

Hemicrania Continua

This fact sheet is intended as a general introduction to hemicrania continua which is a rare form of primary headache more common in women.

What is hemicrania continua?

Hemicrania continua is a primary chronic daily headache disorder. Headache disorder is classified as primary when an underlying organic cause has been carefully ruled out by medical history, physical, neurological examination and/or appropriate investigations.

Hemicrania continua is characterised by a continuous, fluctuating, unilateral pain without shifting sides of the head. The pain is, typically, mild to moderate in intensity and in the majority of patients episodes of severe pain are superimposed on the continuous baseline pain. During these episodes (called exacerbations, which can last from 20 minutes to several days) the sufferer may experience a variable combination of autonomic features such as eye-watering, eye redness, nasal blocking or running, and eye lid drooping. These will occur on the same side of the pain. Also migrainous symptoms such as light or sound sensitivity, nausea and vomiting are also present. The condition is usually unremitting, however, some patients have an episodic or remitting form with distinct headache phases separated by pain-free remission.

The incidence and prevalence of hemicrania continua are unknown. Hemicrania continua is more common in women. The condition usually starts in adulthood, yet the range of onset is from 5 to 67 years of age.

The hallmark of this syndrome is the absolute positive response when treated with a medication called indomethacin.

Are there different types of hemicrania continua?

There is only one type of hemicrania continua. This syndrome is a relatively new entity. In fact it was not described in the first edition of International Classification of Headache Disorders [ICHDs] by the International Headache Society in 1988. The syndrome was officially included in the last update of ICHDs in 2004.

Headache or hemicrania continua?

Distinguishing between different types of headache can be difficult. In fact patients can experience different types of headache at different times of their life for varying reasons. For example, if you have hemicrania continua you may also experience other types of headache such as migraine. It is always very useful to keep a detailed headache diary in order to identify different headache types as it leads to different treatments.

What is the treatment?

The treatment of choice for this condition is indomethacin. The positive response to this drug is an essential criterion for the diagnosis. Nevertheless, other treatments appear to be useful in this condition. There is currently no cure for hemicrania continua.

What causes hemicrania continua?

The cause of this syndrome is not known. It is also not clear whether there is a genetic element as in migraine. There are a lack of known factors that precipitate hemicrania continua. Precipitants that are important in other headaches (e.g. migraine) generally seem to be less important in this condition. This includes stress, lack of food, alcohol, hormonal changes in women, lack of sleep.

The importance of a diagnosis

The diagnosis is based on medical history, neurological examination and the positive improvement in response to indomethacin. It is important that the trial with indomethacin is carried out under the supervision of a doctor as this drug is contra-indicated in some medical conditions, particularly stomach disorders, whereas caution is required in others. The achievement of the right diagnosis is an essential step in order to properly treat hemicrania continua and therefore help patients to control their daily pain.

Hemicrania continua diary

Keeping a headache diary is pivotal for the doctor in order to have accurate information regarding the headache and also to evaluate carefully the response to medical treatments. Useful information that the headache diary may provide include:

- The location of the pain
- When the exacerbation (or worsening) of the pain started
- How often the exacerbations occurred
- How long the exacerbations last
- If there are other symptoms during severe pain, such as eye watering, redness of the eye, nasal blocked, runny nose, being sick and sensitivity to lights, sounds, smell and movement.

In addition, it is useful to record in the diary as many aspects of daily life as possible, such as:

- Medication taken
- Vitamins or health products taken
- Women should record details of their menstrual cycle.

Hemicrania continua and classification

A group of Doctors who are part of The International Headache Society have drawn up a list of broad symptoms for helping diagnose many headaches including hemicrania continua. The following chart highlights the current criteria for this condition in the **International Classification of Headache Disorders (second edition 2004)**.

Hemicrania continua

Description: persistent headache which is always responsive to indomethacin

**Diagnostic
criteria**

A. Headache for more than 3 months fulfilling criteria B-D

B. All of the following characteristics:

- unilateral pain without side-shift
- daily and continuous, without pain-free periods
- moderate intensity, but with exacerbations of severe pain

C. At least one of the following autonomic features occurs during exacerbation and on the same side of the pain;

- conjunctival injection and/or lacrimation
- nasal congestion and/or rhinorrhoea
- ipsilateral miosis and/or ptosis

D. Complete response to therapeutic doses of indomethacin

E. Not attributed to another disorder