Chapter 9
Episodic Syndromes that May Be Associated with Migraine, Pediatric Tension-type Headache, Chronic Daily Headache Syndromes in Children and Pediatric Idiopathic Intracranial Hypertension

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Introduction

This chapter is divided into four sections on pediatric headache and includes episodic syndromes that may be associated with migraine, tension-type headache (TTH), and chronic daily headache (CDH) syndromes. A brief overview on idiopathic intracranial hypertension (IIH) is also included, as this represents one of the most common secondary headaches in children and adolescents. The trigeminal autonomic cephalalgias (TACs) are mentioned briefly, as these are covered in more detail in other chapters of this book.

Episodic Syndromes that may be Associated with Migraine

The term "childhood periodic syndromes" was first introduced by Wyllie and Schlesinger in 1933 to describe stereotypical, recurrent episodes of vomiting, headache, and/or abdominal pain, separated by symptom-free intervals. Several years later, Dr. Charles Barlow described how these periodic syndromes were common precursors of migraine.

The International Classification of Headache Disorders, third edition, beta version (ICHD-3) includes the category *Episodic syndromes that may be associated with migraine (1.6)*. Within this grouping are benign paroxysmal vertigo (BPV), benign paroxysmal torticollis (BPT), as well as a new subcategory, *Recurrent gas-*

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Table 9.1 Secondary causes to be considered in childhood periodic syndromes

• *Central nervous system*
Increased intracranial pressure
Posterior fossa mass
Epilepsy
Infection (meningitis, encephalitis)
• *Inborn errors of metabolism*
Organic acidemias
Urea cycle defects
• *Mitochondrial disorders*
• *Acute intra-abdominal disease*
Bowel obstruction
Ureteropelvic junction obstruction
Hepatitis
Pancreatitis

trointestinal disturbance, covering cyclical vomiting syndrome (CVS) and abdominal migraine (AM).

Although more frequently encountered in the pediatric population, these disorders may also occur in adults. They are considered diagnoses of exclusion (see Table 9.1). Inborn errors of metabolism such as organic acidemias, urea cycle defects, mitochondrial disorders, increased intracranial pressure (ICP), posterior fossa tumors, and acute intrabdominal pathology may present in a similar fashion. Therefore, a thorough evaluation for these disorders is necessary in order to avoid missing causes that untreated could result in significant morbidity and mortality.

Recurrent Gastrointestinal Disturbance

Previously known as functional abdominal pain, chronic abdominal pain, functional dyspepsia, and irritable bowel syndrome, recurrent gastrointestinal disturbance is characterized by recurrent attacks of abdominal pain that may be associated with nausea and/or vomiting, and may occur intermittently with a predictable pattern or in a more chronic fashion. Attacks may be associated with migraine headache as well. The diagnostic criteria for this disorder are summarized in Table 9.2.

Cyclical Vomiting Syndrome

First described by Heberden in 1806, cyclical vomiting syndrome (CVS) is characterized by recurrent episodes of nausea, vomiting, and lethargy separated by symptom-free intervals. Its estimated prevalence is 0.4–1.9%, with girls more affected than boys (see Table 9.3 for diagnostic criteria). Patients of northern European ancestry are more frequently affected.

Table 9.2 Recurrent gastrointestinal disturbance, ICHD-3 diagnostic criteria

A. At least five attacks characterized by abdominal pain and/or discomfort, nausea, and/or vomiting
B. Normal gastrointestinal examination and evaluation
C. Not secondary

Table 9.3 Cyclical vomiting syndrome, ICHD-3 diagnostic criteria

A. ≥5 attacks fulfilling criteria B and C
B. Stereotypical attacks for each patient, with predictable recurrence
C. All of the following:
1. Nausea and vomiting occurring at least four times per hour
2. Attacks lasting 1 h–10 days
3. Attacks occur at least 1 week apart
4. No interictal symptoms
5. Not secondary

Attacks are more common in the early morning hours or soon after waking up. Patients develop multiple episodes of emesis per hour, associated with nausea, retching, pallor, and in some cases, dysautonomia. Symptoms peak between 1 and 2 h after onset, but an individual attack may last from 6 to 48 h. Following the ictal phase, children often fall asleep for several hours, to wake up later back to baseline.

CVS is more frequent in young children between 4 and 5 years old, but is increasingly recognized in adults. Eighty-seven percent of patients have a positive family history of migraine. Episodes tend to subside by 10 years of age, but approximately 75 % of affected patients develop migraine later on. As noted, in some patients, symptoms persist into adulthood.

Although episodes may occur infrequently in some patients, they are very disabling for the child, leading to frequent hospitalizations, emergency room visits, and school absences. Therefore, preventive therapy should be strongly considered for these patients, even if the episodes are infrequent.

Over the last decade, CVS has also been recognized as an important clinical manifestation of other disorders, such as neurometabolic and mitochondrial disease. Some authors have referred to this form as *CVS Plus* (CVS+), in which patients not only manifest stereotypical cyclical vomiting, but may also have additional symptoms such as neuromuscular disease, cognitive delay, or seizures. Patients with CVS+ may develop clinical manifestations earlier in life compared to patients with the migraine-related CVS form and should be thoroughly evaluated to rule out underlying neurometabolic disease (Table 9.4).

In 2008, the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition published a consensus statement for the diagnosis and management of CVS. This publication highlights symptoms and patient characteristics that may increase the risk of a serious underlying disorder as opposed to idiopathic CVS and are summarized in Table 9.5.

Table 9.4 Evaluation of patients presenting with cyclical vomiting syndrome

Serum electrolytes, glucose
Upper GI series
Abdominal US
Long chain fatty acids analysis

GI gastrointestinal, *US* ultrasound

Table 9.5 When to consider underlying organic disease manifesting as cyclical vomiting syndrome

- Patients less than 2 years old are more likely to have surgical or metabolic disease
- Bilious vomiting
- Severe abdominal pain or tenderness
- Attacks precipitated by intercurrent illness, fasting, and/or high-protein meal
- Abnormal neurological evaluation
- Progressively worsening attacks and or conversion into a continuous pattern without symptom-free interval

Abdominal Migraine

Abdominal migraine (AM) was first described by Buchanan in 1921 as recurrent attacks of abdominal pain without headache. Episodes are characterized by disabling abdominal pain, dull in quality, with a location that is either periumbilical or diffuse. Children may also exhibit other symptoms classically associated with migraine such as pallor, flushing, dark circles around the eyes, and anorexia (Table 9.6). Of note, according to the most recent ICHD-3 criteria, episodes must last a minimum of 2 h (treated or unsuccessfully treated). Vomiting may be present, but it is less severe than in CVS. Visual aura may also occur. Headache does not occur during these attacks. Patients are symptom-free between attacks.

Table 9.6 Abdominal migraine, ICHD-3 diagnostic criteria

A. ≥5 attacks of abdominal pain, fulfilling criteria B–D
B. Pain has at least two of the following features:
   1. Midline location, periumbilical, or poorly localized
   2. Dull or described as “sore” in quality
   3. Moderate to severe in intensity
C. At least two of the following during attacks:
   1. Anorexia
   2. Nausea
   3. Vomiting
   4. Pallor
D. Spells last 2–72 h whether treated, or unsuccessfully treated
E. Symptom-free intervals
F. Not secondary

Frequent triggers include psychological (excitement) and physical stress (illness). Motion sickness is frequently reported.

AM is more common in girls. The age of onset is between 3 and 10 (mean 7 years) with an estimated prevalence of 2.4–4.1%. A family history of migraine is common.

Benign Paroxysmal Vertigo

Benign paroxysmal vertigo (BPV) was first described by Basser in 1964; it is characterized by abrupt loss of balance, vertigo, and even falls. The prevalence is 2–2.6%, with equal distribution between boys and girls.

At the beginning of the episode, children may appear frightened, while trying to hold on to furniture or to another person to avoid falling. They refuse to walk and want to lie still. Older children may describe dizziness and nausea.

Associated symptoms include nystagmus, pallor, nausea, diaphoresis, phonophobia, and photophobia. Severe vomiting may also occur. There is no loss of consciousness.

Attacks are typically brief, lasting less than 5 min in most cases, although episodes up to 48 h have been described. They occur once every 1–3 months, with decreasing frequency with advanced age. Movements that stimulate the labyrinth such as swings and roundabouts may trigger the episodes.

Onset is between 2 and 4 years, and BPV symptoms of childhood resolve in most cases by 5 years. A positive family history of migraine is common; Abu-Arafeh and Russell also described a higher prevalence of migraine in children with BPV. It has also been suggested that BPV constitutes a precursor of migraine with brainstem aura. The diagnostic criteria for BPV are summarized in Table 9.7.

BPV is a diagnosis of exclusion; differential diagnosis includes posterior fossa pathology and episodic ataxia, among others. It must also be differentiated from migraine-associated vertigo, which occurs in older children. Later in life, children

Table 9.7 Benign paroxysmal vertigo, ICHD-3 diagnostic criteria

A.	≥5 of attacks fulfilling criteria B–C
B.	No aura or prodrome, precipitous onset vertigo, peak at onset, resolving in minutes to hours without loss of consciousness
C.	At least one of the following:
	1. Nystagmus
	2. Ataxia
	3. Vomiting
	4. Pallor
	5. Fearfulness
D.	Normal interictal audiometry, vestibular testing, and exam between attacks
E.	Not secondary

may develop CVS or migraine. Of note, a normal EEG is no longer required as part of the diagnostic criteria.

Benign Paroxysmal Torticollis

First described by Snyder in 1969, Benign Paroxysmal Torticollis (BPT) is characterized by sudden onset, recurrent dyskinesias involving the neck. During the attack, there is an abnormal rotation of the head and neck toward the affected side, which may be accompanied by vomiting and ataxia. Other symptoms frequently encountered in migraine such as pallor, drowsiness, photophobia, and epiphora may occur.

Each episode may last hours to days and resolves spontaneously without sequelae. Patients develop symptoms between 2 and 8 months of age that resolve by age 3–5 years. The frequency and severity of attacks decrease as children get older. It is more frequently encountered in girls, and a family history of motion sickness and migraine is common.

BPT shares several features with migraine, including its paroxysmal nature, female preponderance, and associated migrainous features. Over the last decade, families with clustering of migraine, other episodic disorders that may be associated with migraine, BPT, and underlying calcium channelopathy CACNA1A mutations have been described, providing evidence that BPT may represent a migraine precursor. BPT is now included in the ICHD-3 as part of the episodic syndromes that may be associated with migraine. Diagnostic criteria for BPT can be found in Table 9.8.

Summary of Childhood Periodic Syndromes

Tables 9.9 and 9.10 provide summaries of the differences and similarities between the different periodic syndromes.

Table 9.8 Benign paroxysmal torticollis, ICHD-3 diagnostic criteria

A.	Recurrent attacks in a young child fulfilling:
B.	Head tilt to one side (variable side), sometimes with slight rotation, remitting spontaneously after minutes to days
C.	At least one of the following associated symptoms or signs:
	1. Pallor
	2. Irritability
	3. Malaise
	4. Vomiting
	5. Ataxia
D.	Interictal normal exam
E.	No secondary cause

Table 9.9 Summary of episodic syndromes that may be associated with migraine

Disease	Age of onset	Prevalence (%)	Predominant symptoms	Duration	Age of resolution (years)	% of patients who develop migraine
CVS	4–5 years	0.4–1.9	Multiple episodes of emesis/hour	1–6 h	10	75
BPV	2–4 years	2–2.6	Vertigo, imbalance	5 min	5	75
AM	3–10 years	2.4–4.1	Epigastric/diffuse abdominal pain	1–72 h	10	70
BPT	2–8 months		Torticollis	Hours–days	3–5	To be determined

CVS cyclical vomiting syndrome, *BPV* benign paroxysmal vertigo, *AM* abdominal migraine, *BPT* benign paroxysmal torticollois

Table 9.10 Episodic syndromes that may be associated with migraine (childhood periodic syndromes): Clinical Pearls

- Female predominance
- Strong family history of migraine and motion sickness
- Sudden onset and spontaneous resolution without sequelae
- Symptoms associated with migraine may be seen (pallor, flushing, nausea, vomiting, photophobia)
- Normal physical and neurological evaluation
- 70–75% of patients develop migraine later in life

Pediatric Tension-Type Headaches

Episodic Tension-Type Headache

Tension-Type Headache (TTH) is considered to be the most frequent headache type encountered in adult series. It is also estimated, based on population studies, to occur in 10–72% of school-age children, although clinic-based studies have reported an incidence of approximately 30%.

Its true prevalence may be underestimated, as many patients with TTH may not need to seek medical attention and, since most studies are done in school-age children, the very young patients (below 7–8 years of age) are not accounted for. Episodic TTH (ETTH) is equally prevalent in boys and girls before puberty, but becomes more prevalent in young women later on.

The pain is usually described as holocephalic or bilateral, pressure-like in quality, of mild to moderate severity. There are no other associated symptoms. Patients can often continue their usual activities and may not take medication for the pain.

Patients with ETTH often have comorbid mood disorders such as anxiety and depression. ETTH may also coexist with migraine in some patients (6%), and the predominant entity may alternate from time to time. The clinical features of ETTH are summarized in Table 9.11.

Table 9.11 Clinical features of pediatric episodic tension-type headaches

Location	Bilateral, holocephalic
Quality	Pressure-like
Intensity	Mild to moderate
According to frequency	
- Infrequent	1 day/month
- Frequent	1 day/month to <15 days/month
Average duration	30 min. –7 days

Pediatric chronic tension-type headache (CTTH) will be covered in the next section.

The Pediatric Chronic Daily Headaches

Chronic Daily Headache (CDH) is not an ICHD-3 term term, but is one of the most common headache entities resulting in referral to a pediatric neurologist or headache specialist in pediatrics. It often results in significant disability, school absenteeism, and economic burden due to frequent emergency room and office visits, hospitalizations, and unnecessary testing.

CDH, as a primary headache disorder, often results in significant anxiety in patients and parents, as the symptom persists, but no specific etiology is identified. Therefore, one of the keys to successful management of these patients is the ability of the treating health-care provider to be able to provide *confident reassurance* and formulate a comprehensive, often multidisciplinary, treatment plan after a careful evaluation of the patient has been accomplished.

All subtypes of CDH are characterized by being present for at least 3 months, with headache occurring in more than 15 days per month. It may be intermittent or continuous; exacerbations and remissions may occur, and it may be chronic from the time of onset (as in new daily persistent headache, NDPH) or evolve from different forms of primary episodic headache (such as migraine or TTH).

CDH is often accompanied by other symptoms, such as anxiety, depressed mood, dizziness, and fatigue, among others. It has a significant impact in quality of life, as these patients often miss school and withdraw from academic and social activities. Complicating factors, such as medication overuse, also need to be addressed.

Key concept: All forms of CDH may be complicated by medication overuse.

The most frequently encountered forms of CDH in children include CTTH, transformed migraine (TM; now known as chronic migraine, CM), and NDPH. The clinical characteristics of each subtype are summarized below.

Chronic Tension-Type Headache

This type of headache often evolves from ETTH. As in ETTH, it is characterized by bilateral or holocephalic pressure-like pain, of mild to moderate severity. Migrainous features are absent. It may be intermittent (with most episodes lasting at least 4 h), or continuous, occurring at least 15 days per month for 3 months. CTTH in adolescents has been associated with unhealthy habits such as smoking, obesity, and a sedentary life style, and these have been have been recently demonstrated as independent risk factors for this disorder.

Transformed Migraine/Chronic Migraine

It is not infrequent, when evaluating children and adolescents with CDH, to encounter "two different types of headache" in their description of symptoms. Patients often describe a daily headache, of moderate to severe intensity that has exacerbations, sometimes several times per month. These worse headaches are often accompanied by classic migrainous features, such as nausea, vomiting, photophobia, and phonophobia; auras may also occur.

The term "transformed migraine" was initially applied to those patients with a history of episodic migraine who later developed CDH with migrainous features accompanying exacerbations. The ICHD-3 includes the term CM to describe a primary CDH with a link to previous episodic migraine.

New Daily Persistent Headache

Increasingly recognized in clinical practice, New Daily Persistent Headache (NDPH) classically presents in a patient without a prior history of frequent headache as "a headache occurring, out of the blue, that just won't go away."

NDPH manifests as a daily headache with onset from a particular day, within 24 h according to ICHD-3. In adults, half of the patients in one case series had tension-type features and the other half had migrainous features. The ICHD-3 does not characterize the phenotype other than to require the CDH to be primary and have the abrupt onset on the specific day.

The key feature is that patients with NDPH can remember the exact date on which the headache starts. This abrupt onset is the most useful aspect of presentation that helps distinguish NDPH from other forms of CDH.

Epidemiological studies have shown NDPH to be more common in children and adolescents when compared to adults and more frequent in females as well. It is not uncommon to identify certain personality traits in these patients. Adolescent females with NDPH are typically "high achievers," overinvolved in extracurricu-

Table 9.12 Chronic daily headache subtypes in children and adolescents

Feature	CTTH	TM/CM	NDPH
Pattern	>4 h < continuous	Daily, continuous	Should be continuous almost from onset
Evolution	From ETTH	From episodic migraine	Daily and persistent from onset
Associated symptoms	Nausea, pericranial tenderness	Migrainous features during exacerbations	Nausea
Precipitating factors	Stress	Same as for migraine	Viral illness, surgery
Prior history of headache	Yes	Yes	No

CTTH chronic tension-type headache, *TM* transformed migraine, *CM* chronic migraine, *NDPH* new daily persistent headache

lar activities. Patients may experience significant disability, with withdrawal from daily activities. Medication overuse, depression, and anxiety can be consequences of this disorder.

A study by Mack et al. in 2004 in pediatric patients with NDPH found that a physical stressor can be identified in 88 % of patients preceding the onset of headache. The most common identified triggers included (in order of frequency): febrile illness (with Epstein–Barr viral infections being most common), minor head trauma, and extracranial surgery. Although only 12 % of patients had no identifiable precipitant in this series, similar studies in the Japanese literature have reported no identifiable trigger in up to 65 % of patients.

Table 9.12 summarizes the most common types of CDH encountered in the pediatric population.

Less Frequent Primary Headaches in Children and Adolescents

The Trigeminal Autonomic Cephalalgias (TACs) constitute another group of primary headache disorders. This group encompasses cluster headache, paroxysmal hemicrania (PH), short-lasting unilateral neuralgiform headache attacks (SUNHA), and hemicrania continua (HC). Features in common among this group of disorders are the ipsilateral, autonomic manifestations such as conjunctival injection, lacrimation, and nasal congestion.

Although the TACs account for less than 1 % of primary headache disorders in children, these entities are important to recognize because specific therapy such as indomethacin may lead to significant improvement and even resolution of the headache in some of the TACs (PH and HC). More extensive diagnostic descriptions of the TACs are contained in Chap. 2. Pediatric cases have similar features to adults.

Idiopathic Intracranial Hypertension

Previously known as pseudotumor cerebri, idiopathic intracranial hypertension or IIH was first identified as a neurological disease in the 1900s, in patients who presented with signs and symptoms of raised intracranial pressure (ICP) but in whom no tumor was identified. In the 1930s, when ventriculography was in use, David and Dyke described patients with symptoms of increased ICP and negative ventriculography for tumor. Those who underwent cranial decompression showed improvement in symptoms of raised ICP, but it was also observed that papilledema itself could persist for several months. The first diagnostic criteria for this syndrome can be attributed to Dandy and are included in Table 9.13.

Although the exact pathogenesis of this disorder is not entirely clear, it has been postulated that alterations in CSF flow may lead to a buildup of fluid causing increased ICP. With the advent of magnetic resonance venography (MRV), venous sinus stenosis has been identified in some patients, leading to an impaired reabsorption of cerebrospinal fluid (CSF). In some adult patients who are refractory to medical management, stenting of the venous sinuses has been performed. Certain medications, in particular the tetracyclines, may also have toxic effects on the arachnoid villi, resulting in impaired CSF reabsorption.

Epidemiology of Idiopathic Intracranial Hypertension

IIH most frequently affects females, with a female to male ratio of 4:1. Ninety percent of these patients are overweight. Although the exact prevalence in the pediatric population is not known, it has become an increasing health problem in children as obesity rates have risen, causing great concern in light of the potential for permanent visual loss.

Clinical Manifestations of Idiopathic Intracranial Hypertension

Headache is the most common presenting symptom of IIH and affects 90 % of these patients. The head pain may have migrainous features such as photophobia, and is typically retro-orbital and bifrontal in location. It is usually worse in the morning, when the patient is supine, and is aggravated by Valsalva maneuvers.

Table 9.13 Dandy's initial diagnostic criteria for idiopathic intracranial hypertension

- Signs and symptoms of increased intracranial pressure (headache, papilledema, transient visual obscurations)
- No localizing neurological signs although cranial nerve VI palsies may occur
- Normal cerebrospinal fluid analysis
- Normal to small ventricles

Transient visual obscurations are present in 70% of patients, and other visual disturbances such as diplopia, blurred vision, and constriction of peripheral vision may also occur. Pulsatile tinnitus, often described by patients as a "whooshing sound" or a sensation of water running, may also be reported. In children, irritability and rarely, facial diplegia may also occur.

Papilledema is almost invariably present in IIH, and is bilateral in 90% of patients. It may also precede the onset of other symptoms such as headache or visual disturbances. *Papilledema is the single most important predictor of visual loss in these patients*.

Visual loss may occur in 10–16% of children at the time of initial presentation. Visual field defects, present in 70–85% of patients, must be carefully looked for. An inferonasal defect and enlargement of the blind spot may be the initial manifestation.

Diagnostic Evaluation of Idiopathic Intracranial Hypertension

When evaluating patients with suspected increased ICP, secondary causes must be excluded first. In the past, when the term pseudotumor cerebri was used, clinicians would use the term "pseudotumor cerebri complex" (PTC) to distinguish between those patients with IIH and those in whom causes other than tumors or space-occupying lesion resulted in raised ICP. Several infectious, vascular, toxic, and metabolic factors are recognized that can mimic IIH or lead to increased ICP, resulting in a clinical syndrome similar to IIH. These are summarized in Table 9.14, with emphasis on those more frequently encountered in the pediatric population.

Table 9.14 Mimics of idiopathic intracranial hypertension

1. Infectious
   a. Otitis media and mastoiditis resulting in venous sinus thrombosis
   b. Lyme aseptic meningitis
   c. Varicella
2. Endocrine
   a. Hypo- and hyperthyroidism, thyroid hormone replacement
   b. Growth hormone replacement
   c. Addison's disease
   d. Malnutrition and refeeding syndrome
   e. Uremia
3. Medications
   a. Tetracyclines, in particular minocycline
   b. Vitamin A and its derivatives
   c. Corticosteroid withdrawal
4. Craniosynostosis

Ancillary Testing in Idiopathic Intracranial Hypertension

Neuroimaging studies should be obtained in all patients suspected of having increased ICP. Magnetic resonance imaging (MRI) with MRV are the preferred evaluations, as the studies may reveal venous sinus stenosis or thrombosis demonstrating both the cause of the patient's symptoms and a potential route to treatment. As a rule, neuroimaging should be negative in IIH patients, eliminating such causes as tumors, abscesses, and other space-occupying lesions. Due to the elevated ICP, findings such as flattening of the posterior pole of the ocular globes, increased CSF signal surrounding the optic nerve sheaths, tortuous optic nerves, flattening of the pituitary gland, an empty sella, and tonsillar ectopia may be identified with imaging in IIH.

All patients with IIH should have formal visual evaluation that includes standardized visual field testing (either Humphrey visual field testing or Goldman perimetry in young patients who cannot complete Humphrey visual fields). Visual acuity and color vision should also be evaluated.

Recently, optic coherence testing (OCT) has also been used to measure the average nerve fiber thickness to detect nerve edema. No standardized values have been established in pediatric patients as of the writing of this chapter.

Laboratory testing including CBC, CMP, ANA titers, and thyroid function studies should also be drawn in all patients. Lyme serology and vitamin A levels should also be obtained if clinical suspicion warrants it.

The diagnosis is confirmed by obtaining a lumbar puncture in the lateral decubitus position to avoid falsely elevated values when the procedure is done with the patient upright.

What Constitutes a Normal Opening Pressure in the Pediatric Population?

For several decades, a normal adult opening pressure was considered to be below 20 cm H_2O. This number was then extrapolated to the pediatric population, but recent studies have revealed that the normal opening pressure can actually be much higher in children.

In 2010, Avery and colleagues conducted a study in pediatric patients aged 1–18 years of age and measured opening pressures in this group. None of these patients had associated conditions or exposure to medications that could alter the CSF opening pressure (OP), and the authors found a mean opening pressure of 19.8 ± 6.8 cm. A subsequent 2011 study by Lee et al. found similar results with a mean OP of 20.3 cm (SD 7.1), which has led to the acceptance of 28 cm H_2O as the upper limit for a normal opening pressure in the pediatric population, significantly higher when compared to adults (25 cm H_2O by ICHD-3).

Table 9.15 Clinical pearl on normal pediatric opening pressure

• Opening pressures up to 28 cm H_2O can be considered normal in children

In 2011, Avery et al. published another study, this time comparing pediatric patients with papilledema on exam and their OP with control subjects without papilledema and their OP. The mean opening pressure in patients with papilledema was 40 cm H_2O, and all but one patient had opening pressures above 28 cm H_2O (Table 9.15). Interestingly, the only patient in the papilledema group with an opening pressure below 28 cm H_2O was a child who had received a renal transplant, and had been recently restarted on steroid therapy. Lastly, CSF analysis should be normal in the IIH patients.

Treatment of Idiopathic Intracranial Hypertension in Children

Acetazolamide remains the mainstay of treatment for patients with IIH. A carbonic anhydrase inhibitor is thought to reduce CSF production, thereby decreasing CSF volume and lowering CSF pressure. Usual dosing ranges from 20 to 25 mg/kg/day in divided doses, although up to 3 g/day may be needed in some patients.

Side effects such as anorexia, nausea, vomiting, and paresthesias may occur. Electrolytes should be carefully monitored in these patients, and supplementation with sodium bicarbonate instituted if needed, as significant metabolic acidosis can occur. Aplastic anemia and Steven–Johnson syndrome have also been rarely reported.

Furosemide, a loop diuretic in doses of 1–2 mg/kg/day in three divided doses in children and 20–40 mg three times daily in adolescents may be used if there are contraindications to acetazolamide or as adjunct therapy.

Topiramate, an anticonvulsant with weak carbonic anhydrase inhibitor effects, has also been used in these patients as second line therapy, but the appropriate doses for IIH have not been well established. Topiramate has the added benefit of being a good headache prophylaxis agent, as it is not infrequent for patients with IIH to have coexistent migraine or TTH as well.

The use of steroids, such as prednisone and methylprednisolone has also been advocated by some, especially in patients with severe visual impairment at the time of presentation. However, these can cause significant side effects as well as the risk of rebound IIH as the steroid is being tapered.

Treatment should be continued until vision normalizes and papilledema has completely resolved. After this, medications need to be slowly tapered with serial ophthalmological exams performed to detect early recurrence.

Surgical Interventions for Idiopathic Intracranial Hypertension

When medical management fails there is impending visual loss, surgical interventions such as ventriculoperitoneal shunting (VPS) and lumboperitoneal shunting may be used as a last resort. In some centers, optic nerve sheath fenestration is also used when vision is threatened. Fenestration is not effective for the headache but preserves the discs.

Prognosis of Idiopathic Intracranial Hypertension

Permanent visual loss is the most feared complication of this disease. Risk factors associated with irreversible visual loss include recent weight gain, severe papilledema, subretinal hemorrhages, decreased visual acuity, visual field loss at the time of presentation, early optic nerve atrophy, and hypertension. Symptom duration, transient visual obscurations, pulsatile tinnitus, headache severity, and the actual number of the opening pressure have not been shown to influence long-term prognosis.

Overall, 10–20 % of patients will have permanent visual impairment despite normalization of the OP and papilledema resolution. Twenty percent of patients will relapse within 3 years of diagnosis, and it is important to keep in mind that relapse may occur even while patients are receiving medical therapy.

Conclusions

- The episodic syndromes that may be associated with migraine are likely precursors to migraine in adulthood. Each has very specific diagnostic criteria
- Childhood daily headaches, as in adult CDH, often have mixed features. Most can be diagnosed as TM or CM, NDPH, or CTTH
- Medication overuse can complicate CDH in the pediatric population
- IIH is an increasingly prevalent headache disorder in children and should be promptly identified and treated to prevent irreversible visual loss
- A normal opening pressure in children and adolescents is considered to be up to 28 cm H_2O
- Acetazolamide remains the first line of treatment in patients with IIH. Furosemide and corticosteroids may be used as adjunctive therapy or second line of treatment

Suggested Reading

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