



Paroxysmal hemicrania

Paroxysmal hemicrania (PH) is a rare form of headache under the classification of Trigeminal Autonomic Cephalalgias (TACs). PH is a debilitating one-sided headache affecting the area around the eye. It is often treated with non-steroidal anti-inflammatory drugs – indomethacin is most commonly effective in stopping symptoms. This information sheet from Great Ormond Street Hospital (GOSH) explains the two types of PH with episodic paroxysmal hemicrania and chronic paroxysmal hemicrania and where to get help.

What are Trigeminal Autonomic Cephalalgias (TACs)?

TACs tend to be short duration severe headaches around the eye, that are boring/drilling in nature, lasting for seconds to minutes and can be incapacitating. They tend to be associated with nasal congestion, swelling of the eyelid, tearing, reddening of the eye (conjunctival injection) and facial swelling.

What is PH?

PH is one type of TAC that normally involves multiple, sudden-onset, severe, short-lasting attacks affecting one side of the head and is more commonly diagnosed in women. Paroxysmal means periodic and hemicrania means 'one side of the head'. There are no associated features such as photosensitivity (sensitivity to light) or nausea/vomiting unlike in migraine. PH is classified by the frequency and duration of attacks experienced by patients.

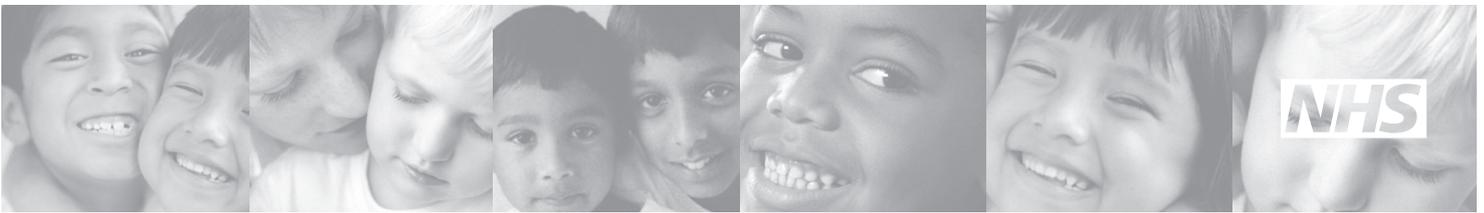
Episodic PH attacks occur at least twice a year and last anywhere from seven days to a year with pain-free periods of a month or longer separating them. Chronic PH attacks occur over the course of more than a year without remission (phases without symptoms) or with remissions lasting less than a month. Chronic PH is the more common form, and although mainly occurring in adults, it is well recognised in children as young as three years of age.

How do we diagnose PH?

To diagnose PH, 20 or more attacks of severe one-sided headache must occur in the child or young person, lasting between five minutes and half an hour. On most headache days, there tend to be five or more attacks. At least one of the following is associated with the attacks:

- Conjunctival reddening or tearing on same side as the headache
- Nasal congestion or runny nose on same side of headache
- Swelling or drooping of the eyelid on the same side of headache
- Forehead or facial swelling on the same side of headache
- Restlessness or agitation

PH may be confused with cluster headache. When people with PH have constant background pain between attacks, there is a considerable overlap in the signs and symptoms of PH and cluster headache. Both are strictly one-sided, relatively brief but frequent headaches that occur in association



with other symptoms affecting the same side of the head. At present, the only way to tell the difference between PH and cluster headache is that PH responds well to treatment with a medicine called indometacin.

How do we treat PH?

This is one of the few primary headache disorders that has a consistent and significant response to medication. The treatment of PH is prophylactic (preventative) and it has a rapid and consistent response to indometacin. The dose for children is around 25mg three times a day. The dose may be adjusted depending on the frequency and severity of symptoms.

When the child or young person is being tried on indometacin, a stomach-protective agent should be added in order to reduce the possible gastric problems, especially if patients require long term treatment.

Unfortunately, there are no trials for the management of PH when indometacin cannot be tolerated. There has been limited success reported with the use of other medications such as cyclooxygenase-2 (COX-2) inhibitors such as rofecoxib and celecoxib, in adults. However, these medicines have been linked to serious side effects and are not recommended in children. Topiramate and greater occipital nerve injection with lidocaine (local anaesthetic) and methylprednisolone (steroids) are helpful in some patients.

What is the long term outcome of PH?

Many people experience complete to near-complete relief of symptoms following supervised medical treatment. PH in some cases may last indefinitely but has been known to go into remission or stop spontaneously.

The headache clinic at GOSH

There is a headache clinic at GOSH, which aims to help children and young people when the diagnosis of headache is not certain and treatment has failed. Referrals need to be made by your local paediatrician to Dr Prabhakar – Consultant in Paediatric Neurology. We encourage family doctors (GPs) to make referrals via their local paediatric team. If you would like to know more about the headache clinic at GOSH, please visit www.gosh.nhs.uk/medical-information/clinical-specialties/neurology-information-parents-and-visitors/clinics-and-wards/headache-clinic or call us on 020 7405 9200 ext 0182.

Further information and support

The Migraine Trust offers support and advice to anyone affected by migraine or headaches. Call them on 020 7462 6602 or visit their website at www.migrainetrust.org. They offer specific information for children and young people.

Migraine Action also offers support and advice, and also has separate sections for children and young people. Their helpline number is 0116 275 8317 and their main website is www.migraine.org.uk. The section for 8 to 10 year olds is at www.migraineadventure.org.uk, the section for 11 to 13 year olds at www.migraineexplorers.org.uk and the section for young people aged 14 to 17 years is at www.migrainenetwork.org.uk.