CHILDHOOD HEADACHE SYNDROMES
(PART II)

Abstract
Objective
Headache is one of the most common reason that children are referred to the Pediatric Neurology Services. It is said that ten percent of children aged 5 to 15 years have migraine. Subsequently, it is essential for clinician to have a thorough, comprehensive and systematic approach to the evaluation and management of the child or adolescent who complains of headache.

This writing aims to explore the symptoms of headache, its epidemiology, classification, appropriate evaluation, differential diagnosis and management. Headaches are divided into primary and secondary categories. Migraine and tension type headaches are prototype of primary headaches without underlying pathology. On the other hand, the type of headache which stems from organic diseases such as brain tumor, increased intracranial pressure, systemic disease, drug toxicity, ear-nose and throat problems are considered secondary.

On the whole, the majority of children with primary headache have two patterns of headache. One is a chronic low-grade and the other is an intermittent disabling headache. The cause of the former is either caffeine or analgesic abuse, and the latter is predominantly migraine.

Traditionally, if a child presents himself with chief complain of headache, caretaker physician begins with history taking followed by thorough physical and neurological examinations.

In the majority of the cases, this initial process leads to a diagnosis or indicate the need for further testing.

Once the diagnosis is made, a management program can be put into place.

Keywords: Headaches, Child, Children, Migraine

Introduction
Different headaches syndromes
The general pediatrician sees both a combination of headache secondary to other conditions such as infectious disorder or systemic disease and primary headache syndromes.

Acute generalized headache is most often seen by general physician or general pediatrician. If acute generalized headache is associated with abnormal neurologic symptoms or signs, then pediatric neurologist will be called. An acute localized headache should arouse concern about a localized pathology such as sinusitis, otitis, ocular, dental or temporal mandibular Joint (TMJ) problems. Astigmatism, refractive error, glaucoma, optic neuritis and orbital cellulitis are rarely the cause of headaches. Headache may follow mild head trauma, which can be one of the many
triggers of migraine, and well-documented trauma can trigger the first attack of migraine (26).

**Acute recurrent headaches**

The migraine syndrome is the classic example of an acute recurrent headache. Attack of migraine is characterized by an episodic, periodic, and paroxysmal throbbing headache which may be unilateral or bilateral. The attacks are separated by pain free intervals. They are often preceded by pallor and behavior change and are often associated with decreased appetite, nausea, vomiting, photophobia and phonophobia. They are frequently relieved by sleep (11).

Migraine surprisingly may begin early in life. Initial complaints are paroxysmal and recurrent abdominal pain, restlessness, head banging or sudden alteration in personality. A history of motion sickness or carsickness can be elicited in approximately two-third of patients (27).

The most common symptoms in children involve: nausea, vomiting, abdominal pain and disturbances of vision, including complete blindness in one eye (amaurosis fugax). A family history of the disorder can be elicited in over one half of the patients and was found in 72% of cases reported by Prensky (28). Other symptoms preceding the headache include numbness and tingling in one arm or over the entire side, hemiplegia, aphasia or apraxia (29).

It is best to think of a patient with migraine and neurologic features as having an underlying neurologic disorder until proven otherwise (11).

**Two types of migraine with and without aura**

Migraine as an acute recurrent headache may be divided into with and without aura (30).

**Common Migraine (Migraine without aura)**

This type of migraine is the most frequent type of migraine in children. The headache which is throbbing or pounding, tends to be unilateral at onset or throughout its duration but may also be bifrontal or located in temporal regions. The headache usually persists for 1-3 hours or may last as long as 72 hours. A characteristic feature of this type of childhood migraine is intense nausea and vomiting. The vomiting which may be more troublesome than headache, may be associated with abdominal pain and fever. These condition may be erroneously confused with surgical abdomen. A positive family history particularly on the maternal side is present in 90% of children who suffer from migraine without aura. Additional symptoms include pallor, photophobia, lightheadedness, phonophobia, osmophobia (aversion to odors) and paresthesia of hands and feet.

**Classic migraine (migraine with aura)**

In this disorder, an aura precedes the onset of the headache. Visual aura are uncommonly described by young children with migraine, but when they occur, they may take the form of blurred vision, scotoma, “photopsia flash of light” fortification spectra (brilliant with zigzag lines) or irregular distortion of objects. Distortion of body image (Alice in wonderland syndrome) may predominate as a prelude to a classic migraine headache.

**Familial Hemiplegic migraine (FHM)**

This condition is considered a migraine aura and transmitted as autosomal dominant trait. It is characterized by the onset of unilateral sensory or motor signs during an episode of contralateral migraine headache. Hemis syndromes are more common in children than in adults and may be characterized by numbness of the face, arm, and leg; unilateral weakness and aphasia. The neurologic signs may be transient or may persist for days. Familial hemiplegic migraine (FHM) is characterized by hemiplegia during the headache and in some kindreds progressive cerebellar atrophy (30). It is said in approximately 20% of patients suffering from FHM, this problem is accompanied by progressive cerebellar ataxia. So far for FHM 2 loci have been mapped to chromosome 19P13 and long arm of chromosome 1q23.

**Basilar artery migraine**

Basilar artery migraine was first described by Bickerstaff (31). It is a recurrent dysfunction which stems from the brain stem, cerebellum and parieto-occipital and inferotemporal cortices. The condition manifests itself by vertigo, tinnitus, ataxia, dysarthria and diplopia that can precede the onset of headache. The symptoms are quite variable and also may include blurred vision or tunnel vision, paresthesia, dizziness, hemiparesis, obtundation,
quadri paresis, loss of consciousness and aphasia. They may be associated with occipital headache, nausea and vomiting. The neurologic symptoms are usually of short duration. The condition is most common in adolescent females with first attack occurring at any time from infancy to adolescence. Although symptoms clear after an hour to several hours, residua after several attacks has been reported (32). The differential diagnosis include: seizure disorder, demyelinating disease, vertebral artery dissection and abnormalities of the bony structures of the occipital cervical junction.

**Ophthalmologic migraine**

Ophthalmologic migraine has been reclassified in the 2004 IHS classification as a “cranial neuralgia”. This condition is manifested by association of orbital or frontal pain with a complete or incomplete third nerve palsy. The headache may precede, accompany or follow the ophthalmoplegia. The third nerve dysfunction and at time fourth and sixth nerve dysfunction frequently outlast the headache. In the first episode, the paralysis lasts for only few hours. With repeated attacks, it can persist for weeks or months or even lasts permanently (33). Acute treatment with steroid reduces both the pain and the duration of ophtalmoplegia. (34).

**Confusional migraine**

This disorder which in the past was classified as “complicated migraine” is not a specific syndrome. Confusion can occur in migraine often in the setting of basilar migraine, hemiplegic migraine and migraine with aura. In some instances a period of confusion is triggered by a relatively minor head injury. This leads to an obvious but inaccurate diagnosis of epidural or subdural hematoma. Migraine with confusion, its neurologic deficit should not last more than 4-6 hours. Confusional attacks tend to recur, but eventually is replaced by typical migraine (35).

**Migraine variants**

Migraine variants imply episodic, recurrent or transient neurologic dysfunction in patients who are known to have migraine, who have family history of migraine or who are destined to have migraine. The most common forms of migraine variants include: benign paroxysmal vertigo of children, benign Paroxysmal torticollis, abdominal migraine and cyclic vomiting. The Alice in wonderland syndrome is now considered an aura and not a migraine variant (11).

**Benign Paroxysmal Vertigo**

Typically, a child between the age of 1 and 2 will develop a sudden unsteady gait, and they will be confused and grab on to an nearly object or person for stability and or falls to the ground. Consciousness is not lost and nystagmus may be noticed. The spells are short, lasting only a few minutes, and the children will often sleep afterwards. Follow up studies showed that benign paroxysmal vertigo often evolves into typical maigraine. Evaluation should include an MRI Scan to rule out posterior fossa lesion. Vestibular dysfunction can be documented in a significant numbers of subjects(36).

**Benign Paroxysmal Torticollis**

This rare variant of migraine consists of paroxysmal attacks of head tilt in an infant which may be associated with vomiting. The etiology of this disorder is felt to be similar to migraine (37). The spells may last longer than those in benign paroxysmal vertigo from hours to days. The differential diagnosis includes gastroesophageal reflux and torsion dystonia. Evaluation of the interventional contents to rule out a posterior fossa on cranio cervical junction abnormalities should be considered.

**Abdominal Migraine**

Abdominal migraine as a poorly understood an even more poorly characterized condition. It present in childhood with repeated stereotyped bouts of unexplained abdominal pain, nausea and vomiting. The diagnosis can only be entertained after exhaustive gastrointestinal and metabolic evaluation have been unrevealing. The condition could be a variant of the cyclic vomiting syndrome (38).

**Cyclic Vomiting**

Important in understanding the nature of this condition is the widespread observation that children with cyclic
vomiting become adult with migraine and have a positive family history of migraine (54). The attacks start as early as 1 year of age; in 82% the first attack occurs before age 6 years (55). Treatment is directed toward maintaining fluid and electrolyte balance. Aside from vomiting, symptoms include headache in 36%, fever in 43% and abdominal pain in 18%. Recent work suggests the presence of mitochondrial component in pathogenesis of cyclic vomiting (56).

Epilepsy and Migraine
Relationship between epilepsy and migraine have been discussed extensively. They have many features in common, including the fact that both are familial and that there is an increased frequency of migraine in epileptic patients and an increased incidence of epilepsy in migraineurs. Both are chronic disorder that are paroxysmal and episodic in nature. The clinical scenarios may be similar including an aura, loss of consciousness, motor dysfunction, and an associated headache. Both have EEG abnormalities of different natures and the etiology has been related to neurotransmitters and channel pathologies in both disorders. Both disorders respond to hormones and both may respond to antiepileptic drugs.

Diagnosis
In the initial evaluation of a patient with headache, the first step is to identify any secondary causes. In general, once a suspected secondary cause is effectively treated, the headache should resolve. The diagnosis of Migraine rests on the periodicity of the paroxysmal headache and at time, their initiation by stress. In the series of children younger than age 7 years evaluated for headaches, Chu and Shinnar found that 75% were experiencing migraine. Common migraine (without aura) was the most frequent form and only 17% of children could relate the presence of an aura (classic migraine) (39). The question as to whether neuroimaging should be part of the work up for a child presenting with recurrent headache, has to be addressed carefully. In older children and adolescents, the incidence of abnormal findings in the face of a normal neurologic examination is very low. Straussberg and Amir recommend performing imaging studies on youngsters, younger than 4 years of age in whom headache is accompanied by vomiting, even when the neurologic examination is normal (40). A thorough history and neurological examination would have served the child better than the imaging study (3). Battistella and colleagues found that headache was the first symptom in 27% of children with brain tumors and the only presenting symptoms in 10% (41). These authors noted that headache associated with brain tumors had a high incidence of projectile vomiting but nausea, photophobia, phonophobia rarely accompanied headache associated with brain tumors. There is a report of migraine attack associated with temporary unilateral sensory symptoms, aphasia or motor deficits. CSF examination showed lymphocytic pleocytosis and an aseptic inflammation of leptomeningeal vasculature (42).

Treatment
The treatment of migraine is symptomatic and prophylactic (11). It is based on the patient’s age, as well as the frequency of attack and the severity and disability caused by the migraine. Often when the patient and his or her family are reassured that this is a migraine and there is no serious underlying disorder, the attacks seem to become fewer and less distressing. Both non-pharmacologic and pharmacologic treatments are useful in dealing with migraine. General nonpharmacologic methods include patient and parent education and the elimination of trigger factors. A regular diet, sufficient sleep, and exercise also can be helpful. Food allergies are commonly believed to trigger attacks. A double-blind study suggests that some food, notably, cow’s milk, eggs, chocolate, orange, wheat, benzoic acid, cheese, tomato, and rye can provoke attacks and that their exclusion results in improvement in the majority of children (43).

Pharmacologic treatment
Rare patients require no pharmacologic intervention, if the attacks are of short duration, not severe and
quickly relieved by vomiting or sleep (11). However, if nausea, vomiting and pain are severe and/or prolonged, symptomatic medications such as analgesics, antiemetics and sedative, play an important role. Acetaminophen and the nonsteroidal anti-inflammatory drugs (NSAID) have been well studied in children and adolescents. In most studies, (NSAID) seen to be more successful than acetaminophene in relieving pain. Acetaminophen 15mg/kg is effective to relieve moderate to severe attacks (44).

Other medications used for severe attacks include: naproxen, ibuprofen, phenacetin or caffeine singly or in combination. The introduction of triptans for the treatment of migraine, represented a significant step in their remediation. Numerous trials have demonstrated both their effectiveness and their safety. However this drug has not been approved by FDA in United States. The recently published practice parameter, has concluded that for acute treatment of migraine in children and adolescents, Ibuprofen is effective and acetaminophen is probably effective. In the past, patients with infrequent episodes were treated with ergotamine. Presently this is not recommended in pediatric and adolescents patients. There is a form of dihydroergotamin (Migranal) that is given nasally and has been successful in adolescents especially if they are unresponsive to triptans(45).

**Prophylactic medication**

Prophylactic therapy should be considered when migraine headache is unresponsive to acute measures or migraine is occurring so frequently that results in days lost from school and other activities. Prophylactic medications that have been utilized include: NSAIDs, antihistamines, beta-blockers, calcium channel blockers, and antiepileptic drugs. Propranolol has been advocated as a preventive for childhood migraine. In children who weigh less than 35kg, the maximum dosage is 20 mg three times daily; in those weighing more than 35 kg, it is 40 mg three times/day (46). This drug should be used with considerable caution in asthmatic children and in childhood diabetes. In one study valproate, 10-20 mg/kg per day was found as effective as propranolol in the prophylax of migraine without aura (47). Another medication that was found successful in adolescents, who may be obese, is Topiramate. Its dosage seems to be lower than those used in treatment of epilepsy (48).

**Prognosis**

The outlook for the patients with migraine headache is excellent and in most instances, the condition does not interfere with school work. In approximately two-thirds of children, attacks persist throughout life, although many patients are intermittently free from them for long period or their headaches are less stressful for family and school environment(11).

**Status migrainosus**

This condition consists of a migraine attack lasting longer than 72 hours. It most commonly occurs in patients with preexisting migraine and only rarely is the first manifestation. Most require parenteral medications as well as intravenous fluids and antiemetics. Patients are initially sedated, then a variety of medications including: magnesium, valproic acid, sedative and dehydroxyergotamin are used.

**Chronic progressive headache**

The presence of a chronic and progressive headache, suggests a disorder which worsens over time. Frequently there are abnormal neurologic symptoms including evidence of increased intracranial pressure. Usually neurologic examination of subject is abnormal. Chronic progressive headache may be a symptom of brain tumors, hydrocephalus, Pseudotumor cerebri, brain abscess and chronic subdural hematoma.

**Chronic Non-progressive headache**

The term Chronic Non-progressive headache implies a headache, that has been present for longer than six weeks to three months (11). Silberstein and Lipton have divided this disorder into chronic tension type headache, hemicranial continual, new onset daily persistent headaches, and transformed or chronic migraine (12).

**Chronic Tension type headache**

Another name for this entity is chronic muscle contraction headache. The pain which is not associated with aura, is
bitemporal or bifrontal, may occur many times per month. The patient describes the headache in a non specific manner and may have mild associated symptoms such as blurred vision, fatigue and dizziness. Manifestation of depression may be concomitant of Chronic Non-progressive headache. In adolescents, depression appears as withdrawal, poor school performance, sleep disturbance, a change in behavior, somatic complaints, lack of energy, mood change, weight loss and school phobia.

If the headache has been present for more than 8 to 12 weeks in the absence of neurologic symptoms and signs and physical and neurologic examination are normal, the headache is usually not secondary to a structural abnormality. Treatment could be tried with low dose of amitriptyline (11).

**Hemicrania continua**

This type of headache which is a strictly unilateral continuous one, with multiple daily painful exacerbations, occurs anywhere from 2 to 3 times per day to 2 to 3 times per week. This type of headache is associated with unilateral autonomic features including tearing, nasal stuffiness, ptosis, conjunctival injection and rhinorrhea as well as migrainous symptoms such as nausea, photophobia and phonophobia (49).

Indomethacin is the drug of choice which provides complete relief of symptom in most cases of hemicrania continua. The usual dosage of 100 to 225 mg given in two to three divided doses.

**Chronic migraine / transformed migraine**

This type of headache begins as episodic migraine and over time become less severe but more frequent until they merge into a daily headache syndrome. Some authorities believe, this type of headache disorder is one of the most frequent headache disorder seen in most large referral headache clinics. It is significant that many of these patients have comorbidities such as low back pain, stomachaches, frequent sore throat, and a high level of psychosocial difficulties (50).

**Miscellaneous headache syndromes**

There are several headache syndromes that do not fit into usual classification and majority of these conditions are not association with structural brain disease. Proper identification may lead to specific treatment, resulting in dramatic relief of symptoms (11).

**Cluster headaches**

Two forms of cluster headache can occur: episodic and chronic. The episodic form is defined by frequent headaches lasting for periods from one to three months, followed by periods of remission lasting from month to years. Episodic cluster forms 80% of cluster headache, and remainder 20% are of chronic type.

Chronic Cluster headaches are defined as headaches that last for more than 1 year without remission or with remissions that last less than 2 weeks. These types of headaches are difficult to treat. Cluster headaches primarily affect males. The typical attack occurs 2 to 10 times daily and lasts 10 minutes to 3 hours. The pain is severe, primarily affect male but not exclusively and unilateral, rarely if ever, changes sides.

It is localized in or about the eye and is associated with ipsilateral lacrimation, rhinorrhea and nasal stuffiness. Ptosis and miosis also may occur. The acute treatment include oxygen, triptan and cafe got. Prophylactic medications have included methysergide, lithium and prednisone.

**Indomethacin- Sensitive Headache**

There are four types of headache syndromes that are specifically responsive to indomethacin even infrequently encountered in pediatric age group (11).

**Chronic Paroxysmal hemicrania**

This type of headache is characterized by multiple daily attacks lasting from 5 to 30 minutes and persistently unilateral. The pain is severe and localized to on eye in forehead above the eye and precipitated by head movement.

**Exertional Headaches**

Any exertion such as running, coughing, swimming and sexual activity may precipitate an episode of headache (51). They can occur during the initiation of an activity, the middle or after activity is completed.
Cyclic Migraine
This is a form of migraine which comes in cycles (52). It also has been called “cluster migraine” but it is not a form of cluster headache. The headache cycle averages anywhere from 2 to 10 to 12 weeks with an average of 6 weeks. During the cycle, the migraine headache occurs daily or several times per week followed by months of headache free interval.

Hemicrania Continua
Is marked by a steady, nonparoxysmal, severe hemicranias localized to the frontal part of the head and not associated with nausea or autonomic symptoms (53). All of the above 4 types of headaches are responsive to indomethacin.

References