# Cigna Medical Coverage Policies – Radiology
# Pediatric Peripheral Nerve Disorders (PND) Imaging Guidelines

Effective October 1, 2021


---

**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





400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

# 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*


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. Update on FDA approach to safety issue of gadolinium retention after administration of gadolinium-based contrast agents available at https://www.fda.gov/media/116492/download.
7. 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.
8. Raybaud C and Barkovich AJ. The Phakomatoses. Pediatric neuroimaging. Chapter 6. Philadelphia, Wolters Kluwer, 5th edition. 2012;569-636.
9. 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.
10. 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.
11. Kang PB, McMillan HJ, Kuntz NL, et al. Utility and practice of electrodiagnostic testing in the pediatric population: An AANEM consensus statement. Muscle & Nerve. 2020;61(2):143-155. doi:10.1002/mus.26752.
12. Orozco V, et al. A Systematic Review of the Electrodiagnostic Assessment of Neonatal Brachial Plexus. Neurol Neurobiol (Tallinn). 2020;3(2): doi:10.31487/j.nnb.2020.02.12.

# 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 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. Studies of NF1 individuals 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 individuals at this time as the positive predictive value is only 60 to 65% even in symptomatic individuals.
- MRI without and with contrast is appropriate for any clinical symptoms suggestive of change in a known plexiform neurofibroma 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 (Malignant peripheral nerve sheath tumor), 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 plexiform neurofibroma 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.
  - The Utility of Whole Body MRI remains unclear in NF-1 at this time, and WBMRI is generally not supported for routine surveillance or screening of individuals with NF-1 nor NF-2.

### *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

## 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 in individuals with 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)** 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.

## *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.
15. Ahlawat S, Blakeley JO, Langmead S, Belzberg AJ, Fayad LM. Current status and recommendations for imaging in neurofibromatosis type 1, neurofibromatosis type 2, and schwannomatosis. Skeletal Radiology. 2019;49(2):199-219. doi:10.1007/s00256-019-03290-1.

# 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.

### *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 www.eviCore.com

# PEDPN-4: Gaucher Disease

## PEDPN-4.1: Gaucher Disease

- 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 individuals on enzyme replacement therapy or to assess 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 to assess 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 to assess disease progression for individuals in surveillance.
- DXA may be approved every 2 years beginning at 6 years of age if requested by an institution with technical and interpretive expertise with DXA in pediatrics. These requests should be sent to Medical Directors Review.
- Individuals with Gaucher disease are at risk for osteonecrosis, osteomyelitis, and bony tumors. Acute bone pain should be imaged with plain 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.

- 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*


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.
3. Degnan AJ, Ho-Fung VM, Ahrens-Nicklas RC, et al. Imaging of non-neuronopathic Gaucher disease: recent advances in quantitative imaging and comprehensive assessment of disease involvement. Insights into Imaging. 2019;10(1). doi:10.1186/s13244-019-0743-5.

# 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.

### *References*


1. Arnold WD, Kassar D and Kissel JT. Spinal Muscular Atrophy: Diagnosis and Management in a New Therapeutic Era. Muscle Nerve. 2015 February ; 51(2): 157–167. doi:10.1002/mus.24497.
2. Glascock J, et al. Treatment Algorithm for Infants Diagnosed with Spinal Muscular Atrophy through Newborn Screening.Research Report. 8. Journal of Neuromuscular Diseases 5 (2018) 145–158. DOI 10.3233/JND-180304 9. Silveira-Moriyama Laura and Paciorkowski AR. Genetic Dianostics for Neurologists. CONTINUUM (MINNEAP MINN) 2018;24(1, CHILD NEUROLOGY):18–36 .
3. Prior TW, Leach ME and Finanger E. Spinal Muscular Atrophy. Gene Reviews. Created: February 24, 2000; Updated: November 14, 2019. Copyright © 1993-2020, University of Washington, Seattle. GeneReviews is a registered trademark of the University of Washington, Seattle. All rights reserved.
4. Arnold WD, Kassar D and Kissel JT. Spinal Muscular Atrophy: Diagnosis and Management in a New Therapeutic Era. Muscle Nerve. 2015 February ; 51(2): 157–167. doi:10.1002/mus.24497