

Cluster Headaches

Cluster headaches occur as attacks of severe, one-sided headaches. Typically, a number of attacks occur over several weeks (a cluster of attacks). The headaches then usually go for weeks, months or years until a next cluster of headaches develops. An injection with a medicine called sumatriptan, or a nasal spray containing a medicine called zolmitriptan, or breathing 100% oxygen, will usually relieve each headache. Some medicines are also used to prevent the headaches. Also, it is recommended not to drink alcohol at all for the duration of a cluster period.

What are cluster headaches and what are the symptoms?

Cluster headaches consist of attacks of severe one-sided pain in the head. It is sometimes called migrainous neuralgia. Each attack develops suddenly, usually without any warning. Typically, you feel the pain mainly in or around one eye or temple. The pain may spread to other areas on the same side of the head. The pain is sometimes described as burning or boring. Each attack lasts 15-180 minutes, but most commonly 45-90 minutes. Attacks may occur from once every two days, to eight times a day.

The pain during an attack is usually severe. It can be so severe that you are likely to become agitated and unable to lie down. Some people even bang their head against the wall in frustration with the pain. Attacks often occur at night and wake you from sleep.

During each attack, one or more of the following symptoms also usually occurs: redness and watering of the eye; a runny and bunged up nose; sweating of the face; swelling of the eyelids; drooping of the eyelid; constriction of the pupil in the eye.

Attacks usually occur in clusters (bouts). That is, a number of attacks of pain occur over a period of time, which then stop. During a cluster, each attack of pain usually occurs on the same side of the head. Each cluster of attacks usually lasts for several weeks or months. Each cluster is then usually separated by months or years of remission (where no attacks occur). However, it can vary greatly from person to person as to how often the clusters occur. For example:

- A fairly typical case is for a cluster to last 6-12 weeks once a year, or once every two years, and at about the same time of year.
- Some people have more frequent clusters, some fewer, than the above.
- In a small number of cases only one cluster of attacks ever occurs.
- In about 1 in 10 cases, attacks continue without any remission periods. (This is called chronic cluster headache.)

Who gets cluster headaches?

Cluster headaches affect about 1 in 1,000 people. They are four times more likely to occur in men than in women. Anyone can be affected. The first cluster typically develops between the ages of 20-40 years, but it can start at any age.

What causes cluster headaches?

Research suggests that a part of the brain called the hypothalamus becomes overactive during each attack. However, it is not known what causes this overactivity. During an attack the hypothalamus is thought to release chemicals (neurotransmitters). These may stimulate nerve cells in the brain to cause the pain and other symptoms. The nerve overactivity may also cause a widening (vasodilation) of the arteries (blood vessels) in the affected side of the brain. One theory is that the pain is caused by the widening of these blood vessels which then press on nearby tissues in the brain, which can cause pain. However, this is just a theory and the exact cause of the pain is not known.

One feature of the hypothalamus is thought to be concerned with circadian rhythms (the body clock). Something to do with this body clock function of the hypothalamus is possibly why in many cases the clusters tend to occur at the same time each year. And, also, why during a cluster, each headache attack often occurs at about the same time each day. (This is often a couple of hours after going to sleep at

night.)

There may be some genetic part, as in about 1 in 20 cases the condition also occurs in some other close relative.

Most cluster headaches occur for no apparent reason. However, some people find that something may trigger a headache. If you find that something triggers a headache, then it is best to avoid it for the duration of a cluster period (until you are in remission). For example:

- Alcohol. Some people find that a headache often occurs within an hour or so of having an alcoholic drink. It is usually advised to stop drinking alcohol completely for the duration of a cluster period.
- Being hot may be a trigger such as exercising in a hot room, or having a hot bath.
- Strong-smelling substances, such as solvents, perfumes, petrol, etc.

Almost all people with cluster headaches have no abnormality of the brain that can be shown by scans or tests. In a very small minority of cases, cluster headaches seem to be triggered by a tumour in the pituitary gland (pituitary adenoma) or other brain tumours. (The pituitary is a small gland at the base of the brain.) It has to be stressed that this is a rare cause of cluster headaches.

How are cluster headaches diagnosed?

The diagnosis is based on the typical symptoms. There is no test that can prove the diagnosis. Tests are sometimes done if the diagnosis is not clear to rule out other causes of headache. Sometimes tests such as a brain scan may be done to rule out a pituitary adenoma (as described above.)

What is the treatment for cluster headaches?

Treatment is divided into treatments to relieve (abort) each headache, and treatments aimed at preventing the headaches.

Treating each headache

Ordinary painkillers do not work. Generally, if you take an ordinary painkiller, it takes too long to work, as the headache will usually have gone before the painkiller takes effect.

Sumatriptan injection

Sumatriptan, given by injection just under the skin, is the commonly used treatment to abort a headache. It relieves pain within 15 minutes in about 3 in 4 people with a cluster headache. You can be shown how to use this injection. Use it as soon as a headache occurs. Sumatriptan is classed as a triptan medicine that is used to treat migraine and cluster headaches. It is not a painkiller. Triptans work by interfering with a brain chemical called 5HT. This chemical is thought to be involved in migraine and cluster headaches.

Some points about sumatriptan injection:

- It works within 5-15 minutes to ease the headache in most affected people.
- The adult dose is a 6 mg injection for each headache. The maximum dose in 24 hours is two 6 mg injections (12 mg) with a minimum interval of one hour between the two doses.
- Side-effects sometimes occur but, if they do, are generally mild and do not last long. They include feeling sick, dizziness, tiredness and dry mouth. A minority of people also develop a warm-hot sensation, tightness, tingling, flushing, and feelings of heaviness or pressure in the face, arms, legs, and occasionally the chest.
- Some people should not take sumatriptan. For example, some people with heart disease, stroke disease or peripheral vascular disease. So, read the leaflet that comes with the medicine packet for a full list of possible side-effects and cautions.

Zolmitriptan nasal spray

Zolmitriptan is also a triptan medicine. Zolmitriptan nasal spray is an alternative to sumatriptan injection. It often works well but possibly not as quickly as sumatriptan injection.

100% oxygen therapy

This is an alternative treatment that may be advised, especially if sumatriptan injections or zolmitriptan nasal spray cannot be used, or do not work. It often works well to relieve pain within 15 minutes, but it does not work in everybody. Its advantage, when it works, is that it can be used as often as necessary. The

oxygen has to be 100% and so needs to be delivered, through a special mask, from an oxygen cylinder. Some people with cluster headaches have an oxygen cylinder and mask at home ready to treat an attack.

100% oxygen treatment may not be suitable for people who also have chronic obstructive pulmonary disease (COPD).

Other treatments

Sumatriptan injections, zolmitriptan nasal spray or oxygen are usually the first-line treatments. Other treatments that are sometimes used include: sumatriptan nasal spray and ergotamine injections. In general, these are often not as good as the first-line treatments.

It is sometimes difficult for doctors to rule out another type of headache called paroxysmal hemicrania. This is a rare condition of unknown cause which causes one-sided headaches similar to cluster headaches. Sometimes a doctor will prescribe a one-week trial of a medicine called indometacin. This will almost always take away the pain of paroxysmal hemicrania, but will have no effect on cluster headaches.

Preventing cluster headaches

Some medicines are used which aim to prevent cluster headaches. Most people with cluster headaches will be prescribed one. It is difficult to say exactly how well they work at reducing the frequency and/or severity of headaches. This is because there is a lack of large research trials which have studied these treatments. However, smaller research studies have shown encouraging results and it seems that they do work for many people with cluster headaches. Briefly:

- Verapamil is the most commonly used treatment. It is a medicine that is normally used to treat heart problems. It is not clear how it works for cluster headaches. The doses used are often higher than those used for heart problems, and you may need heart tests such as ECG before increasing to high doses.
- A steroid medicine such as prednisolone may be given at the beginning of a cluster. This may help quickly to prevent further headaches. However, you should not take it long-term due to side-effects. In some cases, a steroid medicine is started at the same time as another medicine such as verapamil. But, the steroid is stopped after a week or so when the effect of the other medicine has built up.
- Lithium is sometimes used. This medicine is commonly used to treat bipolar disorder. Again, it is not clear how it may help cluster headaches. You need regular blood checks to measure the level of lithium in the blood to make sure the dose is correct.
- Methysergide is used to prevent migraine and cluster headaches. You should not take it for more than six months as there is a risk of possible serious side-effects.
- Various other medicines are sometimes used. Also, research continues to find better treatments for this very painful condition.

Some notes about preventative medicines for cluster headaches

- Preventative treatment is often taken intermittently - that is, just for the duration of a cluster period. Treatment is typically continued for a cluster period until you are headache-free for 14 days. The exception to this is steroids which are normally only prescribed for a short time. Some people take preventative medication indefinitely. It depends on how often, and for how long, your cluster periods occur. This can vary greatly between affected people. In particular, people with chronic cluster headache may take preventative medication indefinitely.
- It is often trial and error to find out which preventative treatment works best for you. Many people start with verapamil and this is often successful. For most medicines, the dose is built up as quickly as possible to the maximum tolerated dose. If this is not found to help within one week, then it may be stopped and another tried. Treatment with a single medicine is preferred, but a combination of medicines may sometimes be necessary.
- Once an effective preventative treatment has been found for an individual, this can be restarted as early as possible after the start of the next cluster period.

However ...

- Preventative treatments that have previously worked well do not always continue to work well for future cluster periods. When this occurs, an alternative preventative treatment can be tried. For example, methysergide is thought to work in up to 7 in 10 cases. However, some people become tolerant to the effect of this medicine and it may then not work after two or three cluster periods.
- All the medicines used to prevent cluster headaches have potential side-effects and may not be suitable for people with certain other conditions. Therefore, always read the leaflet that comes with the medicine packet. However, sometimes a trade-off has to be taken. That is, accepting that some side-effects may be the price to pay for relief of the headaches.

Further help and advice

OUCH (UK) - Organisation For The Understanding Of Cluster Headaches
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