Chapter 130

Childhood Migraine and Related Syndromes

Ishaq Abu-Arefeh and Eric Pearlman

MIGRAINE WITHOUT AURA

Migraine without aurua (MO) in children and adolescents has some features that make it different from that in adults (Chapter 43). The duration of attacks is variable, lasting from 1 to 72 hours; the pulsating quality of pain is not easily described by young children; and headache is, commonly, at the forehead. The headache, as in adults, is moderate or severe in intensity, aggravated by routine physical activity, and associated with nausea and/or vomiting and/or photophobia and phonophobia.

Epidemiology

The prevalence of migraine increases with age, from 0 to 7% in preschool children (under 7 years) to 20% in adolescents (Fig. 130-1), with 75 to 80% being MO (20). The peak age of onset is 12 to 14 years. In younger children, migraine is more common in boys than girls, but is more common in girls after the age of 12 years (2).

Clinical Features

During attacks of migraine the child is pale and quiet and wants to be left alone. Commonly associated symptoms are loss of appetite, nausea, vomiting, and intolerance to light, noise, smell, and physical activities. Dizziness (unreal sensation of movement), abdominal pain, and limb pain may also be present. Short attacks of migraine lasting less than 2 hours are uncommon, but are well recognized as confirmed by prospective diary records (1). The International Headache Society (IHS) criteria for the diagnosis of MO (16) are otherwise as described in earlier chapters.

MIGRAINE WITH AURA

Migraine with aura (MA) in children and adolescents is characterized by distinct and sometimes graphic symptoms that precede the onset of headache. Children describe the symptoms, commonly visual, in words and by drawings (23). Common visual aura includes blurred vision, tunnel vision, blind spots (scotomata), zigzag colored lines in front of the eyes, or image distortion. Less commonly, the aura symptoms are sensory such as tingling sensation or numbness. Motor symptoms are rare and may represent a complicated migraine attack or hemiplegic migraine. The aura is brief and fully reversible. Headache follows within 60 minutes from resolution of aura, but the two phases can be indistinct. The aura symptoms can be intense and frightening to the child and parents, leading to their urgently seeking medical advice. The headache phase is similar to that in MO, but occasionally atypical.

Prognosis of Childhood Migraine

The natural course of migraine is that of remissions and relapses. Remissions of at least 1 year were recorded in up to 58% of patients during periods of follow-up of at least 5 years (19). Long-term follow-up of 73 children showed that 34% became headache free after 6 years, 62% after 16 years, and only 40 to 47% after 22 to 40 years (7,8).

Management of Childhood Migraine

A management plan should follow a careful assessment and accurate diagnosis to alleviate concerns of the child and anxieties of the parents. The treatment starts with reassurance and education of the child and parents about the benign nature of migraine and its remitting–relapsing

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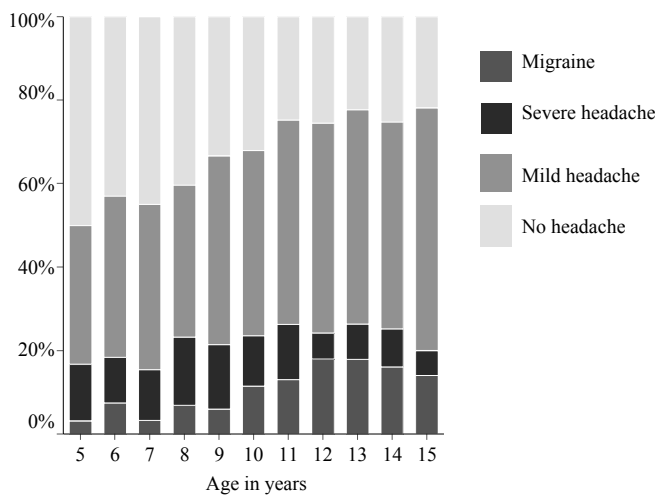

FIGURE 130-1. Epidemiology of headache and migraine in schoolchildren.

course. Diagnostic studies, including neuroimaging (computed tomography or magnetic resonance imaging) is necessary in a small number of patients with features of secondary headaches.

Management of Acute Attacks

Many children and families may try nonpharmacologic strategies (rest and sleep) and over-the-counter analgesics before they consult their doctors, but only one in three parents can determine and measure the correct doses for children (21). Medications are likely to be successful if given soon after the onset of attack in appropriate dosages.

In children and adolescents under 15 years of age, paracetamol (acetaminophen) (15 mg/kg body weight, maximum 1g) and ibuprofen (10 mg/kg) are effective and safe (15).

Aspirin is effective, but its association with Reye syndrome prohibits its use in children under 15 years. No clinical trials are available on other nonsteroidal antiinflammatory drugs. Occasionally, codeine phosphate may be necessary if other acute treatments fail.

Sumatriptan (Imigran/Imitrex) is a selective agonist of 5-$HT_{1B\text{-}D}$ receptors with a good efficacy and safety record in adolescents. It is available as a subcutaneous autoinjection, oral tablets, and a nasal spray. Multicenter, randomized controlled trials of nasal sumatriptan in doses of 10 and 20 mg showed good efficacy and safety profile as compared to placebo (6,27). Nasal sumatriptan is now licensed for use in 12- to 17-year-olds in some countries. Other triptans are also available, but their efficacy in children and adolescents is not established yet.

Metoclopramide (Maxolon/Reglan) or prochlorperazine (Stemetil/Compazine) may alleviate nausea and vomiting but should be used with caution due to possible extrapyramidal symptoms and dystonic reactions.

Prevention

Nonpharmacologic strategies for the prevention of migraine include the avoidance of identified trigger factors and adopting a healthy lifestyle, including a predictable sleep pattern, regular meals, and regular pattern of rest and exercise.

There is limited evidence for the efficacy and tolerability of pharmacologic prophylaxis. It is only indicated if acute treatment is unsuccessful and migraine attacks are frequent (more than two attacks per month), prolonged, or severe enough to interfere with normal life.

Propranolol reduced the frequency of migraine attacks in one trial (18), but not in another (13). In children over 7 years of age, propranolol may be given in a dose of 1 to 2 mg/kg/day in two divided doses (maximum 40 to 50 mg twice daily). Pizotifen (Sanomigran), commonly used in the United Kingdom, may be effective in the prophylaxis of abdominal migraine (24), but not in migraine headache (14). Valproate is effective in adults, but has not been studied in children (9).

Amitriptyline was effective in prevention of chronic headache in a large open observational study (17). However, it showed only a modest benefit in the prevention of migraine in 19 children (6 to 12 years of age), it did not influence the duration of migraine attacks, and drowsiness was a common adverse effect (22). The efficacy of other medications, including topiramate, lamotrigine, magnesium, and feverfew, has yet to be established.

ABDOMINAL MIGRAINE

International Headache Society (IHS) second edition code and diagnosis: 1.3.2 Abdominal migraine
WHO code and diagnosis: G43.820
Short description (Headache Classification Committee, 2004): An idiopathic recurrent disorder seen mainly in children and characterized by episodic midline abdominal pain manifesting in attacks lasting 1 to 72 hours with normality between episodes. The pain is of moderate to severe intensity and associated with vasomotor symptoms, nausea, and vomiting.
Other terms: Functional, nonorganic, or psychogenic abdominal pain, recurrent abdominal pain of childhood, and periodic syndrome

Epidemiology

Eight percent of schoolchildren between 5 and 15 years reported recurrent abdominal pain and 4.1% reported episodes that fulfilled the criteria for the diagnosis of abdominal migraine (AM). AM affects both boys and girls equally with a peak age of onset at 10 years (3).

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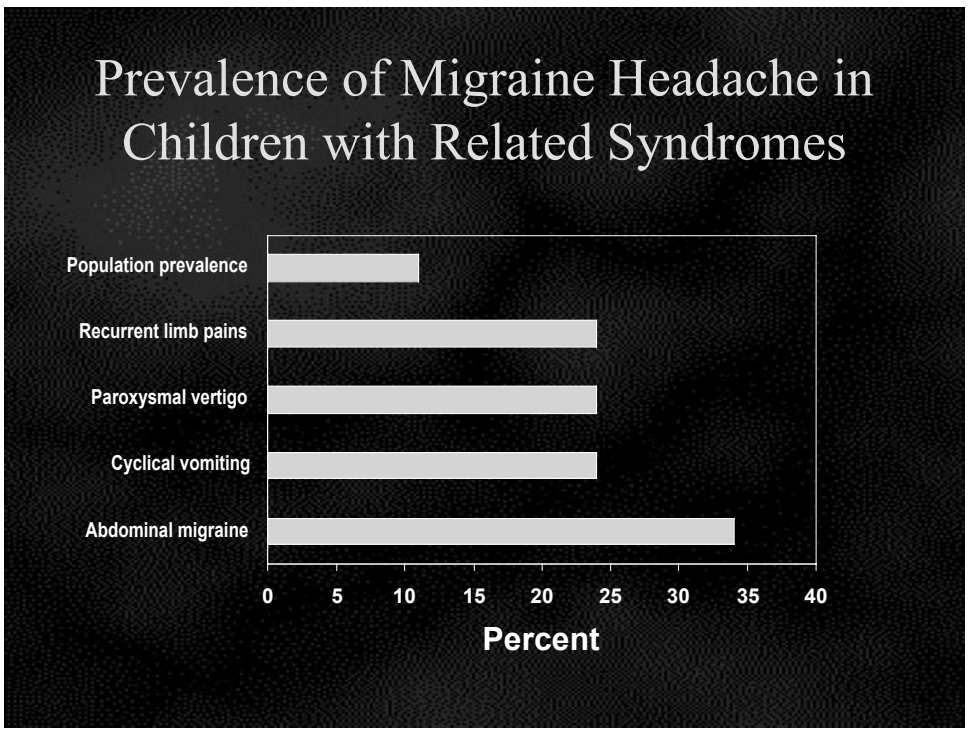

FIGURE 130-2. Prevalence of migraine headache in children with related syndromes.

Genetics

The genetic basis of AM is not known. Population-based studies show an increased prevalence of migraine among children with AM and their first-degree relatives. Also, the prevalence of migraine is statistically higher among children with AM than in control children (Fig. 130-2), suggesting that both conditions may have common genetics.

Clinical Features

Episodes of AM are characterized by central abdominal pain, dull in nature, severe enough to interfere with normal activities, commonly associated with anorexia, nausea, vomiting, and pallor, lasting for at least 1 hour, and with complete resolution between attacks. The child is otherwise well and healthy with normal growth and development. Diagnosis should be made after exclusion of gastroesophageal reflux, peptic ulcers, inflammatory bowel disease, constipation, food intolerance, malabsorption, and renal disorders.

IHS diagnostic criteria for abdominal migraine (Headache Classification Committee 2004) (16) are as follows:

A. At least five attacks fulfilling criteria B through D
B. Attacks of abdominal pain lasting 1 to 72 hours (untreated or unsuccessfully treated)
C. Abdominal pain has all of the following characteristics:
  1. Midline location, periumbilical or poorly localized
  2. Dull or "just sore" quality
  3. Moderate or severe intensity
D. During abdominal pain at least two of the following:
  1. Anorexia
  2. Nausea
  3. Vomiting
  4. Pallor
E. Not attributed to another disorder

Prognosis

At least one third of children with AM continue to have attacks of abdominal pain in late adolescence and early adult life, and around 50% suffer from migraine headache attacks (10).

Management

The management starts with the confirmation of the diagnosis, reassurance to parents, and identification and avoidance of trigger factors. Acute attacks are treated with simple analgesics, rest, and fluid replacement and occasionally with antiemetics. There are no published data on the use of specific antimigraine agents such as sumatriptan. Drug prophylaxis has been evaluated in a randomized double-blind placebo-controlled trial of pizotifen. It was shown to be effective in the prevention of abdominal migraine (24). Evidence for the value of propranolol and cyproheptadine is based on open trials only (28).

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CYCLICAL VOMITING SYNDROME

IHS second edition code and diagnosis: 1.3.1
WHO code and diagnosis: G43.82
Short description (Headache Classification Committee, 2004): Recurrent episodes, usually stereotypical in the individual patient, of intense nausea, vomiting, pallor, and lethargy. There is complete resolution of symptoms between attacks.
Other terms: acephalgic migraine, bilious headache, fitful vomiting, and periodic syndrome

Epidemiology

Around 2% of schoolchildren reported unexplained attacks of vomiting that fulfill the criteria for the diagnosis of cyclical vomiting syndrome CVS (4). CVS commonly presents in infancy and early childhood. Both girls and boys are equally affected.

Genetics

Clinical and epidemiologic studies show a close relationship of CVS to migraine (Fig. 130-2).

IHS diagnostic criteria for CVS (Headache Classification Committee, 2004) (16) are as follows:

A. At least five attacks fulfilling criteria B and C
B. Episodic attacks stereotypical in the individual patient of intense nausea and vomiting lasting from 1 hour to 5 days
C. Vomiting during attacks occuring at least four times an hour for at least 1 hour
D. Symptom-free between attacks
E. Not attributed to another disorder

Clinical Features

Children with CVS present with sudden recurrent episodes of intense anorexia, nausea, vomiting, and lethargy. During the attacks the child has intolerance to light, noise, and exercise. The child looks unwell, pale, and miserable. Each attack lasts from a few hours to a few days and may lead to dehydration. Between attacks the child is completely well. The diagnosis is made on exclusion of other similarly presenting disorders.

Prognosis

A follow-up study, for up to 10 years, showed that 50% of children continued to suffer from vomiting attacks in their teenage and early adult life and 46% suffered from migraine as compared to 12% of a matching control group (11).

Management

The treatment of acute attacks aims to stop vomiting and prevent dehydration. Early administration of oral or intravenous antiemetic drugs such as ondansetrone may abort attacks. Prophylactic treatment with erythromycin as a prokinetic agent may reduce the number and severity of attacks (25).

BENIGN PAROXYSMAL VERTIGO

IHS second edition code and diagnosis: 1.3.3
WHO code and diagnosis: G43.821
Short description (Headache Classification Committee, 2004): Benign paroxysmal vertigo (BPV) is probably a heterogenous disorder characterized by recurrent brief attacks of vertigo that occur without warning and resolve spontaneously in otherwise healthy children.
Other terms: Dizziness, giddiness, and periodic syndrome

Epidemiology

The prevalence of early childhood BPV is not known. Late-onset BPV childhood is estimated to occur in 2.6% of schoolchildren (5).

Genetics

BPV has close clinical and epidemiologic relationships to migraine. Migraine is more common among children with BPV (Fig. 130-2) and among their first-degree relatives than in matching control children (5).

Clinical Features

BPV is characterized by episodes of unreal sensation of movement of the child or the surrounding environment. Each episode lasts for a few minutes but on occasions may last a few hours. BPV is uncommon in early childhood (age of 2 to 4 years). It presents with sudden episodes of pallor, screaming, unsteadiness, nystagmus, nausea, and occasionally vomiting (26). The child either sits or clings to his or her parent in fear. The attacks terminate spontaneously. There is no loss of consciousness and the child is aware and responsive during the event. Episodes of BPV decrease in frequency by age 5 years and may be replaced by episodes of headache typical of migraine.

BPV is more common among schoolchildren (5) who can describe the unreal sensation of movement that lasts for seconds to minutes. The only abnormal finding on physical examination during attacks is horizontal nystagmus. The diagnosis of BPV is based on excluding underlying neurologic causes, vestibular disorders, and adverse reactions of medications and toxic substances.

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IHS diagnostic criteria for BPV (Headache Classification Committee, 2004) (16) are as follows:

A. At least five attacks fulfilling criterion B
B. Multiple episodes of severe vertigo that occur without warning and resolve spontaneously after minutes to hours
C. Normal neurologic examination and audiometric and vestibular functions between attacks
D. Normal electroencephalogram

Prognosis

Prognosis of BPV is usually favorable for resolution. Many children develop migraine in later years.

Management

No specific treatment is necessary, apart from reassurance about the benign nature of the condition. Some children may benefit from the antihistamine betahistine (12) as a preventive agent.

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