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## **Headache Toolbox**

## Cluster Headache

Cluster headache is an unusual, severe type of one-sided headache characterized by eye tearing or redness (90% of people), running of one nostril (84%), sweating or flushing (59%), or swelling of the eyelid. These symptoms all occur on the same side of the head as the searing, stabbing pain, usually behind one eye or at the temple near the forehead. Cluster headache attacks are shorter than migraine, lasting between 15 minutes to 3 hours. Typically, the attacks occur at predictable times, most commonly in the very early morning between midnight and 3 am, and once again later in the day or early evening. Although the average number of attacks is 2 per day, there can be as few as one every other day, or rarely, as many as 8 attacks in a single day.

Cluster pain is described as being at least 10/10 in severity, and typically there is an inability to lie still. People with migraine tend to lie down in a dark, quiet room, while, in contrast, those with cluster will pace, rock in one place, or sometimes even bang their head to distract themselves from the pain. People almost never lie down during a cluster attack.

Cluster attacks come in runs of headaches or bouts called periods or cycles. During the cluster period, a person with cluster will have attacks daily. Typical periods of daily attacks are 6-8 weeks in duration, but for some the attack cycles are much longer. Cluster headache periods often occur 1-2 times per year at predictable times of the year, or they can skip years. Those cluster headache sufferers who have these runs of daily attacks and then have long periods of time with no attacks have the most common type of cluster, called episodic cluster headache.

About 25% of people having one cluster cycle will never have another. An unlucky 15% of cluster



sufferers will have an unending cluster period going on for longer than a year with less than a month of no attacks throughout the year. These individuals are said to have chronic cluster headache. Most individuals with cluster are male, estimated at a 2:1 ratio, unlike migraine, in which 2-3:1 are women.

An interesting survey of 1132 cluster patients was conducted in the United States by Rozen and Fishman. Information about cluster headache is difficult to obtain because there are only about 550,000 cluster sufferers in the United States compared to about 28 million individuals with migraine. People with cluster headaches have some unusual features. Many of them are smokers. About 50% smoke at the time they have cluster headaches, and only about 18% have never smoked. The most reliable trigger for cluster headache attack is alcohol, and in particular, beer. Therefore, most people with cluster will not drink any alcohol when they are having cluster attacks to avoid the pain. More than half of cluster sufferers described a family history of migraine, but only 18% had a close relative with cluster.

Sleep apnea is found in an estimated 30-80% of those with cluster headaches. The cluster generator is located in a part of the brain called the hypothalamus, which is linked to brain sleep-wake cycles.

It is interesting that during sleep apnea episodes, there is a drop of oxygen to the brain, and that oxygen is such an effective treatment for cluster attacks. Despite knowing where oxygen works in the brain, and that oxygen can be a highly effective treatment during a cluster attack, it is not known whether treatment of sleep apnea will directly benefit cluster headaches.

The average time before an individual with cluster headaches is diagnosed is often very long,

generally 5 years, which is a very long time to suffer such severe pain without specific treatment. Perhaps because of this, cluster is a disorder in which the afflicted can become desperate, and in the Rozen and Fishman survey, as many as 55% of respondents had contemplated suicide.

Fortunately, there are specific treatments which can bring relief. At the onset of a cluster period, a 10-14 day taper of steroid tablets or an injection of steroids plus anesthetic to the back of the head on the same side as the pain, called an occipital nerve block, can be tried. The steroids or injections can provide quick temporary relief until the most effective daily oral preventive medicine, verapamil, can take hold.

Verapamil is a calcium channel blocker blood pressure medication that is available in inexpensive generic forms. The dose of verapamil for cluster is often higher than is usually used for blood pressure control, with the dose for cluster starting at 80 mg 3 times per day, and increasing rapidly until there is benefit. At higher doses, an electrocardiogram should be checked as heart rhythm changes can occur with increased amounts of verapamil. To boost the effect of verapamil or, instead of verapamil if it is not tolerated, lithium, or valproate, may be used.

To terminate an attack, high-flow oxygen may be used with a face mask. This is highly effective for most individuals with cluster, and is very safe. Sumatriptan injectable, zolmitriptan nasal spray, or dihydroergotamine (DHE) injections, all originally formulated for migraine, can also help terminate an attack. Sumatriptan and DHE injections are FDA approved for acute treatment of cluster headache. Zolmitriptan nasal spray is approved in the European Union for acute treatment of cluster headache. The three options, sumatriptan, zolmitriptan, and DHE, all result in some temporary narrowing of the blood vessels to the heart and brain, and therefore cannot be used in individuals with known or suspected vascular disease. Using

the pill form of any medicine to stop an individual cluster attack is not recommended, because each cluster headache reaches its peak very, very fast, usually in less than 15 minutes, and each attack generally lasts about an hour, so oral tablets do not provide adequate relief. The pills take too long to kick in, and the cluster stops before the pill has a chance to work. For that reason, oxygen, injection, or nasal sprays are the standard for stopping a cluster headache attack.

Newer treatment options are being tested for cluster, including a mild electrical stimulator applied to the outside of the neck at the onset of an attack (a noninvasive vagal nerve stimulator), an implanted electrical stimulator surgically placed through the roof of the mouth (a sphenopalatine ganglion stimulator), and an injectable antibody targeting the pain chemical calcitonin gene-related peptide. None of these have yet been approved by the Food and Drug Administration at the time of this writing, but headache research centers are testing them throughout the United States and Europe.

Cluster headache is an unusual, severe headache disorder with specific characteristics and treatment. Clues to the diagnosis are short duration of each attack, lasting between 15-180 minutes, one-sided severe pain behind the eye or temple, eye tearing or redness, nose running from one nostril, or one-sided sweating or flushing. A full treatment plan includes a temporary bridge to provide some initial relief with steroids taken by mouth or injected to the back of the head, the starting of a daily preventive medication such as verapamil, and an immediate treatment available for each attack. There are new treatments on the immediate horizon that are likely to provide improved safe and rapid relief.

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