Cluster Headache and Chronic Paroxysmal Hemicrania

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[Cluster Headache 1](#_Toc5997491)

[Pathophysiology 1](#_Toc5997492)

[Epidemiology 1](#_Toc5997493)

[Clinical Features 2](#_Toc5997494)

[Differential Diagnosis 3](#_Toc5997495)

[Evaluation 4](#_Toc5997496)

[Abortive therapy 4](#_Toc5997497)

[Preventive therapy 4](#_Toc5997498)

[Prognosis 5](#_Toc5997499)

[Chronic Paroxysmal Hemicrania 5](#_Toc5997500)

[Hemicrania continua 6](#_Toc5997501)

Cluster Headache

Old synonyms - Raeder syndrome, Horton cephalalgia, histamine cephalalgia, sphenopalatine neuralgia.

Pathophysiology

- not fully determined.

Theories

1. **circadian pacemaker** **alterations** (due to ***hypothalamic dysfunction***).
   * attacks increase following beginning and end of *daylight savings time*.
   * there is *loss of circadian rhythm* (for blood pressure, temperature, hormones - prolactin, melatonin, cortisol, beta endorphins).
   * recently, functional neuroimaging have identified **posterior hypothalamic grey matter** as key area for basic defect.
   * pain is generated at pericarotid / cavernous sinus complex.
2. **neurogenic inflammation**
3. carotid body **chemoreceptor dysfunction**
4. central **parasympathetic & sympathetic imbalance**
5. increased **responsiveness to histamine**.

Epidemiology

Prevalence 0.01-1.5% (≈ 0.3%)

* higher in **men** (male : female ≈ 6-8:1) and in **blacks**.
* family history is rare.

Onset - any age (generally - in ***late twenties***).

* ≈ 10% patients develop cluster in their sixties.

**Peptic ulcer disease** is only known associated medical disorder.

* strong associations with ***smoking***, ***alcohol*** use, and previous ***head / face trauma***.
* certain *personality and physical characteristics* have been associated with cluster headache (e.g. tall and rugged-looking body, leonine facial appearance, multifurrowed and thickened skin with prominent folds, broad chin, vertical forehead creases, nasal telangiectasias).

Clinical Features

* 1. **Episodic** **cluster headache** - remission periods lasting **≥ 14 days** (usually 6 months ÷ 2 years).
  2. **Chronic** **cluster headache** (≈ 10%) - no remissions or remissions last **< 14 days**; headache is occurring for > 1 year.

Either type may transform into other! (in 4-13% patients, **episodic CH** transforms into **chronic CH**)

Multiple episodes of headache:

1. pain begins ***without warning***.
2. **severe** – pain rapidly increases (within 5-15 minutes) to ***excruciating levels***.
3. **short-lived** – if left untreated, attacks usually last 30-90 minutes (15-180 minutes).
4. strictly **unilateral**\*, **periorbital** (orbital / supraorbital / temporal) – distribution of 1st or 2nd divisions of trigeminal nerve. \*usually affects same side in subsequent months
5. ***may radiate*** to forehead, temples, jaws, nostrils, ears, neck, or shoulder.
6. pain is **deep**, **constant** (not throbbing), boring, piercing, burning, explosive.
7. at least one symptom of **unilateral (ipsilateral) autonomic dysfunction**:
   1. conjunctival injection (“red eye”)
   2. lacrimation
   3. miosis
   4. ptosis
   5. eyelid edema
   6. nasal congestion
   7. rhinorrhea
   8. facial sweating

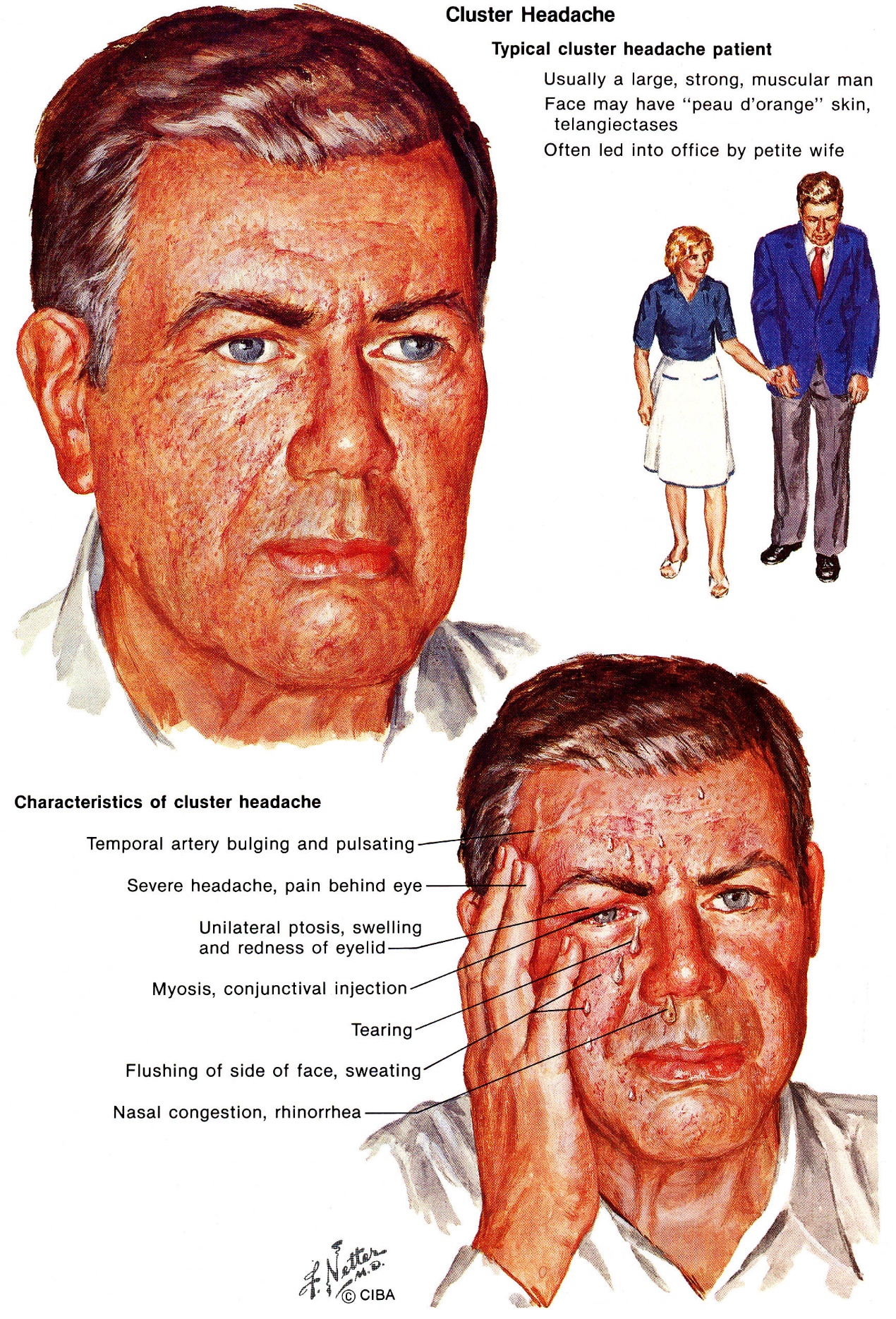
* in variant of cluster headache, full Horner's syndrome can be seen.

1. patients feel ***agitated*** or restless with need to isolate themselves and move around (most sufferers assume upright position to relieve discomfort!); patients have been known to become violent or bang their heads against wall.
2. GI symptoms uncommon.

Attack frequency varies (8 attacks per day ÷ 1 attack every 2 days).

**Periodicity is hallmark of cluster headache**! - attributed to hypothalamic (particularly suprachiasmatic nuclei) hormonal influences

* + - attacks often ***occur at same time*** ***each day*** ("alarm clock headache"); onset is nocturnal in 50% cases (may awaken patients from sleep).
    - attacks occur in **cluster periods** that last 1 week ÷ 1 year (usually 1-3 months) separated by periods of headache-free remission.
    - cluster episodes often appear at ***characteristic times of year*** (particularly around vernal and autumnal equinoxes).
    - attacks may cease *during pregnancy* (but attacks seldom correlate with menses).



Differential Diagnosis

* + - 1. Secondary cluster-like headache (due to structural lesions near cavernous sinuses).
      2. Chronic paroxysmal hemicrania
      3. Migraine
      4. Trigeminal neuralgia
      5. Temporal arteritis
      6. Raeder paratrigeminal syndrome - pain is constant (no distinct attacks)
      7. Tolosa-Hunt syndrome
      8. Sinusitis
      9. Glaucoma

Evaluation

* + - * + strictly clinical diagnosis - careful **history** is all that is needed.
        + **MRI** is justified only in ***atypical cases*** or ***abnormal neurological examination*** (except when abnormality is Horner's syndrome!).

Abortive therapy

Oral preparations are not recommended - absorbed too slowly.

1. Inhaled high-flow oxygen (**12 L/min 100% by mask for full 15 minutes** following headache onset) - treatment of first choice (70-80% effective); postulated mechanism – O2 is vasoconstrictor and increases serotonin synthesis.
2. Parenteral sumatriptan 6 mg s/c
3. Parenteral DHE
4. Topical (intranasal) **local anesthetics** (2-4% lidocaine) - to most caudal aspect of inferior nasal turbinate (patient in supine position) - can deliver *sphenopalatine ganglion block* - remarkably effective!
5. Parenteral **narcotics**
6. **Sphenopalatine ganglion stimulation** with implantable system

* at 15 minutes following stimulation, 55% of treatment group had pain relief compared with 6% in sham treatment group (P < .0001); pain relief was maintained to 90 minutes (50% pain relief for treatment group vs 13% for sham). <http://www.medscape.com/viewarticle/807281>

Preventive therapy

- required for almost all patients!:

* attacks are too short and too severe to treat with only abortive medication;
* prophylactic treatment is most effective among all primary headache disorders! (except **chronic CH** - notoriously resistant to standard prophylactic agents)
* avoid ***alcohol*** and ***nitroglycerin***.

Alcohol provokes attacks in 70% patients but has no effect when cluster bout remits (“***on-off vulnerability to alcohol***” - pathognomonic of cluster headache!!!).

* **Peripheral Nerve Blocks** see [p. S24 >>](S24.%20GENERAL%20-%20Headache.pdf#Peripheral_Nerve_Blocks)

In order of preference (begin early in cluster period and continue until headache-free for at least 2 weeks):

* 1. ergotamine (orally 2 mg × 2/d) - **classic treatment** - most effective when given 1-2 hours before expected attack (for patients with single nocturnal episode, 1 mg suppository at bedtime may be all that is necessary)

Educate regarding early symptoms of ergotism (limb claudication) when ergotamine is used daily (H: weekly limit of 14 mg).

* 1. verapamil
  2. methysergide (no longer available in USA)
  3. lithium (300 mg bid or tid titrated according to serum lithium level) – esp. useful in chronic cluster headache!
  4. prednisone (10-day course, beginning at 60 mg/d for 7 days and rapidly tapering); long-term use not recommended.
  5. divalproex
  6. capsaicin drops to ipsilateral nostril - induces **substance P** release (principal mediator of pain); after repeated applications, depletes neuron of substance P.
  7. indomethacin

N.B. propranolol and amitriptyline are largely ineffective!

* new approach: in refractory CCH + low risk for anticoagulant-related hemorrhagic complications, low-intensity anticoagulation with warfarin (to achieve INR 1.5–1.9) is associated with significantly higher incidence of remission lasting ≥ 4 weeks, as well as significantly less impact of headache on patients' quality of life as compared to placebo.

Surgical therapy:

1. new promising approach - DBS into ***posterior inferior hypothalamus***.
2. **surgical intervention** (for strictly unilateral chronic cluster) - ablation of sensory input of ***trigeminal nerve*** and ***autonomic pathways*** (e.g. percutaneous RF trigeminal gangliorhizolysis, rhizotomy) - effective in 75%.
3. SRS – methodology ***as for trigeminal neuralgia*** but results not as good (no sustained pain relief beyond 2 years F/U); some authors tried to add 8 mm shot to ***sphenopalatine ganglion*** (Pollock BE & Kondziolka D. J Neurosurg 87:450-453, 1997).

Prognosis

- chronic headache that *may last for patient's life*.

* drug therapy may convert from chronic to episodic cluster.
* prolonged, spontaneous remissions have been described in up to 12% patients.

Chronic Paroxysmal Hemicrania

- as **cluster headache** (2% prevalence of cluster headache) with following differences:

* dramatic response to indomethacin – diagnostic criterion!
* **women** > men (7:1)
* shorter headache duration ≈ 13 (5-30) minutes.
* headaches occur ≈ 5-11 times/day.
* 10% attacks may be triggered by flexing / rotating / pressing ***upper portion of neck***.
* typically, no remissions! (rarely, episodic paroxysmal hemicrania with remissions lasting weeks or months).
* MRI / CT should be undertaken to exclude symptomatic causes.
* treatment of choice - indomethacin (up to 200 mg/d); aspirin may also be beneficial, but relief is usually not complete.
* prognosis - may last indefinitely (with frequently reduced indomethacin requirement); spontaneous cures have been described.

Hemicrania continua

- exclusively 1-sided, **constant** moderate ÷ severe headache exquisitely responsive to indomethacin.

Bibliography see [p. S24 >>](http://www.neurosurgeryresident.net/S.%20Symptoms,%20Signs,%20Syndromes\S20-29.%20Pain,%20Headache,%20Opioids,%20Sensory%20Disorders\S24.%20GENERAL%20-%20Headache.pdf)

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