

# Chronic Cluster Headache

Attacks occur for more than 1 year without remission or with remission periods shorter than 3 months.

## Pathophysiology

The pain and autonomic symptoms of cluster headache (CH) result from activation of the trigeminal parasympathetic reflex, mediated through the [sphenopalatine ganglion](#) (SPG).

Low frequency (LF) SPG stimulation may induce cluster-like attacks with autonomic features, which can subsequently be treated by HF SPG [sphenopalatine ganglion stimulation](#). Efferent parasympathetic outflow from the SPG may initiate autonomic symptoms and activate trigeminovascular sensory afferents, which may initiate the onset of pain associated with CH <sup>1)</sup>.

## Clinical features

Recurrent unilateral attacks of severe pain. Usually oculofrontal or oculotemporal with occasional radiation into the [jaw](#), usually recurring on the same side of the head. Ipsilateral autonomic symptoms (conjunctival injection, nasal congestion, rhinorrhea, lacrimation, facial flushing) are common. Partial Horner syndrome (ptosis and miosis) sometimes occurs.

Headaches characteristically have no prodrome, last 30–90 minutes, and recur one or more times daily, usually for 4–12 weeks, often at a similar time of day, following which there is typically a remission for an average of 12 months <sup>2)</sup>.

Individuals typically experience repeated attacks of excruciatingly severe unilateral headache pain.

Cluster headache attacks often occur periodically; spontaneous remissions may interrupt active periods of pain, though about 10–15% of chronic CH never remit. The cause of cluster headache has not been identified.

## Treatment

see [Cluster headache treatment](#).

## Case reports

A patient with disabling cluster headache highly resistant to medical treatment underwent implantation of a peripheral nerve neurostimulation system to stimulate the supraorbital nerves (SON) and greater occipital nerve (GON) in the Pain Unit <sup>3)</sup>

A 41-year-old man presented with a 3-month history of side-locked attacks of excruciating severe stabbing and boring right-sided pain located in the temple and the orbit. The attacks were associated with conjunctival injection and restlessness and migrainous features. The duration of attacks was about 30 minutes and the frequency 4 to 5 per 24 hours. His vital signs and physical and neurological examination were normal. A previous unenhanced brain computed tomography had been normal. A diagnosis of CH was made. The patient responded partially to treatment. Enhanced magnetic resonance imaging after 3 weeks displayed a right-sided parietal glioma with surrounding edema and mass effect. After debulking, the headache attacks resolved completely.

Contrast-enhanced magnetic resonance imaging should always be considered in patients with CH despite earlier normal head computed tomography/examinations. Late-onset CH represents a condition that requires careful evaluation. Parietal glioblastoma multiforme can present as CH. <sup>4)</sup>

<sup>1)</sup>

Schytz HW, Barløse M, Guo S, Selb J, Caparso A, Jensen R, Ashina M. Experimental activation of the sphenopalatine ganglion provokes cluster-like attacks in humans. *Cephalalgia*. 2013 Jul;33(10):831-41. doi: 10.1177/0333102413476370. Epub 2013 Feb 4. PubMed PMID: 23382519.

<sup>2)</sup>

Kittrelle JP, Grouse DS, Seybold ME. Cluster Headache: Local Anesthetic Abortive Agents. *Arch Neurol*. 1985; 42:496-498

<sup>3)</sup>

Dominguez Garcia MM, Abejon Gonzalez D, de Diego Gamarra JM, Cánovas Martinez ML, Balboa Díaz M, Hadjigeorgiou I. Symptoms and pathophysiology of cluster headache. Approach to combined occipital and supraorbital neurostimulation. *Rev Esp Anesthesiol Reanim (Engl Ed)*. 2023 Feb;70(2):83-96. doi: 10.1016/j.redare.2021.10.005. Epub 2023 Feb 21. PMID: 36822404.

<sup>4)</sup>

Edvardsson B, Persson S. Cluster headache and parietal glioblastoma multiforme. *Neurologist*. 2012 Jul;18(4):206-7. doi: 10.1097/NRL.0b013e31825cf181. PubMed PMID: 22735247.

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