



NCC Pediatrics Continuity Clinic Curriculum: **Headache** *Faculty Guide*

Goals & Objectives:

To understand the common causes and the assessment of severe headache in children:

- Know the historical “red flags” in the assessment of headache.
- List the treatment modalities used in the treatment of migraine headache.
- Know the indications for imaging studies and referral in a child with headache.

Pre-Meeting Preparation:

- “Pediatric Headache a Review” (*PIR, 2012*) or alternate review article below
- **Prepare for Headache “Show & Tell” (see below)**

Conference Agenda:

- Review Headache Quiz
- Complete Headache Case
- **Headache “Show & Tell”:** Go around table and residents should present one of the following: (1) an interesting HA patient they have been following; (2) a recent HA RCT or other study; or (3) a current event article on a clinical controversy related to HAs.

Post-Conference: Board Review Q&A

*****Patient Handouts*****

[Headache-Migraine](#) (*Barton-Schmitt*)

[SMART Tips for Migraine](#) (*AHS*)

Extra-Credit:

- [The International Classification of Headache Disorders, 3rd Edition](#) (*IHS, 2016*)
- [Headache intake sheet](#) (*Walter Reed Adolescent Clinic*)
- [Headache in Children](#) (*PIR, 2020*)
- [Primary headache epidemiology in children and adolescents . . .](#) (*Journal of Headache and Pain, 2023*)
- [An Approach to Children with Chronic Daily Headache](#) (*Dev Med&Child Neuro, 2006*)
- [Neuro-imaging in Migraine and Chronic Daily Headache](#) (*Headache, 2000*)
- [The Optimal Management of Headaches in Children and Adolescents](#) (*Therapeutic Advances in Neurologic Disorders, 2016*)
- [Complementary, Holistic, and Integrative Medicine: Headaches](#) (*PIR, 2006*)
- [Practice Guideline Update: Pharmacologic Treatment for Pediatric Migraine Prevention](#) (*AAN, 2019*)

Pediatric Headache: A Review

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Dr Blume has disclosed no financial relationships relevant to this article. This commentary does contain a discussion of an unapproved/investigative use of a commercial product/device.

Educational Gap

Headaches are common in children; while most are caused by a benign problem or primary headache disorder, headaches can be a sign of a serious underlying condition. Pediatricians must be aware of the most recent recommendations for evaluating and managing headaches.

Objectives

After reading this article, readers should be able to:

1. Understand the evaluation of a child who has headache.
2. Recognize the diagnostic criteria for pediatric migraine.
3. Recognize "red flags" for elevated intracranial pressure or other underlying conditions in the child who has headache.
4. Discuss treatment strategies for migraine, tension, and chronic headache disorders.

Introduction

Headaches are common in children and adolescents and are a frequent chief complaint in office and emergency department visits. The vast majority of childhood headaches are due to a primary headache disorder, such as migraine, or an acute, relatively benign process, such as viral infection. However, clinicians also need to consider other causes of headaches in children. Even when headaches are benign, they may cause significant dysfunction for the child and family and must be managed appropriately to minimize disability and optimize function. In this review, we discuss the epidemiology of childhood headache, evaluation of the child who has headaches, when to consider secondary headache syndromes, and the diagnosis and management of primary headache disorders such as migraine and tension-type headaches.

Epidemiology of Childhood Headache

Acute and chronic headaches are relatively common in children and adolescents, although estimates of the precise prevalence of headache and migraine vary widely. Depending on the study definition of headache, population involved, and time periods studied, 17% to 90% of children report headaches, with an overall prevalence of 58% reporting some form of headache in the past year. (1)(2) Headaches are slightly more common in young boys than girls (age <7 years), but around the time of puberty, this ratio begins to change. Although the prevalence of headache increases with age in both genders, the prevalence of headache increases much more sharply in girls until it reaches adult levels in late adolescence, when the prevalence of headache is significantly higher in women than in men. In adolescence, 27% of girls and 20% of boys describe frequent or severe headaches, and 8% of girls and 5% of boys have had a migraine in the past year. (1)(3) In adults, over 80% of women and 60% of men have had a headache, and 15% of women and 6% of men report having had a migraine in the past year. (1)(2)(4)(5)

Abbreviations

CSF:	cerebrospinal fluid
ICH:	intracranial hemorrhage
ICP:	intracranial pressure
IIH:	idiopathic intracranial hypertension
NDPH:	new daily persistent headache
NSAID:	nonsteroidal anti-inflammatory drug
SVT:	sinus venous thrombosis
TAC:	trigeminal autonomic cephalgia

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Childhood Headache Patterns

Although the majority of children who have headaches do not seek medical care, severe or recurrent acute headaches and chronic headaches are common causes of office and emergency department visits for families who are concerned about the cause of the child's headache (often worried about a brain tumor) and are looking for ways to prevent or manage the headaches. Although serious secondary causes of headache are not common, it is important to ensure that there is no significant underlying disorder that is causing the headaches.

Headaches can be divided into four basic patterns, the recognition of which facilitates the evaluation and diagnosis of the headache: 1) acute; 2) acute recurrent (or episodic); 3) chronic progressive; and 4) chronic nonprogressive (Fig. (6)). A thorough history and examination are essential to classify and manage the headache appropriately. Acute recurrent and chronic nonprogressive headaches are most likely related to a primary headache disorder, although other secondary causes of headaches should be considered in the appropriate circumstances. Chronic progressive headaches are the most worrisome type of headache and deserve a thorough evaluation, most often including neuroimaging. A single acute headache is most often benign, usually triggered by an underlying primary headache disorder or viral infection; however, other serious disorders can cause acute severe headaches (Table 1). (7)

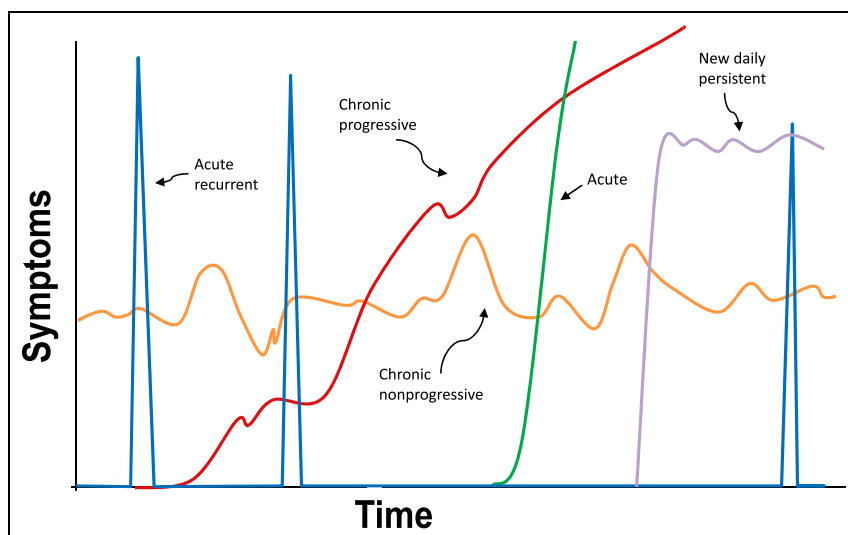


Figure. Headache patterns. (Adapted with permission from Rothner AD. The evaluation of headaches in children and adolescents. *Semin Pediatr Neurol*. 1995;2[2]:109–118.)

Primary Headache Syndromes

Migraine

Migraine is common in childhood, with a prevalence of 1% to 3% in children age 3 to 7 years and 8% to 23% in adolescence, (3) when migraine is much more common in girls than in boys. Migraine is also a common cause of acute headaches that lead to evaluation by a medical professional. The definition of migraine is outlined in Table 2; migraine typically is diagnosed via history and examinations without neuroimaging.

Migraine headaches without aura are more common than those with aura, but both can affect children. It may be difficult to diagnose migraine in very young children because symptoms of vomiting or vertigo may be more prominent than headache. Migraine headache pain may be unilateral or bilateral in children, often is frontal or temporal, and typically is a pounding or pulsing pain. Exclusively occipital pain is unusual and should raise suspicions for another disorder.

Migraine is a primary neurologic disorder. The pathophysiology of migraine is presumed to be the same in children and adults. It is believed that the mechanisms of migraine are based on complex interactions between the neural and vascular systems, including cortical spreading depression, abnormal neuronal excitability, serotonin activity, inflammatory response, and trigeminal neurovascular activation with signal transmission through the thalamus to the cortex. (8) Migraine is no longer believed to be a simple phenomenon of isolated vasoconstriction-triggering pain but a complex cascade of events. We know that mutations in a calcium channel gene (CACNA1A), a sodium/potassium pump (ATP1A2) gene, and a sodium channel (SCN1A) gene and mitochondrial dysfunction all can result in migraine. There are likely to be other genetic differences, yet to be revealed, that can alter neuronal or glial function and lead to the clinical syndrome of migraine. However, migraine is multifactorial; although migraine has a strong genetic component, the heritability pattern is not simple, the clinical manifestations can be different in various family members, and migraine has a very strong environmental component. Although migraines have been described for thousands of years and studied for over 50 years, the

Table 1. Causes of Pediatric Headache

Acute headache
Migraine
Viral respiratory infection, streptococcal pharyngitis
Meningitis/encephalitis
Intracranial hemorrhage
Tumor
Toxic exposures: alcohol, toxins, illicit drugs, medications
Trauma
Stroke
Malignant hypertension
Vasculitis
Episodic recurrent headaches
Tension-type headache
Migraine with or without aura
Fasting/eating disorders
Recurrent toxic exposures: alcohol, toxins, illicit drugs, medications
Recurrent sinus disease
Seizure-associated headache
Mitochondrial disease
Trigeminal autonomic cephalgias
Chronic progressive headaches
Elevated intracranial pressure
Tumor
Vascular malformations
Infection
Sinus venous thrombosis
Idiopathic intracranial hypertension
Endocrine disease: thyroid or parathyroid disease
Chiari malformation
Vasculitis
Chronic nonprogressive headaches
Chronic tension-type headaches
Chronic or transformed migraine
New daily persistent headache
Chronic sinus disease
Dental disease
Sleep apnea
Idiopathic intracranial hypertension
Thyroid disease
Chiari malformation
Fasting/eating disorders
Chronic posttraumatic headache
Chronic trigeminal autonomic cephalgias

exact pathophysiologic mechanisms of migraine remain unknown.

Migraine headaches in children typically are shorter than adult migraine attacks and may last only 30 to 60 minutes. Children often seek a quiet dark place to go during an attack due to phonophobia or photophobia.

Anorexia, nausea, and vomiting also are common during a migraine attack. Sleep often relieves the headache.

Other symptoms that may be associated with a migraine attack include dizziness, blurry vision, difficulty reading, stomach pain, flushing, sweating, pallor, and dark circles around the eyes. Children may have difficulty describing the pain or associated symptoms; asking the child to draw his or her headache can help to define the headache. A family history of migraine or headaches is common, although the pattern of headaches may be different in other family members. Common triggers for a migraine attack include stress, “let-up” from stress, fatigue and poor sleep, illness, fasting, and dehydration. An obvious food trigger is not common but may be a factor for some children who experience migraine.

Migraine With Aura

Most children do not have aura with migraine, and many of those who do have aura sometimes have headaches without aura. Auras usually occur less than 30 minutes before the headache and last only 5 to 20 minutes. A typical visual aura may consist of scotomata, transient blurry vision, zig-zag lines, or scintillations, but more complex visual changes such as those seen in Alice in Wonderland syndrome (visual distortions that include sensation that objects are bigger or smaller than they are; objects appear to be moving when they are still; or objects have shattered like glass) can occur. Other types of aura also occur, including sensory changes (numbness or tingling), confusion, weakness, amnesia, or aphasia. These symptoms are notable, but the onset of symptoms in migraine typically occurs over a longer time period than the symptoms of stroke or seizure. The symptoms of aura are completely reversible, usually last <30 minutes, and often are recurrent over time. Patients who have neurologic symptoms that are prolonged, not related to headache, or not completely reversible should have an evaluation to rule out other underlying conditions.

Other Migraine Headache Syndromes

Less common subtypes of migraine include basilar, confusional, and hemiplegic. In basilar migraine, the aura is characterized by vertigo, ataxia, nystagmus, dysarthria, tinnitus/hyperacusis, bilateral paresthesias, diplopia, or visual disturbance. The aura can be unilateral or bilateral but does not involve motor weakness; the accompanying headache often is occipital.

Confusional migraine is characterized by altered mental status, often accompanied by aphasia or impaired speech and followed by a headache. This state can be triggered by relatively mild head trauma, and the initial

Table 2. **Pediatric Migraine Criteria****Migraine without aura**

- A. At least five attacks fulfilling criteria B–D
- B. Headache attacks lasting 1–72 hours (untreated or unsuccessfully treated)
- C. Headache has at least two of the following characteristics:
 1. Unilateral location, although may be bilateral or frontal (not exclusively occipital) in children
 2. Pulsing quality
 3. Moderate or severe pain intensity
 4. Aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)
- D. During headache at least one of the following:
 1. Nausea and/or vomiting
 2. Photophobia and phonophobia (which may be inferred from behavior)
- E. Not attributed to another disorder

Migraine with aura

- A. At least two attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following:
 1. Fully reversible visual symptoms including positive features (eg, flickering lights, spots, or lines) and/or negative features (ie, loss of vision)
 2. Fully reversible sensory symptoms including positive features (ie, pins and needles) and/or negative features (ie, numbness)
 3. Fully reversible dysphasic speech disturbance
- C. At least two of the following:
 1. Homonymous visual symptoms and/or unilateral sensory symptoms
 2. At least one aura symptom develops gradually over ≥ 5 min and/or different aura symptoms occur in succession over ≥ 5 min
 3. Each symptom lasts ≥ 5 and < 60 min
- D. Headache fulfilling criteria B–D for migraine that begins during aura or follows aura within 60 minutes
- E. Not attributed to another disorder

Adapted with permission from Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders: 2nd edition. *Cephalalgia*. 2004;24(suppl 1):9–160.

episode warrants a complete evaluation to rule out other disorders and intoxication. Hemiplegic migraine is a rare migraine variant that can be familial or sporadic and is characterized by prolonged hemiplegia, numbness, aphasia, and confusion. Genetic testing is available to identify mutations in three genes (CACN1A, ATP1A2, and SCN1A) that have been associated with familial hemiplegic migraine.

Tension Headache

Tension-type headache probably is the most common type of headache in childhood but generally is less disabling than migraine. In contrast to migraine, the pain is mild to moderate, may last for 1 hour or for several days, and often is described as “band-like,” pressure, or tightening. Triggers may be similar to migraine and include stress, fatigue, and illness but also include muscle pain and tension, particularly in the neck and shoulders.

Tension headaches may be episodic (< 15 days per month) or chronic (≥ 15 days per month). A thorough

history to identify stressors, depression, or other factors associated with these headaches is important. Little is known about the pathophysiology of tension-type headaches. Some believe that nociceptive input from cranial/cervical myofascial components triggers these headaches initially, and if this noxious input is sustained, central sensitization can occur, so that an individual becomes more sensitive to these impulses and develops chronic headaches.

Chronic Headache

Chronic daily headache is defined as ≥ 15 headache days per month. There are three major categories of chronic daily headaches in children: chronic migraine, chronic tension-type headaches, and new daily persistent headache (NDPH).

Chronic or transformed migraine is not uncommon in adolescents. These children typically have a history of episodic migraine that becomes more and more frequent until they have more than 15 days per month of headache. Often they have few or no headache-free days. Typically, the serious migraine symptoms that were associated

initially with the headache, such as vomiting and severe head pain or aura, diminish somewhat as the headaches become more frequent, although patients still may have “spikes” of severe head pain at times.

Chronic tension-type headaches may share similarities with chronic migraines once the headaches become daily, and it can be difficult to classify daily headaches as tension or migraine. However, those who have tension-type headaches should not have a history of episodic migraine. Preventive treatment should be considered when the child is having 4 or more days of disabling headache per month. It is also critical to address lifestyle issues such as inadequate or irregular sleep, stress, inadequate or inappropriate food or caffeine intake, inadequate exercise, and poor hydration. Depression and anxiety are common issues for patients who have chronic headaches and may contribute to the headache.

NDPH is characterized by the occurrence of a new headache that becomes daily within 3 days of onset and is not caused by another disorder. Given the abrupt onset of this headache, children who have this type of headache should have an evaluation for secondary disorders. NDPH often is triggered by a viral illness but may be caused by mild head trauma or surgery, or a trigger may be absent.

Other Primary Headache Syndromes

Trigeminal autonomic cephalalgias (TACs) are rare in children. The treatments for migraine or tension headaches may not be effective for TACs, and recognition of these headaches is therefore important. This diagnostic group includes cluster headaches, paroxysmal hemicranias, and SUNCT (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing). These paroxysmal headaches typically are accompanied by autonomic symptoms, such as ipsilateral eye redness, tearing, nasal congestion, rhinorrhea, eyelid swelling, forehead or facial sweating, miosis, or ptosis. Another rare headache that is important to recognize is primary stabbing headache. These patients have stabbing pain in the first division of the trigeminal nerve (orbit, temple, and parietal area) that lasts for a few seconds and recurs in an irregular pattern. Given that secondary causes of TACs and stabbing headaches have been reported, children who have these symptoms should undergo neuroimaging, although most will have normal scan results.

Secondary Headaches

Abnormal Intracranial Pressure

Elevated intracranial pressure (ICP) is an uncommon but important cause of pediatric headaches and has various causes. Hydrocephalus may result from a space-occupying

lesion, blockage of cerebrospinal fluid (CSF) flow via aqueductal stenosis, or impaired CSF absorption. Increasing the volume of tissue or fluids in the cranial vault (eg, mass lesions, edema, inflammation, hemorrhage) also can lead to a dramatic increase in ICP.

Headaches are the most common presenting symptom of elevated ICP. Typically, these headaches are progressive, may cause nighttime waking, and are worse with the Valsalva maneuver or exertion. Children who have elevated ICP often experience persistent vomiting, neurologic deficits, lethargy, or personality change. Other signs of elevated ICP include papilledema and palsies of the third, fourth, or sixth cranial nerves, resulting in eye movement or pupillary abnormalities.

Low ICP also can cause headaches. Intracranial hypotension should be considered if there is a risk for CSF leak (eg, spinal surgery, trauma, connective tissue disease). Meningeal enhancement on brain magnetic resonance imaging may be seen with intracranial hypotension.

Idiopathic Intracranial Hypertension

Idiopathic intracranial hypertension (IIH), sometimes called pseudotumor cerebri, is elevated ICP without evidence of a specific cause. Daily headache is the most common symptom of IIH and may be associated with nausea and vomiting and other migrainous features, but the headache often is poorly characterized. Classic symptoms of IIH include transient obscuration of vision, tinnitus, and diplopia due to cranial nerve dysfunction. In young children, the most common complaints are headache, stiff neck, strabismus, irritability, apathy, somnolence, dizziness, and ataxia.

Children are more likely to have an underlying condition associated with IIH than adults (Table 3); a thorough evaluation in addition to accurate assessment of opening pressure is thus essential when IIH is suspected in a child. The patterns of IIH in adolescence are similar to adult patterns; more female patients than male patients are affected, and obesity is associated with IIH. However, in younger children, when IIH is less common, the genders are affected equally, and obesity is not strongly associated with IIH.

Infection

Acute viral illness with fever is the most common cause of pediatric headache evaluated in the emergency department. (9) Typically, these children will have an acute onset of headache, and the headache resolves as the other viral symptoms dissipate. Many other systemic infections can be associated with headache, including streptococcal pharyngitis, sepsis, Lyme disease, *Bartonella* infection,

Table 3. Conditions Associated With Intracranial Hypertension

Cerebral venous sinus thrombosis
Medications
Thyroid replacement
Corticosteroids (particularly withdrawal)
Growth hormone
Levothyroxine
Cytarabine, cyclosporine
Lithium
Levonorgestrel
Sulfa antimicrobials
Tetracycline antibiotics (minocycline, doxycycline)
Vitamin A
Cis-retinoic acid
Lyme disease
Anemia
Antiphospholipid antibody syndrome
Occult craniosynostosis
Sarcoidosis
Sleep apnea
Systemic lupus erythematosus

rickettsial diseases, and human immunodeficiency virus infection, but headache rarely is the only symptom. Viral infection, particularly with Epstein-Barr virus, can be associated with the onset of NDPH.

Although sinusitis may cause or trigger headaches in some children, the majority of patients diagnosed as having “sinus headaches” have some form of primary headache syndrome. Sinus-related pain generally is pressure-like and dull periorbital pain, worse in the morning, associated with nasal congestion, and lasts for days at a time. It is not associated with nausea, visual changes, phonophobia, or photophobia.

Meningitis or encephalitis are the causes of acute headaches in 2% to 9% of children evaluated for headache in the emergency department. (9) Headaches due to meningitis or encephalitis often are associated with photophobia, nausea, vomiting, and pain with eye movements. These patients typically also have symptoms such as fever, altered mental status, and nuchal rigidity, although fungal meningitis may be more indolent.

Structural Disorders

Although families and patients worry that a brain tumor is causing the headaches, tumors are uncommon causes of headache in children. Tumors and other space-occupying lesions, such as large arachnoid cysts or vascular malformations, can cause headache via hydrocephalus, mass

effect, or hemorrhage. One should consider a space-occupying lesion if the child has “red flag” symptoms noted in the following discussion (Table 4) or if the child has a history of exposure to ionizing radiation or a syndrome (such as tuberous sclerosis or neurofibromatosis) that is associated with tumors.

Chiari I malformation, characterized by the herniation of the cerebellar tonsils >5 mm below the foramen magnum, may cause headache. The classic symptoms include occipital headaches, cough headaches or syncope, sensory disturbance, weakness, ataxia, vertigo, or other cranial nerve dysfunction. Confirming that headaches are due to a Chiari malformation can be challenging because more than 30% of patients with Chiari I malformation on magnetic resonance imaging are asymptomatic, radiologic findings often do not correlate with clinical symptoms, and other causes of headache not related to Chiari malformations are common.

Vascular Disorders

Spontaneous intracranial hemorrhage (ICH) and ischemic stroke are rare causes of headache in children. Although an acute “thunderclap” headache is the classic presenting symptom of ICH, most children who have ICH or ischemic stroke have additional signs or symptoms by the time they present to medical care. ICH should be considered in patients who have an acute onset of severe headache, particularly if the patient has an

Table 4. Red Flags for Secondary Headache

- Progressive pattern of the headache: becoming more severe and/or more frequent
- Increased headache with straining, coughing, or sneezing
- Explosive or sudden onset of severe headache (<6 m duration)
- Systemic symptoms: fever, weight loss, rash, and joint pain
- Secondary risk factors: immunosuppression, hypercoagulable state, neurocutaneous disorder, cancer, genetic disorder, and rheumatologic disorder
- Neurologic symptoms or signs: altered mental status, papilledema, abnormal eye movements, or other abnormalities or asymmetries on neurologic examination
- New or different severe headache, change in attack frequency, severity, or clinical features
- Sleep-related headache, headache waking the patient from sleep, or headache always present in the morning

abnormal result on neurologic examination or a disorder that places him or her at risk for hemorrhage.

In ischemic stroke, neurologic symptoms come on abruptly and persist, typically do not progress from one side of the body to the other, and typically are not recurrent. In contrast, the symptoms of migraine aura usually last less than 30 minutes, may involve both sides of the body or progress from one side of the body to the other, and often are recurrent over months to years.

Sinus venous thrombosis (SVT) is another uncommon cause of secondary headaches in children. The most common presenting symptoms of SVT in children are headache, focal neurologic signs, seizures, decreased level of consciousness, and papilledema. The vast majority have some risk factor for SVT, including head or neck infection, chronic systemic disease, or other prothrombotic state. Thus, SVT should be considered in patients who have headaches and other neurologic symptoms, particularly in those who have underlying conditions that place them at risk for thrombosis.

Trauma

Children who have severe or progressive headache or altered mental status after head injury should be evaluated emergently. Headaches after a head injury may be due to traumatic ICH or fracture but more commonly are due to posttraumatic headache *without* significant structural injury. One study concluded that children older than age 2 years who have normal mental status, no signs of skull fracture, no loss of consciousness, no vomiting, nonsevere mechanism of injury, and nonsevere headache do not need a computed tomography scan after head trauma. (10)

Posttraumatic headaches develop within 1 week of head trauma, concussion, or whiplash. These headaches may have qualities of migraine or tension headaches and often are associated with other postconcussive symptoms, including sleep disturbance, balance abnormalities, cognitive changes, and mood changes. The vast majority of posttraumatic headaches resolve within 2 weeks.

Children and teenagers who have posttraumatic headache (indeed, all who have sustained a concussion) should not return to sports or vigorous exercise until they are symptom-free at rest and while active and have been cleared by a trained medical provider. Once symptom-free, they should return to their regular activities in a step-wise fashion, as outlined in the 2009 Zurich Consensus Statement on Concussion in Sport. (11) Adolescents who have chronic posttraumatic headaches may benefit from returning to low-level “subthreshold” exercise (aerobic exercise that does not trigger worsening of symptoms), with supervision by a trained medical provider. (12)

Substances That Can Cause Headache

There are many substances that can cause headaches, including overuse or withdrawal of caffeine, alcohol use, illicit drug use, carbon monoxide poisoning, and lead toxicity. Medications also can trigger headaches due to the medication’s primary mechanism of action, an idiosyncratic response to the medication, or medication withdrawal. Overuse of medications to treat headaches, especially analgesics, caffeine, opioids, ergotamines, and 5-hydroxytryptamine 1 (5-HT₁) receptor agonists (ie, the triptans), is associated with transformation from episodic to chronic headaches. Some medications that have been associated with headache are listed in Table 5. (13)

Systemic Disease and Headache

Metabolic Derangements and Endocrine Disorders

Fasting is a relatively common cause of headaches in children. Eating disorders also can trigger headaches but may be concealed by the patient. Hypothyroidism can cause headache and should be considered when evaluating for refractory headaches. Hypercapnia and

Table 5. Medications Associated With Headaches

Angiotensin-converting enzyme inhibitors
 α - and β -adrenergic agonists and blockers
 Amphetamines
 Antiarrhythmics
 Calcium channel blockers
 Methylxanthines
 Nitrates
 Phosphodiesterase inhibitors
 Sympathomimetics
 Caffeine
 Ergotamine
 Estrogen
 Opioids
 Acid blockers: including famotidine and ranitidine
 Antimicrobials: amoxicillin, metronidazole, sulfamethoxazole, trimethoprim, ciprofloxacin, gentamicin, nitrofurantoin, ofloxacin, tetracyclines
 Immunoglobulin
 Amiodarone
 Corticosteroids
 Oral contraceptives
 Thyroid hormone replacement
 Vitamin A and retinoic acid

Adapted with permission from Ferrari A, Spaccapelo L, Gallesi D, Sternieri E. Focus on headache as an adverse reaction to drugs. *J Headache Pain*. 2009;10(4):235–239.

hypoxia occur together in sleep apnea or hypopnea due to neuromuscular disease and are associated with headache. A sleep study may be indicated in a patient who has morning headaches and symptoms suggestive of sleep apnea.

Epilepsy

Peri-ictal headaches are common in children who have epilepsy. Typically, the association between the headache and seizure will be obvious. However, some seizures are characterized by episodes of altered mental status or visual disturbances followed by headache, which may be confused with migraine. The visual hallucinations in epilepsy typically are colored and rounded objects, rather than the jagged or scintillating impressions seen in migraine aura.

Mitochondrial Disease

Headache and migraine are frequent findings in mitochondrial disease. However, children who have significant mitochondrial disease typically also have problems with other organ systems and additional neurologic symptoms.

Dental Disease

Temporomandibular disorders have been associated with migraine, tension-type headache, and chronic daily headache. Malocclusion of the jaw and other dental problems also can cause headache.

Sickle Cell Disease

Children who have sickle cell disease are at risk for multiple serious causes of headache, including ischemic stroke, intracerebral hemorrhage, thrombosis, and chronic anemia. However, they also may have migraine or tension headaches, and young children who have sickle cell disease are more likely to have idiopathic headaches than age-matched controls. (14)

Hypertension

Acute, severe headaches may be the initial sign of a hypertensive crisis. However, mild to moderate hypertension typically does not cause significant headaches.

Rheumatologic Disease

Children who have rheumatologic disease often have headache. The causes of headache in rheumatologic disease can include aseptic meningitis, intracranial hypertension, SVT, vasculitis, intracerebral hemorrhage, ischemia, or headache without underlying pathology. Immunosuppressive agents and nonsteroidal anti-inflammatory drugs (NSAIDs) used to treat rheumatologic disorders also can cause headaches. Thus, children who have

diagnosed or suspected rheumatologic disease and headache should have a thorough evaluation for secondary causes of headache.

Psychiatric Disease

Although depression and anxiety disorders are common in patients who have headaches, psychiatric disease usually is an exacerbating rather than a causative factor. Screening for depression or anxiety is beneficial because successful treatment of the headache will be difficult if psychiatric issues remain unaddressed. Headaches should be attributed to somatization disorder, psychotic disorder, major depressive disorder, or anxiety if those symptoms are prominent and if the headaches remit with treatment of the psychiatric disorder.

Evaluation of Headache in Children

History

The patient's history is the single most important factor in the evaluation of headache. Both the patient and the parents should be included in this discussion to gather a complete picture of the patient and his or her symptoms. The history should include characterization of the headache:

- How many different types of headache does the child have?
- When did the headaches begin? Was anything associated with headache onset?
- Are the headaches getting worse, staying the same, or improving? Are they getting more or less frequent? Are they more or less intense?
- Are there any triggers for the headaches?
- What are the headaches like? Where is the pain located? What does the pain feel like?
- Does the headache wake the child from sleep?
- Are there any headache patterns or triggers?
- Are there any other symptoms associated with the headache, or warning signs that a headache is coming (aura)?
- What does the child do during a headache?
- How long does the headache last?
- What makes the headache better or worse? (6)

Adapted from Rothner AD. The evaluation of headaches in children and adolescents. *Semin Pediatr Neurol.* 1995;2(2):109–118.

It is important also to obtain a detailed medical history because headaches may be associated with systemic illnesses and medications. For example, one would have a higher suspicion for secondary headache in a patient who has neurofibromatosis who is at risk for central

nervous system tumors, or in a patient who started minocycline just before headache onset and is at risk for intracranial hypertension. Review of other symptoms related or seemingly unrelated to headache is important because they may suggest systemic disease as a secondary cause of the headache (eg, a malar rash suggestive of systemic lupus erythematosus, cold intolerance and skin changes suggestive of hypothyroidism, a history of episodic torticollis or vertigo as a young child consistent with migraine precursors).

FAMILY HISTORY. Information regarding any family history of headaches, pain, and other medical disorders is important because migraine has a strong genetic component and other disorders related to headache may also run in the family. It is important to ask about family history of *any* type of headaches because other forms may be mentioned by the family when “migraine” is denied. It is helpful to get information about headaches in grandparents, siblings, aunts, and uncles because migraines may skip the parent (often the father) and manifest in the child (often the daughter), and the headache pattern often is different in different family members.

SOCIAL HISTORY. Discussion of social history also is critical because stressors at home, at school, or with friends can trigger or exacerbate headaches. A private conversation with an adolescent often is helpful because the teenager may not wish to discuss some issues in a parent’s presence. This dialogue might include questions about conflict with friends or family, sexual activity, pregnancy, drugs, cutting behavior, physical or sexual abuse, bullying, depression, family finances, alcohol use, or eating disorders, all of which may affect headache.

Lifestyle Factors

Lifestyle factors often affect a headache pattern; it is therefore important to ask about sleep, diet, exercise, caffeine intake, and other activities. A headache diary can help to identify headache triggers or patterns. Inadequate sleep, poor hydration, and poor food choices are common, particularly in teenagers, and these factors often exacerbate or trigger headaches. Children and teenagers who have chronic headaches frequently do not get enough exercise, and regular appropriate exercise is an essential part of a headache management plan (Table 6). Caffeine intake more than 2 to 3 days per week may be a cause of rebound or medication overuse headaches. Clinicians should be aware of the presence of caffeine in soda and energy drinks.

Physical Examination

The evaluation should include measurement of heart rate, blood pressure, weight, and height in the context of a thorough physical examination looking for signs of systemic disease or focal findings that could be related to headaches. This procedure should include palpation of the face, neck, and shoulders, looking for nuchal rigidity, muscular or bony tenderness, trigger points, or allodynia (abnormal pain sensation with light touch, often associated with migraine); skin examination looking for signs of systemic disease, cutting behavior, or neurocutaneous syndromes; and oral evaluation looking for signs of dental disease.

A thorough neurological examination is essential to look for abnormalities in mental status, vision, eye movements, speech, sensation, strength, reflexes, gait, or coordination, particularly noting any focal abnormalities, significant asymmetries, or cranial nerve palsies. Funduscopic examination looking for evidence of papilledema, optic atrophy, or other abnormalities must be included in the examination.

“Red Flags” and Neuroimaging

Several risk factors are associated with an intracranial space-occupying lesion in children who have headache, including sleep-related headache, absence of family history of migraine, headache <6 months’ duration, confusion, abnormal neurologic findings, lack of visual aura symptoms, and vomiting. Children who have more risk factors have a higher risk of having a brain lesion requiring surgery. (15) Other worrisome symptoms include headache associated with cough, urination, or defecation; recurrent and focal headache; exclusively occipital headache; change in headache type; and progressive increase in headache frequency or severity. Table 4 lists factors associated with serious secondary headache.

Table 6. **SMART Headache Management**

Sleep	Regular and sufficient sleep
Meals	Regular and sufficient meals, including breakfast and good hydration
Activity	Regular (but not excessive) aerobic exercise
Relaxation	Relaxation, stress reduction, and management
Trigger avoidance	Avoid triggers such as stress, sleep deprivation, or other identified triggers

One should consider neuroimaging to rule out a structural intracranial lesion if the child has symptoms noted here. Although there are no guidelines regarding the risks of serious secondary causes of headaches in very young children (less than age 3-5), because of the possibility of secondary headaches, neuroimaging should be considered thoughtfully in these young children who have significant recurrent headaches.

Brain magnetic resonance imaging is the modality of choice to investigate potential structural abnormalities, infection, inflammation, and ischemia; however, computed tomography scan is preferred if there is a concern for hemorrhage or fracture.

The American Academy of Neurology practice parameter regarding the role of neuroimaging in the evaluation of children who have recurrent headaches states:

1. Obtaining a neuroimaging study on a routine basis is not indicated in children who have recurrent headaches and normal results on neurologic examination.
2. Neuroimaging should be considered in children who have abnormal results on neurologic examination, the coexistence of seizures, or both.
3. Neuroimaging should be considered in children in whom there are historical features to suggest the recent onset of severe headache, change in the type of headache, or if there are associated features that suggest neurologic dysfunction. (16)

Other Testing

Further testing for children who have headaches should be considered if there is clinical suspicion of an underlying disorder such as meningitis, thyroid disease, or other systemic disease. If there is a suspicion for elevated ICP and the neuroimaging results are normal, lumbar puncture with measurement of opening pressure and measurement of CSF indices is appropriate.

Management of Primary Headache Syndromes

Once a diagnosis of migraine or tension headache has been established and serious secondary causes of headache have been excluded, headache education and management can begin. Education of the patient and family about primary headaches, reassurance that there is no serious underlying disorder, and unification of the patient, family, and provider on a treatment strategy and goal are essential. Successful management of the headaches will be difficult if this foundation is not in place. It is helpful for the patient to keep a headache diary to identify headache patterns and triggers and to evaluate treatment response. The

four major domains of headache treatment include: 1) lifestyle modification; 2) acute headache management; 3) complementary treatments; and 4) preventive treatment. Many children will require only recognition and modification of headache triggers and instruction about appropriate acute headache management, but those who have chronic headaches often require a multifaceted treatment plan.

Lifestyle Changes and Stressors

Any lifestyle factors that could be triggering or exacerbating headaches must be addressed and modified if possible. This modification might include getting adequate

Table 7. Acute Treatment for Childhood Migraine

Drug	Dose
Acetaminophen	10–12.5 mg/kg q 4–6 h Adult: 650–1,000 mg q 6 h Maximum: <4,000 mg/d
Ibuprofen	10 mg/kg q 4–6 h prn Adult: 400–800 mg q 6 h Maximum: 3,000 mg/d
Naproxen sodium	5–7 mg/kg q 8–12 h prn Adult: 250–500 mg q 8 h Maximum: 1,250 mg/d
5-HT ₁ agonists, triptans Rizatriptan ^a	Adult: 5–10 mg may repeat once in 2 h ODT or tablets Maximum: 15 mg/d
Zolmitriptan ^b	Oral (tablet or ODT) or nasal Adult 2.5–5 mg per dose; may repeat once in 2 h Maximum: 10 mg/d
Sumatriptan ^b	Oral: 25–100 mg, maximum 200 mg/d Nasal: • 4–6 y: 5 mg • 7–11 y: 10 mg • >12 y: 20 mg ^c • Adult maximum: 40 mg/d SC: 0.06 mg/kg, >12 y: 6 mg SC, Adult maximum: 12 mg/d SC
Almotriptan ^d	6.25–12.5 mg; may repeat dose once in 2 h Maximum: 25 mg/d

Maximum=maximum dose; ODT=oral disintegrating tablet; prn=as needed; qXh=every X hours; SC=subcutaneous.
^aApproved for treatment of migraine in children 6- to 17-years-old.
^bNot approved for pediatric use.
^cStrong supporting efficacy and safety data in adolescents.
^dApproved for use in children age 12 to 17 years.

and regular sleep and removing electronics from the bedroom at night; eating regular and nutritious meals (including breakfast); limiting caffeine intake to no more than 2 to 3 servings per week; maintaining adequate fluid intake; avoiding or managing stressors; and performing regular aerobic exercise (Table 6). For those with anxiety, depression, or significant stressors, management also should include consultation with a mental health provider to help manage these symptoms because these issues *must* be addressed in conjunction with treating the headaches. Without simultaneous management of depression, anxiety, or stressors, reasonable headache control is likely to be extremely difficult.

Acute Treatments

The most important factor in the acute treatment of migraine or tension headache is early intervention for a significant headache because early treatment is most likely to be effective. However, patients should not use acute treatments more than 2 to 3 days per week (<15 days per month for NSAIDs and <10 days per month for triptans or caffeine) to avoid developing medication overuse headaches.

In children, over-the-counter medications such as ibuprofen, naproxen, and acetaminophen often are effective for the management of migraine (Table 7). The correct dose for the child's weight should be reviewed with the parent because underdosing may result in treatment failure and overdosing can be harmful.

For more severe headaches, a dose of an NSAID can be combined with caffeine (such as a limited amount of soda, tea, or coffee) less than 9 days per month, as long the child is not consuming caffeine on a regular basis. If the child does use caffeine regularly, it should be withdrawn carefully, because caffeine withdrawal can cause rebound headaches.

If NSAIDs are not effective, one may consider using triptans in appropriate circumstances. Although many different triptans are available for use in adults, only rizatriptan has been approved for migraine treatment in children age 6-17 and almotriptan has been approved for use in adolescents. This situation is due primarily to difficulty in study design (ie, overcoming the high placebo response in children) rather than current safety concerns.

There are data to support the use of sumatriptan (subcutaneous and nasal) and rizatriptan in children age >6 years, and zolmitriptan (oral

and nasal) in children age >12 years (Table 7). (17) Opiates and barbiturates are not indicated for the treatment of primary pediatric headache disorders. Opiates may alter the pain response, increasing the risk of chronification of pain, and both compounds can lead to overuse headaches and addiction.

Antinausea medications can be useful adjuncts to the NSAIDs and triptans when patients have significant nausea and vomiting with their migraines. Prochlorperazine and metoclopramide are effective in the acute emergency department setting and may also be helpful in the outpatient setting. Although these medications do pose a risk of dystonic reaction, these dopamine antagonists may help treat the underlying migraine as well as the nausea and vomiting. Ondansetron also has been used as an antiemetic in migraine and has a lower risk of adverse effects than some other antiemetic agents, although it is also more expensive (Table 8).

Complementary Medicine

Complementary therapies can be very helpful in the management of chronic or recurrent episodic headache. Bio-behavioral techniques that may be successful include biofeedback therapy, relaxation techniques, hypnosis, acupuncture, and training in the use of coping mechanisms. Appropriate physical therapy or massage therapy also can be very helpful in some circumstances, particularly if the child has muscle pain or tension.

Preventive Therapy

Most headache specialists agree that daily therapy designed to prevent migraines should be considered when a patient experiences ≥ 4 days of disabling headaches per month. Many families prefer to try supplements or "nutraceuticals" before using prescription medications.

Table 8. **Antiemetic Agents for Pediatric Migraine**

Drug	Dose	Toxicity
Promethazine	0.25–0.5 mg/kg/dose Adult: 12.5–25 mg/dose	Sedation Dystonic reactions ^a
Prochlorperazine	2.5–5 mg bid prn Adult: 5–10 mg q 6–8 h prn Maximum daily dose: 40 mg/d po Adult rectal dose: 25 mg	Sedation Dystonic reactions
Ondansetron	4–8 mg q 8 h • <15 kg: 0.2 mg/kg • 15–30 kg: 4 mg • >30 kg: 4–8 mg	Sedation Dystonic Reactions

prn=as needed; po=oral; q 8 h=every 8 h.
^aOculogyric crisis (managed with intravenous diphenhydramine).

These options may be as effective as some prescription medications and generally have fewer adverse effects. There is also a high placebo response rate in children who have headaches. Before beginning any preventive therapy, it is important to discuss expectations with the family because any preventive therapy typically takes at least 8 to 12 weeks to cause a recognizable effect. Switching preventive treatments every few weeks due to

presumed ineffectiveness without an adequate trial will be unproductive and frustrating for all involved. A headache diary will help to assess the efficacy of the treatment over several months.

Supplements/Nutraceuticals

Although there are few randomized controlled trials of supplements for the management of headaches in

Table 9. Selected Preventive Medications for Pediatric Migraine

Drug	Dose	Toxicity
Cyproheptadine	0.25–1.5 mg/kg per day Adult: 4–20 mg/d tid	Sedation, dry mouth Weight gain
Tricyclic antidepressants		
Amitriptyline	10–50 mg qhs 0.1–1 mg/kg per day Maximum: 50–100 mg for headache	Sedation Weight gain May exacerbate cardiac conduction defects (consider baseline electrocardiogram)
Nortriptyline	10–75 mg qhs	Suicidal thinking, mood changes
Antiepileptics		
Topiramate	1–2 mg/kg per day for headache Typical adult dose: 50 mg bid Maximum: 800 mg bid for seizures	Sedation, paresthesias, appetite suppression/weight loss, glaucoma, kidney stones cognitive changes, word finding difficulty, mood changes, depression
Valproic acid	20–40 mg/kg per day; adult: 500–1,000 mg/d	Weight gain, bruising, hair loss, hepatotoxicity, ovarian cysts, teratogenic, thrombocytopenia, leukopenia, mood changes, depression
Gabapentin	10–40 mg/kg per day Adult: 1,800–2,400 mg/d Maximum: 3,600 mg/d	Fatigue, ataxia, tinnitus, gastrointestinal complaints, mood changes, depression
Antihypertensives		
Propranolol ^a	2–4 mg/kg per day Adult: 160–240 mg/d	Hypotension Sleep disorder Decreased stamina Depression
Verapamil	4–10 mg/kg per day divided tid <12 y: ≤120 mg 13–18 y: 240 mg	Hypotension, nausea, atrioventricular block Weight gain
Selected supplements used for headache prevention		
Riboflavin/vitamin B ₂	50–400 mg	Yellow urine (25 mg may be effective; studies done using 400 mg/d)
Melatonin	1–6 mg before bed	Vivid or disturbing dreams
Magnesium oxide	9 mg/kg per day tid	Diarrhea Modify dose with renal dysfunction
Coenzyme Q10	100 mg/d	Rash, irritability Gastrointestinal symptoms
Migralief or Children's Migralief (B ₂ /magnesium/feverfew)	1 capsule 1–2 times/d	Yellow urine, diarrhea

bid=twice a day; Maximum=maximum dose; qhs=at bedtime; tid=3 times a day.

^aAvoid when patient has asthma or diabetes; use caution when patient has depression.

Adapted from Lewis DW. Headaches in children and adolescents. *Curr Probl Pediatr Adolesc Health Care*. 2007;37(6):207–246.

children, there are several options that may be effective and are unlikely to cause harm. Riboflavin (vitamin B₂) seems to be effective for the treatment of adult migraine, using doses from 25 to 400 mg per day. (18)(19) Magnesium also has been used for the management of migraine. One pediatric study found that children who took magnesium oxide had a significant decrease in headache frequency over time, whereas those given placebo did not. (20) However, the most useful form of magnesium is not clear; other forms of magnesium may be more bioavailable or less likely to cause diarrhea than magnesium oxide, but these forms have not been studied in migraine. The typical adolescent dose of elemental magnesium is 350 to 500 mg per day (Table 9).

Coenzyme Q10 also may be helpful for the management of migraine and has few troublesome adverse effects. Butterbur extract (*Petasites hybridus*) also has been used for the management of migraine, but this plant naturally contains alkaloids that can be hepatotoxic or carcinogenic if not processed correctly; using a reputable source that is “PA free” is thus essential. There are reports of the use of melatonin to treat chronic daily headaches in teenagers and help manage sleep disruption; melatonin may therefore be particularly beneficial for teenagers with headache who have insomnia (Table 9).

Prescription Preventive Medications

There are many options for preventive therapy for migraine, but relatively few have been studied in rigorous, randomized controlled studies in children. In 2004, the American Academy of Neurology practice parameter concluded that flunarizine (a calcium channel blocker not available in the United States) is probably effective, but that there were conflicting or insufficient data to make recommendations for the use of other medications for the management of pediatric migraine. (21)

Since then, several studies have found that topiramate is significantly more effective than placebo for the management of pediatric migraine. (22)(23)(24) Other medications that are used typically for the prevention of pediatric migraine include β -blockers such as propranolol and tricyclic antidepressants such as amitriptyline and nortriptyline. Valproic acid also is used to treat migraines; however, this agent is teratogenic and may have other toxic effects and is therefore not the best first-line treatment for teenage girls. A nightly dose of cyproheptadine may be particularly useful in younger children or those who have environmental allergies and migraine. Other medications that have been used for migraine headache prevention include gabapentin and verapamil (Table 9). (25)

Prescription preventive medications should be started at a low dose and gradually increased to the goal or effective dose to minimize the risk of adverse effects. The choice of prescription preventive headache medication often is based on comorbid factors. For example, amitriptyline may be a reasonable choice for a teenager who is active and has difficulty sleeping at night but would not be a good choice for a child who is sleeping well and fatigued; topiramate may be a good option for an overweight adolescent but would not be a good choice for a thin 12-year-old who has a family history of kidney stones; and propranolol and low-impact aerobic exercise may be a good option for a child who has symptoms of postural orthostatic tachycardia syndrome but is not a good choice for an asthmatic child or competitive athlete.

Treatment of Unusual Headache Syndromes

There are few studies of the therapy of NDPH, but treatment strategies include typical migraine preventive medications (eg, topiramate, tricyclic antidepressants) and complementary therapies such as biofeedback, psychotherapy, physical therapy, massage, and education regarding coping skills. Some have suggested using acne doses of minocycline or doxycycline due to an elevation in the CSF of tumor necrosis factor- α found in a study of adults who have NDPH. These medications may lower levels of this cytokine in CSF. (26) Some TACs are responsive to indomethacin. Primary stabbing headaches often respond to indomethacin.

Summary

- Although most pediatric headaches are due to benign illness or a primary headache syndrome, clinicians must be able to recognize disorders that can cause secondary headache. Failure to identify and treat the underlying cause of a child's headache, whether it is due to migraine, viral illness, or serious systemic disease, can lead to morbidity and prolonged suffering.
- The most important factors in the evaluation and management of a child who has headaches are:
 1. A comprehensive history detailing headache characteristics and any disorders, symptoms, or exposures that may be associated with the headache. Social and family history is important.
 2. A complete physical and neurologic examination with attention to signs or symptoms that could be associated with a secondary cause of headache.
 3. Further testing if indicated by the history and examination findings.
 4. Creation of a multifaceted treatment plan with the child and family that is appropriate for the patient's

headache and is endorsed by the patient, family, and medical provider.

Use of these techniques in combination with a good understanding of primary and secondary headache syndromes will help to provide optimal care for children who have headaches.

- Headaches are common in childhood. Obtaining a neuroimaging study on a routine basis is not indicated in children who have recurrent headaches and normal results on neurologic examination (American Academy of Neurology 2002 Practice Parameter).
- Secondary causes of headache should be considered in children who have abnormal neurologic findings, who have had recent onset of severe headache, or who experience a new severe headache and in those with systemic disease or medication use that increases the risk of abnormal intracranial pressure, hemorrhage, thrombosis, intracranial infection, or structural lesion (according to the American Academy of Neurology 2002 Practice Parameter and strong research evidence).
- Treatment of primary episodic headaches, such as migraine or tension-type headache, should include assessment and modification of problematic lifestyle factors and stressors and appropriate acute headache treatment. Chronic or recurrent headache treatment may also include complementary therapies or preventive daily medications (according to expert opinion and some research evidence).

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Headache Quiz

1. Name the “**red flag**” symptoms of headache.

Box 1 Red flag features leading to serious consideration of further investigation

- ▶ Headache worse in recumbency or with coughing and straining.
- ▶ Headache waking up child (note the difference between headache present on awakening which is not uncommon in migraine).*
- ▶ Associated confusion, and/or morning or persistent nausea or vomiting.
- ▶ Recent change in personality, behaviour or educational performance.
- ▶ Physical signs to include field defect, short stature or cranial bruit; cerebellar signs and signs of raised intracranial pressure.

*Many people with a migraine wake and then feel the gradual onset of head pain as opposed to being woken by severe pain which could well represent a space-occupying lesion.

Arch Dis Child Educ Pract Ed 2008;93:105–111.

2. Using the broadest definition of headache, what percent of children overall will ever report headaches? **90%** (A 1993 study from Stang and Osterhaus listed migraine in the "top five" of all pediatric diseases.)

3. Define **chronic daily headaches**. What are the three major categories of chronic daily headache? **HA present for >15 days/month. Chronic migraine, chronic tension-type headaches, and new daily persistent head-ache (NDPH.)**

4. Match the migraine subtype with its description:

- | | |
|-----------------------------|---|
| confusional C | A. numbness,aphasia,confusion, genetic test available |
| hemiplegic A | B. aura with vertigo, ataxia, speech/vision/hearing disturbance |
| basilar B | C. aura with altered mental status, impaired speech |

5. Fill in the following Migraine Management Table, starting with 1st-line medication options:

Abortive Treatment	Prophylactic Treatment: Indicated when HA freq > 4/month
1. Motrin, TLN, ± Phenergan	1. Periactin (< 6 yrs)
2. Oral Triptans (tablet or dissolvable)	2. Propranolol (older children)
3. Triptan nasal spray (>5 yrs)	3. AEDs (VPA, Topamax)
	4. Amitriptyline (? Depression)

6. As a general rule, at what point should migraine prophylaxis be considered?
Greater than or equal to 4 disabling headaches per month. One might consider nutritional supplements such as riboflavin (B2) or magnesium before offering medications.

Headache Mega-Case

Lucy is a healthy 16 yo female who presents as a walk-in to the Adolescent Clinic with complaint of a "HORRIBLE headache" that started this morning. She describes the headache as sharp, stabbing pain behind her eyes. Since waking up this morning, it has gotten progressively worse. She says that she was unable to eat breakfast this morning due to nausea. She took 200mg of Motrin before school with minimal relief. She called her Mom to pick her up after lunch because she was unable to concentrate in calculus class. She has had similar headaches in the past. On ROS, Lucy admits to nasal drainage for the past 4-5 days and occasional chills. She denies fever, emesis, focal weakness or loss of consciousness.

What general category/pattern of headaches do Lucy's symptoms fall under?

Hard to tell from this initial history . . . Likely **acute and recurrent**, but would need more history to determine whether possibly chronic non-progressive or chronic progressive. Also need more H&P to determine **primary vs. secondary**.

List at least 3 diagnoses to consider in this patient based on the history:

- Migraines
- Tension headache
- Headache associated with a viral illness
- Sinusitis

Less likely causes: space-occupying lesion, refractive error, post-traumatic HA, CNS infection

What additional history do you want to obtain from Lucy?

- Description of prior HA—frequency, severity, association with menses?
- Family history of migraines? (found in **80%** of cases)
- Associated symptoms – nausea, vomiting, photophobia/phonophobia, relieving factors, vision changes (blurry vision, double visions, scotomas), dizziness.
- Stressors in her life?
- Does she wear glasses?
- Red flag symptoms – Did headache wake her from sleep? Are sxs different from prior ones?
- Any recent fevers? Recent travel?
- Any problems with blood pressure in the past?
- HEADSS exam—any drug use?

Lucy's headaches occur "maybe every other week?" since starting her period at 15. She endorses photophobia/phonophobia with her headaches. Generally, she has mild nausea with the headaches and occasionally experiences vomiting. Typically, these headaches last 4-5 hours and resolve with sleep. Lucy's mother has a history of similar headaches for which she treats with "some sort of injection". Lucy's HEADSS exam is unremarkable; although, she provides only brief answers to your questions due to her discomfort.

Her physical exam is as follows:

VS: T 100.5, HR 98, BP 118/77, RR 20, SaO₂ 100%

On exam, she is wearing dark sunglasses – when she removes the sunglasses, you notice dark circles under her eyes. Neuro: PERRLA, EOMI, CN 2-12 intact, normal cerebellar function and DTR's are also normal. No cranial bruits appreciated. VA 20/20 OU.

What is the most likely diagnosis and why? Does she have chronic daily headache?

- **Migraine headaches.** She likely fits “migraine without aura” (at least 5 attacks; lasts 4-72 hrs; characterized by severe pain causing avoidance of physical activity; associated with N/V/photo/phonophobia).
- **Not CDH**—although >3 months of symptoms, not >15 days/month.

Is the location of Lucy's headaches unusual, given your most likely diagnosis? No— the typical unilateral pattern in adults is often not seen in children. Commonly bilateral in young children. Unilateral in 60-70% of young adults and adolescents, and bifrontal or global in 30%.

Is this the end of your evaluation? Why or why not? Yes . . .

- No lab testing is needed to support the diagnosis of migraines.
- Neuroimaging is not needed given none of the following: abnormal neuro exam; recent onset of severe HA; change in type of HA; suspicion of meningitis or other intracranial process; underlying disease that predisposes to intracranial pathology)

What *non-pharmacologic* measures could you suggest to Lucy to manage her symptoms?

- Manage expectations – Educate your patient that the frequency and severity of headaches may decrease over weeks to months of therapy, but the headaches may continue.
- Return to school, if possible.
- Headache diary –helps identify any specific triggers of headaches. *Table 6. SMART Management*
- Identify headache triggers and avoid if possible – inadequate hydration; poor sleep (too much, too little, napping); stress, **certain foods** (e.g. **caffeine, chocolate, aged cheese, alcohol, meats with nitrates, foods with MSG, ice cream**)
- Daily exercise for 20-30 minutes.
- Biofeedback therapy, guided imagery, acupuncture, hypnosis, massage, counseling, etc.

What *pharmacologic* measures will you recommend *today*? What are possible side effects?

- Abortive therapy with **Motrin**—increase initial dose to 400mg and instruct to take q4h with max daily dose of 1200mg. *Do not take more than 2 days/week for risk of medication-overuse headache.*
- Add **anti-emetic** (Phenergan, Reglan, Compazine), given vomiting history.
- If truly refractory to Motrin (or Tylenol), start with PO **Sumatriptan**. Start with 25mg. (*UpToDate* says repeat in 20min; *Harriet Lane* says repeat in 2hr to max 200mg/day).

Does Lucy need long-term therapy for her headaches? If so, what would you recommend?

This is up for discussion. . . According to UpToDate: “Prophylactic agents may be necessary for children with headaches > 4x/mo or headaches that adversely affect the child’s activities.” One could argue that Lucy’s migraines adversely affect her activities. However, it may be prudent to do 2-4 wk trial of adequate abortive therapy before starting prophylaxis.

If you decide to proceed with long-term therapy, what is your duration of treatment?

This is also up for discussion . . . The approach presented in the UpToDate article is 6-12 months of prophylaxis, then tapered over several weeks.

When would you refer and to whom?

- Consider referral to Peds Neuro if HA is refractory to above abortive and prophylactic therapy options. *After referral, however, it is important to “recapture” patient and help with implementation of Neurology plan.*
- Consider referral to Behavioral Health or Adolescent Medicine *IF* mood or anxiety symptoms become more prominent as treatment progresses.

How would your diagnosis change with the following history?

- a) Her temperature is 102.9 and she appears confused. **Consider CNS infection** (e.g. encephalitis, encephalopathy, meningitis).
- b) Her headaches are worse in the morning; she has recently developed problems with peripheral vision. **Consider space-occupying lesion.**
- c) She has unilateral pain with lacrimation on one side; the headaches appear at approximately the same time every day. **Consider Cluster headache.**
- d) She has a 3-day history of fevers ranging 100.2-101.8 and a history of seasonal allergies. **Consider sinusitis.**

Headache Board Review

1. 1. A 17-year-old girl presents with complaints of recurring headaches that are becoming more frequent. She was diagnosed with migraine headaches 2 years ago. Previously she had an average of one migraine per month, but she now has one per week. She has no double vision, vomiting, or awakening from sleep with headaches. Headaches can occur at any time of day, are throbbing and bifrontal, and are associated with nausea and phonophobia. They are relieved by nonsteroidal anti-inflammatory drugs and by lying down in the dark. They rarely last more than 4 hours. The adolescent and her mother are concerned that the increased headache frequency could be due to a brain tumor or aneurysm.

Of the following, the MOST appropriate next step is:

- A. brain magnetic resonance angiography to rule out aneurysm
- B. brain magnetic resonance imaging to rule out brain tumor
- C. head computed tomography scan with contrast to rule out brain tumor
- D. lumbar puncture with manometry to rule out elevated intracranial pressure
- E. perform no diagnostic testing at this time.**

When evaluating a child who has a headache, the first decision point is whether to perform medical diagnostic testing, particularly neuroimaging such as head computed tomography (CT) scan or brain magnetic resonance imaging (MRI). This decision is based on an assessment of the likelihood of a primary headache (eg, migraine, tension, cluster) or a secondary headache (due to disease). Primary headaches are painful paroxysmal events occurring in isolation or, more commonly, as part of a chronic, recurring pain disorder. No underlying lesion is causing the pain. Most children, adolescents, and adults presenting to the primary care physician with headache, including the girl described in the vignette, have primary headaches, and no medical diagnostic testing is needed as part of validated standard of care.

A family history of aneurysms may increase the risk for aneurysms (and anxiety), but it is not an indication for neuroimaging testing for aneurysms in a patient younger than age 30 years. However, because cigarette smoking increases the risk for aneurysmal rupture, this is a good opportunity to remind families about the dangers of smoking.

Most brain tumors are sporadic primary tumors or metastatic, not familial, and, therefore, a family history of brain tumors is not an indication for neuroimaging in most cases. In the absence of symptoms and signs of elevated intracranial pressure or meningitis, lumbar puncture is not needed.

Imaging studies (CT scan, MRI, and occasionally magnetic resonance angiography), sometimes followed by lumbar puncture with manometry, are indicated when there is suspicion of intracranial disease (eg, tumor, aneurysm) or other medical processes, particularly processes that increase intracranial pressure. Symptoms that may herald the presence of secondary headache include pain awakening from sleep, pain relieved by vomiting, and pain increased by the Valsalva maneuver. Additional symptoms such as new binocular double vision, any focal numbness or weakness, or loss of coordination should prompt an investigation. Secondary headaches are more likely in the presence of abnormal signs such as papilledema, acquired (new) ocular malalignment with double vision, stiff neck, limb weakness, or ataxia.

2. A 16-year-old girl who is new to your practice complains of a nearly constant headache for the past year. She describes the pain as a band around her head that often is throbbing and is worse during the middle of the day. She denies nausea or vomiting but reports occasional fatigue. There is no family history of headaches. She has missed more than 20 days of school this year because of the headache, and she is struggling to maintain a C average. She admits to hating school and does not participate in

extracurricular activities because she "doesn't like anything." Findings on her physical examination, including complete neurologic and fundoscopic evaluation, are normal.

Of the following, the BEST next step in the management of this girl's headaches is to

- A. advise her to keep a headache diary and return in 2 months
- B. obtain a lumbar puncture
- C. obtain computed tomography scan of the brain
- D. prescribe oral sumatriptan

E. refer her for psychosocial evaluation and counseling

Chronic headache is a common complaint in children and adolescents. A careful history and complete neurologic examination are indicated in the evaluation of headache. The most important initial consideration is to identify characteristics of the headache that suggest serious diagnoses, such as brain tumors or other diseases causing increased intracranial pressure.

Such characteristics include worsening pain at night or immediately upon awakening, association with vomiting, and worsening pain with coughing or straining. Papilledema or focal neurologic findings may be found in patients who have increased intracranial pressure or the examination results may be normal. Migraine headaches are periodic, may be accompanied by an aura, and typically are relieved by sleep. A family history of migraines usually can be elicited. The neurologic examination typically yields normal results, although complicated migraines can be accompanied by focal neurologic deficits such as hemiparesis, cranial nerve palsies, and visual disturbances.

Pain from stress-related or tension headache generally is diffuse and may be described as "bandlike" or throbbing, as reported for the girl in the vignette. Pain usually occurs on most days, and school absence is frequent. The neurologic examination yields normal results.

It is also very important for the clinician to obtain further information regarding any emotional, social, or academic difficulties the patient may be experiencing. Such data can help to determine both an underlying cause for the headache and the effect the headache is having on the child's quality of life. Family stressors and depression are known causes of headache, and questions regarding sleep patterns, anhedonia, school performance, and relationships with family and friends can help to screen for these conditions. Studies have shown that children and adolescents who have frequent or severe headaches have greater impairment in academic and social functioning than those who do not have headaches. A thorough psychosocial evaluation can aid in evaluating patients for depression and assessing the effect of headaches on their daily functioning. Nonpharmacologic therapies, such as rest, relaxation techniques, and removal of stress from the environment, can be effective once the stress is identified.

Advising the girl in the vignette to keep a diary for 2 months likely would prolong her difficulties and not address the underlying cause of her headaches. Lumbar puncture and computed tomography scan of the brain may play some role in the evaluation of an acute headache, especially if meningitis or a mass lesion of the brain is suspected, but these diagnoses are very unlikely in a patient who has had a headache for a year and normal results on neurologic examination. Oral sumatriptan is used in the treatment of migraine, but this girl's history points to depression or another psychological cause for her headache, and prescribing medications without a psychosocial evaluation probably would not be effective.

3. A 14-year-old girl who has a 1-year history of migraine headaches presents to the emergency department with a severe headache that she calls "the worst headache of my life." The headache occurred suddenly after she lifted a heavy box. Her mother says that the girl has been holding her head stiffly. On physical examination, she appears in severe pain and has meningismus. Other findings on the physical examination are normal.

Of the following, the MOST appropriate initial course of action is

A. emergent noncontrast head computed tomography scan

- B. intravenous administration of ceftriaxone
- C. intravenous administration of dihydroergotamine
- D. lumbar puncture
- E. oral administration of sumatriptan

Although severe headaches in adolescents are often due to migraine, four key findings from the history provided by the girl described in the vignette indicate that she is experiencing a "symptomatic" headache (ie, headache due to an underlying disease) instead of a migraine headache. These findings are: 1) her description of this headache as "the worst ever," 2) its abrupt onset, 3) the onset during a "Valsalva maneuver" (heavy lifting), and 4) the stiff head/neck position. These features are characteristic of a subarachnoid hemorrhage (SAH), likely a small hemorrhage at this point, because the girl is still conscious.

In contrast, migraine headaches, which are common in children and adolescents, are recurrent headaches often associated with nausea, dizziness, and photo- or phonophobia. They represent an "idiopathic" chronic recurrent headache disorder, meaning that no space-occupying lesions (tumors, vascular malformations) are causing the pain. In "symptomatic" headaches, the pain is a symptom of an underlying disease process.

Head computed tomography (CT) scan is the preferred diagnostic modality for the girl in the vignette because it is fast, noninvasive, and highly sensitive to blood and structural lesions causing pain. Of note, CT findings are normal in patients whose headaches are caused by pseudotumor cerebri.

Although this girl has nuchal rigidity, which occurs in meningitis, she is afebrile, and her symptoms began abruptly while lifting, which should not be related to central nervous system infection. Accordingly, ceftriaxone treatment is not appropriate at this point. Intravenous administration of dihydroergotamine might be appropriate treatment for a severe migraine headache, not for the symptomatic headache described for the girl in the vignette. For the same reason, oral sumatriptan is not the appropriate initial therapy. Although a lumbar puncture eventually may be necessary, it should not be the initial diagnostic test because it is invasive and may be unsafe in the presence of a focal lesion. Because the history suggests SAH and the sensitivity of head CT in diagnosing this condition is at least 95%, CT should be the initial diagnostic test. If the suspicion for SAH is high and results of the CT are normal, lumbar puncture should be obtained to look for red blood cells.

4. A 15-year-old girl presents to the emergency department with a 4-week history of nasal drainage and face pain and a 2-week history of frontal headaches and fatigue. Her mother complains that her daughter has an "attitude" and has not been respectful or seemed to care about anything for the past 2 weeks. The daughter awoke this morning with a headache and vomited. On physical examination, the adolescent is afebrile and has normal vital signs. She responds slowly to questions and is not oriented to the date. She complains of pain to palpation of her cheeks and forehead. She has no nuchal rigidity and no focal weakness. The remainder of the physical examination findings are normal.

Of the following, the BEST initial diagnostic procedure is

A. computed tomography scan of the head with intravenous contrast

- B. emergent electroencephalography to rule out nonconvulsive status epilepticus
- C. lumbar puncture to rule out meningitis
- D. nasal swab for bacterial culture
- E. urine drug screen for barbiturates, amphetamines, and cocaine

The subacute onset of mental status changes described for the adolescent in the vignette warrants an emergency evaluation. In most cases, neuroimaging is indicated, along with appropriate laboratory testing.

The relatively nonspecific pain and what her mother perceives as common emotional problems (apathy in a teenager) probably represent early frontal lobe symptoms. The headache on awakening and vomiting are concerning for increased intracranial pressure (ICP). Confusion and psychomotor retardation on the mental status examination indicate involvement of the central nervous system. A focal, ischemic, ictal, infectious/inflammatory, or toxic/metabolic process must be identified urgently. A brain abscess is suggested by the prominent facial pain in this setting; the sinuses are a common source of brain abscesses. Brain abscesses often present only with nonspecific pain and not with fever.

Head computed tomography (CT) scan is preferred for this patient because the constellation of pain, confusion, and morning vomiting makes a focal intracranial mass a possibility. Increased ICP is associated with morning vomiting because ICP is highest in the morning. Contrast is recommended because of the insidious onset, which could indicate either a neoplasm or infectious process. Intravenous contrast is not needed for all neuroimaging procedures. However, it increases the diagnostic yield of imaging studies where either neoplasm or infection is suspected because both typically involve some degradation of the blood-brain barrier or hypervascularity, resulting in contrast enhancement at the site of the lesion.

Magnetic resonance imaging (MRI) with contrast also is a good choice. The advantage of MRI is higher spatial and soft-tissue resolution. Disadvantages of MRI compared with CT include: 1) less availability for emergency department studies; 2) need for pharmacologic sedation in agitated patients because sedation affects mental status, thereby masking disease-related mental status; 3) longer time in the scanner, which could delay treatment decisions; and 4) cost. Thus, in most cases, a head CT scan with contrast is preferred as the initial study in the emergency department.

Electroencephalography (EEG) is an important test for assessment of a patient who has encephalopathy of unclear cause to rule out nonconvulsive status epilepticus (NCSE), particularly if the patient is known to have epilepsy. If an EEG cannot be obtained rapidly, intravenous administration of 0.1 mg/kg lorazepam can treat NCSE immediately, although this would not clear confusion about other causes. For this adolescent, the facial pain makes the diagnosis of NCSE less likely than a brain abscess.

A lumbar puncture may be needed to rule out meningitis or encephalitis, but the pain and morning vomiting more strongly suggest the possibility of an intracranial mass. Lumbar puncture prior to head CT is not advised in this case because it could reduce pressure below the foramen magnum and result in herniation from the supratentorial mass. Toxicology screening and nasal swabs are reasonable but not the preferred initial diagnostic tests because they will not affect emergency management of the increased intracranial pressure.

5. A 10-year-old boy presents after falling from a second floor window onto the pavement 30 minutes ago. There was a 3-minute loss of consciousness initially, but he has been alert and talking to his mother during the ambulance ride to the hospital. On physical examination, the child has tenderness and a hematoma over the right parietal region of the head. Results of an initial complete neurologic examination are normal, but on subsequent examination 15 minutes later, the boy exhibits marked lethargy and slurred speech.

Of the following, the MOST likely explanation for his current symptoms is

- A. Cerebral contusion
- B. Concussion
- C. Epidural hematoma**
- D. Subarachnoid hematoma
- E. Subdural hematoma

The sequence of events described for the boy in the vignette after sustaining the blow to the head is an initial loss of consciousness, followed by a lucid interval, followed once again by an alteration in the level of consciousness. This “waxing and waning” the level of consciousness is typical of an epidural hematoma. The rapid increase in intracranial pressure that results when the origin of the bleeding is a damaged artery can cause acute deterioration of the patient. Prompt recognition of the injury is critical to providing appropriate medical or surgical intervention.

A concussion is a head injury that causes at least temporary neurologic dysfunction, often with loss of consciousness of 1 minute or less. These injuries may be associated with abnormal findings on computed tomography (CT) of the head, but often the findings are normal. A cerebral contusion represents direct injury to the brain parenchyma and can be seen with CT. A subdural hematoma results from bleeding from bridging veins or dura and can lead to increased intracranial pressure, as seen with epidural hematomas. Deterioration generally is not as rapid as with an epidural hematoma and may occur hours to days after the initial injury. Subarachnoid hemorrhage is common with severe brain injury and leads to blood in the cerebrospinal fluid. This type of bleeding can cause vasospasm and ischemic brain injury. All of these injuries can be associated with an initial loss of consciousness and later deterioration, but the rapid progression of the symptoms in the patient in the vignette makes these alternative diagnoses less likely.

6. A 13-year-old girl has had 5 days of unremitting headache that is relieved by vomiting but not sleep and 1 day of double vision. She was previously healthy and has no history of migraine headaches, but she was treated for otitis media 6 weeks ago. Findings on physical examination are normal except for some tenderness over her left mastoid. On neurologic examination, you note normal mental status and normally reactive pupils and vision but florid papilledema. She cannot abduct her right eye fully and has subjective double vision with both eyes open looking to the right but not looking to the left or with either eye covered. Facial sensation and movements are normally symmetric, and the rest of the findings are normal. Head computed tomography scan yields normal results.

Of the following, the diagnostic test or procedure that is MOST likely to be helpful is:

- A. brain magnetic resonance imaging
- B. cerebral angiography
- C. lumbar puncture with manometry**
- D. ocular nerve sheath fenestration

The girl described in the vignette has unremitting head pain that improves transiently after vomiting but not sleep. In addition, she has a new symptom of double vision when looking to the right, not left, an acquired ocular malalignment consistent with a right 6th nerve palsy. The lack of facial weakness or facial sensory loss suggests that this symptom comes from outside of the brainstem. Funduscopic examination reveals bilateral papilledema. This complex of symptoms is characteristic of raised intracranial pressure due to pseudotumor cerebri. Diagnosis and treatment are needed to reduce pain and prevent visual loss.

An urgent head computed tomography (CT) scan in the emergency department showed normal findings. This rules out hydrocephalus or a mass lesion but does not rule out elevated intracranial pressure. The

most helpful procedure to determine this finding is a lumbar puncture with manometry. Manometry pressure measurement should be obtained with the child in the lateral decubitus position, with legs relaxed. A pressure greater than 20 cm H₂O confirms the clinical impression of elevated intracranial pressure and relieves the pressure. The pressure often remains low for several days because cerebrospinal fluid drains into the soft tissues around the lumbar puncture site. Cerebrospinal fluid studies should include assessment of protein, glucose, and cell count. Bacterial, fungal, and Mycobacterium tuberculosis studies as well as cytology for malignancy may be indicated in some cases.

The differential diagnosis of pseudotumor cerebri includes complications of obesity; adverse effects of medications such as minocycline, isotretinoin, or steroids; hypervitaminosis A; venous sinus thrombosis; anemia; renal failure; and hypercalcemia.

The recent history of otitis media and current pain over the mastoid described for the girl are consistent with mastoiditis. The cause for her pseudotumor was venous sinus thrombosis. This may be apparent on head CT scan, but magnetic resonance imaging (MRI) with contrast and with venography is more sensitive for detection than head CT or routine MRI.

Cerebral angiography typically is used to characterize arterial vascular pathology. However, this invasive procedure is undertaken infrequently because of improvements in magnetic resonance angiography. Ocular nerve sheath fenestration is performed occasionally as a longer term treatment for refractory pseudotumor cerebri to reduce pressure on the eyes and preserve vision. Serum vitamin A measurement is reasonable after lumbar puncture as part of the evaluation for causes of intracranial hypertension.

7. A 14-year-old boy who was diagnosed with migraines at age 11 presents to the emergency department with a severe migraine. For the past 2 months, he has had two to three such headaches per week. His mother asks about using stronger pain medications. You are concerned about the possible complications of medication overuse.

Of the following, the class of abortive medications MOST likely to induce chronic headaches is:

- A. caffeine-containing medications
- B. isometheptene compounds
- C. nonsteroidal anti-inflammatory drugs
- D. opiates**
- E. triptans

Opiates and barbiturates are more likely than caffeine-containing medications, isometheptene compounds, nonsteroidal anti-inflammatory drugs (NSAIDs), or triptans to cause chronic headaches due to overuse of medications. Medication overuse headache is the third most common type of chronic headache after migraine and tension headache. Most often, it results from NSAID use because NSAIDs are the most widely used agents for headaches. However, the probability of medication overuse headache is higher with opiates.

Medication withdrawal headache is a huge public health problem and a factor involved in chronic daily headache for many adolescents. The only effective treatment is withdrawal of the medication. Although less common than with opiates, medication overuse headache may occur with over-the-counter analgesics, triptans, ergotamines, and combination analgesics with caffeine.

Other well-known complications of medications used to treat headache include constipation with opiates, tolerance with benzodiazepines, and gastrointestinal distress with NSAIDs.