

## CASE REPORT

 Nevra Öksüz<sup>1</sup>  
 Halit Fidancı<sup>2</sup>  
 Aynur Özge<sup>1</sup>

<sup>1</sup>Mersin University, Faculty of Medicine, Department of Neurology, Mersin, Turkey  
<sup>2</sup>Adana City Training and Research Hospital, Department of Neurology, Division of Clinical Neurophysiology Adana, Turkey

### **Yazışma Adresi:**

Halit Fidancı  
Adana City Training and Research Hospital, Department of Neurology, Division of Clinical Neurophysiology Adana, Turkey  
mail: dr.halitfidanci@gmail.com

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konuralptipdergi@duzce.edu.tr  
konuralptipdergisi@gmail.com  
www.konuralptipdergi.duzce.edu.tr

## Unusual Primary Headaches of Children and Adolescents: Practical Tips for Physicians

### ABSTRACT

Childhood and adolescent headaches somehow look like adulthood headaches but not known as much as them. In this paper eight rare known primary headache disorders of children and adolescents have been given with practical clues for physicians. All of the cases have been selected from Mersin University Faculty of Medicine, Childhood and Adolescent Headache Outpatient Department database and discussed with literature. This paper mainly based on increasing physician awareness about unusual primary headache disorders of children and adolescents.

**Keywords:** Childhood, Headache, Migraine

## Çocukluk Çağı ve Ergenlik Döneminin Nadir Primer Baş Ağrıları: Hekimler için Pratik İpuçları

### ÖZET

Çocukluk çağı ve ergenlik döneminin baş ağrıları bir şekilde erişkin baş ağrılarına benzese de, erişkin baş ağrıları kadar bilinmez. Bu yazıda çocuk ve ergenlere ait sekiz nadir bilinen primer baş ağrısı pratik ipuçları ile birlikte verilmiştir. Olguların tamamı Mersin Üniversitesi Tıp Fakültesi Çocuk ve Adolesan Baş Ağrısı Polikliniği veritabanının arşivlerinden seçilmiş ve literatür eşliğinde tartışılmıştır. Bu makale özellikle çocuk ve ergenlerin nadir primer baş ağrıları hakkında hekimin farkındalığını arttırmayı temel almaktadır.

**Anahtar Kelimeler:** Çocukluk Çağı, Baş Ağrısı, Migren

## INTRODUCTION

Headache is a common complaint in children and adolescents. The frequency is increasing with age and etiologies are changing (1,3-5) The children who complained of headaches are initially treated by their parents or teacher without scientific basis under the danger of a serious problem like a tumor. Also headache sometimes is unapparent to parents of younger children who are crying, and have nausea, vomiting and abdominal pain (6,7). It has been showed that headache disorders in the childhood population is significant not only because of its prevalence but also the burden of pediatric headaches including loss of family function and life standart (4). However most of the cases especially atypical ones were underdiagnosed even in developed centers because of the changing face of headache disorders in this age group.

Accordingly, this paper focuses on pediatric uncommon headache syndromes for which we have recent and substantially clinical information about the headache characteristics. We first described nine case stories and then some important practical clues based on this case.

### Case 1

*A 16 year-old boy, complained headache episodes for four years. He describes a headache once a month. An illusion happening in the middle of the visual field and in seconds this illusion slide to periphery, a few minutes after, headache begins that unilateral from time to time. In the last six months he experienced 5 episodes of unilateral-bilateral and usually frontal-periorbital in location, severe and lasts more than three hours. During one of these episodes he had only conjunctival injection and in all of the five episodes he had photophobia, phonophobia and dizziness. His mother noted that he always feels sick when he is riding in a car. Also, he is known to have pollen allergy. The headache episodes generally start when he wakes up.*

*The physical exam was within normal limits including a normal body mass index (19 kg/m<sup>2</sup>) and cognitive development. His current medication is fluoxetine which he used for several months for obsessive-compulsive disorder. In his history, there was no risk factors which include smoking, head trauma and family history of any type of headaches. His echocardiogram, magnetic resonance imaging (MRI) and MR Angiography were normal.*

*He was diagnosed with 5 episodes of "retinal migraines" in the last six months. He had attacks with fully reversible monocular visual disturbance associated with his headaches. Between the attacks he had a normal ophthalmologic examination.*

To diagnose as "Retinal Migraine" the patient must have had at least two attacks of transient visual loss followed by a migraine headache. Headache fulfilling criteria for 1.1

Migraine without aurabegins during the visual symptoms or follows them within 60 minutes. All underlying diseases and other forms of migraine must be excluded that causes transient monocular blindness. Retinal migraine can be diagnosed only fully reversible monocular positive and/or negative visual phenomena and with a normal ophthalmological examination between attacks. (Adapted from The International Headache Disorders, 2nd edition) (8). Our patient met all these criteria's for retinal migraine.

*Tip 1.* Unilateral unexplained reversible visual field disturbance can be a sign of retinal migraine especially in subject with migraine equivalent or positive family history of migraine.

### Case 2

*A 12 year-old boy presented with atypical headache attacks of six times in the last month, lasting less than one hour during the last five months. Every episode he described before headache an image comes in front of eye like a man and a woman desired to kill and chasing him, it takes about 20 minutes and then he had severe headache with nausea, vomiting, photophobia, phonophobia and a few times of vertigo characterized by a bilateral pressing tightness that occurs in front of the cranium. Episode is getting worse with physical activity.*

*His mother noted that he had cyclic abdominal pain in his first three years of his life. His physical examination and growing history was normal ranges. Fundoscopic exam and mangetic resonance imaging were also normal. In electroencephalography normal background activity and unregular right centrottemporal spikes were determined. He had family history of migraines with his mother, except this he had no other risk factors like smoking, obesity, and cardiovascular disease in his family. He is currently not using any medications. After three month management with valproic acid, he was headache free. After one-year follow-up period, not any headache attack was observed again and electroencephalography had normal limits again.*

Some children like our patient presented to us with a complaint of seeing spots, colors, lights or hallucinations before or as the headache begins. Three dominant visual phenomena were found; binocular visual impairment with scotoma (77%), hallucinations (16%) and monocular visual impairment or scotoma (7%) (9). It is difficult to distinguish if the hallucination with headache is a migraine with aura or benign occipital epilepsy. Also bizarre visual phenomena may be seen in the "Alice in Wonderland" syndrome with or without visual illusions such as micropsia where objects appear smaller; macropsia where objects appear larger; teleopsia, where objects appear far away (10). As our patient has electroencephalography, migrainous patients have been reported to have nonspecific electroencephalographic abnormalities

which often does not require treatment. But when we treated him with valproic acid, he had no any attack again. Final diagnosis was made as “Migraine with Complex Visual Aura” and the patient was treated with prophylactic medication.

*Tip 2.* Unexplained complicated visual hallucination can be a sign of migraine with aura after exclusion of epileptic syndromes and intracranial abnormality.

### Case 3

*A 11-year-old boy had severe headache attacks more than fifteen days a month, for four years. Every day he had more than three attacks. Before the attacks he often had visual aura like broken lights. His aura takes seconds and in minutes he had headache which is unilateral, stabbing and especially located on trigeminal nerve first division. Stabs last no more than a few seconds. He had headaches every time on the same exact location. Photophobia, phonophobia, dizziness also attend to his headaches. His attack is going worse with physical activity.*

*In first three years of his life he had a head trauma that he crushed his face. He didn't have any family history of migraines but both of his grandfathers had coronary artery disease, his mother's cousins had epilepsy. He had asthma and taking medication. His mother had noted that he had anxiety but did not need any medication.*

*Both of his physical and neurological examinations and investigations including MRI, MR angiography and EEG were normal.*

He is diagnosed as “Primary Stabbing Headache (PSH)”. Characteristically associated symptoms are discontinuous. Comorbidity with migraine (about 40%) or CH (about 30%) has been reported in children and adults. In a tertiary center PSH presented 3.2% of children and adults with recurrent headaches (11). Other types of headache syndromes like trigeminal neuralgia, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT syndrome), chronic paroxysmal headache and secondary causes which simulate PSH must be excluded. Trigeminal neuralgia responses to carbamazepine treatment and has trigger points that cause attacks. It is difficult to distinguish SUNCT and chronic paroxysmal headache from the primer stabbing headache because of the duration of episodes (12,13). SUNCT and chronic paroxysmal headache have autonomic features. Arnold-Chiari malformation, colloid cyst and tumors of the third ventricle and pineal region, chronic subdural hematoma and basilar impression were excluded by magnetic resonance imaging (MRI) and MR angiography. Some studies have shown that PSH responses to indomethacin treatment (8,11-14). Our patient was treated by indomethacin and magnesium. In follow-up visits he had less attacks without aura.

*Tip 3.* Unilateral short-lasting stabbing

headache attacks without autonomic features can be a sign of primary stabbing headache after exclusion of secondary causes.

### Case 4

*A 17 year old boy presented to us with repeated stereotyped episodes of unexplained abdominal pain with nausea and vomiting from their childhood. He was examined by pediatrician many times in emergency service because of abdominal pain attacks. All exhaustive gastrointestinal and metabolic evaluations indicated any reason. We observed his headaches in the course of long-term follow up. Headache attacks reported in the last 4 years, unilateral located, generally 4-5 times in a month, severe attacks lasting 5 hours associated with nausea, vomiting, phonophobia and dizziness. He had abdominal pain with or without subsequent headache attack, which is poorly localized, moderate or severe intensity. During the abdominal pain he had nausea, vomiting and pallor many times.*

*He had Dandy-Walker abnormality in his medical history. Also his mother, his grandmother and his grandmother's mother had Dandy walker abnormality. His physical and neurological examinations were normal ranges. His MRI showed hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlargement of the posterior fossa. EEG and other biochemical investigations were normal ranges.*

He is diagnosed as definite “Abdominal Migraine (AM)”. Abdominal migraine occurs in 1% to 4% of children and one of many potential etiologies of recurrent abdominal pain in children (15,16). Diagnosis has been made according to the International Classification of Headache Disorders, Second Edition and American College of Gastroenterology Rome III Diagnostic Criteria for abdominal migraine in children. After puberty ongoing abdominal pain attacks is a rare condition. Also typical migraine attacks with abdominal pain attacks are also rare. However positive family history of migraine (more than 90%) and headache characteristics are supported this diagnosis after exclusion of secondary causes (17-19).

His attacks are still ongoing but after flunarizine and domperidone prophylactic treatment, he has less frequent and lower severity of headache attack without abdominal crisis.

*Tip 4.* Unexplained abdominal pain attacks of childhood and adolescents, even adults, can be a sign of migraine, even abdominal migraine after exclusion of secondary causes.

### Case 5

*A 14 years old girls complained severe vertigo attacks for 1.5 month described movement illusion of the environment putting down all daily activities including school. Their attacks were aggravated by head position changes and associated with nausea sometimes vomiting. She underwent a specific medical management in*

otolaryngology clinic but did not work. Including MRI, MR angiography and venography all investigations were normal ranges. Last 1 month she described 3 headache attack in left or right side of the head, lasting more than 2 hours and associated with nausea, vomiting and phonophobia independent from vertigo periods. Before this vertigo period she described averaged 5-6 headache attacks in a year, described in the same characteristics. Past medical history periodical vomiting attacks in first 3 years of life and travel sickness had been reported. Her mother and aunt diagnosed as migraine.

This girl is diagnosed with “Vestibular migraine (VM)”. Migraine and vertigo are most frequent symptomatology in pediatric age groups and related each others. Vertigo can be an associated feature of migraine or a specific diagnosis as VM, Migraine Stimulated Vertigo (MSV) or Somatoform vertigo (SV). In patients with VM reported vertigo attacks with or without correlated headache attacks. VM have to made a differential diagnosis from Benign Paroxysmal Positional Vertigo (BPPV) by the aspect of continuing vertigo period even head did not move. In children and adolescent age groups the most frequent diagnosis was MRV (28%), followed by MSV (19%), SV (14%) and VM (11.2%). MSV and VM reported as most frequently in adolescent girls. Competent care of childhood migraine should include skill in detecting both the clinical symptoms of vertigo and overlapping somatoform symptoms (20,21). This girl was treated with flunarizine in two month and supported with vestibular rehabilitation. She was headache and vertigo free for last 6 months.

*Tip 5.* Headache associated vertigo attacks can be a sign of Vestibular Migraine, especially in girls with a positive past history of migraine or family history of migraine.

#### **Case 6**

A 16 year-old boy presented to our department with headaches of ten times a months, lasting less than half an hour during the last five years. Every episodes occurred during sleeping and made him woke up. He had no prodromal period, no aura or no any other concomitant symptoms. Episodes are getting more often when he is stressful. His remission period takes ten days. Both of his social history and family history he had no risk factors. He is currently not using any medications. His echocardiogram, EEG and magnetic resonance imaging (MRI) were normal ranges. Polysomnographic investigation did not show additional disturbance.

This boy diagnosed as “Hypnic headache (HH)”. This type of headache disorders is recurrent and sleep related headache which is typically a disorder of the elderly and rare in children. Clinicians should be aware of this possibility. There are recent articles that reported hypnic headache in

one child and one adolescent (22,23). Attack occurs at night during sleeping and awaking the patient at almost the same time like alarm clock. Headache is usually last less than 15 minutes, not well-located and bilateral two thirds of cases and no any other symptoms like autonomic features. The differentiation of nocturnal migraine, trigeminal autonomic cephalalgias and headache attributed to increased intracranial pressure from hypnic headache is very difficult. Also nocturnal migraine has nocturnal awakenings (24). Hypnic headache does not include autonomic phenomena and trigger points (25-31). This boy treated with indomethacine (first month regularly and followed by during attacks) and asetazolamide (regularly) for 6 months.

*Tip 6.* Sleep related short duration headache attacks can be a sign of Hypnic headache after exclusion of secondary causes.

#### **Case 7**

A 14 year-old boy with headaches during three months, almost everyday. Headaches were unilateral, usually located periocular and continuous with fluctuations in pain intensity. He sometimes had severe attacks that lasts more than twelve hours. Conjunctival injection, lacrimation, nasal congestion were associated with exacerbations of pain. He had no prodromal period, no aura or no any other concomitant symptoms. Episodes are getting more often when he is stressful and getting worse in course of physical activity. His physical and neurological examination were normal, also his MRI was normal. Except his father hypertension he had no important history.

He was diagnosed as “Hemicrania Continua (HC)”. HC have onset in the third decade of life, but with a range from first to seventh decades (32). HC is mightily unilateral headache with moderate-severe intensity. Autonomic features like lacrimal injection, nasal congestion, conjunctival tearing, facial flushing attend to episodes of headache (33). The diagnosis of HC in childhood is rare, but when we observe patients’ social histories, we found headaches as same as they have now (34). Clinicians must be aware of this possibility which is completely responsive to indomethacin treatment. He treated with indomethacin for 6 months and was headache free for last 3 months.

*Tip 7.* Unilateral sustained headaches at the same side of the head, associated with cranial autonomic features, can be a sign of Hemicrania Continua after exclusion of secondary causes.

#### **Case 8**

An active eleven years old girl complained from headaches attacks after tennis games. She had headache attacks described as pressure both on her temples for the past one year after vigorous physical exertion. The pain is sometimes worse, sometimes less and lasts anywhere from a few seconds to half an hour. When she had a rest it ended in minutes, also she had no any other concomitant symptoms. Her physical exam and all

her analysis were normal includes MRI, doppler ultrasonography and echocardiogram. She was treated by 50 milligrams of indomethacin at her attacks and magnesium salts were effective prophylactic medication of the attacks. Last 6 months she was headache free.

She was diagnosed with "Benign Exertional Headaches (BEH)". BEH occurs only during or after physical exercise and characterized by pulsating pain episodes that is usually bilateral, throbbing and lasts from 5 minutes to 48 hours (rarely). Nausea and vomiting does not attend to this type of headache (35). Primary exertional headache is seen in younger population than older with a male predominance. Headache usually occurs at the peak of exercise and last in minutes after the end of the exercise. It has many characteristics of migraine headache like photophobia, phonophobia, nausea, vomiting and throbbing. But 60% of the cases is bilateral. Differential diagnosis include hemorrhage due to vascular malformations, middle cerebral artery dissection, intracranial space occupying lesion (12,36,37). This type of headache attacks commonly responsive to effective dose of indomethacin: Some patients with frequent or unresponsive attacks requires prophylactic medication.

*Tip 8.* Headache attacks with triggered by any specific activity such as exercise, cough, sexual activity etc. can be a sign of Benign Exertional Headaches after exclusion of secondary causes.

## DISCUSSION

Headache is one of the more common presenting complaints to neurology polyclinics (1,2,3). Each year, over 10 million patients visit their physician or emergency department with a complaint of headache (38). We will review several types of headaches that do not easily fall into the usual classifications. These headaches are based on the primary headache syndromes. It is important to recognize these syndromes that may lead to specific treatment resulting in dramatic recovery.

The differential diagnosis of unusual

headaches in children requires a systematic approach. In some children, prostration, nausea, repeated vomiting and abdominal discomfort may overshadow the complaint of headache.

Migraine occurs at all age and may begin in infancy (39,40). The disorder begins before age 20 years in 50 percent of cases. We discussed rare variants of migraine like abdominal migraine, retinal migraine and vestibular migraine here which can be treatable if it is recognized.

There are many differences between management of primary headaches and unusual variants of headaches. If the rare variants were not recognized by physician, these syndromes get worse and treatment get hard. We want to point out that, if the rare variant of headaches which do not meet usual classification criteria is recognized by physicians can be treatable. On the contrary if we missed these types of headaches, as the treatment gets harder, the headache may be turns into chronic daily headache. Also these headache disorders showed important effects on young's' daily life activities and happiness depending on headache type (4).

This article is a consideration of unusual headache syndromes. The definition criteria for each disorder are presented in the ICHD-II and associated papers. The consideration can support clinicians to decide quickly and observe well when evaluating the children who have headaches. The practical tips could be help physicians for early and definite diagnosis and differential diagnosis also.

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