

Updated overview of pediatric headache and migraine

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ABSTRACT

Headache is a common complaint, occurring in >90% of school age children. The frequency increases with increasing age and the etiologies range from tension to life-threatening infections and brain tumors. Migraine is the most frequent cause of acute and recurrent headaches in children. The overall prevalence of non-migraine headaches is 10-25%. A thorough history, physical and neurological examination, and appropriate diagnostic testing (if indicated) will enable the physician to distinguish migraine and tension headaches from those of a secondary etiology. In this review, we present an updated overview of childhood headaches. The recently developed International Classification of Headache Disorders, second edition (ICHD-II) will be summarized. The Quality Standards Subcommittee of the American Academy of Neurology (AAN) and the Practice Committee of Child Neurology Society (CNS) recommendations for neuroimaging of children with recurrent headaches concluded that routine neuroimaging is not indicated if the neurological examination is normal. Neuroimaging should be considered in children with recent onset of severe headache, change in the headache type, associated focal neurological features, or seizures. Trends in the management guidelines will be highlighted.

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Headache is a common complaint, occurring in the majority of school age children.¹ The frequency increases with increasing age and the etiologies range from tension to life-threatening infections and brain tumors. Migraine is the most frequent cause of acute and recurrent headaches in children.^{2,3} Children are usually brought to medical attention to exclude serious causes, such as brain tumors or meningitis. A thorough

history, physical and neurological examination, and appropriate diagnostic testing (if indicated) will enable the physician to distinguish tension and migraine headaches from those of a secondary etiology.¹ Children with recurrent headaches are more likely to have certain migraine related phenomena, such as motion sickness, teeth grinding (bruxism), sleep talking and walking (somnambulism), and syncope.^{4,5} Most patients with migraine have type A personality, making them worry allot, perfectionist, and high achievers. This may provide further risks and potential triggers to the recurrent headaches.

Epidemiology. More than 90% of school age children suffer from headaches. Overall, the prevalence of non-migraine headaches is 10-25%.⁶ In the acute setting (emergency room), many children with acute headache have a viral illness or an upper respiratory infection (such as sinusitis, otitis media). In the outpatient department, psychosocial (such as family or school problems) and infectious etiologies are common. Tension-type headache (TTH) and migraine are the 2 most common types in children and adolescents.⁶ Migraine constituted up to 75% of referrals for childhood headache in one series.² The prevalence of migraine headaches is 4-5% among 3-11 year old children with an increasing prevalence with age.⁷ The female-to-male ratio also changes with age. The prevalence is greater in boys before age 7 years, is equivalent in boys and girls between ages 7 and 11 years, and is greater in adult women than in men (ratio 3:1).

Pathophysiology. Muscles attached to the skull are the possible source of pain in TTH, however, the pathophysiology is largely unknown.⁶ The smaller genetic effect on TTH than on migraine suggests that the 2 disorders are distinct. However, many believe that TTH and migraine represent the same pathophysiological spectrum.^{6,8} The old belief that the migraine aura is due to cerebral vasoconstriction and the headache is due to vasodilatation, has been abandoned. Recent evidence suggests spreading cortical depression during the aura and neurogenic inflammation, due to increased cellular permeability,

causing the pulsatile headache. Note that the brain itself is painless. Vascular instability in the pain sensitive structures (dura, larger vessels, roots, and nerves) is responsible for the headache while the aura is a cortical phenomenon. Increased intracranial pressure causes pain mainly by the traction and displacement of intracranial arteries.

Clinical evaluation. Headache may not be apparent to parents of younger children who may present with excessive crying, colic, rocking, or hiding.⁹ Chronic headache may result in developmental regression, social withdrawal, and behavior problems. Older children are better able to perceive, localize, and remember pain. Emotional, behavioral, and personality factors become more important as the child enters adolescence. The history of headache provides most of the necessary diagnostic information in the evaluation of childhood headaches (Table 1). A thorough history helps to focus the physical examination and prevent unnecessary investigations and neuroimaging.¹ The history of headache should be obtained from both parents and the child, who may add valuable diagnostic information (such as visual migraine aura). The following history details should be obtained including age of onset, precipitating and relieving factors, aura, time and mode of onset, location, frequency, severity (effect on activity and school attendance), quality (throbbing or pulsatile in migraine), duration, associated symptoms (photophobia, phonophobia, nausea, vomiting), effect of physical activity (migraine increases with exercise), and response to treatment.¹⁰ A diary, in which the quality, location, severity, timing, precipitating factors, and associated features of the headache are recorded prospectively, is a useful adjunct. A diary is not subject to recall error, may reveal a pattern that is typical for a certain type of headache, and provides important diagnostic information for children who are unable to provide sufficient detail during the clinic visit. In acute headaches, questions concerning symptoms of increased intracranial pressure are particularly important (Table 2). The presence of these features is an indication for further evaluation and/or neuroimaging.^{11,12} The following features are suggestive of raised intracranial pressure including sleep-related or early morning headache (intracranial pressure is higher during lying down), persistently localized headache of recent onset, progressive increase in headache frequency or severity, personality change, persistent vomiting without nausea, and headache worsened by cough, micturition, or defecation. In chronic headaches, it is important to appreciate the potential co-existence of other chronic disorders, such as asthma or diabetes.¹ Headache also can be a side effect of many medications including bronchodilators, stimulants, and some antiepileptic

drugs (namely lamotrigine). Family history of headache or migraine, particularly maternal, is commonly encountered in children with migraine.

The International Classification of Headache Disorders II. The recently developed International Classification of Headache Disorders, second edition (ICHD-II) was the result of many years' work by many headache experts from different countries.¹³ International Classification of Headache Disorders-II criteria are superior to the earlier ICDH-I and International Headache Society (IHS) criteria in identifying definite migraine in children and adolescents.¹⁴⁻¹⁶ These diagnostic criteria are summarized in Table 1. The terms "common" and "classic" migraine are no longer used. Differentiating the first migraine episode from

Table 1 - The International Classification of Headache Disorders II, 2004.¹³

Tension-type headache (TTH)	
A	At least 10 episodes fulfilling B-D criteria Episodic or Chronic (lasting for >15 days)
B	30 minutes to 7 days in duration
C	At least 2 of the following 4 criteria: Non-pulsating (pressing or tightening) Mild to moderate Bilateral Not increased by exercise
D	Both of the following: No nausea or vomiting No photophobia or phonophobia, or one but not both
E	Not attributed to another disorder
Migraine without aura	
A	At least 5 attacks fulfilling B-D criteria
B	4 to 72 hours in duration
C	At least 2 of the following four criteria Unilateral Pulsatile Moderate to severe Increase by exercise
D	At least one of the following 2 criteria: Nausea and/or vomiting Photophobia and phonophobia
E	Not attributed to another disorder
Migraine with aura	
A	At least 2 attacks fulfilling B-D criteria
B	Aura of one of the following criteria, but no motor weakness: Reversible positive or negative visual features Reversible positive or negative sensory features Reversible dysphasic speech disturbance
C	At least 2 of the following criteria: Homonymous visual and/or unilateral sensory symptoms Aura symptoms develops gradually over ≥ 5 min Aura 5 to 60 minutes in duration
D	Migraine without aura begins during the aura or within 60 min
E	Not attributed to another disorder

symptomatic headache (such as caused by infection or neoplasm) may be difficult because the diagnostic criteria would not have been met. Simpler and easier diagnostic criteria were developed earlier by Prensky.¹⁷ It requires recurrent headaches separated by symptom-free intervals and at least 3 of the following 6 symptoms or criteria: 1) abdominal pain, nausea or vomiting, 2) localized unilateral or hemicranial headaches, 3) throbbing, pulsatile quality to the pain, 4) complete relief after a brief period of sleep, 5) visual or sensory aura, and 6) family history of migraine.¹⁷ The Prensky criteria are applicable to children and can be used in the clinic as an initial screen. The ICHD-II is useful for more accurate case definitions and reporting in clinical research.¹⁸ However, some authors suggested that these criteria may be too restrictive to differentiate TTH from migraine without aura in children.⁶

Tension-type headache. Tension-type headache (TTH) is characterized by bilateral pressing or constricting tightness that occurs anywhere on the head (Table 1). The headache is non-throbbing, is of mild to moderate intensity, and lasts from 30 minutes to several days, namely can be prolonged. The patient may have increased tenderness on pericranial manual palpation. Tension-type headache is not accompanied by nausea or vomiting nor aggravated by routine physical activity, which are migraine features. Stress (good or bad) is the usual trigger. Sometimes, the overlap of some of the symptoms of migraine can make the differentiation difficult.^{6,19} As well, the co-occurrence of migraine and TTH is frequent, namely many patients have more than one headache type.⁸

Migraine headache. Migraine is characterized by recurrent episodes of headache that are aggravated by exercise and relieved by sleep (Table 1). Autonomic

symptoms are commonly seen and include photophobia, phonophobia, nausea, and vomiting. Reversible aura can take several forms including visual features that can be positive (for example flickering lights, spots, lines) or negative (vision loss, homonymous field defect), sensory features that also can be positive (for example pins and needles) or negative (numbness), or dysphasic speech disturbance (Table 1). The aura typically develops gradually over 5 minutes and lasts for less than one hour. The headache may occur during or within 60 minutes of the aura symptoms. Occasionally, migraine aura occurs without headache (acephalgic migraine).²⁰ Compared to adults, migraine in children is more likely to be bilateral (bifrontal or bitemporal) with shorter duration.²¹ Migraine in infants and toddlers may be more difficult to recognize, with symptoms of head holding, irritability, vomiting, pallor, and head tilt, all relieved by sleep.⁹ Hemicrania continua is a rare neurological emergency with continuous unilateral headache that usually responds to indomethacin.

Migraine variants. The migraine variants are unique to pediatrics and are a fascinating and challenging group of disorders characterized by several clinical and neurological features.²² Migraine may be complicated by ophthalmoplegia, recurrent vomiting, hemiplegia, ataxia, and confusion. These unusual features are associated with recurrent migraine headaches and family history of migraine. However, all the variants represent diagnoses of exclusion. Therefore, these children should be thoroughly evaluated by careful medical history, physical examination, and appropriate neurodiagnostic studies, to exclude other diagnoses (for example intracranial tumor, hemorrhage, or infection). Often, these unusual features initially lead the clinician in the direction of epilepsy, cerebrovascular, traumatic, or metabolic disorders. Only after detailed evaluation does the diagnosis become apparent.²² Recognition of the typical feature may help in preventing unnecessary investigations and hospital admission of some of these children.

1) Migraine variants with visual features. The visual aura associated with migraine has been discussed earlier in the migraine headache section. Several other visual variants can be associated with migraine including ophthalmoplegic migraine, retinal migraine, and transient post-traumatic cortical blindness.^{23,24} In ophthalmoplegic migraine, ipsilateral oculomotor palsy may last for up to 4 weeks. Retinal migraine is rare in children and manifests by attacks of transient monocular vision loss associated with migraine headaches. Vision loss usually lasts <1 hour; however, irreversible defects may occur with recurrent attacks representing migrainous retinal infarction.²³ Transient cortical blindness is characterized by normal pupillary

Table 2 - Important symptoms and signs that may suggest raised intracranial pressure or a space occupying lesion (Red Flags).

Symptoms
New onset headache
Headache persistent in one site
Headache progressive in severity or frequency
No relief with regular analgesics
Sleep-related or early morning headache
Worsened by cough, micturition, or defecation
Persistent vomiting without nausea
Personality change
Other neurological symptoms (such as seizures, double vision, weakness)
Signs
Macrocephaly
Bulging anterior fontanel
Lethargy
Neck stiffness
Neurocutaneous signs
Papilledema
Focal neurological signs (such as motor, sensory, cerebellar)

response and funduscopy, and follows minor head trauma.²⁴ Associate migraine symptoms occur with vision returning to normal within minutes to hours after the trauma leaving no neurological sequelae.

2) Migraine variants with gastrointestinal features. The so called "abdominal migraine" is no longer recognized as a migraine variant in the latest ICHD-II guidelines. An organic pathology is more likely responsible for symptoms of episodic abdominal pain with nausea and vomiting that are not associated with headaches. Recognized migraine variants with prominent gastrointestinal symptoms include cyclic vomiting, recurrent vomiting following minor head injury, and infant colic. Jan et al²¹ found migraine headaches, motion sickness, and family history of migraine highly predictive of recurrent vomiting after minor head injury. Recognition of this association will reduce extensive investigations and hospitalization of selected children with minor trauma and no focal neurological features.²¹ We also found a strong association between migraine headaches and history of infantile colic.⁹ The pain and crying in some of these genetically predisposed infants could represent a form of migraine with an age specific clinical expression. Favorable response to anti-migraine treatment is also supportive; however, such diagnosis should be made after careful exclusion of organic causes of infant colic, such as milk allergy, otitis media, urinary tract infection, intestinal obstruction, and hypertension.⁹

3) Migraine variants with motor features. Benign paroxysmal torticollis, comprised of recurrent, often short-lived, and spontaneously recovering attacks of head tilt in infants, also has been proposed as a migraine variant. Migraine can also be complicated with hemiplegia (hemiplegic migraine), which can lead erroneously to the diagnosis of stroke.²⁵ The diagnosis is by exclusion. Familial alternating hemiplegia has been also linked to migraine. Such diagnoses should be made only after excluding cerebrovascular disorders.

4) Migraine variants with psychic features. Sudden state of confusion and agitation with migraine characterize acute confusional migraine.²⁶ The episode can follow mild head injury and is more common in boys. Alice in wonderland phenomenon is an interesting syndrome manifesting with recurrent episodes of impaired time sense, body image, and visual analysis of the environment.²⁷ The child would report that objects and people look smaller or larger, closer or farther, and slower or faster than normal. The symptoms occur with a clear state of consciousness and in the absence of any evidence of an encephalitic process, seizures, drug ingestion, or psychiatric illness.²⁷

Physical examination. The physical examination often is normal in children with tension or migraine

headaches. Normal blood pressure should be documented. Measurement of height, weight, and head circumference, auscultation of the head for bruit (a sign of arteriovenous malformation) are important aspects of the examination.²⁸ Detailed neurological examination is needed to exclude focal neurological signs. Lethargy, macrocephaly, focal neurological signs, neck stiffness, neurocutaneous signs, or papilledema should raise the suspicion of a space-occupying lesion (**Table 2**). However, note that most brain tumors in childhood are midline (for example medulloblastoma, cerebellar astrocytoma, and craniopharyngioma) with minimal lateralizing physical findings. Papilledema may be difficult to appreciate in the young or uncooperative child and therefore ophthalmologic assessment is needed.

Laboratory evaluation. Laboratory testing is rarely helpful in the evaluation of childhood headache.¹ Lumbar puncture (LP) is necessary if intracranial infection or pseudotumor cerebri are suspected clinically. Neuroimaging typically is performed before LP because LP is contraindicated in patients with space-occupying lesions. However, in patients with suspected bacterial meningitis, the risks of delaying the LP and administration of antibiotics while awaiting neuroimaging must be considered. Patients in whom pseudotumor cerebri is suspected may require reassurance or sedation before undergoing the lumbar puncture because an accurate opening pressure measurement is crucial for accurate diagnosis. Electroencephalography (EEG) is not useful in evaluating children with headaches unless epilepsy is suspected.²⁹

Neuroimaging. Children who have signs or symptoms of an intracranial process should undergo urgent neuroimaging with computed tomography (CT) or magnetic resonance imaging (MRI) (**Table 2**). These studies may detect a variety of disorders that cause headache, including trauma, infections, neoplasms, or vascular disorders. Computed tomography is typically performed in acute situations in which hemorrhage is suspected or rapid diagnosis of a space-occupying lesion is necessary. Magnetic resonance imaging usually is preferred in other situations, or if there is persistent concern despite a normal CT scan. Magnetic resonance imaging demonstrates sellar lesions, craniocervical junction lesions, white matter abnormalities, and congenital anomalies more accurately than does CT. However, MRI may require heavy sedation and is more expensive and time-consuming than CT. Magnetic resonance venography (MRV) is recommended for children with pseudotumor cerebri to exclude venous sinus abnormalities, including thrombosis. Most patients with migraine headaches do not need neuroimaging; nor do children who have chronic non-progressive

headaches with no signs or symptoms of increased intracranial pressure.¹¹ Studies have documented that all of the children with surgically treatable lesions had abnormal findings on neurological examination, including papilledema, abnormal eye movements, and motor or gait dysfunction.^{11,12} The Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of Child Neurology Society recommendations for neuroimaging of children and adolescents with recurrent headaches unassociated with trauma, fever, or other obvious provocative cause, concluded that routine neuroimaging is not indicated if the neurological examination is normal.¹² Neuroimaging should be considered in children with recent onset of severe headache, change in the headache type, associated focal neurological features, or seizures.¹²

Management. An important step in the management is to exclude serious intracranial pathology and reassure the child and the parents.¹ This by itself may alleviate associated anxieties and result in reduction of the headache severity and frequency. We always advise the older children or the parents of younger children to document in a diary the headache occasions and associated activities and symptoms in the preceding 24 hours. This would help in identifying the triggers and therefore trying to avoid them. Simple analgesics in appropriate doses are usually effective at the beginning of the headache. Typically, acetaminophen (panadol or tylenol) is used at 20 mg/kg/dose. A common pitfall is to use subtherapeutic doses or use the analgesic late, after the symptoms became severe and therefore less responsive to simple analgesics. If not effective, stronger nonsteroidal analgesics (such as Ibuprofen) or abortive therapy (Triptans) can be used. Narcotics should be avoided because of the risk of dependency. Chronic daily headaches (CDH) are often linked to abuse of pain medications.³⁰ Therefore, the physician should enquire about the dose and frequency of pain medications in every patient. Treatment of CDH includes neck physiotherapy for stiffness, withdrawal of the abused drug, and treatment of any withdrawal symptoms.³⁰ Recurrent headaches unresponsive to standard therapy or resulting in significant social limitations or absence from school can be treated prophylactically for 3-6 months. Drugs that can be used ordered in our preference include Cyproheptidine (Periactin), Valproic Acid, Topiramate, or Gabapentin. The parents should understand that these drugs are not analgesics and that they should be taken regularly to decrease headache recurrences. Recently, relaxation exercises (self hypnosis) were found very effective in aborting the headaches in older children and adolescents.³¹

Prognosis. Although the short-term outcome of most children with recurrent headaches appears favorable, few studies have reported long-term outcome.

Recurrent headaches rank third among illness-related causes of school absenteeism.³² Long-term follow-up studies revealed that 30% of the children and adolescents presenting with migraine or TTH become headache free.^{15,19} Another 20-25% shift from migraine to TTH or vice versa. Recently, Brna et al reported the long-term prognosis of a Canadian Cohort 20 years after initial evaluation.³² They also found that most patients (73%) continue to have headache, although the headache classification often changes across time. The child and family needs to understand that migraine is a life long condition and therefore, every effort should be made to prevent its occurrence and negative effects on the quality of their lives. Simply, they need to find the best way that works for them to minimize migraine related morbidity. As our patients grow older, they frequently choose to care for their headaches pharmacologically.

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Related topics

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