

# CIGNA MEDICAL COVERAGE POLICIES – RADIOLOGY

## Pediatric and Special Populations Spine Imaging Guidelines

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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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# General Guidelines (PEDSP-1.0)

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# Procedure Codes Associated with Spine Imaging (PEDSPINE)

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Procedure Codes Associated with Spine Imaging	
MRI	CPT®
MRI Cervical without contrast	72141
MRI Cervical with contrast	72142
MRI Cervical without and with contrast	72156
MRI Thoracic without contrast	72146
MRI Thoracic with contrast	72147
MRI Thoracic without and with contrast	72157
MRI Lumbar without contrast	72148
MRI Lumbar with contrast	72149
MRI Lumbar without and with contrast	72158
MRI Unlisted procedure (for radiation planning or surgical software)	76498
MRA	CPT®
MRA Spinal Canal	72159
CT	CPT®
CT Cervical without contrast	72125
CT Cervical with contrast	72126

Procedure Codes Associated with Spine Imaging	
CT Cervical without and with contrast	72127
CT Thoracic without contrast	72128
CT Thoracic with contrast	72129
CT Thoracic without and with contrast	72130
CT Lumbar without contrast	72131
CT Lumbar with contrast	72132
CT Lumbar without and with contrast	72133
CT Pelvis without contrast	72192
CT Pelvis with contrast	72193
CT Pelvis without and with contrast	72194
CT Guidance for Placement of Radiation Therapy Fields	77014
CT Unlisted procedure (for radiation planning or surgical software)	76497
<b>Ultrasound</b>	<b>CPT®</b>
Ultrasound, spinal canal and contents	76800

# General Guidelines (PEDSP-1.0)

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- A pertinent clinical evaluation since the onset or change in symptoms, 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 imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) can serve as a pertinent clinical evaluation.
  - A thorough neurologic examination should include results of manual motor testing, specific dermatomal distribution of altered sensation, reflex examination, nerve root tension signs (e.g., straight leg raise test, slump test, femoral nerve tension test), and documentation of any specific radicular features.
- For those spinal conditions/disorders for which the Spine Imaging Guidelines require a plain x-ray of the spine prior to consideration of an advanced imaging study, the plain x-ray must be performed after the current episode of symptoms started or changed and results need to be available to the requesting provider of the advanced imaging study.
- 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 patient management or treatment decisions.

# Pediatric Spine Imaging Age Considerations (PEDSP-1.1)

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- 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 patient age, comorbidities, and differences in disease natural history between children and adults.
- Patients who are  $\leq 18$  years old should be imaged according to the Pediatric Spine Imaging Guidelines if discussed. Any conditions not specifically discussed in the Pediatric Spine Imaging Guidelines should be imaged according to the General Spine Imaging Guidelines. Individuals who are  $> 18$  years old should be imaged according to the General Spine Imaging Guidelines, except where directed otherwise by a specific guideline section.

# Pediatric Spine Imaging Appropriate Clinical Evaluation (PEDSP-1.2)

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- See: General Guidelines (PEDSP-1.0)



# Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)

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- 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 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.
    - 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 these 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 is the imaging study of choice in the setting of trauma
    - 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 individuals with fixation hardware which limits utility of MRI.
  - Severe congenital scoliosis with inconclusive MRI.
  - Evaluation of nerve root avulsion in patients 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<sup>®</sup> 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<sup>®</sup> 76800 for intraoperative spinal canal ultrasound as CPT<sup>®</sup> 76998 (intraoperative ultrasonic guidance) is the appropriate code in this circumstance.
  - Spinal canal ultrasound (CPT<sup>®</sup> 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: **Spinal Dysraphism (PEDSP-4)**).
    - 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-1)

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# Pediatric Back and Neck Pain and Trauma (PEDSP-2)

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# Introduction (PEDSP-2.1)

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- 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.

# Back and Neck Pain in Children Age 5 and Under (PEDSP-2.2)

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- A pertinent clinical 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 should be approved.
    - Individuals in this age group will require sedation to complete MRI imaging. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)** 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 patient 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

# Back and Neck Pain in Children Age 6 and Older (PEDSP-2.3)

SPP.TR.0002.3.C

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- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination including results of manual motor testing, the specific dermatomal distribution of altered sensation, reflex examination, and nerve root tension signs (e.g., straight leg raise test, slump test, femoral nerve tension test) and documentation of any specific radicular features, should be performed prior to considering advanced imaging.
- X-rays, while not required prior to conservative treatment, must be obtained before advanced imaging can be approved.
  - The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- 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
  - Constant or radicular pain lasting  $\geq 4$  weeks
  - 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 patients having undergone spinal surgery
  - Associated bowel or bladder dysfunction
- In the absence of any “red flags”, a recent (within 3 months) 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 patients with “red flag” findings, or who remain symptomatic after a 4 week trial of provider-supervised conservative treatment.

- The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- 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 ( $\geq 100^{\circ}$  F)
  - 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 a patient 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 patients but is uncommon in adolescents and rare in children.



## Spondylolysis (PEDSP-2.4)

SPP.TR.0002.4.C

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- A pertinent clinical 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.
  - MRI without contrast of the symptomatic spinal level is indicated to evaluate for stress reaction in bone and visualizing nerve roots if symptoms have continued despite a recent (within 3 months) provider-directed 4 week course of conservative care, or if 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 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.

# Spine Pain Due to Infectious Causes (PEDSP-2.5)

SPP.TR.0002.5.C

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

## **Initial Imaging Studies**

- Plain x-rays should be performed initially.
  - The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study.
- 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 clinically 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 including, but not limited to, discitis and vertebral osteomyelitis, typically present with sudden onset of back pain, fever, and elevated white blood cell count, occurring most commonly in the first decade of life.

# Spine Pain Related To Trauma and Painless Spine Trauma (PEDSP-2.6)

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- Imaging evaluation of traumatic spine injury in children is generally directed based on clinical examination. 60% to 80% of all spinal injuries in children involve the cervical spine as opposed to the thoracic spine and lumbar spine. Common causes are motor vehicle accidents, falls, and sports-related injuries.
- A pertinent clinical evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, should be performed prior to considering advanced imaging.
- When advanced imaging is appropriate, MRI without contrast or CT without contrast of the involved level is indicated as discussed in **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**
  - If the initial CT or MRI study is considered inconclusive, an exam of the other modality may be approved if needed to direct clinical management.

## **Cervical Spine**

- The results of plain x-rays performed after the current episode of symptoms started or changed need to be available to the requesting provider of the advanced imaging study
- Children under 3 years of age should be approved for advanced imaging of the cervical spine following a relevant recent x-ray when one or more of the following “red flags” are present:
  - Glasgow Coma Scale <14
  - Individual does not open eyes regardless of stimulus
  - Motor vehicle collision
- Children ≥3 years of age should be approved for advanced imaging of the cervical spine following a recent (within 60 days) x-ray when one or more of the following “red flags” are present:
  - Altered mental status
  - Focal neurologic findings
  - Neck pain
  - Torticollis not present prior to trauma
  - Substantial torso injury
  - Diving or head-first injury
  - High speed motor vehicle collision
  - Predisposing conditions, e.g. Down Syndrome

- Children older than 2 years of age **SHOULD NOT** be approved for advanced imaging of the cervical spine if they meet **ALL** of the following criteria:
  - Absence of posterior midline cervical pain
  - Absence of focal neurologic deficit
  - Normal level of alertness
  - No evidence of intoxication
  - Absence of other clinically apparent pain which could distract patient from the pain of a cervical injury

### **Thoracolumbar Spine**

- Children should be approved for advanced imaging of the thoracolumbar spine following a recent x-ray when x-rays are inconclusive, or there is an abnormal neurological examination.

### **Suspected Physical Child Abuse**

- In children with suspected physical child abuse and documented findings suggesting abuse (e.g., fractures on skeletal survey or other clinical indicators), MRI Cervical (CPT<sup>®</sup> 72141), Thoracic (CPT<sup>®</sup> 72146), and Lumbar (CPT<sup>®</sup> 72148) Spine without contrast are indicated to search for associated abnormalities.
  - If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast can be approved. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**

# References (PEDSP-2)

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# Kyphosis and Scoliosis (PEDSP-3)

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

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- This condition is also known as Scheuermann Kyphosis, and these individuals generally present with chronic and recurrent back pain.
- A pertinent clinical 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 varying between 20° and 45° 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.

## Scoliosis (PEDSP-3.2)

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- A pertinent clinical 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.
    - MR or CT Spine postoperative when recent postoperative x-rays are inconclusive for managing individual treatment.
      - CT Chest with contrast (CPT<sup>®</sup> 71260) or without contrast (CPT<sup>®</sup> 71250) may be obtained in the perioperative period as well as 2 and 5 years post operatively to assess lung growth in individuals with severe scoliosis 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 in a skeletally mature person 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.

### Congenital Scoliosis

- In infants under 6 months of age spinal ultrasound (CPT<sup>®</sup> 76800) can be approved after initial imaging with plain x-rays.
- MRI Cervical (CPT<sup>®</sup> 72141), Thoracic (CPT<sup>®</sup> 72146), and Lumbar (CPT<sup>®</sup> 72148) Spine without contrast are indicated to search for underlying anomalies.



- 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 approved. See: **Pediatric Spine Imaging Modality General Considerations (PEDSP-1.3)**
- MRI Brain without and with contrast if the clinical evaluation or preliminary imaging studies suggest an associated intracranial anomaly.
- Renal ultrasound (CPT<sup>®</sup> 76770 or CPT<sup>®</sup> 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.

### **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
  - Age younger than 10 years
  - Neurological abnormalities on examination or neurological symptoms
  - Left sided curve (concave to right)
  - Absence of apical segment lordosis/kyphosis
  - Rapid curve progression (>1 degree per month)
  - Pes Cavus (see: **Occult Spinal Dysprahism (PEDSP-4.3)**)
  - Double curves or high thoracic curves
  - Kyphosis
  - 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)
- MRI Cervical (CPT<sup>®</sup> 72141), Thoracic (CPT<sup>®</sup> 72146), and Lumbar (CPT<sup>®</sup> 72148) Spine without contrast is the preferred study for the evaluation of scoliosis and should be approved when any of the above clinical features is present or if imaging is requested for individuals who are being actively evaluated for corrective surgery.

## Background and Supporting Information

Idiopathic scoliosis is the most common form of pediatric scoliosis, and is divided into infantile (0-3 years of age), juvenile (4-9 years of age), and adolescent (10-17 years of age). Idiopathic scoliosis is defined as having no underlying structural abnormality or accompanying syndrome.<sup>10</sup>

### **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 of the cervical, thoracic, and/or lumbar spine can be approved in these individuals with painful neuromuscular scoliosis, or when they are actively being evaluated for spinal deformity corrective surgery.
- Post-surgical considerations are similar to adult post-operative indications (see: **Post-Operative Spinal Disorders (SP-15)** in the General Spine Imaging Guidelines) except as follows:
  - Post-operative CT Chest without contrast (CPT<sup>®</sup> 71250) with 3D reconstruction is indicated for lung volume measurement in children with early onset scoliosis, (e.g. congenital/thoracogenic type), due to risk of restrictive lung disease and thoracic insufficiency syndrome which occur from failure of spine and chest to support normal lung growth.<sup>2</sup>

## 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.

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# Spinal Dysraphism and Tethered Spinal Cord (PEDSP-4)

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# Introduction (PEDSP-4.1)

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## **Spinal Dysraphism**

- Spinal dysraphism refers to a group of disorders characterized by incomplete or absent fusion of posterior midline structures. This includes a range of congenital and/or developmental anomalies of the spinal cord and associated spinal structures that can affect any level of the spine, but most commonly the lumbosacral region.
- Based on clinical classification, dysraphism is grouped into two categories:
  - Open dysraphism (spina bifida aperta), which are non-skin-covered, open neural tube defects (myelomeningocele).
  - Occult spinal dysraphism (also called closed spinal dysraphism), which includes skin-covered defects (either with or without an associated subcutaneous mass).

## **Normal position of spinal cord**

- In newborns, the spinal cord should terminate (at the conus medullaris) at L2-3 or higher.
- By 3 months of age, the conus should lie at or above the L2 level.
- Afterwards, in normal infants and children, the conus medullaris should be positioned at L1-2.
- Of note, however, in premature infants, the conus medullaris may be located at the mid L3-level.
  - If such a finding on an initial spinal ultrasound results in uncertainty as to whether cord termination is low, repeat spinal ultrasound (CPT<sup>®</sup> 76800) can be performed in 4 to 6 weeks, since a normal cord will have “moved” higher within the spinal canal by this time.

## **Tethered cord**

- 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, intraspinal mass, or lipoma.
- If the conus terminates at a normal position (at L2-3 under 3 months of age, at L2 by 3 months of age, at L1-2 in older infants and children), the cord may still be tethered by an abnormal structure. Such tethering of the spinal cord can be found in some (but not all) patients with Occult Spinal Dysraphism. Abnormalities can be found in both lumbosacral and thoracic regions and are often associated with spinal lipomas in either region.
- Open Spinal Dysraphism is frequently associated with tethering of the spinal cord; symptoms of or findings from that tethering may manifest initially or may increase

after the newborn period and the initial imaging evaluation. See: **Open Dysraphism (PEDSP-4.4)**.

### **“Tethered cord Syndrome”**

- “Tethered Cord Syndrome” refers to symptoms and abnormal physical findings (such as low back or leg pain, decreased or absent lower extremity reflexes, urinary urgency, urinary incontinence, bowel incontinence, and constipation) that arise when a pathologic attachment causes abnormal spinal tension (increased by axial growth), with ensuing pathophysiologic effects. Some of these patients do have an abnormally low conus medullaris; other patients have other spinal abnormalities (such as spinal dysraphism) that causes the spinal cord to be abnormally tethered. Other patients with spinal dysraphism who may present with symptoms or findings suggestive of “Tethered Cord Syndrome” may have those clinical manifestations caused by primary dysplasia of neural tissue, instead of being caused by abnormal tethering. See: **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**.
- Not all anatomically tethered spinal cords result in symptoms of “Tethered Cord Syndrome.”

### **Imaging Studies to Evaluate Suspected Occult Spinal Dysraphism and/or Tethered Cord**

- Plain x-rays are not indicated for suspected Occult Spinal Dysraphism and/or Tethered Cord.
- Spina Bifida Occulta, an incomplete fusion of the posterior lumbosacral bony elements (present in about 25% of people), is often discovered as an incidental finding on x-rays and other imaging exams. In asymptomatic individuals it is of no consequence, and is not an indication for further imaging.
- A plain spine x-ray finding suggesting an absent or distorted pedicle (the “winking owl sign”) can be indicative of occult spinal dysraphism, for which an initial MRI without contrast or MRI without and with contrast of the appropriate spinal level can be approved.
- When indicated (See: **Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)**, **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**, and **Open Dysraphism (PEDSP-4.4)** for indications), the following imaging may be approved:
  - Spinal ultrasound (CPT<sup>®</sup> 76800) for initial evaluation in infants up to 6 months of age, in premature infants whose “corrected age” (subtracting the number of weeks of prematurity from the infant’s actual age) is less than or equal to 6 months, or in older individuals with open spinal dysraphism (see: **Open Dysraphism (PEDSP-4.4)**).
  - In a term infant, the diagnosis of tethered cord is likely if the conus terminates below the L2-L3 disc space. 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 in a premature infant, repeat spinal ultrasound (CPT<sup>®</sup> 76800) can be performed in 4 to 6 weeks, since a normal cord will have “moved” higher within the spinal canal by this time.

- MRI Cervical, Thoracic, and Lumbar spine without contrast (CPT<sup>®</sup> 72141, 72146, and 72148) or without and with contrast (CPT<sup>®</sup> 72156, 72157, and 72158) may be approved for initial evaluation in individuals older than 6 months of age.
  - MRI can be approved at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or when MRI imaging prior to 6 months of age has been ordered by (or in consultation with) an appropriate specialist for an indication from **Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)**, **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**, or **Open Dysraphism (PEDSP-4.4)**.
- The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually ordered by (or after consultation with) an appropriate specialist.
- Postoperative MRI is not done routinely but may be indicated if there are recurrent symptoms or findings suggesting recurrent tethering or other deterioration. Contrast level per ordering specialist.
- A complete abdominal ultrasound (CPT<sup>®</sup> 76700) or retroperitoneal ultrasound (CPT<sup>®</sup> 76770) can be approved as an initial evaluation for patients with newly diagnosed neurogenic bladder, myelomeningocele (open spinal dysraphism), or occult spinal dysraphism.
  - A complete retroperitoneal ultrasound (CPT<sup>®</sup> 76770) can be approved every 6 to 12 months for follow-up/surveillance for any of the above conditions.
- CT of the effected spinal level can be approved for surgical planning when a complex bony deformity of the spine is present, or when the Guidelines support doing MRI of the spine in a patient for whom MRI is contraindicated.

# Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.2)

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- More than 80% of individuals with Occult Spinal Dysraphism and/or Tethered Spinal Cord will have a cutaneous lesion overlying the lower spine.
- Spine imaging is NOT indicated in the following situations:
  - Pilonidal cysts below the level of the intergluteal fold.
    - For discussion of imaging in pilonidal cysts, see: **Pilonidal Cyst (PV-21.4)** in the Pelvis Imaging Guidelines
  - Non-specific darkened areas of skin over the sacrum (such as dermal melanosis) unless there are other associated midline cutaneous abnormalities
  - Occult bony dysraphism incidentally noted on x-ray
- Screening with advanced imaging IS recommended in the following clinical conditions which are associated with an increased risk of underlying spinal dysraphism:
  - Spinal dimples (midline soft tissue depression over the spine); or deviated or split (bifid) gluteal cleft
    - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Follow-up of a normal screening spinal ultrasound with ultrasound is not appropriate.
    - MRI of the involved spinal level without contrast or without and with contrast may be approved for initial evaluation in individuals older than 6 months of age. MRI can be approved at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.
    - A screening MRI can be approved after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not appropriate.
  - Dermal sinuses overlying the lumbar, thoracic, or cervical spine, and sacral dermal sinuses, whether manifested by a dermal sinus tract (a small opening in the skin, which leads into a narrow duct; it may be associated with protruding hairs) or a dermal cyst. They may be associated with an overlying or nearby hairy patch or vascular nevus
    - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Follow-up of a normal screening spinal ultrasound is not appropriate.



- MRI of the involved spinal level without contrast or without and with contrast should be approved if an ultrasound shows abnormalities other than a cutaneous dermal cleft, if ordered after 6 months of age, or at a younger age if ordered by (or in consultation with) an appropriate specialist.
- A screening MRI can be approved after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not appropriate.
- Subcutaneous midline masses (including cysts and lipomas) at any level.
  - Plain x-rays are not required to approve other imaging for midline masses overlying the spine when occult spinal dysraphism and/or tethered cord is suspected.
  - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age), but MRI of the involved spinal level without contrast or without and with contrast is the preferred initial imaging for midline masses overlying the spine. Repeat ultrasound follow-up of a normal screening spinal ultrasound is not appropriate.
  - MRI of the involved spinal level without contrast or without and with contrast may be approved for initial evaluation in patients older than 6 months of age. MRI can be approved at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.
  - A screening MRI can be approved after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not appropriate.
- Caudal extensions (including tail-like appendages), midline skin tags, abnormal patches of hair over the spine at any level, infantile hemangiomas overlying any spinal level, and complex midline birthmarks above the upper sacral region.
  - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not appropriate.
  - MRI of the involved spinal level without contrast or without and with contrast may be approved for initial evaluation in individuals older than 6 months of age. MRI can be approved at a younger age when there are symptoms or physical findings or concerning findings on ultrasound showing the need for more prompt MRI imaging, or if ordered by (or in consultation with) an appropriate specialist.
  - A screening MRI can be approved after a normal screening spinal ultrasound exam. Follow-up of a normal screening MRI imaging study is not appropriate.
- Café au lait spots are a marker for type 1 neurofibromatosis
  - See imaging indications in **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** and/or **Neurofibromatosis (PEDPN-2)**.

# Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)

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- Imperforate anus
- VACTERL (vertebral malformations, anal atresia, cardiac anomalies, tracheo-esophageal fistula, renal abnormalities, and limb defects) syndrome
- Currarino triad (sacral dysgenesis, presacral mass, anorectal malformation), OEIS (omphalocele, exstrophy, imperforate anus, spinal defects) syndrome
- Caudal regression syndrome
- Sacral agenesis (when 2 or more of the sacral vertebral bodies are absent; about 20% of children with sacral agenesis are not detected prior to age of 3 years).
- For all of the above conditions, the following imaging is indicated:
  - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not appropriate.
  - The following should be approved when requested: MRI Lumbar Spine without contrast (CPT<sup>®</sup> 72148) or without and with contrast (CPT<sup>®</sup> 72158); and/or MRI Pelvis without contrast (CPT<sup>®</sup> 72195) or MRI Pelvis without and with contrast (CPT<sup>®</sup> 72197).
  - Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
  - Follow-up of a normal screening MRI imaging study is not appropriate, but an initial MRI can be approved if the first screening study was an ultrasound.
  - Postoperative MRI is not done routinely but may be indicated if there are recurrent symptoms or findings suggesting recurrent tethering. Contrast level per ordering specialist.
- Rubinstein-Taybi syndrome (gait abnormalities, short stature, short limbs, characteristic facies, developmental delay, tethered spinal cord)
  - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age (or in premature infants with a “corrected” age up to 6 months of age). Repeat ultrasound follow-up of a normal screening spinal ultrasound is not appropriate.
  - MRI Lumbar spine without contrast (CPT<sup>®</sup> 72148) or without and with contrast (CPT<sup>®</sup> 72158) should be approved.

- Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
- Follow-up of a normal screening MRI imaging study is not appropriate, but an initial MRI can be approved if the first screening study was an ultrasound.
- Individuals with known DiGeorge Syndrome (22q11.2 deletion syndrome), when tethered cord syndrome or occult spinal dysraphism is suspected.
  - Spinal ultrasound (CPT<sup>®</sup> 76800) may be approved for initial evaluation in infants up to 6 months of age.
  - The following should be approved when requested: MRI Lumbar Spine without contrast (CPT<sup>®</sup> 72148) or without and with contrast (CPT<sup>®</sup> 72158)
  - Appropriate MRI (or other modality) imaging (including contrast level) of any other spinal level will depend on the nature of the underlying disease, usually ordered by (or in consultation with) an appropriate specialist.
  - Follow-up of a normal screening MRI imaging study is not appropriate, but an initial MRI can be approved if the first screening study was an ultrasound.
- Neurologic related symptoms and physical exam findings suggestive of occult spinal dysraphism or tethered cord syndrome and/or low lying conus medullaris (see: **Myelopathy (SP-7.1)** and **Myelopathy (PEDSP-6)**, and **Developmental Motor Delay (PEDHD-19.3)** for spinal cord involvement suspected in individuals with developmental motor delay) for which MRI of the involved spinal level without contrast or without and with contrast may be approved when any of the following are present:
  - Asymmetry of the feet, with one smaller foot, a high arch, and/or clawing of the toes. This is sometimes called the “neuroorthopedic syndrome”, and is associated with lack of an ipsilateral ankle jerk deep tendon reflex and calf atrophy.
  - Cavus foot (also called pes cavus or pes cavovarus)
  - Toe walking, when associated with upper motor neuron signs including hyperreflexia, spasticity, and positive Babinski sign
  - Ataxia (see: **Ataxia (PEDHD-20)**)
  - Absent perineal sensation
  - Lower urinary tract dysfunction, including urinary urgency or urinary incontinence. Though not a requirement for advanced imaging, some of these patients will have had abnormal urodynamic studies (such as cystometrography and/or sphincter electromyography).
  - Constipation, especially if there are abnormal physical exam findings related to the spine (such as lower extremity weakness, decreased lower extremity tone, abnormal lower extremity reflexes, a tuft of hair over the spine or covering a pilonidal dimple, a sacral dimple, gluteal cleft deviation, or absent anal or cremasteric reflex), failure of maximal laxative therapy (see: **Constipation, Diarrhea, and Irritable Bowel Syndrome (PEDAB-12)**) and/or bowel

incontinence, when tethered cord syndrome or occult spinal dysraphism is suspected as the cause

- Back or leg pain when tethered cord syndrome or occult spinal dysraphism is suspected as the cause. In this setting, neither a plain x-ray of the spine nor a recent period of provider directed conservative treatment is required to approve an MRI spine).

# Spinal Dysraphism (PEDSP-4.4)

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- Clinically significant dysraphism includes findings ranging from complex vertebral anomalies to myelomeningocele.
- Dysraphism is categorized into 2 major groups:<sup>1</sup>
  - Open Dysraphism - lack of skin covering with exposed neural elements
  - Closed Dysraphism - skin covered
- It is rare to perform MRI in neonates with open dysraphism as the diagnosis is usually made with obstetric ultrasound and confirmed with visual inspection
  - MRI of the entire spine may be approved for preoperative planning if ordered by a specialist.
- MRI Brain without contrast (CPT<sup>®</sup> 70551) or with and without contrast (CPT<sup>®</sup> 70553) is indicated in all cases of open dysraphism as Chiari II malformation will be present<sup>26</sup>
- Closed Dysraphism
  - MRI of the entire spine without contrast or without and with contrast is appropriate at the time of initial diagnosis.
    - MRI Brain without contrast (CPT<sup>®</sup> 70551) or without and with contrast (CPT<sup>®</sup> 70553) or CT without contrast of the brain (CPT<sup>®</sup> 70450) may be approved in cases with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
    - MRI Pelvis without contrast (CPT<sup>®</sup> 72195) or without and with contrast (CPT<sup>®</sup> 72196) may be approved once if there are clinical signs of pelvic malformation or anorectal anomaly.
  - MRI Cervical, Thoracic, and Lumbar spine without contrast (CPT<sup>®</sup> 72141, 72146, 72148) or without and with contrast (CPT<sup>®</sup> 72156, 72157, 72158) when ordered for preoperative planning.
  - Spinal canal ultrasound (CPT<sup>®</sup> 76800) may be approved as an alternative to MRI, if requested, in individuals with open dysraphism as the posterior bony defect provides an acoustic window for ultrasound.
  - MRI of the appropriate spinal level without contrast or without and with contrast may be approved when there are new and/or worsened neurologic symptoms and/or physical exam findings suggestive of new or worsened tethering of the spinal cord, such as any of the following:
    - New or worsened cavus foot
    - New or worsened toe walking and/or upper motor neuron signs (including hyperreflexia, spasticity, and positive Babinski sign)
    - New or worsened leg weakness or numbness or difficulty in ambulation
    - New or worsened loss of perineal sensation

- New or worsened lower urinary tract dysfunction (including urinary urgency or urinary incontinence, or new or worse changes on diagnostic urodynamic studies)
- New or worsened constipation
- New or worsened pain in the back or legs suspected to have been caused by tethering of the spinal cord
  - MRI Brain without contrast (CPT<sup>®</sup> 70551) or without and with contrast (CPT<sup>®</sup> 70553) or CT without contrast of the brain (CPT<sup>®</sup> 70450) may be approved in cases with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
  - MRI Pelvis without contrast (CPT<sup>®</sup> 72195) or without and with contrast (CPT<sup>®</sup> 72196) may be approved once 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.



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# Tethered Cord (PEDSP-5)

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## Tethered Cord (PEDSP-5)

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- See: Spinal Dysraphism and Tethered Spinal Cord (PEDSP-4)

# Myelopathy (PEDSP-6)

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# Myelopathy (PEDSP-6)

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Myelopathy imaging indications in pediatric individuals are similar to those for adult individuals. See: **Myelopathy (SP-7)** in the Spine Imaging Guidelines and/or **Non-Cutaneous Indications to Suspect Occult Spinal Dysraphism (PEDSP-4.3)**

# Other Congenital and Pediatric Spine Disorders (PEDSP-7)

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# General Guidelines - Other Congenital and Pediatric Spine Disorders (PEDSP-7.0)

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- Many congenital spine disorders also affect adults as survival continues to improve for these individuals. Adults with disorders covered in this section may follow these guidelines except where contraindicated by specific statements in the general imaging guidelines.

# Achondroplasia (PEDSP-7.1)

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- The diagnosis of achondroplasia is made clinically. Achondroplasia patients are at risk for hydrocephalus as well as myelopathy from spinal stenosis with increasing age.
- A pertinent clinical 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 or without and with of the symptomatic spinal region can be approved when new or worsening clinical symptoms suggest achondroplasia-related spinal stenosis.
- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) can be approved when new or worsening clinical symptoms suggest hydrocephalus.

# Inflammatory Spondylitis (PEDSP-7.2)

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

For pediatric patients with juvenile idiopathic arthritis:

- MRI without and with contrast or without contrast of the involved levels is appropriate.
- An initial x-ray is not necessary prior to MRI in these patients.



# Atlantoaxial Instability in Trisomy 21 (Down Syndrome) (PEDSP-7.3)

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- The diagnosis of atlantoaxial instability is a recognized complication of trisomy 21, and patients are routinely screened with lateral x-rays of the cervical spine.
- MRI Cervical Spine without contrast (CPT<sup>®</sup> 72141) or without and with contrast (CPT<sup>®</sup> 72156) in individuals where the lateral cervical spine x-ray demonstrates an atlantodental (pre-dens) interval of  $\geq 4.5$  mm, and/or a neural canal width of  $\leq 14$  mm.
- MRI Cervical Spine without contrast (CPT<sup>®</sup> 72141) or without and with contrast (CPT<sup>®</sup> 72156) when new or worsening clinical symptoms suggest myelopathy in a trisomy 21 individual.

## Basilar Impression (PEDSP-7.4)

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See: **Basilar Impression/Basilar Invagination (PEDHD-9.4)** in the Pediatric Head Imaging Guidelines

# Chiari Malformation (PEDSP-7.5)

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See: **Chiari and Skull Base Malformations (PEDHD-9)** in the Pediatric Head Imaging Guidelines

# Klippel-Feil Anomaly (Congenital Fusion of Cervical Vertebrae) (PEDSP-7.6)

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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 of the cervical spine are sufficient 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<sup>®</sup> 72141) or CT Cervical Spine without contrast (CPT<sup>®</sup> 72125) for these indications.

## Marfan Syndrome (PEDSP-7.7)

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Marfan syndrome patients are at risk for scoliosis (see **Scoliosis (PEDSP-3.2)**) and dural ectasias. Dural ectasias are usually asymptomatic but can be associated with other spinal lesions.

- A pertinent clinical 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 can be approved when:
  - New or worsening clinical symptoms suggest a complicated dural ectasia.
  - The individual is under active consideration for surgery.

## Neurofibromatosis (PEDSP-7.8)

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- See: **Neurofibromatosis 1 and 2 (NF1 and NF2) (PEDONC-2.3)** in the Pediatric Oncology Imaging Guidelines for screening recommendations in neurofibromatosis.
- See: **Neurofibromatosis (PEDPN-2)** in the Pediatric Peripheral Nerve Disorders Imaging Guidelines for imaging considerations in neurofibromatosis individuals with known plexiform neurofibromas.
- See: **Non-Rhabdomyosarcoma Soft Tissue Sarcomas (PEDONC-8.3)** in the Pediatric Oncology Imaging Guidelines for imaging in individuals with neurofibromatosis and malignant peripheral nerve sheath tumors.

# Von Hippel-Lindau Syndrome (VHL) (PEDSP-7.9)

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- See: **Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10)** in the Pediatric Oncology Imaging Guidelines for screening recommendations in VHL patients.
- MRI without and with contrast of the affected spinal level can be approved for patients with known spinal hemangioblastomas in the following conditions:
  - Annually for asymptomatic patients with unresected spinal hemangioblastoma(s).
  - Preoperative planning for resection of a hemangioblastoma.
  - New or worsening symptoms suggesting progression of a known hemangioblastoma.

## References (PEDSP-7)

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