

MedSolutions, Inc.
Clinical Decision Support Tool
Diagnostic Strategies

This tool addresses common symptoms and symptom complexes. Imaging requests for patients with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or patient's Primary Care Physician (PCP) may provide additional insight.

PEDIATRIC and CONGENITAL HEAD IMAGING GUIDELINES

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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 HEAD IMAGING GUIDELINES

PACHD-1~GENERAL GUIDELINES

- The Head 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 Head Imaging Guidelines.
- Advanced neuroimaging is only appropriate when there is either evidence of a cranial disorder or a clinically supported reason to search for cranial involvement in a systemic process.
- **PACHD-1.1 Congenital anomalies:** Children and adults with one congenital anomaly are likely to have others. Imaging requests should be considered in this light. Anomalies are too various to permit general rules.
- **PACHD-1.2 Overlapping studies:** If two studies using the same modality both cover the area of clinical interest, only one is generally needed. Certain exceptions are discussed as they arise.
 - **Maxillofacial versus orbital/temporal bone CT:**
 - There is overlap in anatomy between maxillofacial and orbital CT.
 - A temporal bone CT exam requires a separate protocol and should be considered a separate entity.
 - The exception is that the temporomandibular joint (TMJ) may be seen on both temporal bone and maxillofacial CT studies.
 - Unless there is a grounded suspicion of simultaneous involvement of more posterior lesions, especially of the region involving the middle or inner ear, only one of these studies should be approved in a single case.
 - Mild mucosal thickening in the paranasal sinuses or mastoids without other abnormalities is common in healthy individuals and not of itself an indication for advanced imaging.
 - Head MRI provides sufficient visualization of the paranasal sinuses to evaluate for the presence or absence of sinusitis.
 - See PACHD-25 Sinus, Child guideline.
- **PACHD-1.3 Screening**
 - In the following well-defined situations, certain advanced neuroimaging studies may be useful in screening. Otherwise, screening is not indicated.
 - Screening noncontrast head MRI (CPT 70551) can be performed in first degree relatives (parents, siblings, children) of patients with known familial cerebral cavernous malformations (cavernomas).
 - Screening for cerebral aneurysm – See HD-17.8 Screening for aneurysm in the adult Head guidelines.
 - Screening for von Hippel Lindau Disease—See HD-24.12 von Hippel Lindau Disease in the adult Head guidelines
 - Screening in sickle cell disease—See PACHD-16 Sickle Cell Disease in the Pediatric and Congenital Head guidelines
 - Screening based on a family history of a disorder is not appropriate if the disorder is not known to be familial.

- Screening of asymptomatic individuals using advanced imaging is inappropriate in most circumstances, especially those in which the presence of clinical features is required to make the diagnosis (e.g. multiple sclerosis).
- **PACHD-1.4 CT versus MRI:**
 - MRI is usually preferable to CT for brain imaging. However, in some situations, the difference in value between the two is small.
 - When the advantage of MRI is slight, CT is often used in infants and toddlers to avoid anesthesia/heavy sedation.
 - **CT is the initial procedure of choice for the following:**
 - Urgent/emergent settings due to availability and speed of CT
 - Trauma
 - Evaluation for recent hemorrhage, whether traumatic or spontaneous
 - Evaluation of the bony structures of the head
 - Evaluation and follow-up of hydrocephalus
 - In patients dependent on life support
 - CT is normally performed prior to lumbar puncture in patients with cranial complaints.
 - On occasion MRI may be substituted. The need for contrast in these MRI studies depends on the clinical setting.
 - The contrast used for the MRI depends on the clinical setting.
 - In some low yield imaging settings such as dementia in the elderly and headache in patients with normal neurological examinations, CT continues to be useful despite the theoretical superiority of MRI.
 - CT has little role in epilepsy, multiple sclerosis, pituitary disorders, characterization of known tumors, or the evaluation of the late effects of stroke or head trauma.
- **PACHD-1.5 Brain PET:** Should be coded as metabolic brain PET (CPT 78608).
- **PACHD-1.6 References:**
 - Grossman RI, Yousem DM. *Neuroradiology, the requisites*. 2nd Ed. Philadelphia, Mosby, 2003
 - Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005
 - Rowland LP (Ed.). *Merritt's Neurology*. 11th Ed. Philadelphia, Lippincott, 2005
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006

PACHD-2~CONTRAST USE IN HEAD IMAGING

- **PACHD-2.1 Contrast in CT**
 - Head CT is normally performed without contrast **except** in certain situations in which it is being used as a necessary alternative to MRI: e.g. evaluating tumor, abscess, or the pituitary gland in patients who cannot have MRI.
 - **In these guidelines, head CT is without contrast (CPT 70450) unless otherwise specified.**
 - **Sinus CT (CPT 70486) and temporal bone CT (CPT 70480)** are generally performed without using contrast. Exceptions are noted in the appropriate locations below.

- The iodide contrast used in CT reveals breakdown of the blood brain barrier and shows vasculature.
- Mass effect, blood or blood products, and abnormal tissue are shown on noncontrast CT.
- In patients who can have MRI, any abnormality on noncontrast CT is almost always better evaluated by MRI rather than CT with contrast.
 - MRI done in follow-up of an abnormal CT finding is usually done without and with contrast.
- Contrast only head CT (CPT 70460) has almost **no** indications.
 - Unless there has been a noncontrast CT done within a few days with abnormal results (but see the comments in the bullet point above), such requests are almost always made in error (i.e. the request for “CT with contrast” should be interpreted as without and with contrast [CPT 70470]).
- Neurologists, neurosurgeons, ENT specialists, and ophthalmologists should have the option of not using contrast when they believe it to be unnecessary.
- **PACHD-2.2 Contrast in brain MRI**
 - MRI is done without contrast to find masses, simple infarcts, anatomical abnormalities, and blood or blood products. Otherwise, contrast is often useful.
 - MRI contrast (Gadolinium) also shows breakdown of the blood brain barrier (including inflammation), displays blood supply to advantage in certain settings, reveals contrast patterns which make a number of lesions easier to characterize, and can visualize the meninges when this is needed. It often helps to characterize posterior fossa lesions and to characterize known masses.
 - Contrast only MRI (CPT 70552) is never ordered in the head except to follow-up a very recent noncontrast study (within one to two weeks at most). Otherwise, requests for brain MRI with contrast only are almost always made in error and should be coded as without and with contrast (CPT 70553).
 - Neurologists, neurosurgeons, ENT specialists, and ophthalmologists should have the option of not using contrast when they believe it to be unnecessary.
- **References:**
 - Grossman RI, Yousem DM. *Neuroradiology, the requisites*. 2nd Ed. Philadelphia, Mosby, 2003
 - Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005
 - Rowland LP (Ed.). *Merritt’s Neurology*. 11th Ed. Philadelphia, Lippincott, 2005
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006

PACHD-3~CT and MR ANGIOGRAPHY

- Except for stroke, arteriovenous malformation (AVM), sickle cell disease, pre-operative planning, and certain very rare disorders, these procedures are not often useful in childhood, and should be ordered only if there is both an appropriate presumptive diagnosis and an abnormal MRI, CT, or transcranial Doppler ultrasound. The exceptions are separately covered in the guidelines.
 - Review by a medical director is necessary for indications other than stroke, AVM, and sickle cell disease.

- **Reference:**
 - *Pediatr Neurol* 2000;23:307-311
- **CT and MR angiography (CTA and MRA):** These have been regarded as equivalents, but for most uses, CTA seems to be somewhat superior. For *many* purposes, but not all, CTA has replaced catheter angiography.
 - CT angiography of head or neck is often ordered to resolve uncertainties identified on MRA of those regions, and this is acceptable.
 - **Head MRA is these guidelines means without contrast (CPT 70544).**
 - Head MRA is generally done without contrast (CPT 70544). Some cerebrovascular experts prefer contrast MRA (CPT 70545) to evaluate certain strokes and AVM's and to follow known aneurysms, but for technical reasons, the addition of contrast usually has little to offer.
 - Requests for head MRA with contrast (CPT 70545) from neuro specialists are acceptable.
 - In patients with documented marked reduction in cardiac output, head MRA with contrast (CPT 70545) may be useful to improve image quality.
 - **There are no generally recognized indications for head MRA without and with contrast (CPT 70546).**
 - MRA of the neck vessels is usually done with contrast only (CPT 70548), and **“Cervical MRA” or “neck MRA” in these guidelines refers to contrast only MRA (CPT 70548) unless otherwise indicated.**
 - Some specialists use noncontrast MRA of the cervical vessels (CPT 70547) and this is acceptable when specifically requested.
 - A reasonable suspicion of carotid or vertebral dissection is the only clear indication for performing cervical MRA without and with contrast (CPT 70549).
 - **References:**
 - Grossman RI, Yousem DM. *Neuroradiology, the requisites*. 2nd Ed. Philadelphia, Mosby, 2003
 - Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005
 - Rowland LP (Ed.). *Merritt's Neurology*. 11th Ed. Philadelphia, Lippincott, 2005
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006

PACHD-4~ATAXIA

- **Detailed neurological history and recent clinical examination are indicated** prior to selection of neuroimaging in the evaluation of ataxia.
- Neurological consultation is helpful in determining the appropriate imaging pathway in these cases.
- **PACHD-4.1 Differential diagnosis of ataxia in children includes:**
 - tumor
 - ataxia telangiectasia
 - Friedreich's ataxia
 - juvenile lipofuscinoses
 - Refsum's disease
 - Abetalipoproteinemia

- Post viral ataxia of childhood
- Noncontrast brain MRI (CPT 70551) is most often the appropriate imaging study, but MRI without and with contrast (CPT 70553) is reasonable when tumor or multiple sclerosis is being considered.
- Neurological consultation is helpful in determining the appropriate imaging pathway.
- **Cervical spine imaging:** In both adults and children, noncontrast MRI of the cervical spine (CPT 72141) is appropriate when no etiology for the ataxia has been discovered after other evaluation.
- **Reference:**
 - *ACR Appropriateness Criteria, Ataxia, 2006*
- **PACHD-4.2 Hereditary ataxias and sporadic slowly progressive ataxias** are an indication for MRI of the brain (contrast as requested) in both adults and children. If brain MRI is nondiagnostic, noncontrast cervical spine MRI (CPT 72141) is appropriate.
 - DNA probes for many of these conditions exist, but they are not as reliable as that for Huntington's disease and their use does not change the need for imaging.
- **PACHD-4.3 Ataxia telangiectasia:**
 - The most common cause of ataxia after tumor in children under age 10. In essence, inherited via autosomal recessive mechanisms.
 - Cerebellar abnormalities appear by early childhood and progress slowly. Cutaneous and conjunctival telangiectasias appear later.
 - Immune (IgA) deficiencies are usual, and sinopulmonary infections are prominent.
 - Lymphomas and related disorders occur with 50-100 times the expected frequency.
 - *Formes frustes* of Ataxia telangiectasis exist, with milder features occurring later in life.
 - Brain MRI shows cerebellar atrophy. A variety of medical tests and genetic tests are relevant.
 - Sinus and body imaging may be necessary for complications.
 - These children are immunocompromised.
 - Brain MRI, including upper cervical spine is appropriate when the diagnosis is unclear. Contrast is appropriate only when tumor is under consideration.

PACHD-5~PEDIATRIC EPILEPSY/SEIZURE

- Detailed neurological examination and case history, including family history and the accounts of witnesses to seizures, should precede imaging.
- Neurological consultation is helpful in determining the need for advanced imaging in potential pediatric seizure patients.
- When imaging is indicated, brain MRI should be done if at all possible, rather than CT. Contrast should be added to the protocol if there is a progressing neurological deficit, if a noncontrast scan was abnormal, or at the discretion of an evaluating consultant.

- In contemporary American practice, noncontrast brain MRI (CPT 70551) will generally be performed in any child with documented new onset of non-febrile epileptic seizures.
- Repeat imaging for surveillance is not often indicated.
 - The specific indications for repeat imaging are those of adults (see HD-14 Adult Epilepsy/Seizure in the adult Head guidelines).
- **Imaging of children with typical febrile seizures is not medically necessary.** These occur in children from ages 6 months to age five during fevers of $>101^{\circ}\text{F}$ ($>38.4^{\circ}\text{C}$) and are brief generalized seizures. There is very often a family history of febrile seizures.
 - Neurological consultation may be helpful in confirming the diagnosis.
 - **Important:** not all seizures which occur in the presence of fever are febrile seizures.
 - **Reference:**
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.919-922
- **Evaluation for epilepsy surgery and intractable epilepsy:** As in adults, PET (CPT 78608) may be useful in the evaluation of intractable epilepsy in children.
 - In certain rare pediatric syndromes, use of PET tracers other than FDG is growing at academic pediatric epilepsy centers.
- **References:**
 - *Neurology* 2000;55:616-623 Reaffirmed 7-28-2006
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.857-942, esp. 887-890

PACHD-6~HEADACHE, CHILD

- Brain MRI without and with contrast (CPT 70553) is indicated in children with headache accompanied by an abnormal neurological examination, papilledema, or seizures.
- Advanced imaging without contrast is appropriate in children without neurological abnormalities if the headaches:
 - awaken the child from sleep
 - are associated with morning vomiting
 - are associated with diplopia.
 - Brain MRI **contrast as requested** is preferred to evaluate diplopia
 - become severe and progressive within a month of onset
- MRI (CPT 70551) is preferred to CT (CPT 70450) to avoid exposure to ionizing radiation in a low benefit situation. However, in some of these children it may be difficult to obtain the cooperation needed for MRI.
- **Thunderclap headaches in children without a family history of migraine:**
 - Noncontrast head CT (CPT 70450) is the procedure of choice within 12 hours of onset.
 - If more than 12 hours, noncontrast head CT (CPT 70540) or head MRI, contrast as requested, can be performed.
 - If bleeding is seen, brain MRI without and with contrast (CPT 70553) will usually

be appropriate, since in children, arteriovenous malformation (AVM), not aneurysm, is the most common non-traumatic source.

- Head CTA (CPT 70496) or brain MRA without contrast (CPT 70544) should be performed if the initial imaging study shows intracranial bleeding.
 - If AVM is identified, cervical CTA (CPT 70498) or MRA (CPT 70548) is also appropriate.
- Pediatric migraine, tension headaches, and school headaches are not an indication for advanced imaging.
- See HD 17.2 Hyperacute headache in the adult Head guidelines
- **References:**
 - *Neurology* 2002;59:490-498 Reaffirmed 10-15-2005
 - *Radiology* 1997;202:819-824
 - *ACR Appropriateness Criteria, Headache—Child, Revised 2008*

PACHD-7~SUBARACHNOID HEMORRHAGE

- **Children and adolescents:** Arteriovenous malformation (AVM), not aneurysm, is the leading cause of non-traumatic subarachnoid hemorrhage.
 - Even if CT has already been done (to establish the presence of subarachnoid blood), MRI (usually without and with contrast [CPT 70553]) is appropriate along with head and neck CTA (CPT 70496 and 70498) or MRA (CPT 70544 and 70548). Some centers use contrast enhanced MRA of the head for AVM, while others do not.
 - **Reference:**
 - Nelson PK, et. al. *Neuroradiology in the imaging and therapeutic management of brain arteriovenous malformations* (Ch 38). In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier, 2005

PACHD-8~HEAD TRAUMA

- Detailed medical history, recent general physical examination, and recent neurological examination are the initial phase in evaluating patients with potential neurological trauma.
- CT is the primary imaging modality in patients with acute head trauma.
 - MRI is used chiefly in severe acute head trauma when the clinical findings are not explained by the CT results (“the patient is much worse than the CT”) or to evaluate late effects of brain injury.
- When more than evaluation for potential neurosurgical lesions is needed, MRI is superior to CT in recognizing non-hemorrhagic cortical contusions, diffuse axonal injury (“shears”), and brain stem injury.
- **PACHD-8.1 Head CT is appropriate:**
 - After minor acute trauma in patients whose modified Canadian CT Head Rule inventory has any positive feature (see below)
 - Any head trauma patient who is:
 - taking one anticoagulant or two antiaggregants, (e.g. aspirin and Plavix)
 - has a known platelet or clotting disorder
 - has significant renal failure (creatinine>6)

- The modified Canadian CT Head Rule:
 - Positives include:**
 - Glasgow coma scale (GCS) score of <15 within 2 hours of injury
 - >30 minutes of amnesia
 - any “dangerous mechanism of injury”
 - a suspected open skull fracture
 - any signs of basilar skull fracture
 - two or more episodes of vomiting
 - patient >64 years old
 - There must be **no** positives to omit scanning
 - **References:**
 - [JAMA 2005;294:1551-1553](#)
 - [JAMA 2005;294:1511-1518](#)
 - [JAMA 2005;294:1519-1525](#)
 - [N Engl J Med 2000;343:100-105](#)
 - [Lancet 2001;357:1391-1396](#)
- In the six months following such injuries, whether or not there has been an initial scan, head CT or MRI without contrast (CPT 70450 or 70551) is appropriate if the patient develops dementia, alteration of alertness, or focal neurological deficits (e.g. hemiparesis, diplopia). This includes fluctuating problems.
- See SP-13 Mechanical Neck Pain, Trauma in the adult Spine guidelines for guidelines pertaining to cervical spine trauma.
- **PACHD-8.2 Brain MRI is not generally recommended as a first study, but noncontrast brain MRI (CPT 70551) is appropriate in:**
 - Patients (acute or chronic) who after head trauma have neurological features not explained by CT results.
 - As part of a neurological or Pain Management evaluation following non-acute head trauma with documented neurological or neuropsychological deficits.
 - Infants and children suspected of battered child syndrome (see below). These children have multiple non synchronous lesions which are best seen on MRI.
- **PACHD-8.3 Head MRA (CPT 70544) or CTA (CPT 70496) and brain MRI without and with contrast (CPT 70553) can be performed:**
 - If there is high suspicion for vascular injury.
 - To evaluate for post-traumatic aneurysm following penetrating trauma.
- **PACHD-8.4 Battered child syndrome:**
 - In children under age 2 with no neurological abnormalities in whom this situation is suspected, noncontrast brain MRI (CPT 70551) is recommended for evidentiary reasons to identify clinically silent non- synchronous lesions.
 - If there is a history of head trauma, noncontrast head MRI (CPT 70551) is preferred.
 - After age 5, history can be obtained and the situation more nearly resembles adult head trauma.
 - **Reference:**
 - [ACR Appropriateness Criteria, Suspected physical abuse--child, Updated 2005](#)
- **PACHD-8.5 Follow-up of known subdural or epidural hematomas** can be by either head CT or MRI (contrast as requested), and the preference of neurosurgeons and neurologists should be honored.

- There is no precise schedule for follow-up imaging studies. These patients are usually under the care of a neuro specialist.
- **PACHD-8.6 Patients with post-traumatic headache** persistent past the acute phase (a week or two) but without specific findings are best evaluated with noncontrast brain MRI (CPT 70551), but noncontrast head CT (CPT 70450) is acceptable.
- Patients with head trauma are often at risk for associated facial and cervical trauma.
 - Consult the relevant guidelines when such cases are under review.
- **PACHD-8.7 References:**
 - Zee CS, et. al. *Computed tomography and magnetic resonance imaging in traumatic brain injury.* (Ch 3). In Evans RW. *Neurology and Trauma.* 2nd Ed. Oxford, 2006
 - Gutierrez-Cadavid, JE. *Imaging of head trauma.* (Ch 43). In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System.* Philadelphia, Elsevier, 2005

PACHD-9~DYSTONIA

- **Dystonia:** In adults and children with dystonia, brain MRI (contrast as requested) is indicated if there are other neurological features beside the dystonia itself.*
 - **PET:** At this time, there is no firmly established basis for the use of PET in the evaluation or management of dystonia or other movement disorders.

PACHD-10~SUSPECTED MULTIPLE SCLEROSIS (MS)

- **PACHD-10.1 Introduction**
 - MS is notoriously variable in its presentation and course, but there are some useful generalizations.
 - The most common presentation is relapsing: the occurrence of multiple episodes of focal neurological deficit each of which at least partially resolves.
 - Over time, this tends to evolve into a course of either steady progression of deficits (chronic progressive) or of relapses without improvement (progressive relapsing).
 - MS is correctly thought of as a disease of young adults, particularly young women. However, it can present in childhood or in middle age.
 - When it presents in mid-life, a progressive form affecting the spinal cord in particular is not unusual.
 - Symptoms from MS most often arise from involvement of the optic nerves, the brain stem, the cerebellum, and the spinal cord.
- **PACHD-10.2 Diagnosis:**
 - MS is diagnosed by correlation between clinical, laboratory, and imaging data. The medical and social consequences of overdiagnosis of MS in murky clinical situations can be dire.
 - Extremely detailed history and recent neurological examination are indicated before selection of imaging studies.
 - Specialist consultation (neurology, neurosurgery, or, for visual syndromes, ophthalmology) is helpful in determining the appropriate imaging pathway and the significance of what are often difficult-to-interpret findings on imaging studies.

- MS most commonly presents with apparently single episodes of demyelination involving specific areas of the nervous system.
 - However, many patients who experience such a single episode do not go on to develop MS.
- The criterion for a firm diagnosis of MS is the presence of lesions dispersed in time and space (space=different locations in the nervous system).
 - Since treatments which somewhat affect the course of the disease have become available, the use of MRI to anticipate dispersion in either space or time has become widespread. This allows for earlier treatment.
 - Various MRI diagnostic criteria for this purpose, which include findings on both brain and spinal cord imaging, are discussed in HD-22 Evidence Based Clinical Support section in the adult Head guidelines.
- **General remarks on advanced imaging in MS:**
 - CT, CTA, MRA are **not** useful in the evaluation of either new onset or established MS unless there is documentation of a grounded concern regarding a concurrent and unrelated diagnosis for which any of these studies would be of value.
 - Orbital MRI is not generally indicated, except for atypical cases of optic neuritis.
 - At this time, the value of newer imaging techniques such as diffusion tensor imaging and magnetic resonance spectroscopy in patients with multiple sclerosis remains to be established.
 - Newer MRI diagnostic criteria lay greater stress on the results of spinal cord imaging, and inclusion of the spinal cord in the initial imaging battery is appropriate for most situations other than clinically pure optic neuritis.
 - **Spinal cord imaging in MS:**
 - Cervical and thoracic spine MRI scans visualize the entire spinal cord, and lumbar spine MRI is not needed.
 - Screening spinal MRI consisting only of sagittal views of the entire spinal cord using a phased array detector coil may occasionally be requested and is appropriate. Screening spinal MRI should be coded as one spine segment (CPT 72141 or 72146)
- **PACHD-10.3 Isolated clinical syndromes:**
 - **Optic neuritis:** MRI brain without and with contrast (CPT 70553) is indicated initially for patients with optic neuritis
 - MRI of the spinal cord (cervical spine with or without imaging the thoracic spine), contrast as requested, can be approved if brain MRI is suggestive of MS but not firmly diagnostic.
 - **Other cerebral isolated clinical syndromes:** MRI of the brain without and with contrast (CPT 70553) should be performed initially.
 - MRI of the spinal cord (cervical spine with or without imaging the thoracic spine), contrast as requested, and can be approved if brain MRI is suggestive of MS but not firmly diagnostic.
 - In certain patients, neurological findings are such that the likelihood of a normal brain MRI is very low, and in such cases, on specialist request, spinal cord imaging may be done simultaneously with head imaging. The same is

- true if the clinical setting indicates a high probability of abnormalities on spinal cord imaging.
- **Transverse myelitis:**
 - Another “isolated clinical syndrome”
 - Spinal cord imaging (cervical and thoracic spine MRI [contrast as requested]) is appropriate initially.
 - If the clinical presentation is typical of a demyelinating process, it is acceptable to include brain MRI without and with contrast (CPT 70553) in the initial imaging battery.
 - If spinal imaging does not show a non-inflammatory origin (spinal tumor or compression), brain MRI without and with contrast (CPT 70553) is also appropriate to rule out Multiple sclerosis if that has not already been done.
 - **References:**
 - [Neurology 2003;61:602-611](#)
 - [Ann Neurol 2005;58:840-846](#)
 - [Ann Neurol 2001;50:121-127](#)
 - [Lancet Neurology 2007;6:677-686](#)
 - [AJNR 2006;27:455-461](#)
 - [Eur J Neuroradiol 2006;13:313-325](#)
 - **PACHD-10.4 Migratory Paresthesias:**
 - Patients with normal examinations who have either attacks of wandering paresthesias **lasting at least a full day** or a history of a recovered isolated clinical syndrome may be approved for imaging using the guidelines in PACHD-10.3 Isolated clinical syndromes.
 - Also see HD-26 Paresthesia in the adult Head guidelines
 - **PACHD-10.5 Repeat of initial negative studies:**
 - In the settings covered by PACHD-10.3 and 10.4, if the initial imaging studies are diagnostic, repeat studies are not indicated.
 - If the initial scans are not diagnostic, repeat studies at 3 months, and, if again negative, at one year can be approved.
 - Some centers prefer a repeat at 6 months in patients not started on treatment after a single isolated clinical episode, and this is acceptable.
 - Under the unusual circumstances detailed in the HD-22 Suspected MS Evidence Based Clinical Support section in the adult Head guidelines, repeat studies at one month may be appropriate.
 - These requests should be sent for Medical Director review.
 - **PACHD-10.6 Familial MS and screening**
 - The lifetime risk of MS in first degree relatives of MS patients is about 4% (higher for female relatives).
 - Identical twins have a 35% concordance rate for MS.
 - Offspring of two MS patients have a 30% concordance rate for MS.
 - Regardless of family history, screening based on family history in the absence of a clinical indication is not appropriate since the diagnosis cannot be made without a clinical component.
 - **Reference:**
 - [Lancet Neurology 2004;3:104-110](#)

- **Also see PACHD-20 Optic Neuritis**
- **PACHD-10.7 Neuromyelitis optica (DeVic's disease):**
 - A demyelinating syndrome characterized by involvement of optic nerves and spinal cord without symptomatic cranial lesions.
 - Most patients have normal brain MRI but some have hypothalamic lesions or non-specific features.
 - While spinal cord lesions of MS involve two or fewer segments, those in Neuromyelitis optica involve three or more.
 - Recently, a specific putative serum immune marker for this disease has been discovered.
 - Initial imaging includes brain and spine MRI.
 - Any needed follow-up can be limited to spine MRI in typical cases.
 - **Reference:**
 - [*Neurology* 2006;66:1485-1489](#)

PACHD-11~ESTABLISHED MULTIPLE SCLEROSIS (MS)

- Detailed interval history and recent neurological examination are the first steps in any re-evaluation of patients with MS.
- **PACHD-11.1 Baseline imaging of the brain or brain and spinal cord** (contrast as requested) before starting immunomodulating treatment of MS is appropriate.
 - Use of the agent natalizumab (Tysabri) requires baseline brain MRI without and with contrast (CPT 70553).
 - Repeat brain MRI without and with contrast (CPT 70553) is appropriate if symptoms consistent with PML occur while on Tysabri (PML = progressive multifocal leukoencephalopathy). Symptoms can include a rapidly progressive subacute dementia or a series of apparent strokes.
 - For all patients taking Tysabri, clinical evaluation at 3 months of treatment and then semiannually is required.
 - Head MRI without and with contrast (CPT 70553) is acceptable at any of these re-evaluations if the treating physician requests it.
- **PACHD-11.2 Repeat imaging in established MS (MRI contrast as requested) is appropriate:**
 - If there is a new spinal episode (imaging should be limited to the spinal cord).
 - If the patient is being evaluated for the use of immunomodulating therapy. (see PACHD-11.1 Baseline imaging of the brain or brain and spinal cord)
 - glatiramer = Copaxone
 - natalizumab = Tysabri
 - mitoxantrone = Novantrone This agent may cause cardiotoxicity and MUGA scans may be useful (see CD-3.7 MUGA study in the adult Cardiac guidelines)
 - Beta-interferons = Avonex, Betaseron, and Rebif are the “standard” ones at present.
 - If the patient develops what seems to be a new and unrelated disorder, imaging appropriate to the potential new disorder should be performed.

- **PACHD-11.3 Annual surveillance scans of established MS patients** require that the patient be on immunomodulating therapy or be a candidate for such therapy.
 - Imaging can include:
 - Brain MRI (contrast as requested)
 - Cervical and thoracic spine MRI (contrast as requested) if spinal cord findings are likely.
 - The value of surveillance scanning in established MS is uncertain at this time.
 - In the progressive spinal form of MS, if prior brain imaging has been negative, spinal MRI (contrast as requested) rather than brain MRI may be sufficient for surveillance.
- **PACHD-11.4 Other Issues**
 - Specialist evaluation (neurology, neurosurgery, or, for visual syndromes, ophthalmology) is helpful in determining the need for advanced imaging in established MS.
 - In patients with severe spinal cord disorders, including MS, clinical evaluation of abdominal disorders may be very difficult because impaired cord function affects expected signs and symptoms. Requests for abdominal and pelvic imaging studies should be evaluated in this light.
 - MS patients on immune therapy of any sort must be regarded as immunocompromised, and this may be relevant to extra-neurologic imaging requests.
 - The practical difficulty of arranging imaging sessions in patients who are litter- or wheelchair-bound should be weighed carefully before recommending a serial approach to imaging in such patients.
- **PACHD-11.5 References**
 - [AJNR 2006;27:455-461](#)
 - [Eur J Neuroradiol 2006;13:313-325](#)

PACHD-12~NEURO-ONCOLOGY-BRAIN TUMORS

- **PACHD-12.1 General remarks**
 - Brain MRI without and with contrast (CPT 70553) is indicated for both characterization and follow-up of brain tumors. However, occasionally neurologist, neurosurgeons, and oncologists treating such patients will find appropriate use for CT or noncontrast MRI. Their preferences should be honored in such cases.
 - In general, MRA/CTA scans are not necessary for the diagnosis or management of brain tumors without a clear documented indication.
 - Postoperative brain MRI is standard, usually 24 to 72 hours following brain tumor surgery.
 - Repeat imaging is appropriate when patients deteriorate or develop new features.
 - The management of pediatric brain tumors presents issues not readily addressed by general guidelines.
 - Reasonable imaging protocols in use by dedicated pediatric neuro-oncology centers should be considered acceptable.

- MRI of the entire neural axis without and with contrast includes CPT 70553, 72156, 72157, and 72158.
- **PACHD-12.2 Neurofibromatosis**
 - **Neurofibromatosis, type 1 (von Recklinghausen's Disease, NF 1)**
 - Autosomal dominant. Incidence 1 per 5000. Only half have family history.
 - Imaging to screen children without symptoms is not generally appropriate.
 - Subcutaneous neurofibromas and multiple café au lait spots are typical.
 - Kyphoscoliosis is common and may cause cord compression. Spinal dural ectasias and meningoceles occur.
 - Intraspinial tumors are frequent.
 - Screening those without symptoms or signs is usually not useful, since most occult neurofibromas do not grow aggressively.
 - Optic nerve and brain stem gliomas are common (brain MRI without and with contrast [CPT 70553] and also orbits [CPT 70543] for those with optic nerve lesions).
 - These tumors require monitoring when present, but do not behave as malignantly as their names suggest.
 - Growth can be heralded by precocious puberty.
 - Headache is common, and because of elevated tumor risk and a high incidence of aqueductal stenosis (hydrocephalus), prompt brain MRI without and with contrast (CPT 70553) is appropriate.
 - Neurofibromatosis I is a known cause of strokes and of Moya moya disease.
 - Imaging should follow guidelines appropriate for pediatric stroke (see PACHD-17 Pediatric Stroke).
 - Imaging to screen family members without signs of the disease is generally inappropriate since the clinical picture is readily recognized.
 - **Neurofibromatosis, type II** is a separate and extremely rare disease characterized by either bilateral acoustic neuromas or a combination of familial acoustic neuroma and another brain tumor. The tumors determine the imaging. It is mentioned only to avoid confusion with NF 1.
 - **Reference:**
 - Maria BD, Menkes JH. Neurocutaneous syndromes. In Menkes JH, Sarnat HB, Maria BL. Child Neurology. 7th Ed. Philadelphia, Lippincott, 2006, pp.803-824
- **PACHD-12.3 Grade I-II astrocytoma and benign oligodendroglioma (low grade)**
 - After initial biopsy or other treatment, repeat MRI brain without and with contrast (CPT 70553) is appropriate.
 - Surveillance for posterior fossa tumors in this class is by brain MRI without and with contrast (CPT 70553) repeated every 3 to 6 months for 5 years and then annually.
 - Supratentorial (cerebral proper) tumors should be re-imaged at approximately 3 months, 6 months, and then annually.
 - Frequent imaging is used in the follow-up of low grade astrocytomas of childhood, with imaging in the range of every 3-4 months in the first two years, every 6 months in the third year, and every 6 to 12 months thereafter. Further imaging may be appropriate upon specific indication.
 - Magnetic Resonance Spectroscopy (MRS) can be useful in following the course

of pediatric low-grade astrocytomas, especially in regions such as the cerebellum, brain stem, and diencephalon.

- Certain payers consider the use of MRS to be investigational, and their coverage policies will take precedence over MedSolutions' guidelines. Prior authorization does not guarantee payment of the study.
- **PACHD-12.4 Glioblastoma and other malignant glial tumors (including grade III astrocytoma).**
 - Following surgery and radiation therapy (XRT) with or without adjuvant chemotherapy, brain MRI without and with contrast (CPT 70553) is usually performed 2 to 6 weeks following completion of treatment, and then every 2 to 3 months.
 - During chemotherapy treatments or a course of XRT, MRI brain without and with contrast (CPT 70553) every 8 to 10 weeks is usual.
 - PET: see PACHD-12.8 PET in brain tumor
- **PACHD-12.5 Ependymoma**
 - These tumors usually occur below the tentorium in children and above in adults.
 - The more malignant ones can seed the entire neural axis.
 - Postoperatively, MRI of the entire neural axis is appropriate (brain and entire spine without and with contrast).
 - Surveillance scanning should be every 3 to 4 months the first year, every 6 months the next year, and then every 6 to 12 months depending on the malignancy of the tumor.
 - For malignant ependymoma, entire neural axis scans are appropriate, but for benign ependymomas, imaging limited to the level of the tumor is appropriate.
 - While the child remains under active treatment with radiation and/or chemotherapy, bimonthly imaging is acceptable.
 - Magnetic Resonance Spectroscopy MRS may be useful to evaluate response to therapy.
 - Certain payers consider the use of MRS to be investigational, and their coverage policies will take precedence over MedSolutions' guidelines. Prior authorization does not guarantee payment of the study.
- **PACHD-12.6 Medulloblastoma**
 - This tumor of childhood (also referred to as **PNET**--primitive neuro-ectodermal tumor) also seeds the entire neural axis, and imaging guidelines are the similar to those for malignant Ependymoma (see PACHD-12.5 Ependymoma).
 - As long as the child remains under treatment with chemotherapy and/or radiation, imaging at bimonthly intervals is acceptable.
- **PACHD-12.7 Meningiomas** are tumors of the dura and are usually benign.
 - Initial imaging should be a brain MRI without and with contrast (CPT 70553).
 - In selected cases, noncontrast head CT (CPT 70450) may also be required to evaluate bony involvement.
 - As a preoperative study, CTA/MRA of head (CPT 70496/70544) and/or neck (CPT 70498/70548) may be appropriate.
 - Meningiomas that are being observed (no resection):
 - Imaging can be performed if new signs/symptoms/neurological findings related to the meningioma occur.

- In asymptomatic patients, imaging can be performed once a year for ten years and then every five years
- Following documented complete resection, repeat imaging at 6 months, 2 years, and 5 years is sufficient.
 - For skull base meningiomas or any meningioma subtotally resected, follow-up imaging every 3 to 6 months for 2 years and then annually for life is recommended.
- Malignant meningiomas (by pathology): re-image at 3 and 6 months post resection and then annually for life.
- Meningiomas in children are unusual, but very aggressive, and more frequent MRI imaging is appropriate, especially during the two years following diagnosis.
- **PACHD-12.8 PET in brain tumor (metabolic brain PET – CPT 78608)**
 - Certain payers consider the use of brain PET in tumors to be investigational, and their coverage policies will take precedence over MedSolutions’ guidelines. Prior authorization does not guarantee payment of the study in that situation.
 - The chief use of PET in the management of brain tumor is to aid in distinguishing recurrent tumor from radiation cerebritis in patients with known anaplastic tumors of glial origin (glioblastoma, anaplastic astrocytoma, and anaplastic oligodendroglioma) and prior XRT.
 - Candidates for PET will have had a very recent brain MRI showing enhancing new lesions compatible with either recurrent tumor or radiation necrosis.
 - On rare occasion, brain PET (or Magnetic Resonance Spectroscopy) may be useful in resolving a differential diagnostic issue in a patient when MRI and clinical course cannot distinguish tumor from “tumefactive” MS plaque, non-acute inflammation, or infarct. Such cases require review by a Medical Director.
 - Magnetic Resonance Spectroscopy (MRS) is sometimes used to distinguish recurrent tumor from radiation cerebritis, and this is an acceptable alternative to PET
 - Certain payers consider MRS investigational, and their coverage policies will take precedence over MedSolutions’ guidelines. Prior authorization does not guarantee payment of the study.
 - Brain, especially gray matter, takes up FDG avidly, and only very metabolically active tumors are more “PET avid” than this.
 - FDG-PET is therefore generally not useful in the evaluation of most metastatic deposits and well-differentiated brain tumors.
 - **Reference:**
 - *Central Nervous System Cancers. NCCN Practice Guidelines in Oncology v.1.2008*
- **PACHD-12.9 Evaluation of suspected retinoblastoma (including leukokoria):**
 - Orbital CT without and with contrast (CPT 70482) is preferred initially.
 - Once a retinoblastoma has been confirmed, MRI scans without and with contrast of brain (CPT 70553) and orbits (CPT 70543) are indicated.

PACHD-13~PAPILLEDEMA–PSEUDOTUMOR CEREBRI

- Pseudotumor cerebri is also called benign intracranial hypertension.
- The first step in evaluation is a detailed history and recent neurological examination.
- Papilledema indicates the presence of elevated intracranial pressure.
 - Brain MRI without and with contrast (CPT 70553) is indicated.
 - The vast majority of alert neurologically normal patients will have idiopathic intracranial hypertension (pseudotumor cerebri) and normal imaging studies.
 - Brain MRI is performed in these cases to exclude cerebral mass lesions, obstructive hydrocephalus, and occult meningeal disease.
 - Patients with papilledema will generally require lumbar puncture, but for reasons of patient safety, lumbar puncture is done after the initial brain imaging study.
- MRV/CTV (CPT 70544/70496) is appropriate to exclude venous sinus thrombosis in atypical cases of pseudotumor.
 - Typically, pseudotumor occurs in overweight women of childbearing years and responds to medical treatment.
 - Atypical cases include:
 - male patients.
 - slender patients
 - women > age 45
 - children (< age 16) unless there is an apparent cause
 - patients with known intrinsic system clotting disorders
 - patients who fail to respond to pharmacologic treatment
- MRA/CTA is not generally appropriate in these cases without a specific indication. Since MRA/CTA share the same CPT codes with MRV/CTV, it will be necessary to know which study is being requested.
- Orbital MRI or CT is not indicated initially unless there is a documented concern for orbital pseudotumor or other primary bilateral orbital disorder
- Ophthalmology or Neurology consultation may be helpful to:
 - distinguish papilledema from papillitis
 - distinguish pseudopapilledema from genuine papilledema
 - establish the presence of mild papilledema
- Re-imaging is infrequently indicated unless done to evaluate possible shunt dysfunction in those patients who have had ventriculoperitoneal (VP) or lumboperitoneal (LP) shunts or because of distinct clinical deterioration.
- Also see HD-16.6 Chronic intractable headaches in the adult Head guidelines
- **Reference:**
 - *Headache Currents* 2005;2:1-10

PACHD-14~HYDROCEPHALUS

- **Hydrocephalus:**
 - Head ultrasound to screen for hydrocephalus is appropriate in infants less than six months of age (important for radiation exposure considerations).
 - Head CT (CPT 70450) or MRI (CPT 70551) without contrast is appropriate to screen for hydrocephalus in children with accelerated head growth.

- A positive study will usually require follow-up with brain MRI without and with contrast (CPT 70553).
- Head CT without contrast (CPT 70450) is appropriate to check shunt integrity in patients with ventricular shunts in whom shunt obstruction is suspected (children and adults).
 - Occasionally, there may be a role for abdominal CT to evaluate distal ventriculoperitoneal (VP) shunt problems.
- In some patients with obstructive hydrocephalus, CSF flow studies may be helpful in making treatment decisions (see HD-35.5 CSF flow imaging in the adult Head guidelines). These studies are generally part of a preoperative evaluation of the patient, and therefore, evaluation by a neurological or neurosurgical specialist is appropriate.

PACHD-15~OTHER PEDIATRIC HEAD GUIDELINES (NOT ELSEWHERE COVERED)

- **PACHD-15.1 Behavioral disorders**
 - Behavioral disorders of childhood or adolescence generally require no imaging.
 - **Exception:**
 - Requests from neurologists and psychiatrists for MRI (contrast as requested) in children who show features of major psychoses and intellectual decline, and who have not responded to treatment, are often appropriate, since schizophrenia is uncommon before mid-adolescence.
- **PACHD-15.2 Cerebral palsy**
 - A non-progressive motor impairment dating from infancy and usually antenatal in origin.
 - MRI or CT can identify a treatable problem in about 5% of those cases in which the cause was not determined in the newborn period (usually by ultrasound). In addition, it can prove the timing of the insult in most of the remaining cases.
 - MRI is superior, but generally requires sedation or anesthesia.
 - Brain MRI without and with contrast (CPT 70553) or head CT (CPT 70470) is appropriate for cases of cerebral palsy of undetermined origin or if a fixed deficit worsens.
 - **Reference:**
 - *Neurology* 2004; 62:851-863
- **PACHD-15.3 Craniostenosis**
 - Noncontrast head CT (CPT 70450) is indicated in the diagnosis of craniostenosis (craniosynostosis), and 3-D rendering (CPT 76376 or 76377) may be needed for surgical planning.
- **PACHD-15.4 Developmental delay/mental retardation**
 - Brain MRI without contrast (CPT 70551) is appropriate to evaluate for congenital abnormalities whether cerebral palsy is noted or not. If necessary, noncontrast head CT (CPT 70450) may be substituted.
 - **Reference:**
 - *Neurology* 2003;60:367-380. Reaffirmed 10-15-2005

- **PACHD-15.5 Macrocephaly**
 - Defined as head circumference over two standard deviations above the mean for age and sex (as measured by standard growth charts).
 - Accelerated head growth of more than a standard deviation from prior measurements is also significant.
 - MRI scans should be approved contrast as requested to minimize the need for re-imaging.
 - **Age less than 6 months:** ultrasound is preferred initially. If ultrasound is abnormal (or unsuccessful), head CT to evaluate for hydrocephalus and calcifications is preferred, but MRI will also often be indicated after CT images are reviewed and, at times, will be done first.
 - **Age 6 months to 2 years:** MRI is recommended initially, since in this age group, uncomplicated hydrocephalus is less likely than in early infancy. CT is sometimes appropriate because of difficulty obtaining MRI in patients of this age.
 - **Over two years of age:** MRI is recommended initially.
- **PACHD-15.6 Megalencephaly**
 - This term refers to excessive size of brain constituents rather than just head size. MRI is appropriate when further imaging is required (the use of the term suggests there has been prior imaging).
 - **Reference:**
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.327-329
- **PACHD-15.7 Microcephaly**
 - The definition is similar to that for macrocephaly in reverse. MRI is recommended initially since CT may not detect the relevant anatomical abnormalities.
 - **Reference:**
 - Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.322-325
- **PACHD-15.8 Sturge-Weber syndrome**
 - Port-wine nevus of the upper face (forehead or upper eyelid typical) combined with a meningeal vascular anomaly, usually with developmental delay, premature glaucoma, seizures, and stroke-like events. Only 5% of patients with the nevus have the cerebral disorder.
 - This condition is not familial and familial screening is not indicated.
 - **Imaging:** Brain MRI without and with contrast (CPT 70553). Head CT without and with contrast (CPT 70470) can be useful at times.
 - Cortical resections can be used to treat intractable seizures, and brain PET (CPT 78606) can be useful for preoperative mapping.
 - **Reference:**
 - Maria BD, Menkes JH. *Neurocutaneous syndromes*. In Menkes JH, Sarnat HB, Maria BL. *Child Neurology*. 7th Ed. Philadelphia, Lippincott, 2006, pp.803-824
- **PACHD-15.9 Tuberos scleriosis**
 - Transmitted as an autosomal dominant with prevalence of ~1 per 7500.
 - Nervous system manifestations include cortical tumors, gliotic areas which resemble astrocytomas in appearance but not in behavior (tubers), and subependymal nodules. Seizures and mental retardation are typical.
 - Calcification of tubers is seen in about half of the cases.

- A peculiar tumor (giant cell astrocytoma) occurs in about 15% of patients.
- Multiple small angiofibromas of the face usually enable visual recognition of the syndrome.
- Extra-cranial neoplasms are also common, esp. cardiac rhabdomyomas, renal cysts, and benign tumors.
- **Imaging:** Brain MRI without and with contrast (CPT 70553) is sufficient for confirmation of the diagnosis and to follow giant cell astrocytomas.
- Cardiac echocardiogram and abdominal CT without and with contrast (CPT 74170) are appropriate when there is concern for tumors in those regions.
- Female patients often develop lymphangiomatosis of the lungs, and high resolution chest CT (CPT 71250) for screening is appropriate in adult female patients with tuberous sclerosis.
- **Familial screening:** Careful clinical evaluation without imaging is generally sufficient.
- **Reference:**
 - [N Engl J Med 2006;355:1345-1356](#)
- **PACHD-15.10 von Hippel Lindau Disease**
 - Autosomal dominant disorder
 - Principal features are retinal angiomas and hemangioblastoma of the cerebellum.
 - Pheochromocytomas (10%) and renal carcinoma are also relatively frequent.
 - The hemangioblastomas are benign cystic tumors and may be associated with secondary polycythemia.
 - Hemangiomas in other regions and benign renal and hepatic cysts occur.
 - DNA testing can identify family members **not** at risk.
 - No screening imaging is needed for those members.
 - For those at risk, abdominal screening by ultrasound should be done during the teenage years.
 - If the ultrasound is abnormal, CT of the abdomen with contrast (CPT 74160) can be performed.
 - MRI of the brain and spine without and with contrast are recommended annually during the teenage years and then every two years.
 - Temporal bone CT (CPT 70482) or MRI (CPT 70543) to rule out tumors of the endolymphatic sac is appropriate if hearing loss is present.
 - **References:**
 - [von Hippel Lindau Alliance. Screening recommendation. <http://www.vhl.org/handbook/vhlhb4/htm>. Accessed November 29, 2006](#)
 - [von Hippel Lindau syndrome, *Online Mendelian Inheritance in Man*. <http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=OMIM&dopt=Detail>. Accessed November 29, 2006.](#)
- **PACHD-15.11 Tourette's syndrome**
 - The diagnosis of Tourette's syndrome is made clinically and advanced neuroimaging is generally not of value for either diagnosis or management.
 - If the presentation is atypical and there is an unresolved differential diagnostic issue, MRI brain, generally done without contrast (CPT 70551) may be helpful. CT is not often helpful and MRA/CTA is generally not indicated unless justified by specific MRI findings.
 - Repeat or surveillance imaging for this disorder is not generally necessary.

PACHD-16~SICKLE CELL DISEASE

- Stroke is common and is an indication for active treatment. Half of these strokes are asymptomatic.
- Many centers follow the cerebral circulation of children with sickle cell disease (SS) with transcranial Doppler. Positive findings are further evaluated with brain MRI/MRA.
 - Transcranial Doppler is not reliable in those over age 20.
- At the preference of the treating physicians, brain MRI without and with contrast (CPT 70553) and noncontrast brain MRA (CPT 70544) can be used to follow these children even when neurologically normal.
 - Annual repeat studies are acceptable.
- A Moya-moya disease effect may arise. Noncontrast brain MRA (CPT 70544) is indicated, since CTA is unsafe (iodide based contrast is contraindicated in these patients).
- Sickle cell carriers (SA) are not at risk for these complications and do not require imaging.
- Many patients with S-Thalassemia and some patients with S-C disease require the same management as SS patients.

PACHD-17~PEDIATRIC STROKE

- **PACHD-17.1 General Considerations**
 - The differential diagnosis is similar to that discussed for young adults (see HD-31.5 Premature stroke in the adult Head guidelines), although large vessel atherosclerosis is less likely.
 - The differential diagnosis includes cranio-cervical dissections, fibromuscular dysplasia, arteritis, venous infarction, cardioembolic stroke, MELAS, Moya moya disease, congenital heart disease, and arteriopathy related to a variety of congenital malformation syndromes, etc.
 - Specialty consultation is strongly supported.
 - Brain MRI without and with contrast (CPT 70553) is appropriate even if an initial head CT to exclude hemorrhage was performed.
 - Brain and neck MRA, or CTA will generally be indicated as well.
 - Neck MRA should be without and with contrast (CPT 70549) when dissection is suspected.
 - Sickle cell anemia is a major cause of strokes in children (see PACHD-16 Sickle Cell Disease).
 - **References:**
 - *J Child Neurol* 2005;20:194-197
 - *Ann Neurol* 2003;53:167-173
 - Rowland LP (Ed.). *Merritt's Neurology*. 11th Ed. Philadelphia, Lippincott, 2005. pp.309-310
- **PACHD-17.2 Venous infarcts**
 - These are a small percentage of strokes (incidence ~3 per million per year vs ~2000 per million for all stroke), but most occur in children or young adults (75% of those being in women).

- Venous infarcts can arise either from cortical vein or venous sinus thrombosis. Those from sinus thrombosis typically cause elevated intracranial pressure.
- The most common outpatient presentation is intracranial hypertension with papilledema from venous sinus thrombosis.
- Brain MRI without and with contrast (CPT 70553) should be performed initially. MRV (CPT 70544) is appropriate when the typical pattern of venous infarction is seen on MRI.
- Children or young adults who present with a stroke in which headache and seizures are prominent, or who are known to have an intrinsic system clotting disorder, can have brain MRI (70553) and MRV (CPT 70544) initially.
- Head CT is often the first procedure done in stroke, and will usually indicate the presence of venous infarcts, but MRI/MRV will still be required if CT shows a venous infarct. Most of these cases are treated in hospital.
- **Reference:**
 - *N Engl J Med* 2005;352:1791-1798
- **PACHD-17.3 Kawasaki syndrome**
 - Kawasaki syndrome (mucocutaneous lymph node syndrome) can cause aseptic meningitis and occasionally pediatric strokes
 - Most patients with this disease are under age 12
 - Coronary aneurysms are the most feared complications (see CD-8.6 Other Indications for CTCA in the adult Cardiac guidelines)
- **PACHD-17.4 Takayasu’s arteritis (“pulseless disease”)**
 - Suspected in patients under age 40 with loss of at least one peripheral pulse, symptoms of limb claudication, and blood pressure asymmetries between limbs.
 - About half of patients have recurrent syncope.
 - Strokes, transient ischemic attacks (TIA’s), amaurosis fugax, and cardiovascular events are common.
 - The illness is seen in young children also.
 - The site of involvement is the aorta and its major branches, including the coronary arteries (see CD-8.6 Other Indications for CTCA in the adult Cardiac guidelines).
 - MRA or CTA is useful for diagnosis and follow-up, and multiple studies (brain to lower limbs) are commonplace.
 - Brain MRI (CPT 70553) is appropriate if there are focal neurological complaints or substantial changes on head or cervical MRA or CTA.
 - Periodic re-evaluation with extensive MRA of the aorta and its primary branches is standard (annual studies are acceptable).
 - **Reference:**
 - *Practical Neurology* 2002;2:80-93

PACHD-18~PITUITARY

- **PACHD-18.1 General Considerations**
 - The initial step in the evaluation of all potential pituitary masses is a detailed history, recent physical examination, and thorough neurological exam, including evaluation of the visual fields.

- Endocrine laboratory studies should be performed prior to considering advanced imaging.
- Pituitary imaging is accomplished by brain MRI, generally done without and with contrast (CPT 70553). Noncontrast MRI (CPT 70551) or MRI Orbit, Face, Neck (CPT 70543) is used at times. CT head without and with contrast (CPT 70470) is acceptable in patients who cannot have MRI.
 - Head CT without and with contrast (CPT 70470) is also occasionally used in addition to MRI to visualize parasellar bony structures in the preoperative evaluation of certain sellar tumors.
 - One study (either brain MRI [CPT 70553] or MRI Orbit, Face, Neck [CPT 70543]) is adequate to image the pituitary. **The ordering physician should specify that the study is specifically to evaluate the pituitary gland. The use of two CPT codes to image the pituitary is not indicated.**
- Pituitary adenomas are uncommonly found in children.
- **PACHD-18.2 Growth hormone (GH) deficiency** (pituitary short stature)
 - In patients suspected of growth hormone deficiency who have failed at least two standard growth hormone stimulation tests, imaging to evaluate the pituitary-hypothalamic area is recommended.
 - One abnormal GH test is sufficient for patients with defined central nervous system pathology, history of irradiation, multiple pituitary hormone deficiency or genetic defect affecting the growth hormone axis.
 - **Reference:**
 - *Endocr Pract* 2003;9:65-76
- **PACHD-18.3 Pediatric diabetes insipidus**
 - The most common assigned causes are intracranial tumor, Langerhans' cell histiocytosis, and "idiopathic."
 - Brain MRI without and with contrast (CPT 70553) is appropriate initially in pediatric diabetic insipidus.
 - **Reference:**
 - *N Engl J Medicine* 2000;343:998-1007
- **PACHD-18.4 Other pituitary region tumors**
 - Craniopharyngiomas arise in the parasellar area, and are the most common tumor of that region in children. Over half of these tumors present by about age 20. Few general rules can be given for follow-up, especially for the adamantinomatous variety generally seen in children.
 - **Meningiomas:** About 10% of meningiomas arise in this area. Evaluation may require CT in addition to MRI at times to evaluate for hyperostosis. Follow-up imaging is as for basal meningiomas in general (see PACHD-12.7 Meningiomas).
- **PACHD-18.5 Precocious puberty**
 - More common in girls than in boys. Defined as the appearance of secondary sexual characteristics before age 8 in girls and before age 9 in boys.
 - Most cases, especially in girls, are of no known cause. However, brain tumors, especially those of the diencephalon, need to be excluded. The most common tumor is a hypothalamic hamartoma.
 - Brain MRI without and with contrast (CPT 70553) is appropriate and usually sufficient.

PACHD-19~MAGNETIC RESONANCE SPECTROSCOPY-PEDIATRIC

- **Magnetic resonance spectroscopy (MRS)**
 - Analysis of the levels of certain chemicals in pre-selected voxels (small regions) on an MRI scan done at the same time (see discussion in HD-35 Newer Imaging Techniques, Evidence Based Clinical Support section in the adult Head guidelines).
 - Certain payers consider MRS investigational, and their coverage policies will take precedence over MedSolutions' guidelines. Prior authorization does not guarantee payment of the study in this situation.
 - **Pediatric uses in neuro-oncology:** MRS is often useful in the management of pediatric brain tumors to determine the need for further therapy. Such cases require referral to a Medical Director.
 - **Reference:**
 - *Neuroimag Clin N Am* 2006;16:169-192
 - MRS is clearly useful in the diagnosis and subsequent management of certain rare inborn errors of metabolism affecting the central nervous system, including adrenoleukodystrophy, creatinine pathway disorders, and others. Cases should be referred for Medical Director review.
 - **References:**
 - *Neuroimaging Clin N Am* 2006;16:87-116. An encyclopedic review of current knowledge regarding use of MRS in inborn errors, disease by disease.
 - *Neurology* 2005;64:434-441
 - *Neurology* 2005;64:406-407
 - MRS produces highly variable results in MS, varying with the pathological process. It does not appear to be useful in distinguishing multiple sclerosis plaques from tumors, since both can produce similar results.
 - The use of MRS in multiple sclerosis, especially in making the differential diagnosis of MS versus tumor, is experimental at this time.
 - Use of MRS in patients with cerebral metastases of systemic cancers is currently regarded as experimental.

PACHD-20~OPTIC NEURITIS

- Also see PACHD-10 Suspected MS and PACHD-11 Established MS
- The diagnosis of optic neuritis can be made clinically - without imaging - with over 99% accuracy.
 - Imaging is done to find associated evidence of Multiple Sclerosis (MS); therefore, brain MRI without and with contrast (CPT 70553) is indicated on initial presentation.
 - Spinal cord imaging (cervical and thoracic spine) may be useful if brain imaging is neither normal nor firmly diagnostic of MS, but in apparently isolated optic neuritis, spinal cord imaging is not often useful.
 - **Reference:**
 - *Neurology* 2004;62:226-233
- Dedicated orbital imaging will usually show demyelination/inflammation of the optic nerve. However, this information is rarely clinically useful and in patients with optic

neuritis, it is not relevant to McDonald criteria scoring for MS. **Orbital MRI is appropriate only in atypical cases.**

- **MRI of the orbits without and with contrast (CPT 70543) is appropriate in the presence of at least one of the atypical features listed below:**
 - Visual loss progressing in severity for more than 10 days.
 - Patient age >45.
 - Lack of *any* pain or soreness with the visual loss.
 - Severe disc edema on clinical examination. Mild disc edema is common in optic neuritis, but severe edema with hemorrhages and exudates is not.
 - Evidence of iritis or uveitis (eye disease not limited to the optic nerve).
 - Failure to manifest at least some improvement in visual acuity within a month of onset.
- **References:**
 - *Magn Reson Imaging Clin N Am* 2002;10:663-678
 - *Lancet Neurology* 2005;4:111-121
- In adults, optic neuritis is generally unilateral. In children bilateral involvement is seen in about 40% of cases.
 - **Reference:**
 - *Neurology* 2006;67:258-262

PACHD-21~EPISTAXIS

- Initial evaluation of epistaxis (nose bleed), including recurrent epistaxis, is by direct or endoscopic visualization of the relevant portions of the upper airway.
 - If the initial clinical evaluation is unrevealing, Ear, Nose, and Throat (ENT) examination may be helpful.
 - Maxillofacial CT may be useful in individual cases, depending upon the findings during the initial clinical evaluation.

PACHD-22~FACIAL TRAUMA

- CT without contrast is the preferred imaging study in facial trauma.
- Coding of Facial imaging:
 - **Maxillofacial versus orbital/temporal bone CT:** both orbital/facial bone CT (CPT 70480, 70481, and 70482) and maxillofacial CT (CPT 70486, 70487, and 70488) cover the structures of the orbits, sinuses, and face. Unless there is a grounded suspicion of simultaneous involvement of more posterior lesions, especially of the region involving the middle or inner ear, one of these studies only should be sufficient.
- Mild mucosal thickening in the paranasal sinuses or mastoids without other abnormalities is common in healthy individuals and not of itself an indication for imaging.
- Ear, Nose, and Throat (ENT), Plastic surgery, or other relevant specialist evaluation is helpful in determining the appropriate imaging pathway.

- Maxillofacial CT (CPT 70486) is the usual study (except in orbital or temporal bone trauma), but the preference of a requesting ENT or neurologist/neurosurgeon should be honored.
- Patients with facial trauma are often at risk for associated injury of both the cranial contents and the cervical spine.

PACHD-23~HEARING LOSS

- Otoscopic and audiological examination is the initial step in evaluating hearing loss of all types.
- **Conductive hearing loss:**
 - Advanced imaging is generally inappropriate in patients with hearing loss caused by benign impaction of one or both external auditory canals.
 - In patients with unilateral conductive hearing loss, especially those with abnormal otoscopic findings, temporal bone CT without contrast (CPT 70480) may be useful.
 - When advanced imaging is necessary in patients with bilateral conductive hearing loss, CT of the temporal bone (70480) is usually appropriate.
 - ENT physicians often use contrasted CT or MRI when malignancy is identified, and this is acceptable.
- **Cochlear hearing loss:**
 - ENT consultation is of benefit in patients with unexplained bilateral cochlear hearing loss.
 - In patients with unilateral cochlear loss, imaging with either brain MRI without and with contrast (CPT 70553) or temporal bone CT (CPT 70480) may be appropriate.
 - MRI (CPT 70553) is generally preferred when a retrocochlear lesion cannot be definitely excluded by other means.
- **Retrocochlear hearing loss:**
 - Brain MRI with attention to the internal auditory canals and without and with contrast (CPT 70553) is helpful in both unilateral and bilateral cases.
- **Cochlear implants:**
 - The surgeon's choice among preoperative craniofacial studies should be honored.
- **Cholesteatoma**
 - If cholesteatoma is suspected by clinical exam and/or symptoms (e.g. painless drainage from the ear, conductive hearing loss, chronic/recurrent ear infections), CT of the temporal bone, contrast as requested (CPT 70480, 70481, or 70482), can be performed.
 - 3D rendering can be performed in conjunction with the temporal bone CT if requested.
 - Brain MRI, contrast as requested, is used if specific problems involving the surrounding soft tissues are suspected such as dural involvement, abscess, herniated brain into the mastoid cavity, inflammation of the membranous labyrinth or facial nerve, or sigmoid sinus thrombosis.

- Also see HD-24.9 Acoustic neuroma and other cerebellopontine angle tumors in the adult Head guidelines since acoustic neuroma is often in the differential diagnosis with cholesteatoma.
- The following imaging studies can be performed for preoperative planning of a known cholesteatoma if ordered by the operating surgeon:
 - CT of the temporal bone, contrast as requested (CPT 70480, 70481, or 70482), with or without 3D rendering
 - Brain MRI, contrast as requested
- **Reference:**
 - Roland PS. *Middle Ear, Cholesteatoma*. eMedicine, June 6, 2006. <http://www.emedicine.com/ent/topic220.htm>. Accessed December 1, 2008

PACHD-24~EAR PAIN (OTALGIA)

- A recent detailed history and physical examination, including an otoscopic examination, should be performed initially.
- Common causes of ear pain include ear infections, dental problems, sinus infection, neck problems, tonsillitis, and pharyngitis.
 - Advanced imaging is not indicated in patients with improvement of symptoms following an episode of one of these common causes of ear pain, including otitis media.
 - Advanced imaging is not indicated in patients with otitis externa.
- If ear pain persists with no obvious cause, CT scan of the temporal bone, contrast as requested (CPT 70480, 70481, or 70482), is the usual initial advanced imaging study.
- In selected cases, brain MRI, contrast as requested, may be necessary
 - Usually brain MRI is considered if a cerebellopontine angle or other intracranial tumor is suspected
 - Also see HD-24.9 Acoustic neuroma and other cerebellopontine angle tumors in the adult Head guidelines
 - Brain MRI, contrast as requested, can be performed in cases of nervus intermedius neuralgia in order to exclude a structural lesion
- **Reference:**
 - Li JC and Brunk J. *Otalgia*. eMedicine. August 26, 2008. <http://www.emedicine.com/ent/TOPIC199.HTM>. Accessed December 1, 2008

PACHD-25~SINUS, CHILD

- **PACHD-25.1 Indications for advanced imaging of the sinuses**
 - Maxillofacial or sinus CT is performed without contrast (CPT 70486) unless specifically indicated otherwise.
 - **Apparent sinusitis in an immunocompromised patient:** in this setting, sinus CT without and with contrast (CPT 70488) may be appropriate, since occult neoplasm and ill-contained infection are often issues.
 - Sinus CT without and with contrast (CPT 70488) is appropriate for sinusitis complicated by local spread of the infection into orbital or facial cellulitis or for other complications of sinusitis.

- Suspected sinus infections should be treated empirically, generally with some combination of antihistamines, steroids, and antibiotics. If there has been no clear cut response within 10 days, especially if fever persists, sinus CT is indicated.
- Sinus CT is also indicated if there is recurrence of a treated infection within 8 weeks of treatment.
- Asthma with upper respiratory symptoms which responds poorly to empirical treatment for at least a week is a potential indication for sinus CT.
- Sinus CT is indicated in the initial evaluation of fungal sinusitis.
- Sinus CT may be indicated prior to upper respiratory endoscopy and as part of a specialty evaluation (ENT or Allergist) of either a persistent sinus problem or pediatric obstructive sleep apnea (see PACHD-25.4 Sleep apnea).
 - Children are not often referred to see ENT and Allergy consultants unless the problem is persistent, so sinus imaging is often a reasonable part of the consultant's initial evaluation.
- CT stereotactic localization (CPT 77011) is sometimes used to direct surgical planning in patients undergoing surgery in this body region. When ordered by the operating surgeon for this purpose, such an operative study is appropriate.
- If there is orbital or intracranial involvement, brain MRI (contrast as requested) may also be useful, typically following maxillofacial CT.
- **PACHD-25.2 Combined head and sinus imaging.**
 - Head CT does not visualize all of the sinuses.
 - Head MRI provides excellent visualization of the sinuses sufficient to recognize sinusitis, and addition of sinus CT for this purpose is unnecessary.
 - In patients being evaluated for potential sinus surgery, separate sinus CT is often appropriate even after a head MRI in order to visualize obstructions to spontaneous mucous flow.
 - Separate head imaging is not generally indicated in patients with a nonfocal neurological examination who have headaches associated with sinus symptoms.
 - Sinus CT or MRI is not indicated for the evaluation of headaches without a more specific indication pointing to a sinus etiology.
- **PACHD-25.3 Repeat sinus imaging:** Repeat sinus CT or MRI is appropriately ordered to address a specific issue in management, and the reasons for the repeat study should be documented. Generally, re-imaging of a patient who has responded satisfactorily to treatment is not appropriate unless needed for preoperative planning of an interventional procedure
- **PACHD-25.4 Sleep apnea:** At times either maxillofacial or neck (soft tissue) CT may be useful to evaluate for structures compressing the nasopharynx. Lateral radiographs should be done initially.
- **PACHD-25.5 References:**
 - *Pediatrics* 2001;108:798-808
 - *ACR Appropriateness Criteria, Sinusitis – child, Updated 2006*
 - *Otolaryngol Clin N Am* 2005;38:1279-1299
 - *N Engl J Med* 1997;337:254-259
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PACHD-7~Subarachnoid Hemorrhage

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