

# Pediatric Aspects of Headache Classification in the International Classification of Headache Disorders—3 (ICHD-3 beta version)

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**Abstract** This analysis looks at the applicability of the International Classification of Headache Disorders—3 beta (ICHD-3 beta) to various headache syndromes of children and adolescents. Areas of similarities and differences between adult and pediatric headaches are addressed as they relate to the ICHD-3 beta.

**Keywords** Children · Adolescents · Headache · International Classification of Headache

## Introduction

Headache disorders are common among the adult and pediatric populations. Given the absence of biological markers, establishing clear diagnostic criteria for headache syndromes is paramount for many reasons including the selection of appropriate treatment regimens as well as the recruitment of homogenous

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samples of patients for research purposes. The aim of this article is to explore the applicability of the International Classification of Headache Disorders—3 (ICHD-3) criteria [1] to various pediatric headache syndromes.

Determining whether the adult-based criteria in the ICHD-3 are applicable to pediatric patients is a difficult task. Some of these headache syndromes are rare in children and there are a limited number of publications from which one can analyze the headache characteristics. In some reports, the children are classified as having a specific headache syndrome, yet complete symptom details are not provided to determine if they would have fulfilled prior ICHD criteria for the headache syndrome. Also, some of these headache types have been reported in very young children who may be incapable of adequately describing the headache characteristics to an examiner. Often the examiner is establishing the diagnosis based solely on a witness's observation of the child's behavior. Studies that have assessed other clinical pediatric headache guidelines have noted them to be flawed based on their lack of adherence to evidence-based medicine [2].

Systematic assessment of the ICHD-3 criteria for the pediatric population is currently not available. We have analyzed the ICHD-3 (beta) criteria applicability to children and adolescents regarding various types of headache syndromes.

## Migraine

Migraine without aura and tension-type headache are the two most common types of headaches in children and adolescents. The prevalence of definite migraine varies from 5.9 to 82 % in pre-adolescent children depending on the definition criteria and reaches the adult figure of 17 % among women and 5.6 % among men in older adolescents [3, 4]. The frequency of probable migraine is more elusive in both children

and adults but thought to be at least as common as definite migraine [5].

The published pediatric studies are not uniform in the criteria used for diagnosis of migraine when comparing the utility of the previous ICHD classification systems. For instance, one study demonstrated the ICHD-2 to have higher specificity and sensitivity when compared to the ICHD-1 [6]. Another study demonstrated the ICHD-2 to have a lower specificity and sensitivity when compared to the ICHD-1 when short headache duration was excluded as a criterion [7].

Although studies are few in number, very high predictive value has not been established when the ICHD-3 was utilized to confirm the diagnosis of pediatric migraine. In a study of 783 students aged 12 to 18 years diagnosed with migraine by ICHD-3 criteria, a diagnosis of migraine was confirmed in 65.71 % of students who were re-evaluated 7 months after initial diagnosis [8••]. In this study, the predictive value was 76.88 % for definite migraine and 44.71 % for probable migraine. Another prospective study of 150 children and adolescents using ICHD-3 criterion noted that there was a 58 % sensitivity for pediatric migraine headache of greater than 2 h duration and a 94 % sensitivity for attacks less than 2 h duration using ICHD-3 criteria [9••].

The on-going debate about the lack of sensitivity, specificity, and predictive value in application of the ICHD-3 to pediatric migraine revolves around two main factors: unilaterality and duration of the headache. Most studies report that children and adolescents frequently report that their migraine headaches are bilateral rather than unilateral [10]. Also, the ICHD-3 increased the duration for migraine in children from 1 h in the ICHD-2 to 2 h noting that the evidence for untreated duration of less than 2 h in children has still not been substantiated. However, pediatric neurologists will frequently diagnose migraine in children with headache duration of less than 2 h, and even in very young children with a headache duration of less than 1 h. Studies in children with a clinical diagnosis of migraine demonstrate that 11 to 81 % have a headache duration of less than 2 h and 8 to 25 % have a headache duration of less than 1 h [10–13]. One study noted that these children with briefer headaches of less than 2 h would also fulfill the ICHD-1 criteria for migraine [14]. One recent study noted duration of clinically diagnosed migraine from 5 to 45 min [15•]. To correlate with this, some migraine equivalents in children are very brief, lasting seconds to minutes [16]. One study of 1134 children and adolescents with chronic headache (73.2 % with migraine) found migraine equivalents (abdominal migraine, cyclical vomiting, benign paroxysmal vertigo, and benign paroxysmal torticollis) to be so common that these should potentially be considered as part of the migraine syndrome [17]. The ICHD-3 lists these equivalents as separate disorders (1.6–1.6.3). Approximately 40 % of vertigo syndromes in pre-adolescents are considered to be related to migraine [18].

Other features of pediatric migraine warrant consideration for modification of ICHD-3 diagnosis in children. Family

history of migraine is so commonly elicited that it should be potentially considered as a criterion for pediatric migraine [19, 20]. One study noted that pulsating quality, an ICHD-3 criterion, was infrequent in younger children but was increasingly reported with increasing age [21]. There has been a longstanding suspicion that anxiety and depression are frequent co-morbid conditions associated with pediatric migraine and that a common neurobiological pathophysiology exists [22, 23]. This raises the issue whether it should be considered as a criterion for diagnosis. However, a more recent review questions this association between pediatric migraine and co-morbid psychiatric disorders [24]. Because of the scarcity of published studies in children, this merits further study.

In very young children, the diagnosis of migraine can be particularly challenging due to their inability to verbalize symptoms. Thus, diagnosis based on a witness's observations is often at the core of diagnosis. Our proposed diagnostic criteria for migraine in younger children, which emphasize objective findings rather than verbal reports, are summarized in Table 1.

There are also rare episodic syndromes, such as acute confusional migraine (ACM) and Alice in Wonderland syndrome that have not been included in the ICHD-3 and remain unclassified. Acute confusional migraine of childhood is almost exclusively seen in late childhood to adolescence, characterized by episodes of confusion with prominent impairment of sensorium and disorientation occurring with or without headaches. There may be complete or partial amnesia of this episode, and the attacks are usually relieved by sleep. There is insufficient data to estimate the prevalence of ACM, but some suggest that it is the rarest form of migraine variants [25]. Alice in Wonderland syndrome is an intense visual aura manifested by a distortion in size of objects (i.e. micropsia, macropsia) or time (sense of increased or decreased speed of movement or speech) that generally does not occur with headache, but usually occurs

**Table 1** Proposed criteria for migraine in children 5 years old and younger

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- A. At least five headache attacks fulfilling the criteria B thru D
  - B. Headaches lasting 30 min or longer (untreated or treated)
  - C. Unilateral or bilateral headaches with at least one of the following characteristics:
    1. Pain of at least moderate severity, indicated by disruption of normal activities or lack of engagement in playing
    2. Exertion intolerance; avoidance of walking; desire to rest
  - D. The headache is associated with at least one of the following:
    1. Inability to eat, presence of nausea and/or vomiting, stomach discomfort
    2. Sensitivity to light and sounds as indicated by inability to watch TV or play on the computer or other electronics
  - E. Not better accounted for by other diagnoses
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in persons with a history of, or the eventual development of, migraine [26].

### Tension-Type Headache

Similar to adults, the existing literature of tension-type headache (TTH) in children and adolescents report a wide range of prevalence [27]. Many argue that the diagnosis of TTH lacks sensitivity and specificity as the phenotype is defined by the absence of migraine features [28]. Moreover, many children and adolescents exhibit a mixed phenotype of migraine and TTH over time [28]. Longitudinal studies note that the diagnosis of TTH may convert to migraine, or vice versa, by the end of the teenage years [29]. Only one study has analyzed the applicability of the ICHD-3 criteria to children and found a predictive value of 69.71 % for definite TTH and 46.1 % for probable TTH [8••]. When examined 7 months later, 62.66 % had the diagnosis of TTH confirmed [8••].

There are several features of TTH that are probably unique to children and adolescents that, if confirmed in other studies, should be considered in future revisions of the ICHD. In a large study of 1738 children and adolescents that focused on meningeal signs in primary headache, 97 % of the 731 children with TTH had the entire set of meningeal signs that were analyzed, including nuchal rigidity, Kernig's sign, Brudzinski's signs, the "tripod" sign, and Guillain's and facial signs [30]. This has not been the experience of the current authors, but if confirmed, meningismus could be considered a cardinal feature of pediatric TTH. Another interesting association reported is pediatric TTH and constipation, with improvement of the headache with resolution of the constipation, which needs further study [31].

### Trigeminal Autonomic Cephalgia

Trigeminal autonomic cephalgias (TACs) are divided into five headache syndromes: cluster headache (CH), paroxysmal hemicranias (PH), short-lasting unilateral neuralgiform headache attacks with conjunctival tearing and injection (SUNCT), and short-lasting unilateral headache attacks with cranial autonomic symptoms (SUNA). The ICHD-3 has re-classified hemicrania continua (HC) to the TAC category. The hallmark of these headache syndromes is the presence of autonomic manifestations during the headache episode.

### Cluster Headache

There are several differences regarding cluster headache when comparing the pediatric and adult populations. From a review of pediatric CH, it was noted that the inter-cluster interval is

usually very prolonged in children and there is a shorter duration of the cluster bout which makes the decision about long-term treatment more difficult [32–34]. As the child enters the adult years, the frequency of cluster bouts and the duration of bouts increase [33]. Although children do have increased restlessness which helps distinguish it from migraine, the restlessness seems less than reported in adults with CH [33–37].

The restlessness may be difficult to characterize and has been reported simply as irrational or thrashing behavior leading to a misdiagnosis of hyperactive syndrome with psychogenic headaches [37]. Also, the autonomic features may be less prominent [32, 33]. Lacrimation of the eye is noted to be the most common autonomic symptom followed by conjunctival injection and nasal discharge [33].

In the ICHD-3, criterion D for general cluster, criterion B for episodic cluster, and criterion B for chronic cluster, which all define the duration and frequency of attacks, may not be applicable to children. We propose the following amendment to the ICHD-3 criteria for CH: compared to adults, the attacks may be less frequent and of shorter duration in younger children. Restlessness may not be severe and difficult to characterize.

### Paroxysmal Hemicranias

Both episodic and chronic paroxysmal hemicranias have been reported in children [38]. The hallmarks of these headaches are the strict unilateral location of the pain, the brief duration (ICHD-3 notes duration of up to 30 min) and the dramatic response to indomethacin. From the few reports, there are some pediatric features not in conformity with the ICHD-3. In very young children, the pain has been reported as midline in location [38]. In one author's (GNM) experience with six children, the pain is usually unilateral and on the same side at least 90 % (but not 100 %) of the time. Shifting sides (with one side predominant) and bilaterality of the pain have been reported in adults [39, 40]. Some reports noted an absence of autonomic symptoms required by the ICHD-3 [38, 41, 42]. Facial paleness rather than facial flushing (an autonomic criterion in the ICHD-3) has been noted in children [38]. The attacks often last longer than the maximum 30 min noted by the ICHD-3. Duration has been reported up to 45 min in children [38, 43–45]. Although the headaches are typically severe in children, the lack of missed school days in the 4 weeks prior to diagnosis raises questions whether all PH attacks are moderate to severe in intensity as required by the ICHD-3 [38]. Also, the response to indomethacin may be incomplete. Pediatric cases of incomplete response to indomethacin but with additional improvement with verapamil [41], aspirin [47], topiramate [45] and valproic acid [45] have been reported. Children with PH who had less than five attacks per day for more than half of the time as specified by the ICHD-3 have been reported [45,

46, 48, 49]. All other characteristics of the headache syndrome were consistent with adult PH. Also, migraine features, including vomiting, throbbing in nature, photophobia, osmophobia, as well as a family history of migraine, have also been reported; this is not mentioned in the ICHD-3 [38, 45].

### Hypnic Headache

Reports of hypnic headache in children are scarce. The hallmark of the headache is the arousal from sleep, often at the same time nightly. The ICHD-2 age requirement of onset after age 50 years was eliminated in the ICHD-3. A recent literature review of five children with hypnic headache (HH) [50], as well as one unreported child who is a patient of one of the authors (GNM), was used to evaluate the validity of the ICHD-3 criteria in children. Two of the children had duration of less than 2 months and would not fulfill ICHD-3 criteria requiring duration of 3 months or more; however, they were included in the analysis because their clinical features were so striking. The differences primarily relate to the frequency of the headache. In 3/6 children, criteria C (occurs >10 times per month for >3 months) could not be fulfilled. These children had one and two attacks per month. The other children varied from having 8–12 monthly attacks to near daily attacks. Similarities between adults and children with HH include bilaterality (5/6 children), frontal to frontal-temporal location (5/6), absence of autonomic symptoms (6/6) and brief duration (<30 min) (5/6).

### Exertional Headaches

Chen et al. utilized a questionnaire to study exertional headaches among 13–15-year-old adolescents [51]. They recruited 1963 students in Taiwan and estimated a prevalence of 30.4 %. More than half of the cases reported bilateral and pulsating headaches which lasted less than 1 h. Migraineurs were more likely than non-migraineurs to suffer from exertional headaches (EHs). EH in those who had fulfilled the ICHD-2 criteria for migraine were migraine-like, of longer duration and more likely to necessitate pain killers. They concluded that only 59.4 % of these adolescents fulfilled the ICHD-2 criteria A (pulsating pain), and only 54 % fulfilled criteria B for duration (5 min to 48 h). The criteria offered in ICHD-3 for exertion headaches have lifted these restrictive features and would accept headaches with any quality that occur during or immediately after exercise and last for less than 48 h. Even though there have been no studies assessing the ICHD-3 criteria, extrapolating from the above study, it may be concluded that the current criteria are applicable to children. It must be added that the current criteria are too inclusive and will not differentiate between true EH and

migraines provoked by exertion, a limitation that equally applies to children and adults.

### Primary Stabbing Headache

The core manifestations of primary stabbing headache are the same in children and adults. However, there are some ICHD-3 criteria differences. Children and adolescents have been reported to have a higher incidence of co-morbid migraine, TTH, and EH [52–54] although others disagree [42, 55–57]. The ICHD-3 criteria include duration of a few seconds, but some pediatric reports note duration of up to 15 min [52, 57]. Co-morbid migraine and TTH as well as a family history for migraine are not uncommon in this age group [52, 54]. An association with migraine is noted by the ICHD-3. Also, extracephalic pain (abdominal, low back, chest, and knee) has been reported as co-morbidity in a child [58].

### New Daily Persistent Headache

The reported pediatric series have used an ICHD classification, or a modified version, for diagnosis [59–63]. However, the reported pediatric series note two additional issues associated with childhood NDPH. First, many children have either an antecedent infection or head injury prior to onset [59–61]. Although various types of infection have been reported, the most common are Epstein-Barr virus or an upper respiratory virus [59]. Second, there have been many secondary causes associated with NDPH such as cerebral venous sinus thrombosis, low or high cerebrospinal fluid pressure syndrome, carotid/vertebral dissection, vasculitis, toxins, congenital anomalies, metabolic disorders, and neoplasms [59]. One study analyzed 27 adolescents with new daily persistent headache (NDPH) without medication overuse and noted that there were too many migraine features to fulfill ICHD-2 criteria (which are more specific than the ICHD-3 criteria are) [62]. In that study, only 51.2 % would be classified as NDPH according to the ICHD-2 criteria [62]. Other studies using a modified version of the ICHD-2 criteria have confirmed the frequent co-existence with migraine-associated symptoms, a feature noted in the ICHD-3 [63].

### Brain Tumor Headaches

Headaches associated with pediatric brain tumors would likely fulfill ICHD-3 criteria. But, more specific predictable factors exist in children. In a study of 3000 children with brain tumors, 98 % had at least one of the following findings: papilledema, ataxia, hemiparesis, abnormal eye movement, or depressed reflexes [64]. Also, because 2/3 of brain tumors

in children are in the posterior fossa, nocturnal awakening is common [65]. These are issues that should be considered future ICHD.

## Structural Lesions

The ICHD-3 notes that the primary headache syndromes are not associated with structural lesions. However, structural lesions have been reported in children who fulfilled the criteria for primary headaches. Pontine tumor has been reported in pediatric TAC with resolution of the attacks with debulking of the tumor [66], posterior fossa tumor in a child with SUNCT with partial improvement after surgery [67], and ipsilateral occipital infarction in a child with PH [44]. Many secondary causes of NDPH have been reported. [59] The occurrence of these structural lesions and other disorders in primary headache syndromes in children could be incidental.

## Conclusion

We have addressed several of the headache types that occur in children and adolescents regarding similarities and differences relating to the ICHD-3 criteria. Some of the features are distinguishable and should be considered in future revisions of the ICHD. In the case of very rare headache syndromes in children and adolescents, we are unable to offer specific comments due to the limitation of existing reports. Pediatric neurologists should be encouraged to report the unusual and rare types of headache in order to expand our knowledge in this area.

## Compliance with Ethical Standards

**Conflict of Interest** Gary N. McAbee, Anne Marie Morse, and Mitra Assadi declare that they have no conflict of interest.

**Human and Animal Rights and Informed Consent** This article does not contain any studies with human or animal subjects performed by any of the authors.

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