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.

This version incorporates MSI accepted revisions prior to 07/22/11
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PACHD-1~GENERAL GUIDELINES

- **PACHD-1.1 General Considerations**
  - 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.
  - These head imaging guidelines cover a variety of medical conditions and the imaging studies generally appropriate to them. Imaging studies that are not mentioned in conjunction with a particular clinical condition are generally not indicated, but their value will be reviewed on a case by case basis.
  - **Clinical Evaluation**
    - Prior to considering advanced imaging, patients should undergo a recent detailed history, physical examination, including a neurological examination, appropriate laboratory studies, and a clinical plan relevant to the problem should be documented.
  - **Clear Indications**
    - Advanced neuroimaging should only be considered in patients who have or are reasonably suspected of having a neurological disorder (or a neurological extension of a general disorder).
  - **Limitations of Screening**
    - The use of advanced neuroimaging to screen for neurological disorders is not appropriate when the disorder in question is either not known to be familial, or is known to definitely not be familial (e.g. Chiari malformation, ordinary AVM).
    - Screening of asymptomatic individuals using advanced imaging is inappropriate for disorders in which the presence of clinical features is required to make the diagnosis (e.g. multiple sclerosis).
    - At times, age is an important factor in deciding the appropriateness of screening studies.
    - Specific indications for familial screening are discussed in individual guidelines in these PACHD guidelines.
  - **Avoidance of Duplicate Studies**
    - Duplicate or largely overlapping imaging studies are to be avoided unless there is a very well defined reason for performing multiple studies. If two
studies using the same modality both cover the area of clinical interest, only one is generally needed.

- Examples of inappropriate duplicate imaging requests include the following (see HD-1.1 Anatomic Issues in the adult Head Imaging Guidelines for the correct coding of these studies):
  - Requests for imaging both the sinuses and orbits
  - Requests for imaging both the TMJ and either the facial bones or the soft tissues of the neck
  - Requests for two imaging studies to image the brain and pituitary gland

- **Coding Issues**
  - **Brain PET and MR Spectroscopy**
    - Brain PET should be reported as metabolic brain PET (CPT®78608).
    - Brain MR spectroscopy should be reported with CPT®76390
  - **Echoencephalography** is coded as CPT®76506
    - Echoencephalogram (CPT®76506) is an ultrasound study primarily performed on a child or infant less than 12 to 14 months of age. It is used to determine intracranial pathology such as intracranial hemorrhage, fluid collections, masses, or other structural abnormalities.
  - **Transcranial Doppler study**
    - CPT®93886 Transcranial Doppler study of the intracranial arteries; complete study
    - CPT®93888 Transcranial Doppler study of the intracranial arteries; limited study
    - CPT®93890 Transcranial Doppler study of the intracranial arteries; vasoreactive study
    - CPT®93892 Transcranial Doppler study of the intracranial arteries; emboli detection without intravenous microbubble injection
    - CPT®93893 Transcranial Doppler study of the intracranial arteries; emboli detection with intravenous microbubble injection

- **PACHD-1.2 Issues Specific to Pediatric Neuroimaging**
  - Many differences characterize the optimal approach to neuroimaging in the pediatric population vs the optimal approach in adults. Some of these differences arise due to the probability of various illnesses in children vs adults.
  - The differential diagnoses and relative frequency of disorders often differ considerably between children and adults, and between infancy, early childhood, “middle childhood”, and adolescence.
  - **Issues Specific to the Pediatric Population:**
    - **Congenital anomalies**: this is an especially prominent disease category in infancy and childhood. Certain anomalies tend to cluster, and those who have one congenital anomaly are more likely to have certain others, as well.
    - **Radiation**: children are more sensitive to the harmful effects of ionizing radiation, and can expect longer life spans in which to experience those effects. In general, this favors the use of MRI/MRA over CT/CTA if either modality would otherwise be satisfactory (although see **Need for sedation** bullet point below). Exceptions will be mentioned in specific guidelines as necessary.
Need for sedation: the scanning times involved for MRI/MRA generally require that infants and young children be sedated or anesthetized for these procedures.

- CT can usually be performed successfully without sedation (short scanning times).
- Therefore, CT may be performed in children under age 6 rather than MRI if CT will reasonably provide the imaging information needed.
- If MRI is felt to be necessary in these very young children, contrast and coding approvals should be geared toward avoiding a second imaging session.

- Neuroimaging is sometimes needed with infants and young children due to limitations in gathering an accurate history and symptoms, as well as difficulties in interpreting the neurological exam.

**PACHD-1.3 Head CT versus Brain MRI in Childhood**

- **Head CT:**
  - Head CT is normally performed without contrast (CPT®70450)
    - With modern CT scanners, the addition of contrast adds little useful diagnostic information in most settings, doubles the radiation dose, and adds the risk of a contrast reaction.
  - **Head CT is used for the following:**
    - Urgent/emergent settings due to availability and speed of CT
    - Trauma
    - Evaluation for recent hemorrhage, whether traumatic or spontaneous
    - Evaluation for calcifications
    - Evaluation of the bony structures of the head
    - Evaluation and follow-up of hydrocephalus
    - In patients dependent on life support
    - Uncooperative patients (including very young children—see Need for sedation bullet in PACHD-1.2)
    - Prior to lumbar puncture to rule out mass or bleeding
      - If requested, brain MRI without contrast (CPT®70551) can be substituted for head CT

- **Brain MRI**
  - Brain MRI is superior to head CT in evaluating known tumors, the pituitary region, epilepsy, white matter, and inflammation within the brain, and is the appropriate imaging study to use in these conditions, even in very young children.

**PACHD-1.4 Contrast Use in Head Imaging**

- **Head CT**
  - Head CT is normally performed without contrast (CPT®70450)
    - **Exception:** a child with a known or strongly suspected intracranial abscess and an absolute inability to have an MRI can undergo head CT without and with contrast (CPT®70470).

- **Brain MRI**
  - In children, brain MRI is generally performed without contrast (CPT®70551)
Brain MRI without and with contrast (CPT®70553) is indicated for the following (especially in young children who require sedation):
- Well-grounded concern regarding the pituitary region, or
- Known brain tumor, or
- Ataxia, or
- Presence of focal neurological signs (including stroke)

**PACHD-1.5 MRA/CTA in Children (including MRV/CTV)**
- Indications for CT and MR angiography and venography are well defined, and it is inappropriate to request these studies as simply an “add-on” to head CT or brain MRI without clinical information that supports performing the angiography or venography studies.
- Head MRA, CTA, MRV, CTV should not be used as the only initial neuroimaging study(ies) in children prior to review of a recent head CT or brain MRI.
- **Coding:**
  - Head MRA is normally performed without contrast (CPT®70544)
    - 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 (CPT®70548)
    - For stroke or reasonable suspicion of carotid or vertebral artery dissection, neck MRA without and with contrast (CPT®70549) can be performed
    - Some specialists use noncontrast MRA of the neck vessels (CPT®70547) and this is acceptable when specifically requested.
  - Head MRV: report as CPT®70544
- **MRA vs CTA**
  - MRA is usually preferred in children in order to avoid radiation exposure, but CTA may be needed in specific conditions (e.g. vasculitis)
- **Indications for MRA/CTA in Childhood:**
  - Surveillance imaging for children with sickle cell disease or moyamoya disease (head MRA [CPT®70544] should be used)
    - Many patients with S-Thalassemia and some patients with S-C disease require the same management as SS patients.
  - Evaluation for moyamoya disease in children who have had irradiation to the skull base for treatment of neoplasms (head MRA [CPT®70544] or CTA [CPT®70496] can be used)
  - Pediatric stroke (head and neck MRA [CPT®70544 and CPT®70548] or head and neck CTA CPT®70496 and CPT®70498] along with brain MRI without and with contrast (CPT®70553) or noncontrast head CT (CPT®70450) if not already done
  - Evaluation of children with known AVM(s) or with known non-traumatic subarachnoid hemorrhage. This includes children who have brain AVM(s) discovered on MRI or CT (head MRA [CPT®70544] or CTA [CPT®70496] can be used)
    - See PACHD-2.2 AVM
  - Evaluation of infants with aneurysm of the Great Vein of Galen, which is a type of complex vascular malformation (head MRV [CPT®70544] or CTV
[CPT®70496] plus brain MRI, contrast as requested, [if not already performed] can be used)

- See PACHD-2.2 AVM

  - Evaluation of papilledema or pseudotumor of childhood (head MRV [CPT®70544] or CTV [CPT®70496] plus brain MRI, contrast as requested, [if not already performed] can be used)

  - **Berry Aneurysm**
    - Significant-sized or symptomatic berry aneurysms are rare below age 10 and very uncommon below age 20.
    - Advanced imaging for occult berry aneurysms will rarely be appropriate prior to age 20.
    - Also see PACHD-2.3 Aneurysm

- **References:**
PACHD-2~ANEURYSM, ARTERIOVENOUS MALFORMATION

- **PACHD-2.1 Thunderclap Headache or Headache of Explosive Onset**
  - These types of headaches are much less common in children than in adults. Often, it is very difficult to obtain a clear history describing these headaches.
  - Within the first 12 hours, noncontrast head CT (CPT®70450) is appropriate when a headache is documented as thunderclap or explosive onset.
  - After the first 12 hours, noncontrast brain MRI (CPT®70551) can be performed rather than head CT, if head imaging has not already been performed.

- **PACHD-2.2 Arteriovenous Malformation (AVM)**
  - Also see HD-17.3 Arteriovenous Malformations in the adult Head Imaging Guidelines
  - These are uncommon lesions (annual incidence is 1.5 per 100,000 persons)
  - Hemorrhage from an AVM is the most common source of non-traumatic subarachnoid hemorrhage in the first decade of life. In teenagers, it is somewhat less common than aneurysmal hemorrhage.
  - In newborns, AVM’s may present with high output heart failure due to the large volume of blood traversing the shunt.
  - Brain MRI without and with contrast (CPT®70553) can be performed for evaluation of AVM.
  - “Aneurysms” of the Great Vein of Galen are complex venous malformations involving that structure. Clinical presentations vary and include high output heart failure in early infancy and hydrocephalus or subarachnoid hemorrhage in later infancy or childhood.
    - Brain MRI, contrast as requested plus brain MRV/CTV can be performed.
  - **Screening for AVM’s**
    - AVM’s are not familial lesions except in conditions such as hereditary hemorrhagic telangectasia syndrome (OWR).
    - Head imaging for screening is not indicated except for a family history of familial cavernoma

- **PACHD-2.3 Aneurysm**
  - Only 1%-2% of aneurysmal bleeds per year in the United States occur in individuals under age 20, and only one tenth of those bleeds occur in individuals under age 10.
  - Most berry aneurysms in children under age 10 are found by age 2 and involve the junction of the anterior cerebral and internal carotid arteries. Many seem to be associated with anterior midline cerebral anomalies.
  - Berry aneurysms are uncommon in the second decade of life.
  - Emergency noncontrast head CT (CPT®70450) or noncontrast brain MRI (CPT®70551) plus head CTA (CPT®70496) can be performed to evaluate suspected ruptured aneurysm.
  - Head MRA to screen for berry aneurysms is rarely indicated before age 20. Exceptions may apply to a teenager with a family history of aneurysm either in both parents or in an identical twin.
References:
Ataxia refers to an abnormally ill-coordinated or unsteady gait for age.
- “Limb ataxia” refers to impaired coordination (for age) of limbs, especially arms.
- **Note:** developmental failure to acquire the ability to walk is a form of developmental retardation, not ataxia
  (See **PACHD-9~Developmental Delay Mental Retardation**)

Detailed neurological history and recent clinical examination, including a neurological examination, are indicated prior to considering advanced imaging in the evaluation of ataxia.

**PACHD-3.1 Differential Diagnosis of Ataxia in Childhood**
- The differential diagnosis of ataxia in childhood includes:
  - In children less than 2 years old, a normal variation of development
  - Neuraxis tumor
  - Ataxia telangiectasia
  - Friedreich’s ataxia
  - Juvenile lipofuscinosis, Refsum’s disease, Abetalipoproteinemia, and other uncommon genetic progressive ataxias.
  - Sex-linked adrenoleukodystrophy may present atypically, as progressive ataxia in affected males, usually during adolescence
  - Benign ataxia of childhood (postviral)—this has become infrequent since the widespread use of Varicella vaccine.

**PACHD-3.2 Advanced Imaging Studies**
- Brain MRI, contrast as requested, can be performed to evaluate ataxia.
- Cervical spine MRI, contrast as requested, can be performed if brain MRI is non-diagnostic.

**PACHD-3.3 Hereditary and Sporadic Slowly Progressive Ataxia**
- DNA probes or other laboratory tests exist for many of the genetic conditions, but often they are not definitive. Therefore, brain MRI, contrast as requested, can be performed.
  - If brain MRI is non-diagnostic, cervical spine MRI, contrast as requested, can be performed.
    - In younger children, brain MRI and cervical spine MRI can be performed together as initial studies.
- **Ataxia Telangiectasia**
  - After tumor, ataxia telangiectasia is the most common cause of ataxia in children under age 10.
  - Usually transmitted via autosomal recessive mechanisms, and is a systemic disorder with prominent neurological manifestations.
  - Immune (IgA) deficiencies are usual, and frequent sinopulmonary infections are prominent. These children should be considered immunocompromised.
    - Sinus CT, chest x-ray, and sometimes chest CT may be needed
  - Lymphomas and related disorders occur with 50-100 times the expected incidence.
Ataxia and other cerebellar abnormalities appear in early childhood and progress slowly.
The typical conjunctival telangiectasias usually appear after the ataxia is present.
Formes frustes of ataxia telangiectasia occur, with milder features occurring later in life.
- Brain MRI, contrast as requested, can be performed if there is uncertainty about the diagnosis.

Friedreich’s Ataxia
- In the United States, Friedreich’s ataxia is the most common cause of progressive ataxia in adolescents.
- Onset is usually in the teens or early 20’s.
- A GAA expansion is usually identified on DNA testing. The gene is typically an autosomal recessive.
- Type I diabetes and cardiac disturbances are often present.
- Brain MRI, usually without contrast (CPT®70551), is appropriate.

References:
- ACR Appropriateness Criteria, Ataxia, 2009

PACHD-4~AUTISM and AUTISM SPECTRUM DISORDERS

The members of this group, including Asperger syndrome, are classified as pervasive development disorder (PDD).
This is a clinical diagnosis. No lab or imaging study can confirm the diagnosis of autism.
Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.
- In addition, selected lab tests such as lead level, genetic tests, metabolic tests, brain MRI, and EEG may be useful.
- Brain MRI without contrast (CPT®70551) may be useful if there is still uncertainty regarding the diagnosis after the above evaluation has been done.
- There is insufficient evidence-based data to support the use of PET in patients with autism

PACHD-5~BEHAVIORAL DISORDERS

Behavioral disorders of childhood or adolescence generally require no advanced imaging for diagnosis or management.
- 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 the major psychoses are uncommon before mid-adolescence.
PACHD-6~CEREBRAL PALSY

- 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 without contrast (CPT®70450) is appropriate for cases of cerebral palsy of undetermined origin or if a fixed deficit worsens.
- Reference:
  - Neurology 2004;62:851-863

PACHD-7~CHIARI MALFORMATIONS

- **Classification**
  - Chiari malformations involve abnormally hindward positioning ("herniation") of the cerebellum so that the tips of the cerebellar tonsils lie at least 5 mm below the foramen magnum
  - There may be associated abnormalities of the skull base, the cranio-cervical junction, the upper cervical vertebrae, or the spinal cord
  - **Chiari I**
    - The most common type; involves the cerebellar tonsils and is often associated with syringomyelia
    - Usually does not include overt hydrocephalus
    - Usually not symptomatic during childhood, but can present later in the teens
    - Most cases are discovered accidentally on a head scan performed for another indication
    - Symptoms are usually nonspecific, but can include lower cranial nerve palsies or sleep apnea
  - **Chiari II**
    - Incidence is one in 5000 live births; presents in infancy with profound neurological abnormalities and hydrocephalus
    - Includes positioning of the cerebellar vermis, fourth ventricle, and brain stem below the foramen magnum
    - Hydrocephalus occurs
    - Tethered cord and lumbar meningomyelocele are also generally present, and midline cerebral anomalies are common
  - **Chiari III**
    - A more profound malformation which includes an occipital or high cervical encephalocele, extensive syringe/hydromyelia, and cerebral malformation
    - Presents in infancy with profound neurological abnormalities and hydrocephalus
• **Initial Imaging**  
  o **Chiari I**  
    - Brain MRI without contrast (CPT® 70551) and MRI of entire spine without contrast (CPT® 72141, 72146, 72148)  
  o **Chiari II and III**  
    - Brain MRI and MRI of the entire spine, contrast as requested.  
    - Noncontrast head CT (CPT® 70450) to visualize the skull, and/or CT of involved spinal regions may also be necessary  

• **Follow-up Imaging**  
  o **Chiari I**  
    - If initial spine imaging was normal, repeat spine studies should not be necessary unless signs or symptoms appear that suggest a spinal cord disorder  
    - Follow-up spine and head imaging in individuals with syringomyelia/hydromyelia is individualized and should be at the discretion of the specialist following the patient  
    - Patients who have had an MRI confirming Chiari I should not need repeat brain imaging unless there are new clinical abnormalities or a surgical procedure is actively being considered  
  o **Chiari II and III**  
    - Follow-up imaging is individualized and should be at the discretion of the specialist following the patient  
  o **Familial screening is not indicated**

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**PACHD-8~CRANIOSTENOSIS**

• Noncontrast head CT (CPT® 70450) is indicated in the diagnosis of craniostenosis (craniosynostosis), and 3-D rendering (CPT® 76376 or CPT® 76377) may be needed for surgical planning.  
• Maxillofacial CT is not generally necessary to evaluate craniostenosis (craniosynostosis) either pre- or post-op.

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**PACHD-9~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.  

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**PACHD-10~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-11 ~ EAR

11.1 Hearing Loss

Retrocochlear hearing loss is very uncommon in children, so these guidelines will focus on conductive hearing loss and sensory-neural (cochlear) loss. Mixed hearing loss can also occur.

Both conductive hearing loss and sensory-neural (cochlear) loss can either be acquired or congenital.

- Conductive hearing loss is usually acquired but can be congenital.
  - The most common acquired cause is fluid in the middle ear
  - Other causes include anomalies, cholesteatoma, tumors, and disorders of the temporal bone

- Sensory-neural (cochlear) loss is equally divided between acquired and congenital.
  - Congenital causes include infections in utero, especially by CMV. The onset of hearing loss in these cases may be delayed
  - There are many genetic syndromes associated with congenital sensory-neural (cochlear) loss
  - Acquired causes include trauma and complications of bacterial meningitis or other infections

Diagnosis

History, otoscopic examination, and hearing tests such as audiology, speech recognition testing, and auditory evoked responses should be performed prior to considering advanced imaging.

The selection of tests will depend on the age of the child and local factors.

Initial Imaging

- Temporal bone CT without contrast (CPT® 70480) can be performed initially to help determine etiology.
- Children with hearing loss of congenital origin may have brain anomalies as well, and brain MRI without contrast (CPT® 70551) can be performed if the need to evaluate for associated cerebral anomalies is specifically stated.
- In children with unilateral or bilateral sensory-neural (cochlear) loss, high resolution temporal bone CT without contrast (CPT® 70480) is appropriate for initial imaging.
- Brain MRI without and with contrast (CPT®70553) can be useful in selected cases
  - In the rare case of documented retrocochlear hearing loss, brain MRI without and with contrast (CPT®70553) can be performed
- Cochlear implants
  - The surgeon’s choice of preoperative craniofacial studies should be honored
- PACHD-11.2 Earache
  - A recent detailed history and physical examination, including an otoscopic examination, should be performed initially
  - Common causes of ear pain include external and middle 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 without contrast (CPT®70480) is the usual initial advanced imaging study
- PACHD-11.3 Otitis Media
  - Acute otitis media is a very common infection of young children, typically presenting with ear pain and fever
  - The diagnosis is typically made by otoscopic examination, and advanced imaging is generally not indicated
  - CT of the temporal bone without contrast (CPT®70480) is appropriate if any of the following is present:
    - Multiple recurrent attacks of acute otitis media
    - Clinical signs of mastoiditis (can do CT of the temporal bone without and with contrast (CPT®70482)
    - Otitis media with effusion (or serous otitis media) that does not respond to medical treatment
- PACHD-11.4 Cholesteatoma
  - Cholesteatomas are expansive cysts of the middle ear filled with cellular debris
  - Can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane.
  - Hearing loss is conductive, although if the lesion is large enough, mixed type hearing loss may be found
  - When cholesteatoma is suspected by otoscopic exam or symptoms such as painless drainage from the ear, conductive hearing loss, chronic/recurrent ear infections, CT of the temporal bone (CPT®70480 or CPT®70482) can be performed initially
    - 3D rendering (CPT®76376 or CPT®76377) can be approved in conjunction with the temporal bone CT if requested
    - Temporal bone MRI (CPT®70543) can be performed instead of temporal bone CT if requested
    - Brain MRI without and with contrast (CPT®70553) can be performed for pre-operative planning, especially if there is evidence of intracranial
involvement

- Brain MRV (CPT®70544) can be performed for pre-operative planning if there is concern for straight sinus thrombosis
- Post-operative imaging (same studies that were performed pre-op) can be performed one time to ensure complete removal of the cholesteatoma

**PACHD-11.5 Vertigo**

- Vertigo is an uncommon complaint during childhood
- History and physical examination, including otoscopic exam, should be done prior to considering advanced imaging
- Middle ear/Eustachian tube problems are the most common cause of vertigo in children
  - Otoscopic examination is usually very helpful in establishing the diagnosis
  - If the neurological examination is otherwise normal and the vertigo responds to treatment, advanced imaging is not indicated
    - If vertigo persists, CT of the temporal bone without contrast (CPT®70480) can be performed
- Vertigo in children with a normal otoscopic examination and no history of middle ear problems usually reflects migraine, benign paroxysmal vertigo of childhood, benign positional vertigo, or head trauma
  - Benign paroxysmal vertigo of childhood is characterized by episodes (lasting for minutes) of pallor with vertigo and ataxia. Many children later develop typical migraine
  - Other causes are uncommon, and advanced imaging is only useful in selected, persistent cases

**References:**

- The Laryngoscope 1999;109:1642-1647
- Internat J Ped Otolaryngol 2006;70:1547-1554
- Internat J Pediatr Otorhinolaryngol 2003;67:889-894
A detailed clinical assessment and EEG should be performed prior to considering advanced neuroimaging.

- This clinical assessment must be very thorough and should include neurological and general physical examination, case and family history, and, whenever possible, the accounts of eye witnesses of the events.

**Imaging**

- When advanced imaging is indicated, brain MRI (usually without contrast—CPT®70551) is usually sufficient and is strongly preferred over CT. Contrast should be added to the MRI protocol only if there is a progressive neurological deficit or a relevant abnormality identified on a noncontrast scan.
  - Head CTA/MRA, neck CTA/MRA, and cervical spine MRI do not generally add valuable information initially and are not indicated.
  - Noncontrast brain MRI (CPT®70551) will generally be performed in any child with documented new onset of epileptic seizures other than simple febrile seizures.

**Repeat Imaging**

- **Surveillance imaging:**
  - Not generally necessary in generalized epilepsies
  - In focal or partial epilepsies, repeat noncontrast brain MRI (CPT®70551) can be performed at one and two years after diagnosis and then every few years.

- **Repeat imaging on indication:**
  - Repeat brain MRI is indicated for any of the following:
    - There is a change in the type of seizures, or
    - Persistent worsening in the frequency of seizures, or
    - New neurological findings

**Evaluation for Epilepsy Surgery**

- Patient must have had a previous brain MRI and documentation of intractable epilepsy for which surgical treatment or another interventional modality is under active consideration.
  - If these criteria are met, PET (CPT®78608) can be performed.
  - MR spectroscopy (CPT®76390) and metabolic PET using tracers other than FDG are acceptable alternatives to FDG PET if requested by the patient’s referring epileptologist.
    - **NOTE:** Certain payers consider MR Spectroscopy investigational, and their coverage policies will take precedence over MedSolutions’ guidelines.
  - Brain SPECT is another acceptable alternative. Currently, MedSolutions does not prior authorize brain SPECT.

**Febrile Seizures**

- Neuroimaging is not medically necessary in children who have typical febrile seizures.
- Simple febrile seizures occur in children only in the presence of a body temperature of >101°F (>38.4°C) and are brief generalized seizures, usually
lasting no more than several minutes.

- Ninety-five percent of cases occur between the ages of 6 months and 3 years.
  - The incidence of simple febrile seizures in this age group is about 2% per year.
- There is usually only one seizure per febrile episode.
- Focal onset or prolonged seizure activity are not seen in simple febrile seizures, and postictal palsies do not occur.
- Neurological exam after a simple febrile seizure is normal.
- Eighty percent of children with simple febrile seizures have a family history of febrile seizures (a parent or sibling in 25% of cases). Only a few percent have a family history of other types of seizures.
- Not all seizures that occur during a febrile illness are simple febrile seizures. The details are part of the diagnosis.

**References:**
- *Brain* 2009;132:2785-2797

**PACHD-13~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.
2011 PEDIATRIC AND CONGENITAL HEAD IMAGING GUIDELINES

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PACHD-14~HEADACHE, CHILD

- A complete headache history and neurological examination should be performed prior to considering advanced imaging.

- **PACHD-14.1 Children of School Age (over 5 years old)**
  - Headache is a very common complaint in this age group. Many of these children have a family history of one of the primary headache disorders, such as migraine or tension headache.
  - **Stable Headaches**
    - Neuroimaging is very unlikely to be of value in a child with a normal neurological examination who has stable migraine, tension, or school headaches. This is especially true when there is a family history of these disorders.
  - **New Onset Headaches**
    - A four week trial of symptomatic treatment prior to considering advanced imaging is usually appropriate in children with a normal neurological exam.
  - **Indications for Imaging:**
    - Abnormal neurological examination (including papilledema and ataxia)—brain MRI, contrast as requested
    - Concomitant seizure history—brain MRI, contrast as requested
    - Recent head trauma—noncontrast head CT (CPT®70450)
    - Current use of anticoagulants—noncontrast head CT (CPT®70450)
    - Congenital heart disease—noncontrast head CT (CPT®70450)
    - New onset, clearly documented thunderclap headache—noncontrast head CT (CPT®70450) or noncontrast brain MRI (CPT®70551)
    - Morning vomiting—noncontrast brain MRI (CPT®70551)
    - Frequent nocturnal awakening by headaches—noncontrast brain MRI (CPT®70551)
    - Diplopia—noncontrast brain MRI (CPT®70551)
    - Headaches which become severe and progressive within a month of onset—noncontrast brain MRI (CPT®70551)
    - Failure of improvement after four weeks of symptomatic treatment—noncontrast brain MRI (CPT®70551)
  - **MRA/CTA**
    - MRA/CTA is not generally medically necessary in the evaluation of headache in children unless an AVM has been seen on a prior brain MRI or head CT.
  - **MRV**
    - Head MRV (CPT®70544) may be useful in patients with papilledema (See PACHD-10)
• **PACHD-14.2 Preschool Children (age 5 and under)**
  o Headache is a relatively uncommon complaint at this age. The prevalence of the primary headache disorders (migraine headache, tension headache, etc.) is about 2% at age 5.
  o Headache history and an accurate neurological exam are difficult to obtain in this age group.
  o Headache guidelines appropriate to older children must be applied cautiously.
    ➢ Noncontrast head CT (CPT®70450) or noncontrast brain MRI (CPT®70551) will often be appropriate, especially when there is no family history of migraine.

• **References:**
  o *Radiology* 1997;202:819-824
  o ACR Appropriateness Criteria, Headache—Child, Revised 2008
  o *Pediatrics* 2009;124:e12-e17
  o *Neurology* 1992;42:1657-1662
These guidelines apply to adults as well as the pediatric population.

**PACHD-15.1 Macrocephaly**
- Defined as head circumference that is more than two standard deviations above the mean for age, sex, and body size, established by use of measurements and standard growth charts
  - The normal curve dictates that 2.5% of any random group will be over two standard deviations above the mean
- Accelerated head circumference growth by more than one standard deviation from the child’s previous standing can also indicate macrocephaly
- Imaging
  - *Birth to 6 months old:*
    - Ultrasound of the head (CPT®76506) should be performed initially
    - If ultrasound is abnormal, either head CT (CPT®70450) or brain MRI, contrast as requested, can be performed. CT is often chosen initially to see calcifications and to view skull anatomy, but MRI shows more anatomical detail and may be chosen instead.
    - If head CT shows hydrocephalus, then brain MRI, contrast as requested, is appropriate
  - *Age 6 months and older:*
    - Brain MRI, contrast as requested, is preferred initially to CT since uncomplicated hydrocephalus is less likely after early infancy
    - In younger children, head CT is acceptable (if requested) reasons listed in PACHD-1.3 Head CT vs Brain MRI in Childhood

**Megalencephaly**
- Increased head size caused by increased brain size, rather than by excess fluid, tumor, or cyst formation
- Can be either a normal variant or can arise from one of several uncommon disorders
- This term can only be applied after there has been a brain imaging study
- Brain MRI, contrast as requested, is appropriate if it was not done initially

**Hydranencephaly**
- Defective development of the cerebral hemispheres, which are replaced by large cystic structures. The infants seem normal at birth, but then have an abnormally enlarging head and profound failure to achieve milestones.
- This condition is seen on cerebral ultrasound (CPT®76506), but once see,
requires further evaluation by brain MRI, contrast as requested

- **PACHD-15.2 Microcephaly**
  - The definition is similar to that for macrocephaly in reverse.
  - Brain MRI, contrast as requested, is recommended initially since CT may not detect the relevant anatomical abnormalities

- **PACHD-15.3 Hydrocephaly (hydrocephalus)**
  - Most common identifiable cause of macrocephaly
  - Incidence is about 1 per 4000 live births
  - Almost all hydrocephalus is obstructive, except hydrocephalus due to choroid plexus papillomas
  - Hydrocephalus is traditionally divided into non-communicating (the obstruction lies within the course of the brain’s ventricular system) and communicating (the obstruction is distal to the ventricular system)

- **Etiologies**
  - Aqueductal stenosis or gliosis
    - Accounts for about 20% of hydrocephalus cases
    - Risk of occurrence in siblings is a few percent
    - Symptoms are of insidious onset and can begin at any age
    - Acute exacerbations can occur after minor head trauma
  - Chiari malformation
    - Accounts for about 40% of cases
    - See PACHD-7 Chiari Malformations
  - Dandy-Walker malformation
    - Accounts for 10% of cases
    - Hydrocephalus generally becomes evident during the third to twelfth month of life
    - Triad of findings includes cystic dilatation of the fourth ventricle, full or partial agenesis of the cerebellar vermis, and upward displacement of tentorium and associated venous structures
    - Must be distinguished from an isolated finding of an enlarged cistern magna, which is a normal anatomical variant
  - Communicating hydrocephalus
    - Accounts for 30% of cases
    - Can present at any age, and actual macrocephaly may be minimal or absent if the event causing the hydrocephalus occurs after mid-childhood
    - Underlying causes include prior bacterial meningitis, prior subarachnoid hemorrhage, and more rarely, malformations of the straight sinus or vein of Galen and subarachnoid infiltration by lymphoma or leukemia.

- **Initial Imaging**
  - First 6 months of life:
    - Screening head ultrasound examination (CPT® 76506)
    - If ultrasound is abnormal, brain MRI, contrast as requested, can be performed
  - Greater than 6 months old:
    - Brain MRI, usually without contrast CPT® 70551, but can be contrast as requested. Head CT without contrast (CPT® 70450) can be substituted if
requested

- **Spine imaging**
  - Spine MRI may be indicated in individuals with Chiari malformation (multiple spine segments), Dandy-Walker malformation (cervical spine only), or malignant infiltration of the meninges

- **Surveillance Imaging after Shunting**
  - Noncontrast brain MRI (CPT®70551) or head CT (CPT®70450) can be performed a few months after shunt placement and then once a year
    - MRI provides more anatomical detail and does not involve radiation exposure, but many providers use head CT, especially in individuals younger than school age
    - Complications after shunting include obstruction, disconnection, infection, catheter tip migration in the brain, and the need for revision created by the child’s growth. These complications are difficult to recognize clinically.

- **Shunting into the peritoneum (VP shunts)** can give rise to abdominal complications, but these are generally symptomatic, so surveillance imaging of the abdomen is not indicated

- **Re-imaging for Indication**
  - Head CT without contrast (CPT®70450) or brain MRI, contrast as requested, can be performed for new signs or symptoms such as sepsis, altered level of awareness, protracted vomiting, visual or neurological deterioration, decline of mentation after initial improvement, or new onset of seizures

- **Familial Screening**
  - Not generally indicated except in siblings of individuals with aqueductal stenosis (noncontrast head CT (CPT®70450) or MRI (CPT®70551))

- **Reference:**
  - Neurology 2009;73:887-897
**PACHD-16~MAGNETIC RESONANCE SPECTROSCOPY-PEDIATRICS**

- **Magnetic Resonance Spectroscopy (MRS)—CPT®76390**
  - 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-34 Newer Imaging Techniques, Evidence Based Clinical Support section in the adult Head Imaging Guidelines).
    - Certain payers consider MRS investigational, and their coverage policies will take precedence over MedSolutions’ guidelines.
  - **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.
  - 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.
  - 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.

- **References:**
  - *Neurology* 2006;64:434-441

**PACHD-17~MULTIPLE SCLEROSIS**

- Multiple sclerosis is distinctly uncommon in children. About 4% of MS cases begin before age 17, and only approximately 0.5% begin before age 10.

- **Initial Diagnosis**
  - Ataxia, optic neuritis, diplopia, and transverse myelitis are common presentations.
    - MS can present as an acute encephalitis-like illness, especially in childhood.
  - Among children with suspected demyelinating diseases, the principal differential diagnosis is often between MS and one particular isolated clinical syndrome—acute disseminated encephalomyelitis.
    - Other demyelinating disorders, such as various leukodystrophies and DeVic’s disease (neuromyelitis optica) may also need to be considered.
    - Non-demyelinating diseases such as brain tumor may also need to be
considered in the differential.

- **Advanced Imaging**
  - MRI is necessary to confirm a demyelinating disease and to help distinguish between the different demyelinating diseases.
  - CT, CTA, and MRA are generally not of value.
    - CT may occasionally be useful to exclude a mass lesion.
  - PET and MR spectroscopy are not generally indicated in evaluating MS.
    - On a rare occasion, PET may aid in distinguishing “tumefactive” demyelinating lesions from tumor.
  - **Initial imaging for MS**
    - Brain MRI without and with contrast (CPT®70553) and MRI of the spinal cord without and with contrast can be performed.
      - Cervical and thoracic spine MRI scans visualize the entire spinal cord, and lumbar spine MRI usually is not needed.
      - Sagittal MRI of the spinal cord with phased array detector coil (CPT®72156 or CPT®72157) OR full MRI of the cervical (CPT®72156) and thoracic spine (CPT®72157) can be performed to visualize the entire spinal cord.
  - **Follow-up imaging in established MS**
    - Surveillance MRI, contrast as requested, of the brain and spinal cord can be performed twice a year.
    - MRI, contrast as requested, of the brain and spinal cord can be performed whenever an episode with significant new neurological deficit occurs.
  - **Screening studies for MS**
    - Screening studies are inappropriate in asymptomatic children and in those whose clinical picture does not support a diagnosis of MS.

- **References**
  - McAlpine ed. 4, pp.343-346
  - *Neurology* 2010;74:1412-1415
  - *Neurology* 2009;72:961-967
  - *Neurology* 2009;72:968-973
  - *Neurology* 2010;1404-1405
  - *Neurology* 2006;66:1485-1489
  - *AJNR* 2006;27:455-461
PACHD-18 GENERAL REMARKS

Pediatric versus Adult Neuro-Oncology

- Nervous system tumors are the most common non-hematologic tumors of childhood. There are about 1500 cases diagnosed annually in the United States.
- Tumor types commonly seen in the pediatric population are different from those typical in adults.
  - Pediatric brain tumors are more commonly seen in the cerebellum and brain stem; adult brain tumors more commonly occur in the cerebral hemispheres.
- It is more useful to classify tumors in the pediatric population by location rather than by tumor type (cell of origin).
- There are a number of tumor-associated genetic syndromes which are important in the pediatric population.
- In the United States, children with known nervous system tumors are managed by neurologists, neurosurgeons, and oncologists who specialize in pediatric neuro-oncology and use internationally standardized treatment and imaging protocols. Requests for imaging studies made within this setting are generally appropriate unless there is a conflict with the coverage policy of the payer regarding certain studies.

Imaging Studies

- For all pediatric brain tumors, brain MRI without and with contrast (CPT® 70553) is appropriate initially, for surveillance studies, and if there are clinical indications of recurrence.
- CTA, MRA, and catheter angiography are at times useful for preoperative planning.
- Head CT is occasionally useful to evaluate calcifications or bony structures, or to look for hemorrhage.
- Imaging studies may be needed if the location of the tumor makes biopsy very hazardous

### PACHD-18.2 Cerebellar Tumors
- Symptoms typical of tumors in this group include vomiting, headache, ataxia, and visual impairment with papilledema.
- **Medulloblastoma or PNET (Primitive Neuro-Ectodermal Tumor)**
  - Age of onset is early to middle childhood (3 to 9 years old), but there is a secondary peak in the early 20’s.
  - This tumor often spreads throughout the neuraxis by subarachnoid seeding.
  - **Initial Evaluation:**
    - Brain MRI without and with contrast (CPT®70553) and MRI of entire spine (CPT®72156, CPT®72157, and CPT®72158)
  - **Treatment:**
    - In most cases, treatment with a variable combination of radiation therapy and chemotherapy is intended to be curative, and often is.
  - **Surveillance Imaging:**
    - A typical surveillance imaging protocol includes repeat brain MRI without and with contrast (CPT®70553) every 3 months for the first year, every 4 months for the second year, every 6 months for the third year, and then brain MRI (CPT®70553) at year 4, year 5, year 7, and year 10.
    - Indications for repeat spine imaging will depend upon the case.
  - **Recurrence or the presence of complications** may dictate the need for additional studies and restaging studies.
  - **Extra-neural metastases**
    - PNET is an exception to the “rule” that brain tumors do not metastasize outside of the nervous system. In a few percent of cases, PNET deposits are found outside the nervous system, most often in bone and sometimes in peritoneum.
    - The need for extra-neural imaging will be infrequent and very case-specific.
    - PNET systemic metastases seem to be FDG-avid (PNET is biologically similar to Ewing’s sarcoma) and PET scan can be considered on a case-by-case basis.
- **Ependymoma**
  - Usually arise from the floor of the fourth ventricle.
  - Common tumor of childhood (usually first decade of life) and occasionally are found in adults
  - Ependymoblastomas (“malignant ependymomas”) appear to be a variation of PNET, and have occasionally been found to metastasize outside of the nervous system.
  - **Surveillance imaging** will depend on the histological type and clinical behavior of the tumor, and may be as intense as that for PNET (see **Surveillance Imaging** for Medulloblastoma or PNET above)
    - If there is no known spread of tumor to the spinal cord, MRI of the spine
(CPT®72156, CPT®72157, and CPT®72158) every year for three years can be performed.

- **Pilocytic (or cystic or grade 1) Astrocytoma of the Cerebellum**
  - A non-infiltrating tumor usually arising in a cerebellar hemisphere.
  - Clumsiness and limb ataxia are commonly the earliest symptoms.
  - These tumors are mostly resectable.

  - **Surveillance imaging**
    - Following complete resection: brain MRI (CPT®70553) every 6 months for the first year or two, then brain MRI (CPT®70553) at year 7 and year 10.
    - Following partial resection: brain MRI (CPT®70553) every year from years 2 to 10.
  - Infiltrating cerebellar astrocytomas (grade 2 or above) are much less common than pilocytic astrocytomas.
    - Surveillance imaging will at least be the same as for partially resected pilocytic astrocytoma.
    - Further treatment after surgery may create the need to perform restaging MRI.

- **Hemangioblastoma of the Cerebellum**
  - A manifestation of von Hippel-Lindau disease
  - Usually resectable, but new tumors may form
  - See also **HD-23.12 von Hippel Lindau Disease** in the adult Head Imaging Guidelines

- **PACHD-18.3 Brain Stem Glioma (astrocytoma)**

  - **Clinical**
    - Comprise about 10% of pediatric brain tumors
    - Peak incidence occurs at 6-8 years old
    - Presentation is typically with progressive cranial nerve palsies, long tract signs, and cerebellar signs, with later and variable appearance of increased intracranial pressure
    - The pons is the most common location
    - Long term survival is uncommon
  
  - **Imaging**
    - Brain MRI without and with contrast (CPT®70553) initially
    - **Re-staging scans** (usually brain MRI-CPT®70553) may be needed during radiation therapy or chemotherapy
    - **Surveillance imaging:**
      - Brain MRI without and with contrast (CPT®70553) every 4 months for the first two years, twice a year in the third year, then once in year 4, year 5, year 7, and year 10.
      - For low grade astrocytomas, whole spine imaging is not generally useful unless there is a documented reason to suspect subarachnoid seeding. Cervical spine MRI without and with contrast (CPT®72156) may be useful when there is reason to suspect cervical cord infiltration.
      - For high grade tumors, spine imaging once a year for 4 years can be performed.
● **PACHD-18.4 Midline Supratentorial Tumors**
  ○ **Pituitary Adenomas**
    ➢ Very uncommon in children
    ➢ Usually hormonally active if found in the pediatric population
    ➢ See also **PACHD-20.5 Secreting Adenomas**
  ○ **Craniopharyngioma**
    ➢ The most common supratentorial tumor of childhood
    ➢ A suprasellar tumor which extends into the sella in about 50% of cases
    ➢ In children, these tumors almost always contain calcifications and cysts
    ➢ **Presentation**
      ▪ Increased intracranial pressure is the most common presentation in the very young
      ▪ Older children tend to present with blurry vision and, on examination, bitemporal hemianopsia
      ▪ Endocrine abnormalities, diabetes insipidus, and features of hypothalamic compression can also be seen
    ➢ **Imaging**
      ▪ Either brain MRI, contrast as requested, or noncontrast head CT (CPT®70450), or both can be performed initially
        ➢ Head CT is performed to show the cysts and calcifications
      ▪ Other imaging studies are not often helpful
        ➢ **Exception:** thin-cut brain MRI, contrast as requested, plus either head MRA (CPT®70544) or head CTA (CPT®70496) can be performed for surgical planning
      ▪ **Surveillance imaging**
        ➢ Complete resection is achieved in about 75% of those cases treated surgically. Surveillance brain MRI, contrast as requested, or noncontrast head CT (CPT®70450) is generally needed, but with individualized schedules per the treating specialist’s preference
  ○ **Optic Glioma**
    ➢ This is a grade 1 astrocytoma
    ➢ Uncommon tumor (about 2% of all pediatric brain tumors), but is the most common intracranial tumor seen in Neurofibromatosis, Type 1.
      ▪ About half of patients with optic glioma have Neurofibromatosis, Type 1
    ➢ **Presentation**
      ▪ In very young children, the diagnosis is often delayed
      ▪ In older children, loss of vision, abnormal fundoscopic examination, and exophthalmos (when present) lead to early diagnosis.
    ➢ **Imaging**
      ▪ Brain MRI without and with contrast (CPT®70553) initially
      ▪ Orbital MRI without and with contrast (CPT®70543) can also be performed initially if requested by an ophthalmologist or neurologist.
    ➢ **Surveillance imaging**
      ➢ Management of optic gliomas varies greatly from case to case, and surveillance depends on management choices and requires individualized schedules per the treating specialist’s preference.
Pineal Tumors
- Comprise about 5% of all pediatric intracranial tumors
- **Four main basic types:**
  - Germ cell tumors
    - Germ cell tumors tend to occur in males in their teenage years or twenties
  - Pinealomas
  - Teratomas
  - Gliomas
- **Presentation**
  - Parinaud’s sign (paralysis of upward gaze and poor accommodation) (75%)
  - Argyll Robertson pupil (50%)
  - Diabetes insipidus (30%)
  - Diplopia (30%)
  - Hypopituitarism (25%)
  - Precocious puberty (10%)
- **Imaging**
  - Brain MRI without and with contrast (CPT®70553) initially
  - Noncontrast head CT (CPT®70450) can also be performed initially to find calcifications, which aid in identifying teratomas.
  - Whole spine MRI may be appropriate if requested, since spread of tumor through the CSF can occur with some of these tumors
- **Subsequent imaging**
  - Requires individualized schedules per the treating specialist’s preference

Ventricular Tumors
- **Choroid plexus papilloma**
  - Generally benign tumor of the choroid plexus of one of the four ventricles (usually the lateral ventricles)
  - Can present with hydrocephalus caused by increased CSF production
  - Appearance on brain MRI without and with contrast (CPT®70553) is typical, and they are usually treated by excision
  - Occasionally can be malignant (usually patients are 2-3 years old). Follow-up imaging will be similar to that for a malignant brain tumor
- **Colloid cysts of the third ventricle**
  - Comprise less than 1% of all brain tumors and are not specific to any age group
  - Cause hydrocephalus with headaches, which may be explosive, intermittent, and postural
  - Brain MRI without and with contrast (CPT®70553) and head CT (CPT®70450) appearance is diagnostic and surgery is usually curative.
  - Prolonged imaging surveillance is rarely needed following resection, although one repeat noncontrast head CT (CPT®70450) or brain MRI (CPT®70551) can be performed 6 months to a year after resection
• **PACHD-18.5 Tumors of the Cerebral Hemispheres**
  o These tumors comprise a large minority of pediatric brain tumors (compared with 85% in adults)
  o **Meningiomas**
    ➢ Rare in childhood, but often behave like aggressive meningiosarcomas rather than benign tumors
    ➢ Brain MRI without and with contrast (CPT®70553) for initial imaging
    ➢ Noncontrast head CT (CPT®70450) can also be performed initially to visualize skull involvement
    ➢ Surveillance brain MRI without and with contrast (CPT®70553) can be performed every 3-6 months during the first two years and then once a year for life
    ➢ Re-staging brain MRI without and with contrast (CPT®70553) may be needed after radiation treatment or chemotherapy
  o **Astrocytomas (including oligodendroglioma and glioblastoma)**
    ➢ Brain MRI without and with contrast (CPT®70553) for initial imaging
    ➢ Noncontrast head CT (CPT®70450) can also be performed initially to detect calcifications, which occur in about 25% of pediatric hemispheric astrocytomas
    ➢ Surveillance imaging depends on the grade of the tumor and the specifics of treatment and requires individualized schedules per the treating specialist’s preference
      ▪ Annual re-imaging for 5 years will be required at a minimum
  o **Ganglioglioma**
    ➢ Usually not found until the teenage years
    ➢ Comprise 2%-3% of pediatric brain tumors
    ➢ 75% occur in the temporal lobes
    ➢ Presents with history of intractable temporal lobe epilepsy
    ➢ Brain MRI without and with contrast (CPT®70553) for initial imaging
    ➢ This tumor is not seen on CT unless calcified
    ➢ Can have a cystic appearance, and if it also does not enhance, it can be confused with an arachnoid cyst

• **PACHD-18.6 PET and Newer Imaging Techniques**
  o **PET**
    ➢ Certain payers consider the use of brain PET in tumors to be investigational, and their coverage policies will take precedence over MedSolutions’ guidelines.
    ➢ FDG-PET is used to distinguish late post-radiation necrosis from tumor recurrence when MRI is inconclusive (which it often is)
    ➢ FDG-PET is only reliable in very metabolically active tumors (PNET, grade 3 or 4 astrocytomas, ependymoblastomas)
      ▪ There are experimental PET ligands which can overcome this limitation
    ➢ FDG-PET is not useful in distinguishing early phase radiation-related necrosis from tumor recurrence, since recently irradiated tissue can be expected to be FDG-avid
Magnetic Resonance Spectroscopy (CPT®76390)
- Certain payers consider MR Spectroscopy investigational, and their coverage policies will take precedence over MedSolutions’ guidelines.
- Useful as an inexpensive and radiation-free alternative to FDG-PET to distinguish late post-radiation necrosis from tumor recurrence when MRI is inconclusive.
- In well-differentiated astrocytomas and related tumors located in brain areas for biopsy (mainly the diencephalon and brain stem), FDG-PET is of limited use, whereas MR spectroscopy remains useful.

PACHD-18.7 Inherited Syndromes that include Brain Tumor
- Neurofibromatosis, type 1 (von Recklinghausen’s Disease, NF 1)
  - Autosomal dominant, but a positive family history is obtained in only half of cases.
  - Subcutaneous neurofibromas and multiple café au lait spots are typical.
  - Kyphoscoliosis is common and may cause cord compression.
  - Spinal dural ectasias and meningoceles occur.
  - Intraspinal tumors are frequent.
  - Screening individuals who do not have symptoms or signs is usually not useful, since most occult neurofibromas do not grow aggressively.
  - Optic nerve and brain stem gliomas are common and can be evaluated with brain MRI without and with contrast (CPT®70553)
    - MRI of the orbits (CPT®70543) can be performed if optic nerve lesions are known or suspected.
    - 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 prompt brain MRI without and with contrast (CPT®70553) should be performed due to elevated tumor risk and high incidence of aqueductal stenosis (hydrocephalus).
  - Neurofibromatosis, type 1 is a known cause of strokes and moyamoya disease. Imaging should follow guidelines for pediatric stroke.
    (See PACHD-24 STROKE)
  - Imaging to screen family members without signs of the disease is generally inappropriate since the clinical picture is readily recognized.

- Neurofibromatosis, Type II
  - This 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 that are present determine the imaging.

- Tuberous Sclerosis
  - See PACHD-30 Tuberous Sclerosis

- von Hippel Lindau Syndrome
  - See PACHD-31 von Hippel Lindau Disease

PACHD-18.8 Retinoblastoma (including evaluation of leukokoria)
- Orbital CT without and with contrast (CPT®70482) is preferred initially.
- Once a retinoblastoma has been confirmed, MRI of the brain and orbits without...
and with contrast (CPT®70553 and CPT®70543) can be performed

- **PACHD-18.9 Unexplained Vomiting in Children**
  - A thorough evaluation for GI causes should be performed initially
  - If no GI cause is found, noncontrast head CT (CPT®70450) or MRI (CPT®70551) can be performed to exclude an intracranial mass

- **References:**
  - *Cancer* 1983;53:974-981
  - *Cancer* 1985;56:1782-1785
Also see PACHD-17 Multiple Sclerosis

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.

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.

In adults, optic neuritis is generally unilateral. In children bilateral involvement is seen in about 40% of cases.

References:

- Neurology 2004;62:226-233
- Lancet Neurology 2005;4:111-121
- Neurology 2006;67:258-262
### PACHD-20~PITUITARY

#### PACHD-20.1 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.
- When pituitary imaging is indicated, brain MRI without and with contrast (CPT® 70553) is preferred.
  - 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 reporting of two CPT® codes, to image the pituitary, is not indicated.**

#### PACHD-20.2 Panhypopituitarism/somatotrophin (growth hormone) Deficiency (pituitary dwarfism)
- Affected children will be normal size for gestational age at birth, but their growth charts will display failure to have normal growth after birth, and bone maturity will lag. Over time, other signs of pituitary failure may become apparent.
- Endocrine testing should be performed initially, and will reveal growth hormone deficiency with or without associated deficiency of other pituitary hormones
  - For isolated growth hormone deficiency, two measurements of growth hormone with stimulation are performed.
    - In children, normal stimulated values are greater than 10 ng/ml
    - Laron syndrome is caused by a rare inherited mutation of the growth hormone receptor. Affected children have the features of growth hormone deficiency, but have normal or elevated growth hormone levels
- Primary failure—15%-20% of all cases, with autosomal recessive, X-linked, and dominant patterns of transmission. Many specific DNA defects are known.
Secondary failure—causes can include mass of the sellar region (see [PACHD-18.4 Midline Supratentorial Tumors](#)), histiocytosis, trauma, hemorrhage, sarcoidosis, bacterial meningitis, etc.

- Radiation of the skull base for brain tumor or leukemia requires special mention. Growth hormone deficiency can be a result of dose-related radiation damage and these children must be monitored for this.

**Initial Imaging**

- Brain MRI, contrast as requested, with special attention to the pituitary, can be performed when cause of the clinical presentation is in doubt

**Re-imaging**

- The need for re-imaging depends on the particular condition and should be at the preference of the specialist following the patient

### PACHD-20.3 Diabetes Insipidus (DI) and Other Disorders of Anti-Diuretic Hormone

- Vasopressin (ADH) acts on the kidneys as the principal regulator of serum osmolality. In its absence, excess water is excreted and serum sodium rises.
- Vasopressin (ADH) is secreted by the paraventricular and supraoptic nuclei of the hypothalamus, but is released from the posterior pituitary
- The principal evaluation of ADH deficiency is by urine and blood electrolyte and osmolality testing—serum osmolality greater than 300 with urine osmolality less than 300
- Deficiencies in ADH can either be central or nephrogenic

#### Central Diabetes Insipidus (DI)

- Causes include congenital defects (pure or associated with midline anomalies), prior neurosurgery, trauma to the skull base, and hypothalamic/pineal region tumors

- **Imaging**
  - Brain MRI without and with contrast (CPT®70553) is usually appropriate initially
  - Noncontrast head CT (CPT®70450) with attention to the skull base may be useful in traumatic cases
  - If the cause remains uncertain based on the initial brain MRI, serial brain MRI scans (CPT®70553) at 6 month to one year intervals can be performed, since germinomas and other hypothalamic region tumors can initially be difficult to identify on MRI

#### Nephrogenic DI

- Once this diagnosis is firmly established, further advanced imaging is usually not indicated

#### Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH)

- SIADH is rare in childhood
- When present, it is usually in the context of skull base neurosurgery, major head trauma, or severe intracranial disease
- Laboratory studies should be obtained prior to considering advanced imaging—urine osmolality should be high and serum osmolality low

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• **PACHD-20.4 Precocious Puberty**  
  o 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.  
  o Most cases, especially in girls greater than 2 years old, 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-20.5 Secreting Adenomas**  
  o **Pituitary Adenomas**  
    - These are rare in childhood  
    - Endocrine work-up should be performed initially. If the indications for imaging are met (see **HD-27.1 Microadenomas** and **HD-27.2 Macroadenomas** in the adult Head Imaging Guidelines) then brain MRI without and with contrast with attention to the pituitary (CPT®70553) can be performed.  

  o **Prolactinomas and Galactorrhea in Adolescent Girls**  
    - Prolactinoma is the most common functioning pituitary adenoma of childhood  
    - As in adults, it presents with unexplained galactorrhea  
    - Brain MRI without and with contrast with attention to the pituitary (CPT®70553) is appropriate if there is unexplained, confirmed elevation of serum prolactin, or if galactorrhea persists for at least 6 months, regardless of serum prolactin level.

  o **ACTH-producing Tumors**  
    - These tumors usually present with growth failure caused by adrenal over-activity  
    - Endocrine work-up to measure hormone levels should be performed initially and usually point to pituitary Cushing’s disease rather than primary adrenal over-activity  
    - If endocrine work-up supports pituitary Cushing’s disease, then brain MRI without and with contrast with attention to the pituitary (CPT®70553) is appropriate.

  o **Pituitary Gigantism**  
    - This condition is part of the differential diagnosis of overly tall stature  
    - The diagnosis is often not made until there is a delay in the appearance of sexual maturation  
    - Endocrine work-up should be performed initially—failure of growth hormone levels to be depressed to below 5 ng/ml by glucose loading is diagnostic of pituitary hyperactivity. Since growth hormone is normally released in pulses, a random blood level of growth hormone is not sufficient.

  - **Imaging**  
    - Brain MRI without and with contrast with attention to the pituitary (CPT®70553) can be performed to identify or exclude a pituitary adenoma if endocrine work-up supports the diagnosis of pituitary hyperactivity.

    - Reactive hypertrophy of the growth-hormone-producing cells in the pituitary can occur in a variety of illnesses which interrupt the normal feedback inhibition of growth hormone production. This can be extensive enough to
enlarge or erode the sella.

- **Soto’s Syndrome (cerebral gigantism)**
  - Characterized by excessive height, frontal bossing of the skull, a particular facial appearance, and variably present clumsiness and mental retardation. Skeletal maturation is usually normal.
  - Brain MRI without and with contrast with attention to the pituitary (CPT®70553) can be performed if this diagnosis is suspected

  - **Thyrotropin-producing Tumors**
    - These are rare tumors in which both TSH and thyroid hormone levels are elevated (ordinary hyperthyroidism presents with elevated thyroid hormones and low TSH levels)
    - Gonadotropin-producing tumors are not known to occur in children

- **PACHD-20.6 Pituitary Region Masses**
  - See [PACHD-18.4 Midline Supratentorial Tumors](#)

- **References:**
  - Endocr Pract 2003;9:65-76
  - N Engl J Medicine 2008;358:2366-2377
PACHD-21~PSEUDOTUMOR CEREBRI (BENIGN or IDIOPATHIC INTRACRANIAL HYPERTENSION)

- Presents with headaches, a normal neurological examination, and papilledema
- Pediatric cases are often associated with veno-occlusive disorders such as lateral sinus thrombosis, or with the use of certain drugs (vitamin A, isoretinoin, tetracyclines, steroid withdrawal)
- **Imaging**
  - Brain MRI, contrast as requested (to exclude a mass) and brain MRV (CPT®70544) can be performed initially in children
  - Brain MRV may not be needed in overweight females who are age 17 or older
  - MRI of the orbits, contrast as requested, can be performed if there is concern about orbital pseudotumor. This concern should be documented, but it can be assumed if the request is from an ophthalmologist
  - Repeat imaging studies are generally not necessary
- **Reference:**
  - *Headache Currents* 2005 Jan/Feb;2(1):1-10
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**PACHD-22~SINUS and FACIAL IMAGING**

- **PACHD-22.1 Sinusitis in Immune-Competent Children**
  - Suspected sinus infections should be treated empirically, generally with some combination of steroids, and antibiotics
  - When advanced imaging is indicated, the study should be CT of the sinuses without contrast (CPT®70486) unless otherwise specified
  - **CT of the sinuses without contrast (CPT®70486) can be performed if any of the following is present:**
    - No improvement after 10 days of treatment (7 days if the child also has asthma)
    - Recurrence of a treated infection within 8 weeks of treatment
    - Fungal sinusitis
    - Evidence of spread of sinusitis such as orbital or facial cellulitis, or features suggesting intracranial extension—sinus CT either without (CPT®70486) or without and with contrast (CPT®70488) can be performed
      - **NOTE:** children are not generally referred to see ENT or Allergy specialists unless the problem is persistent, so sinus imaging is often a reasonable part of the specialist’s initial evaluation
  - Mild mucosal thickening in the paranasal sinuses or mastoids is often incidentally noted on head imaging studies done for other indications. If there are no other abnormalities of facial structures noted, this finding is not an indication for advanced imaging of the sinuses or temporal bone

- **PACHD-22.2 Sinusitis in Immune-Compromised Children**
  - Suspected sinus infections in children with compromised immune systems require a more aggressive diagnostic approach, since occult neoplasm and spread of infection are possibilities
  - Immune-compromised children include children with cystic fibrosis, known malignancies, long-term treatment with steroids or other immune suppressants, HIV disease, hypogammaglobulinemias, ataxia telangiectasia, severe congenital heart disease, etc.
  - Initial imaging can include sinus CT without contrast (CPT®70486) or without
and with contrast (CPT®70488)

- **PACHD-22.3 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-22.4 Additional Uses of 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.
  - Facial trauma—CT sinus without contrast (CPT®70486)
    - See **PACHD-29.2 Facial Trauma**
  - Congenital anomalies of facial structures—CT or MRI of the sinuses without contrast (CPT®70486 or CPT®70540)
  - Tumors or other disorders of facial structures—CT or MRI of the sinuses without and with contrast (CPT®70488 or CPT®70543)
  - Obstructive sleep apnea—see **PACHD-23 Sleep Disorders of Childhood**
  - Repeated attacks of sinusitis—CT of the sinuses without contrast (CPT®70486) or if immune-compromised, CT of the sinuses without and with contrast (CPT®70488)

- **PACHD-22.5 Stereotactic CT Localization Scan (CPT®77011)**
  - A stereotactic CT localization scan is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The information provides exact positioning of surgical instruments with regard to the patient’s 3D CT images.
  - In most cases, the preoperative CT is a technical-only service that does not require interpretation by a radiologist.
    - The imaging facility should report CPT®77011 when performing a scan not requiring interpretation by a radiologist.
    - If a diagnostic scan is performed and interpreted by a radiologist, the appropriate diagnostic CT code (e.g. CPT®70486) should be used.
    - It is not appropriate to report both CPT®70486 and CPT®77011 for the same CT stereotactic localization imaging session.
    - 3D Rendering (codes CPT®76376 or CPT®76377) should not be reported in conjunction with CPT®77011 (or CPT®70486 if used). The procedure inherently generates a 3D dataset.
    - When ordered by the operating surgeon for this purpose, such an operative study is appropriate

- **PACHD-22.6 Requests for both 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 normal neurological examination who have headaches associated with sinus symptoms.
  - Sinus CT or MRI is not indicated for the evaluation of headaches or neurological
complaints without a more specific indication pointing to a sinus etiology.

- **References:**
  - *Pediatrics* 2001;108:798-808
  - *ACR Appropriateness Criteria, Sinusitis – child, Updated 2006*
  - Prim Care Clin Office Pract 2007;34:39-58
  - *BMJ* 2007;334:358-61
Parasomnias
- Includes sleep terrors, nightmares, sleep walking, myoclonic jerks during sleep
- These are variants of normal development which can be recognized clinically
- Advanced imaging is not indicated

Bed Wetting
- Advanced imaging is not indicated if the child is otherwise neurologically normal

Insomnia
- Advanced imaging is not indicated

Narcolepsy
- An adult disorder which is very uncommon in children, but can begin in adolescence
- Advanced imaging is generally not indicated

Sleep Apnea
- Both central and obstructive types of sleep apnea occur in children
- Polysomnography should be performed initially
  - **Obstructive sleep apnea**
    - Can be seen in association with anomalies or trauma involving the upper airway
      - Hypertrophy of the adenoids and tonsils is the most common finding
      - Obstructive sleep apnea is especially common in children with Down's syndrome
      - Snoring is not particularly abnormal in children
    - Endoscopic examination of the upper airway should be performed initially
    - Lateral upper airway x-rays or maxillofacial CT (CPT®70486) may be helpful if requested by the specialist evaluating the patient
- **Central sleep apnea**
  - Can arise from a variety of posterior fossa lesions
  - Brain MRI, contrast as requested, can be performed if the clinical picture and polysomnography study suggests central sleep apnea

Restless Legs Syndrome
- This condition can begin in childhood
- Polysomnography is useful
- Advanced imaging is not indicated
2011 PEDIATRIC AND CONGENITAL HEAD IMAGING GUIDELINES

PACHD-24 STROKE

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**PACHD-24~STROKE**

- **PACHD-24.1 General Considerations**
  - Incidence of pediatric stroke is low—about 5 per 100,000 per year
  - Clinical recognition of stroke in infants and very young children can be difficult
  - TIA is very infrequently recognized in children
  - **Neonatal Strokes**
    - 25% of all pediatric strokes occur in newborns
    - Birth trauma, anomalous circulation, congenital heart disease, and maternal cocaine use are some of the etiologies
    - Imaging is the same as in older children (see PACHD-24.2 and PACHD-24.3)
  - **Etiology of Pediatric Strokes**
    - There is a high frequency of embolic stroke in children with congenital heart disease (25% of all pediatric strokes)
    - Other causes include sickle cell disease, coagulation disorders or thrombocytopenia, venous infarcts, congenital anomalies of the cranial circulation, complications of bacterial meningitis, vasculitis, fibromuscular dysplasia, and cervico-cranial arterial dissection
    - About 25% of pediatric strokes have no explained cause
  - 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.

- **PACHD-24.2 Initial Imaging**
  - Noncontrast head CT (CPT®70450) can be performed if requested, but brain MRI, contrast as requested, and MRA of the head and neck (CPT®70544 and CPT®70548) or CTA of the head and neck (CPT®70496 and CPT®70498) are preferred and can be performed even if an initial head CT was done.
• **PACHD-24.3 Subsequent Imaging**
  o Repeat brain MRI, contrast as requested, is indicated if seizures or new neurological findings appear once the acute phase of the stroke is past
  o Surveillance imaging is generally not appropriate except for the following:
    ➢ Children with sickle cell disease or known moyamoya disease from another cause (See **PACHD-24.6 Sickle Cell Disease** and **PACHD-24.5 Moyamoya Disease**)
    ➢ Progressive vasculitis—brain MRI without and with contrast (CPT® 70553) can be performed every few months or as course and treatment dictate (preference of the specialist following the patient)
    ➢ Veno-occlusive disease—brain MRI, contrast as requested, and/or MRV (CPT® 70544) can be performed at 3 months and one year

• **PACHD-24.4 Kawasaki Syndrome**
  o Kawasaki syndrome (mucocutaneous lymph node syndrome) can cause aseptic meningitis and occasionally pediatric strokes
  o Most patients with this disease are under age 12
  o Coronary aneurysms are the most feared complications (See CD-8.6 Other Indications for CCTA in the adult Cardiac Imaging Guidelines)

• **PACHD-24.5 Moyamoya Disease**
  o Progressive stenosis of the supraclinoid carotids of unclear cause
  o In adults, this condition often presents with small, otherwise unexplained deep cerebral hemorrhages, but in children, strokes and mental decline are the more typical presentation.
  o Causes or predisposing conditions include sickle cell anemia, β-thalassemia, hemoglobin SC disease, tuberous sclerosis, type 1 neurofibromatosis, and irradiation of the skull base
  o **Initial imaging**
    ➢ Brain MRI, contrast as requested, and MRA of the head (CPT® 70544) or MRA of the head and neck (CPT® 70544 and CPT® 70548)
  o **Surveillance imaging**
    ➢ Head MRA (CPT® 70544) once a year
    ➢ Noncontrast brain MRI (CPT® 70551) can also be performed once a year if requested

• **PACHD-24.6 Sickle Cell Disease**
  o Stroke is common and is an indication for active treatment. Half of these strokes are asymptomatic, so surveillance imaging is needed to find them.
  o These children may develop progressive moyamoya disease
  o Many patients with S-Thalassemia and some patients with S-C disease require the same management as SS patients.
  o Sickle cell carriers (SA) are not at risk for these complications and do not require imaging.
  o **Imaging**
    ➢ Noncontrast brain MRI (CPT® 70551) and brain MRA (CPT® 70544) can be performed once a year for surveillance imaging
    ➢ Many centers follow the cerebral circulation of children with sickle cell disease
(SS) with transcranial Doppler (CPT® 93886 for complete study, CPT® 93888 for limited study, CPT® 93890 for vasoreactive study). Positive findings are further evaluated with brain MRI/MRA.

- **PACHD-24.7 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 CCTA** in the adult Cardiac Imaging 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).
  - **References:**
    - *J Child Neurol* 2005;20:194-197
    - *Practical Neurology* 2002;2:80-93
    - *Practical Neurology* 2002;2:80-93
PACHD-25~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.
- **Imaging:** Brain MRI without and with contrast (CPT®70553). Head CT without contrast (CPT®70450) can be useful in children greater than 2 years old to detect calcifications, which appear after that age and are cortical, not meningeal.
- Cortical resections can be used to treat intractable seizures, and brain PET (CPT®78606) can be useful for preoperative mapping.
- This condition is not familial and familial screening is not indicated
- **Reference:**

PACHD-26~SYNCOPE

- Syncope and lightheadedness are uncommon in children, but are seen in adolescents, mostly girls
- If the neurological examination is normal, advanced imaging is not indicated
- Syncope provoked by exercise should lead to cardiac evaluation for conditions such as short QT syndrome and aberrant coronary artery (see CD-11 Syncope in the adult Cardiac Imaging Guidelines)
  - Syncopal episodes from cardiac disease can be provoked by emotional stress

PACHD-27~TEMPOROMANDIBULAR JOINT (TMJ) IMAGING IN CHILDREN

- TMJ MRI (CPT®70336) can be useful in detecting silent TMJ arthritis in children with juvenile rheumatoid arthritis (JRA), juvenile idiopathic arthritis (JIA), or psoriatic arthritis.*
  - There are generally no indications for advanced neuroimaging in these conditions
  - There is a paucity of clinical symptoms and poor sensitivity of conventional x-rays in diagnosing TMJ arthritis in this population
- **References:**
  - *Current Opinion in Rheumatology* 2006;18:490-495
  - *AJR* 2007;188:182-186
- However, state Medicaid policy and other payer coverage policies will take precedence over MedSolutions’ guidelines.
  - **Alabama Medicaid Agency Policy** (19-01) states the following:
    - PA Policy Number: 19-01 Updated: October 5, 2009
MRIs will only be covered when a medical condition or injury is present which has caused or is suspected to have caused the TMJ as evidenced by:

A. Progress notes that demonstrate the medical necessity of the procedure with documentation that **one or more** of the following conditions exist:
   - Progress notes indicating significant pain i.e. affects daily activities; or
   - Progress notes indicating that function i.e. eating or speaking has been impeded; or
   - Progress notes which demonstrate traumatic injury; or
   - Documentation of prior TMJ surgery with continued pain and symptoms.

   AND

B. Each of the following is present:
   - Copy of x-ray report obtained in the last 3 months; and
   - There is evidence that other forms of treatments (such as splints and devices) have not worked (not necessary if acute trauma present).

   OR

C. Diagnosis of Juvenile Idiopathic Arthritis
   - Limitations:
     - MRIs to diagnosis TMJ will only be covered after other treatment options have proven unsuccessful (except with diagnosis of Juvenile Idiopathic Arthritis.)

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**PACHD-28~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-29.1 Head Trauma**
  o In patients with recent head trauma, a history focused on the accident itself and careful examination of the head, neck, and neurological function should be performed prior to considering advanced imaging.
  o Noncontrast head CT (CPT®70450) is the primary advanced imaging study in patients with acute head trauma.
    ➢ CT of the facial structures (CPT®70486 or CPT®70480) or cervical spine CT without contrast (CPT®72125) may also be needed if there has been associated injury to those structures.
    ➢ **Brain MRI without contrast (CPT®70551)** can be useful as a secondary test to evaluate the following:
      ▪ Children with an abnormal neurological exam that is not explained by the CT findings.
      ▪ Children suspected of being the victims of repeated assaults (battered child)
        ❖ Under age 2, brain MRI without contrast (CPT®70551) is superior in confirming the presence of lesions of varying ages in these children.
        ❖ History can be used in older children to determine the timeline of injuries, so head CT without contrast (CPT®70450) is usually sufficient.
  o Currently there is no well-validated pediatric version of the Canadian or New Orleans Head CT Rule to aid in deciding which children seen after recent head trauma would benefit from head CT.
    ➢ It is generally unnecessary to obtain advanced imaging if there is no history of concussion, no clinical sign of skull fracture, no period of abnormal mental status, no very large scalp hematoma, no persistent symptoms, and no focal deficits on neurological examination.
  o During the six months following a head injury, a repeat noncontrast head CT (CPT®70450) or brain MRI (CPT®70551) can be performed if the child develops fixed or fluctuating diminished mental acuity or alertness, or new abnormalities on neurological examination.
  o **Follow-up of known or treated subdural or epidural hematoma:**
    ➢ No general guidelines can be given for this situation. Imaging should be dictated by the neurosurgeon or neurologist who has treated and is following the patient.

• **PACHD-29.2 Facial Trauma**
  o CT without contrast is the preferred imaging study in facial trauma.
  o **Coding of Facial imaging:**
    ➢ **Maxillofacial versus orbital/temporal bone CT:** both orbital/facial bone CT (CPT® 70480, and CPT®70482) and maxillofacial CT (CPT®70486, and CPT®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.

- Maxillofacial CT (CPT®70486) is the usual study (except in obvious 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.

**References:**
- CMAJ 2010;182:341-348
- Pediatrics 2009;124:e145-e154

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**PACHD-30~TUBEROUS SCLEROSIS**

- A genetic disorder characterized by epilepsy, variable degrees of mental retardation, and skin lesions.
- Transmitted as an autosomal dominant with prevalence of ~1 per 7500.
- Autism and pervasive developmental disorder are common in children with this disorder
- Tuberous sclerosis is a common cause of infantile spasms (a severe epilepsy occurring during the first year of life) and other types of epilepsy
- Adenoma sebaceum (angiofibroma), the characteristic facial cutaneous lesion of tuberous sclerosis, appears between ages 1 to 5 years old. De-pigmented areas of skin are common

**Initial Imaging**
- Tuborous sclerosis is a clinical diagnosis, but advanced imaging is appropriate to confirm the diagnosis. Brain MRI without and with contrast (CPT®70553) is preferred, but noncontrast head CT (CPT®70450) can also be performed in order to identify typical calcifications.
- Other typical nervous system manifestations of tuberous sclerosis besides the signature finding of subependymal nodules, include tubers (gliotic areas which resemble astrocytomas in appearance but not in behavior), giant cell astrocytomas, and hamartomas
- Extra-cranial neoplasms are common, especially cardiac rhabdomyomas, renal cysts, and various benign tumors
  - Echocardiogram (CPT®93306) and/or CT abdomen with contrast (CPT®74160) can be performed if there is concern for these neoplasms
- Pulmonary complications: females often develop lymphangiomatosis of the lungs
  - High resolution chest CT (CPT®71250) can be performed as a screening study in adult female patients with tuberous sclerosis

**Repeat Imaging**
- Repeat imaging is needed to follow the status of giant cell astrocytomas, and is
also indicated if clinical deterioration occurs. The frequency of repeat imaging should be individualized and based on the preference of the specialist following the patient.

- Periodic surveillance imaging with brain MRI without and with contrast (CPT®70553) can be performed to evaluate for the presence of giant cell astrocytomas. The frequency of surveillance imaging should be based on the preference of the specialist following the patient.

- **Familial Screening**
  - Careful clinical evaluation without imaging is generally sufficient
  - Screening neuroimaging studies have not been shown to be helpful

- **References**
  - *Semin Neurol* 1988;8:83-96

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**PACHD-31~von HIPPEL LINDAU DISEASE**

- Autosomal dominant disorder with a frequency of about 1/50,000
- Rarely manifests itself before the teenage years
- 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. Just over half are cerebellar, but spinal and brainstem hemangioblastomas do occur.
- Hemangiomas in other regions and benign renal and hepatic cysts occur.
- DNA testing can identify family members not at risk, and no screening imaging is needed for those members

- **Screening Studies for Those at Risk:**
  - Abdominal screening by ultrasound (CPT®76700 or CPT®76705) 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.*
  - *von Hippel Lindau syndrome, Online Mendelian Inheritance in Man.*
**PEDIATRIC and CONGENITAL HEAD IMAGING REFERENCES**

**PEDIATRIC AND CONGENITAL HEAD IMAGING GUIDELINE REFERENCES**

**PACHD-1~General Guidelines**

**PACHD-2~ANEURYSM, ARTERIOVENOUS MALFORMATION**

**PACHD-3~ATAXIA**

**PACHD-6~CEREBRAL PALSY**

**PACHD-9~DEVELOPMENTAL DELAY MENTAL RETARDATION**

**PACHD-10~DYSTONIA**

**PACHD-11~EAR**

**PACHD-12~EPILEPSY and OTHER SEIZURE DISORDERS**

**PACHD-14~Headache, Child**

**PACHD-15~Macrocephaly, Microcephaly, Hydrocephaly, And Related Conditions**

**PACHD-16~Magnetic Resonance Spectroscopy**

**PACHD-17~MULTIPLE SCLEROSIS**


**PACHD-18~Neuro-Oncology**


**PACHD-19~OPTIC NEURITIS**


**PACHD-20~Pituitary**


PACHD-21~Pseudotumor Cerebri

PACHD-22~SINUS and FACIAL IMAGING

PACHD-24~Stroke

PACHD-25~Sturge-Weber Syndrome

PACHD-27~TEMPOROMANDIBULAR JOINT (TMJ) IMAGING IN CHILDREN

PACHD-29~TRAUMA - HEAD and FACE


**PACHD-30~TUBEROUS SCLEROSIS**


**PACHD-31~von HIPPEL LIINDEAU DISEASE**
