International Headache Society – Diagnostic Criteria for Cluster Headache
(Source: http://ihs-classification.org/en/02_klassifikation/02_teil1/03.01.00_cluster.html, 2014)

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Previously used terms

Ciliary neuralgia, erythro-melalgia of the head, erythroprosopalgia of Bing, hemicrania angio-paralytica, hemicrania neuralgiformis chronica, histaminic cephalalgia, Horton’s headache, Harris-Horton’s disease, migrainous neuralgia (of Harris), petrosal neuralgia (of Gardner), Sluder’s neuralgia, sphenopalatinal neuralgia, vidian neuralgia

Coded elsewhere

Symptomatic cluster headache is coded to the underlying causative disorder.

Description:

Attacks of severe, strictly unilateral pain which is orbital, supraorbital, temporal or in any combination of these sites, lasting 15-180 minutes and occurring from once every other day to 8 times a day. The attacks are associated with one or more of the following, all of which are ipsilateral: conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis, eyelid oedema. Most patients are restless or agitated during an attack.

Diagnostic criteria:

A. At least 5 attacks fulfilling criteria B-D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes if untreated¹
C. Headache is accompanied by at least one of the following:
   1. ipsilateral conjunctival injection and/or lacrimation
   2. ipsilateral nasal congestion and/or rhinorrhoea
   3. ipsilateral eyelid oedema
   4. ipsilateral forehead and facial sweating
   5. ipsilateral miosis and/or ptosis
   6. a sense of restlessness or agitation
D. Attacks have a frequency from one every other day to 8 per day²
E. Not attributed to another disorder³

Note:

1. During part (but less than half) of the time-course of cluster headache, attacks may be less severe and/or of shorter or longer duration.
2. During part (but less than half) of the time-course of cluster headache, attacks may be less frequent.
3. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

Comment:

Acute attacks involve activation of the posterior hypothalamic grey matter. Cluster headache may be inherited (autosomal dominant) in about 5% of cases. Attacks usually occur in series (cluster periods) lasting for weeks or months separated by remission.
periods usually lasting months or years. However, about 10-15% of patients have chronic symptoms without remissions. In a large series with good follow-up, 27% of patients had only a single cluster period. These should be coded as 3.1 Cluster headache.

During a cluster period, and in the chronic subtype, attacks occur regularly and may be provoked by alcohol, histamine or nitroglycerine. Pain is maximal orbitally, supraorbitally, temporally or in any combination of these sites, but may spread to other regions of the head. Pain almost invariably recurs on the same side during an individual cluster period. During the worst attacks, the intensity of pain is excruciating. Patients are usually unable to lie down and characteristically pace the floor.

Age at onset is usually 20-40 years. For unknown reasons prevalence is 3-4 times higher in men than in women.

Cluster headache with coexistent trigeminal neuralgia (cluster-tic syndrome):

Some patients have been described who have both 3.1 Cluster headache and 13.1 Trigeminal neuralgia. They should receive both diagnoses. The importance of this observation is that both conditions must be treated for the patient to be headache free.