TO: Friends, Employers, and Colleagues

FROM: Cluster Headache Support Group (CHSG.com)

REF: A note to those who know a person who is a CH sufferer

A person you know has given you this note to explain the neurological disorder called “Cluster Headaches” (CH). Very few people know of this disorder. Cluster Headaches only affect 1/10 of 1% of the population. Because it is so rare, sufferers often feel isolated. Isolation occurs because many even in their community misunderstand the disorder. Many misunderstand the impact the disorder has on the sufferer and their supporters.

This letter is an attempt to clear-up some of the confusion.

What is a “cluster headache”?

1. First; the word “headache” is the only similarity between “cluster headaches” and the other headaches you know about. They are nothing like a migraine.

2. “Episodic” sufferers will have a period that lasts several weeks while “Chronic” sufferers are “in-period” constantly, without remission.

3. An attack will reach devastating pain levels on one side of the head within a few minutes. Some other associated effects will also occur.

4. Attacks last an hour or longer and occur several times a day, on schedule every day (“in a cluster”) during a period.

How is your friend/colleague affected?

1. When an attack occurs, sufferers may be crying or wailing uncontrollably. They might not respond to question/comments. If they do, it will be “one-word” answers if at all.

2. Sufferers have been known to pace or rock in place, even “act way out of character”. The pain is one of the worst known to man. Worse than amputation, childbirth, even gun-shot.

3. Other effects the sufferer may experience are tearing from the eye, visual and aural disturbances, flushing, nausea, swollen or droopy eyelid and possibly dizziness.

How can you help your friend/colleague?

1. During the attack - give the sufferer lots of space, calling 911 unfortunately will not help. The reason is by time a sufferer is seen by a nurse, doctor, prescribed medication – the attack will resolve itself. The patients themselves are expert in how best to treat this illness.

2. After the attack – offer them sips of water, if they want to talk it is okay. Please don’t equate any other headache you or your friends had with CH. It is nothing like CH, not even close.

The Cluster Headache Support Group – www.CHSG.org
Cluster headache is a stereotyped primary pain syndrome characterised by strictly unilateral severe pain, localised in or around the eye and accompanied by ipsilateral autonomic features. The syndrome is characterised by the circadian rhythmicity of the short-lived attacks, and the regular recurrence of headache bouts, which are interspersed by periods of complete remission in most individuals. Headaches often start about 1–2 h after falling asleep or in the early morning, and show seasonal variation, suggesting that the hypothalamus has a role in the illness. Consequently, the vascular theory has been superseded by recognition that neurovascular factors are more important. The increased familial risk suggests that cluster headache has a genetic component in some families. Neuroimaging has broadened our pathophysiological view and has led to successful treatment by deep brain stimulation of the hypothalamus. Although most patients can be treated effectively, some do not respond to therapy. Fortunately, time to diagnosis of cluster headache has improved. This is probably the result of a better understanding of the pathophysiology in combination with efficient treatment strategies, leading to a broader acceptance of the syndrome by doctors.

Clinical features

“Imagine, your eye is pushed out of its socket and your right eyelid is beginning to swell shut. You start squinting and your eye is tearing, you are convinced there was blood pouring out. A red-hot knife is crushed into your head, excruciating, horrible pain. Your only saving grace is to pace from room to room, crying, flinging yourself to the floor, until eventually the pain drains from you. Waiting for the next attack to happen is a terrible, scary feeling. I sometimes think that I will go mad. I’m exhausted but then the next one hits.”

This is an example of how a cluster headache patient might describe his pain in an outpatient setting. Cluster headache, one of the most severe pain syndromes—female patients describe each attack as being worse than childbirth—is still underdiagnosed and suboptimally managed in primary care. Results of a recent health-related quality-of-life study in 56 patients suggest that cluster headache has substantial effects on patients’ ability to function, even when appropriate treatments are used. Typically, attacks can strike up to eight times a day, are relatively short-lived, and are characterised by strictly unilateral severe head pain accompanied by autonomic symptoms. A side shift is mentioned in only about 15% of cases. Unlike individuals with migraine, patients with cluster headache are restless and prefer to pace about or sit and rock back and forth. Some patients will exert pressure on the painful area with a hand over the affected eye and temple. Many will isolate themselves during the headache or leave the house to get into cold or fresh air, and tend to become aggressive during an attack.

The unilateral autonomic symptoms such as ptosis, miosis, lacrimation, conjunctival injection, rhinorrhoea, and nasal congestion occur only during the pain attack and are ipsilateral to the pain, indicating parasympathetic hyperactivity and sympathetic impairment (figure I). In some patients, the signs of sympathetic paralysis (miosis and ptosis) persist indefinitely, but intensify during attacks. Sweating and bloodflow to the skin also increase on the painful side, particularly in areas of sympathetic deficit. About 3% of patients have no autonomic symptoms, and in rare cases sympathetic disturbances persist on the previously affected side of the face in patients whom cluster headaches have switched sides.

Another clinical feature of the syndrome is the circadian rhythmicity of the painful attacks, which are relatively short (15–180 min). In the episodic form, headaches occur daily for some weeks followed by a period of remission. On average, a cluster period lasts 6–12 weeks, and remissions can last up to 12 months. In the chronic form, attacks occur without substantial periods of remission. When chronic cluster headache is unresponsive to medical treatments, it becomes a serious problem and surgical options may have to be considered.

Epidemiology and genetics

Compared with migraine, cluster headache is uncommon. The disorder has a prevalence of less than 1%, and mostly affects men. The episodic form is most common, affecting 80–90% of cluster headache patients. It is characterised by periods of headaches (clusters or bouts) and periods of remission. During a
Note the Horner syndrome ipsilateral to the headache and increased facial sweating exclusively around the left eye.

Figure 1: Patient soon after a left-sided cluster headache attack
Note the Horner syndrome ipsilateral to the headache and increased facial sweating exclusively around the left eye.

bouts, patients may experience one to eight attacks per day, and bouts can last from 7 days to 12 months. While in remission, patients are usually asymptomatic. The chronic form of cluster headache lacks remissions, and is diagnosed after a year without remission or if remission has lasted less than 30 days. A chronic cluster may arise de novo (primary chronic cluster headache) or evolve from the episodic type (secondary chronic cluster headache).

One of the most urgent questions patients put to their doctors is whether, as in migraine, the cluster attacks decline with age. Longitudinal data for cluster headache are anecdotal and only recently have data from larger epidemiological studies become available. Overall, the authors of these reports assume that within the natural course of the condition, the symptoms remit with age.

The largest epidemiological study to date reports data for about 550 patients with episodic and chronic cluster headache over an observation period of more than 30 years (1963–97). In this study, there was a trend towards a decreasing male preponderance; the male-to-female ratio was substantially higher among patients with onset before 1970 than in those with onset after 1970, whereas the proportion of episodic to chronic form did not change during the study period. According to prospective studies, the syndrome occurs about three times more often in men than in women, and is clinically identical in both sexes. Mechanisms associated with sex hormone regulation, and environmental factors related to lifestyle, have been suggested to account for the increase in numbers of women diagnosed with cluster headache since 1970. The nature of the sex-related and age-related pattern of cluster headache onset is unclear. However, the increase in the diagnosis in women might also be the result of increased awareness and acceptance of the disorder by doctors, due to improved understanding of cluster headache pathophysiology.

The medical history often reveals a high incidence of head trauma with brain concussion, but it is hard to prove a cause-and-effect relation. Interestingly, up to 85% of patients with chronic headache are also chronic cigarette smokers. Quitting smoking has no effect on the disease. The question arises whether chronic nicotine consumption is needed as a trigger to initiate the syndrome, possibly on the basis of some genetic background.

Before 1990, cluster headache was not generally thought to be an inherited disorder. However, reports of cluster headache in monozygotic twins and familial occurrence of cluster headache in 7% of families, resulting in a 14-fold increase in risk of cluster headache in first-degree relatives and a two-fold increased risk for second-degree relatives, show that genetic factors should be considered. In a study of 186 index patients and 624 first-degree relatives, investigators showed a positive family history of cluster headache in 11% of the index patients. They concluded that no precise mode of inheritance could be ascertained. A complex segregation analysis of cluster headache has suggested that an autosomal dominant gene has a role in some families, although some evidence exists for autosomal recessive or multifactorial inheritance in others. However, future studies should take into account that since cluster headache can start between the ages of 7 years and 83 years, the distinction between affected and unaffected individuals is clearly provisional. To date, the increased familial risk strongly supports the hypothesis that cluster headache has a genetic component, at least in some families. However, no clear molecular genetic clues have yet been identified. In view of the paroxysmal character and circadian and circannual rhythmicity of the disease, future studies need to focus on ion channel genes and clock genes.

Pathophysiology
Although the syndrome is well defined from a clinical point of view and has been recognised for more than two centuries, its pathophysiology is still poorly understood. However, the past decade has seen remarkable progress toward solving the pathophysiological puzzle. Any pathophysiological model needs to explain the three major features of cluster headache: trigeminal distribution of the pain, ipsilateral cranial autonomic features, and (circadian) episodic pattern of attacks. The vascular theory, which is based on an inflammation of the walls of the cavernous sinus (the only peripheral anatomical location where a single pathology could involve trigeminal C-fibres and sympathetic fibres), has been superseded by recognition that neurovascular events and some central impulse generator or oscillator seem to be more important. The severe unilateral pain is likely to be mediated by activation of the first (ophthalmic) division of the trigeminal nerve, whereas the autonomic symptoms such as lacrimation are due to activation of the cranial parasympathetic outflow from the seventh cranial nerve.