Cigna Medical Coverage Policies – Radiology
Peripheral Nerve Disorders (PND)
Imaging Guidelines

Effective February 01, 2024

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.

CPT® (Current Procedural Terminology) is a registered trademark of the American Medical Association (AMA). CPT® five digit codes, nomenclature and other data are copyright 2023 American Medical Association. All Rights Reserved. No fee schedules, basic units, relative values or related listings are included in the CPT® book. AMA does not directly or indirectly practice medicine or dispense medical services. AMA assumes no liability for the data contained herein or not contained herein.

Table of Contents

Guideline Page

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines (PN-1)

Abbreviations for Peripheral Nerve Disorders Imaging Guidelines

PN.GG.Abbreviations.A
v1.0.2024

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

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines (PN-1.0)

PN.GG.0001.0.A
v1.0.2024

- 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 **Peripheral Nerve Disorders (PND) 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.

©2023 eviCore healthcare. All Rights Reserved.

References (PN-1)

v1.0.2024

1. Walker FO. Ultrasonography in Peripheral Nervous System Diagnosis. *Continuum (Minneap Minn)*. 2017;23(5):1276-1294. doi: 10.1212/con.0000000000000522.
2. Ohana M, Moser T, Moussaouï A, et al. Current and future imaging of the peripheral nervous system. *Diagn Interv Imaging*. 2014;95(1):17-26. doi: 10.1016/j.diii.2013.05.008.
3. Stoll G, Wilder-Smith E, Bendszus M. Imaging of the peripheral nervous system. *Handb Clin Neurol*. Published online 2013:137-153. doi: 10.1016/b978-0-444-52902-2.00008-4.
4. Russell JA. General Approach to Peripheral Nerve Disorders. *Continuum (Minneap Minn)*. 2017;23(5):1241-1262. doi: 10.1212/con.0000000000000519.
5. 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.
6. London ZN. A Structured Approach to the Diagnosis of Peripheral Nervous System Disorders. *Continuum (Minneap Minn)*. 2020;26(5):1130-1160. doi: 10.1212/con.0000000000000922.

©2023 eviCore healthcare. All Rights Reserved.

Focal Neuropathy (PN-2)

Focal Neuropathy (PN-2.1)

PN.FN.0002.1.A
v1.0.2024

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 Wrist without contrast (CPT® 73221) is indicated. • 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 Non Joint (Forearm or Hand) without contrast (CPT® 73218)
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 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.		

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Focal Disorder	EMG/NCV Initially?	Advanced Imaging

Pudendal Neuropathy (7-12)	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 from 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.

©2023 eviCore healthcare. All Rights Reserved.

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.		
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
		• 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 easily diagnosed based on a careful history and physical exam. EMG/NCV testing is often technically difficult and not required.		
Peroneal Neuropathy	YES	• MRI Knee without contrast (CPT® 73721) **OR** MRI Lower Extremity other than joint without contrast (CPT® 73718) in severe cases when surgery is considered.
Tarsal Tunnel Syndrome	N/A	• See **Foot (Tarsal Tunnel Syndrome) (MS-27)** in the Musculoskeletal Imaging Guidelines.

©2023 eviCore healthcare. All Rights Reserved.

References (PN-2)

v1.0.2024

1. Andreisek G, Crook DW, Burg D, Marincek B, Weishaupt D. Peripheral Neuropathies of the Median, Radial, and Ulnar Nerves: MR Imaging Features. *RadioGraphics*. 2006;26(5):1267-1287 doi: 10.1148/rg.265055712.
2. Linda DD, Harish S, Stewart BG, Finlay K, Parasu N, Rebello RP. Multimodality Imaging of Peripheral Neuropathies of the Upper Limb and Brachial Plexus. *RadioGraphics*. 2010;30(5):1373-1400. doi: 10.1148/rg.305095169.
3. Hobson-Webb LD, Juel VC. Common Entrapment Neuropathies. *Continuum (Minneap Minn)* . 2017;23(2):487-511. doi: 10.1212/con.0000000000000452
4. Tsivgoulis G, Alexandrov AV. Ultrasound in Neurology. Continuum (Minneap Minn). 2016;22(5):1655-1677. doi: 10.1212/con.0000000000000374.
5. Buchanan BK, Maini K, Varacallo M. Radial Nerve Entrapment. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; September 4, 2022. https://pubmed.ncbi.nlm.nih.gov/28613749/.
6. Kalia V, Jacobson JA. Imaging of Peripheral Nerves of the Upper Extremity. *Radiol Clin N Am*. 2019;57(5):1063-1071. doi: 10.1016/j.rcl.2019.04.001.
7. Kaur J, Leslie SW, Singh P. Pudendal Nerve Entrapment Syndrome. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; May 30, 2023.
8. Ploteau S. Cardaillac C, Perrouin-Verbe MA, Riant T, Labat JJ. Pudendal Neuralgia Due to Pudendal Nerve Entrapment: Warning Signs Observed in Two Cases and Review of the Literature. *Pain Physician*. 2016; 19(3): E449-E454.
9. Aoun F, Alkassis M, Tayeh GA, et al. Sexual dysfunction due to pudendal neuralgia: a systematic review. *Transl Androl Urol*. 2021;10(6):2500-2511. doi: 10.21037/tau-21-13.
10. Luesma MJ, Galé I, Fernando J. Diagnostic and therapeutic algorithm for pudendal nerve entrapment syndrome. Algoritmo diagnóstico y terapéutico del síndrome de atrapamiento del nervio pudendo. *Med Clin (Barc)*. 2021;157(2):71-78. doi: 10.1016/j.medcli.2021.02.012.
11. Labat JJ, Riant T, Robert R, Amarenco G, Lefaucheur JP, Rigaud J. Diagnostic criteria for pudendal neuralgia by pudendal nerve entrapment (Nantes criteria). *Neurourol Urodyn*. 2008;27(4):306-310. doi: 10.1002/nau.20505.
12. Levesque A, Bautrant E, Quistrebert V, et al. Recommendations on the management of pudendal nerve entrapment syndrome: A formalised expert consensus. *Eur J Pain*. 2022;26(1):7-17. doi:10.1002/ejp.1861.

©2023 eviCore healthcare. All Rights Reserved.

Polyneuropathy (PN-3)

Polyneuropathy (PN-3.1)

PN.PN.0003.1.A
v1.0.2024

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)	Examples: Guillain-Barré syndrome and Lyme disease

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments

AIDS-Related Cytomegalovirus 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)
			• Urinary retention and a clinically confusing picture in the legs.
			• For myelopathic signs and symptoms, see **Myelopathy (SP-7.1)**.

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	YES	MRI Lumbar Spine without and with contrast (CPT® 72158) if uncertain following EMG/NCV. • See **Brachial Plexus (PN-4.1)**, **Lumbar and Lumbosacral Plexus (PN-5.1)**, and **Muscle Diseases (PN-6.2)**	

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Poly-Disorder	EMG/NCV Initially?	Advanced Imaging	Comments
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 Spine) 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]

©2023 eviCore healthcare. All Rights Reserved.

References (PN-3)

v1.0.2024

1. Harrison TB, Smith B. Neuromuscular Manifestations of HIV/AIDS. *J Clin Neuromuscul Dis*. 2011;13(2):68-84. doi: 10.1097/cnd.0b013e318221256f.
2. Eftimov F, Lucke IM, Querol LA, Rajabally YA, Verhamme C. Diagnostic challenges in chronic inflammatory demyelinating polyradiculoneuropathy. *Brain*. 2020;143(11):3214-3224. doi: 10.1093/brain/awaa265.
3. Saperstein D. Chronic Acquired Demyelinating Polyneuropathies. *Semin Neurol*. 2008;28(2):168-184. doi: 10.1055/s-2008-1062268.
4. Binks S, Uy C, Honnorat J, Irani SR. Paraneoplastic neurological syndromes: a practical approach to diagnosis and management. *Pract Neurol*. 2022;22(1):19-31. doi: 10.1136/practneurol-2021-003073.
5. Stoll G, Wilder-Smith E, Bendszus M. Imaging of the peripheral nervous system. *Handb Clin Neurol*. Published online 2013:137-153. doi: 10.1016/b978-0-444-52902-2.00008-4.
6. London ZN. A Structured Approach to the Diagnosis of Peripheral Nervous System Disorders. *Continuum (Minneap Minn)*. 2020;26(5):1130-1160. doi: 10.1212/con.0000000000000922.

©2023 eviCore healthcare. All Rights Reserved.

Brachial Plexus (PN-4)

Brachial Plexus (PN-4.1)

PN.BP.0004.1.A
v1.0.2024

- 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) **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**	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)	
Brachial plexitis (Parsonage-Turner Syndrome or painful brachial amyotrophy) **Traumatic injury[13]**	• Any **ONE** of the above studies **AND** • If concern for radiculopathy, 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**.

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 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)

©2023 eviCore healthcare. All Rights Reserved.

- **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.

©2023 eviCore healthcare. All Rights Reserved.

References (PN-4)

v1.0.2024

1. Wittenberg KH, Adkins MC. MR Imaging of Nontraumatic Brachial Plexopathies: Frequency and Spectrum of Findings. *RadioGraphics*. 2000;20(4):1023-1032. doi: 10.1148/radiographics.20.4.g00jl091023
2. Bykowski J, Aulino JM, Berger KL, et al. ACR Appropriateness Criteria® Plexopathy. *J Am Coll Radiol*. 2017;14(5S):S225-S233. doi: 10.1016/j.jacr.2017.02.002.
3. Kori SH, Foley KM, Posner JB. Brachial plexus lesions in patients with cancer=100 cases. *Neurology*. 1981;31(1):45-50. doi: 10.1212/WNL.31.1.45.
4. Qayyum A, MacVicar AD, Padhani AR, Revell P, Husband JES. Symptomatic Brachial Plexopathy following Treatment for Breast Cancer: Utility of MR Imaging with Surface-Coil Techniques. *Radiology*. 2000;214(3):837-842. doi: 10.1148/radiology.214.3.r00mr11837.
5. Miller JD, Pruitt S, McDonald TJ. Acute brachial plexus neuritis: an uncommon cause of shoulder pain. *Am Fam Physician*. 2000;62(9):2067-2072.
6. Ohana M, Moser T, Moussaouï A, et al. Current and future imaging of the peripheral nervous system. *Diagn Interve Imaging*. 2014;95(1):17-26. doi: 10.1016/j.diii.2013.05.008.
7. Szaro P, McGrath A, Ciszek B, Geijer M. Magnetic resonance imaging of the brachial plexus. Part 1: Anatomical considerations, magnetic resonance techniques, and non-traumatic lesions. *Eur J Radiol Open*. 2022;9:100392. doi:1 0.1016/j.ejro.2021.100392.
8. Szaro P, Geijer M, Ciszek B, McGrath A. Magnetic resonance imaging of the brachial plexus. Part 2: Traumatic injuries. *Eur J Radiol Open*. 2022;9:100397. doi: 10.1016/j.ejro.2022.100397.
9. Magill ST, Brus-Ramer M, Weinstein PR, Chin CT, Jacques L. Neurogenic thoracic outlet syndrome: current diagnostic criteria and advances in MRI diagnostics. *Neurosurg*. 2015;39(3):E7. doi: 10.3171/2015.6.focus15219.
10. Mallouhi A, Marik W, Prayer D, Kainberger F, Bodner G, Kasprian G. 3T MR tomography of the brachial plexus: Structural and microstructural evaluation. *Eur JRadiol*. 2012;81(9):2231-2245. doi: 10.1016/j.ejrad.2011.05.021.
11. Gilcrease-Garcia BM, Deshmukh SD, Parsons MS. Anatomy, Imaging, and Pathologic Conditions of the Brachial Plexus. *RadioGraphics*. 2020;40(6):1686-1714. doi: 10.1148/rg.2020200012.
12. Wade RG, Takwoingi Y, Wormald JCR, et al. MRI for Detecting Root Avulsions in Traumatic Adult Brachial Plexus Injuries: A Systematic Review and Meta-Analysis of Diagnostic Accuracy. *Radiology*. 2019;293(1):125-133. doi: 10.1148/radiol.2019190218.

©2023 eviCore healthcare. All Rights Reserved.

Lumbar and Lumbosacral Plexus (PN-5)

Lumbar and Lumbosacral Plexus (PN-5.1)

PN.LP.0005.1.A
v1.0.2024

- 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 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)
- 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)**
- For PET imaging, see **PET Imaging in Oncology (ONC-1.4)** in the Oncology Imaging Guidelines.
  - However, EMG is not needed if concern for malignant infiltration
  - If MRI is contraindicated, CT offers the next highest level of anatomic visualization and can characterize local osseous or vascular anatomy and injury.

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

©2023 eviCore healthcare. All Rights Reserved.

References (PN-5)

v1.0.2024

1. Bykowski J, Aulino JM, Berger KL, et al. ACR Appropriateness Criteria® Plexopathy. *J Am Coll Radiol*. 2017;14(5S):S225-S233. doi: 10.1016/j.jacr.2017.02.002.
2. Pinto MV, Ng PS, Howe BM, et al. Lumbosacral Radiculoplexus Neuropathy. *Neurology*. 2021;96(16):e2098-e2108. doi: 10.1212/wnl.0000000000011799.
3. Dyck PJB, Thaisetthawatkul P. Lumbosacral Plexopathy. *Continuum (Minneap Minn)*. 2014;20:1343-1358. doi: 10.1212/01.con.0000455877.60932.d3.

©2023 eviCore healthcare. All Rights Reserved.

Muscle Disorders (PN-6)

Neuromuscular Junction Disorders (PN-6.1)

PN.MD.0006.1.A
v1.0.2024

Myasthenia Gravis (MG)

Myasthenia Gravis (MG) is associated with thymic disease.

- After an established diagnosis of MG or when MG is suspected by a neurologist, rheumatologist, or ophthalmologist, ONE of the following is indicated[1,4]:
  - 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)[5,6]
  - 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.[27]

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.[7,8]

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- 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

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.[5]
- 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).

©2023 eviCore healthcare. All Rights Reserved.

Muscle Diseases (PN-6.2)

PN.MD.0006.2.C
v1.0.2024

Imaging for Muscle Disease		
Disease	**Indication**	
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

Background and Supporting Information

- 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.

©2023 eviCore healthcare. All Rights Reserved.

Gaucher Disease (Storage Disorders) (PN-6.3)

PN.MD.0006.3.A
v1.0.2024

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) • 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)
  • 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)
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 x-ray is non-diagnostic or indicates the need for further imaging, such as equivocal for osteonecrosis, infection, or malignancy

©2023 eviCore healthcare. All Rights Reserved.

- PET/CT imaging is considered **investigational** 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

©2023 eviCore healthcare. All Rights Reserved.

References (PN-6)

v1.0.2024

1. National Comprehensive Cancer Network® (NCCN®). NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®): Thymoma and Thymic Carcinomas Version 1.2023. December 22, 2022. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Thymoma and Thymic Carcinomas V.1.2023. ©National Comprehensive Cancer Network, Inc. 2022. All rights reserved. Accessed July 28, 2023. The NCCN Guidelines® and illustrations herein may not be reproduced in any form for any purpose without the express written permission of the NCCN. To view the most recent and complete version of the NCCN Guidelines®, go online to NCCN.org.
2. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthenia Gravis: 2020 Update. *Neurology*. 2021;96(3):114-122. doi: 10.1212/WNL.0000000000011124.
3. Hehir MK 2nd, Li Y. Diagnosis and Management of Myasthenia Gravis. *Continuum (Minneap Minn)*. 2022;28(6):1615-1642. doi: 10.1212/CON.0000000000001161.
4. Tuan PA, Vien MV, Dong HV, Sibell D, Giang BV. The Value of CT and MRI for Determining Thymoma in Patients with Myasthenia Gravis. *Cancer Control*. 2019;26(1):1073274819865281. doi:1 0.1177/1073274819865281.
5. Raja SM. Lambert-Eaton Myasthenic Syndrome and Botulism. *Continuum (Minneap Minn)*. 2022;28(6):1596-1614. doi: 10.1212/CON.0000000000001205.
6. Kesner VG, Oh SJ, Dimachkie MM, Barohn RJ. Lambert-Eaton Myasthenic Syndrome. *Neurol Clin*. 2018;36(2):379-394. doi: 10.1016/j.ncl.2018.01.008.
7. Newsome SD, Johnson T. Stiff person syndrome spectrum disorders; more than meets the eye. *J Neuroimmunol*. 2022;369:577915. doi: 10.1016/j.jneuroim.2022.577915.
8. Dade M, Berzero G, Izquierdo C, et al. Neurological Syndromes Associated with Anti-GAD Antibodies. *Int J Mol Sci*. 2020;21(10):3701. Published 2020 May 24. doi: 10.3390/ijms21103701.
9. Manousakis G. Inflammatory Myopathies. *Continuum (Minneap Minn)*. 2022;28(6):1643-1662. doi: 10.1212/CON.0000000000001179.
10. Malartre S, Bachasson D, Mercy G, et al. MRI and muscle imaging for idiopathic inflammatory myopathies. *Brain Pathol*. 2021;31(3):e12954. doi: 10.1111/bpa.12954.
11. Goyal NA. Inclusion Body Myositis. *Continuum (Minneap Minn)*. 2022;28(6):1663-1677. doi: 10.1212/CON.0000000000001204.
12. Maas M, Poll LW, Terk MR. Imaging and quantifying skeletal involvement in Gaucher disease. *Br J Radiol*. 2002;75(suppl_1):A13-A24. doi: 10.1259/bjr.75.suppl_1.750013.
13. Giraldo P, Pocoví M, Pérez-Calvo J, Rubio-Félix D, Giralt M. Report of the Spanish Gaucher's disease registry: clinical and genetic characteristics. *Haematologica*. 2000;85(8):792-799.
14. London ZN. A Structured Approach to the Diagnosis of Peripheral Nervous System Disorders. *Continuum (Minneap Minn)*. 2020;26(5):1130-1160. doi: 10.1212/CON.0000000000000922.
15. Silvestri NJ. A Symptoms and Signs Approach to the Patient with Neuromuscular Weakness. *Continuum (Minneap Minn)*. 2022;28(6):1580-1595. doi: 10.1212/CON.0000000000001150.
16. Pastores GM, Hughes DA. Gaucher Disease. In: Adam MP, Ardinger HH, Pagon RA, et al., eds. *GeneReviews® [Internet]*. 2000 Jul 27 [Updated 2018 Jun 21]. Seattle (WA): University of Washington, Seattle; 1993-2022. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1269/.
17. 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 Imaging*. 2019;10(1). doi: 10.1186/s13244-019-0743-5.
18. Kaplan P, Baris H, De Meirleir L, et al. Revised recommendations for the management of Gaucher disease in children. *Eur JPediatr*. 2012;172(4):447-458. doi: 10.1007/s00431-012-1771-z.
19. Simpson WL, Hermann G, Balwani M. Imaging of Gaucher Disease. *World J Radiol*. 2014;6(9):657-668. doi: 10.4329/wjr.v6.i9.657.
20. Leeuwenberg KE, van Alfen N, Christopher-Stine L, et al. Ultrasound can differentiate inclusion body myositis from disease mimics. *Muscle Nerve*. 2020;61(6):783-788. doi: 10.1002/mus.26875.
21. Tudorancea AD, Ciurea PL, Vreju AF, et al. A Study on Dermatomyositis and the Relation to Malignancy. *Curr Health Sci J*. 2021;47(3):377-382. doi: 10.12865/CHSJ.47.03.07.
22. Lancaster E. Paraneoplastic Disorders. *Continuum (Minneap Minn)*. 2017;23(6, Neuro-oncology):1653-1679. doi: 10.1212/CON.0000000000000542.
23. Guimaraes JB, Cavalcante WCP, Cruz IAN, et al. Musculoskeletal Ultrasound in Inclusion Body Myositis: A Comparative Study with Magnetic Resonance Imaging. *Ultrasound Med Biol*. 2021;47(8):2186-2192. doi: 10.1016/j.ultrasmedbio.2021.04.019.
24. Day JA, Bajic N, Gentili S, Patel S, Limaye V. Radiographic patterns of muscle involvement in the idiopathic inflammatory myopathies. *Muscle Nerve*. 2019;60(5):549-557. doi: 10.1002/mus.26660.

©2023 eviCore healthcare. All Rights Reserved.

25. Pilania K, Jankharia B. Role of MRI in idiopathic inflammatory myopathies: a review article. *Acta Radiol*. 2022;63(2):200-213. doi: 10.1177/0284185121990305.
26. Guimarães JB, Nico MA, Omond AG, et al. Diagnostic Imaging of Inflammatory Myopathies: New Concepts and a Radiological Approach. *Curr Rheumatol Rep*. 2019;21(3):8. Published 2019 Feb 14. doi: 10.1007/s11926-019-0807.
27. Flanagan EP. Paraneoplastic Disorders of the Nervous System. *Continuum (Minneap Minn)*. 2020; 26(6): 1602-1628. doi: 10.1212/CON.0000000000000941.

©2023 eviCore healthcare. All Rights Reserved.

Magnetic Resonance Neurography (MRN) (PN-7)

Magnetic Resonance Neurography (MRN) (PN-7.1)

PN.MR.0007.1.A
v1.0.2024

- 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.

©2023 eviCore healthcare. All Rights Reserved.

References (PN-7)

v1.0.2024

1. Chhabra A, Belzberg AJ, Rosson GD, et al. Impact of high resolution 3 tesla MR neurography (MRN) on diagnostic thinking and therapeutic patient management. *Eur Radiol*. 2016;26(5):1235-1244. doi: 10.1007/s00330-015-3958-y.
2. Chhabra A, Madhuranthakam AJ, Andreisek G. Magnetic resonance neurography: current perspectives and literature review. *Eur Radiol*. 2018;28(2):698-707. doi: 10.1007/s0030-017-4976-8.
3. Holzgrefe RE, Wagner ER, Singer AD, Daly CA. Imaging of the peripheral nerve: concepts and future direction of magnetic resonance neurography and ultrasound. *J Hand Surg Am*. 2019;44(12):1066-1079. doi: 10.1016/i.ihsa.2019.06.021.
4. Sneag DB, Queler S. Technological advancements in magnetic resonance neurography. *Curr Neurol Neurosci Rep*. 2019;19(10):75. Published 2019 Aug 24. doi: 10.1007/s11910-019-0996-x.

©2023 eviCore healthcare. All Rights Reserved.

Neuromuscular Disorders (PN-8)

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

PN.ND.0008.1.A
v1.0.2024

- 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:
    - Monomelic Amyotrophy (Hirayama disease)
    - Spinal Bulbar Muscular Atrophy (Kennedy Disease)

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- 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]

©2023 eviCore healthcare. All Rights Reserved.

Spinal Muscular Atrophy (PN-8.2)

PN.ND.0008.2.A
v1.0.2024

- 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)**.

©2023 eviCore healthcare. All Rights Reserved.

Fasciculations (PN-8.3)

PN.ND.0008.3.A
v1.0.2024

Fasciculations are spontaneous, erratic movements of muscle that may be secondary to benign and non-benign etiologies.

- **ALL** of the following evaluations are required prior to advanced imaging:
  - **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.
  - **EMG/NCV evaluation**
    - In the setting of clinical concern for radiculopathy, neuromuscular disorders, or muscle disorders, see the following imaging guidelines:
      - **Neuromuscular Junction Disorders (PN-6.1)**
      - Muscle Diseases (PN-6.2)
      - **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)**
  - **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.)
- For the presence of upper motor neuron signs (e.g. increased tone; hyperreflexia; presence of Babinski or Hoffman signs) to exclude mimics of non-benign etiologies of muscle fasciculations (i.e. motor neuron disease), **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)
- **Lumbar Spine**: 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 the following Spine Imaging Guidelines:
  - **Red Flag Indications (SP-1.2)**
  - **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)**

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

References (PN-8)

v1.0.2024

1. Kollewe K, Körner S, Dengler R, Petri S, Mohammadi B. Magnetic resonance imaging in amyotrophic lateral sclerosis. *Neurol Res Int*. 2012;2012:608501. doi: 10.1155/2012/608501.
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;17(4):526-e20. doi: 10.1111/j.1468-1331.2010.02951.x.
3. Agosta F, Spinelli EG, Filippi M. Neuroimaging in amyotrophic lateral sclerosis: current and emerging uses. *Expert Rev Neurother*. 2018;18(5):395-406. doi: 10.1080/14737175.2018.1463160.
4. Garg N, Park SB, Vucic S, et al. Differentiating lower motor neuron syndromes. *J Neurol Neurosurg Psychiatry*. 2017;88(6):474-483. doi: 10.1136/jnnp-2016-313526.
5. Shefner JM, Al-Chalabi A, Baker MR, et al. A proposal for new diagnostic criteria for ALS. *Clin Neurophysiol*. 2020;131(8):1975-1978. doi: 10.1016/j.clinph.2020.04.005.
6. Glascock J, Sampson J, Haidet-Phillips A, et al. Treatment Algorithm for Infants Diagnosed with Spinal Muscular Atrophy through Newborn Screening. *J Neuromuscul Dis*. 2018;5(2):145-158. doi: 10.3233/JND-180304.
7. Silveira-Moriyama L, Paciorkowski AR. Genetic Diagnostics for Neurologists.*Continuum (Minneap Minn)*. 2018;24(1, Child Neurology):18-36. doi: 10.1212/CON.0000000000000556.
8. Hatcher-Martin JM, Adams JL, Anderson ER, et al. Telemedicine in neurology: Telemedicine Work Group of the American Academy of Neurology update. *Neurology®* . 2020;94(1):30-38. doi:10.1212/WNL.0000000000008708
9. Prior TW, Leach ME, Finanger E. Spinal Muscular Atrophy. In: Adam MP, Everman DB, Mirzaa GM, et al., eds. *GeneReviews® [Internet]*. 2000 Feb 24; [Updated 2020 Dec 3]. Seattle (WA). University of Washington, Seattle. 1993-2022.
10. Filippakis A, Jara J, Ventura N, et al. A prospective study of benign fasciculation syndrome and anxiety. *Muscle Nerve*. 2018;58(6):852-854. doi: 10.1002/mus.26193.
11. Quinn C, Elman L. Amyotrophic Lateral Sclerosis and Other Motor Neuron Diseases. *Continuum (Minneap Minn)*. 2020;26(5):1323-1347. doi: 10.1212/CON.0000000000000911.

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

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

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

PN.NS.0009.1.A
v1.0.2024

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 PNST is suspected, the following advanced imaging is indicated:
  - 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.
  - Suspected 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)
- Routine follow-up imaging is **NOT** indicated except in the following scenarios:
  - New symptoms or neurological findings develop
  - 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
  - Malignant transformation is known **or** suspected. **ANY** of the following imaging is indicated for metastatic work-up:
    - CT Chest with contrast (CPT® 71260), **AND/OR**
    - CT Abdomen with contrast (CPT® 74160)
- For guidelines related to known malignancies in individuals with 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 well established yet.[8]
- Malignant transformation may be present in approximately 5% of Peripheral Nerve Sheath Tumors.

©2023 eviCore healthcare. All Rights Reserved.

References (PN-9)

v1.0.2024

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. *Skelet Radiol*. 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. *Semin Musculoskelet Radiol*. 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. *Quant Imaging Med Surg*. 2018;8(2):232-251. doi: 10.21037/qims.2018.03.03.
4. Rosser T. Neurocutaneous Disorders. *Continuum (Minneap Minn)*. 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-Oncol Adv*. 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. *Childs Nerv Syst*. 2020;36(10):2229-2268. doi: 10.1007/s00381-020-04758-5.
8. Assadi M, Velez E, Najafi MH, Matcuk G, Gholamrezanezhad A. PET Imaging of Peripheral Nerve Tumors. *PET Clinics*. 2019;14(1):81-89. doi: 10.1016/j.cpet.2018.08.013.

©2023 eviCore healthcare. All Rights Reserved.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com