



# Cluster headache

## Focus on fast-acting intervention

BY MAREK GAWEL, MD

Cluster headache is a distinct clinical entity that is classified with other headache syndromes as Trigeminal Autonomic Cephalgias (TACs), which involve both pain (trigeminal) and autonomic features. The prevalence of cluster headache is 0.09%, while the incidence has been calculated at 15.5 and 4.0 per 100,000 men and women, respectively. Cluster headaches affect more men than women with a ratio of 5:1. In most patients, the attacks occur several times a day, often disrupting sleep. The episodes may last weeks or months at a time, and 10% of individuals who suffer from cluster headache experience no remission.

Although the exact cause of cluster headaches isn't known, their occurrence at regular intervals suggests some involvement of the suprachiasmatic nucleus, which controls circadian rhythms. Recently, a positron emission tomography study of nine patients experiencing an attack has shown activation of the hypothalamic grey matter occurring on the same side as the headache, but only during the pain episode.

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### Risk factors

- age (onset typically occurs in the third or fourth decades)
- stress
- alcohol use
- smoking
- history of migraine
- sleep apnea
- family history (evidence is weak, but genetics may play a small role)

### Triggers

- alcohol (only in a cluster period)
- bright light
- exertion
- heat
- high altitudes
- certain foods (high in nitrates)
- certain meds (nitroglycerin, histamine and some hypertension meds)

### Symptoms

- rapid onset; usually between 5 and 15 minutes
- attacks often occur episodically; untreated bouts of headache can last from 15 minutes to three hours
- the pain is usually unilateral, but quite severe and is accompanied by autonomic features, of which at least one of the following must be present:
  - conjunctival injection
  - lacrimation
  - nasal congestion
  - rhinorrhea
  - forehead/ facial sweating
  - miosis
  - ptosis
  - eyelid edema

### Differential diagnosis

**The most important step is to exclude any secondary causes.**

- head trauma (most common)
- also consider a variety of lesions that involve:
  - the occipital lobe
  - the vertebral and the carotid artery
- it's often not possible to predict these by a physical assessment alone; consider ordering an MRI with gadolinium in all new cases and in patients who don't respond to treatment
- on examination, there may be signs of Horner syndrome and conjunctival injection, but the rest of the neurologic exam is normal
- **Other TACs to consider:**
  - cluster tic — a combination of cluster and trigeminal neuralgia
  - paroxysmal hemicrania — a continuous hemicrania with autonomic features
  - SUNCT syndrome — short-lasting unilateral neuralgiform pains with conjunctival injection and tearing

### Prophylactic therapy

- educate patients regarding the diagnosis and nature of the problem
- advise susceptible individuals to avoid alcohol and other potential triggers (e.g. nitrates)
- begin with prednisone 60 mg/day for two weeks then taper by 5 mg/day
- try another cycle if cluster returns
- in resistant patients, try solumedrol 1g/day IV for three days followed by prednisone
- **Other options include:**
  - verapamil 180-240 mg/day
  - lithium carbonate 300 mg/2-3 times a day (check lithium levels and thyroid function)
  - methysergide 2 mg/three times a day (stop for a month every 4-6 months and check for retroperitoneal and cardiac fibrosis)
  - valproate 250-500 mg/three times a day
  - gabapentin 400 mg (dose can go as high as 3,600 mg/day)
  - topiramate 100-300 mg/day
  - botulinum toxin 100 units injected in aliquots over the area of the pain

### The refractory patient

- polytherapy
- DHE IV 1 mg/three times a day for 3-4 days
- histamine desensitization
- occipital block with lidocaine/methylprednisolone

### Acute treatment

**Initiate abortive treatment for acute attacks; episodes are generally short so there's a need for fast-acting intervention.**

**Pharmacologic therapy**

- 100% oxygen 8 L/min for 15 minutes
- sumatriptan 6 mg SC
- zolmitriptan nasal spray 5 mg
- dihydroergotamine (DHE) 1 mg IM or IV
- DHE 2 mg nasal spray
- nasal lidocaine 4%

### Surgery

**Criteria for surgery includes:**

- strictly unilateral headache
- resistance or complications with full trial of medical therapy
- stable psychologic profile; ensure patient has no addictive traits

**Procedures**

- sphenopalatine ganglionectomy
- vidian nerve section
- radiofrequency thermocoagulation of the trigeminal ganglion
- glycerol injection of trigeminal ganglion
- electrical stimulation of the occipital nerve by implanted electrode