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Hemicrania continua

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Abstract Hemicrania continua was described in 1984 by Sjastaad and Spierings as a unilateral, almost always side-locked headache that is continuous and fluctuating in intensity. There are added pain exacerbation periods of varying frequency, consisting of more severe headache, of at least moderate intensity, lasting from hours to days. The condition is either absolutely responsive to indomethacin or associated with autonomic signs or idiopathic stabbing headaches (jabs and jolts) during pain exacerbations. There is a lack of precipitating mechanisms.

Key words Hemicrania continua •
Chronic daily headache •
Trigemino-autonomic cephalalgia •
Treatment

Hemicrania continua (HC) was first recognized and named by Ottar Sjastaad of Trondheim, Norway, and Egilius Spierings of Boston, USA, in 1984 [1]. The authors initially considered the condition to be rare but it may be more common than they thought, with approximately 100 cases reported in the literature [2, 3]. While it affects both men and women, it is twice as common in women, unlike cluster headache, another unilateral trigemino-autonomic cephalalgia which is over twice as common in men. It could be the most misdiagnosed of all the primary headache disorders, which are the headache

conditions for which no organic cause is found.

Clinically, HC presents as a unilateral headache that is continuous and usually of mild or moderate intensity. There may be added pain exacerbation periods of varying frequency consisting of severe headache that lasts for hours or days with associated migrainous and/or autonomic symptoms. There are two forms, the continuous form which accounts for 85% of cases, and the relapsing and remitting form which accounts for the remaining 15%. The continuous form may start out as continuous or transform from the remitting form [2, 3].

The International Headache Society has a new classification of headache entitled *International Classification of Headache Disorders* (2nd edition) published in 2004 [4]. The criteria for diagnosing HC can be seen in Table 1. Note that HC is classified under Chap. 4, "Other Primary Headaches", and not with the more common forms like migraine and tension-type headache, nor with the other trigemino-autonomic cephalgias (cluster headache and paroxysmal hemicranias).

Most patients with HC have mild pain most of the time which can be interrupted by exacerbations of more severe pain. During the mild pain, patients describe a continuous, unilateral pain, usually in the temple, periorbital, or ocular area. It is almost invariably on the same side throughout the course of the illness. On rare occasions it has been known to switch sides or occur bilaterally, which does not fit with its name.

During exacerbations, patients have a variety of associated symptoms. Some of them are termed migrainous symptoms such as nausea, vomiting, photo- and phonophobia. Others are autonomic symptoms including ptosis, conjunctival injection, lacrimation, nasal congestion, and rhinorrhea. Other key symptoms are eyelid edema, eyelid twitching, ice pick headaches

Table 1 Classification of hemicrania continua (HC) according to the IHS criteria (2004)

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- A. Headache present for at least 2 months fulfilling B–D
- B. All of the following characteristics:
- Unilateral pain without side shift
 - Daily and continuous without pain-free periods
 - Moderate intensity (can be severe during exacerbations)
- C. At least one of the following during pain exacerbations and ipsilateral to the pain:
- Conjunctival injection and (or) lacrimation
 - Nasal congestion and (or) rhinorrhea
 - Ptosis and (or) miosis
- D. Complete response to indomethacin
- E. Not attributed to another disorder
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Table 2 Clinical features of ten subjects with HC evaluated at a headache center (from [6])

Unilateral without side shift:	100%
Absolute response to indomethacin:	100%
Able to tolerate prolonged treatment with indomethacin:	70%
Continuous pain:	100%
Moderate pain:	80%
(two patients had mild pain without exacerbations)	
Autonomic features during exacerbation:	70%
Migraine symptoms during exacerbation:	30%
Response to antimigraine drugs:	10%

(also termed "jabs and jolts" or "idiopathic stabbing headache") and the sensation that there is a foreign body in the eye. Peres and colleagues reported some cases that developed auras [5].

We performed a descriptive study on ten consecutive patients in our headache center in 2002 [6]. The results can be seen in Table 2. All ten patients had continuous, unilateral, side-locked headache responsive to indomethacin. The great majority had autonomic features: 40% had lacrimation and conjunctival injection; 30% had nasal stuffiness, rhinorrhea, ptosis, and eyelid edema.

The etiology of HC is usually idiopathic. A few reports exist of post-traumatic forms, following a surgical procedure or organ transplant. There are rare secondary mimics including cervical disc herniation, mesenchymal tumor involving the sphenoid bone, and HIV [7].

The evaluation of a patient with a unilateral headache should usually include magnetic resonance imaging (MRI) of the head, possibly an MRI of the cervical spine, and even magnetic resonance angiography of the neck if there are associated focal neurologic symptoms (to rule out carotid dissection).

The treatment for HC comprises indomethacin. Indomethacin should be started at a dose of 25 mg p.o. t.i.d. for 3 days. If the patient is not 100% pain-free within that time frame, we increase the dose to 50 mg p.o. t.i.d. for another 3 days. If there is only a partial response, we increase the dose by 25 mg every 2–3 days up to a maximum of 75 mg p.o. t.i.d. We rarely raise the dose higher. In special circumstances, doses can be raised to 300 mg/day.

Once the patient has an adequate response, we look for the lowest effective dose and maintain the patient at that level for an extended period of time. For breakthrough pain we try an additional dose of indomethacin 25–50 mg p.o., every 4h, p.r.n. If the patient remains on indomethacin we either add a histamine type-2 blocker plus misoprostol (100–200 µg q.i.d.) or one dose of a proton pump inhibitor daily.

Antonaci described the Indotest in countries where indomethacin was available by injection. He administered 50 mg intramuscularly and noted that patients with HC were pain-free within 73 min on average and the improvement lasted for 13 h [8].

Other medications have been tried in open studies and found to be occasionally successful. They include rofecoxib, gabapentin, lamotrigine, methysergide, corticosteroids, and dihydroergotamine [9–11].

In conclusion, HC is an uncommon primary headache that can be easily missed. Once the diagnosis is made and other organic conditions are excluded, the patient should do well with the proper dose of indomethacin.

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