Headache represents one of the most common reasons why children and adolescents are referred to pediatric neurology practices where the most common headache syndromes diagnosed are migraine and its variants, and chronic daily headache. The bulk of recent literature regarding headache in children has focused on these two clinical entities even though large epidemiologic studies have demonstrated that tension-type headache may be two to three times more common in children. Why has so little attention been given to these other disorders? The purpose of this review is to examine the “other” primary headache disorders in children and adolescents. © 2005 by Elsevier Inc. All rights reserved.


Introduction

The International Headache Society has recently published its revised International Classification of Headache Disorders which are available online (www.i-h-s.org/) [1]. This document serves as the gold standard for defining the clinical criteria upon which specific headache diagnoses are established. Part One focuses on the primary headaches, migraine, tension-type, cluster, and other trigeminal autonomic cephalgias, and Part Two outlines the secondary headache syndromes such as headache caused by trauma, vascular disorders, substance use, infection, disorders of homoeostasis, or psychiatric disorders.

This new edition incorporates many welcomed revisions to the 1988 version and contains more developmentally sensitive features within its narratives regarding pediatric migraine. These revisions were adopted based upon solid, evidence-based investigations wherein the 1988 International Headache Society criteria were “field tested” over a 12-year period. Although work remains to be done, these new criteria will provide a valuable foundation upon which future pediatric clinical trials may be performed.

Unfortunately, little evidence exists regarding the other primary headaches in children, so the adult International Classification of Headache Disorders criteria must be used for children. But are they valid? Given the infrequent nature for many of these “other” primary headaches in children, it is possible that different names for the same headache condition or the same name for different headache patterns are being applied. It is imperative that attention be given to the other primary headache syndromes so that future revisions of the International Classification of Headache Disorders may also incorporate the features that will permit broader application for children.

The purpose of this article is to provide a review of the literature regarding the other primary headaches in children and adolescents and highlight the areas that require future exploration. Proponents of the “continuum theory” of headache might argue that this is a futile exercise because most primary headaches represent the spectrum of clinical expression of a common pathophysiology. However, because these entities are clinically distinct and adult criteria have been established for their diagnosis, few would disagree that having criteria that are applicable for children would be valuable.

For this review, the searches were conducted using National Library of Medicine PUBMED and MEDLINE for each headache syndrome.

Tension-Type Headache

The 2004 International Classification of Headache Disorders has divided tension-type headache into three categories: infrequent episodic tension-type headache; frequent episodic tension-type headache; and chronic...
tension-type headache. Each of these types is further subdivided based upon the presence or absence of “pericranial tenderness”.

The diagnostic criteria state:

**Tension-type headache**

A. At least 10 episodes fulfilling the criteria B-D.
B. Headache lasting 30 minutes to 7 days
C. Headache has at least two of the following characteristics:
   1. bilateral location
   2. pressing/tightening (nonpulsating) quality
   3. mild to moderate intensity
   4. not aggravated by routine physical activity such as walking or climbing stairs
D. Both of the following:
   1. no nausea or vomiting
   2. no more than one of photophobia or phonophobia
E. Not attributed to another disorder.

The distinction between infrequent, frequent, and chronic relates to the number of headaches per month: infrequent, less than 1 per month; frequent, more than one per month but not more than 15 per month; and chronic, more than 15 per month.

The key element of these criteria is the absence of migrainous features; absence of unilateral, pulsating quality, severe intensity, aggravation by activity, nausea or vomiting, as well as photophobia and phonophobia. Essentially, these criteria describe a recurring pattern (≥10 episodes) of nonmigrainous headache as being the diagnostic criteria for tension-type headache. Is this valid in children?

**Tension-Type Headache in Children**

**Epidemiology**

Establishing the prevalence of tension-type headache in children has proved a challenge. Professor Bo Bille’s classic epidemiologic study of more than 9000 Scandinavian children found 48% had “infrequent non-migraine headache” and another 7% had “frequent non-migraine” suggesting a prevalence of approximately 50% for tension-type headache in this population, but specific criteria or case definition for “non-migraine” headaches were not provided [2]. Subsequent studies undertaken in various European and developing nations have reported widely divergent rates. Strategies employed to collect the data include door-to-door surveys of small villages, questionnaires distributed to families of school-aged children, recruitment of subjects from primary care offices and headache clinics, and prospective studies monitoring children for years with periodic evaluation for headache. The prevalence estimates range from 11% to 72.8% [3-7]. The largest of these series (n = 8255) that included school children ages 13-15 years found a 1-year prevalence of tension-type headache was 18%; migraine, in this series, had a 1-year prevalence of 7% [7].

Several difficulties have been encountered in collecting and interpreting the data, including differing demographics, methods used to collect data, and reliance upon the accuracy of parental report of the frequency and characteristics of headache for their children. Additionally, as with migraine, the International Headache Society criteria were established to classify headache in adults, not children [8]. It is possible that strict adherence to International Headache Society criteria may fail to identify children with tension-type headache because children are less able to articulate their headache experience and have some differences in symptoms when compared with adults.

**Clinical Features**

Children and adolescents who suffer from tension-type headaches report similar symptoms as adults with some slight modifications. Four large series have been published regarding the clinical phenomenology of tension-type headache in children and adolescents [8-11]. Compiling these series (n = 576) permits description of the typical features of childhood tension-type headache. In one of these series, Gallai (n = 244) found that 52% were episodic tension-type headache, 16% were chronic tension-type headache, and 33% had tension-type headache that did not fulfill criteria for either [8].

The duration of attacks may vary from 5 to 30 minutes or last greater than 48 hours. Gallai identified 36.7% of children with tension-type headache that lasted less than 30 minutes. If International Classification of Headache Disorders criteria for duration were strictly adhered to, a significant number of children with tension-type headache would be missed. By the same token, the location of headache is often difficult for young children to determine and describe [12]. Bilateral location was identified in 57-86% of patients. Wober-Bingol et al. observed that adolescents more often fulfill the criterion for location than younger children. This observation holds true for both the unilateral location of headache in migraine as well as the bilateral character of tension-type headache [9].

The quality of headache was most often described as pressing or tightening (74%) rather than pulsating (16%). In one series, only 15% of children described exacerbation with routine physical activity. Seventy-five percent described the headache as mild to moderate intensity [2]. Wober-Bingol et al. identified intensity as the most relevant headache characteristic for differentiating migraine from tension-type headache in a group of 409 children [9]. In questionnaires distributed by Aromaa et al. to families of children comparing migraine and tension headache, children with tension-type headache were less likely to report abdominal pain, nausea, vomiting, vertigo, visual
disturbances, sweating, vomiting, or using a darkened room for pain relief (9-30%) [10].

Therefore, the “non-migraine” features of quality (pressing or tightening), intensity (mild-moderate), and lack of associated symptoms may be more specific for tension-type headache (vs migraine) than location or duration in pediatric patients. Large, prospective series are needed.

Management

We were able to discover only two published trials investigating the pharmacologic management of tension-type headache in children or adolescents. One clinical trial, still in progress, compares relaxation training (“limited contact format”) compared with amitriptyline (10 mg daily) for children ages 8-16 years (n = 19) for 3 months of a planned 12-month program. Clinical improvement was observed in both groups. For the amitriptyline group, baseline headaches were 17 ± 11 migraine headaches/month and reduced to 5.6 ± 6.7 headaches per month after 3 months. This result is comparable to the behavioral group where baseline was 12.1 ± 10.1 headaches/month down to 6.4 ± 9.6 per month (not significant) [13]. The second trial (n = 48) compared the efficacy of relaxation to a muscle relaxant chlormezanone in the treatment of adolescent tension-type headache in a randomized, double-blind, crossover design and found significant improvement in the home-based relaxation therapy without further improvement with addition of drug treatment [14].

Bio-behavioral therapies (e.g., relaxation treatment and thermal biofeedback) have been assessed. Two studies comparing relaxation therapy for adolescents with migraine, migraine plus tension-type headache, or tension-type headache alone found significant improvement in migraine patients but not the tension-type headache group [15,16].

In another study, children (n = 37; mean age 12 years) with episodic tension-type headache who were examined in small group settings once per week for 8 weeks and taught coping skills and progressive relaxation techniques were found to have statistically significant and sustained (~1 year) reduction in headache days, “state and trait” anxiety, and depression scales [17].

One small study of five children (ages 8-14 years) with tension-type headache used six thermal biofeedback sessions and found significant reduction in headache frequency, duration, and intensity with sustained (6-month) headache-free state in four of the five children [18]. A combination approach with electromyographic biofeedback and progressive muscle relaxation therapy was found to be effective (86% improvement) vs the control group (50%) in 20 children, mean age 11 years, with sustained efficacy for 6 and 12 months [19].

Prognosis

There is a paucity of information regarding the prognosis of tension-type headache in children. No longitudinal series have been published.

Comorbidity has been evaluated. In a study of Finnish children with tension headaches identified by questionnaire, Anttila et al. described a higher frequency of depressive symptoms when compared with headache-free children [20]. A study by Fearon and Hotopf theorized that children with headache are at increased risk for both headache and psychiatric symptoms as adults [21].

Cluster and Other Trigeminal Autonomic Cephalgias

Cluster headache and the trigeminal autonomic cephalgias share the clinical features of repetitive attacks of intense headache accompanied by prominent cranial parasympathetic signs and symptoms. Cluster and paroxysmal hemicrania are further subdivided into chronic or episodic based upon their presence more or less than 1 year. The common pathophysiology relates to activation of the “trigeminal-parasympathetic reflex”.

Table 1 compares the clinical features of these three entities.

Cluster

With an estimated prevalence of 0.1-0.9% of the general population, cluster headache is an uncommon primary headache. Reports indicate the onset of cluster headaches occurs between 20-50 years of age, with a slight male predominance [22]. Approximately 7% of first-degree relatives of those with cluster headaches also suffer from the disorder, and studies have demonstrated a 100% concordance in five pairs of identical twins [23]. There are few case series regarding cluster headaches in the literature and only isolated reports of cluster headache in pediatric patients.

Often described as excruciating, boring pain, cluster headaches are strictly unilateral and localize to the temporal and periorbital regions. In contrast to migraine headaches, cluster headaches do not become bilateral or switch sides during an attack. Headaches may, however, occur on opposite sides with different cycles. Although those with cluster headache rarely describe migraine symptoms such as nausea, vomiting, or photophobia, many experience autonomic features such as ipsilateral rhinorrhea and tearing nasal congestion [24]. Attacks are relatively brief, lasting from 15 minutes to 3 hours. Headaches longer than 3 hours should raise questions as to whether cluster headache is the correct diagnosis.

Most patients experience from one to several attacks per day occurring in cycles or clusters that last from weeks to months followed by spontaneous pain-free periods that last from 6 months to 2 years [23]. Cluster headaches may also have circadian and circannual components. Many
report that headaches often occur at night or toward the end of a sleep cycle. Similarly, there is a predilection of cycle onset within several weeks of the summer or winter solstices. There is limited information regarding the prognosis of cluster headaches. Like migraines, however, there is a decline in the prevalence of the disorder after age 50, and chronic headaches are rarely observed in individuals over age 70 years [24].

The management of cluster headaches has been the subject of two recent reviews, dividing management into acute measures and preventative strategies [23,24]. While a wide variety of treatments are proposed for the treatment of cluster headaches, few double-blind, placebo-controlled studies exist. Acute measures include oxygen (100% at 8-10 L/min for 15 minutes), lidocaine (4% aqueous drops intranasally), olanzapine (2.5-10 mg orally at onset), dihydroergotamine (1 mg intravenously, subcutaneously, or intramuscularly at onset with repeated doses), or sumatriptan (6 mg subcutaneously at onset). Inhaled 100% oxygen has been demonstrated to be a successful abortive treatment; however, access to this therapy poses clear limitations in its use. For adults, there are controlled, masked data (Class I) demonstrating efficacy of subcutaneous sumatriptan acute relief of cluster headache [22-24].

Because attack periods can occur repeatedly for weeks to months, suppressive or preventative therapies are essential to the management of cluster headache. Corticosteroids (prednisone 40 mg/day for a 3-5 day pulse followed by 3-week gradually withdrawn) are valuable because they can rapidly suppress attacks. Other options include: ergotamine tartrate (2 mg at bedtime), sumatriptan (100 mg three times daily for up to 7 days), naratriptan (2.5 mg twice daily for 7 days), lithium (300 mg two to four times daily), verapamil (80-240 mg three times a day), sodium valproate (250-1000 mg twice daily), topiramate (25-200 mg twice daily), or melatonin (10 mg daily). Prednisone is the most commonly employed agent for short-term pulses to suppress attacks. Verapamil may be the most commonly used agent for maintenance preventive therapy. Histamine “desensitization” or combination approaches are used for patients with refractory symptoms and, infrequently, surgical ablation may be indicated for medically intractable patients [22-26].

**Cluster Headache in Children and Adolescents**

Information regarding cluster headaches in children is limited. The prevalence of childhood onset of cluster is estimated to be 0.1%. One large Scandinavian series found the prevalence of cluster in 18-year-old men to be 0.09% [25].

One series of 35 patients with onset of cluster headache before age 18 years found that 7 of 35 experienced the onset of symptoms before age 10. All patients met the International Headache Society criteria for either episodic or chronic cluster headache, and all patients had clinical feature and symptoms that were “consistent with adult forms” of cluster. Long-term follow-up determined that 14 of 35 had gradually increasing frequency and duration of their symptoms through adult life [26].

Sporadic case reports of isolated cases exist. One report of recurring pain and irritability with ocular symptoms in a 1-year-old child monitored for several years suggests an onset as early as infancy [27]. Another report describes a

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**Table 1. Key clinical features of cluster, paroxysmal hemicrania, and SUNCT**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Cluster</th>
<th>Paroxysmal Hemicrania</th>
<th>SUNCT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of attacks</td>
<td>5</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Location of pain</td>
<td>Unilateral</td>
<td>Unilateral</td>
<td>Unilateral</td>
</tr>
<tr>
<td></td>
<td>Orbital</td>
<td>Orbital</td>
<td>Orbital</td>
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<tr>
<td></td>
<td>Supraorbital</td>
<td>Supraorbital</td>
<td>Supraorbital</td>
</tr>
<tr>
<td>Duration of attacks</td>
<td>15-180 minutes</td>
<td>2-30 minutes</td>
<td>5-240 seconds</td>
</tr>
<tr>
<td>Frequency of attacks</td>
<td>qod-8/day</td>
<td>5/day</td>
<td>3-200/day</td>
</tr>
<tr>
<td>Autonomic symptoms</td>
<td>CI, L, NC EE, FS, M, P</td>
<td>CI, L, NC EE, FS, M, P</td>
<td>CI, L</td>
</tr>
<tr>
<td>Indomethacin response</td>
<td>negative</td>
<td>positive</td>
<td>negative</td>
</tr>
<tr>
<td>Occurrence in children</td>
<td>++</td>
<td>+++</td>
<td>2 reported</td>
</tr>
</tbody>
</table>

**Abbreviations:**
- CI = Conjunctival injection
- EE = Eyelid edema
- FS = Forehead or facial sweating
- M = Miosis
- NC = Nasal congestion
- L = Lacrimation
- P = Ptosis
- R = Rhinorrhoea
- qod = Every other day

SUNCT = Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing
7-year-old female who suffered from daily attacks located in the right orbital region. These headaches lasted approximately 30 minutes and were invariably unilateral with associated rhinorrhea, conjunctival injection, and agitation. There was no identified precipitating event. Additionally, family history was negative for migraine or cluster headaches. Indomethacin provided no relief from these headaches, but the attacks responded to intravenous prednisolone [22]. Neubauer et al. describe a 12-year-old female with an anhistamine-responsive form of cluster headache who achieved sustained, significant relief for 3 years with astemizole and loratadine [28].

The management of cluster headaches is not well documented in children. No controlled series for either the acute or preventive management of cluster exist. Furthermore, because the inter-cluster period described with children may be prolonged, the utilization of long-term therapy is less desirable.

**Paroxysmal Hemicrania**

Paroxysmal hemicrania is a rare headache with a prevalence of 0.021% [29]. Paroxysmal hemicrania generally begins in adulthood with onset generally in the third decade of life. Characterized by brief, unilateral attacks of intense pain around the superorbital and temporal region, afflicted patients may have from usually 5-6 to as many as 30 attacks per day that last from 2 to 45 minutes. Like other trigeminal autonomic cephalgias, paroxysmal hemicrania is associated with autonomic symptoms such as tearing, conjunctival injection, rhinorrhea, ptosis, and eyelid edema. A key element defining paroxysmal hemicranias is their exquisite sensitivity to indomethacin. This disorder can therefore be distinguished from cluster headaches by their shorter duration, higher frequency, female predominance, and clear response to indomethacin [30].

The prognosis for paroxysmal hemicrania is good, and long-term remission has been reported [31].

Relatively few pediatric cases have been reported in the literature. Children as young as 3 years of age have been described with the disorder. Shabbir and McBee reported two teenaged females (13 and 14 years) with chronic paroxysmal hemicrania (symptoms > 1 year) with repetitive attacks (8/day) of lancinating unilateral pain without mention of autonomic symptoms, both of whom had partial response to indomethacin, but achieved “nearly complete” relief with verapamil [32]. Gladstein et al. reported an 8-year-old male with typical features of chronic paroxysmal hemicrania who responded to indomethacin and was symptom-free for 1 year [33].

The author monitored three adolescents, all males, with paroxysmal hemicrania. All had episodes of brief, excruciating, disabling attacks of retrobulbar pain with at least one autonomic component, and all responded immediately to indomethacin at a dose of 25 mg orally twice a day. One patient discontinued the medicine after 2 months, and attacks recurred within 1 week.

**Short-Lasting Unilateral Neuralgiform Headache Attacks With Conjunctival Injection and Tearing (SUNCT)**

First described in 1978, short-lasting unilateral neuralgiform headache with conjunctival injection and tearing syndrome (SUNCT) is an uncommon trigeminal autonomic cephalgia described in only approximately 50 complete case reports and series, overwhelmingly in adults [31,34,35].

The age of onset of this syndrome generally begins between the ages of 35-65 years, with a range of 10-77 years. There is a high variability in the frequency of attacks, which begin abruptly and last from 5 to 250 seconds [36]. The pain is unilateral in the distribution of the trigeminal nerve, particularly around the orbital, peri-orbital, and temporal region. Patients may have up to 30 attacks per hour, though most report around 5-6 episodes per hour. As quickly as the onset of these headaches, the pain often rapidly abates. Most cases occur in an episodic manner with 1-2 symptomatic periods per year that last from days to months. As the name implies, the most notable autonomic feature of SUNCT syndrome is the conjunctival injection and tearing. Other concomitant features include rhinorrhea and forehead sweating. In contrast to migraine headaches, there is no associated nausea, vomiting, or photophobia. Additionally, SUNCT syndrome lacks the association with Horner’s syndrome that is observed with cluster headaches.

Unlike paroxysmal hemicrania, SUNCT syndrome is unresponsive to indomethacin, and neither oxygen nor other nonsteroidal anti-inflammatory drugs provide relief. Although many therapies that are used to treat other short lasting headaches are ineffective with SUNCT syndrome, partial or complete (open label) success has been achieved with sumatriptan, intravenous lidocaine, and some antiepileptics (e.g., lamotrigine, gabapentin, topiramate, carbamazepine) [34,35].

As with other short lasting headaches, the prognosis of SUNCT syndrome is poorly understood.

Case reports in children are rare. Blatter et al. described “symptomatic SUNCT” in an 11-year-old female with right-sided paroxysmal headaches associated with marked autonomic activation. The symptoms began after a febrile upper respiratory tract infection. The patient described the pain as moderate to severe and located at the retromandibular fossa with radiation to the cheek with up to 20 attacks a day, each episode lasting from 30 to 60 seconds. The pain was always associated with conjunctival injection, tearing of the right eye, and salivation. She denied photophobia and rhinorrhea. A trial of indomethacin decreased the frequency of attacks from 20 to 10 per day; however, the intensity of the pain remained unchanged. A magnetic resonance imaging scan revealed a mass lesion of the right cerebellum near the entry zone of the right trigeminal nerve root. An exploratory operation revealed a growing tumor with prominent vascularity that was
histologically identified as a pilocytic astrocytoma. Sub-total removal of the mass resulted in shorter and less intense pain attacks [37].

Other Primary Headaches

Primary Stabbing Headache

Also known as ice-pick pains, jabs, and jolts, and ophthalmodynia periodica, primary stabbing headache is uncommon in children. The diagnostic criteria are:

A. Head pain occurring as a single stab or a series of stabs;
B. Exclusively or predominantly felt in the distribution of the first division of the trigeminal nerve;
C. Stabs last for up to a few seconds and recur with irregular frequency ranging from one to many per day;
D. No accompanying symptoms;
E. Not attributed to another disorder.

In adult series, the migrating stabs last approximately 3 seconds, occur repetitively over days, and may occur comorbidly with migraine or cluster headaches.

Two reports described idiopathic stabbing headache in children. The first report included 83 patients, each of whom had brief attacks lasting less than 1 second to few minutes, of intense stabbing pain. In follow-up, there was no association with other primary headache syndromes [38]. The second report characterized a group of 23 children, average age 9 years, 12 males and 11 females, with attacks lasting less than 15 seconds, with 60% of attacks being bilateral and 40% unilateral. During a 2-year follow-up, no other comorbid headache was evident. Correlation with the International Headache Society criteria was difficult [39].

Primary Cough Headache (Céphalée d’effort)

Also known as benign cough headache or Valsalva-maneuver headache, the diagnostic criteria are:

A. Headache fulfilling B and C;
B. Sudden onset, lasting 1 second to 30 minutes;
C. Brought on by and occurring only in association with coughing, straining, and/or Valsalva maneuver;
D. Not attributed to another disorder.

There are no reported series of primary cough headache in children. The cause is thought to be related to brief increases in intracranial pressure that accompany coughing, sneezing, laughing, straining, or performance of the Valsalva maneuver. There are reports attributing cough headache to increases in intraocular pressure [40]. The first author (D.W.L.) has identified this entity in many children with cystic fibrosis and other chronic pulmonary diseases.

Structural processes, particularly those referable to the “crowded” posterior fossa, brain tumors, Chiari malformation, syringobulbia, and vascular malformations must be excluded [41-43]. The first attack after straining must also raise suspicions for subarachnoid hemorrhage, although accompanying meningeal signs ought to be present. Favorable response to indomethacin and beta-blockers has been reported [44].

Primary Exertional Headache

Primary exertional headache likely represents a similar spectrum of effort-induced headache. Exertional headache is pulsing in quality, lasts 5 minutes to 48 hours, and is brought on by or occurs during physical exertion. This entity must be viewed within the spectrum of exertional migraine if autonomic features are present.

Management options may include pre-participation medications (nonsteroidal anti-inflammatory drugs, “triptan”) or indomethacin.

Primary Headache With Sexual Activity

This condition (also known as coital headache, sexual headache, benign vascular sexual headache) also falls within the spectrum of exertional headaches. The diagnostic criteria are:

A. Dull ache in the head and neck associated with awareness of neck and/or jaw muscle contraction;
B. Occurs during sexual activity and increases with sexual excitement.

The typical duration is 1 minute to 3 hours.

A separate and distinct form of headache related to sexual activity is orgasmic headache wherein the patient experiences a sudden, explosive headache at the moment of orgasm. As with exertional headache, the first attack while straining during intercourse must also raise suspicions for subarachnoid hemorrhage or arterial dissection [45].

There are no reported cases of sexual activity headache in children; however, the first author (D.W.L.) has diagnosed this entity in sexually active teenagers and teenage males during masturbation.

A recent report of 51 patients with headache associated with sexual activity found a mean age of 39 years, but two peaks of age, one of which was between the ages of 20-24 years (n = 13) approaching the late adolescent years. A male predominance was found (3:1), and the mean duration of headache was 4 hours. Three subsets were identified: (1) a dull pain, which slowly intensifies as sexual excitement increases; (2) an explosive subtype, which occurs suddenly (“vascular-type”); (3) postural headache which begins after orgasm [46].

Hypnic Headache

Also known as “alarm clock” headache, this uncommon entity is characterized by attacks of dull, bilateral pain that
always awakens the patient from sleep. By definition, the entity occurs first after 50 years of age, so it is not discussed further here except to reinforce the idea that headaches which awaken the patient from sleep warrant investigation for increased intracranial pressure.

**Primary Thunderclap Headache**

Thunderclap headache represents the explosive onset of high-intensity headache mimicking rupture of a cerebral aneurysm. The diagnostic criteria are as follows:

A. Severe head pain;
B. Both of the following:
   1. sudden onset reaching maximum intensity in <1 minute
   2. lasting 1 hour to 10 days
C. Does not recur regularly over subsequent weeks or months;
D. Not attributed to another disorder.

To establish this diagnosis, neuroimaging and cerebrospinal fluid examination must be normal.

There are no reported series in children or adolescents. The incidence is 43 per 100,000 patients >18 years of age, but no data exist for the pediatric age range [47]. The International Headache Society states that “evidence that thunderclap headache exists as a primary condition is poor.” Therefore, exhaustive efforts must be undertaken to exclude other conditions such as intracranial hemorrhage, venous thrombosis, vascular malformation, arterial dissection, central nervous system angiitis or angiopathy, colloid cysts of the third ventricle, cerebrospinal fluid hypotension, acute sinusitis, barotrauma, or pituitary apoplexy [1].

**Cranial Neuralgias**

The cranial neuralgias represent a group of disorders, uncommon in children, in which the patients experience brief attacks of excruciating pain localized to a particular anatomic distribution. All represent diagnoses of exclusion with particular attention to posterior fossa neoplastic, demyelinating, or inflammatory processes.

Rare entities such as supraorbital neuralgia, superior laryngeal neuralgia, nasociliary (Charlin’s neuralgia), and nervus intermedius neuralgia will not be discussed.

Ophthalmoplegic migraine has recently been added to the group of cranial neuralgias, but will not be addressed in this report.

**Trigeminal Neuralgia**

Also known as “tic douloureux,” classic trigeminal neuralgia is characterized by brief, shock-like pain limited to one or more distributions of the trigeminal nerve, not crossing the midline. The most commonly affected single division is the mandibular branch. The pain is commonly precipitated by trivial stimuli such as washing, shaving, talking, or tooth brushing. The pain may occur spontaneously and may cause a spasm or tic-like movement of the adjacent facial muscles.

Posterior fossa abnormalities including tumors, inflammatory processes, vascular malformations (“neurovascular compression”), Chiari I malformation, and demyelinating disease must be considered in the differential diagnosis for trigeminal neuralgia.

The diagnostic criteria are:

A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C;
B. Pain has at least one of the following characteristics:
   1. intense, sharp, superficial, or stabbing
   2. precipitated from trigger areas or by trigger factors
C. Attacks are stereotyped in the individual patient.

Trigeminal neuralgia occurs in approximately 1 in 25,000 in the general population and is uncommon before the third decade, with only 1% of the cases occurring before 20 years of age. Four reports were found regarding trigeminal neuralgia in children (Table 1) [48-52]. Perhaps the key point is the high proportion with defined organic pathology. The medical management of choice is carbamazepine [53]. Surgical decompression may be necessary in selected instances.

**Glossopharyngeal Neuralgia**

This neuralgia is characterized by intense attacks of pain in the ear, base of the tongue, tonsils, or angle of the jaw, the sensory distribution of the auricular and pharyngeal branches of the vagus nerve. Common precipitants include swallowing, coughing, or talking.

The diagnostic criteria include:

A. Paroxysmal attacks of facial pain lasting from a fraction of a second up to 2 minutes and fulfilling B and C;
B. Pain has all of the following characteristics:
   a. Unilateral location
   b. Distribution within the posterior part of the tongue, tonsillar fossa, pharynx, or beneath the angle of the jaw or ear
   c. Sharp, stabbing pain
   d. Precipitated by swallowing, chewing, talking, coughing, or yawning;
C. Attacks are stereotyped in the individual patient;
D. There is no clinically evident neurologic disorder.

A large, population-based study in Rochester, Minnesota from 1945 to 1984 found that the annual incidence of glossopharyngeal neuralgia was 0.7/100,000 with no sex predilection. This study suggested that glossopharyngeal neuralgia is “generally a mild disease,” with single attacks being common and the annual recurrence rate for second episodes low at 3.6%. Only one fourth of the cases required surgical intervention [54]. There is an extensive
literature regarding cardiac syncope in association with glossopharyngeal neuralgia [55]. This association may be due to the close anatomic relationship between IX and X (vagus) cranial nerves as they exit through the jugular foramen.

Reports in children are rare. Five references were found. One described a 13-year-old female with glossopharyngeal neuralgia who presented with episodes of paroxysmal pain in the right ear from infancy. Magnetic resonance angiography revealed a prominent, looping, right posterior inferior cerebellar artery, compressing the right glossopharyngeal and vagal nerve complex as it exited from the medulla [56]. Another report describes glossopharyngeal neuralgia following amygdalectomy or tonsillectomy [57]. A third report found an association with Chiari I malformation [58,59]. “Otalgie” glossopharyngeal neuralgia in a 13-year-old male was refractory to medical management and required cervical section of the glossopharyngeal nerve and its tympanic branch to provide complete relief of symptoms [60].

Occipital Neuralgia

Not uncommon in children, occipital neuralgia is characterized by intense, jabbing painful episodes localized to the distribution of the greater or lesser occipital nerve in the occipital region. An emergency department-based study identified 12 patients with occipital neuralgia whose symptoms, in addition to occipital pain, included visual disturbances (76%), dizziness (50%), nausea (42%), scalp paresthesias (33%) and tinnitus (33%). In this study, 80% of the patients experienced “significant relief” with local anesthetic injection [61].

Care must be taken to exclude anatomic abnormalities of the upper cervical region and posterior fossa.

The diagnostic criteria are as follows:

A. Paroxysmal stabbing pain with or without persistent aching between paroxysms, in the distribution of the greater, lesser, or third occipital nerves;
B. Tenderness over the affected nerve;
C. Pain is eased temporarily by local anesthetic block of the nerve.

There is one case series in children or adolescents, but from the pre–computed tomography era [62].

Neck-Tongue Syndrome

Neck-tongue syndrome is an unusual clinical entity related to occipital neuralgia, characterized by brief episodes of intense upper cervical or occipital pain accompanied by ipsilateral numbness of the tongue precipitated by movement or rotation of the head. The attacks are brief, stabbing pain, lasting only seconds up to 1 minute, located unilaterally in the upper neck or occipital region, accompanied by transient paresthesias or numbness of the tongue, ipsilateral to the pain and may include lingual “pseudo-athetosis,” dysarthria, and lingual paralysis.

The proposed mechanism was related to irritation of the C2-3 root, with tongue involvement due to afferent impulses traveling from the lingual nerve via the hypoglossal nerve to the C2 root [63].

The diagnostic criteria are:

A. Pain lasting seconds or minutes, with or without simultaneous dysesthesia, in the area of distribution of the lingual nerve and second cervical root and fulfilling B and C;
B. Onset of pain is acute;
C. Pain is commonly precipitated by sudden turning of the head.

Nearly 25 children and adolescents (ages 8-15 years) have been reported with neck-tongue syndrome. The majority of children (79%) have no identifiable anatomic abnormalities, whereas about two thirds of adults with neck-tongue syndrome have cervical abnormalities such as ankylosing spondylitis, degenerative disc disease, or osteoarthritis. A benign, familial (autosomal dominant) pattern has been observed in approximately five pedigrees [63].

Once structural pathology is excluded, the management of neck-tongue syndrome includes conservative treatments, with avoidance of trauma coupled with nonsteroidal anti-inflammatory drugs and, if necessary, agents (e.g., carbamazepine, gabapentin) to limit neuropathic pain.

Cold-Stimulus Headache (Ice Cream Headache or “Brain Freeze”)

A sudden, intense, brief (<5-minute) frontal or frontal vertex headache after ingestion or inhalation of cold substances is extremely common in children, but there is no case series reported. The substances may be liquid, solid, or gaseous. Popsicles, ice cream, snow cones, crushed ice, and ice slurries seem to be the most common offenders.

The diagnostic criteria are:

A. Acute frontal nonpulsatile headache fulfilling C and D;
B. Cold stimulus to the palate or posterior pharyngeal wall resulting from ingestion of cold food, drink, or inhalation of cold air;
C. Headache develops immediately and only after cold stimulus;
D. Headache resolves within 5 minutes after removal of cold stimulus.

This phenomenon has been known to precipitate migraine-like attacks.
Figure 1. Recurrent headache with normal neurologic examination in children and adolescents.‡

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*Indomethacin responsive* Prompt decrease in headache within 1-2 days following introduction of 25 mg bid up to 50 tid

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*Autonomic symptoms*
- Conjunctival injection and/or tearing (C, PH, Sumct)
- Nasal congestion (C, PH)
- Eyelid edema (C, PH)
- Facial sweating (C, PH)
- Miosis or ptosis (C, PH)
- C= cluster
- PH= paroxysmal hemicrania

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*No history to suggest the recent onset of severe headache, change in the type of headache or associated features that suggest neurological dysfunction.*

*Absence of focal findings, signs of increased intracranial pressure, significant alteration of consciousness and/or seizures.*
Conclusion

While migraine and the various forms of chronic daily headache are the most frequent headache syndromes referred for neurologic consultation, other primary headache syndromes may begin in the pediatric years. Figure 1 provides a clinical decision tree for pediatric headache incorporating the latest International Classification of Headache Disorders criteria.

Tension-type headaches are mild to moderate in intensity, often frontal in location, last from minutes to hours, and lack the autonomic and disabling features of migraine. The diagnosis may be made on clinical grounds. Behavioral measures and simple analgesics may be the most useful therapies, though no masked, controlled trials have been reported.

The majority of the “other” primary headaches and cranial neuralgias are brief attacks, with or without autonomic components. Some have characteristic periodicity (e.g., cluster and trigeminal autonomic cephalgias) or clear precipitating phenomena (e.g., activity, cold, head turning, cough, awakening), whereas others have specific locations (e.g., occipital, oropharyngeal). Some have dramatic, near-diagnostic, responsiveness to indomethacin (e.g., paroxysmal hemicrania). Because these “other” entities are uncommon and may be symptomatic of underlying organic pathology, ancillary diagnostic testing may be considered.

References


