

# What is cluster headache?

## Fact sheet for patients and their families. A publication to mark Cluster Headache Day 2016



In the last year the European Headache Alliance (EHA) has launched the “What’s Under the Hat?” story-telling campaign, encouraging headache patients to share their stories of the impact that headache disorders have on their life. The goal is to inspire public compassion for headache sufferers by making the invisible visible, and also to provide a platform to give patients a voice (<http://www.europeanheadachealliance.org/under-the-hat>).

On 21st March 2016 the EHA celebrated the first Cluster Headache Day as part of the *What’s Under the Hat?* public awareness campaign. Even though cluster headache is considered the worst pain known to man it is largely underestimated. In fact more than 600,000 people in Europe live with cluster headache, with less than 50% of these seeing a specialist and more than a third having to miss work because of the condition, at a cost of 7 billion euros per year. This fact sheet has been prepared by expert headache patients and headache specialists to educate patients and their families about what cluster headache is.

### What is cluster headache?

- Cluster headache is a condition considered to be one of the worst pains known to man.
- It affects up to 4 in 1000 people, similar to the incidence of multiple sclerosis and Parkinson’s disease.
- It affects more men than women, making it unusual among headache disorders.
- A family history of cluster headache is rare (only 3–5% of cases are hereditary).
- The word ‘cluster’ refers to a period of time (weeks or months) during which the individual suffers recurrent attacks (cluster periods or bouts); the start of bouts often coincides with season changes, and with the times of year with the most or fewest hours of daylight.
- Cluster headache presents in two distinct clinical forms: episodic and chronic. The most common is the episodic form which affects 80 to 90% of patients. In episodic cluster headache, subjects have cluster periods, or bouts, during which they experience one to eight attacks per day (with or without pain-free days) and periods of remission lasting more than 30 days. The typical patient has one bout per year, often at the same time of year. The chronic form is diagnosed after one year of pain without remission, or in subjects with remission periods of less than one month.

Attacks consist of a severe stabbing pain rapidly peaking at an unbearable intensity, usually lasting between 15 and 180 minutes, affecting one side (and nearly always the same side) of the head, and usually located in and around the orbit and in the temporal area. The pain is often likened to a ‘hot poker’ penetrating one eye.

During bouts, attacks recur at typical times, such as the middle of the night and around 3 and 9 p.m.

Out of a number of symptoms at least one will also occur during an attack:

- Eyes may become red and watery.
- Nasal congestion.
- Running from the nostril.
- Forehead and facial sweating.
- Constriction of the pupil.
- Drooping or swelling of the eyelid.
- Physical agitation and tendency to move around.

The exact cause of cluster headache is unknown, but recent findings favor a dysfunction of the pain network.

### What is the difference between migraine and cluster headache?

One of the main differences between migraine and cluster headache is that the person with cluster headache, unlike the person with migraine, becomes agitated during an attack and is unable to sit or lie down or find relief in sleeping. Where a migraine can last up to 72 hours, a cluster headache lasts less than 3 hours with the intensity of the pain peaking within a few minutes and reducing quickly at the end of the attack.

## Diagnosis

There is no special test to diagnose cluster headache and so your doctor will need to take a very detailed history of all your symptoms in order to make the correct diagnosis. You may be referred for an MRI or CT scan to rule out other causes of the sudden onset of pain.

You are likely to be referred to a headache specialist in a hospital or headache clinic. This is because this is an infrequent condition, which GPs or family doctors do not often treat, not because it is dangerous or life threatening.

## Triggers

Alcohol is one well-known trigger of cluster headache, often bringing on the pain within an hour of drinking. If you have cluster headache you should not drink any alcohol during a cluster period. Once the bout is over you will be able to drink alcohol again.

A significant number of people find that strong smelling substances such as petrol, paint fumes, perfume, bleach or solvents can trigger an attack. Nitroglycerin and similar chemicals are well known triggers of attacks during a bout but not outside a bout. During a cluster period you should try to avoid these things.

Research has shown that nearly all cluster headache patients are heavy smokers. Although stopping smoking does not ease the cluster headache, heavy smokers are at an increased risk of developing chronic cluster headache, so giving up smoking or cutting down is worth considering.

Some patients find cold water or cold pads applied on the pain location or short-lasting intense physical activity helpful to alleviate the pain.

## Treatment

Whilst there is currently no cure for cluster headache, the available treatment has become much more effective in the last 20 years.

*Acute treatment* is used to stop the pain once it has started. Treating cluster headache can be tricky because the pain becomes extremely severe very quickly – usually within a few minutes. Thus the key to treating a cluster headache attack is speed, to reduce the excruciating pain as fast as possible.

Ordinary painkillers that you can buy over the counter are not usually effective, as the pain of cluster headache is too intense and they take too long to work. Opioids are not effective either.

*Oxygen* is one of the safest ways to treat cluster headache. You need to breathe the oxygen in at a rate of between 10 and 15 liters per minute using a non-rebreathing mask, i.e. one without holes, for 15 minutes at the start of the attacks. The treatment usually starts to work within 10 to 15 minutes.

### *Triptans*

*Sumatriptan injections* during an attack have been found to reduce the pain within 10 minutes. In general tablets are not effective if you have cluster headache because of the time they take to work.

*Sumatriptan and zolmitriptan nasal sprays* do help the majority of people although the onset of action may be slower than with an injection.

Research on the effectiveness of a number of medical devices is ongoing.

## Prevention

### *Lifestyle*

Keeping a headache diary to identify trigger factors can be helpful in terms of self-management; by helping the affected individual to regain control it can empower him or her during a cluster period. Due to the severity of cluster headache, a cluster period can be distressing and isolating; therefore, peer-to-peer support is vital for improving coping skills. Patient organizations should have a good medical advisory board and be equipped to provide accurate, reliable information and support to all those bearing this heavy burden.

### *Preventative treatment*

Preventative treatment is used throughout the entire course of the cluster period in an attempt to reduce the frequency and severity of cluster attacks. A wide range of preventative treatments is available and research on the effectiveness of possible new targeted prophylactic medications is ongoing.

The most common currently used preventative treatments are:

**Verapamil:** verapamil may be prescribed for cluster headache as research has shown that a daily dose can be effective in both episodic and chronic cluster headache.

As verapamil can disturb heart rhythms, patients may need ECG testing whilst the correct dose is being established. Any patients with an underlying condition will require careful ongoing monitoring should they require high doses of verapamil.

**Corticosteroids, as tablets or local injections:** these are given because they are fast acting. They can be used in a short burst, for 2 to 3 weeks, in decreasing amounts as a first step to break the cycle. They are often used alongside other treatments, which take longer to work. They can only be recommended as transitory treatments in short cycles, as long-term treatments are associated with significant side effects.

Other options that can be considered are lithium carbonate, topiramate, valproic acid, gabapentin and baclofen.

As with any drug treatment you may need to work with your doctor to determine what works best for you. You may need to try several treatment regimens before you discover the best one for you and a referral to a headache specialist can be recommended.

## **Prognosis**

Many people living with cluster headache may experience periods of remission, and chronic cluster headache can turn into the episodic form, however there is no significant predicting factor of long-lasting remission. The unpredictable nature of the disorder can have a significant impact and place a considerable burden on persons living with cluster headache, and their families.

## **Commandments for Health Professionals**

Cluster headache patients have issued the following 7 Commandments to Health Professionals for better treatment of those who live with cluster headache:

1. Ensure early and accurate diagnosis and treatment.
2. Know and educate colleagues about the clinical signs of cluster.
3. Provide accurate and reliable information and dispel the myths.
4. Listen to the person, acknowledge the condition, and reassure them as you provide the care.
5. Recognize the impact and burden on all those affected, both at home and at work.
6. Encourage the person to speak about their disorder in all aspects of their life and engage with their patient community.
7. Ensure easy access to headache specialists and provide ongoing support.

If you have met a doctor who observes these Commandments you are in good hands ... You can recommend him/her to others with the same problem.

*Paolo Rossi<sup>a</sup>, Jane Whelan<sup>b</sup>, Audrey Craven<sup>b</sup>, Elena Ruiz De La Torre<sup>b</sup>*

<sup>a</sup>Headache Clinic INI Grottaferrata, Grottaferrata (Rome), Italy

<sup>b</sup>European Headache Alliance