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Uncommon Headache Syndromes in the Pediatric Population

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Abstract Headache is one of the most common symptoms in children and adolescents, and headache syndromes are an important reason for medical consulting. According to the second edition of the International Classification of Headache Disorders, there are 196 possible headache diagnoses, of which 113 have been described in pediatric population. Herein, we focus on unusual pediatric headache syndromes. We group them as headaches with migraine features, short-duration headaches with autonomic features, short-duration headaches without autonomic features, and potentially ominous forms of headaches. Although rare as single entities, providers focusing on pediatric headaches certainly will face some of these headaches and need to be comfortable on the diagnostic approach.

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Introduction

Headache is one of the most common symptoms in children and adolescents, and headache syndromes are an important reason for medical consulting. We recently conducted a population study that showed a lifetime prevalence of headache in children and adolescents of 81%. Although migraine and tension-type headaches were the most common diagnoses [1••], a sizable 1.7% of the children had chronic daily headaches, and 4.2% of the children had 10 or more days of headache per month [2••].

The study of headache disorders in the pediatric population is important not only because of its prevalence; the burden of pediatric headaches has been well established as impacting the families and the children, including school performance, family function, social life, mental health, and quality of life. Also, the phenotype of particular syndromes, as well as their differential diagnoses, varies as a function of age, and this may pose diagnostic and therapeutic challenges. These outstanding peculiarities are enhanced by the fact that children often are incapable of describing important clinical features, and providers are not comfortable diagnosing unusual headache syndromes in the pediatric population.

The second edition of the International Classification of Headache Disorders (ICHD-II) [3] describes 14 headache categories subdivided into a total of 196 possible headache diagnoses, of which 113 have been described in the pediatric population (Arruda and Bigal, unpublished data). For the vast majority of these conditions, the headache

characteristics are not described in detail, especially for the secondary headache groups (5–12). Furthermore, some of these disorders are so rare in the pediatric population (prevalence less than 1:1500; eg, Takayasu's arteritis, pheochromocytoma, posterior reversible encephalopathy syndrome, systemic lupus erythematosus, Chiari malformation type I, craniocervical arterial dissection, and cavernous angioma) that a comprehensive review of them is beyond the scope of any article. Accordingly, this paper focuses on pediatric uncommon headache syndromes for which we have recent and substantially clinical information about the headache characteristics. We first describe a brief algorithm on how to approach a child with headache as the main symptom. We then describe specific unusual syndromes in the pediatric population.

Approaching Children with Pediatric Headaches of Difficult Recognition

The diagnosis of the headache disorders in the pediatric population is challenging, as a consequence of their limitations in describing the numerous features of the symptoms. Moreover, it may be complicated by the fact that one individual may have more than one disorder.

An orderly approach is required for the proper diagnosis. Crucial elements include a thorough history (based on the information given by the parents and child) supplemented by general medical and neurological examinations, as well as laboratory testing and neuroimaging in selected patients. If multiple headache disorders occur concurrently, the conceptual process needs to be repeated for each headache.

An important first step is to distinguish primary from secondary headaches. In brief, the approach is to spot "red flags" that suggest the possibility of secondary headache (Fig. 1) [4]. Once these features are identified, the physician must conduct the workup indicated by the red flag (Table 1) and thereby diagnose any secondary headache disorder that is present [4].

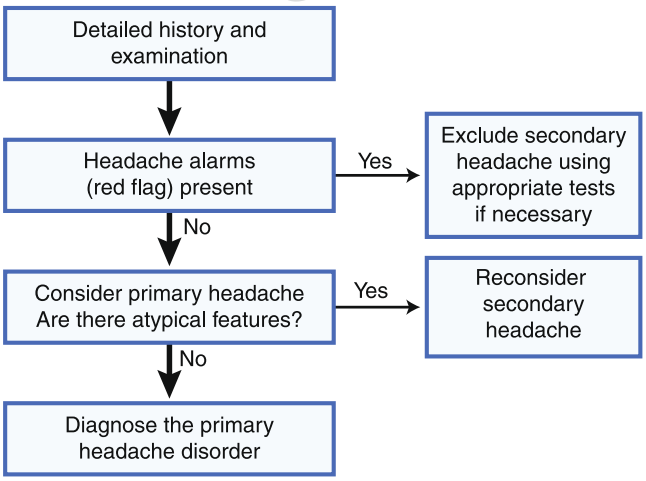

Fig. 1 Algorithm for headache diagnosis: flow assessment for patients with headache disorders. (Modified from Evans and Purdy [4].)

In the absence of secondary headache, the clinician proceeds to diagnosing a primary headache disorder. If the headache is atypical or difficult to classify, the possibility of secondary headache should be reconsidered, although the modifying effect of any treatment being taken should be kept in mind.

In children, once secondary headache disorders are excluded and common primary headache disorders (eg, migraine with or without aura, tension-type headache) have not been diagnosed, it is useful to frame the differential diagnoses by grouping headaches according to common features. Herein, we group them in the following categories: long-duration headaches with migraine features, short-duration headaches with autonomic features, short-duration headaches without autonomic features, and potentially ominous forms of headaches.

Long-Duration Headaches with Migraine Features

Hemiplegic Migraine

Hemiplegic migraine (HM) is a rare migraine-with-aura subtype. It is characterized by fully reversible attacks of motor weakness with or without other aura symptoms (positive or negative visual features; positive or negative sensory symptoms; or dysphasic speech). Symptoms of basilar aura occur in up to 70% of the patients [3]. HM is subdivided into familial or sporadic forms.

In familial hemiplegic migraine (FHM), there is at least one first- or second-degree relative with migraine aura including motor weakness, and in sporadic hemiplegic migraine (SHM) there is not. The prevalence of both forms of HM is similar (about 1:10,000) [5].

Gene mutations associated with these disorders have been identified (eg, *CACNA1A*, *ATP1A2*, and *SCN1A*). In FHM type 1, there are mutations in the *CACNA1A* gene on chromosome 19, and in FHM type 2, mutations occur in the *ATP1A2* gene on chromosome 1. Other pedigrees have been identified.

Aura symptom develops gradually over 5 min, lasting less than 24 h. The headache begins during the aura or follows onset of aura within 60 min, and its characteristics fulfill the criteria for migraine without aura. Hemiparesis or single-limb paresis must be present, and bilateral paresis provides negative evidence against the diagnosis. In the course of HM, common symptoms include severe and prolonged attacks of hemiplegia, fever, seizure, confusion, and sometimes even coma. Therefore, we emphasize that, for the most symptomatic cases (when diagnosis has not

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t1.1 **Table 1** Red flags in the diagnosis of headache

t1.2 Red flag	Consider	Possible investigation(s)
t1.3 Sudden-onset headache	Subarachnoid hemorrhage, bleed into a mass or AVM, mass lesion (especially posterior fossa)	Neuroimaging; lumbar puncture (after neuroimaging evaluation)
t1.4 Worsening-pattern headache	Mass lesion, subdural hematoma, medication overuse	Neuroimaging
t1.5 Headache with cancer, HIV, or other systemic illness (fever, neck stiffness, cutaneous rash)	Meningitis, encephalitis, Lyme disease, systemic infection, collagen vascular disease, arteritis	Neuroimaging; lumbar puncture; biopsy; blood tests
t1.6 Focal neurologic signs, or symptoms other than typical visual or sensory aura	Mass lesion, AVM, collagen vascular disease	Neuroimaging; collagen vascular evaluation
t1.7 Papilledema	Mass lesion, pseudotumor, encephalitis, meningitis	Neuroimaging; lumbar puncture (after neuroimaging evaluation)
t1.8 Triggered by cough, exertion, or Valsalva	Subarachnoid hemorrhage, mass lesion	Neuroimaging; considerer lumbar puncture
t1.9 Headache during pregnancy or postpartum	Cortical vein/cranial sinus thrombosis, carotid dissection, pituitary apoplexy	Neuroimaging

AVM—arteriovenous malformation; HIV—human immunodeficiency virus.

(Modified from Evans and Purdy [4].)

been previously described or when cognitive symptoms occur), a diagnosis of HM can occur only after exclusion of secondary conditions. The differential diagnosis of HM in children and adolescents includes basilar migraine, stroke (mainly transient ischemic attacks), Todd's paralysis, epilepsy, and the syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL syndrome).

The pharmacological treatment includes prophylaxis with antiepileptic agents and/or acetazolamide in the aura period. Triptans and ergotamine compounds are contraindicated in these children.

Retinal Migraine

Retinal migraine (RM) is characterized by recurrent attacks of fully reversible monocular visual disturbances (scintillations, scotomata, or blindness) that are temporally associated (within 60 min) with headache fulfilling criteria for migraine without aura [3].

The diagnosis of RM in children is difficult. We usually ask the child to draw their symptoms or to indicate in a chart to confirm the monocular visual phenomena during the attack. Appropriate ophthalmologic and neurologic investigations are needed to exclude other very rare causes of transient monocular blindness in children, such as retinal detachment, optic neuropathy, carotid dissection, and vasculitis.

The prevalence of RM in children is yet to be determined. In a multicenter hospital-based study in France involving 398 children with chronic headache, only one case of RM was reported [6•].

The treatment of RM in the pediatric population is similar to the treatment in older patients, consisting of therapeutic doses of antiepileptic drugs, calcium-channel blockers, or flunarizine. Anecdotal evidence suggests that magnesium or vitamin B2 may be of help as well.

Chronic Headaches

Chronic Migraine

Chronic migraine (CM) is a significant worldwide problem not only in adults, but also in children and adolescents [7–9], although few pediatric studies were conducted in representative samples. We conducted a population study, wherein 1.7% of the children younger than 11 years had chronic daily headaches (although formal CM diagnosis was not established) and another 2.52% had headaches on 10 days or more [2••].

The diagnosis of CM in children is identical to the diagnosis in adults. Patients should have headaches on at least 15 days per month, and at least half of these headaches should be migraine. Nonetheless, substantial differences exist between CM in adults and in children. For example, while acute medication overuse seems to be an important risk factor in adults, this is not the case for children. This issue is of particular importance. It may be speculated that those with special biological vulnerability for frequent pain would develop CM early and without the need of particular risk factors; those with intermediate predisposition would develop the disease later in the presence of specific risk factors such as medication overuse, obesity, and stressful life events among others. Those without predisposition would not develop CM even when exposed to risk factors for migraine progression. They would persist with episodic migraine and could eventually remit [10].

Also worth mentioning is that CM is more refractory when developed early in life, or when the time from onset of episodic migraine to CM is less than 1 year [10]. Finally, adolescents with CM have more migraine attacks than adults. It is well established that during the process of migraine progression, as attack frequency increases, the number of migraine features diminish during the transformation period. Accordingly, it is natural that adolescents with chronic daily headache have more migraine days compared to adults, as demonstrated by a study conducted in the specialty care [11].

Describing the treatment of CM is beyond the scope of this review and the readers are referred to the review by Termine et al. [12••] for further information

Short-Duration Headaches

Short-Duration Headache with Autonomic Features

Cluster Headache

The phenotype of cluster headache (CH) in children is not substantially different than what is seen in adults, being characterized by attacks of severe and sometimes excruciating unilateral pain, mainly orbital, supraorbital, and/or temporal. Attacks typically last from 15 to 180 min and happen from every other day to up to eight attacks per day. They are accompanied by at least one of the following autonomic ipsilateral features: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, eyelid edema, forehead and facial sweating, miosis, and ptosis. Along the time course of CH, the attacks may become less severe, of shorter or longer duration, and less frequent [3]. Both forms of CH, episodic (pain-free periods lasting 1 month or longer) and chronic (no remissions or remissions that are shorter than 1 month), are seen in children, although about a quarter of the patients have only a single cluster period [3].

The diagnosis of CH in children often is misleading because incidence peaks between the ages of 20 and 40 years. CH is rare, and population-based studies have estimated the lifetime prevalence as being 0.1% [13]. At younger ages, prevalence becomes significantly lower. In 18-year-old Swedish Army recruits, prevalence was only 0.09% [14]. In a multicenter pediatric study, only two cases of CH were seen among 6629 children and adolescents attended in 27 Italian headache centers [15]. Nonetheless, if a first-degree relative is affected, chances of CH increase by up to 14-fold.

A recent systematic review found that only 80 cases of CH in children have been reported [15–20, 21••], and many of them did not meet the full ICHD-II diagnostic criteria for CH. In many, pain was described as bilateral, while others responded to propranolol or other migraine preventive medications, raising the question whether CH has a different phenotype in children or if diagnostic issues exist. In our experience, we have followed only one atypical case of CH in a 12-year-old boy with bilateral autonomic manifestations and fulfilling all the other diagnostic criteria for CH. The headache attacks remitted after a period of 6 months and did not recur after 10 years of follow-up.

Paroxysmal Hemicrania

Paroxysmal hemicrania (PH) is characterized by short attacks (2–30 min) of excruciating unilateral pain in the first trigeminal division. Pain is accompanied by at least one ipsilateral autonomic feature, such as conjunctival injection, lacrimation, nasal congestion, rhinorrhea, eyelid edema, forehead and facial sweating, miosis, and ptosis. Attacks are frequent (>5 per day) and remit completely after indomethacin. The shorter duration and higher frequency of the attacks, as well as the absolute treatment response to indomethacin, differentiate PH and CH [3].

Different than CH, most cases of PH start during childhood. The first pediatric case was reported by Kudrow & Kudrow [22] in 1989 and described a 9-year-old boy that had PH since the age of 6 years. He evolved from episodic to chronic PH and successfully responded to aspirin prophylaxis.

Based on a literature review, most described cases of PH have longer duration and lower frequency of headache attacks, as well as only partial response to indomethacin, making the differential diagnosis with CH sometimes cumbersome [23–26].

Short-Lasting Unilateral Neuralgiform Headache Attacks with Conjunctival Injection and Tearing

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is ultrashort trigeminal autonomic cephalalgias. It is a very rare headache syndrome characterized by short-lasting attacks (5–240 s) of unilateral pain (orbital, supraorbital, or temporal) of stabbing quality, accompanied by ipsilateral conjunctival injection and lacrimation (nasal congestion, rhinorrhea, ptosis, or eyelid edema may occur). Its frequency ranges from 3 to 200 per day [3].

This syndrome was first described by Sjaastad et al. [27] in 1989 and has been reported in only three children since then [28–30]. Of them, two were idiopathic and one symptomatic (pilocytic astrocytoma).

When facing such a dramatic headache presentation, the clinician must exclude lesions of the posterior fossa or of

the pituitary gland, even when the case is typical SUNCT and without neurologic deficits. Possible underlying pathologies include posterior fossa tumors, cavernous hemangioma, arteriovenous malformation, dorsolateral brainstem stroke, and basilar impression.

SUNCT responds poorly to triptans and indomethacin. Preventive options include lamotrigine and gabapentin. Spontaneous remission may occur.

Hemicrania Continua

Continuous headache without pain-free intervals is a rare condition in children and adolescents. Hemicrania continua (HC) is a primary headache characterized by continuous and strictly unilateral headache of moderate intensity accompanied by autonomic features (conjunctival injection, lacrimation, nasal congestion, rhinorrhea, ptosis, and miosis). It is completely responsive to indomethacin. The diagnosis of HC in childhood is extremely rare [31], but most adults with HC report that their headaches started at childhood, suggesting that HC is indeed underdiagnosed and missed at earlier ages. It may be prudent to try indomethacin once secondary headaches have been ruled out in children with unilateral and chronic headaches.

Short-Duration Headache Without Autonomic Features

Primary Stabbing Headache

Primary stabbing headache (PSH; formerly known as ophthalmodynia periodica, ice-pick headaches, or jabs & jolts syndrome) is characterized by one or multiple well-localized attacks of stabs in the distribution of the first division of the trigeminal nerve. Stabs last no more than a few seconds, from one or few to several per day. Characteristically, associated symptoms are absent. Nonetheless, comorbidity with migraine (about 40%) or CH (about 30%) has been reported in adults.

In a case series, PSH happened in 83 of 2543 children and adolescents with recurrent headaches seen in a tertiary center (3.2% of headache cases) [32]. In other series, PSH responded to a lower relative frequency among headache cases (0.5%) [33, 34]. As with other pediatric headaches, only a small proportion of children with probable PSH fulfilled full ICHD-II diagnostic criteria [35].

To the best of our knowledge there is no populational study concerning PSH in children and adolescents.

The differential diagnosis of PSH includes colloid cyst and tumors of the third ventricle and pineal region, Arnold-Chiari malformation, platybasia and basilar impression, chronic subdural hematoma, and pheochromocytoma.

Some uncontrolled studies have shown a positive response to indomethacin, while others have shown only partial or no responses.

Hypnic Headache

Hypnic headache (HH) typically is a disorder of the elderly and is very rare in children. It is characterized by headache attacks happening exclusively during sleep, awakening the patient. Attacks last less than 15 min and occur on most days [3]. Two recent papers reported HH in one child [36] and one adolescent [37], and clinicians should be aware of this rare possibility. Differential diagnosis includes nocturnal migraine, trigeminal autonomic cephalalgias, and headache attributed to increased intracranial pressure. Indeed, in our personal experience, over one quarter of the children with migraine have nocturnal awakenings caused by headache. Because migraineurs also are more likely to have periodic syndromes characterized by sleep disturbances, the differentiation may be very difficult sometimes.

The treatment of HH in children is not described. Caffeine (a cup of coffee at bedtime) and lithium seem to be effective. Other prophylactic agents (eg, flunarizine, gabapentin, acetazolamide, and indomethacin) may be tried.

Cranial Neuralgias

Trigeminal neuralgia Trigeminal neuralgia (TN) is characterized by paroxysmal and stereotyped attacks of intense and sharp pain lasting a fraction of a second to 2 min, affecting one or more divisions of the trigeminal nerve (mostly the second and the third), being precipitated by trivial stimuli (including washing, shaving, smoking, talking, and brushing the teeth) or occurring spontaneously. A spasm of the facial muscles on the affected side may be evoked by the pain, the so called *tic douloureux* [3].

TN may be idiopathic or symptomatic (compression of the trigeminal root by tortuous or aberrant vessels). TN is very rare in children and symptomatic until proven otherwise (Chiari type I, embryonal rhabdomyosarcoma, lipoma, and pilocytic astrocytoma).

Accordingly, in children with probable TN, complete investigation should be performed, including magnetic resonance imaging (MRI) and MR angiography.

Glossopharyngeal neuralgia Glossopharyngeal neuralgia (GN) is characterized by paroxysmal attacks of lancinating unilateral pain in the distributions of the auricular and pharyngeal branches of the vagus nerve, as well as of the glossopharyngeal nerve (posterior part of the

tongue, tonsillar fossa, pharynx, or beneath the angle of the lower jaw and/or in the ear), lasting from a fraction of a second to 2 min. The attacks are commonly precipitated by swallowing, chewing, talking, coughing, and/or yawning. As with TN, GN may be idiopathic or symptomatic. In symptomatic GN, there is evidence of sensory impairment in the distribution of the glossopharyngeal nerve at the neurologic examination, and a causative lesion may be identified with a proper investigation and/or surgery [3]. As with TN, GN is typically secondary in children (posterior fossa tumors, multiple sclerosis, vascular malformation, and neurovascular compression) [38].

Nervus intermedius neuralgia Nervus intermedius neuralgia (NIN), also known as geniculate neuralgia, is an extremely rare condition that causes pain that is very similar to that of TN, but with the distribution of nervus intermedius. Paroxysms are felt in the auditory canal and phenotype often resembles otological disorders. Herpes zoster, temporomandibular joint disease, carcinoma of the nasopharynx, osteoma of the petrous bone, and neuroborreliosis may mimic NIN.

In 2006, we reported a case of a secondary NIN caused by a trigeminal schwannoma in a 2-year-old girl [39]. She had a 4-month history of left auricular pain, with no precipitating factor, lasting about 8 s. During attacks, the child suddenly would cry and place her left hand above the left ear, although not touching it. The frequency gradually increased over 2 months, ranging from 10 to 20 attacks per day. There were diurnal and nocturnal attacks and no autonomic features. Neurologic examination was normal and MRI disclosed a mass suggestive of a left trigeminal tumour. Surgery revealed a schwannoma. She remains asymptomatic after 8 years of follow-up.

Potentially Ominous Conditions

Thunderclap Headache

Thunderclap headache (TH) is a dramatic headache presentation characterized by abrupt onset of a very severe headache (maximum intensity in <1 min) that lasts from 1 h to days. In the vast majority of the cases, an underlying cause is identified (subarachnoid or intracerebral hemorrhage, cerebral venous thrombosis, unruptured vascular malformation, arterial dissection, central nervous system angiitis, reversible segmental cerebral vasoconstriction, and pituitary apoplexy) and little evidence exists that TH may exist as a primary condition. Accordingly, children with TH should be handled urgently.

In children, TH may happen secondary to reversible segmental cerebral vasoconstriction [40, 41], a recognizable clinical and radiographic syndrome consisting of TH with or without focal neurologic symptoms combined with reversible segmental vasoconstriction of proximal cerebral blood vessels.

Headache Attributed to Brain Tumor

Brain tumors are the most common solid tumor in children and one of the most frequent causes of cancer deaths in childhood. Despite the fact that headache secondary to this condition is a rare diagnosis among children with chronic headache (<1%) [25], headache is the most frequent, first-presenting symptom in children with brain tumor [42].

Accordingly, the clinician should look carefully for red flags in children with headaches, which include progressive and continuous pattern of the headache, strictly localized pain, significant worsening during the night (awakening the child), or occurrence in the early morning, as well as headaches aggravated by maneuvers known to increase intracranial pressure (such as Valsalva maneuver, coughing, sneezing, and bending forward) and associated with prominent nausea and/or vomiting.

In a case series of 200 children with brain tumor, initial diagnosis was migraine in nearly a quarter of the cases, and almost 10% were diagnosed as having tension-type headaches [42]. The clinician also should keep in mind that having migraine or tension-type headache does not "immunize" the child against a brain tumor.

Detailed physical and neurologic examinations are of utmost importance in children with headaches. In most children with brain tumor, some neurologic abnormality is found within 6 months of headache onset [43].

Neck–Tongue Syndrome (N-TS)

The neck–tongue syndrome (N-TS) is characterized by sudden onset of pain upon neck rotation. Pain happens in the occiput or upper neck and is associated with dysesthesia in the same side of the tongue. The duration of the pain is usually short, lasting seconds or minutes, and residual numbness may persist in the tongue for a while [3, 44].

To the best of our knowledge, there are seven cases reported in children and adolescents, five of them with a familial occurrence, suggesting an autosomal dominant inheritance pattern [45, 46].

In the Vågå study, Sjaastad and Bakketeig [44] have described three interesting cases of N-TS in which headache attacks have started in childhood and adolescence.

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Clinical and surgical evidence link N-TS to C2 root involvement, particularly with subluxation of the atlantoaxial joint.

Painful Ophthalmoplegia

Tolosa-Hunt Syndrome

Tolosa-Hunt syndrome (THS) is characterized by a relapsing and remitting course of episodic and unilateral orbital pain, associated with paralysis of one or more of the C3, C4, and C6 cranial nerves. It persists for weeks if untreated but resolves within 72 h when treated with adequate doses of corticosteroids (prednisolone, 1–2 mg/kg/day) [3].

Demonstration of the typical granulomatous lesion involving the cavernous sinus by MRI is mandatory. Other possible causes of painful ophthalmoplegia (eg, ophthalmoplegic migraine, vascular malformation, vasculitis, basal meningitis, diabetes mellitus, meningioma, sphenoid sinusitis, lymphoma and tuberculous pachymeningitis) must be excluded.

The first two cases of THS in children were reported by Terrence and Samaha in 1973 [47], and at least five additional cases have been documented [48, 49].

Ophthalmoplegic "Migraine"

Ophthalmoplegic "migraine" (OM) is diagnosed when migraine attacks are followed within 4 days by paresis of one or more of C3, C4, and C6 nerves (mostly C3) in the absence of demonstrable intracranial lesion other than MRI changes involving the affected nerve [3].

OM formerly was classified as a variant of migraine, but it now is seen as a cranial neuralgia. Of notice, preceding headache attacks often are prolonged (>1 week) and there is a latent period of up to 4 days from the onset of headache to the onset of ophthalmoplegia; tendency for recurrent episodes with more severe and persistent nerve involvement; evidence of permanent neurologic deficits with recurrent episodes (30% of patients); rapid improvement and shortened duration in response to corticosteroid therapy; and transient, reversible MRI contrast enhancement of the affected cranial nerve (86% of patients) [50].

At least 40 cases of OM have been reported. Onset ranges from 3 months to 18 years (mean age: 4.2 yrs), and 73% of the patients were younger than 5 years [49]. The estimated incidence of OM is 0.7 per million, but it may respond to 7% of all childhood oculomotor nerve palsies [50].

Common signs are ptosis, adduction defects, skew deviations, and pupillary involvement (77%) [50].

Conclusions

The differential diagnosis of unusual headaches in children requires a systematic approach. Herein, we presented an orderly approach to differential diagnosis. The precise criteria for each disorder are presented in the ICHD-II.

Table 2 Uncommon characteristics of headache and facial pain in children and adolescents and possible related conditions

Headache characteristic	Possible related conditions
Abrupt onset reaching the maximum intensity in <1 min	Thunderclap headache, AVM, aneurysm, and primary stabbing headache
Short-lasting (seconds or <5 min)	SUNCT, cranial neuralgias, and neck–tongue syndrome
Many attacks in the same day	Cluster headache (<8), paroxysmal hemicrania (>5), SUNCT (3–200), primary stabbing headache, and cranial neuralgias
Continuous headache (without pain-free interval)	Hemicrania continua, chronic daily headache, and intracranial hypertension
Unilateral autonomic signs	Cluster headache, paroxysmal hemicrania, hemicrania continua, and SUNCT
Headache accompanied by fully reversible neurologic symptoms: motor weakness plus sensitive, visual, or speech disturbance	Hemiplegic migraine, basilar migraine, stroke, Todd's paralysis, and HaNDL syndrome
Nocturnal headache awakening the child	Migraine, cluster headache, hypnic headache, and intracranial hypertension
"Nuchal jabs" upon neck rotation and ipsilateral numbness in the tongue	Neck–tongue syndrome and cervicogenic headache
Pain associated to ophthalmoparesis	Tolosa-Hunt syndrome, ophthalmoplegic migraine, AVM, vasculitis, diabetes mellitus, meningioma, sphenoid sinusitis, lymphoma, and tuberculous pachymeningitis
Transient monocular blindness	Retinal migraine, optic neuropathy, carotid dissection, and vasculitis

AVM—arteriovenous malformation; SUNCT—short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing; HaNDL syndrome—syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis.

The approach should help physicians to move forward quickly and safely when assessing children with headaches. Table 2 summarizes some of the differential diagnoses discussed in this paper.

Disclosures Dr. M. A. Arruda: none; Dr. R. Albuquerque: none. Dr. Marcelo E. Bigal is an employee of Merck and Co. and holds stock interest in the company.

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- • Of importance
- •• Of major importance

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