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Seminars in PEDIATRIC NEUROLOGY

Approach to the Diagnosis of Pediatric Headache

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Headache in children and adolescents is a common symptom that can be worrisome to patients, their parents and clinicians due to the myriad of underlying etiologies, both benign and life-threatening. The evaluation of headache must be directed primarily to exclude secondary causes. A detailed headache history, recognition of headache patterns and red flags and thorough physical and neurological examinations are essential in the diagnosis; and identifies patients requiring further workup. Considerations for neuroimaging and ancillary testing are also discussed.
Semin Pediatr Neurol 40:100920

Introduction

Headache is a common complaint in a pediatric emergency department (ED) or ambulatory clinic. The spectrum of pediatric headaches is broad, and the underlying etiology can be as simple as a viral illness or as serious as a brain tumor. The clinician's role is to determine primary from secondary causes. In this article, we discuss an approach to pediatric headache diagnosis. A knowledge of headache epidemiology, formulation of semi-structured history taking, recognition of red flags and thorough examination will lead the clinician to the correct diagnosis, appropriate diagnostic testing if necessary and proper therapy.

Epidemiology of Pediatric Headache

The global one year prevalence for headache disorder is up to 85%.[1] The overall prevalence of headache in the pediatric population varies, with one-year prevalence from most epidemiologic studies being at least 30%-40%,[2] while in Austria and Norway, it exceeds 75%.[2,3] Approximately 30% of the pediatric population experiences recurrent headache[2] and the occurrence of headache among children increases with age.[4]

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Among pediatric headache disorders, migraine followed by tension-type headache (TTH) and then by medication overuse headache (MOH) are the most common.[3,5,6] Headache disorders are more common in girls than in boys. For migraine, female sex becomes a risk factor in the post-pubertal years, whereas in the pre-pubertal years, migraine is predominant in boys.[4] Trigeminal autonomic cephalalgias (TACs) such as cluster headache are encountered far less often in pediatric clinical practice.

In contrast, the international incidence of central nervous system (CNS) tumors in childhood is 1.12-5.14 cases per 100,000 persons or about 0.005%.[7] The annual incidence of primary intracranial hypertension (PIH, previously known as idiopathic intracranial hypertension or IIH) is even lower, estimated by pediatric studies to be < 1 per 100,000.[8,9,10] An exhaustive list of the epidemiologic statistics of every cause of secondary pediatric headache is beyond the scope of this article however, are significantly less common than primary headache disorders. While non-life-threatening diseases such as self-limiting infection and minor head trauma are more common culprits,[11] secondary headaches must not be missed.

Headache History

History is the cornerstone in the diagnosis of pediatric headache. A detailed history is essential to achieve the most important goal of distinguishing between a primary and secondary headache disorder.

https://doi.org/10.1016/j.spen.2021.100920
1071-9091/11/

History taking in pediatric patients can be challenging. Some children may be too young to answer questions. Important information may not always be volunteered. It is imperative that headache history must be elicited not given.

The following series of standard questions can provide adequate information to reach a diagnosis and provide a systematic approach to ensure that secondary headaches are not overlooked:

How often and when did your headache start?

This question establishes the headache temporal pattern. Rothner classified pediatric headaches into five temporal patterns: acute, acute recurrent, chronic progressive, chronic non-progressive, mixed pattern.[12] Each of these patterns provide its own set of differential diagnosis that can guide further diagnostic testing (Table 1).

Acute headache pattern represents a single episode or new onset of headache. It is typically associated with systemic or neurologic symptoms. In children, viral illness is a frequent cause of an acute headache. A worst headache following Valsalva maneuver, or a thunderclap headache raises concern for an aneurysmal subarachnoid hemorrhage (SAH).

Acute recurrent headache is a pattern with periods of complete freedom from symptoms between headache attacks. This is the pattern of primary headache disorders such as migraine and TTH. It is also a pattern for TACs and cranial neuralgias which are uncommon among pediatric patients; hence the possibility of underlying CNS lesion must be excluded. When this pattern has been ongoing for at least 6 months and neurological examination is normal, further work-up is not indicated.[13]

Chronic progressive headache is a pattern that gradually increases in frequency and severity over several months. It is worrisome as it indicates the likelihood of increased intracranial pressure (ICP). Other neurologic symptoms and abnormal neurologic examination usually accompany this pattern.

Chronic non-progressive headache is a pattern of frequent intermittent or daily constant headache with normal neurologic examination. Chronic daily headache, a terminology still used by many clinicians, fits into this pattern and is defined as a headache of >15 days per month for >3 months with each headache lasting >4 hours. Psychosocial stressors are commonly present.

Mixed headache pattern refers to a long-standing headache such a chronic migraine with a superimposed acute and/or acute recurrent headache. The superimposed headache can be another primary headache disorder (eg, chronic TTH and migraine) or a secondary headache. The latter implies a change in headache pattern and requires diagnostic evaluation.

Where is the location of your headache?

A brief side-locked headache is characteristic for TACs. Due to its rarity in pediatric patients, pituitary lesions mimicking TAC must be excluded.[14] Occipital headache, specifically when associated with abnormal neurologic examination, is an ominous sign for posterior fossa tumors or lesion in the craniocervical junction. However, a recurrent occipital headache in a child without atypical history and who has a normal neurologic examination is not more likely to be associated with intracranial pathology than headaches in other locations.[15] Localized pain can be seen in otitis media, temporomandibular joint dysfunction, optic neuritis or maxillary sinusitis.

What does your headache feel like?

Migraine is typically described as throbbing or pounding; while pressure or dull are descriptors used for TTH. Sharp, stabbing, jabbing are words used to describe primary stabbing headaches or TACs. Shooting and/or shock-like pain is common in trigeminal neuralgia. For younger children, allowing them to draw their pain provides invaluable information.

How long does your headache last?

Distinguishing headache duration between long-lasting headaches from short-lasting headaches (seconds to minutes) can distinguish migraine from TACs and cranial neuralgias. Be cognizant that migraine in younger children can be as short as 2 hours.[16]

Table 1 Pediatric Headache Patterns and Potential Causes

Headache Pattern	Potential Causes
Acute Headache	Infection (Paranasal infection, URI with fever, Sinusitis, Otitis media, Pharyngitis, Meningitis), Subarachnoid/Intracranial hemorrhage, Stroke, Hypertension, Metabolic causes (hypoglycemia, hypercapnia, hypoxia), Post-seizure, Substance abuse, Intoxication, Trauma
Acute Recurrent Headache	Migraine, Tension type headache, Cluster headache, Paroxysmal hemicrania, Exercise headache, Cranial neuralgias
Chronic Progressive Headache	CNS neoplasm, brain abscess, subdural hematoma, hydrocephalus, chronic meningitis, PIH
Chronic Non-Progressive Headache	Chronic migraine, Chronic tension type headache, Post-concussion, Medication overuse headache, depression
Mixed Headache	Primary headache disorder (eg chronic migraine, chronic tension type headache) with superimposed secondary cause

Are there other symptoms accompanying your headache?

This question focuses on searching for systemic and neurologic symptoms. Fever with behavioral changes can indicate infectious process. Early morning or nocturnal vomiting in a child with a headache that is progressively worsening may indicate intracranial mass. Pulsatile tinnitus in an obese child can be from PIH. Numbness and/or paresthesia, dysarthria, vertigo and motor weakness must be carefully dissected. While these are aura symptoms of migraine, vascular or demyelinating processes must be excluded. Autonomic symptoms such as lacrimation, facial flushing and/or sweating, nasal congestion and ptosis are associated symptoms of TACs. These can also be symptoms of pediatric migraine.[17]

What triggers your headache, and what makes it better or worse?

Any headache provoked by straining, cough, exercise, physical exertion or Valsalva requires exclusion of secondary cause. Postural aggravation on upright position especially following lumbar puncture (LP) indicates intracranial hypotension, while worsening of headache on supine position indicates intracranial hypertension.

Do you take headache medications or other medications for other medical conditions?

Reviewing the patient's medication list can provide clues to the diagnosis. A headache with visual obscuration in a patient taking minocycline or growth hormone may indicate secondary intracranial hypertension. A teenager with headache and seizure while on oral contraceptives can have cerebral venous sinus thrombosis (CVST). Frequent use of analgesics or triptans may lead to MOH. Adolescents should be screened for substance use such as THC that can cause reversible cerebral vasoconstriction syndrome.

Do you have other medical conditions?

Current or previous medical conditions can identify headache etiology. New onset headache in a patient with known malignancy can indicate metastatic disease. Opportunistic infection can occur in an immunocompromised patient presenting with headache and fever. Connective tissue disorders such as Ehlers-Danlos may have cervical arterial dissection presenting as thunderclap headache. Presence of anxiety or depression can contribute to perpetuation or worsening of headaches.

Is there a family history of headache or neurologic disorder?

Migraine is an inherited disorder, and absence of a family history of migraine has been regarded a predictor for space-occupying lesion and the need for neuroimaging.[18] Consider that not all parents know their family history. Therefore, in the absence of a migraine family history in a child with headache consistent with migraine and normal neurologic examination, obtaining a brief parental headache history can disclose an undiagnosed migraine. Vascular malformations are heritable as well as genetic conditions with predisposition to tumors such neurofibromatosis and tuberous sclerosis.

What do you think is causing your headache?

While this question doesn't identify a secondary headache, it identifies the level of parental anxiety about the headache and sets the discussion on confident reassurance for the clinical diagnosis of a primary headache disorder.

Physical and Neurologic Examination

A thorough physical and neurological examination are essential in a headache evaluation. Table 2 summarizes the important aspects of the examination of a child presenting with headache.

The value of a careful examination cannot be overemphasized as an abnormal neurologic exam is highly indicative of a secondary headache pathology. The combination of detailed history and examination also uncovers headache red flags.

A simplified approach to red flags for headache is the mnemonic SNOOP introduced by Dodick in 2003.[19] Following the review of the literature by Kabbouche and Cleves on when to image children with acute headache,[20] this mnemonic was adapted to SNOOP^{4}Y for use in pediatric patients[21] (Table 3).

Following the clinical evaluation, a clinician must be able to clinch a diagnosis of a primary headache disorder, narrow down the differential diagnosis and identify patients who will need neuroimaging or further investigations.

Neuroimaging Considerations

A common question to arise in the evaluation of a pediatric patient presenting with headache is whether or not to obtain neuroimaging. As previously discussed, thorough clinical evaluation can determine a primary headache disorder from a possible secondary headache syndrome. It should be noted that most children with brain tumors, the most feared diagnosis, have abnormal neurologic examination at the time of diagnosis.[22]

Neuroimaging is typically not appropriate for children whose presentation is consistent with a primary headache disorder.[23] In the study of Yilmaz et.al, only a minority of patients with migraine and TTH who underwent neuroimaging had magnetic resonance imaging (MRI) abnormalities.[24] Most of these abnormalities were incidental findings with $<1\%$ of the patients having relevant imaging findings that explained their headache. Notably, parental concern was the reason for imaging in the majority of these patients. Likewise, an occipital headache location in the absence of concerning

Table 2 Physical and Neurological Examinations in the Evaluation of Pediatric Headache

Examination	Worrisome signs and clinical relevance
Vital signs (blood pressure, heart rate, temperature, BMI)	- Hypertension may be the cause of headache or a sign of increased ICP. - Fever suggests an infectious process. - Elevated BMI raises suspicion for PIH.
Head Circumference	Macrocephaly may indicate an elevated ICP that is slowly progressing
Head and Neck (Palpate and auscultate)	- Pericranial tenderness may suggest migraine and tension type headache. - Tenderness on the back of the head may indicate occipital neuralgia. - Paranasal sinus tenderness may indicate sinusitis. - Nuchal rigidity in the proper setting can signify meningitis. - Cranial, orbital and cervical bruits may indicate vascular malformation
Fundoscopic exam	- Papilledema, absent venous pulsations, hemorrhages may indicate increased ICP
Mental Status	- Fluctuating mental status may indicate elevated ICP, intracranial hemorrhage or encephalitis. - Dysarthric speech may indicate stroke or vascular event
Neurologic examination	- Abnormal pupillary reaction, ocular motility, visual fields, asymmetric facial movements, motor and sensory deficits, abnormal deep tendon reflexes, cerebellar signs may indicate intracranial pathology
Skin	- Neurocutaneous stigmata (eg Neurofibromatosis, tuberous sclerosis) can be highly associated with intracranial tumors and vasculopathy. - Presence of rash may suggest autoimmune and infectious processes. - Presence of bruises may indicate trauma or bleeding disorder - Needle tracks can be seen in substance use.

features and in the setting of a normal neurologic examination does not require neuroimaging.[15]

When secondary headache is suspected, the most appropriate imaging modality will be dependent upon the acuity of the presentation and the diagnoses being considered. In most cases, MRI is the more medically appropriate imaging study over computed tomography (CT).[23] Lateef et.al. demonstrated that even in the ED setting, CT seldom yields a diagnosis or alters management acutely in children presenting with non-worrisome headache history and normal neurologic exam.[11]

Table 3 SNOOP[4]Y Mnemonic for Evaluation of Pediatric Headache and Need for Investigations*

S	Systemic symptoms or illness: fever, altered level of consciousness, anticoagulation therapy, pregnancy, cancer, HIV infection
N	Neurologic symptoms and signs: papilledema, asymmetric cranial nerve function, asymmetric motor function, abnormal cerebellar function, new seizure, focal findings at examination
O	Onset recently or suddenly (thunderclap headache)
O	Occipital localization of pain†
P	Pattern: precipitated by Valsalva maneuver
P	Pattern: Positional
P	Pattern: Progressive
P	Parents: lack of family history‡
Y	Years: < 6 years

*Adapted from Gofshteyn JS, Stephenson DJ. Diagnosis and management of childhood headache. Curr Probl Pediatr Adolesc Health Car. 2016; 46: 36-51.

†Occipital headache alone with normal neurologic examination can be due to primary headache disorder and not more likely to be associated with secondary pathology[14]

‡May require obtaining brief headache history from parents to establish undiagnosed migraine

Radiographs (plain film x-rays) have little role in diagnosing headache unless to evaluate for skull fracture in cases of head trauma or suspected physical abuse.[23]

Head CT without intravenous (IV) contrast is appropriate in the ED setting for the following clinical scenarios:

- *Acute onset "thunderclap" headache* — a severe, explosive headache with peak intensity at onset, often referred to as the "worst headache of one's life": CT can evaluate for SAH and is superior to MRI in detection of acute SAH.[23,25] It is important to note that aneurysmal hemorrhage is uncommon in children, approximately 35 times more common in the adult population than the pediatric population.[26] Other potential etiologies of SAH include bleeding arteriovenous malformation (AVM) or trauma.
- *Traumatic brain injury*: The Pediatric Emergency Care Applied Research Network Head Injury and/or Trauma Algorithm, commonly known as the PECARN rule, can be used to identify patients at low risk of clinically-important brain injuries following head trauma in order

Table 4 PECARN Pediatric Head Injury/Trauma Algorithm: Findings associated with very low risk of significant traumatic brain injury in children

Age (years)	Clinical criteria
<2	Normal mental status
	Normal behavior per routine caregiver
	No loss of consciousness (LOC) >5 seconds
	No severe mechanism of injury*
	No non-frontal scalp hematoma
	No evidence of skull fracture
2-18	Normal mental status
	No LOC
	No severe mechanism of injury
	No vomiting
	No severe headache
	No signs of basilar skull fracture

*Defined as motor vehicle crash with patient ejection, death of another passenger, or rollover; pedestrian or bicyclist without helmet struck by a motorized vehicle; fall from >0.9m/3ft for child under 2 or fall from >1.5m/5ft for child 2-18; head struck by a high-impact object

to determine for whom it is safe and appropriate to defer CT in favor of clinical observation.[27] The PECARN rule can only be applied to patients with a Glasgow Coma Score (GCS) of 14 or greater. Head CT is not recommended for patients who meet all of the clinical criteria (Table 4). For those in whom individual criteria are not met, observation or neuroimaging may be indicated.

- *Signs or symptoms of hydrocephalus*: Examples include rapidly increasing head circumference, downward deviation of the eyes ("sunset sign"), excessive vomiting, somnolence or irritability. It is rare that such a child's initial presentation will be to the ED unless the child has a history of shunted hydrocephalus. More likely, these symptoms will be brought to the attention of the primary care provider who should then direct the patient to the ED for evaluation.
- *Suspected intracranial infection*: CT is beneficial in ensuring that it is safe to proceed with LP for evaluation of suspected CNS infection by excluding risks of herniation such as mass lesions or cerebral edema.[23]
- *Focal deficits on neurologic examination or history otherwise concerning for a brain tumor or increased ICP*: MRI remains the preferred imaging study in these situations. MRI offers a more detailed evaluation of brain structure, particularly of the posterior fossa anatomy that is more limited on CT images.[23,28]

A word on sinusitis: Typically, CT is not required as sinusitis is a clinical diagnosis. Notably, patients with migraine are frequently misdiagnosed as having "sinus headache." Clinicians must have a high index of suspicion for migraine given its frequency in the pediatric population, especially for patients presenting with recurrent stereotyped episodes of headache accompanied by autonomic symptoms like rhinorrhea but without more concerning signs of infection like fever.[16,23]

Some important considerations for MR imaging by clinical situation are as follows:

- For patients in whom PIH is suspected, the recommended imaging studies include MRI to rule out intracranial mass and MR venography (MRV) to rule out CVST.[23] MRI can also detect signs of increased ICP including an empty sella, dilated optic sheaths and flattening of the posterior aspects of the globes. CT venography can be considered if MRV is not available, however it is less preferred due to radiation exposure.
- For patients in whom Chiari I malformation is suspected, the recommended imaging study is MRI with sagittal T2-weighted sequence of the craniocervical junction and CSF flow study at the cranio-cervical junction.[23]
- MR angiography (MRA) is indicated in only a few clinical scenarios. These include: for patients with sickle cell anemia presenting with headache; as a secondary study after SAH or intraparenchymal hemorrhage has been identified on initial imaging; for patients with "thunderclap" headache; and for patients in whom arterial dissection is suspected.[23]

In most cases, the initial MRI study can be performed without IV contrast except in cases of intracranial infection, for which MRI with and without IV contrast is indicated.[23]

The neuroimaging considerations outlined in this review are not all-inclusive. For detailed recommendations regarding choice of imaging modalities in pediatric patients with headache, the authors suggest the practice guideline by the American College of Radiology as an excellent resource to guide medical decision-making in clinical practice.[23]

Ancillary Testing

Lumbar Puncture (LP)

For patients in whom intracranial infection or PIH are suspected, LP is indicated after initial neuroimaging confirms that it is safe to proceed with the procedure. Appropriate cerebrospinal fluid (CSF) studies include body fluid cell count, protein, glucose and culture. The diagnosis of PIH requires that other etiologies of intracranial hypertension have been excluded, and the Modified Dandy criteria specify that there can be no CSF cytologic abnormalities.[29]

Opening pressure should be obtained for any patient with signs or symptoms of, or suspected to have, elevated ICP. While intracranial hypertension is often recognized as a complication of bacterial or fungal meningitis, cases of aseptic or viral meningitis have been reported.[30,31] When feasible, opening pressure should be measured on all patients undergoing LP, because signs and symptoms of increased ICP may not be evident immediately.

Electroencephalogram (EEG)

Regardless of the presence or absence of EEG abnormalities, EEG offers minimal if any contribution to the diagnosis of

patients presenting with headache,[32,33] and rarely alters clinical management. The American Academy of Neurology practice parameter on the evaluation of pediatric patients with recurrent headache notes that among children with recurrent headache found to have paroxysmal abnormalities on EEG, the risk of future seizures is negligible. The authors further state that for these patients "investigation for epilepsy or treatments aimed at preventing future seizures is not indicated".[28] An exemption is a patient presenting with headache and seizure, for whom a primary headache disorder cannot be assumed, and for whom further diagnostic workup including EEG and neuroimaging is indicated.[23,34]

Laboratory Evaluations

Laboratory testing is rarely useful in the evaluation of pediatric headaches and is indicated when a secondary cause such as infection or metabolic disease is suspected.[28] In these cases, headache will not likely be the sole presenting symptom. The American Academy of Neurology guidelines affirm that there is insufficient evidence to support any recommendation regarding the appropriateness of routine laboratory studies in the evaluation of pediatric patients with headache.[28]

Conclusion

Pediatric headaches are common and most often due to a primary headache disorder or a benign self-limited cause. The astute clinician can usually reach the diagnosis with a thorough history and examination. The history and examination are imperative to the clinician's ability to determine when to be concerned for a possible secondary cause of headache, which subsequently guides further diagnostic evaluation.

When neuroimaging is indicated, a non-contrasted MRI of the brain is often the most valuable study. There are specific situations, particularly in the ED setting, for which a non-contrasted head CT would be indicated. It is important to balance the risks of neuroimaging, such as radiation exposure from CT or sedation for very young children, with the potential benefits of aiding in diagnosis and treatment.

LP and laboratory tests are done depending upon the diagnoses being considered. In the absence of clinical seizures or altered mental status, an EEG is never indicated in the evaluation of pediatric headache.

Conflict of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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