Primary Stabbing Headache in Children and Adolescents

Subjects: Pediatrics

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Primary Stabbing Headache (PSH) is characterized by brief, focal, and paroxysmal pain ("stab"), occurring sporadically or in clusters. Data on pediatric cases are poor. The prevalence of PSH and probable PSH varies from 2.5 to 10% among children with primary headaches and it is higher among children aged less than 6 years old. The mean age of onset is between 7 and 11 years of age. Attack duration greatly varies, ranging from a few seconds to several minutes. The intensity of pain is usually from moderate to severe. Associated symptoms are infrequent but may be observed (mainly vomiting). Neuroradiological findings photophobia, vertigo, nausea, and are usually unremarkable; Electroencephalographic (EEG) may show sporadic epileptiform abnormalities (up to 30% of cases). Preventive therapy is anecdotal, including treatment with indomethacin, trazodone, valproate, and amitriptyline.

Keywords: primary stabbing headache ; other primary headaches ; children ; adolescents

1. Prevalence

Previous studies show that the prevalence of PSH and probable PSH, among pediatric patients with primary headache, varies from 2.5 to 9.97% ^{[1][2]}, confirming that PSH is a common entity among children and adolescents. Data available in the literature are insufficient to estimate an overall prevalence among the pediatric population.

2. Mean Age of Onset

The mean age of onset is between 7 and 11 years of age ^{[2][3]}. A recent study by Saygi found that 16.9% of patients with PSH had clinical onset before 6 years of age ^[2]. Some cases in which the onset occurred as early as 2 years of age have also been described ^{[4][5]}. In a study cohort conducted by Raieli et al. in 2002, the mean age of onset among girls was 10.8 years (range, 2.6–17.8), while it was 8.8 years (range, 5.0–13.0) among boys ^[6]. Differently, Saygi found that the mean age of onset among girls was 10.1 years (SD ± 3.7), while it was 11.4 years (SD ± 3.4) among boys, not showing a statistically significant difference between genders ^[2].

A retrospective study by Raieli et al. analyzed the frequency of primary headache subtypes in a population of 105 cephalalgic children below 6 years of age; interestingly, the prevalence of PSH (12.4%) was significantly higher in the study cohort compared with a population of 100 cephalalgic children older than 6 years of age (3%) ^[Z]. These data suggest that PSH prevalence could be higher among young children.

3. Gender Distribution

The gender distribution among children with PSH greatly varies. In most studies, males and females were roughly equally represented $\frac{1}{3}\frac$

4. Presence of Other Headache Disorders

Moreover, the overlapping presence of other headache disorders was also taken into consideration by some authors. Ahmed et al. found that 12 patients out of 42 (29%) presented other headache disorders, mostly migraine, which is in line with the ICHD-3 comments on PSH ^[1]. Contrarily, in the study by Fusco et al., no previous history of other types of headache was reported among patients ^[8].

In the case study presented by Hofstadter-Duke et al., the patient was diagnosed with two additional types of primary headaches, other than PSH, including tension-type headache, occurring daily, and migraine, occurring with a weekly

frequency ^[11]. The patient identified Stabbing Headache, which occurred daily, as the most debilitating type of headache pain.

5. Family History of Headache Disorders

Most studies also analyzed the prevalence of a family history of headache disorder, finding it positive in 31-58% of patients, mainly represented by a positive history of migraine: in the case study by Takeshita et al., three out of five children (60%) also manifested migraine and had a relative who suffered from migraine ^[12]. The study by Ahmed et al. found that only 2 patients out of 42 (5%) had a relative who suffered from PSH ^[1].

6. Clinical Features

6.1. Type of Pain, Intensity, and Localization

According to the ICHD-3 definition of PSH, pain is characterized by a single stab or series of stabs. Therefore, the pain is most often described by patients as having a stabbing quality. However, in one study by Saygi up to 34% of patients reported a "throbbing" type of pain ^[2]; this may be due to the fact that in the pediatric population, it is more difficult to precisely describe the pain, but still the duration pattern and the associated symptoms were highly suggestive of PSH.

The intensity of pain ranges usually from moderate to severe, even if some patients with PSH in the studies by Soriani et al. and Saygi described mild to moderate pain, overall not affecting daily activities ^{[2][3]}.

Pain is often described as unilateral [1][3][5][7][9] and it is most often localized in the frontal region [1][2][3][6]. Occipital pain is also described among pediatric patients with PSH [10].

6.2. Duration and Frequency

The attack duration is undoubtedly the most difficult criterion to meet to make an ICHD-3 diagnosis of PSH because of its strict temporal range. In our experience, it is particularly challenging when interviewing pediatric patients because young children have difficulties distinguishing a single stab from a series of stabs. According to the ICHD-3 definition, each stab should last for up to a few seconds, specifying that most studies (on the adult population) report stabs that last 3 s or less and rarely for 10–120 s ^[13]. This limitation highlights the importance of "possible PSH", a diagnostic category introduced in the ICHD-3; "possible PSH" can be diagnosed even without meeting the duration criteria.

Considering the pediatric population, a remarkable variability in attack duration emerged when comparing different studies from the literature, ranging from a fraction of a second to several minutes. Raieli et al. reported that in most PSH attacks, pain lasts a few seconds, but 20% of patients reported some attacks lasting several minutes ^[6]. Fusco et al. reported a mean duration of 5 min ^[8]. Moreover, Ahmed et al. reported that some patients (23%) had symptoms lasting up to 15 min ^[1]. These patients, according to the definition of ICHD-3, would not fall under the diagnosis of PSH; however, as they do not fall under any other type of headache and meet the other criteria of PSH, they can be considered as "probable PSH" ^[13].

Concerning the duration of attacks, Fusco et al. reported a mean duration of 5 min, which reached 15 min in the Saygi et al. study ^{[2][8]}.

All studies investigated the frequency of attacks, which is more than once per week in most cases $\frac{11(2)(3)(6)(7)(8)}{100}$. A monthly frequency was reported in 15 to 37% of patients $\frac{11(6)}{100}$.

6.3. Associated Symptoms

Overall, in all studies, most patients did not report any accompanying symptoms. When complained, the most common symptoms were: nausea (7–14.3%), photophobia and/or phonophobia (2.6–19%), vertigo (1.3–8%), and vomiting (1.3–5%) ^{[1][2][3]}. Soriani et al. reported a high frequency (in 47% of patients) of periodic syndromes: mainly cyclic vomiting and recurrent abdominal pain, preceding the PSH onset ^[3]. Fusco et al. noticed that motion sickness was present in 8/23 subjects and vertigo in only one patient independently of the headache ^[8]. In the study by Vieira et al., patients had no associated signs or symptoms apart from slight pallor probably due to a vagal reaction (two out of 17 cases) ^[7]. Furthermore, a retrospective clinical study by Raieli et al. reported that headaches are associated with sudden painful grimaces (numbers not specified) ^[7].

6.4. Children vs. Adults

To better characterize PSH among children and adolescents, the researchers compared the pediatric studies included in the review with data from recent studies on the adult population, selecting some of the most recent reviews, aiming to find potential differences in clinical presentation between pediatric and adult patients. Stabbing quality of pain with variable intensity (mostly from moderate to severe) is described among adult PSH patients $^{[14][15]}$, which is in line with what arefound in the pediatric population. As reported for children, adults also present unilateral pain in most cases $^{[14][15][16][17]}$ [18]. Differently from our data, localization is often described in the occipital region among adult patients $^{[14][16][17]}$. Duration of pain is usually less than or equal to three seconds $^{[14][16]}$, and only in a minority of adult cases up to 60 s $^{[127]}$. These findings differ from what is reported in pediatric cases since long-lasting PSH attacks, sometimes up to 15 min, are described. Regarding associated symptoms, studies on adults report the presence of jolts (which are sudden movements such as grimacing or shrugging) following the jabs, body jabs, and allodynia as relatively common clinical findings $^{[14][16]}$ [127]. A clinic-based study by Fuh et al. also reported the presence of vocalization in 18% of patients $^{[127]}$. Interestingly, these symptoms are rarely reported in children: only Raieli et al. described a painful grimace occurring with a headache $^{[6]}$. Accompanying symptoms such as nausea or vomiting, dizziness, and photophobia/phonophobia during attacks have similar frequency among children and adults $^{[127][18][19]}$.

7. Triggering Factors

Six out of the twelve selected papers deal with triggering factors in pediatric PSH.

In most studies, no precipitating or triggering factors were clearly identified ^{[2][7][10][20]}. Soriani et al. found a psychogenic triggering event in 18 of 83 patients (22%) ^[3]. In the population studied by Ahmed et al., 7.1% of patients (n = 3) reported attacks triggered by exercise (n = 1) or stress (n = 2). The authors considered alternative diagnoses for these patients, such as primary exercise headache, but they did not match the required diagnostic ICHD-3 crieria regarding the frequency and features of headache attacks ^[1].

Studies in the adult population have found a precipitating factor in up to 50% of patients, particularly stress, recent illness, extreme weather conditions, or sleep disturbance/fatigue ^[21]. Interestingly, in the adult population, possible infectious triggers have been identified; for example, a case report described an adult patient who developed several attacks of PSH after COVID-19 infection ^[22].

8. Neuroradiological Findings

Seven out of twelve analyzed articles provide neuroradiological data about pediatric PSH.

Brain imaging, mainly performed as magnetic resonance imaging (MRI), was normal in almost all patients [1][2][3][7][10][20]. Fusco et al. reported one patient with cerebellar vermis hypoplasia with mild ventricular dilation ^[8]. In another study, radiological evidence of sphenoidal or ethmoidal sinusitis was detected (in 3/17 patients) in the absence of symptoms or signs of acute respiratory infection during periods of headache ^[7]. In the Mukharesh et al. study, a detailed vasculitis workup, MRI, and magnetic resonance venography were performed in one patient, and all results were within normal limits ^[10].

Considering studies on the adult population, similar data were found regarding the negativity of radiological examinations: many authors agree that, as in other primary headaches, radiological investigations should be performed to consider and rule out possible secondary causes of secondary aetiologies ^{[23][24][25][26]}. If PSH attacks have atypical features (such as longer duration, substantial background pain, or other red flags), computed tomography, or MRI of the brain or cervical spine is mandatory, especially in children under 6 years of age ^{[21][23][27]}.

9. Electroencephalographic (EEG) Findings

The diagnostic value of EEG is unclear and its use for the diagnosis or differential diagnosis of PSH is generally believed to be unnecessary ^[23].

Seven out of twelve selected articles provide EEG data about pediatric PSH.

EEG recordings were typically normal ^{[3][10]}. In some studies, interictal epileptiform abnormalities were reported (4–40% of cases) ^{[2][<u>6</u>][<u>7</u>][<u>8</u>]</sub>. In particular, in the cohort studied by Vieira et al., EEG was performed in 5/17 patients: three were normal, one had epileptiform abnormalities in the occipital region, and one in the frontal region ^[5]. In one of the studies, occasional}

posterior slow waves of high amplitude were observed in 24% of the cases, with marked slowing during hyperventilation ^[8]. A similar result was found by Raieli et al. (2002): epileptiform abnormalities (focal sharp waves, spikes, slow waves, and rolandic spikes) were observed in 20% of the performed EEGs (n = 6 patients). Of these patients, one reported focal epilepsy and intellectual disability, another had suffered a mild head injury about thirty days earlier, another patient had mild psychomotor retardation, and another one had a family history of epilepsy ^[6]. In another study, EEG abnormalities were found in 30.7% of PSH patients, but the difference with other primary headaches was not statistically significant ^[7].

It is difficult to interpret EEG data in PSH; in our opinion, it is not possible to state if these could be relevant for the diagnosis since not all patients systematically underwent electroencephalographic investigation.

13. Treatment

Five out of twelve analyzed studies provide findings about prophylaxis therapies in pediatric PSH and none provide findings about on-demand therapies. Concerning prophylaxis therapy, Mukharesh et al. 2011 found that most of the patients did not receive any treatment (3/5), and two patients were treated with amitriptyline (a single dose of 10–25 mg at bedtime) because of their frequency of stabbing pain headaches and associated migraine headaches, with significant improvement within the first months of treatment $\frac{120}{1}$. In the Raieli et al. study, seven female patients (23%) were selected for prophylactic therapies because of frequency (at least weekly) and severity of PSH attacks or because of other coexisting primary headaches requiring preventive treatment. Indomethacin (75 mg/day, in two patients), trazodone (0.5 mg/kg, in one patient), and L-5 hydroxytryptophan + riboflavin (100 mg + 100 mg, in two patients) were used with a reduction of more than 50% in headache attacks. One patient, suffering from 40–50 attacks per day, treated with carbamazepine (10 mg/kg/day) for two months achieved a total resolution of PSH attacks. In one patient treated with flunarizine (5 mg/day), no changes in the frequency or intensity of PSH attacks were observed ^[6].

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