Introduction

Idiopathic stabbing headache (ISH) is defined as transient stabs of pain confined to the head, predominantly in the distribution of first branch of the trigeminal nerve, lasting from fractions of a second to several seconds, and occurring in the absence of organic disease of the underlying structures or cranial nerves [1]. In the International Headache Society (IHS) classification, it is included in group 4 (miscellaneous headaches unassociated with structural lesions), with code 4.1.

Although described since 1964 [2], this disorder needs some clarifications about its clinical characteristics. Its prevalence in the adult population is controversial [3, 4] and the underlying pathophysiological mechanisms are unknown.

Patients and methods

During the period 1997–2000, a total of 838 children were referred for headache to the Department of Child Neuropsychiatry. Diagnosis was made in accordance with IHS criteria. Inclusion criteria were a history of recurrent (more than 5 attacks) episodes of local short headaches lasting from a fraction of a second to a few seconds, without accompanying autonomic signs and absence of organic disease.
Out of 838 children with headache, there were 30 subjects with suspect ISH. Electroencephalography (EEG), neurological examinations and psychological interviews were performed on in all 30 patients, while some patients were only asked to perform projective tests (e.g. the children’s apperception, the thematic apperception and the Rorschach tests) and the psychometric test. Other investigations, including routine laboratory testing (e.g. blood count, sedimentation rate), radiography, evoked potentials, and computed tomography (CT) of the brain, were carried out only in some patients to exclude other diseases.

Results

A total of 30 young patients (20 girls) referred to our observation for ISH were included in the study. The prevalence in the general population of headache sufferers (n=838) was 3.57%. The mean age of patients at referral was 10 years 1 month (range, 2.6–17.8) and the mean age at onset of attacks was 9.6 years (range, 2.2–16.1). For girls, the mean age was 10.8 years (range, 2.6–17.8) while for boys it was 8.8 years (range, 5.0–13.0). The quality of pain was stabbing in all patients.

The pain was bilateral in 30.0%, unilateral in 53.3%, unilateral alternating in 10.0% and central in 6.6% (Table 1). The main cranial localizations of pain were temporal (36.6%) and frontal (26.6%), but in 13.3% and 3.3% they were vertex and occipital, respectively; 23.3% of patients reported several sites of headache.

The duration of pain paroxysms was less than 2 seconds in 53.3% and a few seconds in 46.6% of cases. A portion of the subjects (20%) had some attacks that lasted for several minutes. The attacks occurred both as a single stab and as a series of stabs. The most symptomatic period lasted from several days to several weeks.

Table 1 Characteristics of the 30 children with idiopathic stabbing headache (ISH)

| Patients, n (%) |
|----------------|
| Side of pain   |
| Bilateral      | 9 (30.0) |
| Unilateral     | 16 (53.3) |
| Unilateral alternating | 3 (10.0) |
| Central        | 2 (6.7)  |
| Main localization of pain  |
| Frontal        | 8 (26.7) |
| Temporal       | 11 (36.7) |
| Parietal       | 3 (10.0) |
| Vertex         | 4 (13.3) |
| Occipital      | 1 (3.3)  |
| Nasal          | 1 (3.3)  |
| Variable/multiple | 7 (23.3) |
| Duration of attacks |
| ≤2 s           | 16 (53.3) |
| Seconds        | 14 (46.7) |
| Minutes        | 6 (20.0) |
| Intensity of pain |
| Intolerable    | 6 (20.0) |
| Moderate-severe| 19 (63.3) |
| Mild           | 5 (16.6) |
| Frequency of attacks during symptomatic period |
| Several times daily | 14 (46.7) |
| Several times weekly | 5 (16.7) |
| Several times monthly | 11 (36.7) |
| Other types of associated headache |
| Migraine without aura | 7 (23.3) |
| Tension-type headache | 6 (20.0) |
| Exertional headache | 1 (3.3)  |
| Electroencephalography |
| Focal spikes, spike and slow waves, rolandic spikes | 6 (20.0) |
| Aspecific alterations (e.g. focal or general slow waves) | 9 (30.0) |
The intensity of headache was intolerable in 20.0% of subjects and moderate to severe in 63.3%. The frequency of attacks reported for the symptomatic period was several times daily in 46.6%, several times weekly in 16.6%. Overall, patients reported a wide range of frequencies, from a few times monthly to 40–50 attacks daily.

No reported symptoms were generally associated with nausea, vomiting, photophobia, phonophobia or local autonomic features. A family history for migraine was present in 11 patients (36.7%). No personal history of IHS childhood syndrome-related migraine [1] was found.

In this population, other types of headaches were coexisting in 46.6% (7 had migraine without aura, 6 had tension-type headache and 1 had exertional headache). The neurological examination for other headache causes was normal. The psychiatric examination and psychological tests (as the family’s design) disclosed anxious-depressive traits in 50% of subjects.

EEG was carried out also to investigate the contradictory results reported in literature regarding the prevalence of altered EEG in the juvenile ISH population [5, 7]. In 6 subjects (20.0%), epileptiform anomalies (focal sharp waves, spike and slow waves, rolandic spikes) were observed. One patient had partial epilepsy and intellectual deficits, another had had a mild head injury about thirty days before his referral, a third patient had mild psychomotor deficits and a last patient had a positive family history for epilepsy.

In 7 female patients (23.3%), prophylactic therapy was started (Table 2). Patients were selected for prophylaxis if they had high frequency and severe intensity of attacks or when another invalidating primary headache was associated with ISH. Therapy was successful, especially in an 8-year-old girl who had been suffering from 40–50 attacks daily for two months and whose treatment with carbamazepine achieved a quick response with the disappearance of attacks.

At follow-up by phone interview (range, 2–48 months), we found a general improvement in the frequency of attacks and in the intervals between disappearance and appearance of attacks for variable periods (data not shown); however the attacks persisted in almost all patients (83.3%).

### Discussion

ISH has been defined as a rare form of headache, with a prevalence of about 2.0% [3] in the general adult population. Recently, however, a study that specifically investigated the very brief, short pain paroxysms reported a prevalence of 35.2% in the general population [4]. The authors of this study explained that this wide difference is probably due to the fact that the first study was designed more appropriately for major headache forms and did not completely detect less frequent headaches. To the best of our knowledge, there are no prevalence studies on ISH in the pediatric population. In the only extensive clinical study by Soriani et al. [5], and in another study [7], the prevalence of IHS among headache patients was 3.35% and 5.1%, respectively, similar to that found by Rasmussen and Olesen in adults [3]. In our study the prevalence was 3.57%, similar to the two other pediatric studies. Yet, we agree with Sjaastad et al.’s opinion [4] that ISH is underestimated in the general population and also in the juvenile age. In fact, in our population, as in Soriani et al.’s [5], ISH was the main cause for referral, while in the general population ISH can fail to be noticed because of the low frequency as well as the brief duration of attacks which do not cause a severe morbidity in comparison with other primary headaches. Moreover, not all such patients are referred to neurological services and the diagnosis may be ignored because of the poor knowledge of the disorder.

In our study all patients had very brief attacks, as required by the IHS criteria, while in the other two studies [5, 6] patients with episodes lasting some minutes were included. This could affect the differential diagnosis with

### Table 2 Prophylactic therapy utilized in 7 girls with ISH

| Age, years | Diagnosis | Frequency of attacks | Intensity | Treatment | Efficacy       |
|------------|-----------|----------------------|-----------|-----------|----------------|
| 15.7       | ISH + episodic TTH | 4–5/day               | Severe    | Indomethacin (75 mg/day) | Attacks reduced >50% |
| 17.1       | ISH       | 6–7/day               | Severe    | Indomethacin (75 mg/day) | Attacks reduced >50% |
| 8.0        | ISH       | 40–50/day             | Severe    | Carbamazepine (10 mg/kg) | Attacks abolished |
| 7.1        | ISH + TTH | 4–5/day               | Severe    | Trazodone (0.5 mg/kg)    | Attacks reduced <50% |
| 9.2        | ISH + migraine | Weekly              | Severe    | L-5 hydroxytryptophan (100 mg) + riboflavin (100 mg) | Attacks reduced >50% |
| 8.3        | ISH + migraine | Weekly              | Moderate/severe | Flunarizine (5 mg/day)       | No change            |
| 17.8       | ISH + migraine | Weekly              | Intolerable | L-5 hydroxytryptophan + riboflavin | Attacks reduced >50% |

**TTH**, tension-type headache
other primary headaches, such as episodic tension-type headache that in children can show a brief duration of painful attack (from 5 to 20 minutes). The frequent psychogenic triggering events (22%) reported in Soriani et al.’s patients [5] could be due to misdiagnosed tension-type headaches. However, other authors have also reported that some stabbing attacks can be precipitated by emotional stress [8]. Moreover, in the paediatric age it is not easy to establish the exact duration of a headache; therefore the only precise reference is the determination of a clear temporal cutoff (seconds vs. minutes or hours).

In our opinion, in order to explain and better understand the clinical and neurophysiological characteristics of this headache with respect to the phase of our study, we think it is convenient to comply to the strictly temporal criteria of ISH (maximum duration, a few seconds) and also suggested by Sjaastad et al. [4], who established a maximum duration of 3 s. In a later stage, other studies could verify if patients with longer pain duration are in the same group or in a different group.

The age at onset and at first consultation of patients in our study are similar to those in the study by Soriani et al. [5], but the sex ratio is different (1:1 in Soriani et al.’s study). Our findings are similar to those of Kramer et al. [7] and those in adults [3, 4], but further studies are needed to give clearer answers.

The frequency of attacks was at least daily or weekly in 63.3% of individuals during the active period, similar to those reported in the adult age [6]. The major symptomatic period lasted from some weeks to 2–3 months and the headache could be very upsetting, especially in those patients with attacks of intolerable intensity and more daily frequencies, besides the brief duration of pain.

In Soriani et al.’s study [5], the intensity of attacks was mild in 40% of cases; in our population only 17% reported mild pain and 20% of cases described the pain to be intolerable.

ISH shows also a localization in extratrigeminal regions exactly as reported by other studies [5, 6, 9] and therefore the topographical IHS criterion does not seem to be mandatory.

This type of headache exhibited an irregular temporal pattern with erratic alternation of more symptomatic periods and almost silent periods at follow-up. However, is spite of a general improvement, the attacks reoccur through the years, as reported by Soriani et al. [5].

An interesting feature in our sample is the large prevalence of anxious-depressive traits, identified by psychological examination and projective tests. This can account for the observed reduction of frequency of the attacks after the first neurological examination as well as the benign nature and course of the disorder. Other studies are needed to understand the relationship between this headache and psychological factors and to identify possible different subgroups of subjects.

In our study the high prevalence of coexisting migraine, reported in literature for adults [7, 10, 11], was not confirmed. It is difficult to explain these contradictory results. A possible explanation is that ISH is different in juveniles and adults. Another hypothesis is that ISH has pathophysiological mechanisms, frequently involved in migraine, which account for the high prevalence of coexisting migraine in the ISH population. However, the incomplete maturation, during development, of the involved cerebral structures could allow a more frequent dissociation between ISH and migraine in the juvenile age. Nevertheless, the strong association with hemicrania continua, a clinical form of headache well distinguished from migraine, the typical response to indomethacin in adult ISH and the prevalence of ISH reported by Sjaastad et al. [4] show, in our opinion, that in adult age the association between migraine and ISH, too, needs confirmation with further studies in the general population.

For some patients a short prophylactic therapy was necessary; in two subjects age dever 14 years, indomethacin was used with a good efficacy. In a younger girl, reported previously more in detail [12], who suffered from 40–50 attacks per day, we observed a dramatic improvement with the use of carmamazepine, after several attempts with NSAIDs. This could suggest a paroxysmal neuronal discharge, like a trigeminal neuralgia [13]. However, the bilateral and extratrigeminal localization, and the alternating side suggest a neuronal discharge by a disorder of the central pain control and not a local neuralgic disorder [14]. Another possible demonstration of altered central pain control by hyperexcitability comes from Kramer et al. [7] who reported a higher number of interictal EEG abnormalities in children with very brief pain episodes. In our study, 6 patients showed interictal epileptiform alterations, in a way similar to this study. However, the presence of a patient with a partial epilepsy and the limited number of individuals in our sample make it difficult to provide clearer explanations. Another study does not support this higher prevalence of EEG abnormalities in juvenile ISH [5].

ISH is easily diagnosed with respect to other short-lasting headaches and cranial neuralgias, for its very brief duration, not rare bilateral localization, lack of trigger points and/or involvement of local and general autonomic disorder. We excluded trigeminal neuralgia, or SUNCT (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing) and ophthalmic disorders by clinical examination, clinical history and laboratory and neurophysiological tests [6, 8, 14, 15].

Neuroradiological examinations are not usually necessary for typical and univocal clinical characteristics, the benign course and the certain absence of neurological signs. However, a recent report [16] described ISH associated with monocular visual loss with complete resolution of symptoms after treatment with oxygen inhalation and indomethacin.
The benignity of ISH is underlined by the fact that, to our knowledge, secondary stabbing headaches have never been described as having only an acute and short stabbing pain with negative neurological and objective examinations. A recent report [17] evidenced the presence of ISH in two patients with meningioma, but both were also suffering from presented autonomic signs and continuous headache between attacks. Finally, in the study by Soriani et al. [5], the neuroimaging studies were negative.

This type of pain in adults can be concomitant to other disorders like hemicrania continua, cluster headache, chronic paroxysmal hemicrania, migraine and tension-type headache [7], but that is not confirmed in juvenile age [5].

The pathophysiology of ISH is unknown. A vascular basis has been proposed, especially for the suggested association with migraine [13], but the reported response to indomethacin, the very brief duration of pain, the association with other forms of headache like hemicrania continua and cluster [6] do not support it and a neurogenic mechanism is more likely. After examination of the clinical characteristics (relatively frequent bilateral, lack of local autonomic signs and trigger points and frequent multi-pain foci) an abnormal central control pain appears more likely than local neurogenic mechanisms.

In conclusion, ISH is not rare in the pediatric age group and in some cases it can be so invalidating to require a treatment. The prognosis is usually benign and complex examinations are not necessary. Further studies are needed to explain the mechanisms of pain and the role of psychological factors in this disorder.

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