



CLINICAL GUIDELINES

Pediatric PND Imaging Policy

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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.

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**PEDIATRIC PERIPHERAL NERVE DISORDER (PND) IMAGING
GUIDELINES**

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PEDIATRIC PND IMAGING GUIDELINES

PEDPN-1~GENERAL GUIDELINES

Procedure Codes Associated with Musculoskeletal Imaging	
MRI	CPT®
Neck MRI without contrast	70540
Neck MRI without and with contrast	70543
Cervical MRI without contrast	72141
Cervical MRI without and with contrast	72156
Brachial plexus MRI without contrast (unilateral)	73218
Brachial plexus MRI without and with contrast (unilateral)	73220
Brachial plexus MRI without contrast (bilateral)	71550
Brachial plexus MRI without and with contrast (bilateral)	71552
Chest MRI without contrast	71550
Chest MRI without and with contrast	71552
Thoracic MRI without contrast	72146
Thoracic MRI without and with contrast	72157
Lumbar MRI without contrast	72148
Lumbar MRI without and with contrast	72158
Abdomen MRI without contrast	74181
Abdomen MRI without and with contrast	74183
Pelvis MRI without contrast	72195
Pelvis MRI without and with contrast	72197
Upper Extremity MRI non-joint without contrast	73218
Upper Extremity MRI non-joint with contrast (rarely used)	73219
Upper Extremity MRI non-joint without and with contrast	73220
Upper Extremity MRI joint without contrast	73221
Upper Extremity MRI joint with contrast (rarely used)	73222
Upper Extremity MRI joint without and with contrast	73223
Lower Extremity MRI non-joint without contrast	73718
Lower Extremity MRI non-joint with contrast (rarely used)	73719
Lower Extremity MRI non-joint without and with contrast	73720

Lower Extremity MRI joint without contrast	73721
Lower Extremity MRI joint with contrast (rarely used)	73722
Lower Extremity MRI joint without and with contrast	73723
Unlisted MRI procedure (for radiation planning or surgical software)	76498
MRA	CPT®
Upper Extremity MRA	73225
Lower Extremity MRA	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
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
Nuclear Bone Scan SPECT	78320
Radiopharmaceutical Imaging of Inflammatory Process Limited Area	78805
Radiopharmaceutical Imaging of Inflammatory Process Whole Body	78806
Radiopharmaceutical Imaging of Inflammatory Process SPECT	78807

PEDIATRIC PND IMAGING GUIDELINES

PEDPN-1~GENERAL GUIDELINES

PEDPN-1.1 Pediatric Peripheral Nerve Disorders Imaging 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 age <18 years old should be imaged according to the Pediatric Peripheral Nerve Disorders Imaging Guidelines, and patients age \geq 18 years should be imaged according to the Peripheral Nerve Disorders Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDPN-1.2 Pediatric Peripheral Nerve Disorders Imaging Appropriate Clinical Evaluation

- ✓ A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, 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-1.3 Pediatric Peripheral Nerve Disorders Imaging 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 for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age <7 years), as

well as older children with delays in development or maturity. In this patient population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:

- MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the patient already has intravenous access for anesthesia.
- 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 et al, *Magnetic Resonance Imaging of the Peripheral Nervous System*. In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005, po.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;130:e476-e485.
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PEDIATRIC PND IMAGING GUIDELINES

PEDPN-2~NEUROFIBROMATOSIS

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)**. 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.
- ✓ 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, and CT imaging should only be used for patients who have an absolute contraindication to MRI.
- ✓ PET imaging is not supported for PN surveillance in asymptomatic patients at this time as the positive predictive value is only 60-65% even in symptomatic patients.
- ✓ MRI imaging without and with contrast is appropriate for any clinical symptoms suggestive of change in a known PN in a patient with NF1.
- ✓ Although PET imaging has a positive predictive value of only 61-63% in NF1 patients with suspected transformation to MPNST, the negative predictive value is high (96-99%).
 - PET imaging is indicated for evaluating NF1 patients with clinical symptoms concerning for malignant transformation of a known PN 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.

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**.
- ✓ Patients with NF2 and known ependymoma should be imaged according to guidelines in **PEDONC-4.8 Ependymoma**.

References

1. Friedman JM, Neurofibromatosis 1, *GeneReviews*[™][Internet] eds. Pagon RA, Adam MP, Ardinger HH et al, version September 4, 2014.
2. Williams VC, Lucas J, Babcock MA et al, Neurofibromatosis Type I Revisited, *Pediatrics* 2009;123:124-133.
3. Karajannis MA and Ferner RE, Neurofibromatosis-related tumors: emerging biology and therapies, *Curr Opin Pediatr* 2015; 27:26-33.
4. Meany H, Dombi E, Reynolds J et al, 18-Fluorodeoxyglucose-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;60:59-64.
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;9:e85954.
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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.
 - 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 of the 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.
 - If there is clinical suspicion for cervical nerve root avulsion, MRI Cervical Spine without contrast (CPT[®] 72141) is indicated.

References

1. Wittenberg KH and Adkins MC, MR Imaging of Nontraumatic Brachial Plexopathies: Frequency and Spectrum of Findings, *Radiographics* 2000; 20:1023-1032.
2. Wippold FJ, Cornelius RS, Aiken AH et al, Plexopathy, *ACR Appropriateness Criteria*[®], 2013:1-16.
3. van Es HW, MRI of the brachial plexus, *Eur Radiol* 2001;11:325-336.
4. 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;204:W199-W206.

PEDPN-4~GAUCHER DISEASE

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.

- ✓ MRI without contrast of the lumbar spine (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 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 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 disease progression for patients in surveillance.
- ✓ 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.

References

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2. Anderson H, Kaplan P, Kacena K, and Yee J, Eight-Year Clinical Outcomes of Long-Term Enzyme Replacement Therapy for 884 Children With Gaucher Disease Type I, *Pediatrics* 2008;122:1182-1190.
3. Sidransky E. *Gaucher disease*. Medscape, updated November 24, 2014, available at: <http://emedicine.medscape.com/article/944157-overview>.