

Cluster headache and other autonomic cephalopathies

Background

Cluster headache is arguably one of the most painful conditions a GP will ever see. It is invariably misdiagnosed and usually inadequately treated. Often sufferers will have been to eye Departments with suspected glaucoma, ENT Departments with suspected sinusitis and Dental departments to have their wisdom teeth extracted. Cluster headache is very distinct from migraine and is one of a group of headaches known as the trigeminal autonomic cephalalgias characterised by strictly unilateral pain and autonomic features. The pathophysiology is unknown.

Epidemiology

- 0.1- 0.2%
- Male: female ratio is 5:1
- It can begin at any age though the most common age of onset is the third or fourth decade.
- 10% of patients have chronic cluster headache (CCH) where the attacks are usually constant and remissions last less than one month
- Cluster headache is a life long disease but attacks sometimes get less frequent with age

Making the diagnosis

The cluster attack

- The attacks are strictly unilateral although the headache may alternate sides with attacks.
- The pain is excruciatingly severe and is associated with restlessness or agitation.
- Located mainly around the orbital and temporal regions.
- Headache lasts from 15 minutes to 3 hours. It has an abrupt onset and cessation.
- The cluster attack frequency varies from one on alternate days to up to eight daily. The condition can have a striking circadian rhythmicity, with some patients reporting that the attacks occur at the same time each day.
- There are associated cranial autonomic symptoms on the side of the pain and lasting with it. For example, conjunctival injection, lacrimation, miosis, ptosis, eyelid oedema, rhinorrhoea, nasal blockage and forehead or facial sweating.
- Nausea, photophobia and phonophobia usually absent.
- Alcohol, exercise, and elevated environmental temperature can precipitate an attack but not outside a cluster period. Allergies, food sensitivities, reproductive hormonal changes and stress do not appear to have any significant role in precipitating attacks.

The Cluster Period or Bout

A cluster period is an episode during which there are frequent cluster attacks following which the individual is in remission.

- The average cluster period lasts between 6 and 12 weeks but there is considerable variation between patients.
- Most patients have one or two annual cluster periods, each lasting between one and three months. Often, a striking circannual periodicity is seen with periods occurring in same month of the year, often spring or autumn.

Important differences between Migraine and Cluster Headache	
Migraine	Cluster Headache
Prodrome or aura can occur	Prodrome or aura very rare
Pain can occur in any location	Pain is mainly periorbital
Pain is severe and throbbing. <u>Patients want to lie down.</u>	Pain is very severe and piercing. <u>Patients pace the room.</u>
Attack lasts 4-72 hours	Attack lasts 15-180 minutes and come in clusters
No autonomic features	Autonomic features around the eye on side of pain
Nausea, vomiting, photophobia or phonophobia	Rare

Differential diagnosis of cluster headache – other rarer autonomic cephalopathies					
Diagnosis	Type of pain	Severity	Location	Duration	Frequency
Cluster headache	Boring	High	Orbital	15-180 minutes	1-8 a day
Paroxysmal hemicrania	Boring	High	Orbital	1-30 minutes	3-30 a day
*SUNCT	Stabbing	Moderate	Orbital	15-240 seconds	1 a day to 30 an hour

*Short acting unilateral neuralgia form headaches with conjunctival injection and tearing

Investigation

Approximately 1% of cluster headache presentations will have an underlying pathology. Pituitary tumours are most common. All new cluster headache should be imaged. If a patients presents with a history of many years of stable cluster this can be relaxed. Oxygen should inhaled for 10 to 20 minutes depending on the clinical response.

Treatment

Acute treatment

- Subcutaneous sumatriptan 6mgs is the drug of choice. Unlike in migraine, it can be prescribed at a frequency of twice daily without reduction in effectiveness, side effects or rebound.
- Oral triptans are ineffective but there is evidence to support the nasal route.
- Oxygen. The mechanism of its action is unknown. (See information for prescribing oxygen).

Patients should be made aware of the dangers of continuing to smoke in the presence of oxygen therapy. The majority of cluster patients are smokers at presentation and smoking cessation

intervention should be given. Smoking is a relative contraindication but if you prescribe to a smoker it is good practice to obtain written consent indicating an understanding of the risks.

For other co-existing pulmonary conditions where 100% oxygen may be harmful, advice should be taken from a respiratory physician.

For portable use, an ambulatory cylinder can be ordered providing up to 40 minutes of treatment but this is very expensive and difficult to obtain.

Preventative Treatment

Short term prevention

- Steroids give rapid relief and are useful where there are only 2-3 attacks each year. Prednisolone 1mg/kg, to a maximum of 60mg once daily for 4 days and thereafter decrease over a 3-4 week period. Relapse can occur as the dose is tapered and in this case steroids are used as an initial therapy in conjunction with preventatives until the latter are effective.

Long term prevention

- Verapamil is the preventative drug of choice in both episodic and chronic cluster headache but higher doses than those used in cardiological indications are needed. After performing a baseline ECG, start on 80mgs three times daily and thereafter the total daily dose is increased in increments of 80mgs every 10-14 days until the cluster attacks are suppressed with an ECG performed prior to each increment up to a maximum of 960mgs daily. (See patient information sheet)
- Lithium, Topiramate, Melatonin, Sodium Valproate, Gabapentin are used but their impact is often marginal. Lithium is the most effective aiming for a serum level in the upper part of the therapeutic range.
- Occipital nerve injection can be helpful.
- Surgery is a last-resort measure, either destructive procedures or neuromodulatory procedures with implanted electrodes.