

**Name of Disorder:** Cluster Headache

**Essay Title:** Cluster Headache – A Medical Enigma

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Cluster headache is a relatively rare disorder which often has devastating impact on sufferers' quality of life. Given its high impact, it is fortunate that it is rare; a major capital city in Australia would only have a few hundred patients to a thousand or so. It is frequently referred to as the "suicide headache" as surveys of sufferers have indicated that a majority have contemplated suicide to escape the condition.

The symptoms are so characteristic that any health care professional, or indeed any person who is familiar with the condition, can generally readily recognise it within a few minutes of starting to take the history. However, its under-recognition by health care professionals is reflected in consistent findings in Europe and the United States that the average time to diagnosis is around seven years. Given that cluster headache requires specific treatment, the delays in diagnosis contribute to the poor quality of life of many of these patients. Patients with cluster headache are often overcome the first time they come across a doctor who tells them "I know what this is and we may be able to help".

The key symptoms are short, severe, one-sided headaches located around an eye, usually associated with watering of the eye. Although that sounds superficially somewhat like migraine, the key striking difference is the pattern over time. As the name indicates, episodes of cluster headache tend to come in runs of weeks to months, often at the same times of the year or season. During the "on" phase, patients typically have several attacks per day, usually around 3-5, of which one is very likely to be occurring in the small hours of the morning, around 2-3am. There are very few headache conditions in which a person goes to bed well and wakes with severe pain at around that time. Given the relationship to sleep, some patients in the active phase try to avoid sleep entirely to avoid precipitating attacks.

Other key distinguishing features from migraine are: the relatively short episodes (typically around 20 minutes to 1 hour); the lack of nausea, vomiting, light sensitivity or sound sensitivity and the strictly one-sided location. Additionally, patients with migraine will generally use their whole hand to indicate where in the head they feel their pain, whereas cluster headache patients will often point with one finger to the eye.

Because of the relative rarity of the disease, it has not been as well researched as more common conditions. Although the condition is different from migraine, some of the treatments that work in migraine also work in cluster headache.

The two most effective treatments for the acute attacks are inhaled 100% oxygen and the "triptan" family of medications. Unfortunately, the kind of oxygen that family doctors are used to using for people with lung disease will not work; it needs to be nearly 100% oxygen and specific equipment and training is needed to do this. Cluster headache support groups often help patients access such treatment. Oxygen treatment is very safe provided the patient has stopped smoking, otherwise there is a risk of burning the house down. In our clinic, to see whether oxygen helps we ask the patient to come to our day procedures area and then we deliberately trigger an attack using either alcohol (which the patient has to supply) or a tablet

of nitroglycerin, used for heart disorders. Although the triptan family of medications are effective, given the short and sudden onset of attacks, treatment with injections or intranasal formulations may be better than oral for speed of onset.

To suppress or prevent the attacks, there is a variety of treatments. High doses of steroids (prednisolone) often work but this can only be really used to gain control in the first few days of a run as the doses cause too many side effects when continued. It is my experience that individual patients respond to a consistent dose and it is an almost all-or-none effect and so if it is not working you may have to jump the dose up. More suitable long-term preventative drugs include the heart drug verapamil. Because this drug penetrates the brain poorly, we often have to go to doses higher than the maximum licenced dose for the usual heart and blood pressure conditions and hence the patient needs to give informed consent that this is outside usual treatment guidelines, but is endorsed by the specialists working in the treatment of cluster headache. Verapamil is usually very well tolerated but careful attention needs to be paid to the ECG at high doses as it can cause slowing of heart rhythm and heart conduction. This sounds more frightening than it really is. Unfortunately, if verapamil is not effective there is no reliable well-tolerated drug. The anti-epileptic drug topiramate seems to be effective as can be methysergide, another anti-migraine preventative drug which has potential for serious long-term side effects.

Outside of drug treatment, electrical stimulation of the nerves at the back of the head through a pacemaker-like device shows some promise and trials are underway to see whether this is really effective.

Although many patients have a good response to treatment, sadly there are still a significant number who do not. Some of the ideas for testing have been suggested by patients themselves through ideas swapped in online forums. The cause of cluster headache is really unknown. There is no well accepted theory of how these attacks occur. Brain scanning studies have shown that the part of the brain which regulates the body clock, the hypothalamus, is involved as would be expected by the time clustering of attacks but this is as far as we have got.

Cluster headache patients will do best under the care of a doctor knowledgeable and interested in this enigmatic condition but which is so rewarding when a patient can be helped.