



# CLINICAL GUIDELINES

## Pediatric Spine Imaging Policy

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eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies: This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

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## **Pediatric Spine Imaging Guidelines**

<b>Procedure Codes Associated with Spine Imaging</b>	<b>3</b>
<b>PEDSP-1: General Guidelines</b>	<b>5</b>
<b>PEDSP-2: Pediatric Back Pain</b>	<b>9</b>
<b>PEDSP-3: Kyphosis and Scoliosis</b>	<b>15</b>
<b>PEDSP-4: Spinal Dysraphism</b>	<b>19</b>
<b>PEDSP-5: Tethered Cord</b>	<b>23</b>
<b>PEDSP-6: Other Congenital and Pediatric Spine Disorders</b>	<b>25</b>

<b>Procedure Codes Associated with Spine Imaging</b>	
<b>MRI</b>	<b>CPT®</b>
Cervical MRI without contrast	72141
Cervical MRI with contrast	72142
Cervical MRI without and with contrast	72156
Thoracic MRI without contrast	72146
Thoracic MRI with contrast	72147
Thoracic MRI without and with contrast	72157
Lumbar MRI without contrast	72148
Lumbar MRI with contrast	72149
Lumbar MRI without and with contrast	72158
Unlisted MRI procedure (for radiation planning or surgical software)	76498
<b>MRA</b>	<b>CPT®</b>
Spinal Canal MRA	72159
<b>CT</b>	<b>CPT®</b>
Cervical CT without contrast	72125
Cervical CT with contrast	72126
Cervical CT without and with contrast	72127
Thoracic CT without contrast	72128
Thoracic CT with contrast	
Thoracic CT without and with contrast	72130
Lumbar CT without contrast	72131
Lumbar CT with contrast	72132
Lumbar CT without and with contrast	72133
Pelvis CT without contrast	72192
Pelvis CT with contrast	72193
Pelvis CT without and with contrast	72194
CT Guidance for Placement of Radiation Therapy Fields	77014
Unlisted CT procedure (for radiation planning or surgical software)	76497
<b>Nuclear Medicine</b>	<b>CPT®</b>
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging; skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging; whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
Bone Marrow Imaging Limited Areas	78102
Bone Marrow Imaging Multiple Areas	78103
Bone Marrow Imaging Whole Body	78104
Nuclear Bone Scan Limited	78300

Nuclear Bone Scan Multiple Areas	78305
Nuclear Bone Scan Whole Body	78306
Bone Scan Three Phase	78315
Nuclear Bone Scan SPECT	78320
Radiopharmaceutical Imaging of Inflammatory Process Limited Area	78805
Radiopharmaceutical Imaging of Inflammatory Process Whole Body	78806
Radiopharmaceutical Imaging of Inflammatory Process SPECT	78807
<b>Ultrasound</b>	<b>CPT®</b>
Ultrasound, spinal canal and contents	76800

**PEDSP-1: General Guidelines**

<b>PEDSP-1.1: Pediatric Spine Imaging Age Considerations</b>	<b>6</b>
<b>PEDSP-1.2: Pediatric Spine Imaging Appropriate Clinical Evaluation</b>	<b>6</b>
<b>PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations</b>	<b>6</b>

### **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 patient age, comorbidities, and differences in disease natural history between children and adults.

- Patients who are < 18 years old should be imaged according to the Pediatric Spine Imaging Guidelines, and patients 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 patient is undergoing guideline-supported scheduled imaging evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the spine is not supported. Advanced imaging of the spine should only be approved in patients 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.

### **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 patient 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 patient 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 patient 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 save 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 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® 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 patients with normal spinal ultrasound findings.

- Nuclear Medicine
  - ◆ Nuclear medicine studies are rarely used in the evaluation the spine, but are indicated in the following circumstances:
    - Bone scan (CPT® 78315 or CPT® 78320) is indicated for evaluation of suspected loosening of orthopedic prostheses when recent plain x-ray is nondiagnostic.
- 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.

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**PEDSP-2: Pediatric Back Pain**

<b>PEDSP-2.1: Introduction</b>	<b>10</b>
<b>PEDSP-2.2: Back Pain in Children Age 5 and Under</b>	<b>10</b>
<b>PEDSP-2.3: Back Pain in Children Age 6 and Over</b>	<b>11</b>
<b>PEDSP-2.4: Spondylolysis</b>	<b>12</b>
<b>PEDSP-2.5: Spine Pain Due to Infectious Causes</b>	<b>13</b>

## **PEDSP-2.1: Introduction**

- Currently, only about 40% 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 Pain in Children Age 5 and Under**

- A recent (within 60 days) 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 patients in this age group except those with mild and transient back pain.
  - ◆ MRI of the symptomatic spinal region should be approved
    - Patients 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 may be approved 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.
  - ◆ Bone scan is indicated for evaluation of suspected spinal fracture when x-ray is negative using any of the following CPT® code combinations:
    - CPT® 78300, CPT® 78305, or CPT® 78306 as a single study
    - CPT® 78315 or CPT® 78320 can be approved as a single study when stress fracture is suspected.
  - ◆ Bone scan is indicated for evaluation of suspected spondylolysis using any of the following CPT code combinations: SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period.
    - CPT® codes: CPT® 78300, CPT® 78305, CPT® 78306, CPT® 78315, or CPT® 78320 as a single study
    - CPT® 78305 and CPT® 78320 concurrently
    - CPT® 78306 and CPT® 78320 concurrently

### **PEDSP-2.3: Back Pain in Children Age 6 and Over**

Radicular back pain is common in adult patients but is uncommon in adolescents and rare in children.

- A recent (within 60 days) 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.
- Advanced imaging should be approved 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 patients 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 patients 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 a patient with an established diagnosis of cancer
- CT without contrast of the symptomatic spinal region may be approved 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 patient care decision making.

- Bone scan is indicated for evaluation of suspected spinal fracture when x-ray is negative using any of the following CPT® code combinations:
  - ◆ CPT® codes: CPT® 78300, CPT® 78305, or CPT® 78306 as a single study
  - ◆ CPT® 78315 or CPT® 78320 can be approved as a single study when stress fracture is suspected.

### **PEDSP-2.4: Spondylolysis**

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 patients.
- Surgical treatment is only recommended for patients with disabling symptoms that have not responded to non-surgical care.
- A recent (within 60 days) 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.
    - SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period.
  - ◆ 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 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. 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**

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.

- 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. Nuclear medicine imaging also can be positive as soon as 1 to 2 days after the onset of symptoms
- Any of the following studies are indicated for initial evaluation of suspected osteomyelitis:
  - ◆ Bone scan (one of CPT® codes: CPT® 78300, CPT® 78305, CPT® 78306, or CPT® 78315)
  - ◆ Nuclear Bone Marrow imaging (one of CPT® codes: CPT® 78102, CPT® 78103, or CPT® 78104)
  - ◆ Radiopharmaceutical inflammatory imaging (one of CPT® codes: CPT® 78805, CPT® 78806, or CPT® 78807)

#### **Follow-Up Imaging Studies**

- Follow-up plain x-rays may show disc space narrowing and bony changes of osteomyelitis.
- CT with contrast (including myelography) may be useful in follow-up for evaluating bony changes of osteomyelitis or concern for epidural abscess.
- Any of the following studies are indicated for evaluation of response to treatment in established osteomyelitis:
  - ◆ Bone scan (one of CPT® codes: CPT® 78300, CPT® 78305, CPT® 78306, or CPT® 78315)
  - ◆ Nuclear Bone Marrow imaging (one of CPT® codes: CPT® 78102, CPT® 78103, or CPT® 78104)
  - ◆ Radiopharmaceutical inflammatory imaging (one of CPT® codes: CPT® 78805, CPT® 78806, or CPT® 78807)

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## **PEDSP-3: Kyphosis and Scoliosis**

<b>PEDSP-3.1: Juvenile Thoracic Kyphosis (Scheuermann Disease)</b>	<b>16</b>
<b>PEDSP-3.2: Scoliosis</b>	<b>16</b>

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)**

- This condition is also known as Scheuermann Kyphosis, and these patients generally present with chronic and recurrent back pain.
- A recent (within 60 days) 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 patients is high.
- MRI without contrast of the thoracic spine (CPT® 72146) can be approved preoperatively to rule out any associated spinal cord problems.
- MRI without contrast of the lumbar spine CPT® 72148) can be approved preoperatively to rule out any associated spinal cord conditions when there is clinical or radiographic evidence of lumbar abnormalities.

### **PEDSP-3.2: Scoliosis**

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 patients 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.
- A recent (within 60 days) 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.
  - In addition, MR and CT are useful to identify an underlying cause of scoliosis, such as congenital and developmental anomalies.
  - Concurrent requests for both MRI and CT will be forwarded for Medical Director Review.

### **Congenital Scoliosis**

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.

- In infants, spinal ultrasound (CPT® 76800) can be approved after initial imaging with plain x-rays.
- MRI of the 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 can be approved 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 patients 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.

### **Idiopathic Scoliosis**

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

- 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 patients with typical idiopathic scoliosis (with none of the above clinical features present).
  - ◆ Noncontrast MRI or CT of the cervical, thoracic, and/or lumbar spine can be approved in these patients when they are being actively evaluated for corrective surgery.

### **Neuromuscular Scoliosis**

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.

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 patients when they are actively being evaluated for spinal deformity corrective surgery.
- Painful Scoliosis
- Bone scans are useful to evaluate cases of painful scoliosis and to identify tumors or infections. They are more sensitive than plain radiography.

### **References**

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## **PEDSP-4: Spinal Dysraphism**

<b>PEDSP-4.1: Introduction</b>	<b>20</b>
<b>PEDSP-4.2: Cutaneous Lesions of the Back</b>	<b>20</b>
<b>PEDSP-4.3: Spina Bifida Occulta or Closed Spinal Dysraphism</b>	<b>21</b>

## **PEDSP-4.1: Introduction**

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

## **PEDSP-4.2: Cutaneous Lesions of the Back**

- 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
- 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  $\leq 2.5$  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) may be approved for initial evaluation in infants up to 6 months of age.
    - MRI of the involved spinal level without and with contrast should be approved if the ultrasound shows abnormalities other than a cutaneous dermal cleft.

- MRI of the involved spinal level without and with contrast may be approved for initial evaluation in patients 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) may be approved 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 may be approved for initial evaluation in patients 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) should be approved 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)**

### **PEDSP-4.3: Spina Bifida Occulta or Closed Spinal Dysraphism**

These guidelines apply to adult as well as pediatric patients.

- 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.
  - ◆ Tracts, pits, or lesions above the gluteal fold should be evaluated further for underlying spinal pathology using MRI of the involved spinal level without contrast or without and with contrast.
- Clinically significant dysraphism includes findings ranging from complex vertebral anomalies to meningocele.
  - ◆ MRI of the involved spinal level without contrast or without and with contrast is appropriate.
  - ◆ MRI of the cervical, thoracic, and lumbar spine without contrast or without and with contrast may be approved in patients with open neural tube defects, or when ordered for preoperative planning.

- ◆ MRI Brain or CT Head without contrast of the brain may be approved in cases with associated hydrocephalus, signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
- ◆ MRI of the pelvis without contrast or without and with contrast may be approved 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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## PEDSP-5: Tethered Cord

### **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, intraspinal mass, or lipoma.

### **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 of the lumbar spine without or without and with contrast may be approved for initial evaluation in patients older than 6 months of age.
  - ◆ If a tethered cord is found, follow-up MRI studies to complete imaging of the entire spine (cervical, thoracic, and lumbar) without and with contrast should be approved to rule out associated spinal cord deformities such as syringomyelia. See **PEDSP-4: Spinal Dysraphism** for additional information.
  - ◆ For patients requiring general anesthesia to complete MRI, MRI without and with contrast of the cervical (CPT® 72156), thoracic (CPT® 72157), and lumbar (CPT® 72158) spine can be approved for initial evaluation.
  - ◆ 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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## **PEDSP-6: Other Congenital and Pediatric Spine Disorders**

<b>PEDSP-6.1: Achondroplasia</b>	<b>26</b>
<b>PEDSP-6.2: Inflammatory Spondylitis</b>	<b>26</b>
<b>PEDSP-6.3: Atlantoaxial Instability in trisomy 21 (Down Syndrome)</b>	<b>26</b>
<b>PEDSP-6.4: Basilar Impression</b>	<b>26</b>
<b>PEDSP-6.5: Chiari Malformation</b>	<b>26</b>
<b>PEDSP-6.6: Klippel-Feil Anomaly (congenital fusion of cervical vertebrae)</b>	<b>27</b>
<b>PEDSP-6.7: Marfan Syndrome</b>	<b>27</b>
<b>PEDSP-6.8: Neurofibromatosis</b>	<b>27</b>
<b>PEDSP-6.9: Von Hippel-Lindau Syndrome (VHL)</b>	<b>27</b>

### **PEDSP-6.1: Achondroplasia**

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 recent (within 60 days) 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 achondroplasia-related spinal stenosis.
- Brain MRI without contrast (CPT® 70551) or Head CT without contrast (CPT® 70450) can be approved when new or worsening clinical symptoms suggest hydrocephalus.

### **PEDSP-6.2: Inflammatory Spondylitis**

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

For pediatric patients with juvenile idiopathic arthritis:

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

### **PEDSP-6.3: Atlantoaxial Instability in trisomy 21 (Down Syndrome)**

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 of the cervical spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) can be approved in patients where the lateral cervical spine x-ray demonstrates a pre dens interval of  $\geq 5$  mm, and a neural canal width of  $\leq 14$  mm.
- MRI of the cervical spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) can also be approved when new or worsening clinical symptoms suggest myelopathy in a trisomy 21 patient.

### **PEDSP-6.4: Basilar Impression**

See **PEDHD-9.4: Basilar Impression** for imaging guidelines.

### **PEDSP-6.5: Chiari Malformation**

See **PEDHD-9: Chiari and Skull Base Malformations**

### **PEDSP-6.6: 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 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.
- Either MRI cervical spine without contrast (CPT® 72141) or CT cervical spine without contrast (CPT® 72125) can be approved for these indications.

### **PEDSP-6.7: Marfan Syndrome**

Marfan syndrome patients 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.

- A recent (within 60 days) 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 patient is under active consideration for surgery

### **PEDSP-6.8: 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 patients with known plexiform neurofibromas

See **PEDONC-8.3: Non-Rhabdomyosarcoma Soft Tissue Sarcomas** for imaging in patients with neurofibromatosis and malignant peripheral nerve sheath tumors.

### **PEDSP-6.9: 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 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

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