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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

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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 Spine Imaging Guidelines

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## Procedure Codes Associated with Spine Imaging

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PEDSP-1: General Guidelines

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PEDSP-1.1: Pediatric Spine Imaging Age Considerations
Many conditions affecting the spine 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 Spine Imaging Guidelines, and individuals who are ≥ 18 years old should be imaged according to the Adult Spine Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDSP-1.2: Pediatric Spine Imaging Appropriate Clinical Evaluation
- A recent (within 60 days) face-to-face 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, 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 spine is not supported. Advanced imaging of the spine should only be approved in individuals who have documented active clinical signs or symptoms of disease involving the spine.

- Unless otherwise stated in a specific guideline section, repeat imaging studies of the spine 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.

PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations
- MRI
  - MRI is the preferred modality for imaging the pediatric spine 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 due to concerns regarding the use of gadolinium, 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 generally inferior to MRI for imaging the pediatric spine, but has specific indications in which it is the preferred modality listed in specific sections of these guidelines.
  - CT should not be used to replace MRI in an attempt to avoid sedation unless it is listed as a recommended study in a specific guideline section.
- Myelogram with post-myelogram CT imaging is rarely indicated in children except in certain limited indications (usually requested after specialist consultation), including:
  - Evaluation of spine in patients with fixation hardware which limits utility of MRI.
  - Severe congenital scoliosis with inconclusive MRI.
  - Evaluation of nerve root avulsion in individuals with a brachial plexus injury and inconclusive MRI.
  - Evaluation of paraspinal cyst to assess continuity with the subarachnoid space.
  - Coding note: CT of appropriate spinal level with or without contrast may be appropriate. If the radiologist performs the myelogram the exam should be coded with contrast. If a clinician performs the myelogram the exam should be coded without contrast.

**Ultrasound**
- Spinal canal ultrasound (CPT® 76800) describes the ultrasonic evaluation of the spinal cord (canal and contents) and should not be reported multiple times for imaging of different areas of the spinal canal.
- Do not use CPT® 76800 for intraoperative spinal canal ultrasound as CPT® 76998 (intraoperative ultrasonic guidance) is the appropriate code in this circumstance.
- Spinal canal ultrasound (CPT® 76800) is generally limited to infants up to 6 months of age because of the bone mass surrounding the spinal cord limits evaluation of the intraspinal contents in older infants.
  - **Exception**: the persisting acoustic window in children with posterior spinal defects of spinal dysraphism enables spinal canal ultrasound to be performed at any age (See **PEDSP-4: Spinal Dysraphism**).
- In general, additional imaging studies of the spine are not indicated in asymptomatic individuals with normal spinal ultrasound findings.
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

PEDSP-2: Pediatric Back and Neck Pain

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**PEDSP-2.1: Introduction**

Currently, only about 20% of back pain in children over age 5 is from a discoverable cause. Scoliosis, spondylitic disorders, Scheuermann disease, tumor, and trauma are the most common causes.

Back pain in children under age 5 is uncommon and often reflects underlying serious disease when present.

Disc herniations are rare in children, but become more frequent as activity increases during adolescence.

**PEDSP-2.2: Back and Neck Pain in Children Age 5 and Under**

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.

- Advanced imaging is appropriate in all individuals in this age group except those with mild and transient back pain.
  - MRI of the symptomatic spinal region
    - Individuals in this age group will require sedation to complete MRI imaging. See [PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations](#) for contrast and body area considerations.
  - CT without contrast of the symptomatic spinal region when:
    - Plain x-rays suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group).
    - A recent MRI does not provide sufficient detail of the bony anatomy to allow for acute individual care decision making.

**Background and Supporting Information**

SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period.
PEDSP-2.3: Back and Neck Pain in Children Age 6 and Over

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.

- Advanced imaging should be approved following a recent x-ray when one or more of the following pediatric “red flags” are present:
  - Accompanying systemic symptoms (fever, weight loss, etc.)
  - Functional disability (daily limitation in normal activities because of pain)
  - Pain which is extremely severe or worse at night
  - Early morning stiffness
  - Pain which worsens despite an attempt at symptomatic treatment
  - Neurological symptoms or abnormal neurological examination findings
  - An established diagnosis of cancer other than leukemia
  - Abnormal x-rays
  - Spinal imaging for individuals having undergone spinal surgery
  - Associated bowel or bladder dysfunction

- In the absence of any “red flags”, a 4 week trial of provider-supervised conservative treatment should be attempted before advanced imaging can be approved.
  - It can be assumed that children who are being evaluated by a pediatric spine surgeon have failed a reasonable trial of conservative treatment under the care of the primary care provider as this is by far the most common reason for such referrals.

- X-rays of the involved regions should be obtained prior to advanced imaging in individuals with “red flag” findings, or who remain symptomatic after a 4 week trial of provider-supervised conservative treatment.

- MRI without contrast of the symptomatic spinal region is the preferred study for the evaluation of pediatric spine pain, and should be approved unless one of the following conditions applies, in which case MRI without and with contrast should be approved:
  - Fever (100° F or higher)
  - Clinical suspicion of infection (discitis, osteomyelitis, paraspinous or epidural abscess)
  - Physical examination or plain x-ray suggests a mass lesion
  - New or worsening pain in an individual with an established diagnosis of cancer

- CT without contrast of the symptomatic spinal region when:
  - The request is for re-evaluation of a known vertebral bony disorder.
  - Plain x-rays show spondylotic changes or suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group).
  - A recent MRI does not provide sufficient detail of the bony anatomy to allow for acute individual care decision making.
Background and Supporting Information
Radicular back and neck pain is common in adult individuals but is uncommon in adolescents and very rare in children.

PEDSP-2.4: Spondylolysis
- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- Spondylolysis is best recognized on plain x-rays, and advanced imaging is generally not indicated.
  - If additional imaging is needed because of radiological uncertainty or associated spondylolisthesis, 99mTc-MDP SPECT bone scan (CPT® 78320) is indicated to identify stress reaction in early spondylolysis cases which are radiographically occult. Bone scan has been demonstrated to be superior to MRI in detecting active spondylolysis.
  - MRI without contrast of the symptomatic spinal level is indicated to evaluate for stress reaction in bone and visualizing nerve roots, if bone scan is negative, symptoms have continued despite a recent 4 week course of conservative care, or there is a documented need for preoperative planning.
  - CT without contrast of the symptomatic spinal level is indicated to provide detailed evaluation of bony anatomy, if bone scan is negative or there is a documented need for preoperative planning.

Background and Supporting Information
- Most cases of childhood spondylolysis are believed to be caused by repeated microtrauma, resulting in stress fracture of the pars interarticularis. Heredity is also believed to be a factor in some cases. It is the most common cause of low back pain in children older than age 10.
- Activity modification, NSAID treatment, physical therapy, and/or immobilization with various braces are the initial treatments for symptomatic individuals.
- Surgical treatment is only recommended for individuals with disabling symptoms that have not responded to non-surgical care.
- SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period.
- CT scans have been considered the criterion standard for characterizing fractures and for detailing bone morphology and anatomy.
PEDSP-2.5: Spine Pain Due to Infectious Causes

- A detailed history and physical examination with thorough neurologic examination and plain x-rays should be performed initially.

**Initial Imaging Studies**
- MRI without and with contrast of the symptomatic spinal level is very sensitive at detecting early changes and can be approved when discitis or osteomyelitis is suspected.

**Follow-Up Imaging Studies**
- Follow-up plain x-rays may show disc space narrowing and bony changes of osteomyelitis.
- MRI without and with contrast of the symptomatic spinal level or CT with contrast (including myelography) may be useful in follow-up for evaluating bony changes of osteomyelitis or concern for epidural abscess.

**Background and Supporting Information**
- Entities include discitis and vertebral osteomyelitis, and typically present with sudden onset of back pain, fever, and elevated white blood cell count, occurring most commonly in prepubescent children.

**References**

### PEDSP-3: Kyphosis and Scoliosis

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The term “kyphosis” refers to a curve convex posteriorly. Kyphosis generally affects the thoracic spine.

The term “lordosis” refers to a curve convex anteriorly.

The term “scoliosis” refers to a lateral curvature.

### PEDSP-3.1: Juvenile Thoracic Kyphosis (Scheuermann Disease)

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.

- X-rays will typically show anterior wedging in three or more adjacent vertebral bodies.
  - Lower thoracic kyphosis from developmental vertebral wedging with thoracic kyphosis totaling over 15° to 20° should be identified by plain x-rays before considering advanced imaging.
  - MRI is not an effective diagnostic modality for this condition since the incidence of false positive vertebral changes in normal individuals is high.

- MRI thoracic spine without contrast (CPT® 72146) preoperatively to rule out any associated spinal cord problems.

- MRI lumbar spine without contrast (CPT® 72148) preoperatively to rule out any associated spinal cord conditions when there is clinical or radiographic evidence of lumbar abnormalities.

### Background and Supporting Information

- This condition is also known as Scheuermann Kyphosis, and these individuals generally present with chronic and recurrent back pain.

### PEDSP-3.2: Scoliosis

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, detailed examination of the spine in different body positions, and plain radiography should be performed prior to considering advanced imaging.

  - Standing posteroanterior (PA) and lateral x-rays of the spine are the initial imaging studies and are used for follow-up. If anteroposterior (AP) x-rays are to be performed, breast shields should be used to reduce breast radiation exposure.

  - Spine surgical specialists sometimes appropriately request both MRI and CT together for preoperative planning of scoliosis surgery.
    - Concurrent requests for both MRI and CT will be forwarded for Medical Director Review.
    - MR or CT spine postoperative when recent postoperative x-rays are inconclusive for managing individual treatment.
- Chest CT with contrast (CPT® 71260) or without contrast (CPT® 71250) in the perioperative period as well as 2 and 5 years post operatively to access lung growth in individuals with severe scoliosis who may have compromised lung development.

**Background and Supporting Information**

Scoliosis is an abnormal lateral curve of the thoracic or thoraco-lumbar spine in the frontal plane. A small lateral curve is not uncommon and generally does not require further investigation.

- Using the Cobb technique for measuring these curves, a curve of under 10° is normal, a curve from 10 to 20° is mildly abnormal, a curve over 20° is significantly abnormal, and a curve > 40° is severely abnormal.
- Most individuals with significant scoliosis have some element of kyphosis as well.
  - There are many ways of classifying scoliosis. These guidelines will classify scoliosis as congenital, idiopathic, and neuromuscular scoliosis.
- In addition, MR and CT are useful to identify an underlying cause of scoliosis, such as congenital and developmental anomalies.
- If anteroposterior (AP) x-rays are to be performed, breast shields should be used to reduce breast radiation exposure.

**Congenital Scoliosis**

- In infants, spinal ultrasound (CPT® 76800) after initial imaging with plain x-rays.
- MRI cervical (CPT® 72156), thoracic (CPT® 72157), and lumbar (CPT® 72158) spine without and with contrast is indicated to search for underlying anomalies.
- Brain MRI without and with contrast if the clinical evaluation or preliminary imaging studies suggest an associated intracranial anomaly.
- Renal ultrasound (CPT® 76770 or CPT® 76775) should be performed, since nearly one-third of individuals also have genitourinary anomalies.
- CT, MRI, or nuclear medicine studies of the genitourinary tract may be necessary if the ultrasound is abnormal. These requests should be forwarded for Medical Director Review.

**Background and Supporting Information**

Cases are recognized in infancy or early childhood. Most cases arise from anomalies of vertebral development, and many are associated with anomalies of the genitourinary system or of other organs.
**Idiopathic Scoliosis**

The following clinical features are associated with an increased risk of underlying vertebral or spinal cord abnormality:

- Associated back pain
- Neurological abnormalities on examination or neurological symptoms.
- Left sided curve (concave to right)
- Double curves or high thoracic curves
- Spinal x-ray abnormalities other than the curve itself (widened spinal canal, dysplastic changes in spine or ribs, etc.)
- Midline spinal cutaneous markers (esp. sacral) such as dermal tracts, tufts of hair, skin tags, etc.
- Abnormal number or size of café au lait spots (neurofibromatosis)—these requests should be forwarded for Medical Director Review.

MRI without contrast of the symptomatic spinal region is the preferred study for the evaluation of scoliosis and should be approved when any of the above clinical features is present.

There is uncertainty regarding the clinical value of MRI in the routine evaluation or preoperative work-up of individuals with typical idiopathic scoliosis (with none of the above clinical features present).

- Noncontrast MRI or CT cervical, thoracic, and/or lumbar spine in these individuals when they are being actively evaluated for corrective surgery.

**Background and Supporting Information**

Idiopathic scoliosis is the most common form of pediatric scoliosis, and typically has its onset in late childhood or adolescence.

**Neuromuscular Scoliosis**

The appropriate spinal level, modality, and contrast level of advanced imaging will depend on the nature of the underlying disease.

- MRI without contrast or without and with contrast or CT without contrast cervical, thoracic, and/or lumbar spine in these individuals when they are actively being evaluated for spinal deformity corrective surgery.

- MRI without contrast or without and with contrast or CT without contrast of the symptomatic spinal region can be approved in individuals with painful neuromuscular scoliosis.

**Background and Supporting Information**

Scoliosis can result from many disorders of the nervous system. In some conditions, including (but not limited to) cerebral palsy, muscular dystrophy, and spinal muscular atrophy, associated scoliosis may develop over time.
References
PEDSP-4.1: Introduction

- A complete abdominal ultrasound (CPT® 76700) or retroperitoneal ultrasound (CPT® 76770) as an initial evaluation for individuals with newly diagnosed neurogenic bladder, myelomeningocele (open spinal dysraphism), hydronephrosis, or spina bifida.
  - A complete retroperitoneal ultrasound (CPT® 76770) every 6 to 12 months for follow-up/surveillance for any of the above conditions.

Background and Supporting Information

The term spinal dysraphism refers to a group of disorders characterized by incomplete or absent fusion of posterior midline structures, including neural, mesenchymal and cutaneous structures. Based on clinical classification, dysraphic are grouped into two categories: (a) open dysraphism (spina bifida aperta) which are non-skin-covered, open neural tube defects (myelomeningocele) and (b) closed or occult spinal dysraphism. The latter group includes skin-covered defects associated with a subcutaneous mass.

PEDSP-4.2: Cutaneous Lesions of the Back

- Screening MRI or Ultrasound is not necessary in the following clinical conditions, which are not significantly associated with spinal dysraphism:
  - “Simple dimple” which is defined as a midline soft tissue depression ≤ 2.5 cm above the anus (regardless of size or depth).
  - Deviated gluteal fold which is defined as any abnormal gluteal fold (including bifid or split gluteal cleft) without an underlying mass.
  - Coccygeal pits and pilonidal cysts at or below the level of the intergluteal fold.
  - Strawberry nevi
  - Non-specific darkened areas of skin over the sacrum (such as dermal melanosis) unless there are associated midline cutaneous abnormalities.

- Screening with advanced imaging is recommended in the following clinical conditions which are associated with an increased risk of underlying spinal dysraphism:
  - Dermal sinuses overlying the lumbar, thoracic, or cervical spine, and sacral dermal sinuses.
    - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age.
    - MRI of the involved spinal level without and with contrast if the ultrasound shows abnormalities other than a cutaneous dermal cleft.
    - MRI of the involved spinal level without and with contrast for initial evaluation in individuals older than 6 months of age.
    - Follow-up of a normal screening imaging study is not appropriate.
    - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.
Subcutaneous midline masses at any level, caudal extensions, midline skin tags, abnormal patches of hair over the spine, and complex midline birthmarks above the upper sacral region:
- Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age, but if a mass is present it is appropriate to proceed directly to MRI of the involved spinal level without and with contrast.
- MRI of the involved spinal level without and with contrast for initial evaluation in individuals older than 6 months of age.
- Follow-up of a normal screening imaging study is not appropriate.
- The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

Congenital anorectal abnormalities are often associated with dysraphism
- Lumbar spine MRI without and with contrast (CPT® 72158) when these are present.
- Follow-up of a normal screening imaging study is not appropriate.
- The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

Café au lait spots are a marker for type 1 neurofibromatosis
- See imaging indications in PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)
- Toe walking, when associated with upper motor neuron signs including hyperreflexia, spasticity, and positive Babinski sign

Background and Supporting Information
- The spinal cord arises from an infolding of the skin of the back, so certain lesions of the overlying skin are associated with an underlying spinal deformity, which include:
  - High risk dimples (greater than 5 mm in diameter and more than 2.5 cm above the anus)
  - Skin tags or tails
  - Hairy patches
  - Sinus tracts

PEDSP-4.3: Spina Bifida Occulta or Closed Spinal Dysraphism
These guidelines apply to adult as well as pediatric individuals.
- Unless additional abnormalities described above are present, routine advanced imaging is not indicated.
  - Cutaneous lesions below the gluteal crease are often pilonidal sinuses and need no further evaluation.
  - MRI of the involved spinal level without contrast or without and with contrast for tracts, pits, or lesions above the gluteal fold to evaluate further for underlying spinal pathology.


**PEDSP-4.4: Open Dysraphism**

- Clinically significant dysraphism includes findings ranging from complex vertebral anomalies to meningomyelocele.
  - MRI of the involved spinal level without contrast or without and with contrast is appropriate.
  - MRI cervical, thoracic, and lumbar spine without contrast or without and with contrast in individuals with open neural tube defects, or when ordered for preoperative planning.
  - MRI Brain or CT Head without contrast with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
  - MRI pelvis without contrast or without and with contrast if there are clinical signs of pelvic malformation or anorectal anomaly.
  - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

**References**

**Imaging Studies to Evaluate Tethered Cord**

- Spinal ultrasound (CPT® 76800) may be approved for initial evaluation in infants up to 6 months of age.
  - If the conus terminates below the L2-L3 disk space in a term infant the diagnosis of tethered cord is likely. Of note, however, in premature infants, the conus medullaris may be located at the mid L3-level if there is uncertainty as to whether cord termination is low, repeat spinal ultrasound can be performed in 4 to 6 weeks, since a normal cord will have "moved" higher within the spinal canal by this time.

- MRI lumbar spine without or without and with contrast may be approved for initial evaluation in individuals older than 6 months of age.
  - MRI studies to complete imaging of the entire spine (cervical, thoracic, and lumbar) without and with contrast if a tethered cord is found, to rule out associated spinal cord deformities such as syringomyelia. See **PEDSP-4: Spinal Dysraphism** for additional information.
  - MRI without and with contrast cervical (CPT® 72156), thoracic (CPT® 72157), and lumbar (CPT® 72158) spine for initial evaluation for individuals requiring general anesthesia to complete MRI,
  - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

**Background and Supporting Information**

**Normal position of spinal cord**
The conus medullaris in newborns should terminate at L2-3 or higher. After 3 months of age, the conus should lie at or above the L2 level. The spinal cord normally ends in the conus medullaris, which is positioned at L1-2 in normal infants and children.

**Tethered cord**
If the conus terminates below L2-3, the cord may be tethered by an abnormal structure. Abnormalities can be found in both lumbosacral and thoracic regions and are often associated with spinal lipomas in either region. Tethering is certain when the cord terminates at or below L4 and there is other supporting evidence of tethering such as limited spinal cord pulsatility, posterior positioning in the spinal canal, thick filum terminale, intraspinous mass, or lipoma.
References
PEDSP-6: Myelopathy

- Myelopathy imaging indications in pediatric individuals are similar to those for adult individuals. See **SP-7: Myelopathy** for imaging guidelines.
# PEDSP-7: Other Congenital and Pediatric Spine Disorders

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PEDSP-7.1: Achondroplasia

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- MRI without contrast of the symptomatic spinal region when new or worsening clinical symptoms suggest achondroplasia related spinal stenosis.
- Brain MRI without contrast (CPT® 70551) or Head CT without contrast (CPT® 70450) when new or worsening clinical symptoms suggest hydrocephalus.

Background and Supporting Information
The diagnosis of achondroplasia is made clinically. Achondroplasia individuals are at risk for hydrocephalus as well as myelopathy from spinal stenosis with increasing age.

PEDSP-7.2: Inflammatory Spondylitis

Except as listed below, imaging considerations in pediatric and adult individuals are identical for this condition, and these individuals should be imaged according to SP-10.2: Inflammatory Spondylitis.

For pediatric individuals with juvenile idiopathic arthritis:
- MRI without and with contrast is appropriate.
- An initial x-ray is not necessary prior to MRI in these individuals.
- SPECT bone scan (CPT® 78320) is indicated for evaluation of facet arthropathy in individuals with ankylosing spondylitis, osteoarthritis, or rheumatoid arthritis.

PEDSP-7.3: Atlantoaxial Instability in trisomy 21 (Down Syndrome)

The diagnosis of atlantoaxial instability is a recognized complication of trisomy 21, and individuals are routinely screened with lateral x-rays of the cervical spine.

- MRI cervical spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) in individuals where the lateral cervical spine x-ray demonstrates a pre dens interval of ≥ 5 mm, and a neural canal width of ≤ 14 mm.
- MRI cervical spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) when new or worsening clinical symptoms suggest myelopathy in a trisomy 21 individual.

PEDSP-7.4: Basilar Impression

See PEDHD-9.4: Basilar Impression for imaging guidelines.
**PEDSP-7.5: Chiari Malformation**
See **PEDHD-9: Chiari and Skull Base Malformations**

**PEDSP-7.6: Klippel-Feil Anomaly (congenital fusion of cervical vertebrae)**
This is generally an incidental finding. A detailed history and physical examination with thorough neurologic examination, and plain x-rays should be performed initially. Klippel-Feil can occur in conjunction with platybasia and/or Chiari malformation.

- Plain x-rays cervical spine to establish the diagnosis. Advanced imaging is indicated if there are acute or worsening neurologic symptoms (including pain), or if multiple levels are involved.
- MRI cervical spine without contrast (CPT® 72141) or CT cervical spine without contrast (CPT® 72125) for these indications.

**PEDSP-7.7: Marfan Syndrome**

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- MRI without contrast of the symptomatic spinal region when:
  - New or worsening clinical symptoms suggest a complicated dural ectasia
  - The individual is under active consideration for surgery

*Background and Supporting Information*
Marfan syndrome individuals are at risk for scoliosis (See **PEDSP-3.2**) and dural ectasias. Dural ectasias are usually asymptomatic but can be associated with other spinal lesions.

**PEDSP-7.8: Neurofibromatosis**
See **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines for screening recommendations in neurofibromatosis

See **PEDPN-2: Neurofibromatosis** for imaging considerations in neurofibromatosis individuals with known plexiform neurofibromas

See **PEDONC-8.3: Non-Rhabdomyosarcoma Soft Tissue Sarcomas** for imaging in individuals with neurofibromatosis and malignant peripheral nerve sheath tumors.
PEDSP-7.9: Von Hippel-Lindau Syndrome (VHL)

See PEDONC-2.10: Von Hippel-Lindau Syndrome (VHL) in the Pediatric Oncology Imaging Guidelines for screening recommendations in VHL individuals.

- MRI without and with contrast of the affected spinal level for individuals with known spinal hemangioblastomas in the following conditions:
  - Annually for asymptomatic individuals with unresected spinal hemangioblastoma(s)
  - Preoperative planning for resection of a hemangioblastoma
  - New or worsening symptoms suggesting progression of a known hemangioblastoma

References