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# I) Introduction

Headache disorders are among the <u>most prevalent medical problems</u> worldwide

The <u>World Health</u> <u>Organization</u> estimates that 50% to 75% of all adults between the ages of 18 and 65 years have headaches

This high prevalence results in <u>significant</u> <u>disability</u> and <u>lost</u> <u>productivity</u>: headache disorders are the 3<sup>rd</sup> highest cause of years lost to disability worldwide Accordingly, headaches are one of the <u>most common reasons</u> patients present to physicians in <u>primary care</u> settings, in the <u>emergency</u> department (ER), or in <u>neurologists</u>' <u>offices</u>

For clinicians evaluating a patient with headaches, the 1<sup>st</sup> responsibility is to <u>diagnose</u> the <u>type</u> of <u>headache</u> correctly, and then <u>treat</u> <u>appropriately</u>, because most headache disorders have <u>excellent</u> <u>treatments</u> available, which can <u>reduce</u> the burden of <u>disability</u> <u>Headache disorders</u> fall into <u>2 categories</u>, <u>primary headache disorders</u> ( those caused by the headache disorder itself, not due to other causes) and <u>secondary headache disorders</u>, those caused by ( or "symptomatic of") another underlying medical problem

The pain can be due to the involvement of <u>pain-sensitive</u> <u>structures</u> in the <u>head</u>, including cranial nerves, cervical nerve roots, blood vessels, meninges, scalp, temporomandibular joint (TMJ), teeth, pericranial and cervical muscles, and paranasal sinuses Patients may also have <u>multifactorial headaches</u>, so a <u>detailed history</u> and <u>examination</u> are <u>necessary</u> to identify the contributing factors

Headache disorders may <u>remain refractory</u> to <u>treatment</u> or have an insufficient response to treatment if these <u>comorbidities</u> are <u>not</u> <u>identified</u> and <u>addressed</u>



A detailed <u>history</u> and <u>examination</u> are vital in understanding the headache's cause

There are <u>no biomarkers</u> currently available for <u>primary headache</u> <u>disorders</u>

#### Key points from the history

When obtaining the history the <u>following information</u> must be elicited:

- Onset
- Precipitants and triggers
- Duration
- Location( unilateral or bilateral; frontal, lateral, vertex ,or occipital)
- Quality and severity
- Frequency
- Alleviating and exacerbating factors
- Positional influences (better or worse when supine)
- Waking the patient from sleep, or occurring upon awakening
- Associated with menses
- Associated symptoms

<u>Additional aspects</u> of the <u>history</u> important in evaluating a patient with headache are:

- Analgesic use
- Caffeine use
- Medical history
- Current or recent pregnancy
- Medications( including asking specifically about contraceptive use, over-the –counter treatments, and supplements)
- Social history, including detailed screening for illicit drugs
- Family history
- Sleep, including a history of insomnia and snoring, symptoms suggestive of <u>obstructive</u> <u>sleep</u> <u>apnea</u>

The <u>semiology</u> of the headache helps to <u>differentiate</u> a <u>primary</u> from a <u>secondary headache</u> disorder

The <u>history</u> also allows a clinician to identify <u>red flags</u> that <u>suggest</u> a <u>secondary</u> headache disorder

BUA

Acute onset or progressive worsening from

- baseline
  New or different headache
- Systemic symptoms:
  - Fever, weight loss
- Risk factors:

Malignancy Immunosuppression IV illicit substance use Hypercoagulability, including pregnancy

- Smoking
- Age>50, or no prior headache history
- Features of increased ICP:
  - Waking patient from sleep
    - Worsening with Valsalva maneuver
  - Supine worsening of pain
- Focal features:
  - Seizures
  - Mental status abnormality
  - **Cranial nerve deficits**
  - Weakness
  - Sensory changes (loss of sensation, pares-

thesias; location and pattern of spread)
 Precipitants:

Trauma Newly prescribed medications Infection

#### RED FLAGS

- Acute onset or progressive worsening from baseline
- New or different headache
- <u>Systemic symptoms</u>: fever, weight loss
- <u>Risk factors</u>: malignancy, immunosuppression, IV illicit substance use, hypercoagulability, including pregnancy
- Smoking

- Age more than 50 years, or no prior headache history

- Features of <u>increased ICP</u>: waking patient from sleep, worsening with Valsalva maneuver, supine worsening of pain
- <u>Focal features</u>: seizures, mental status abnormality, cranial nerve deficits, weakness, sensory changes( loss of sensation, paresthesias, location of pattern of spread)
- <u>Precipitants</u>: trauma, newly prescribed medications, infection

#### <u>Key points</u> for the <u>neurological</u> <u>examination</u>

Patients with <u>primary headache</u> <u>disorders</u> usually have <u>normal</u> general medical and neurological <u>examinations</u>, although an acutely symptomatic patient with an <u>autonomic cephalalgia</u> may have <u>signs</u> <u>suggesting that disorder</u>

Some patients with <u>chronic</u> <u>headaches</u> have findings of <u>temporomandibular joint (TMJ)</u> tenderness on palpation, evidence of dental wearing, or pain with palpation of the cervical muscles or the occipital ridge to suggest <u>comorbid causes</u> of headache such as <u>cervicalgia</u> In the era of the <u>smartphone</u>, patients may bring <u>pictures</u> of themselves to a clinician for review if they have <u>paroxysmal symptoms</u> and <u>signs(</u> such as <u>ptosis</u> or <u>lacrimation</u>), which can aid in the diagnosis

Patients should have a general <u>medical</u> and <u>neurological</u> <u>examination</u> to <u>assess</u> for <u>secondary</u> <u>causes</u> of <u>headache</u>

Attention to <u>vital signs</u> is important: patients with significant <u>hypertension</u> may be susceptible to developing <u>certain secondary</u> <u>headaches</u> outlined below

Fever may suggest an underlying infection, including a CNS infection

A <u>cardiovascular</u> <u>exam</u> can evaluate for arrhythmia or carotid stenosis, which can cause secondary headache syndromes

A detailed <u>head</u> and <u>neck exam</u> includes evaluating for nuchal rigidity, cervical myofascial pain, <u>occipital Tinel sign</u>(evaluated by eliciting tenderness or tingling when palpating near the occipital protuberance along the occipital nerve), and <u>palpation</u> of the <u>TMJ</u>, assessment of <u>dental wearing</u> or <u>chipping</u> to suggest <u>bruxism</u>, and observing the <u>oropharynx</u> for narrowing that could suggest <u>obstructive sleep apnea</u> A full <u>neurological</u> <u>exam</u> should also be performed, with emphasis on the <u>funduscopic</u> exam to assess for <u>papilledema</u>

The <u>cortical sensory exam</u> can suggest cortical dysfunction that may occur with venous sinus thrombosis

<u>Focal neurological deficits</u>, including field cuts, cranial nerve palsies, weakness, or sensory symptoms, often <u>suggest</u> a <u>secondary headache</u>

## III) <u>Primary headache disorders</u>

Primary headache disorders are those <u>not due</u> to <u>another</u> <u>medical</u> <u>condition</u>

<u>Diagnosis</u> is established by history and exam

<u>Migraine</u> is by far the most prevalent primary headache disorder

The <u>table</u> outlines <u>common</u> <u>primary</u> <u>headache</u> <u>disorders</u> based on key features of the history

The <u>figure</u> shows the <u>common</u> <u>locations</u> of <u>pain</u> in the <u>primary</u> headache disorders, <u>compared</u> to that of headaches caused by <u>sinus</u> <u>disease</u>

	Episodic Migraine	Episodic Tension	Episodic Cluster	Hemicrania	SUNCT and SUNA
				Female > male	Male > female
Sex	Female > male	Female > male	Male > lemale	Unilateral	Unilateral
Location	Unilateral > bilateral	Bilateral (band around the head)	Unilateral (behind or around the eye)	(behind or around the eye)	(behind or around the eye)
Quality	Throbbing, pulsatile	Dull pressure or tightening (vice-like)	Stabbing, burning, boring	Stabbing, burning, throbbing	Stabbing, burning
Severity	Moderate to severe	Moderate	Severe	Severe	Severe
Attack duration	4–72 h	30 min–7 d	15–180 min	2–30 min	1 s-10 min
Attack frequency	Variable	Variable	From 1 every other day to 8/d	>5/d to 40/d	From 1/d to 200/d
Autonomic eatures	No	No	Yes	Yes	Yes

\*ADAM

FIGURE 10-1. Location of pain associated with primary headache disorders. *Left* to *right*: sinus headache, cluster headache, tension headache, migraine headache. (Used with permission of A.D.A.M.)

withdrawal and over acc,

#### \* <u>MIGRAINE</u>

## A) <u>Migraine without aura</u>

It is estimated that <u>1 in 7 adults</u> worldwide has migraine

It impacts women more than men in a 2:1 ratio

Migraine may start in <u>childhood</u> and manifest occasionally with abdominal symptoms( "<u>abdominal migraine</u>")

<u>Motion sickness</u> in <u>children</u> is a <u>risk factor</u> for the development of migraine

# <u>Migraine</u> headaches are most likely to develop in <u>adolescence</u> and <u>early</u> <u>adulthood</u>

They can be <u>episodic</u> or <u>chronic</u>

The <u>disability</u> and <u>lost productivity</u> from migraine are <u>substantial</u>, because it impacts people in their prime working years

Migraine has numerous <u>identified</u> triggers, including <u>weather</u> <u>changes</u>, <u>menses</u> and <u>caffeine(</u> both <u>withdrawal</u> and <u>overuse</u>)

Many patients identify <u>foods</u> and <u>drinks</u> such as <u>alcohol</u>(most commonly red wine), <u>soft cheeses</u>, and nitrite-heavy foods, such as <u>processed meats</u>, as precipitants, but data are sparse in this area and <u>many migraine attacks</u> occur <u>without</u> identifiable <u>triggers</u> <u>To diagnose migraine</u>, a patient must have at least <u>5 attacks</u> with the <u>following characteristics</u>:

- 1. The headache lasts for 4 to 72 hours if untreated
- 2. It must include at <u>least 2</u> of the following <u>features</u>:
  - a. Throbbing
  - b. Unilateral headaches
  - c. Worsening with activity, such as walking
  - d. Moderate to severe pain
- 3. It must be <u>associated with at least one</u> of the following:
  - a. Nausea, vomiting, or both
  - b. Photophobia and phonophobia

## B) Migraine with aura

Migraine headaches are often <u>preceded</u> by focal neurological symptoms known as <u>auras</u>

These are also called <u>classic migraine</u> or <u>complicated migraine</u>

<u>Auras</u> are <u>defined</u> as fully reversible neurologic symptoms with a gradual onset, usually followed by a headache

The <u>aura</u> usually <u>lasts</u> 5 to 60( often <u>20</u>) <u>minutes</u> and is typically unilateral

It usually <u>resolves</u> <u>without</u> lingering neurologic <u>deficits</u>

Patients are diagnosed with this disorder when they have an <u>aura</u> <u>followed</u> by a <u>headache</u> that meets the criteria for <u>migraine</u>, as above

Some auras occur <u>without</u> a <u>headache</u> ("<u>acephalgic migraine</u>"), but these symptoms usually require <u>additional investigation</u> for a definite diagnosis

<u>Visual auras</u> are by far the <u>most common</u>

Some include a" <u>fortification</u>" <u>spectrum</u>( zigzag lines off the central vision, usually spreading gradually) or a <u>scintillating</u> ( or flickering) <u>scotoma</u> ( an area of decreased visual acuity surrounded by preserved vision)

<u>Migraine auras</u> can also involve the <u>sensory symptoms</u>, most commonly paresthesias (tingling or pins-and-needles sensation)

The <u>paresthesias</u> often "<u>march</u>" or <u>spread</u> <u>gradually</u> over the course of several minutes along a limb or extend from an arm to the leg or face

<u>Migraine auras</u> can also include a <u>gradual</u> onset of <u>weakness</u>, a variant known as <u>hemiplegic migraine</u> when <u>severe</u>

<u>Hemiplegic migraine</u> may be <u>sporadic</u> but there is also a syndrome of <u>familial</u> hemiplegic migraine, sometimes associated with wellcharacterized <u>genes</u> Migraine auras are believed to be due to "<u>cortical spreading</u> <u>depression</u>" in which there is a spread of <u>hyperpolarization</u> of the cortex <u>followed by</u> a wave of <u>depolarization</u>

<u>Imaging studies</u> have shown <u>decreased</u> <u>regional</u> <u>cerebral</u> <u>blood</u> <u>flow</u> in the cortex during migraine aura, but not to the level of worrisome ischemia

# C) <u>Complications associated with migraine</u>

\*<u>Status</u> migrainosus

When <u>migraine lasts</u> for more than <u>72 hours</u>, the condition is known as status migrainosus

This is often caused by <u>abortive medication overuse</u> (often referred to as <u>rebound headache</u>) and frequently requires <u>intravenous</u> (IV) <u>treatment</u> or a brief <u>course</u> of <u>oral steroids</u> to break the headache cycle

#### \* <u>Stroke risk associated with migraine</u>

Patients with <u>migraine</u> with <u>aura</u> have an <u>increased</u> <u>cardiovascular</u> <u>risk</u> when compared to healthy controls

The use of <u>oestogen-based</u> <u>contraceptives</u> is therefore <u>contraindicated</u> in patients with <u>migraine</u> <u>with</u> <u>aura</u>, as the combination results in a substantially <u>increased</u> <u>stroke</u> <u>risk</u>

## \* Migraine and menses

Women of reproductive age frequently have <u>exacerbation</u> of <u>migraine</u> <u>during menses</u>, most commonly 1 to 2 days prior to onset of bleeding, often persisting for up to 3 days into bleeding

This is thought to be due to the <u>withdrawal</u> of <u>estrogen</u> that occurs with menses

Some women have migraine at the time of menstruation only, a condition known as pure <u>menstrual migraine</u>

Most , however, have a <u>few episodic headaches</u> at <u>other</u> times of the month, or menstrually related migraine

It is important to <u>identify</u> the <u>relationship</u> of <u>menses</u> to <u>migraine</u> because there are <u>specific</u> <u>treatments</u> that may be helpful for patients with a clear exacerbation around their menses

# \* <u>Chronic migraine</u>

Patients who have a <u>headache more</u> than <u>15 days/month</u> for <u>more</u> than <u>3 months</u> are diagnosed with chronic migraine

Some patients with chronic migraine do <u>not</u> have <u>typical features</u> of <u>migraine</u> with <u>all headaches</u>, but they must have at <u>least</u> 8 <u>days</u> of <u>headache</u> consistent with <u>migraine</u> to be diagnosed with chronic migraine

<u>If</u> the headaches are <u>not</u> consistent with <u>migraine</u>, <u>other</u> <u>diagnoses</u> must be considered Patients often describe a history of <u>gradually progressive episodic</u> <u>migraines</u> that increase in frequency to the point of meeting criteria for chronic migraine

With frequent headaches, many patients with chronic migraine have some <u>component</u> of <u>medication</u> <u>overuse</u> <u>headache</u> (MOH)

Importantly, patients with chronic migraine can <u>revert</u> to <u>episodic</u> <u>migraine</u> after effective treatment

#### D) <u>Migraine treatments</u>

#### 1. <u>Abortive treatments</u>

Abortive treatments, also called <u>rescue</u> <u>medications</u>, are medications used to stop a migraine at the onset

All abortive treatments are <u>most effective</u> if the patient is treated <u>at</u> the <u>onset</u> of the headache

<u>Delay</u> in <u>treatment</u> results in more <u>prolonged</u> <u>disability</u>, so patients must be counseled on the appropriate use of abortive treatments

Nonsteroidal anti-inflammatory drugs (<u>NSAIDs</u>) and <u>triptans</u> (<u>serotonin 1b/1d agonists</u>) are the mainstay of abortive treatments

Many patients respond to <u>NSAIDs</u> alone

For some patients, however, they are <u>insufficient</u>; some patients have <u>contraindications</u> to using <u>NSAIDs</u>. In these cases, <u>triptans</u> can be highly <u>effective</u>

There are numerous <u>different</u> types of <u>triptans</u>, with different rates of onset of action and half-lives
There are 2 <u>long-acting</u> triptans (naratriptan and frovatriptan) and 5 <u>fast-acting</u> triptans (almotriptan, eletriptan, sumatriptan, rizatriptan, and zolmitriptan)

There are also numerous <u>different</u> formulations, including oral pills, disintegrating tablets, nasal sprays, and injectables

Historically, <u>ergotamines</u> were prescribed as abortive treatments, but they carry a <u>higher cardiovascular risk</u> and have been <u>replaced by</u> <u>triptans</u>

<u>Triptans</u> and <u>NSAIDs</u> can be <u>combined</u> when needed and may have a <u>synergistic</u> effect in treating migraine pain

<u>Caffeine</u> is also often added to many migraine treatments because it <u>can help abort</u> the <u>pain</u>; many over-the-counter " migraine preparations" contain caffeine

<u>Triptans</u> are currently <u>not</u> known to be <u>safe</u> in <u>pregnancy</u> and have a <u>cardiovascular</u> <u>risk</u>

<u>Triptans</u> also <u>interact</u> with selective serotonin reuptake inhibitors and serotonin-norepinephrine reuptake inhibitors , with a <u>low risk</u> of <u>serotonin syndrome</u>

Patients must be <u>counseled</u> on <u>side</u> <u>effects</u> of all treatments

<u>Identifying</u> the <u>right abortive treatment</u> requires <u>careful</u> <u>consideration</u> of the patient's headache features , comorbidities, concurrent medication use, cost, and family planning goals Using <u>abortive</u> treatments on a chronic basis <u>more</u> than <u>twice</u> a <u>week</u> can result in <u>medication</u> <u>overuse</u> <u>headache</u> (MOH), so patients should be counseled to <u>not use</u> any of these treatments chronically <u>more</u> than <u>twice</u> a <u>week</u> to prevent this complication

# 2. Adjuvant treatments

Because nausea and emesis are frequently associated with migraine, many patients benefit from <u>antiemetics</u>

Interestingly, <u>prochlorperazine</u> and <u>metoclopramide</u> are more effective than ondansetron, both in alleviating the nausea and in reducing the severity of the pain

Antiemetics may also be useful in <u>preventing</u> patients from <u>vomiting</u> their <u>abortive</u> therapies

They are frequently <u>used</u> in <u>emergency</u> <u>room</u> (ER) and urgent care settings for patients with <u>severe</u> or refractory <u>migraine</u>

They are often <u>combined</u> with <u>ketorolac</u> and <u>diphenhydramine</u> for patients with <u>status</u> <u>migrainosus</u>

## 3. <u>Preventive</u> treatments

Preventive treatments, also called <u>prophylactic treatments</u>, are used for patients with <u>chronic migraine</u> or <u>frequent</u> and <u>disabling headaches</u> that do not respond sufficiently to abortive treatments

Preventive therapy aims to <u>reduce</u> the <u>frequency</u> and <u>severity</u> of migraine, although patients are <u>unlikely</u> to <u>become</u> <u>completely</u> <u>headache-free</u> and should be counseled accordingly

All prophylactic treatments take <u>some time to</u> have an <u>effect</u> ;patients should remain on treatment for <u>at least a month</u> ( barring significant side effects or other concerns) <u>before assuming</u> that the treatment is <u>ineffective</u>

There are <u>3 primary categories</u> of <u>preventive</u> <u>oral</u> <u>medications</u>: antihypertensives, antiseizure medications, and antidepressants

Within each category, there are <u>specific</u> <u>drugs</u> with the most evidence of efficacy( table)

In addition to oral therapies, <u>onabotulinum toxin A</u> (often referred to simply as <u>Botox</u>) was also approved as migraine prophylaxis for <u>chronic migraine</u> in 2010



In 2018, a new class of <u>preventative</u> <u>therapy</u> for <u>chronic</u> <u>migraine</u>, <u>Calcitonin</u> <u>Gene</u> –<u>Related</u> <u>Peptide</u> (CGRP) <u>antagonists</u> was approved by the FDA

<u>Erenumab</u> is an injectable <u>human</u> <u>monoclonal</u> <u>antibody</u> that <u>antagonizes</u> <u>CGRP</u> receptor function

As with abortive treatments, <u>selecting</u> the <u>right</u> <u>prophylactic</u> <u>medication</u> <u>requires</u> careful <u>consideration</u> <u>of</u> the patient's comorbidities, concomitant medications, cost, and family planning goals

Patients must be <u>counseled</u> <u>about</u> <u>treatment</u> <u>options</u> and <u>side</u> <u>effects</u>, including teratogenicity and impact on contraceptives

Patients who require preventive therapies <u>also require</u> <u>abortive</u> <u>treatments</u>

Some <u>abortive treatments interact</u> with <u>prophylactic medications</u> ( such as antidepressants and triptans) which should be taken in consideration

## 4. <u>Lifestyle modifications</u>

Lifestyle factors are important to identify

A <u>comorbid sleep disorder</u> (insomnia, obstructive sleep apnea ...) makes patients more susceptible to migraine

Skipping meals, insufficient fluid, excessive caffeine intake, and lack of exercise make <u>susceptible</u> patients more <u>prone to migraine attacks</u>

Patients should be counseled on these factors

\* <u>TENSION-TYPE</u> <u>HEADACHE</u>

Tension-type headache (often referred to as tension headaches, <u>stress</u> <u>headaches</u>, or <u>ordinary headaches</u>) are the next most prevalent primary headache disorders, occurring in 30% to 70% of adults worldwide

Pain is usually <u>bilateral</u> and described as <u>pressure</u> or <u>tightness</u>

It is usually <u>mild</u> to <u>moderate</u> and <u>lasts</u> for under an hour to several <u>days</u>

Unlike migraine, it is <u>not associated</u> with <u>photophobia</u>, phonophobia, <u>nausea</u>, or <u>vomiting</u>

The <u>examination</u> is generally <u>normal</u>, but some patients have <u>pericranial</u> <u>tenderness</u> to palpation of the scalp, neck, or shoulder muscles

Tension-type headaches can be <u>episodic</u> or <u>chronic</u>( occurring more than 15 days/month)

Interestingly, patients with <u>infrequent tension-type headaches</u> generally <u>do not seek medical attention</u>, because they do not have significant disability from their symptoms

Patients with <u>frequent</u> or <u>chronic</u> <u>tension-type</u> <u>headaches</u> <u>benefit</u> from <u>treatment</u>

#### Tension-type headache treatment

#### 1. Abortive treatments

Many patients with tension headaches <u>do not require abortive treatments</u> because the pain is generally <u>mild</u> and does not interfere with the patient's functioning

# For those with <u>moderate</u> to <u>severe</u> pain, <u>NSAIDs</u> are the mainstay of treatment

<u>Aspirin</u> and <u>acetaminophen</u> may also be used, but the latter is often less effective than NSAIDs

Patients should be <u>counseled</u> about the <u>development</u> of <u>MOH</u> and advised to not use analgesics more than twice a week for long periods

### 2. <u>Preventive</u> treatments

<u>Antidepressants</u> are the first-line preventive therapy for chronic tension headache

The tricyclic <u>amitryptiline</u> is the most studied to date and has good evidence for efficacy

<u>Other antidepressants</u>, including mirtazapine and venlafaxine are second-line therapies

<u>Muscle relaxants</u> such as <u>tizanidine</u> are helpful sometimes, particularly in patients with a <u>cervicogenic component</u>

## 3. <u>Adjuvant treatments</u>

Tension headaches are often reported to be <u>triggered</u> by <u>stress</u> (physical or emotional); addressing these triggers, if chronic, is important

<u>Biofeedback</u> (a mind-body technique used to teach patients greater body awareness and how to control some physical reactions to pain and stress) <u>can be effective</u>

<u>Poor posture</u> and <u>neck muscle spasm</u> are also frequent contributors to chronic tension-type headaches, and <u>physical therapy</u> can help

# \* <u>TRIGEMINAL</u> <u>AUTONOMIC</u> <u>CEPHALALGIAS</u>

Trigeminal autonomic cephalalgias (TACs) are the <u>3<sup>rd</sup> major category</u> of primary headache disorders

They are characterized by <u>unilateral</u> <u>pain</u> associated with <u>cranial</u> <u>autonomic</u> <u>symptoms</u>

The <u>diagnosis</u> is made by careful evaluation of the <u>pattern</u> of the <u>pain</u> and its <u>associated</u> <u>features</u> ( table shown on a previous slide)

# A) <u>Cluster headache</u>

Cluster headaches are <u>severe headaches</u> characterized by unilateral pain involving the <u>orbit</u>, <u>supraorbitally</u>, at the <u>temple</u>, or combination of these( previous figure)

Cluster periods are <u>bouts</u> of <u>recurrent</u> <u>attacks</u> of <u>pain</u>, generally lasting weeks to months

These periods are <u>followed</u> by <u>remission</u> lasting anywhere from months to years

The pain is often excruciating

During an attack, patients are often <u>restless</u> and <u>pacing</u>, unlike in migraine where activity exacerbates the pain

The pain must be <u>associated</u> with <u>one</u> of the <u>following cranial</u> <u>autonomic symptoms</u>:

- Conjunctival injection, lacrimation, or both
- Nasal congestion, rhinorrhea, or both
- Eyelid edema
- Forehead and facial sweating or flushing
- Sensation of fullness in the ear
- Miosis, ptosis, or both

Cluster headaches typically last between 15 and 120 minutes

During a cluster period, headaches can occur <u>several times</u> a <u>day</u> or as infrequently as <u>every other day</u>

Cluster headaches are relatively uncommon but are <u>3 times more</u> likely to occur in <u>men</u>

The <u>age</u> of <u>onset</u> is typically in <u>early</u> to <u>mid-adulthood</u> (20-40 years)

The <u>cause</u> is <u>unknown</u>, but <u>activation</u> of the <u>posterior hypothalamic</u> <u>gray matter</u> has been seen in some patients during attacks

<u>Alcohol</u>, <u>histamines</u>, and <u>nitroglycerine</u> are <u>triggers</u> in susceptible patients

A <u>Horner syndrome</u> caused by <u>carotid dissection</u> may <u>mimic</u> a <u>cluster</u> <u>headache</u> (figure) but does not usually have the pain characteristics of this primary headache disorder

Cluster headaches may be <u>episodic</u> or <u>chronic</u>

<u>Chronic cluster</u> headache is defined as intractable cluster headaches with <u>less than 1 month</u> of <u>remission</u> before the recurrence of symptoms

Fortunately, less than <u>15%</u> of cluster patients have <u>chronic cluster</u>



FIGURE 10-2. Cluster headache. Horner syndrome. Note mild unilateral ptosis (on the patient's right side), anisocoria with a smaller pupil on the side of the ptosis, and redness from associated cluster headache. The common "upsidedown" ptosis (i.e., elevation) of the lower lid is masked by coexisting eyelid laxity and blepharitis, which can be confounding factors in older adults. <u>Cluster headache treatments</u>

### 1. <u>Abortive treatments</u>

A first-line abortive treatment for cluster headaches is <u>100%</u> oxygen, delivered at <u>12</u> to <u>15L/min</u>

Patients may receive this treatment in an <u>urgent care</u> or <u>ED</u> <u>setting</u> and if effective, be prescribed a <u>home oxygen tank</u>

For patients <u>who do not respond</u>, or who do not have access to home oxygen, <u>triptans</u> are prescribed

<u>Sumatriptan</u> and <u>zolmitriptan</u> are effective as abortive therapies

In the past, <u>dihydroergotamine</u> (<u>DHE</u>) was prescribed as an abortive treatment , but triptans are <u>favored</u> over DHE <u>given their</u> <u>safety</u> <u>profile</u>

Patients with cluster headache are very <u>susceptible</u> to <u>MOH</u> and must be counseled appropriately

Occipital nerve block can also be effective to abort a cluster cycle

## 2. <u>Preventive</u> treatments

Preventive treatments for cluster headache are <u>similar</u> to <u>those</u> used <u>for migraine</u> and include antihypertensive, antiseizure, and psychiatric medications

<u>Verapamil</u> is the first-line therapy for cluster headache prophylaxis

If <u>not tolerated</u> or if there are <u>contraindications</u>, <u>glucocorticoids</u> ( prednisone or dexamethasone) are also effective

<u>Lithium</u> and <u>topiramate</u> are often used as <u>second-line</u> <u>agents</u> or as addon therapy when needed

# IV) <u>Opioids in headache treatment</u>

Opioids are <u>not more effective</u> than alternative therapy and are generally <u>strongly discouraged</u> for use in headache medicine

Most headache conditions are <u>recurrent</u> <u>disorders</u>, so to use opiods in this setting <u>risks</u> development of a <u>secondary opioid</u> <u>use</u> disorder

Opioids also tend to cause MOH

# V) <u>SECONDARY HEADACHE DISORDERS</u>

Secondary headache disorders are headaches <u>caused</u> by a <u>medical</u> <u>condition</u> or <u>medication</u>

They have a <u>broad</u> <u>differential</u> for <u>causes</u>, ranging from preeclampsia and pheochromocytoma to fever and medication side effects

Most secondary headache disorders are <u>associated with other features</u> in the history, examination, or laboratory assessment, which <u>aid in the</u> <u>diagnosis</u>

<u>Treatment</u> is based on <u>addressing</u> the <u>underlying</u> <u>disorder</u>

There are <u>6</u> major <u>categories</u> of secondary headaches that may present with <u>headache</u> <u>only</u> and must be considered

## 1. <u>Vascular causes</u>

There are <u>numerous</u> vascular <u>causes</u> of headache

## All <u>cerebral hemorrhages</u> can cause headache

<u>This includes</u> subarachnoid hemorrhage(SAH), intraparenchymal hemorrhage, and subdural and epidural hematomas

These hemorrhages may be <u>spontaneous</u> (associated with stroke or hypertension) or <u>traumatic</u>

Patients with intracerebral hemorrhages typically present with what is referred to as <u>thunderclap headache</u>; the <u>onset</u> is <u>abrupt</u> and <u>severe</u>

<u>Emergency imaging</u>, usually with a <u>noncontrast CT</u> <u>Brain scan</u>, is needed to evaluate abrupt-onset headache( figure)

<u>Cerebral vessel</u> imaging is also warranted if a <u>SAH</u> is identified , to assess for an <u>aneurysm</u>



s of intracerebral hemorrhages on CT scans. **(A)** Epidural hemorrhage. **(B)** Su hemorrhage. **(D)** Subarachnoid hemorrhage. Arrows point to the subdural he Their semiology is nonspecific, but typically <u>abrupt</u> in <u>onset</u>

Patients may have <u>focal neurologic deficits</u> which aid in the diagnosis

<u>Cerebral thrombosis</u>, either <u>arterial</u> or <u>venous</u>, can also cause headache

Patients with <u>venous sinus</u> <u>thrombosis</u> often have headaches with features of <u>increased</u> <u>intracranial</u> <u>pressure(ICP)</u>

Thrombosis should be considered particularly in patients with <u>hypercoagulability states</u>, including <u>pregnancy</u>

The <u>diagnosis</u> is made on <u>imaging</u>, including that of <u>cerebral vessels</u> (figure)

![](_page_67_Picture_0.jpeg)

E 10-4. Deep venous sinus thrombosis in a pregnant woman. Sagittal image from a magnetic resonance ram (MRV) demonstrating occlusion of the deep venous system, including the straight sinus (red arrow). The or sagittal sinus (green arrow) and right transverse sinus (blue arrow) and sigmoid sinus (orange arrow) are paten

<u>Cerebral vasculitis</u> frequently causes a nonspecific headache

When part of a systemic vasculitis, it is considered a secondary angiitis

If the vasculitis occurs in the cerebral vessels alone, it is referred to as primary CNS angiitis

In addition to <u>headache</u>, patients often have paroxysmal <u>focal</u> neurological <u>deficits</u>

<u>Cerebral arterial vessel imaging</u> and lumbar puncture (<u>LP</u>) are often required to make this diagnosis

# Giant cell arteritis (GCA),

also called <u>temporal arteritis</u>, is a peripheral cranial arterial vasculitis that often presents with <u>unilateral headache</u>

Patients are generally <u>above</u> the <u>age</u> of <u>50</u> and report additional symptoms including vision changes( <u>amaurosis</u> <u>fugax</u>), <u>jaw</u> <u>claudication</u>, <u>fever</u>, and <u>scalp</u> <u>tenderness</u>

Involvement of the <u>branches</u> of the <u>external</u> <u>carotid</u> <u>artery</u>, and occasionally the <u>ophthalmic</u> <u>artery</u>, can result in <u>blindness</u> if <u>not</u> readily identified and <u>treated</u> <u>promptly</u> Patients usually have <u>elevated</u> inflammatory <u>markers</u>( erythrocyte sedimentation rate and C-reactive protein)

<u>Empiric steroids</u> should be started in any patient with a <u>high</u> <u>clinical</u> <u>concern</u>

<u>Temporal artery</u> biopsy is the gold standard, but GCA can cause "<u>skip</u> <u>lesions</u>" and may require <u>serial</u> <u>biopsies</u> to identify the pathology

## 2. Infectious or inflammatory causes

Intracranial infections, such as <u>encephalitis</u> and <u>meningitis</u>, usually present with <u>headache</u> and often <u>fever</u>

They may also have <u>nuchal rigidity</u> and <u>altered mental status</u>

As the infection progresses , <u>seizures</u> and <u>focal</u> neurological <u>deficits</u> may occur

Infections can be <u>bacterial</u>, <u>viral</u>, <u>fungal</u>, or <u>parasitic</u>; the <u>headache</u> <u>semiology does not help</u> to <u>differentiate</u> the underlying cause
<u>LP</u> is the <u>crucial diagnostic test</u> and necessary in any patient for whom there is concern for a CNS infection

There are often <u>other signs</u> of <u>infection</u>, and the <u>headache</u> has a temporal <u>correlation</u> to the infection

<u>CNS</u> inflammatory and <u>autoimmune</u> <u>conditions</u> such as <u>sarcoidosis</u> and <u>lupus</u> frequently present with headache

<u>Intracranial neoplasms</u> may present with headaches, especially when there is significant <u>mass</u> <u>effect</u>

The <u>headache semiology</u> may be nonspecific but may have <u>features</u> of <u>intracranial hypertension</u>, including wakening the patient from sleep , being worse when supine, and worse with Valsalva maneuver

The headache may <u>occur</u> <u>early</u> or <u>late</u> with neoplasms and with any type of primary cancer

<u>Head</u> and <u>neck</u> injuries often result in headache

To be attributed to trauma, the <u>headache</u> must develop with a <u>temporal association</u> with the injury

The <u>severity</u> of the <u>injury</u> does <u>not</u> necessarily <u>correlate</u> with the <u>severity</u> of the <u>headache</u>; even minor head injuries or whiplash may cause headaches

There is <u>no specific</u> headache <u>semiology</u> that helps with the diagnosis

In patients with a history of significant <u>head</u> or <u>neck</u> injury, it is important to assess for <u>intracranial hemorrhage</u> or <u>dissection</u> of <u>cervical vessels-</u> which may require additional treatment

# 5. Intracerebral pressure disorders

<u>Intracranial hypertension</u> and <u>hypotension</u> can cause headaches, but with markedly different semiologies

### A) Intracranial hypertension

Intracranial hypertension may be "<u>idiopathic</u>"(most common in <u>obese</u> young <u>woman</u>) or due to <u>medications</u> or <u>systemic</u> <u>disorders</u>

The headache is often described as <u>worse</u> when <u>supine</u> or <u>sleeping</u> (awakening the patient from sleep), or with <u>Valsalva</u> maneuver

Patients often have <u>associated</u> <u>features</u>, including <u>papilledema</u>, <u>pulsatile</u> <u>tinnitus</u>, or <u>visual</u> symptoms

# Patients should have <u>imaging</u> to <u>exclude</u> a <u>mass</u> <u>lesion</u> or <u>venous</u> sinus <u>thrombosis</u>

If the <u>imaging</u> is <u>unrevealing</u>, the <u>diagnosis</u> is made with an <u>LP</u> when the patient is in the lateral decubitus position with legs extended

Intracerebral pressure (ICP) is elevated above 200 mm CSF

<u>Acetazolamide</u> is the first-line treatment for idiopathic intracranial hypertension (IIH)

Patients require <u>monitoring</u> of their <u>visual fields</u>, and <u>treatment</u> for <u>obesity</u> is warranted when present

## B) Intracranial hypotension

Intracranial hypotension may be <u>spontaneous</u> or <u>traumatic</u>

The headache <u>improves</u> when the <u>patient</u> is <u>supine</u> but worsens with standing

The pain is often most <u>severe</u> at the <u>vertex</u> and can be associated with <u>neck pain</u> or <u>tinnitus</u>

It is <u>frequently traumatic-</u> occurring after an attempted <u>epidural</u> <u>puncture</u> or <u>LP</u>

Generally, the <u>dural leak</u> causing the headache <u>heals</u> gradually, without intervention, but <u>when symptoms</u> <u>persist</u> or when the headache <u>etiology</u> is <u>unclear</u>, <u>brain MRI</u> may be helpful ;it may show evidence of <u>sagging</u>(figure)



FIGURE 10-5. Intracranial hypotension. Gadolinium enhanced MRI scan of a patient with intracranial hypotension. There is widespread, symmetric meningeal enhancement (arrows).

# The <u>definitive</u> <u>diagnosis</u> is made with an <u>LP</u> showing an opening pressure below 60 mm CSF

If a <u>patient remains symptomatic</u> from intracranial hypotension, a <u>blood patch</u> may be attempted to cover the dural leak( if it can be found)

### 6. <u>Medication</u> causes

### <u>Medications</u> associated with headaches are <u>numerous</u>

They range from <u>hormonal</u> therapies, including <u>contraceptives</u>, to nitric oxide

<u>Withdrawal</u> of a <u>medication</u> or other treatment may also cause a headache; the most common example is a <u>caffeine</u> <u>withdrawal</u> headache

Headache <u>semiology</u> is <u>nonspecific</u>, but the <u>temporal</u> <u>association</u> to medication change <u>helps</u> establish the <u>diagnosis</u>

<u>Medication overuse headache</u> (MOH), also referred to as <u>rebound</u> <u>headache</u> or <u>drug-induced headache</u>, is a chronic headache occurring in patients with a primary headache disorder

The <u>baseline headache</u> disorder is typically markedly <u>exacerbated</u> i.e., more frequent, severe, or both) <u>when</u> the <u>medication</u> is <u>overused</u>

<u>MOH</u> is diagnosed when patients use an <u>abortive therapy 10</u> or <u>more</u> <u>times</u> per <u>month</u> for <u>more than 3 months</u> with an increase in headaches

#### VI) <u>Head</u> and <u>neck</u> <u>disorders</u>

There are many <u>different structural</u> <u>disorders</u> that can cause headache

<u>Treatments</u> are based on the specific causes identified, and these disorders may <u>occur simultaneously with</u> other <u>primary</u> or <u>secondary</u> headache <u>disorders</u>

Sinusitis is commonly associated with a headache

It may be <u>bifrontal</u> or <u>unilateral</u>

<u>Acute</u> <u>sinusitis</u> is often associated with other symptoms of a respiratory tract infection

<u>Temporomandibular joint disorder</u> (TMD) is another common cause of headache and may be unilateral or bilateral

<u>On exam</u>, there may be evidence of <u>dental</u> <u>wearing</u>( chipped and flattened teeth) and <u>discomfort</u> on <u>palpation</u> of the <u>joint</u>

<u>Cervicogenic headaches</u> are also common and may be identified by palpation of <u>myofascial trigger points</u> in the <u>neck</u>

<u>Cervical range</u> of <u>motion</u> is often <u>reduced</u>

#### VII) <u>Trigeminal</u> <u>neuralgia</u>

The <u>pain</u> of trigeminal neuralgia is <u>shock-like</u>, occurring in <u>one</u> or <u>all</u> <u>branches</u> of the <u>trigeminal</u> nerve

The pain is usually <u>paroxysmal</u> and <u>recurrent</u>

It may be triggered by common activities such as brushing hair or teeth

It may be <u>idiopathic</u> or due to <u>structural causes</u> such as a mass or vascular lesion, or a demyelinating lesion of multiple sclerosis

<u>Carbamazepine</u> is a common <u>first-line</u> <u>treatment</u>

## VIII) SUMMARY

<u>Migraines, tension headaches</u>, and <u>TACs</u> are the 3 most common types of <u>primary</u> headache <u>disorders</u>

The International Headache Society maintains an evidence-based categorization of primary and secondary headache disorders

If a <u>headache</u> history and pattern is <u>not consistent</u> with a <u>primary</u> <u>headache</u> disorder, the clinician should <u>consider</u> a <u>secondary headache</u> disorder, <u>review</u> the <u>rare</u> types of <u>primary</u> headache disorders, or consider that the <u>headache</u> may be a <u>combination</u> of <u>more</u> <u>than</u> <u>one</u> headache <u>disorder</u>