Cigna Medical Coverage Policies – Radiology
Pediatric Spine Imaging Guidelines

Effective February 17, 2020

Instructions for use

The following coverage policy applies to health benefit plans administered by Cigna. Coverage policies are intended to provide guidance in interpreting certain standard Cigna benefit plans and are used by medical directors and other health care professionals in making medical necessity and other coverage determinations. Please note the terms of a customer's particular benefit plan document may differ significantly from the standard benefit plans upon which these coverage policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a coverage policy.

In the event of a conflict, a customer's benefit plan document always supersedes the information in the coverage policy. In the absence of federal or state coverage mandates, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of:

1. The terms of the applicable benefit plan document in effect on the date of service
2. Any applicable laws and regulations
3. Any relevant collateral source materials including coverage policies
4. The specific facts of the particular situation

Coverage policies relate exclusively to the administration of health benefit plans. Coverage policies are not recommendations for treatment and should never be used as treatment guidelines.

This evidence-based medical coverage policy has been developed by eviCore, Inc. Some information in this coverage policy may not apply to all benefit plans administered by Cigna.

These guidelines include procedures eviCore does not review for Cigna. Please refer to the Cigna CPT code list for the current list of high-tech imaging procedures that eviCore reviews for Cigna.

Pediatric Spine Imaging Guidelines

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

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
Unlisted MRI 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
CT Cervical without and with contrast	72127
CT Thoracic without contrast	72128
CT Thoracic with contrast	
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
Unlisted CT procedure (for radiation planning or surgical software)	76497
Ultrasound	**CPT®**
Ultrasound, spinal canal and contents	76800

PEDSP-1: General Guidelines

PEDSP-1.1: Pediatric Spine Imaging Age Considerations

Many conditions affecting the spine in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to individual age, comorbidities, and differences in disease natural history between children and adults.

- Individuals who are <18 years old should be imaged according to the Pediatric Spine Imaging Guidelines, and individuals who are ≥18 years old should be imaged according to the Adult Spine Imaging Guidelines, except where directed otherwise by a specific guideline section.

PEDSP-1.2: Pediatric Spine Imaging Appropriate Clinical Evaluation

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with a thorough neurologic examination, appropriate laboratory studies, and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MRI, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the spine is not supported. Advanced imaging of the spine should only be approved in individuals who have documented active clinical signs or symptoms of disease involving the spine.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the spine are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect individual management or treatment decisions.

PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations

- MRI
  - MRI is the preferred modality for imaging the pediatric spine unless otherwise stated in a specific guideline section.
  - Due to the length of time 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 bases 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 eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.

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

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

- The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

References

1. Berland LL, Cernigliaro JG, Ho VB, et al. ACR Practice parameter for performing and interpreting magnetic resonance imaging (MRI). *American College of Radiology*. Revised 2017. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-perf-interpret.pdf?la=en.
2. Biassoni L, Easty M. Pediatric nuclear medicine imaging. Br Med Bull 2017;123:127-48.
3. Karmazyn BK, Dillman JR, Epelman MS, et al. ACR–ASER–SCBT-MR–SPR Practice parameter for the performance of pediatric computed tomography (CT) Revised 2014 (Resolution 3). *American College of Radiology*. 2014. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/ct-ped.pdf?la=en.
4. Ing C, DiMaggio C, Whitehouse A, et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. *Pediatrics.* 2012;130: (3). doi:10.1542/peds.2011-3822.
5. Monteleone M, Khandji A, Cappell J, et al. Anesthesia in children: perspectives from nonsurgical pediatric specialists. *J Neurosurg Anesthesiol.* 2014 Oct;26(4):396-398. doi:10.1097/ana.0000000000000124.
6. DiMaggio C, Sun LS, and Li G. Early childhood exposure to anesthesia and risk of developmental and behavioral disorders in a sibling birth cohort. *Anesth Analg.* 2011 Nov;113(5):1143-1151. doi: 10.1213/ANE.0b013e3182147f42.
7. Donohoe KJ, Brown ML, Collier D, et al. Society of nuclear medicine procedure guideline for bone scintigraphy, version 3.0 approved June 20, 2003. *Society of Nuclear Medicine procedure guidelines manual*. 2003 Aug. http://snmmi.files.cms-plus.com/docs/pg_ch34_0403.pdf.
8. Hochman MG, Melenevsky YV, Metter DF, et al. *ACR Appropriateness Criteria®.* Imaging after total knee arthroplasty. *American College of Radiology*. Date of origin: 1986. Last reviewed*:* 2017. https://acsearch.acr.org/docs/69430/Narrative/.
9. Cook GJR and I Fogelman. Bone single photon emission computed tomography. *The British Institute of Radiology.* 2001;13(3):149-154. doi:10.1259/img.13.3.130149.
10. Fraum TJ, Ludwig DR, Bashir MR, et al. Gadolinium-based contrast agents: a comprehensive risk assessment. *J Magn Reson Imaging.* 2017 Aug;46(2):338–353. doi:10.1002/jmri.25625.
11. FDA Medical Imaging Drug Advisory Committee meeting 9/8/17 Minutes. https://www.fda.gov/downloads/AdvisoryCommittees/CommitteesMeetingMaterials/Drugs/MedicalImagingDrugsAdvisoryCommittee/UCM574746.pdf.
12. Siegel MJ. Spinal Ultrasonography. Pediatric sonography. 5th ed. Philadelphia. Wolters Kluwer. 2018; 653-76.

PEDSP-2: Pediatric Back and Neck Pain and Trauma

PEDSP-2.1: Introduction

- Currently, only about 20% of back pain in children over age 5 is from a discoverable cause. Scoliosis, spondylitic disorders, Scheuermann disease, tumor, and trauma are the most common causes.
- Back pain in children under age 5 is uncommon and often reflects underlying serious disease when present.
- Disc herniations are rare in children, but become more frequent as activity increases during adolescence.

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

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- Advanced imaging is appropriate in all individuals in this age group except those with mild and transient back pain.
  - MRI of the symptomatic spinal region
    - Individuals in this age group will require sedation to complete MRI imaging. See **PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations** for contrast and body area considerations.
  - CT without contrast of the symptomatic spinal region when:
    - Plain x-rays suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group).
    - A recent MRI does not provide sufficient detail of the bony anatomy to allow for acute individual care decision making.

Background and Supporting Information

SPECT bone scans are especially sensitive for detecting spondylolysis, revealing areas of bone turnover; and the findings are generally positive for a prolonged period

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

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- X-rays, while not required prior to conservative treatment, must be obtained before advanced imaging can be approved.
- Advanced imaging should be approved following a recent x-ray when one or more of the following pediatric "red flags" are present:
  - Accompanying systemic symptoms (fever, weight loss, etc.)
  - Functional disability (daily limitation in normal activities because of pain)
  - Pain which is extremely severe or worse at night
  - Early morning stiffness
  - Pain which worsens despite an attempt at symptomatic treatment
  - Neurological symptoms or abnormal neurological examination findings
  - An established diagnosis of cancer other than leukemia
  - Abnormal x-rays
  - Spinal imaging for individuals having undergone spinal surgery
  - Associated bowel or bladder dysfunction
- In the absence of any "red flags", a 4 week trial of provider-supervised conservative treatment should be attempted before advanced imaging can be approved.
  - It can be assumed that children who are being evaluated by a pediatric spine surgeon have failed a reasonable trial of conservative treatment under the care of the primary care provider as this is by far the most common reason for such referrals.
- X-rays of the involved regions should be obtained prior to advanced imaging in individuals with "red flag" findings, or who remain symptomatic after a 4 week trial of provider-supervised conservative treatment.
- MRI without contrast of the symptomatic spinal region is the preferred study for the evaluation of pediatric spine pain, and should be approved unless one of the following conditions applies, in which case MRI without and with contrast should be approved:
  - Fever (≥100° F)
  - Clinical suspicion of infection (discitis, osteomyelitis, paraspinous or epidural abscess)
  - Physical examination or plain x-ray suggests a mass lesion
  - New or worsening pain in an individual with an established diagnosis of cancer
- CT without contrast of the symptomatic spinal region when:
  - The request is for re-evaluation of a known vertebral bony disorder.
  - Plain x-rays show spondylotic changes or suggest an isolated vertebral bone abnormality without any concern for spinal canal or cord abnormalities (which is rare in this age group).

- A recent MRI does not provide sufficient detail of the bony anatomy to allow for acute individual care decision making.

Background and Supporting Information

Radicular back and neck pain is common in adult individuals but is uncommon in adolescents and very rare in children.

PEDSP-2.4: Spondylolysis

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- Spondylolysis is best recognized on plain x-rays, and advanced imaging is generally not indicated.
  - If additional imaging is needed because of radiological uncertainty or associated spondylolisthesis, SPECT Radiopharmaceutical Localization Imaging (CPT® 78803) is indicated to identify stress reaction in early spondylolysis cases which are radiographically occult. Bone scan has been demonstrated to be superior to MRI in detecting active spondylolysis.
  - MRI without contrast of the symptomatic spinal level is indicated to evaluate for stress reaction in bone and visualizing nerve roots, if symptoms have continued despite a recent 4 week course of conservative care, or there is a documented need for preoperative planning.
  - CT without contrast of the symptomatic spinal level is indicated to provide detailed evaluation of bony anatomy, if there is a documented need for preoperative planning.

Background and Supporting Information

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

PEDSP-2.5: Spine Pain Due to Infectious Causes

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

Initial Imaging Studies

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

Follow-Up Imaging Studies

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

Background and Supporting Information

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

PEDSP-2.6: Spine Pain Related to Trauma

Imaging evaluation of traumatic spine injury in children is generally directed based on clinical examination.

- 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.
- Children under 3 years of age should be approved for advanced imaging of the cervical spine following a 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
- Older Children should be approved for advanced imaging of the cervical spine following a recent 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 injury
  - Motor Vehicle Collision

- 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 individual from the pain of a cervical injury
- 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.
- When advanced imaging is appropriate, MRI without contrast or CT without contrast of the involved level may be approved as discussed in **PEDSP-1.3: Pediatric Spine Imaging Modality General Considerations**
  - If the initial imaging study is considered inconclusive, an exam of the other modality may be approved if needed to direct clinical management

References

1. American College of Radiology.ACR Appropriateness Criteria® back pain-child; 2-16.
2. Booth TN, Iyer RS, Falcon RA, et al. ACR Appropriateness Criteria® back pain–child, American College of Radiology. 2016 Nov.
3. Calloni SF, Huisman TA, Poretti A, Soares BP. Back pain and scoliosis in children: When to image, what to consider. The neuroradiology journal. 2017 Oct;30(5):393-404.
4. Eckel T, Lehman R, and Paik H. Spondylolisthesis. *Scoliosis Research Society. Scoliosis Research Society E-Text* ©, 2017. http://etext.srs.org/book/.
5. Faingold R, Saigal G, Azouz EM, et al. Imaging of low back pain in children and adolescents. *Semin Ultrasound CT MR.* 2004 Dec;25(6):490-505. doi:10.1053/j.sult.2004.09.005.
6. Kjaer P, Leboeuf-Yde C, Sorensen JS, et al. An epidemiologic study of MRI and low back pain in 13-year-old children. *Spine.* 2005 Apr;30(7):798-806. doi:10.1097/01.brs.0000157424.72598.ec.
7. MacDonald J, Stuart E, Rodenberg R. Musculoskeletal low back pain in school-aged children: a review. JAMA pediatrics. 2017 Mar 1;171(3):280-7
8. Matesan M, Behnia F, Bermo M, et al. SPECT/CT bone scintigraophy to evaluate low back pain in young athletes: common and uncommon etiologies. J Ortho Surg 2016;11:76.
9. Mistovich RJ and Spiegel DA. The spine. *Nelson Textbook of Pediatrics*, *Chapter 679*. eds. Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition 2016;19:3283-3296.
10. Ramirez N, Flynn JM, Hill BW, et al. Evaluation of a systematic approach to pediatric. J Pediat Ortho 2015;35-28-32.
11. Rodriguez DP and Toussaint TY. Imaging of back pain in children. *AJNR Am J Neuroradiol.* 2010;31(5):787-802.
12. Taxter AH, Chauvin NA, Weiss PF. Diagnosis and treatment of low back pain in the pediatric population. Phys Sportsmed 2014;42:94-104.
13. Trout AT, Sharp SE, Anton CG, et al. Spondylolysis and beyond: value of SPECT/CT in evaluation of low back pain in children and young adults. Radiographic 2015;35:819-34.
14. Kadom N, Palasis S, Pruthi S, et al ACR Appropriateness Criteria® Suspected Spine Trauma–Child, American College of Radiology 2018.
15. Kim H, Crawford C, Ledonio C, et al. Current Evidence Regarding the Diagnostic Methods for Pediatric Lumbar Spondylolisthesis: A Report From the Scoliosis Research Society Evidence Based Medicine Committee. Spine Deform. 2018 Mar - Apr;6(2):185-188.
16. Oetgen ME. Current Use of Evidence-Based Medicine in Pediatric Spine Surgery. Orthopedic Clinics. 2018 Apr 1;49(2):191-4.

PEDSP-3: Kyphosis and Scoliosis

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

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

The term “scoliosis” refers to a lateral curvature.

PEDSP-3.1: Juvenile Thoracic Kyphosis (Scheuermann Disease)

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, and plain radiography should be performed prior to considering advanced imaging.
- X-rays will typically show anterior wedging in three or more adjacent vertebral bodies.
  - Lower thoracic kyphosis from developmental vertebral wedging with thoracic kyphosis totaling over 15˚ to 20˚ should be identified by plain x-rays before considering advanced imaging.
  - MRI is not an effective diagnostic modality for this condition since the incidence of false positive vertebral changes in normal individuals is high.
- MRI Thoracic Spine without contrast (CPT® 72146) preoperatively to rule out any associated spinal cord problems.
- MRI Lumbar Spine without contrast (CPT® 72148) preoperatively to rule out any associated spinal cord conditions when there is clinical or radiographic evidence of lumbar abnormalities.

Background and Supporting Information

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

PEDSP-3.2: Scoliosis

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with thorough neurologic examination and documentation of any specific radicular features, detailed examination of the spine in different body positions, and plain radiography should be performed prior to considering advanced imaging.
  - Standing posteroanterior (PA) and lateral x-rays of the spine are the initial imaging studies and are used for follow-up. If anteroposterior (AP) x-rays are to be performed, breast shields should be used to reduce breast radiation exposure.
  - Spine surgical specialists sometimes appropriately request both MRI and CT together for preoperative planning of scoliosis surgery.
    - Concurrent requests for both MRI and CT will be forwarded for Medical Director Review.
    - MRI or CT Spine postoperative when recent postoperative x-rays are inconclusive for managing individual treatment.

- CT Chest with contrast (CPT® 71260) or without contrast (CPT® 71250) in the perioperative period as well as 2 and 5 years post operatively to access lung growth in individuals with severe scoliosis who may have compromised lung development.

Background and Supporting Information

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

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

Congenital Scoliosis

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

Background and Supporting Information

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

Idiopathic Scoliosis

- The following clinical features are associated with an increased risk of underlying vertebral or spinal cord abnormality:
  - Associated back pain
  - Neurological abnormalities on examination or neurological symptoms.
  - Left sided curve (concave to right)
  - Double curves or high thoracic curves
  - Spinal x-ray abnormalities other than the curve itself (widened spinal canal, dysplastic changes in spine or ribs, etc.)
  - Midline spinal cutaneous markers (esp. sacral) such as dermal tracts, tufts of hair, skin tags, etc.
  - Abnormal number or size of café au lait spots (neurofibromatosis) - these requests should be forwarded for Medical Director Review.
- MRI without contrast of the symptomatic spinal region is the preferred study for the evaluation of scoliosis and should be approved when any of the above clinical features is present.
- There is uncertainty regarding the clinical value of MRI in the routine evaluation or preoperative work-up of individuals with typical idiopathic scoliosis (with none of the above clinical features present).
  - MRI or CT Cervical, Thoracic, and/or Lumbar Spine without contrast in these individuals when they are being actively evaluated for corrective surgery.

Background and Supporting Information

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

Neuromuscular Scoliosis

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

- MRI without contrast or without and with contrast or CT without contrast Cervical, Thoracic, and/or Lumbar Spine in these individuals when they are actively being evaluated for spinal deformity corrective surgery.
- MRI without contrast or without and with contrast or CT without contrast of the symptomatic spinal region can be approved in individuals with painful neuromuscular scoliosis.

Background and Supporting Information

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

References

1. ACR-SPR-SSR Practice parameter for the performance of radiography for scoliosis in children. Revised 2014.
2. Alsharief AN, El-Hawary R, Schmit P. Pediatric spine imaging post scoliosis surgery. Pediatric radiology. 2018 Jan 1;48(1):124-40.
3. Calloni SF, Huisman TA, Poretti A, Soares BP. Back pain and scoliosis in children: When to image, what to consider. The neuroradiology journal. 2017 Oct;30(5):393-404.
4. El-Hawary R, Chuckwunyerenwa C. Update on evaluation and treatment of scoliosis. Pediat Clin N Am. 2014;61:1223-41.
5. Kim H, Kim HS, Moon ES, et al. Scoliosis imaging: what radiologist should know. Radiographics 2010;30:1823-42.
6. Mayfair D, Flemming AK, Dvorak MR, et al. Radiographic evaluation of scoliosis: review. AJR 2010;194:S8-S22
7. Mistovich RJ and Spiegel DA. The Spine. *Nelson Textbook of Pediatrics*, *chapter 679*. eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition 2016;3283-3296.
8. Shafa E and Shah SA. Scheuermann Kyphosis. *Scoliosis Research Society E-Text* ©. 2019.
9. Oetgen ME. Current Use of Evidence-Based Medicine in Pediatric Spine Surgery. Orthopedic Clinics. 2018 Apr 1;49(2):191-4.

.

PEDSP-4: Spinal Dysraphism

PEDSP-4.1: Introduction

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

Background and Supporting Information

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

PEDSP-4.2: Cutaneous Lesions of the Back

- Screening MRI or Ultrasound is not necessary in the following clinical conditions, which are not significantly associated with spinal dysraphism:
  - "Simple dimple" which is defined as a midline soft tissue depression ≤2.5 cm above the anus (regardless of size or depth).
  - Deviated gluteal fold which is defined as any abnormal gluteal fold (including bifid or split gluteal cleft) without an underlying mass.
  - Coccygeal pits and pilonidal cysts at or below the level of the intergluteal fold.
  - Strawberry nevi
  - Non-specific darkened areas of skin over the sacrum (such as dermal melanosis) unless there are associated midline cutaneous abnormalities.
- Screening with advanced imaging is recommended in the following clinical conditions which are associated with an increased risk of underlying spinal dysraphism:
  - Dermal sinuses overlying the lumbar, thoracic, or cervical spine, and sacral dermal sinuses.
    - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age.
    - MRI of the involved spinal level without and with contrast if the ultrasound shows abnormalities other than a cutaneous dermal cleft.
    - MRI of the involved spinal level without and with contrast for initial evaluation in individuals older than 6 months of age.
    - Follow-up of a normal screening imaging study is not appropriate.
    - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

- Subcutaneous midline masses at any level, caudal extensions, midline skin tags, abnormal patches of hair over the spine, and complex midline birthmarks above the upper sacral region:
  - Spinal ultrasound (CPT® 76800) for initial evaluation in infants up to 6 months of age, but if a mass is present it is appropriate to proceed directly to MRI of the involved spinal level without and with contrast.
  - MRI of the involved spinal level without and with contrast for initial evaluation in individuals older than 6 months of age.
  - Follow-up of a normal screening imaging study is not appropriate.
  - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.
- Congenital anorectal abnormalities are often associated with dysraphism
  - MRI Lumbar Spine without and with contrast (CPT® 72158) when these are present.
  - Follow-up of a normal screening imaging study is not appropriate.
  - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.
- Café au lait spots are a marker for type 1 neurofibromatosis
  - See imaging indications in **PEDONC-2.3: Neurofibromatosis 1 and 2 (NF1 and NF2)** in the Pediatric Oncology Imaging Guidelines
- Toe walking, when associated with upper motor neuron signs including hyperreflexia, spasticity, and positive Babinski sign

Background and Supporting Information

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

PEDSP-4.3: Spina Bifida Occulta or Closed Spinal Dysraphism

These guidelines apply to adult as well as pediatric individuals.

- Unless additional abnormalities described above are present, routine advanced imaging is not indicated.
  - Cutaneous lesions below the gluteal crease are often pilonidal sinuses and need no further evaluation.
  - MRI of the involved spinal level without contrast or without and with contrast for tracts, pits, or lesions above the gluteal fold to evaluate further for underlying spinal pathology.

PEDSP-4.4: Open Dysraphism

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

References

1. Badve C, Phillips GS, Khanna PC, et al. MRI of closed spinal dysraphisms. Pediat Radiol. 2011;41:1308-20.
2. Ellenbogen RG. Neural tube defects in the neonatal period. *Medscape*. Version January 2, 2015. https://emedicine.medscape.com/article/1825866-overview.
3. Haynes, KB, Wimberly RL, VanPelt JM, et al. Toe walking: a neurological perspective after referral from pediatric orthopaedic surgeons. Journ of Ped Ortho. 2018;38(3):152-6.
4. Kim SM, Chang HK, Lee MJ, et al. Spinal dysraphism with anorectal malformation: lumbosacral magnetic resonance imaging evaluation of 120 patients. *J PediatrSurg.* 2010 Apr;45(4):769-776. doi: 10.1016/j.jpedsurg.2009.10.094.
5. Kinsman SL and Johnson MV. Congenital anomalies of the central nervous system. Nelson Textbook of Pediatrics, chapter 591. eds Kliegman RM, Stanton BF, St. Geme JW III, et al, 20th edition. 2016;2802-2819.
6. Kucera JN, Coley I, O'Hara, et al. The simple sacral dimple: diagnostic yield of ultrasound in neonates. Pediat Radiol. 2015;45:211-6.
7. Wang LL, Bierbrauer KS. Congenital and hereditary diseases of the spinal cord. Semin Ultrasound CT, MRI. 2017;38:105-25.
8. Warder DE. Tethered cord syndrome and occult spinal dysraphism. Neurosurg Focus. 2001 Jan;10(1):1-9. doi: 10.3171/foc.2001.10.1.2.

PEDSP-5: Tethered Cord

Imaging Studies to Evaluate Tethered Cord

- Spinal ultrasound (CPT® 76800) may be approved for initial evaluation in infants up to 6 months of age.
  - If the conus terminates below the L2-L3 disk space in a term infant the diagnosis of tethered cord is likely. Of note, however, in premature infants, the conus medullaris may be located at the mid L3-level if there is uncertainty as to whether cord termination is low, repeat spinal ultrasound can be performed in 4 to 6 weeks, since a normal cord will have "moved" higher within the spinal canal by this time.
- MRI Lumbar Spine without or without and with contrast may be approved for initial evaluation in individuals older than 6 months of age.
  - MRI studies to complete imaging of the entire spine (Cervical, Thoracic, and Lumbar) without and with contrast if a tethered cord is found, to rule out associated spinal cord deformities such as syringomyelia. See **PEDSP-4: Spinal Dysraphism** for additional information.
  - MRI Cervical (CPT® 72156), Thoracic (CPT® 72157), and Lumbar (CPT® 72158) Spine without and with contrast for initial evaluation for individuals requiring general anesthesia to complete MRI.
  - The appropriate spinal level, modality, and contrast level of follow-up advanced imaging will depend on the nature of the underlying disease, usually requested after specialist consultation.

Background and Supporting Information

Normal position of spinal cord

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

Tethered cord

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

Clinical findings which can be associated with tethered cord include low back or leg pain, decreased or absent lower extremity reflexes, urinary urgency and incontinence.

References

1. Farmakis SG and Siegel MJ. Spinal ultrasonogarphy. *Clinical Sonography: a practical guide*. eds. Sanders RC, and Hall-Terracciano B, 5th edition. 2016;657-669.
2. Halevi PD, Udayakumaran S, Ben-Sira L, et al. The value of postoperative MR in Childs Nerv Syst. 2011;27:2159-62.
3. Hertzler DA, DePowell JJ, Stevenson CB, et al. Tethered cord syndrome: a review of the literature from embryology to adult presentation. *Neurosurg Focus.* 2010 Jul;29(1):E1. doi: 10.3171/2010.3.FOCUS1079.
4. Hervey-Jumper SL, Garton HJL, Wetjen NM, et al. Neurosurgical management of congenital malformations and inherited disease of the spine. *Neuroimaging Clin N Am*. 2011 Aug;21(3):719-731. doi: 10.1016/j.nic.2011.05.009.
5. Ladino Torres MF, and DiPietro MA. Spine ultrasound imaging in the newborn. Seminars in Ultrasound, CT, and MRI. 2014;35(6)652-661. doi: 10.1053/j.sult.2014.08.001.
6. Siegel MJ. Spinal ultrasonography. *Pediatric sonography*. 5th ed. Philadelphia. Wolters Kluwer. 2018;653-76.
7. Rekate, HL Tethered Cord. Nelson Textbook of Pediatrics, Chapter 606. eds. Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition 2016:2952-2953.
8. Moore, KR. “Congenital Abnormalities of the Spine (Chapter 43)” in Caffey’s Pediatric Diagnostic Imaging. 13th edition Brian Coley editor, Elsevier Saunders, Philadelphia PA, 2019. 408-418.

PEDSP-6: Myelopathy

- Myelopathy imaging indications in pediatric individuals are similar to those for adult individuals. See **SP-7: Myelopathy** in the Spine Imaging Guidelines

PEDSP-7: Other Congenital and Pediatric Spine Disorders

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

PEDSP-7.1: Achondroplasia

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

Background and Supporting Information

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

PEDSP-7.2: Inflammatory Spondylitis

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

For pediatric individuals with juvenile idiopathic arthritis:

- MRI without and with contrast of the involved levels is appropriate.
- An initial x-ray is not necessary prior to MRI in these individuals.
- SPECT Radiopharmaceutical imaging (CPT® 78803) is indicated for evaluation of facet arthropathy in individuals with ankylosing spondylitis, osteoarthritis, or rheumatoid arthritis.

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

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

- MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) in individuals where the lateral cervical spine x-ray demonstrates a pre dens interval of ≥4.5 mm, and a neural canal width of ≤14 mm.
- MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) when new or worsening clinical symptoms suggest myelopathy in a trisomy 21 individual.

PEDSP-7.4: Basilar Impression

See **PEDHD-9.4: Basilar Impression** in the Pediatric Head Imaging Guidelines.

PEDSP-7.5: Chiari Malformation

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

PEDSP-7.6: Klippel-Feil Anomaly (congenital fusion of cervical vertebrae)

This is generally an incidental finding. A detailed history and physical examination with thorough neurologic examination, and plain x-rays should be performed initially. Klippel-Feil can occur in conjunction with platybasia and/or Chiari malformation.

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

PEDSP-7.7: Marfan Syndrome

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

Background and Supporting Information

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

PEDSP-7.8: Neurofibromatosis

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

See **PEDPN-2: Neurofibromatosis** in the Pediatric Peripheral Nerve Disorders Imaging Guidelines for imaging considerations in neurofibromatosis individuals with known plexiform neurofibromas

See **PEDONC-8.3: Non-Rhabdomyosarcoma Soft Tissue Sarcomas** in the Pediatric Oncology Imaging Guidelines for imaging in individuals with neurofibromatosis and malignant peripheral nerve sheath tumors.

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

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

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

References

1. Child AH. Non-cardiac manifestations of marfan syndrome. Ann Cardiothorac Surg. 2017;6:599-609.
2. Frantzen C, Klasson TF, Links TP, et al. Von Hippel-lindau disease. *GeneReviews*™.[Internet] eds. Pagon RA, Adam MP, Bird TD et al. https://www.ncbi.nlm.nih.gov/books/NBK1463/.
3. Jaremko JL, Liu L, Winn NJ, et al. Diagnostic utility of magnetic resonance imaging and radiography in juvenile spondyloarthritis: evaluation of the sacroiliac joints in controls and affected subjects. J Rheumatol. 2014;41:963-70. doi:10.3899/jrheum.131064.
4. Kao SC, Waziri MH, Smith WL, et al. MR imaging of the craniovertebral junction, cranium, and brain in children with achondroplasia. American Journal of Roentgenology. 1989 Sep: 153(3):565-9. doi: 10.2214/ajr.153.3.565.
5. Lambert RG, Bakker PA, van der Heijde D, et al. Defining active sacroiliitis on MRI for classification of axial spondyloarthritis: update by the ASAS MRI working group [epub ahead of print]. Ann Rheum Dis. 2016. doi:10.1136/annrheumdis-2015-208642.
6. Lin C, MacKenzie JD, Courtier JL, et al. Magnetic resonance imaging findings in juvenile spondyloarthropathy and effects of treatment observed on subsequent imaging. Ped Rheumat. 2014;12:25. doi:10.1186/1546-0096-12-25.
7. Rossi A. Pediatric spinal infection and inflammation. Neuroimaging Clinics. 2015 May 1;25(2):173-91.
8. Restropo R, Lee EY, and Babyn PS. Juvenile idiopathic arthritis: Current practical imaging assessment with emphasis on magnetic resonance imaging. *Radiol Clin N Am*. 2013 Jul;51(4):703-719. doi:10.1016/j.rcl.2013.03.003.
9. Smoker WRK and Khanna G. Imaging the craniocervical junction. *Childs Nerv Syst.* 2008 Oct; 24(10):1123-1145. doi: 10.1007/s00381-008-0601-0.
10. Vezina G, Barkovich AJ. Neurocutaneous disorders. In: Barkovich AJ, Raybaud C, eds. Pediatric Neuroimaging, 6th ed. Philadelphia PA. Wolters Kluwer. 2015;633-702.
11. White KK, Bompadre V, Goldberg MJ, et al. Best practices in the evaluation and treatment of foramen magnum stenosis in achondroplasia during infancy. Am J MedGenet A. 2016;170A:42-51.
12. Dweck J, Lachman RS “Skeletal Dysplasias and Selected Chromosomal Disorders (Chapter 132)” in Caffey’s Pediatric Diagnostic Imaging. 13th edition Brian Coley editor, Elsevier Saunders, Philadelphia PA, 2019. 1258-1295.