

PEDIATRIC AND CONGENITAL IMAGING GUIDELINES
SPINE and PERIPHERAL NERVE DISORDERS
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MedSolutions, Inc. Clinical Decision Support Tool
for Advanced Diagnostic Imaging

Common symptoms and symptom complexes are addressed by this tool. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician may provide additional insight.

This version incorporates MSI accepted revisions prior to 11/30/08

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PEDIATRIC AND CONGENITAL SPINE IMAGING GUIDELINES

PACSP-1~GENERAL GUIDELINES

- The Spine Imaging Guidelines are the same for both the pediatric population and the adult population, unless there are specific guidelines listed here in the Pediatric and Congenital Spine Imaging Guidelines.

PACSP-2~PEDIATRIC BACK PAIN

PACSP-2.1 Introduction

- Traditionally, it has been thought that back pain in children and adolescents, especially in those under age 10, was both very uncommon and usually a sign of serious disease. In the past twenty years, back pain in this population has become more common, and at the same time the incidence of benign back pain of no discoverable cause has risen sharply, though not yet to adult levels.
- Currently, 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 appears to remain quite uncommon and often to reflect underlying serious disease when it occurs.
- Disc herniations are rare in children, but become less infrequent during adolescence.
- **References:**
 - *Southern Med J* 1997;90:789-792
 - *Arch Dis Child* 2005;90:312-316
 - *Spine* 2005;30:798-806

PACSP-2.2 Back pain in children age 5 and under

- A detailed physical examination and plain x-rays should be performed initially.
- Advanced imaging is appropriate in all patients in this age group except those with mild and transient back pain
 - MRI (contrast as requested) of the symptomatic spinal region is usually appropriate, but noncontrast CT may be preferred based on the x-ray findings.
 - At times, CT is performed because the child cannot cooperate for MRI.

PACSP-2.3 Simple back pain in children age 6 or over

- A detailed physical/neurological examination should be performed initially.
- Plain x-rays should be performed before advanced imaging is considered.
- **Advanced imaging is appropriate when certain pediatric “red flags” are present:**
 - Accompanying systemic symptoms (fever, weight loss, etc.)
 - Pain which is extremely severe or worse at night.
 - Pain which worsens despite an attempt at symptomatic treatment.
 - Neurological symptoms or abnormal neurological examination findings.
 - The known presence of systemic cancer.
 - Abnormal x-rays.

- MRI without contrast of the symptomatic spinal region is usually the appropriate study.
- MRI without and with contrast should be performed in the setting of fever, clinical suspicion of infection (discitis, osteomyelitis, paraspinous abscess), or in patients with cancer.
- CT without contrast may be appropriate when the request is based on abnormal plain x-ray results, especially spondylitic changes.
- For diagnosis of pars defects, NM Tc-99m MDP is very sensitive and specific and is an appropriate alternative to CT or MRI.
- In the absence of any “red flags” (see above), a 4 week trial of physician-directed conservative treatment should be attempted before considering advanced imaging.
 - It can be assumed that children who are being evaluated by a pediatric spine specialist have failed a reasonable trial of conservative treatment. This is by far the most common reason for such referrals.
 - Preferences of such specialists to perform CT over MRI are generally acceptable.

PACSP-2.4 Spondylolysis

- Spondylolysis is believed to be caused by repeated microtrauma, resulting in stress fracture of the pars interarticularis. Heredity is also believed to be a factor.¹
 - Immobilization with various corsets or braces and activity restriction are the initial treatment for symptomatic patients.²
 - Surgical treatment is only recommended for very symptomatic patients in whom symptoms are disabling and have not responded to non-surgical care.²
 - Spondylolysis is best recognized on plain x-rays, and advanced imaging is generally not indicated.
 - If imaging is needed because of radiological uncertainty or associated spondylolisthesis, noncontrast CT or MRI can be performed.
 - MRI must be performed at minimum on a 1.0 Tesla scanner with 3 mm cuts and at relatively high resolution.¹
- ¹ Weinberg E. Spondylolysis. eMedicine. <http://www.emedicine.com/radio/topic650.htm>. Accessed October 13, 2007
- ² Spinal Deformity Information, Spondylolisthesis. Scoliosis Research Society. <http://www.srs.org/professionals>. Accessed October 13, 2007

PACSP-2.5 Spinal radiculopathy (cervical, thoracic, or lumbar)

- This is uncommon in adolescents under age 18 and rare in children.
- Documentation of specific radicular features by detailed history and neurological examination should be performed initially.
- Specialty consultation is very useful.
- Since this is an unusual diagnosis in children and younger adolescents, establishing the cause from the onset is important.
- Once the diagnosis is clinically confirmed, spinal MRI of the involved level is appropriate in patients ages 17 and younger.
 - In adolescents, MRI without contrast is usually sufficient
 - MRI without and with contrast is acceptable in children under age 12.

PACSP-3~KYPHOSIS AND SCOLIOSIS

- The term “kyphosis” refers to a curve convex posteriorly
 - Kyphosis generally affects the thoracic spine
- The term “lordosis” implies a curve convex anteriorly
- “Scoliosis” refers to lateral curvature

PACSP- 3.1 Thoracic Kyphosis and Scheuermann Disease

- These patients generally present with chronic and recurrent back pain.
- Careful physical/neurological examination and x-rays should be performed initially.
- Lower thoracic kyphosis from developmental vertebral wedging with thoracic kyphosis totaling over 15°-20° should be identified by plain x-rays before considering advanced imaging.
- If advanced imaging is indicated, noncontrast thoracic MRI (CPT 72146) can be performed.
 - Noncontrast lumbar MRI (CPT 72148) can be added when there are low back complaints as well.
- MRI of the thoracic spine is carried out preoperatively to rule out any associated spinal cord problems.¹
 - This modality is not a diagnostic tool since the incidence of false positive vertebral changes in normal patients is high.²

¹ Byrne LJ and McCormack MS. *The Orthopaedic management of Scheuermann's kyphosis*. <http://www.iol.ie/~rcsiorth/journal/volume5/issue1/sch.htm>. Accessed December 21, 2007

² *J Bone Joint Surg Am* 1995 Nov;77(11):1631-1638

PACSP- 3.2 Scoliosis

- Scoliosis an abnormal lateral curve of the thoracic or thoraco-lumbar spine in the frontal plane. A small lateral curve is not uncommon.
 - Using the Cobb technique for measuring these curves, a curve of under 10° is clearly normal, 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, developmental, and neuromuscular scoliosis.
 - Developmental scoliosis (onset in childhood or adolescence) is the most common.
 - Most developmental scoliosis, especially in adolescents, is idiopathic (no known cause).
- The initial step in the evaluation of all patients with scoliosis is a careful neurological examination, including detailed examination of the spine in different body positions.
- Spinal anteroposterior (AP) and lateral x-rays are the initial imaging study.
- **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 is often useful as a second imaging procedure (following x-rays).
- MRI of the entire spine, generally without contrast, can be performed to search for anomalies.
 - Sagittal/coronal screening of the spine may be appropriate for this, if available (see SP-2 Imaging Techniques in the adult Spine guidelines).
 - By convention, sagittal/coronal screening studies of the entire spine are coded as one segment (cervical [CPT 72141], thoracic [CPT 72146], or lumbar [72148]), whichever is most appropriate.
- Brain MRI, usually without contrast (CPT 70551), is appropriate if the clinical evaluation or preliminary imaging studies suggest an associated cerebral anomaly.
- Renal ultrasound should be performed, since about a third of patients also have genitourinary anomalies.
 - Advanced imaging of the genitourinary tract will be appropriate in many cases, especially if the ultrasound is abnormal.
- **Developmental scoliosis** (especially adolescent and late childhood scoliosis)
 - Most cases are idiopathic, especially in adolescent girls.
 - The following clinical features have been identified which make the presence of an underlying spinal or spinal cord abnormality more likely:
 - Associated prominent back pain
 - Neurological abnormalities on examination or striking 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)
 - Spinal MRI, generally without contrast, should be performed when any of the above clinical features is present.
 - Imaging of the cervical spine may or may not be included depending on spine specialist preference.
 - **Typical idiopathic scoliosis**
 - There is uncertainty regarding the value of MRI in the evaluation or preoperative work-up of patients with typical idiopathic scoliosis (with none of the above clinical features present).
 - Noncontrast MRI of the entire spine or thoracic/lumbar spine can be performed in these patients when they are actively being evaluated for corrective surgery.
- **Neuromuscular scoliosis**
 - Scoliosis can result from many disorders of the nervous system. In some, the presence of scoliosis suggests local disease in the spine or spinal cord, but not in others.
 - The appropriateness of advanced imaging will depend on the nature of the underlying disease.

- **References:**

- Boas SR. *Kyphoscoliosis: Adolescent Idiopathic Scoliosis and Congenital Scoliosis* (Chapter 416.5). In Kliegman RM, Behrman RE, Jenson HB, et al. (Eds.) *Nelson Textbook of Pediatrics*. 18th Ed. Philadelphia, Elsevier, 2007, pp. 1843-1844
- Spiegel DA, Hosalkar HS, Dormans JP. *The Spine*. (Chapter 678). In Kliegman RM, Behrman RE, Jenson HB et al. (Eds.). *Nelson Textbook of Pediatrics*. 18th Ed. Philadelphia, Elsevier, 2007, pp.2811-2815
- *J Bone Joint Surg Am* 2001;83:577-579
- *J Bone Joint Surg Am* 2002;84:2230-2234

PACSP-4~OTHER CONGENITAL AND PEDIATRIC SPINE DISORDERS

PACSP- 4.1 Achondroplasia

- The diagnosis of achondroplasia is made clinically.
- Achondroplastic patients are at risk for hydrocephalus, and with age, myelopathy from spinal stenosis.
- Noncontrast spine MRI directed at the appropriate clinical level can be performed.
- Brain MRI without contrast (CPT 70551) is appropriate if hydrocephalus is reasonably suspected.
- This guideline also applies to adults.

PACSP- 4.2 Ankylosing spondylitis

- Also see SP-9.5 Ankylosing Spondylitis in the adult Spine guidelines
- 97% of patients are HLA B-27 positive
- Both a positive HLA test result and plain x-rays should precede consideration of advanced imaging.
- Advanced imaging is not generally useful in this condition.
- If there are specific neurological problems, imaging by noncontrast MRI of the relevant spinal level is appropriate to the problem.

PACSP- 4.3 Basilar impression

- Basilar impression involves malformation of the occipital bone in relation to C1/2 (cervical vertebrae 1 and 2). The top of the spinal cord is inside the posterior fossa and the foramen magnum is undersized. Over time this can lead to brain stem and upper spinal cord compression.
- Basilar impression can also be associated with the Chiari malformation, producing very complex anatomical abnormalities.
- Noncontrast MRI of the brain (CPT 70551) and cervical spine (CPT 72141) are appropriate, and if surgery is contemplated, noncontrast CT of both may also be appropriate.
- Basilar impression appears to be partly genetic, and familial screening using noncontrast head MRI (CPT 70551) may be appropriate.
- **Reference:**
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.306-307

PACSP- 4.4 Chiari malformation

- Klippel-Feil anomaly (see PACSP-4.5 below) is often also seen
- **Chiari malformation (Chiari I; formerly called Arnold-Chiari)** is location of the cerebellar tonsils at least 5 mm below the foramen magnum.
 - Most patients have no or very vague symptoms, and diagnosis is usually made on head MRI done for other purposes.
 - A significant minority of these patients have an associated syringomyelia or hydromyelia.
 - A small number have hydrocephalus.
 - Noncontrast brain MRI (CPT 70551) is appropriate if not already performed.
 - If the Chiari malformation has been identified, noncontrast MRI spine (CPT 72141 with or without also performing CPT 72146) is recommended to exclude syrinx.
 - Follow-up spinal MRI without and with contrast (CPT 72156 with or without CPT 72157) will be needed if hydro/syringomyelia is seen (see SP-15 Syringomyelia in the adult Spine guidelines).
 - Spinal CT is inferior to spinal MR in the evaluation of syringomyelia.
 - MRA and CTA of either head or neck are not generally indicated in the evaluation of syringomyelia unless ordered by the operating surgeon for preoperative planning.
 - Once the diagnosis has been established by MRI, repeat brain MRI is generally appropriate only in patients with increasing symptoms or signs, or as a preoperative study.
 - CSF flow studies may be appropriate in selected patients with evidence of hydrocephalus (see HD-35.5 CSF flow imaging in the adult Head guidelines), but the coverage policy of the involved health plan regarding this study should be consulted.
 - Chiari malformation is not itself familial, and family screening is not appropriate.
- Chiari II, III, and IV are very rare and involve much more extensive malformations at all levels of the neural axis which are not further discussed in these guidelines.

PACSP- 4.5 Klippel-Feil anomaly (congenital fusion of cervical vertebrae)

- Generally an incidental finding.
- A detailed neurological examination should be performed initially in both adults and children.
- Plain x-rays of the cervical spine should have been performed initially to make the diagnosis.
- Advanced imaging is indicated if there are symptoms, or if multiple levels are involved.
 - Noncontrast cervical spine MRI (CPT 72141) is appropriate, and sometimes cervical spine CT (CPT 72125) may be of value.

PACSP- 4.6 Marfan syndrome

- Spine MRI (contrast as requested) is an appropriate study for Marfan syndrome if dural ectasias are suspected.

PACSP- 4.7 Neurofibromatosis type I

- see PACHD-12.2 Neurofibromatosis in the Pediatric Head guidelines

PACSP- 4.8 Platybasia

- Malformation of the skull base: the clivus is too horizontal.
- Symptoms are not frequent, but noncontrast brain MRI (CPT 70551) or head CT (CPT 70450) can be performed for further evaluation.

PACSP- 4.9 Spine in von Hippel-Lindau (H-L) syndrome

- H-L syndrome is associated with spinal hemangiomas and syrinx: level-appropriate MRI without and with contrast may be indicated.
- Many authorities perform MRI of the entire neural axis every other year in these patients. The value of this is currently unclear since the risk of bleeding from the hemangiomas of von H-L syndrome is low.
- Also see PACHD-15.10 von Hippel-Lindau Disease in the Pediatric Head guidelines
- von Hippel-Lindau syndrome does not usually become symptomatic until the early adult years.

PACSP-5~SPINAL DYSRAPHISM

- **PACSP- 5.1 Introduction:** Embryological abnormalities of the structures which create the spinal cord and vertebrae can be either trivial or catastrophic. Such anomalies can be either obvious or clinically occult.
 - Imaging of the most severe such lesions must be individualized to the case and is beyond the scope of this guideline.
- **PACSP- 5.2 Cutaneous lesions of the back:** the spinal cord arises from an infolding of the skin of the back, and so certain lesions of the overlying skin raise questions about an associated spinal deformity. Others are not associated with a meaningful risk of hidden spinal disorders.
 - **Not significantly associated with spinal dysraphism:**
 - Coccygeal pits and pilonidal cysts:
 - There is strong and extensive evidence that these lesions at or below the level of the intergluteal fold carry no increased risk of associated spinal abnormalities.
 - Advanced imaging is not generally indicated.*
**HK J Paediatr 2007;12(2):93-95*
 - Strawberry nevi are benign in significance and advanced imaging is not generally appropriate.
 - Non-specific darkened areas of skin over the sacrum:
 - There is no increased incidence of spinal anomalies in these children, and advanced imaging is not appropriate unless there are associated midline cutaneous abnormalities.
 - **Sometimes associated with spinal dysraphism:**
 - Dermal sinuses overlying the lumbar, thoracic, or cervical spine.
 - Initial spinal ultrasound is appropriate in neonates and infants.

- MRI of the relevant spinal level without and with contrast should be done if the ultrasound shows abnormalities other than a cutaneous dermal cleft.
 - MRI of the appropriate spinal level (contrast as requested) is appropriate initially in children or young adults (< age 30).
 - Sacral dermal sinuses which point cephalad (up) should be imaged as above.
- Subcutaneous midline masses at any level, caudal extensions, deviation of the gluteal fold, midline skin tags, abnormal patches of hair over the spine, and complex midline birthmarks above the upper sacral region:
 - MRI of the relevant spinal level (contrast as requested) is appropriate in children.
 - In infants, ultrasound is appropriate initially, but with masses, it is acceptable to proceed directly to MRI of the appropriate spinal level (contrast as requested).
- Congenital ano-rectal abnormalities are often associated with dysraphism.
 - Lumbar MRI without contrast (CPT 72148) is appropriate when these are present.
- Café au lait spots are a marker for type 1 neurofibromatosis.
 - See PACHD-12.2 Neurofibromatosis in the Pediatric Head guidelines
- **PACSP- 5.3 Spina bifida occulta** is seen in 20%-30% of the normal population, usually at L5 or S1 and is commonly an incidental finding on spinal x-rays.
 - Unless additional abnormalities are present, advanced imaging is not indicated.*
 *Kahn AN and Turnbull I. *Spinal Dysraphism/Myelomeningocele*. eMedicine, Feb. 20, 2007, <http://www.emedicine.com>. Accessed October 18, 2007
- Clinically significant dysraphism includes findings ranging from complex vertebral anomalies to meningocele.
 - MRI of the involved spinal level is appropriate, and in many cases, MRI of the entire spine will be indicated, often with sagittal and coronal reformatted images. The use of contrast is sometimes necessary.
- Spinal dysraphism does not usually include the brain.
 - Advanced imaging of the brain is not indicated in cases without hydrocephalus, affirmative signs of cerebral involvement, or the presence of multiple hydromyelia (which suggests hydrocephalus).
- Advanced imaging of the pelvis is not commonly necessary in these cases unless there are clinical signs of pelvic or rectal involvement.
- **References:**
 - *Dorsal Dermal Sinus*. <http://www.arnirsys.com>. Accessed November 20, 2006
 - *Pediatrics* 2001;108: e101
 - *Pediatrics* 2000;105:e69
 - *Pediatrics* 2003;112:641-647
 - *Pediatr Neurosurg* 2002;37:137-147
 - *Surgical Neurology* 2005;63(S1):S8-S12
 - *J Neurosurg* 2003;98(3 Suppl):247-250
 - *J Pediatr Orthopaedics* 2001;21:525-531

PACSP-6~TETHERED CORD

- **Tethered cord:**
 - The conus medullaris in newborns should terminate at L2-3 or higher. If the conus is below L2-3, it may be considered tethered.
 - The spinal cord normally ends in the conus medullaris, which is positioned at L1-2 in normal infants and children.
 - If the conus is as low as L2-3, the cord can be presumed to 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.
 - Noncontrast lumbar MRI (CPT 72148) is usually performed, but thoracic spine MRI, cervical spine MRI, and the use of contrast may be appropriate when a tethered cord has been found.
 - These cases should be sent for Medical Director review, especially if the patients have already had recent spine imaging.
- Also see PACSP-5 Spinal Dysraphism
- **References:**
 - Ropper AH and Brown RH. *Adams and Victor's Principles of Neurology*. 8th Ed. New York, McGraw-Hill, 2005, pp.860-861
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.285-290
 - *AJR* 1989;152:1029-1032
 - *Radiographics* 2000;20:923-938

**Evidence Based Clinical Support
PACSP-5~SPINAL DYSRAPHISM**

- **Pilonidal cysts/Sacral dimpling/Dorsal dermal sinus**
 - Skin indentations at the base of the spine, which in the case of pilonidal cysts and sacral dimpling are always benign and never communicate with the subarachnoid space.
 - Dorsal dermal sinus frequently extends into the subarachnoid space and is associated with intraspinal pathology including spinal abscess, tethered cord, and epidermoid tumor.
 - The important finding is the location of the dimple/sinus. If it is below the upper end of the intergluteal crease, the tract never communicates with the spinal canal.
 - In dimples/sinuses above the upper end of the intergluteal crease, spine MRI without contrast is indicated.
 - Ultrasound can be performed in infants under 12 months of age, but MRI should be done if ultrasound is positive.*
 - **Dorsal Dermal Sinus. <http://www.amirsys.com>.*
 - Accessed November 20, 2006
 - Other suggestive findings of dorsal dermal sinus are hair and capillary hemangioma around the opening.
 - MRI without contrast is also appropriate in any dimple with positive lower extremity neurological findings.

PEDIATRIC AND CONGENITAL SPINE IMAGING GUIDELINE REFERENCES

PACSP- 2~Pediatric Back Pain

- Combs JA and Caskey PM. Back pain in children and adolescents. *Southern Med J* 1997;90:789-792.
- Jones GT and Macfarlane GJ. Epidemiology of low back pain in children and adolescents. *Arch Dis Child* 2005;90:312-316.
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PACSP- 3~Kyphosis and Scoliosis

- Byrne LJ and McCormack MS. *The Orthopaedic management of Scheuermann's kyphosis*. <http://www.iol.ie/~rcsiorth/journal/volume5/issue1/sch.htm>. Accessed December 21, 2007.
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- Spiegel DA, Hosalkar HS, Dormans JP. *The Spine*. (Chapter 678). In Kliegman RM, Behrman RE, Jenson HB et al. (Eds.). *Nelson Textbook of Pediatrics*. 18th Ed. Philadelphia, Elsevier, 2007, pp.2811-2815.
- Do T, et. al. Clinical value of routine preoperative MRI in adolescent idiopathic scoliosis. *J Bone Joint Surg Am* 2001;83:577-579.
- Dobbs MB, et. al. Prevalence of neural axis abnormalities in patients with infantile idiopathic scoliosis. *J Bone Joint Surg Am* 2002;84:2230-2234.

PACSP- 4~Other Congenital and Pediatric Spine Disorders

- Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.306-307.

PACSP- 5~Spinal Dysraphism

- Lee ACW, Kwong NS, and Wong YC. Management of sacral dimples detected on routine newborn examination: A case series and review. *HK J Paediatr* 2007;12(2):93-95.
- Kahn AN and Turnbull I. *Spinal Dysraphism/Myelomeningocele*. eMedicine, Feb. 20, 2007, <http://www.emedicine.com>. Accessed October 18, 2007.
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- Santiago L, et.al. Newborns with suspected occult spinal dysraphism: A cost effectiveness analysis of diagnostic strategies. *Pediatrics* 2001;108: e101.
- Weprin BE and Oakes WJ. Coccygeal pits. *Pediatrics* 2000;105:e69.
- Ackerman LL and Menezes AH. Spinal congenital dermal sinus: A 30 year experience. *Pediatrics* 2003;112:641-647.
- Ackerman LL, et.al. Cervical and thoracic dermal sinus tracts. *Pediatr Neurosurg* 2002;37:137-147.
- De Brito Henriques JG, et.al. Minor skin lesions as markers of occult spinal dysraphisms—A prospective study. *Surgical Neurology* 2005;63(S1):S8-S12.

- Allen RM, et.al. Ultrasonographic screening in infants with isolated strawberry nevi. *J Neurosurg* 2003;98(3 Suppl):247-250.
- Suh SW, et.al. Evaluating congenital spine deformities for intraspinal anomalies with MRI. *J Pediatr Orthopaedics* 2001;21:525-531.

PACSP- 6~Tethered Cord

- Ropper AH and Brown RH. *Adams and Victor's Principles of Neurology*. 8th Ed. New York, McGraw-Hill, 2005, pp.860-861.
- Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.285-290.
- Wilson DA and Prince JA. MR imaging determination of the location of the normal conus medullaris throughout childhood. *AJR* 1989;152:1029-1032.
- Unsinn KM, Geley T, Freund MC, et.al. US of the spinal cord in newborns: Spectrum of normal findings, variants, congenital anomalies and acquired diseases. *Radiographics* 2000;20:923-938.

PACSP- 5~Spinal Dysraphism, Evidence Based Clinical Support

- *Dorsal Dermal Sinus*. <http://www.amirsys.com>. Accessed November 20, 2006.

PEDIATRIC AND CONGENITAL PERIPHERAL NERVE DISORDERS (PND) IMAGING GUIDELINES

PACPN-1~GENERAL GUIDELINES

- The Peripheral Nerve Disorders Imaging Guidelines are the same for both the pediatric population and the adult population, unless there are specific guidelines listed here in the Pediatric and Congenital Peripheral Nerve Disorders Imaging Guidelines.
- The peripheral nerves can be damaged by a multitude of causes, including trauma, infection, tumors, and metabolic disorders such as diabetes.
- The initial work up of a suspected peripheral nerve disorder should include a detailed neurological history and examination followed by electromyography and nerve conduction (EMG/NCV) studies.
- Advanced imaging plays a limited role in the diagnosis and management of disorders of peripheral nerves and muscles. The extent of that role is currently being defined.
 - **NOTE:** many disorders of these structures are associated with systemic diseases in which there are well-established indications for advanced imaging.
- When imaging of peripheral nervous tissue or muscles is indicated, MRI is used. In general, CT is not an acceptable alternative (occasional exceptions will be mentioned below).
- MRI is sometimes useful as a preoperative procedure since surgical decisions often depend on the presence or absence of **anatomic** integrity of the nerves (EMG evaluates functional integrity).
- **Reference:**
 - Bowen BC et. al. *Magnetic Resonance Imaging of the Peripheral Nervous System*. In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005, pp.1479-1497

PACPN-2~BRACHIAL PLEXUS

- Disorders of the brachial plexus can generally be identified and distinguished from lesions in other locations by clinical and electromyography and nerve conduction (EMG/NCV) examination. If the diagnosis remains unclear, advanced imaging can be helpful.
- Advanced imaging can be helpful as a preoperative study to evaluate the anatomy of brachial plexus lesions which should have already been defined by clinical examination.
- MRI is the preferred modality. CT is not often useful and should generally not be used as a substitute for MRI to image the brachial plexus.
 - Brachial plexus studies can be coded either as upper extremity other than joint MRI (CPT 73218) or as chest MRI (CPT 71550).
 - Occasionally, for upper trunk lesions, neck MRI (CPT 70540) may be requested.
 - Chest MRI will image both brachial plexi and is useful for comparing one plexus with the other.

- Rarely, more than one CPT code may be necessary to adequately image the brachial plexus area of interest.
- MRI studies should be without and with contrast (CPT 73220 or 71552) when tumor is part of the differential diagnosis.
- **References:**
 - *Radiographics* 2000;20:1023-1032
 - *ACR Appropriateness Criteria, Plexopathy Variant: Brachial, 2008*
 - *Eur Radiol* 2001;11:325-336
- **Trauma:** the cause and extent are generally obvious, but noncontrast MRI of the brachial plexus (CPT 73218 or 71550) is often useful, especially when surgical repair is being considered.
- **Birth trauma:** injury to the baby's upper (Erb's palsy) or lower (Klempke's palsy) plexus can occur during birth.
 - Noncontrast MRI of the brachial plexus (CPT 73218 or 71550) can be useful to define the defect.
 - If there is clinical suspicion for cervical nerve root avulsion, noncontrast cervical spine MRI (CPT 72141) may be useful.

PACPN-3~GAUCHER'S DISEASE

- Gaucher's disease is group of autosomal recessive inborn errors of metabolism characterized by lack of the enzyme acid 3-glucuronidase with destructive ceramide storage in various tissues.
 - Gaucher's disease is a treatable disorder (enzyme replacement) in which the liver, spleen, and bone marrow/bones are the most affected organs.
 - This guideline addresses Type I Gaucher's disease, which is by far the most common type in North America.
 - MRI is used to follow progression of disease in order to make treatment decisions, to monitor the results of treatment, and to evaluate complications as they occur.
 - Liver and spleen size are followed by annual noncontrast abdominal MRI (CPT 74181).
 - Annual noncontrast thigh MRI (CPT 73718) is used to follow marrow replacement by the disease and to monitor response to treatment.
 - MRI of a single thigh should be sufficient.
 - These patients often develop avascular necrosis of the hips and compression fractures of the spine, and relevant noncontrast MRI scans are appropriate when the clinical setting suggests these complications. In addition, many experts routinely perform MRI of the hips in untreated patients.
 - **References:**
 - *BJR* 2002;75 suppl 1:A13-A24
 - *Haematologica* 2000;85:792-799
 - McGovern M. *Gaucher Disease*. Updated October 15, 2003, <http://www.emedicine.com/PED/topic837.htm> Accessed November 22, 2006

PACPN-4~MUSCLE DISORDERS

- **Inflammatory muscle diseases**
 - Includes dermatomyositis, polymyositis, and sporadic inclusion body myositis.
 - Advanced imaging is used in these disorders for three purposes:
 - 1) Selection of biopsy site
 - 2) Treatment monitoring
 - 3) Detection of occult malignancy (for patients with dermatomyositis and polymyositis)
 - **Dermatomyositis in children:**
 - In children with dermatomyositis, MRI is often used to confirm a clinical diagnosis and thus avoid a biopsy.
 - An issue specific to this disorder is the presence of progressive calcification in muscles.
 - Noncontrast CT of the thighs (CPT 73700) is the procedure of choice to follow this condition, but MRI (CPT 73718) is often used since it permits assessment of the primary muscle disease as well.
 - **Search for occult neoplasm in patients with polymyositis:**
 - Lung and ovarian tumors are the most common, but lymphomas and other carcinomas can also be found.
 - Chest CT with contrast (CPT 71260) and pelvic ultrasound (in females) should be done initially.
 - CT abdomen and pelvis with contrast (CPT 74160 and 72193) are indicated if the above fail to make a diagnosis.
 - Tumors may remain occult for months to several years after the onset of the myositis.
 - **Reference:**
 - *Lancet* 2001;357:96-100

PEDIATRIC AND CONGENITAL PERIPHERAL NERVE DISORDERS IMAGING GUIDELINE REFERENCES

PACPN-1~General Guidelines

- Bowen BC et. al. *Magnetic Resonance Imaging of the Peripheral Nervous System*. In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005, pp.1479-1497.

PACPN- 2~Brachial Plexus

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- *ACR Appropriateness Criteria, Plexopathy Variant: Brachial*, 2008.
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PACPN- 3~Gaucher's Disease

- Maas M, Poll LW, Terk MR. Imaging and quantifying skeletal involvement in Gaucher disease. *BJR* 2002;75 (suppl 1):A13-A24.
- Giraldo P, Pocom M, Perez-Calvo JI, et al. Report of the Spanish Gaucher's Disease Registry: clinical and genetic characteristics. *Haematologica* 2000;85:792-799.
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<http://www.emedicine.com/PED/topic837.htm>. Accessed November 22, 2006.

PACPN- 4~Muscle Disorders

- Hill CL, Zhang Y, Sigurgeirsson B, et al. Frequency of specific cancer types in dermatomyositis and polymyositis: a population-based study. *Lancet* 2001 Jan;357:96-100.