Cluster headache is a relatively rare type of headache that affects less than ½ percent of the population. Unlike migraine, it is more common in men than women, although the proportion of women diagnosed with cluster headache has risen over the past 20 years. The headaches can start at any age and often begin in the 20s or 30s. Most patients with cluster headaches are smokers or ex-smokers. Cluster headaches are unique for their location, associated features, and periodicity.

The cluster headache attack
Cluster headache pain comes on quickly and usually without a warning. The pain is located around the eye and temple but can extend to the jaw, cheek, teeth, nose, and side of the head. In most patients, the attacks are always on the same side of the head during a cluster period. The pain is excruciating, boring, stabbing, knife-like, or burning. It is one of the most painful conditions experienced by humans and is sometimes called “suicide headache.” The signature feature of the attack is the “autonomic symptoms,” which are involuntary and caused by activity of specific nerves in the brain and head. They include a droopy eyelid, a small pupil, eyelid swelling, tearing, bloodshot eye, stuffy or runny nose, flushing, or sweating on the same side as the pain.

Some patients with cluster headache also have symptoms that are more typical of migraine, such as aura, sensitivity to light or noise, nausea, vomiting, or other vague symptoms prior to an attack (mood changes, yawning, food cravings).

Alcohol, odors (solvents, perfume), and sleep are common triggers of cluster headache. Some patients have other triggers, such as smoking or eating certain foods (similar to migraine triggers).

Most patients experience agitation or become restless during a cluster headache and prefer to pace, rock back and forth, go outdoors, or strike their head to distract themselves from the pain. Others prefer to sit still; it is uncommon to want to lie down during an attack. The attack can last from 15 minutes to three hours.

The cluster period
Cluster headaches got their name because they come in “clusters.” They tend to occur daily for weeks to months at a time and then disappear for a month or more (termed “episodic cluster headache”). They are often seasonal. About 10%-20% of patients have chronic cluster headaches that are continuous from their onset or have remission periods of less than one month.

The attacks usually occur like clockwork and they often awaken people from sleep at the same time each night. Most people experience more than one attack each day, often in the late afternoon or in the evening.

How is cluster headache diagnosed?
The diagnosis is based on a typical history and normal neurological examination. Rarely, brain abnormalities (such as a tumor of the pituitary gland) can cause headaches that are very similar to cluster headaches. Brain imaging is recommended for all patients with cluster headaches.

How is cluster headache treated?
There are three aspects of treatment and they are generally all started at the same time:

Treatment of the acute attack
Since cluster headaches are relatively brief, the acute treatment must act rapidly. Therefore, oral medication is usually ineffective. Treatment options include:
• **Oxygen** – 100% oxygen through a non-re-breather face mask at 7-12 liters per minute relieves the headache in over half of patients. The oxygen should be used for 15-20 minutes. However, about 25% of patients have incomplete relief with a delay in their headache after using oxygen.

• **Triptans** – Injectable sumatriptan, sumatriptan nasal spray, and zolmitriptan nasal spray are often effective. Since cluster headaches occur multiple times a day, these treatments are expensive and often not covered by insurers in the quantity needed. Frequent use can lead to “rebound” headaches that are difficult to treat.

• **Ergots** – Dihydroergotamine (DHE) injections and intravenous DHE are effective treatments. DHE nasal spray may also be considered. Other ergots are useful.

• **Lidocaine** – 10% solution (a local anesthetic) administered into the nose on a cotton swab or by nasal spray is effective in many patients.

**Stopping the cluster period**
Steroids such as prednisone, prednisolone, and dexamethasone usually stop the headaches within days. They are started at a moderate to high dose (prednisone 40-80 mg daily) and the dose is reduced every few days. Sometimes the headaches return as the dose is tapered, requiring an increase in dosage with re-tapering. Common side effects of steroids are insomnia, increased appetite, and stomach pain. Long-term side effects include ulcer, osteoporosis, fracture, diabetes, weight gain, glaucoma, and easy bruising. Because of the side effects, steroids cannot be used indefinitely—preventive treatment is also needed. Greater occipital nerve injection of a long-acting local anesthetic, combined with a steroid, sometimes breaks the cycle.

**Long-term prevention**
Verapamil is perhaps the most effective long-term preventive treatment for cluster headaches. The dose needed for cluster headache is substantially higher than the dose used for treating blood pressure (up to 960 mg daily). Constipation is a common side effect, although the drug is usually very well tolerated in people with cluster headaches. Because Verapamil occasionally causes abnormal electrical conduction in the heart (prolonged Q-T interval), intermittent EKG monitoring is performed when using high doses.

Lithium (600-1,200 mg daily) has been successfully used for many years as a preventive treatment for cluster headache. It generally works within days. Short-term side effects include weakness, nausea, tremor, and slurred speech. Lithium blood levels, kidney function, and thyroid function must be monitored during treatment.

Topiramate is FDA-approved for migraine prevention and is also useful in cluster headache. It is started at a low dose and increased as tolerated; the effect is seen in one to four weeks. Common side effects are drowsiness, weight loss, memory problems, and tingling. Kidney stones, the sudden onset of glaucoma, and allergy are rare but serious side effects.

Gabapentin, administered in doses of 900 mg daily, may be effective as quickly as one week after starting treatment. Drowsiness and dizziness are the most common side effects.

Several studies of valproate showed mixed results but it seems to be effective. Common side effects are weight gain, tremor, hair loss and mood change. It may not be used during pregnancy.

Although methysergide can be very effective, this medication is no longer available.

Testosterone replacement in men with low testosterone levels may improve the headaches. Botulinum toxin injections have not been studied, but there are reports of their usefulness when oral medications fail.

**Surgical treatment of cluster headache**
When the headaches persist despite medical treatment, surgical options are considered.

Occipital nerve stimulation is often effective. A lead is implanted over the occipital nerve in the back of the head/neck, which is connected to a battery-powered stimulator. The intensity of attacks seems to decrease sooner than the frequency, and improvement occurs in days to weeks.

Hypothalamic stimulation is a new approach to cluster headache treatment. The hypothalamus is located deep within the brain and is the part of
the brain that regulates hunger, thirst, and circadian rhythms. Several studies using functional MRI and PET scans show that it is activated in patients with cluster headache. This procedure has significant risk and is not done at most medical centers.

Destructive surgery is a last resort and has serious risk.

What is the prognosis?
Cluster headache seems to be a lifelong condition in most patients, although remission periods tend to get longer with age. A small percentage of patients with episodic cluster headache evolve into having chronic cluster headache.

For further information, visit www.americanheadachesociety.org