Childhood Periodic Syndromes (CPS) – Learning from Triggers and Family

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ABSTRACT

Objective: To document childhood periodic syndromes and to suggest two clinically useful markers for diagnosing CPS.

Methods: Children and adolescents aged 3 to15 years presenting with recurrent head pain and migraine mothers were interviewed over a period of 5 years. ICHD2 (International Classification of Headache Disorders – edition 2) diagnostic criteria applied in all patients to diagnose migraine and CPS.

Results: Brief, benign paroxysmal episodes of vertigo were reported by 84, abdominal migraines in 62 and cyclical vomiting in 36.The most significant finding in the history was that, 138 were getting these symptoms when exposed to common migraine triggers like bus traveling, hunger, certain food, strenuous physical exercises, tension anxiety states and sleep disturbances. Either a single trigger or combination of triggers precipitated these manifestations suggestive of CPS. 131 first or second degree siblings were suffering from migraine (1.1,1.2,1.6).

Conclusion: CPS are not rare. Documenting common migraine triggers and family history of migraine will make CPS diagnosis easier in children and adolescents.

Keywords: Cyclical vomiting, Abdominal migraine, BPV of childhood, Common triggers, Family history

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INTRODUCTION

Childhood periodic syndromes are considered to be precursors to or associated with migraine head pain. International classification of headache disorders published in 2004 (ICHD2)[1] have included 3 clinical entities as CPS. 1) Cyclical vomiting (ICHD2 code 1.3.1) 2) Abdominal migraine (1.3.2) and 3) Benign paroxysmal vertigo of childhood (1.3.3). No significant studies have been done in India or other Asian countries regarding these syndromes. These syndromes were previously known as Migraine Equivalents.[2] Alternating hemiplegia of childhood was considered as one of the periodic syndromes in ICHD1[3] classification, but omitted from official ICHD2 diagnostic criteria and given in the appendix as it is not sufficiently validated by scientific studies.

Cyclical vomiting is a symptom complex that occurs in infants, children and less commonly in adolescents and adults. It is a self limiting episodic condition of childhood, with periods of complete normality between episodes. The clinical features of this syndrome resemble those found in association with migraine headaches, and multiple threads of research over the last years have suggested that cyclical vomiting is a condition related to migraine.

Abdominal migraine is described as an idiopathic recurrent disorder seen mainly in children and characterized by episodic midline umbilical pain manifesting in attacks lasting 1-72 hours with normality between episodes. The pain is of moderate to severe intensity and associated with vasomotor symptoms, nausea and vomiting.

Benign paroxysmal vertigo of childhood is characterized by recurrent brief episodic attacks of vertigo occurring without warning and resolving spontaneously in otherwise healthy children. Often associated with nystagmus or vomiting and unilateral throbbing headache may occur in some attacks.

MATERIALS AND METHODS

This 5 year study prospectively analyzed all the children and adolescents aged 3 to 15 years with recurrent headaches attending two clinics (Eye and Migraine clinic, Cherthala and St Sebastian's visitation hospital, Arthunkal) and charity camps. Migraine mothers

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attending these two clinics also were interviewed to find out the details about their children's headaches. Particular attention was given to the patients presenting with features suggestive of childhood periodic syndromes. Some children were referred by ENT Specialists to rule out ocular cause for vertigo (ocular vertigo). Migraine mothers were requested to bring their children for examination if history revealed any symptoms suggestive of CPS. Diagnosis was based on ICHD2 criteria and a headache questionnaire suiting to this region of India. Children diagnosed with cyclical vomiting and abdominal migraines were advised referral to pediatricians, surgeons or gastroenterologists to rule out gastrointestinal or renal disease and paroxysmal vertigo patients were persuaded to consult a neurologist and ENT specialist. Most of them with periodic syndrome symptoms had already consulted appropriate specialists as recommended by their family practitioners.

RESULTS

The study included 182 children and adolescents presented with features diagnostic of Childhood periodic syndromes. CPS alone were the first manifestation in life (precursors of migraine) only in 43 and in others it was associated with migraine.

Table 1. Summarizes the periodic syndromes	
Benign paroxysmal vertigo of childhood	84
Abdominal migraine	62
Cyclical vomiting	36
?probable abdominal migraine	269

Many children with migraine origin head pain (269) other than the 62 diagnosed as abdominal migraines were complaining of short duration abdominal pain of less than one hour duration (with normal pediatric and surgical assessment) and they were given a diagnosis of ? probable abdominal migraines (Unlike migraine with and without aura and tension type headaches, a diagnosis of PROBABLE is not recognized by ICHD2 in the case of CPS along with retinal migraines.)

Benign paroxysmal vertigo of childhood

Diagnostic criteria

A. At least 5 attacks fulfilling criterion B

B. Multiple attacks of severe vertigo, occurring without warning and resolving spontaneously after minutes to hours.

C. Normal neurological examination and audiometric and vestibular functions between attacks

D. Normal electroencephalogram

Our study population consisted of 84 children and adolescents. Age of onset of disorder varied from 3 to 14 years. More than 80% (68) of the children were from the age group 8 to 15. 69 had family history of migraine and 63 reported (by patients and parents) one of the common migraine triggers or combination of triggers as precipitating factors. All cases presented with duration of 3-10 seconds to 3 hours. Majority reported the attacks lasting less than 5 minutes. Most of them (61) were also getting probable migraine attacks (unilateral or bilateral throbbing pain, moderate to severe in intensity and precipitated by triggers) along with vertigo or independently. No patient was examined during a vertiginous attack. Examination done in between attacks were normal so also EEG[7] and audiometry[3] in some cases. 57 were seen by both Neurologists and ENT specialists before attending to our centers.

Collateral symptoms other than headaches accompanying vertigo attacks were pallor, nausea, vomiting, phonophobia, photophobia. and blurring of vision (all migraine symptomatology). The evolution of vertigo in our patients was as follows. All were advised avoidance of common migraine triggers whenever possible. All with recurrent vertigo attacks were put on prophylactic agents like flunarizine, cyproheptadine etc. 37 presented with complete remission of vertigo episodes at their last follow up. 31 reported considerable improvement in their symptoms. The rest were either not satisfied with the treatment outcome or got significant relief after first consultation and lost to follow up.

Abdominal migraine

Diagnostic criteria

A. At least 5 attacks fulfilling criteria B-D

B. Attacks of abdominal pain lasting 1-72 hours (untreated or unsuccessfully treated)

C. Abdominal pain has all of the following characteristics

   1. midline location, peri umbilical or poorly localized
   2. dull or just sore in quality
   3. moderate to severe intensity

D. during abdominal pain at least 2 of the following

   1. anorexia
   2. nausea
   3. vomiting
   4. pallor

E. not attributed to another disorder

Out of 62 children, 49 had central or peri umbilical pain, 13 with generalized pain. All became motionless (activity affected) during the pain period.48 had nausea or vomiting and 14, nausea and pallor. Anorexia was reported whenever the episodes were prolonged. Duration varied from one hour to one day. The diagnosis as told to them by their previous practitioners were appendicitis, gastritis, worm infestation, tension anxiety, vitamin and iron deficiency, food intolerance and functional. Appendicectomy done in 4, but were getting same pain after a short duration (up to one year) relief. 49reported triggers like certain food ingestion, hunger, traveling, physical exertion, sleep disturbances and tension anxiety situations at home or class. 51parents were migraineurs with similar triggers along with sun exposure.

During the study period, many migraine children (269) were reporting short duration pain abdomen (less than one hour) suggestive of probable abdominal migraine but not able to include them in the diagnosis of CPS, as ICHD 2 does not recognize PROBABLE childhood periodic syndromes of less than one hour duration.

Cyclical Vomiting

Diagnostic criteria

A. at least 5 attacks fulfilling criteria B and C
B. episodic attacks of intense nausea and vomiting lasting from 1 hour to 5 days
C. vomiting during attacks occurs at least 4 times / hour for at least 1 hour
D. symptom free between attacks
E. not attributed to another disorder

14 had repeated vomiting after every episode of fever or upper respiratory tract infections (infection as a trigger). Each bout lasting 1hour to 2 days. Diagnosis was confirmed after 5 such episodes were noted from the history. 20 reported long distance traveling (Palani, Thirupathi, Guruvayoor etc pilgrimage and picnic trips) as the precipitating factor. 14 children had less than 5 episode striggered by combination of triggers like sunlight, examination, funerals and long distance traveling. All of them had one of the parents or siblings suffering from migraine head pain. Sun exposure alone triggered vomiting episodes in 6, but not fully fitting into the ICHD 2 diagnostic criteria of 5 episodes and 4 times / hour for 1 hour. No complications like dehydration and electrolyte imbalance, hemetemesis etc were reported and only in one child the episodes lasted up to one week.

Abdominal migraine and Cyclical vomiting were diagnosed only after complete assessment by pediatricians, surgeons or gastroenterologists and ruling out other causes.

27 migraineur girls aged 12 to 15 were getting severe lower abdominal pain, recurrent vomiting and vertigo (all periodic syndrome manifestations in one)[4] during the first one or two days of their menstrual cycles. 9 children on follow up now, had cyclical vomiting as the first Periodic Syndrome in life (6 months to 18 months), followed by BPV and currently getting occasional probable migraine without aura episodes.

DISCUSSION

Many experts still having the opinion that it is not possible to propose criteria for delineation of the multiple heterogenous and undefined disorders comprised under the term Childhood Periodic Syndromes and it is unlikely that any progress will be made in this uncertain area until useful markers[3,5] are found.

The commonest of these syndromes diagnosed in this study is Benign paroxysmal vertigo of childhood. It is considered as an early manifestation of migraine related vertigo. In some population based studies the prevalence of recurrent vertigo probably related to migraine was estimated at 2.8% in children between 6 and 12 years. It remains an open question, if the addition of the newly proposed entity, Vestibular migraine or Migrainous vertigo[6] by Neuhauser and Lempert will lead to more clarity or more confusion as opined by T Brandt in his editorial commentary in Cephalalgia.[7] Basilar type migraine (previously Bickerstaff migraine, Basilar artery migraine or Syncopal migraine), where vertigo is one of the most significant diagnostic features and is considered as a fully reversible aura symptom, may create more confusion if all three are compared, especially in adolescent and adult migraineurs.

Vertigo/ dizziness attacks in migraine has extreme variation especially their duration, lasting from seconds to days, without associated headaches in about one-third of attacks, and their initial manifestation, occurring at anytime throughout life. The main features presented by our patients with BPV are in agreement with those reported in the literature[8,9,10] except the very high incidence of family history and triggers. Studies show that Benign recurrent vertigo in adults are occasionally precipitated by migraine triggers like alcohol, lack of sleep hormonal changes, food and emotional

stress but no study has so far properly documented triggers in children. The absence of impaired consciousness during the vertiginous attacks (develop sudden unsteadiness and grab on to whatever is near them), negative CNS exam and imaging, normal or non significant EEG findings are indicators in excluding a relationship between BPV and epilepsy. ENT exam performed in between attacks also were normal. Age of onset of 5 months as reported by Ulla Lindskong et al[9] may be very rare in Asian children. In her study only 39% had family history and none reported vertigo or balance disorder during the follow up period of 13 to 20 years. This study showed more than 80% with family history, though shorter duration, did get patients with recurrent vertigo during the follow up period of 5 years (in some patients) even with all possible trigger avoidance measures and multiple migraine prophylactic agents.

BPV of childhood is considered to be a rare and unknown disorder. Other than epilepsies, another important differential diagnosis which must be ruled out is posterior fossa tumors. Infections of the labyrnth or vestibular nerve is a diagnostic consideration if the episode is single and prolonged. Neuroimaging done in 17(13 CT and 4 MRI) were negative. Some of the atypical presentations were advised neuroimaging by us and others in the past, but couldn't get it done due to financial reasons.

CVS can affect both children and adults. Recent studies indicate that CVS affects 1.9% school going children and most often starts between ages 3 to 7. It was first described in the 19th century with one of the earliest ref being that of Samuel Gee in 1888.[11] This disorder was not included as a childhood periodic syndrome in the first edition of the International classification of Headache Disorders (ICHD1 1988). The clinical features of this syndrome resemble those found in association with migraine headaches, and multiple threads of research[12] over the last years have suggested that cyclical vomiting is a condition related to migraine. Majority of sufferers can identify triggers and this study documented triggers in more than 90% of the patients. The most common triggers documented are infections like viral fever, common cold, sinusitis, psychological stress, strenuous physical exercises, lack of sleep and certain foods. Hot weather, menstruation and traveling have been reported as triggers in the previous studies. Motion sickness is very common in migraineur children but a relation with cyclical vomiting is not clearly documented in the past. In the present study, cyclical vomiting was the first CPS manifestation in life as reported by 4mothers (recurrent vomiting during long distance traveling occurring during the first 6 months of life). In fact, motion sickness can be considered as a migraine trait[13] as many children with motion sickness develop migraine origin pain (moderate to severe throbbing) along with nausea or vomiting later in life while traveling. All triggers reported by CVS patients were identical to known migraine triggers. Either a single trigger or combination of triggers precipitated the attacks. CVS can be disruptive and frightening not just to children who have it but to family members as well.

Many studies[14,15] have recorded the relationship of migraine to recurrent abdominal pain. Similar pattern of attacks noted in this study have been recognized by most of them. The pain is located in the midline, peri umbilical or poorly localized and dull or just sore in quality. Pain is severe enough to interfere with normal daily activities and some children finding it difficult to distinguish anorexia from nausea especially when the pain episodes are prolonged. This condition is often confused with non specific or psychogenic abdominal pain. The literature[14] underlines the presence of functional recurrent abdominal pain (FRAP) as very common in day to day pediatric, surgical and gastroenterology clinical practice (may affect about 15% of middle and high school students) and some diagnostic features of this disorder have similarities with abdominal migraines. There is evidence to suggest that, in children, RAP occurring in the absence of headache may be a migrainous equivalent or an early expression of adult migraine. The data provided by Russel G, Symon DN and Mavromichalis I[14,16] give further evidence for a causal link and continuity between these two disorders. Family history of migraine is recorded in some previous studies but not included in the diagnostic criteria. The results of this study clearly show the significance of family history and known migraine triggers in diagnosing abdominal migraines especially in confusing clinical situations like short duration pain and generalized or non midline abdominal pain presenting without any clinical signs. The only rare entity that can be confused with brief abdominal pain episodes are abdominal epilepsies. There should be no difficulty in distinguishing this condition. Pain is of abrupt in onset, lasts only a few minutes and frequently associated with a change in consciousness such as disorientation or confusion. In this study, many children reported less than one hour duration of abdominal pain which is not officially recognized by the Inter-

national Headache Society as probable abdominal migraines. Accurate clinical diagnosis of abdominal migraine and probable abdominal migraine may spare children with this disorder many unnecessary gastrointestinal investigations and even laparotomy as well as long periods of psychiatric treatment in rare cases.

There are many other migraine equivalents (old terminology for periodic syndromes) described in the literature like hemianopia, scintillating scotomas, ophthalmoplegia, oculo sympathetic palsy, blurring of vision, amaurosis fugax, diplopia, mydriasis, hemiplegia, periodic fever, confusion-stupor etc that have not been sufficiently validated by research studies and thus not included in ICHD2. Benign paroxysmal torticollis (may evolve to Benign paroxysmal vertigo or migraine with aura) also is an appendix criteria in ICHD 2 as sufficient evidence has still not been published. No case of Alternating hemiplegia was detected during the study period of 6 years, thus justifying its removal from ICHD2 as a periodic syndrome and only one child presented with Benign paroxysmal torticollis which after 4 years started complaining of brief and probable migraine episodes.

CONCLUSION

Childhood periodic syndromes deserve further research as it is common and clinically relevant. In addition, it may help to clarify the pathomechanisms of migraine itself. To summarize,

1. CPS are not rare in this part of the world.
2. If one diagnostic criterion is missing from the present ICHD2 CPS criteria, including family history of migraine and common and known migraine triggers will be of immense help in diagnosing these entities.
3. Probable abdominal migraines lasting less than one hour are more common than current definite abdominal migraines(ICHD2, 1.3.2)
4. Menstrual cycles periodically triggering severe lower abdominal pain, recurrent vomiting and vertigo to be considered as classical examples of periodic syndromes and to be used in teaching models to highlight these disorders.
5. Many childhood periodic syndromes are associated with migraine head pain (ICHD1) and considering them as common precursors of migraine(ICHD2), especially in adolescents, is questionable.

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END NOTE

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