CLINICAL GUIDELINES

Pediatric Peripheral Nerve Disorders Imaging Guidelines

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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 Disorders (PND) Imaging Guidelines

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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

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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

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PEDPN-1: General Guidelines

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PEDPN-1.0: General Guidelines

- A pertinent clinical evaluation including a detailed history, physical examination with a thorough neurologic examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
  - EMG may not be of clinical utility or obtainable in infants or individuals with severe developmental delay
  - EMG/NCS results may not be abnormal until 10 days after injury.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the peripheral nervous system is not supported. Advanced imaging of the peripheral nervous system should only be approved in individuals 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 individual 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 individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are <18 years old should be imaged according to the Pediatric Peripheral Nerve Disorders Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Peripheral Nerve Disorder Imaging Guidelines should be imaged according to the General Peripheral Nerve Disorder Imaging Guidelines. Individuals who are ≥18 years old should be imaged according to the General 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**

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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 individual 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 individual 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.

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- 3D Rendering
  - 3D Rendering indications in pediatric PND imaging are identical to those in the general imaging guidelines. See **Preface-4.1: 3D Rendering** in the Preface Imaging Guidelines.
- 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.

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PEDPN-2: Neurofibromatosis

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PEDPN-2.2: Neurofibromatosis 2	**10**

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PEDPN-2.0: Neurofibromatosis – General Information

- This guideline section includes imaging indications for individuals 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 Peripheral Nerve Sheath Tumors, See **PND-9: Peripheral Nerve Sheath Tumors (PNST)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.
- For guidelines related to known malignancies in individuals with NF1, see the appropriate imaging guideline for the specific cancer type.

PEDPN-2.1: Neurofibromatosis 1

- See **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines.
- For Peripheral Nerve Sheath Tumors, See **PND-9: Peripheral Nerve Sheath Tumors (PNST)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.
- For guidelines related to known malignancies in individuals with NF1, see the appropriate imaging guideline for the specific cancer type

PEDPN-2.2: Neurofibromatosis 2

- See **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines.
- Individuals with NF2 and known meningioma should be imaged according to guidelines in **ONC-2.8: Meningiomas (Intracranial and Intraspinal)** in the Oncology Imaging Guidelines.
- Individuals with NF2 and known ependymoma should be imaged according to guidelines in **PEDONC-4.8: Ependymoma** in the Pediatric Oncology Imaging Guidelines.

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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. 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.
  - MRI Upper Extremity Other Than Joint without contrast (CPT® 73218) or without and with contrast (CPT® 73220) is indicated for unilateral brachial plexus.
  - MRI Chest without contrast (CPT® 71550) or without and with contrast (CPT® 71552) is indicated for bilateral brachial plexus studies. MRI Neck without contrast (CPT® 70540) is indicated for upper trunk lesions.
  - It is rare for more than one CPT® code to be necessary to adequately image the brachial plexus area of interest.
  - 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 individuals 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
  - MRI Cervical Spine without contrast (CPT®72141) is indicated if there is clinical suspicion for cervical nerve root avulsion.
  - PET/CT skull base to mid-thigh (CPT® 78815) may be approved if there is a contraindication to MRI in individuals with a known malignancy or post-treatment syndrome.

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PEDPN-4: Gaucher Disease

PEDPN-4.1: 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. 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 rapidly progressive neurological involvement with onset by 2 years of age and death by 2 to 4 years of age; hepatomegaly, splenomegaly, is also present (usually evident by 6 months of age)
  - **Type III**: type I with neurological involvement and slowly progressive disease. Onset may be present before two years of age with survival to the third or fourth decade of life.
  - Additionally, there is a perinatal-lethal and a cardiovascular form. The cardiovascular form involves the heart, spleen and eyes. Note that cardiopulmonary complications may be present, with varying frequency and severity, in all subtypes.

Imaging for Gaucher Disease
Initial Imaging
➢ MRI Lumbar Spine without contrast (CPT® 72148) ➢ MRI Bilateral Femurs without contrast (CPT® 73718) ➢ MRI Abdomen without contrast (CPT® 74181) ➢ DXA scan ➢ CT Chest without contrast (CPT® 71250) for individuals with new or worsening pulmonary symptoms
Every 12 months
➢ To assess treatment response for individuals on enzyme replacement therapy or assess disease progression for individuals in surveillance ◆ MRI Lumbar Spine without contrast (CPT® 72148) ◆ MRI Bilateral Femurs without contrast (CPT® 73718) ◆ MRI Abdomen without contrast (CPT® 74181) ◆ CT Chest without contrast (CPT® 71250) for individuals with documented pulmonary involvement
New or worsening pulmonary symptoms
➢ CT Chest without contrast (CPT® 71250)
Follow-up DXA scans
➢ Every 12-24 months until it is normal ➢ Enzyme replacement therapy dose change ➢ Every 3 years

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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. ^{18}F-FDG does not reliably detect Gaucher disease in the marrow, and other isotopes are not yet FDA-approved for clinical use.

Practice Notes

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

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PEDPN-5: Spinal Muscular Atrophy

PEDPN-5.1: Spinal Muscular Atrophy

- 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 not indicated
  - See **PEDHD-19.3: Developmental Motor Delay** in the Pediatric Head Imaging Guidelines for presentation of weakness or a loss of skills.

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