Cigna Medical Coverage Policies – Radiology
Pediatric Peripheral Nerve Disorders
Imaging Guidelines

Effective February 1, 2022

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.

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 non-joint without contrast	73218
MRI Upper Extremity non-joint with contrast (rarely used)	73219
MRI Upper Extremity non-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 non-joint without contrast	73718
MRI Lower Extremity non-joint with contrast (rarely used)	73719
MRI Lower Extremity non-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

PEDPN-1: General Guidelines

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 follow-up 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**

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.
- 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.

References

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PEDPN-2: Neurofibromatosis

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)**.
- 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).**
- For guidelines related to known malignancies in patients 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.

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.
  - 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. 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 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

Imaging for Gaucher Disease

Initial Imaging

- MRI Lumbar Spine without contrast (CPT® 72148)
- MRI Bilateral Femurs without contrast (CPT® 73718)
- MRI Abdomen without contrast (CPT® 74181)
- DXA scan
- CT Chest without contrast (CPT® 71250) for patients with new or worsening pulmonary symptoms

Every 12 months

- To assess treatment response for patients on enzyme replacement therapy or assess disease progression for patients in surveillance
  - MRI Lumbar Spine without contrast (CPT® 72148)
  - MRI Bilateral Femurs without contrast (CPT® 73718)
  - MRI Abdomen without contrast (CPT® 74181)
  - CT Chest without contrast (CPT® 71250) for patients with documented pulmonary involvement

New or worsening pulmonary symptoms

- CT Chest without contrast (CPT® 71250)

Follow-up DXA scans

- Every 12-24 months until it is normal
- Enzyme replacement therapy dose change
- Every 3 years

Acute bone pain

- X-ray
  - MRI of affected areas with and without contrast may be approved if xray is non-diagnostic or indicates the need for further imaging, such as equivocal for osteonecrosis, infection or malignancy

- PET/CT imaging is considered investigational in the evaluation of Gaucher disease. ^{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 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
- Patients 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** for presentation of weakness or a loss of skills.

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