

OTHER PRIMARY HEADACHES: TENSION-TYPE, STABBING, HYPNIC AND NUMMULAR HEADACHE

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TENSION-TYPE HEADACHE

The most common primary headache disorder in the general population
In adults, affects slightly more women than men with peak prevalence between 30-39 years
In children, girls and boys are equally affected

Risk factors: Female sex, arterial hypertension, history of head trauma, family history of headache, being a student

Risk factors in medical students: dissatisfaction with study (males), depressed mood (females)
High prevalence of temporomandibular joint disease and widespread pain sensitivity
Trigger points in the head, neck and shoulder, while common, are not related to headache frequency

Features:

Usually bilateral pain
If unilateral, consider hemicrania continua, mild migraine, nummular headache or giant cell arteritis in the elderly
Pressing, tightening, “vice like”, “tight band”, or aching pain
May involve the neck and shoulders
Usually mild to moderate in intensity
Not aggravated by routine physical activity (helps distinguish from migraine)
No nausea or vomiting
Lasts 30 minutes to 7 days

When severe, tension-type headache may have one of the following “mild migrainous” features:

Photophobia
Phonophobia
Nausea
Throbbing
No aura or premonitory symptoms
May be triggered by stress, lack of sleep, fatigue, missing meals

Frequency < 1 day per month (<12 days per year) = Infrequent tension-type headache

Frequency < 15 days monthly for at least 3 months = Episodic tension-type headache

Frequency ≥ 15 days monthly for at least 3 months = Chronic tension-type headache

Chronic tension-type headache

Treatment:

Acute:

Acetaminophen/paracetamol (1000 mg)
Aspirin (500-1000 mg) – associated with less frequent rescue medication compared to placebo but there is limited evidence from clinical studies
NSAIDs: ibuprofen 200-800 mg, ketoprofen 25 mg, naproxen (250-500 mg)
Combination medications: Caffeine/acetaminophen/aspirin, butalbital compound (high risk of medication overuse)

Preventive:

- Tricyclic antidepressants (amitriptyline up to 75 mg, highest level of evidence)
- Tizanidine (12-24 mg daily)
- Selective serotonin/norepinephrine reuptake inhibitors (venlafaxine 150 mg daily)
- Mirtazepine (30 mg HS)
- Behavioral treatments (biofeedback, cognitive-behavioral therapy, relaxation training)
- Combined medical and behavior treatment seems most effective
- Acupuncture has moderate to low quality evidence supporting its use (mostly because of blinding) and may decrease the number of headache days

References:

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PRIMARY STABBING HEADACHE

May occur in patients with other types of headaches (migraine, cluster, hemicranias continua, paroxysmal hemicranias)

Onset at any time during life; childhood onset is not uncommon

Population prevalence <2%

Features:

- Described as stabbing, pricking, sharp, "ice pick", "jabs and jolts", "lightning pain", severe pain
- Sudden, spontaneous attacks of fleeting head pain lasting 1-10 seconds
- Usually in V1, but may occur anywhere on the head
- Unilateral, bilateral, often varying in location
- Frequency varies widely
- Usually no associated symptoms or autonomic features
- Although brief, the abruptness and severity causes distress that there is a serious underlying problem

When associated with cluster, stabs are usually ipsilateral to the side of the cluster pain and may resemble trigeminal neuralgia. The stabbing pains may occur with partially effective treatment of cluster headache or just before a cluster bout begins.

Secondary causes:

- Giant cell arteritis
- Cerebral or pituitary neoplasms
- Encephalitis
- Autoimmune disorders
- Pseudotumor cerebri syndrome

Treatment:

Preventive treatment is only needed if frequency and patient distress are high

May subside spontaneously over time

Indomethacin is first choice

Others: cyclo-oxygenase type 2 inhibitors, melatonin, gabapentin, amitriptyline, onabotulinumtoxin A

References:

- Robbins MS, Evans RW. Primary and secondary stabbing headache. *Headache* 2015;55:565-570.
- Mukharesh LO, Jann MM. Primary stabbing “ice pick” headache. *Pediatr Neurol* 2011;45:268-70.

HYPNIC HEADACHE

Rare, usually begins after age 50 (mean 61 years, range 30-83 years)

65% are women

Rare reports in childhood

Features

Develops only during sleep (not circadian, per se)

Headache occurs at a consistent time each night, usually 0100-0300, and rarely after a daytime nap

“Alarm clock headache”

Abrupt onset of diffuse, throbbing pain lasting 15-180 minutes (rarely lasts up to 10 hours)

Mild to moderate in intensity

May be unilateral

Usually located anteriorly but may involve the occiput or neck

Most patients have at least 4 attacks weekly (range 1 per week to 6 per night)

Rarely accompanied by associated symptoms or autonomic features

Reports in children: Pain was usually bilateral, lasting up to 30 minutes, throbbing, severe and usually occurred less than 15 days monthly

A retrospective study of 40 adult with hypnic headache found that the mean age at onset was 62, 80% were women, and most described their headaches as severe and burning (Tariq). Headaches occurred with a mean frequency of 21 days monthly (range 5-30) lasting a mean of 186 minutes (range 30-720). Only 3 patients had trigeminal autonomic features but 9 had migrainous features. Over half had a prior history of headache, most frequently migraine. Comorbidities included hypertension (52%), hypothyroidism (25%), a history of smoking (37%), vitamin D deficiency (28%) and history of concussion or traumatic brain injury (27%). 22% noted that physical activity or exercise immediately after awakening decreased the intensity of their headaches. Patients were followed for a mean of 3 years. Of the many medications tried, the only ones that produced a complete response (in some patients) were caffeine, lithium and melatonin. However, caffeine responsiveness was often not sustained over time. Neuroimaging in 33 patients showed no secondary cause.

Hints in the history

Nocturnal awakening in cluster headache also has an alarm clock quality and may have similar duration.

Differentiate from cluster by severity (hypnic is less severe), lack of agitation and lack of autonomic symptoms.

The paroxysmal hemicranias are less likely to cause nocturnal awakening than cluster, but it can occur.

Headaches are briefer than cluster.

Secondary causes:

Nocturnal hypertension – this has been found in patients with hypnic headache

Medication overuse – a common cause with the medication wears off

Sleep apnea – also associated with lack of the normal nocturnal “dip” in blood pressure.

Nocturnal Hypoglycemia

Brain tumor or increased intracranial pressure – definitely consider when patient is under age 50. It is rare for brain tumors to present with headache only. Funduscopy and imaging are recommended for all patients who do not have the typical phenotype of hypnic headache.

Treatment:

Caffeine (100-200 mg at bedtime, e.g. a strong cup of coffee)
Lithium carbonate (150-600 mg HS)
Melatonin (3-6 mg)
Indomethacin
Topiramate

References:

- Silva-Néto RP, Almeida KJ. Hypnic headache in childhood: A literature review. *J Neurol Sci* 2015;356:45-8.
- Silva-Néto RP, Almeida KJ. Hypnic headache: a descriptive study of 25 new cases in Brazil. *J Neurol Sci* 2014;338:166-8.
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- Traiq N, Estemalik E, Vij B et al. Long-term outcomes and clinical characteristics of hypnic headache syndrome: 40 patient series from a tertiary referral center. *Headache* 2016;56:717-24
- Lasaosa SS, Calzada JN, Benito AC, Lázaro CP. Nighttime blood pressure in cluster headache. *Headache* 2011;51:1445-9
- Caminero AB, Martin J, Sánchez del Río M. Secondary hypnic headache or symptomatic nocturnal hypertension? Two case reports. *Cephalalgia* 2010;30:1137-9
- Forsyth PA, Posner JB. Headaches in patients with brain tumors: a study of 111 patients. *Neurology* 1993;43:1678-83

NUMMULAR HEADACHE

Female:male = 1.8:1

Age at onset 4-82 years (mean 45 years)

Duration of symptoms 1 month to 50 years

Mean duration prior to presentation is about 4 years; almost half are misdiagnosed with a different primary headache disorder first

A peripheral origin is suspected

Must exclude secondary cause

A type of "epicrania" – arising from superficial or extracranial structures

Features:

Focal and well-circumscribed area of mild, moderate or severe head pain ~2-6 cm in diameter
Discrete fixed area that is round, elliptical, oval or bean-shaped (patient can outline it with their fingers)
May have superimposed exacerbations lasting seconds to days

Pain is pressure-like, sharp, stabbing, aching, throbbing, burning, stinging and itching

The involved area is often tender to palpation

Unilateral and side-locked (rarely bilateral or multifocal)

Usually located in parietal area, followed by occipital area

Distortion of sensation in affected area may occur (hypoesthesia, allodynia, paresthesias)

Triggers of exacerbations: touching the area, Valsalva, change in weather, sexual activity, coughing, menstruation

2/3 of patients have chronic, continuous pain and 1/3 have remissions lasting more than 3 months

Associated and autonomic symptoms are rare

Secondary causes:

After craniotomy

Head trauma

Pituitary lesions

Scalp lesions or skull lesions (Paget's disease, meningiomas, linear scleroderma)

Fusiform arterial aneurysms of a scalp vessel

Chronic = continuous pain from onset, or continuous pain that evolved from episodic pain (≥ 15 days monthly)
Episodic = 30 min – 6 days, less than 15 days monthly, may occur multiple times daily

Treatment:

- Gabapentin (some effectiveness in >50% of published cases)
- Tricyclic antidepressants (some effectiveness in >40% of published cases)
- OnabotulinumtoxinA (10-25 U) (at least partial benefit in all published cases)
- Indomethacin, carbamazepine, cyclobenzaprine (case reports)

References:

- Schwartz DP, Robbins MS, Grosberg B. Nummular headache update. *Curr Pain Headache Rep*
- The International Classification of Headache Disorders, 3rd edition (beta version). *Cephalalgia* 2013;33:629-808.
- Rammohan K, Mundayadan SM, Mathew R. Nummular headache: Clinico-epidemiological features in South Indian population. *J Neurosci Rural Pract* 2016;7:532-6.

EPICRANIA FUGAX

First described in 2008

Female:Male 1.7:1, mean age at onset 42 years (range 19-84)

Features:

Starts in a focal area of the parietal, parieto-occipital or temporal region and moves across the head surface in territories of different nerves

Rapidly spreads to the ipsilateral eye or nose in a linear or zigzag trajectory ("forward" EF) – most common type
Variants reported with a reverse trajectory ("backward EF") as well as unusual trajectories in the face, coronal or diagonal

Strictly unilateral and usually side locked (~72%)

Described as electric, shooting, stabbing, pressing and moderate to severe in intensity

Sometimes provoked by touching the site of origin, neck movements, eye movements, Valsalva and emotional stress

Lasts one to 10 seconds

About 1/3 have ipsilateral lacrimation (most common), conjunctival injection, itching eye, rhinorrhea or eyelid edema

May have mild interictal pain at the site of origin

Frequency varies from less than once daily to 10 times daily

Categorized as episodic or chronic, similar to other headache disorders. To date, about half are chronic.
Spontaneous remissions have been reported.

Often associated with other headaches: migraine, tension-type, cluster, trigeminal neuralgia

Differential diagnosis: Nummular headache, primary stabbing headaches, SUNCT, SUNA

Secondary causes: occipital neuralgia, trigeminal neuralgia, giant cell arteritis, structural lesions

Evaluation:

ESR, C-reactive protein (as applicable for clinical setting), neuroimaging

Treatment:

If treatment desired, success has been reported with gabapentin, lamotrigine, pregabalin, carbamazepine, levetiracetam, eslicabazepine, amitriptyline, indomethacin, GON blocks, supraorbital nerve block and trochlear block

References:

- Pareja JA, Cuadrado ML, Fernández-de-las-Peñas C, et al. Epicrania fugax: an ultrabrief paroxysmal epicranial pain. *Cephalalgia* 2008;28:253-63

- Guerrero AL, Cuadrado ML, Porta-Etessam J, et al. Epicrania fugax: ten new cases and therapeutic results. *Headache* 2010;50:451-8.
- Cuadrado ML, Guerrero AL, Pareja JA. Epicrania fugax. *Curr Pain Headache Rep* 2016;20:21

ICHD -3 BETA CRITERIA

2 Episodic Tension Type Headache

Episodes of headache, typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days. The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

Diagnostic criteria:

- A. At least 10 episodes of headache occurring
 - (2.1 Infrequent) <1 day per month and < 12 days per year
 - (2.2 Frequent) 1-14 days per month on average for 3 months (≥ 12 and < 180 days per year) and fulfilling criteria B-D
- B. Lasting 30 minutes to 7 days
- C. At least two of the following four characteristics
 1. Bilateral location
 2. Pressing or tightening (non-pulsating) quality
 3. Mild or moderate intensity
 4. Not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following
 1. No nausea or vomiting
 2. No more than one of photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis

2.3 Chronic Tension Type Headache

Evolves over time from frequent chronic tension type headache. If daily and unremitting from onset, it is termed new daily persistent headache (4.10). If onset is not remembered or is uncertain, it is termed chronic tension type headache.

Diagnostic criteria:

- A. Headache occurring on ≥ 15 days per month on average for > 30 months (≥ 180 days per year) fulfilling B-D
- B. Lasting hours to days or unremitting
- C. At least two of the following for characteristics
 1. Bilateral location
 2. Pressing or tightening (non-pulsating) quality
 3. Mild or moderate intensity
 4. Not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following
 1. no more than one of photophobia, phonophobia or mild nausea
 2. neither moderate or severe nausea nor vomiting
- E. Not better accounted for by another ICHD-3 diagnosis

All types of tension type headache are further defined by the presence or absence of pericranial tenderness:

Increased pericranial tenderness on manual palpation or
No increase in pericranial tenderness

4.8 Nummular Headache

Pain of highly variable duration, but often chronic, in a small circumscribed area of the scalp in the absence of any underlying structural lesion.

Diagnostic Criteria:

- A. Continuous or intermittent head pain fulfilling criterion B
- B. Felt exclusively in an area of the scalp, with all of the following four characteristics

1. Sharply contoured
 2. Fixed in size and shape
 3. Round or elliptical
 4. 1-6 cm in diameter
- C. Not better accounted for by another ICHD-3 diagnosis

4.9 Hypnic Headache

Frequently recurring headache attacks developing only during sleep, causing waking and lasting for up to 4 hours, without characteristic associated symptoms and not attributed to other pathology

Diagnostic criteria:

- A. Recurrent headache attacks fulfilling B-E
- B. Developing only during sleep, and causing waking
- C. Occurring ≥ 10 days per month for >3 months
- D. Lasting ≥ 15 minutes and for up to 4 hours after waking
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-3 diagnosis

4.7 Primary Stabbing Headache

Transient and localized stabs of pain in the head that occur spontaneously in the absence of organic disease or underlying structures or of the cranial nerves

Diagnostic criteria:

- A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B-D
- B. Each stab lasts for up to a few seconds
- C. Stabs recur with irregular frequency from one to many per day
- D. No cranial autonomic symptoms
- E. Not better accounted for by another ICHD-3 diagnosis

A3.11 Epicrania Fugax

Brief paroxysmal head pain, with stabbing quality, describing a linear or zig-zag trajectory across the surface of one hemicranium.

Diagnostic Criteria:

- A. Recurrent stabbing head pain attacks lasting 1-10 seconds fulfilling criterion B
- B. The pain is felt to move across the surface of one hemicranium in a linear or zigzag trajectory, commencing and terminating in the territories of different nerves
- C. Not better accounted for by another ICHD-3 diagnosis