causing common peroneal neuropathy. *Neurology*. 2005 Dec 13;65(11):1829-1831.
3. Cartwright MS, Walker FO. Neuromuscular ultrasound in common entrapment neuropathies. *Muscle & Nerve*. 2013 Sep 2;48(5):696-704.
4. Linda DD, Harish S, Stewart BG, et al. Multimodality imaging of peripheral neuropathies of the upper limb and brachial plexus. *RadioGraphics*. 2010 Sep;30(5):1373-1400.
5. Hobson-Webb LD and Juel VC. Common Entrapment Neuropathies. *Continuum*. 2017 Apr;23(2):487-511.
6. Tsvigoulis G and Alexandrov AV. Ultrasound in neurology. *Continuum*. 2016 Oct;22(5)Neuroimaging:1655-1677.

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	

References

- Anders HJ, Goebel FD. Cytomegalovirus polyradiculopathy in patients with AIDS. Clin Infect Dis. 1998 Aug 27;27(2):345-352.
- Duggins AJ, McLoed JG, Pollard JD, et al. Spinal root and plexus hypertrophy in chronic inflammatory demyelinating polyneuropathy. Brain. 1999 July 1;122(7):1383-1390.
- Amato AA, Barohn RJ, Katz JS, et al. Clinical spectrum of chronic acquired demyelinating polyneuropathies. Muscle & Nerve. 2001 Mar;24(3):311-324.
- Darnell RB, Posner JB. Paraneoplastic Syndromes Involving the Nervous System. N Engl J Med. 2003;349:1543-1554.
- Antoine JC, Bouhour F, Camdessanche JP. [18F] fluorodeoxyglucose positron emission tomography in the diagnosis of cancer in patients with paraneoplastic neurological syndrome and anti-Hu antibodies. Ann Neurol. 2000 July;48(1):105-108.
- Maravilla KR, Bowen BC. Imaging of the peripheral nervous system: evaluation of peripheral neuropathy and plexopathy. AJNR Am J Neuroradiol. 1998;19(6):1011-1023.
- Ohana M, Moser T, Moussaoui A, et al. Current and future imaging of the peripheral nervous system. Diagnostic and Interventional Imaging. 2014;95(1):17-26. doi:10.1016/j.diii.2013.05.008.

PN-4: Brachial Plexus

- 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

References

1. Adkins MC, Wittenberg KH. MR imaging of nontraumatic brachial plexopathies: frequency and spectrum of findings. *RadioGraphics*. 2000 July;20(4):1023-1032.
2. Bykowski J, Aulino JM, Berger KL, et al. (2016). ACR Appropriateness Criteria[®] Plexopathy. American College of Radiology (ACR).
3. Van Es HW. MRI of the brachial plexus. *Eur Radiol*. 2001 Jan;11(2):325-336.
4. Foley KM, Kori SH, Posner JB. Brachial plexus lesions in patients with cancer: 100 cases. *Neurology*. 1981 Jan;31(1):45-50.
5. Cascino TL, Harper CM, Thomas JE, et al. Distinction between neoplastic and radiation-induced brachial plexopathy, with emphasis on the role of EMG. *Neurology*. 1989 April;39(4):502-506.
6. Husband JE, MacVicar AD, Padhani AR, et al. Symptomatic brachial plexopathy following treatment for breast cancer: Utility of MR imaging with surface-coil techniques. *Radiology*. 2000 March;214(3):837-842.
7. McDonald TJ, Miller JD, Pruitt S. Acute brachial plexus neuritis: an uncommon cause of shoulder pain. *Am Fam Physician*. 2000 Nov 1;62(9):2067-2072.
8. Ohana M, Moser T, Moussaoui A, et al. Current and future imaging of the peripheral nervous system. *Diagnostic and Interventional Imaging*. 2014;95(1):17-26. doi:10.1016/j.diii.2013.05.008.
9. American College of Radiology ACR Appropriateness Criteria[®] Plexopathy. Revised 2016. <https://acsearch.acr.org/docs/69487/Narrative/>.

PN-5: Lumbar and Lumbosacral Plexus

- 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 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 Oncology** in the Oncology Imaging Guidelines

References

1. Brejt N, Berry J, Nisbet A, et al. Pelvic radiculopathies, lumbosacral plexopathies, and neuropathies in oncologic disease: A multidisciplinary approach to a diagnostic challenge. *Cancer Imaging*. 2013 Dec 30;13(4):591-601.
2. McDonald JW, Sadwosky C. Spinal-cord injury. *The Lancet*. 2002 Feb 2;359(9304):417-425.
3. American College of Radiology ACR Appropriateness Criteria® Plexopathy. Revised 2016. <https://acsearch.acr.org/docs/69487/Narrative/>.
4. Maravilla KR, Bowen BC. Imaging of the peripheral nervous system: evaluation of peripheral neuropathy and plexopathy. *AJNR Am J Neuroradiol*. 1998;19(6):1011-1023.

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 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)

PN-6.2: Muscle Diseases

- 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 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 **CH-11.1: Interstitial Disease** 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 **ONC-30.3: Paraneoplastic Syndromes** 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.

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

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 patients with new or worsening pulmonary symptoms

Every 12 months

- To assess treatment response for patients on enzyme replacement therapy or assess disease progression for patients 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 patients 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 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.

Background and Supporting Information

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

References

1. Darnell R, Posner J. Paraneoplastic syndromes involving the nervous system. *N Engl J Med*. 2003 Oct;349:1543-1554.
2. Schweitzer M, Fort J. Cost-effectiveness of MR imaging in evaluating polymyositis. *Am J Roentgenol*. 1995;165:1469-1471.
3. Adams E, Chow C, Premkumar A, Plotz P. The idiopathic inflammatory myopathies: spectrum of MR imaging findings. *RadioGraphics*. 1995;15(3):563-574.
4. Park J, Olsen N. Utility of magnetic resonance imaging in the evaluation of patients with inflammatory myopathies. *Curr Rheumatol Reports*. 2001 Aug;3(4):334-345.
5. Sekul E, Chow C, Dalakas M. Magnetic resonance imaging of the forearm as a diagnostic aid in patients with sporadic inclusion body myositis. *Neurolog*. 1997 April;48(4):863-866.
6. Lundberg I, Chung Y. Treatment and investigation of idiopathic inflammatory myopathies. *Rheumatology*. 2000 Jan;39(1):7-17.
7. Park J, Olsen N. Utility of magnetic resonance imaging in the evaluation of patients with inflammatory myopathies. *Curr Rheumatol Reports*. 2001 Aug;3(4):334-345.
8. Hill C, Zhang Y, Sigurgeirsson B, et al. Frequency of specific cancer types in dermatomyositis and polymyositis: a population-based study. *Lancet*. 2001 Jan 13;357(9250):96-100.
9. Maas M, Poll L, Terk M. Imaging and quantifying skeletal involvement in Gaucher disease. *B J Radiol*. 2002;75 suppl1:A13-A24.
10. Giraldo P, Pocovi M, Perez-Calvo J, et al. Report of the Spanish Gaucher's disease registry: clinical and genetic characteristics. *Haematologica*. 2000 Jan;85:792-799.
11. Rosow et al. The Role of Electrodiagnostic Testing, Imaging, and Muscle Biopsy in the Investigation of Muscle Disease. *Continuum*. 2016 Dec;22(6):1787-1802.
12. Somashekar DK, Davenport MS, Cohan RH, et al. Effect of intravenous low-osmolality iodinated contrast media on patients with myasthenia gravis. *Radiology*. 2013 Jun;267(3):727-734.
13. Pastores GM, Hughes DA. Gaucher Disease. *GeneReviews*TM [Internet] Adam MP, Ardinger HH, Pagon RA, et al. eds. Last Revision: June 21, 2018. <https://www.ncbi.nlm.nih.gov/books/NBK1269/>.
14. Degnan AJ, Ho-Fung VM, Ahrens-Nicklas RC, et al. Imaging of non-neuronopathic Gaucher disease: recent advances in quantitative imaging and comprehensive assessment of disease involvement. *Insights into Imaging*. 2019;10(1). doi:10.1186/s13244-019-0743-5.
15. Arnold WD, Kassab D and Kissel JT. Spinal Muscular Atrophy: Diagnosis and Management in a New Therapeutic Era. *Muscle Nerve*. 2015 February ; 51(2): 157–167. doi:10.1002/mus.24497.
16. Pastores GM, Hughes DA. Gaucher Disease. 2000 Jul 27 [Updated 2018 Jun 21]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. *GeneReviews*[®] [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2021. Gaucher Disease - *GeneReviews*[®] - NCBI Bookshelf (nih.gov).
17. Kaplan P, Baris H, De Meirleir L, et al. Revised recommendations for the management of Gaucher disease in children. *European Journal of Pediatrics*. 2012;172(4):447-458. doi:10.1007/s00431-012-1771-z.

PN-7: Magnetic Resonance Neurography (MRN)

- 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

1. Noguero TM, Barousse R, Cabrera MG, Socolovsky M, Bencardino JT, Luna A. Functional MR Neurography in Evaluation of Peripheral Nerve Trauma and Postsurgical Assessment. *RadioGraphics*. 2019;39(2):427-446. doi:10.1148/rg.2019180112.

PN-8: Neuromuscular Disorders

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PN-8.1: Amyotrophic Lateral Sclerosis (ALS)

- For an established patient with ALS, a 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 diagnosis in children, unless other diseases are being considered.
- In patients 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 **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** in the Spine Imaging Guidelines).
- ◆ 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, **PN-6.1: Neuromuscular Junction Disorders** and **PN-6.2: Muscle Diseases** respectively, should follow requirements under Spine Imaging Guidelines, and this includes Lumbar Radiculopathies, See **SP-6.1: Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain** in the Spine Imaging Guidelines.

References

1. Kollwee K, Korner S, Dengler R, et al. Magnetic resonance imaging in amyotrophic lateral sclerosis. *Neurology Research International*. 2012;v2012.
2. Filippi M, Agosta F, Abrahams S, et al. EFNS guidelines on the use of neuroimaging in the management of motor neuron diseases. *Eur J Neurol*. 2010 Apr;17(4):526-e20.
3. Agosta F, Spinelli EG, Filippi M. Neuroimaging in amyotrophic lateral sclerosis: current and emerging uses. *Expert Review of Neurotherapeutics*. 2018;18(5):395-406. doi:10.1080/14737175.2018.1463160.
4. Arnold WD, Kassar D and Kissel JT. Spinal Muscular Atrophy: Diagnosis and Management in a New Therapeutic Era. *Muscle Nerve*. 2015 February ; 51(2): 157–167. doi:10.1002/mus.24497.
5. Garg N, Park SB, Vucic S, et al. Differentiating Lower Motor Neuron Syndromes. *J Neurol Neurosurg Psychiatry* 2017;88:474–483.
6. Shefner JM, et al. A Proposal For New Diagnostic Criteria for ALS. *Clinical Neurophysiology* 131 (2020) 1975–1978. <https://doi.org/10.1016/j.clinph.2020.04.005>
7. Glascock J, et al. Treatment Algorithm for Infants Diagnosed with Spinal Muscular Atrophy through Newborn Screening. *Research Report. Journal of Neuromuscular Diseases* 5 (2018) 145–158. doi: 10.3233/JND-1803048.
8. Silveira-Moriyama Laura and Paciorkowski AR. Genetic Diagnostics for Neurologists. *CONTINUUM (MINNEAP MINN)* 2018;24(1, CHILD NEUROLOGY):18–36.
9. Hatcher-Martin, JM ET al. Telemedicine in Neurology. *Telemedicine Work Group of the American Academy of Neurology Update. Neurology*® 2020;94:30-38. doi:10.1212/WNL.00000000000008708
10. Prior TW, Leach ME and Finanger E. Spinal Muscular Atrophy. *Gene Reviews*. Created: February 24, 2000; Updated: November 14, 2019. Copyright © 1993-2020, University of Washington, Seattle. GeneReviews is a registered trademark of the University of Washington, Seattle. All rights reserved.
11. Filippakis A, et al. A Prospective Study of Benign Fasciculation Syndrome and Anxiety. *Short Reports. Muscle & Nerve*. 58:582-852, 2018.

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 Schwannomas 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 is known or suspected. (Malignant transformation may be present in approximately 5% of Peripheral Nerve Sheath Tumors.) This may include a metastatic work-up with CT Chest and Abdomen with contrast (CPT® 71260 and CPT® 74160)
- For guidelines related to known malignancies in patients with NF1, see the appropriate imaging guideline for the specific cancer type.

References

1. Ahlawat S, Blakeley JO, Langmead S, Belzberg AJ, Fayad LM. Current status and recommendations for imaging in neurofibromatosis type 1, neurofibromatosis type 2, and schwannomatosis. *Skeletal Radiology*. 2019;49(2):199-219. doi:10.1007/s00256-019-03290-1.
2. Soldatos T, Fisher S, Karri S, Ramzi A, Sharma R, Chhabra A. Advanced MR Imaging of Peripheral Nerve Sheath Tumors Including Diffusion Imaging. *Seminars in Musculoskeletal Radiology*. 2015;19(02):179-190. doi:10.1055/s-0035-1546823.
3. Zhang J, Li Y, Zhao Y, Qiao J. CT and MRI of superficial solid tumors. *Quantitative Imaging in Medicine and Surgery*. 2018;8(2):232-251. doi:10.21037/qims.2018.03.03.
4. Rosser T. Neurocutaneous Disorders. *CONTINUUM: Lifelong Learning in Neurology*. 2018;24(1):96-129. doi:10.1212/con.0000000000000562.
5. Dare AJ, Gupta AA, Thipphavong S, Miettinen M, Gladdy RA. Abdominal neoplastic manifestations of neurofibromatosis type 1. *Neuro-Oncology Advances*. 2020;2(Supplement_1):i124-i133. doi:10.1093/noajnl/vdaa032.
6. Bruno F, Arrigoni F, Mariani S, et al. Advanced magnetic resonance imaging (MRI) of soft tissue tumors: techniques and applications. *Radiol Med*. 2019;124(4):243-252. doi:10.1007/s11547-019-01035-7.
7. Ruggieri M, Polizzi A, Marceca GP, Catanzaro S, Praticò AD, Di Rocco C. Introduction to phacomatoses (neurocutaneous disorders) in childhood. *Child's Nervous System*. 2020;36(10):2229-2268. doi:10.1007/s00381-020-04758-5.

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