CLINICAL GUIDELINES

Pediatric Peripheral Nerve Disorders (PND) Imaging Guidelines

Version 1.1

Effective October 15, 2020

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

Pediatric Peripheral Nerve Disorders (PND) Imaging Guidelines

Procedure Codes Associated with Peripheral Nerve Disorders (PND) Imaging	3
PEDPN-1: General Guidelines	5
PEDPN-2: Neurofibromatosis	9
PEDPN-3: Brachial Plexus	13
PEDPN-4: Gaucher Disease	14

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Procedure Codes Associated with Peripheral Nerve Disorders (PND) Imaging

MRI	CPT®
MRI Neck without contrast	70540
MRI Neck without and with contrast	70543
MRI Cervical without contrast	72141
MRI Cervical without and with contrast	72156
MRI Brachial Plexus without contrast (unilateral)	73218
MRI Brachial Plexus without and with contrast (unilateral)	73220
MRI Brachial Plexus without contrast (bilateral)	71550
MRI Brachial Plexus without and with contrast (bilateral)	71552
MRI Chest without contrast	71550
MRI Chest without and with contrast	71552
MRI Thoracic without contrast	72146
MRI Thoracic without and with contrast	72157
MRI Lumbar without contrast	72148
MRI Lumbar without and with contrast	72158
MRI Abdomen without contrast	74181
MRI Abdomen without and with contrast	74183
MRI Pelvis without contrast	72195
MRI Pelvis without and with contrast	72197
MRI Upper Extremity Other Than Joint without contrast	73218
MRI Upper Extremity Other Than Joint with contrast (rarely used)	73219
MRI Upper Extremity Other Than Joint without and with contrast	73220
MRI Upper Extremity Joint without contrast	73221
MRI Upper Extremity Joint with contrast (rarely used)	73222
MRI Upper Extremity Joint without and with contrast	73223
MRI Lower Extremity Other Than Joint without contrast	73718
MRI Lower Extremity Other Than Joint with contrast (rarely used)	73719
MRI Lower Extremity Other Than Joint without and with contrast	73720
MRI Lower Extremity Joint without contrast	73721
MRI Lower Extremity Joint with contrast (rarely used)	73722
MRI Lower Extremity Joint without and with contrast	73723
Unlisted MRI procedure (for radiation planning or surgical software)	76498
MRA	**CPT®**
MRA Upper Extremity	73225
MRA Lower Extremity	73725
Nuclear Medicine	**CPT®**
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging: skull base to mid-thigh (this code not used in pediatrics)	78812
Nuclear Medicine	**CPT®**
PET Imaging: whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815

PET with concurrently acquired CT; whole body	78816
Bone Marrow Imaging Limited Areas	78102
Bone Marrow Imaging Multiple Areas	78103
Bone Marrow Imaging Whole Body	78104
Nuclear Bone Scan Limited	78300
Nuclear Bone Scan Multiple Areas	78305
Nuclear Bone Scan Whole Body	78306
Bone Scan Three Phase	78315
Radiopharmaceutical Localization Imaging Limited area	78800
Radiopharmaceutical Localization Imaging Whole Body	78802
Radiopharmaceutical Localization Imaging SPECT	78803

PEDPN-1: General Guidelines

PEDPN-1.0: General Guidelines	**6**
PEDPN-1.1: Age Considerations	**6**
PEDPN-1.2: Appropriate Clinical Evaluation	**6**
PEDPN-1.3: Modality General Considerations	**6**

PEDPN-1.0: General Guidelines

- A recent (within 60 days) evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the peripheral nervous system is not supported. Advanced imaging of the peripheral nervous system should only be approved in patients who have documented active clinical signs or symptoms of disease involving the peripheral nervous system.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the peripheral nervous system are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect patient management or treatment decisions.

PEDPN-1.1: Age Considerations

- Many conditions affecting the peripheral nervous system in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to patient age, comorbidities, and differences in disease natural history between children and adults.
- Patients who are <18 years old should be imaged according to the Pediatric Peripheral Nerve Disorders Imaging Guidelines, and patients who are ≥18 years old should be imaged according to the Adult Peripheral Nerve Disorders Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDPN-1.2: Appropriate Clinical Evaluation

- See **PEDPN-1.0: General Guidelines**

PEDPN-1.3: Modality General Considerations

- MRI
  - MRI without and with contrast is the preferred modality for pediatric peripheral nerve imaging unless otherwise stated in a specific guideline section.
  - Due to the length of time required for MRI acquisition and the need to minimize patient movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this patient population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition based guidelines. If intravenous access will

already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
    - Recent evidence based literature demonstrates the potential for gadolinium deposition in various organs including the brain after the use of MRI contrast.
    - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
  - If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.

- CT
  - CT is rarely used in the evaluation of pediatric peripheral nerve disorders. See specific guideline sections for indications.
- Ultrasound
  - Ultrasound is rarely used in the evaluation of pediatric peripheral nerve disorders. See specific guideline sections for indications.
- Nuclear Medicine
  - Nuclear medicine studies are not generally indicated in the evaluation of peripheral nerve disorders. See **PEDPN-2: Neurofibromatosis** for specific imaging guidelines regarding PET/CT in evaluation of peripheral nerve tumors.
- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

References

1. Bowen BC. Magnetic resonance imaging of the peripheral nervous system. *Imaging of the Nervous System. eds.* Latchaw RE, Kucharczyk J, Moseley ME, et al. Philadelphia, Elsevier. 2005;1479-1497.
2. Ing C, DiMaggio C, Whitehouse A, et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. *Pediatrics.* 2012 Sep;130(3):e476-e485.
3. Monteleone M, Khandji A, Cappell J, et al. Anesthesia in children: perspectives from nonsurgical pediatric specialists. *J Neurosurg Anesthesiol.* 2014 Oct;26(4):396-398.
4. DiMaggio C, Sun LS, and Li G. Early childhood exposure to anesthesia and risk of developmental and behavioral disorders in a sibling birth cohort. *Anesth Analg.* 2011 Nov;113(5):1143-1151.
5. Fraum TJ, Ludwig DR, Bashir MR, et al. Gadolinium-based contrast agents: a comprehensive risk assessment. *J Magn Reson Imaging.* 2017;46(2):338–353.
6. FDA Medical Imaging Drug Advisory Committee meeting 9/8/17. https://www.fda.gov/downloads/AdvisoryCommittees/CommitteesMeetingMaterials/Drugs/MedicalImagingDrugsAdvisoryCommittee/UCM574746.pdf.
7. Raybaud C and Barkovich AJ. The Phakomatoses. Pediatric neuroimaging. Chapter 6. Philadelphia, Wolters Kluwer, 5th edition. 2012;569-636.
8. Soderlund KA, Smith AB, Rushing EJ, et al. Radiologic-pathologic correlation of pediatric and adolescent spinal neoplasms: Part 2. Intradural extramedullary spinal neoplasms*. AJR Am J Roentgenol*. 2012;198 (1):44-51.
9. Blumfield E, Swenson DW, Iyer RS, Stanescu AL. Gadolinium-based contrast agents — review of recent literature on magnetic resonance imaging signal intensity changes and tissue deposits, with emphasis on pediatric patients. Pediatric Radiology. 2019;49(4):448-457. doi:10.1007/s00247-018-4304-8.

PEDPN-2: Neurofibromatosis

PEDPN-2: Neurofibromatosis – General Information	**10**
PEDPN-2.1: Neurofibromatosis 1	**10**
PEDPN-2.2: Neurofibromatosis 2	**11**

PEDPN-2: Neurofibromatosis – General Information

This guideline section includes imaging indications for patients with neurofibromatosis and known benign lesions. For cancer screening guidelines, See **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines. For guidelines related to known malignancies in patients with NF1, see the appropriate imaging guideline for the specific cancer type.

PEDPN-2.1: Neurofibromatosis 1

- Most cutaneous neurofibromas and deep plexiform neurofibromas do not cause symptoms, and routine surveillance imaging of these lesions has not been shown to improve outcomes.
  - The decision to obtain testing such as imaging studies depends upon the history and physical findings. Clinical evaluation appears to be more useful to detect complications than are screening investigations in asymptomatic patients.
  - The Genetics Committee of the American Academy of Pediatrics have published diagnostic and health supervision guidelines for children with NF1. Surveillance includes:
    - Annual physical examination
    - Annual ophthalmologic examination in children
    - Regular developmental assessment of children
    - MRI for follow-up of clinically suspected tumors
- MRI without and with contrast of a known body area containing a neurofibroma is indicated for any of the following:
  - Every 3 months for treatment response in patients receiving active treatment
  - New or worsening clinical symptoms suggesting progression
  - Preoperative planning
- NF1 patients are more susceptible to damaging effects of ionizing radiation. Studies of NF1 patients irradiated for optic pathway gliomas have reported increased risks for developing another cancer associated with radiotherapy. This risk is associated with radiotherapy, not diagnostic imaging.
- PET imaging is not supported for plexiform neurofibroma surveillance in asymptomatic patients at this time as the positive predictive value is only 60 to 65% even in symptomatic patients.
- MRI without and with contrast is appropriate for any clinical symptoms suggestive of change in a known plexiform neurofibroma in a patient with NF1.
- Although PET imaging has a positive predictive value of only 61 to 63% in NF1 patients with suspected transformation to MPNST, the negative predictive value is high (96 to 99%).
  - PET imaging is indicated for evaluating NF1 patients with clinical symptoms concerning for malignant transformation of a known plexiform neurofibromas when all of the following conditions exist:
    - Recent MRI is inconclusive regarding transformation or progression.

    - Negative PET will result in a decision to avoid biopsy in a difficult or morbid location.
  - Inconclusive PET findings should lead to biopsy of the concerning lesion.
    - Repeat PET studies are not indicated due to the poor positive predictive value in this setting.
  - CT or three-dimensional CT reconstructions may be necessary when surgical treatment of bony lesions is being planned

PEDPN-2.2: Neurofibromatosis 2

- MRI Brain without and with contrast (CPT® 70553) is indicated for patients with known vestibular schwannomas in the following circumstances:
  - Annual imaging for progression in unresected tumors
  - New or worsening clinical symptoms, including hearing loss
  - Preoperative planning
- Patients with NF2 and known meningioma should be imaged according to guidelines in **ONC-2.8: Meningiomas (Intracranial and Intraspinal)** in the Oncology Imaging Guidelines.
- Patients with NF2 and known ependymoma should be imaged according to guidelines in **PEDONC-4.8: Ependymoma** in the Pediatric Oncology Imaging Guidelines.

References

1. Friedman JM. Neurofibromatosis 1. *GeneReviews*™ [Internet] Adam MP, Ardinger HH, Pagon RA, et al. eds. Last Revision: May 17, 2018.
2. Williams VC, Lucas J, Babcock MA, et al. Neurofibromatosis Type I revisited. *Pediatrics.* 2009 Jan;123(1):124-133.
3. Karajannis MA and Ferner RE. Neurofibromatosis-related tumors: emerging biology and therapies. *Curr Opin Pediatr.* 2015 Feb;27(1):26-33.
4. Meany H, Dombi E, Reynolds J, et al. 18-Fluorodeoxyglocose-Positron Emission Tomography (FDG-PET) evaluation of nodular lesions in patients with Neurofibromatosis Type I and Plexiform Neurofibromas (PN) or Malignant Peripheral Nerve Sheath Tumors (MPNST). *Pediatr Blood Cancer.* 2013 Jan;60(1): 59-64.
5. Corbemale P, Valeyrie-Allanore L, Giammarile F, et al. Utility of 18F-FDG PET with a semi-quantitative index in the detection of sarcomatous transformation in patients with Neurofibromatosis Type 1, *PLOS One*. 2014 Feb;e85954.
6. Chirindel A, Chaudhry M, Blakeley JO, et al. 18F-FDG PET/CT qualitative and quantitative evaluation in neurofibromatosis Type 1 patients for detection of malignant transformation: comparison of early to delayed imaging with and without liver activity normalization. *J Nucl Med.* 2015 Mar;56(3):379-385.
7. Evans DG. Neurofibromatosis 2. *GeneReviews*™[Internet] Adam MP, Ardinger HH, Pagon RA, et al. eds. Last Update: March 15, 2018.
8. Blakeley JO, Evans DG, Adler J, et al. Consensus recommendations for current treatments and accelerating clinical trials for patients with neurofibromatosis Type 2. *Am J Med Genetic A.* 2012 Dec;158(1):24-41.
9. Hersh JH. American Academy of Pediatrics Committee on Genetics. Health supervision for children with neurofibromatosis. *Pediatrics*. 2008;121:633.
10. Hirbe AC, and Gutmann DH. Neurofibromatosis Type 1: a multidisciplinary approach to care. *Lancet Neurol.* 2014;13:834–843..

11. Evans DGR, Salvador H, Chang VY, et al. Cancer and Central Nervous System Tumor Surveillance in Pediatric Neurofibromatosis 1. Clinical Cancer Research. 2017;23(12):e46-53.
12. Evans DGR, Salvador H, Chang VY, et al. Cancer and Central Nervous System Tumor Surveillance in Pediatric Neurofibromatosis 2 and Related Disorders. Clinical Cancer Research. 2017;23(12):e54-61.
13. Kleinerman RA. Radiation-sensitive genetically susceptible pediatric sub-populations. Pediatric Radiology. 2008;39(S1):27-31. doi:10.1007/s00247-008-1015-6.
14. Vezina G, Barkovich AJ. Neurocutaneous disorders. In: Barkovich AJ, Raybaud C, eds. Pediatric Neuroimaging. 6th ed. Wolters Kluwer Philadelphia. 2019; 633-702.

PEDPN-3: Brachial Plexus

Disorders of the brachial plexus can generally be identified and distinguished from lesions in other locations by clinical, electromyography and nerve conduction (EMG/NCV) examination. If the diagnosis remains unclear, advanced imaging can be helpful as a preoperative study to evaluate the anatomy of brachial plexus lesions which should have already been defined by clinical examination.

- MRI is the preferred modality for imaging the brachial plexus. The goal of imaging is to visualize the entire course of the neural network from the preganglionic to the postganglionic segments.
  - CT is not often useful and should not be used as a substitute for MRI.
  - Unilateral brachial plexus studies should be ordered as MRI Upper Extremity Other Than Joint without contrast (CPT® 73218) or without and with contrast (CPT® 73220).
  - Bilateral brachial plexus studies should be ordered as MRI Chest without contrast (CPT® 71550) or without and with contrast (CPT® 71552). For upper trunk lesions, MRI Neck without contrast (CPT® 70540) is indicated.
  - It is rare for more than one CPT® code to be necessary to adequately image the brachial plexus area of interest. These requests should be forwarded for Medical Director Review.
  - MRI Shoulder without contrast (CPT® 73221) or without and with contrast (CPT® 73223) is indicated in infants with brachial plexopathy due to birth trauma if requested for preoperative planning. These patients often have glenohumeral dysplasia and require shoulder surgery.
  - Ultrasound also may be indicated in infants with brachial plexus injury to show the glenoid dysplasia and associated shoulder subluxation
  - If there is clinical suspicion for cervical nerve root avulsion, MRI Cervical Spine without contrast (CPT®72141) is indicated.
  - In patients with a known malignancy or post-treatment syndrome, PET/CT skull base to mid-thigh (CPT® 78815) may be approved if there is a contraindication to MRI.

References

1. Wippold FJ, Cornelius RS, Aiken AH, et al. Plexopathy. ACR Appropriateness Criteria®. *American College of Radiology.* 2016:1-10.
2. Menashe SJ, Tse R, Nixon JN, et al. Brachial plexus birth palsy: multimodality imaging of spine and shoulder abnormalities in children. *AJR Am J Roentgenol*. 2015 Feb;204(2):W199-W206.
3. Somashekar DK, DiPietro MA, Joseph JR, et al. Ultility of ultrasound in noninvasive preoperative workup of neonatal brachial plexus palsy. *Pediatr Radiol*. 2016;46:695-703.
4. Volpe JJ. Injuries of extracranial, cranial, intracranial, spinal cord and peripheral nervous system structures. In: Volpe's Neurology of the Newborn, 6th ed. Elsevier. Philadelphia 2018; 1093-1123.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

PEDPN-4: Gaucher Disease

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.

- Three types of Gaucher disease are recognized:
  - **Type I** (non-neuropathic form or adult form): progressive hepatomegaly, splenomegaly, anemia and thrombocytopenia, and marked skeletal involvement; lungs and kidneys may also be involved, but central nervous system is spared
  - **Type II** (acute neuropathic form or infantile form): severe progressive neurological involvement with death by 1 to 2 years of age; hepatomegaly, splenomegaly, is also present (usually evident by 6 months of age)
  - **Type III**: type I with neurological involvement
- MRI Lumbar Spine without contrast (CPT® 72148) and Bilateral Femurs (CPT® 73718) is indicated to evaluate bone marrow involvement at initial diagnosis.
  - Repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- MRI Abdomen without contrast (CPT® 74181) is indicated to assess liver and spleen involvement at initial diagnosis.
  - Repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- Pulmonary involvement is less common, but CT Chest without contrast (CPT® 71250) is indicated for patients with new or worsening pulmonary symptoms.
  - For patients with documented pulmonary involvement, repeat imaging is indicated every 12 months, to assess treatment response for patients on enzyme replacement therapy or to assess disease progression for patients in surveillance.
- 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.

References

1. Simpson WL, Hermann G, and Balwani M. Imaging of Gaucher disease. *World J Radiol.* 2014 Sep;6:657-668.
2. Anderson H, Kaplan P, Kacena K, et al. Eight-year clinical outcomes of long-term enzyme replacement therapy for 884 children with Gaucher disease Type I. *Pediatrics.* 2008;122(6):1182-1190.