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.

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## Pediatric Peripheral Nerve Disorders (PND) Imaging Guidelines

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**Procedure Codes Associated with Musculoskeletal Imaging**

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<tr>
<td>Neck MRI without contrast</td>
<td>70540</td>
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<td>Neck MRI without and with contrast</td>
<td>70543</td>
</tr>
<tr>
<td>Cervical MRI without contrast</td>
<td>72141</td>
</tr>
<tr>
<td>Cervical MRI without and with contrast</td>
<td>72156</td>
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<tr>
<td>Brachial plexus MRI without contrast (unilateral)</td>
<td>73218</td>
</tr>
<tr>
<td>Brachial plexus MRI without and with contrast (unilateral)</td>
<td>73220</td>
</tr>
<tr>
<td>Brachial plexus MRI without contrast (bilateral)</td>
<td>71550</td>
</tr>
<tr>
<td>Brachial plexus MRI without and with contrast (bilateral)</td>
<td>71552</td>
</tr>
<tr>
<td>Chest MRI without contrast</td>
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</tr>
<tr>
<td>Chest MRI without and with contrast</td>
<td>71552</td>
</tr>
<tr>
<td>Thoracic MRI without contrast</td>
<td>72146</td>
</tr>
<tr>
<td>Thoracic MRI without and with contrast</td>
<td>72157</td>
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<tr>
<td>Lumbar MRI without contrast</td>
<td>72148</td>
</tr>
<tr>
<td>Lumbar MRI without and with contrast</td>
<td>72158</td>
</tr>
<tr>
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<td>Pelvis MRI without contrast</td>
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<td>Upper Extremity MRI non-joint without contrast</td>
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<td>Upper Extremity MRI non-joint with contrast (rarely used)</td>
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PEDPN-1: General Guidelines

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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, and individuals 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

- 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 (CT, MR, Nuclear Medicine), unless the individual 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 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-1.3: 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 for image acquisition and the need for the individual to lie still, 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 individual population, MRI imaging sessions should be planned with a goal of minimizing anesthesia exposure adhering to the following considerations:
    - MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the individual already has intravenous access for anesthesia.
    - 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 requesting clinicians indicate that a non-contrast study is being requested with specific concern for gadolinium retention, the exam can be approved.

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.

References
## PEDPN-2: Neurofibromatosis

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**PEDPN-2: 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)**. 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**

- 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 individuals receiving active treatment
  - New or worsening clinical symptoms suggesting progression
  - Preoperative planning
- NF1 individuals are more susceptible to damaging effects of ionizing radiation, and CT imaging should only be used for individuals who have an absolute contraindication to MRI.
- PET imaging is not supported for PN surveillance in asymptomatic individuals at this time as the positive predictive value is only 60 to 65% even in symptomatic individuals.
- MRI imaging without and with contrast is appropriate for any clinical symptoms suggestive of change in a known PN in an individual with NF1.
- Although PET imaging has a positive predictive value of only 61 to 63% in NF1 individuals with suspected transformation to MPNST, the negative predictive value is high (96 to 99%).
  - PET imaging is indicated for evaluating NF1 individuals 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.
    - CT imaging or three-dimensional CT reconstructions may be necessary when surgical treatment of bony lesions is being planned.
**Background and Supporting Information**

- 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 individuals.
- 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 and other.

**PEDPN-2.2: Neurofibromatosis 2**

- MRI Brain without and with contrast (CPT® 70553) is indicated for individuals 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
- Individuals with NF2 and known meningioma should be imaged according to guidelines in **ONC-2.8: Meningiomas (Intracranial and Intraspinal)**
- Individuals with NF2 and known ependymoma should be imaged according to guidelines in **PEDONC-4.8: Ependymoma**
References


Pediatric PND Imaging

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 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 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.
  ➤ Whole body PET/CT (CPT® 78816) may be approved if there is a contraindication to MRI in individuals with a known malignancy or post-treatment syndrome.

References


PEDPN-4: Gaucher Disease

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 individuals on enzyme replacement therapy or disease progression for individuals 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 individuals on enzyme replacement therapy or disease progression for individuals in surveillance.

CT Chest without contrast (CPT® 71250) is indicated for individuals with new or worsening pulmonary symptoms.
- For individuals with documented pulmonary involvement, repeat imaging is indicated every 12 months, to assess treatment response for individuals on enzyme replacement therapy or disease progression for individuals 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.

Background and Supporting Information
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.

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
References

