Evaluation of headache in adults

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All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: Mar 2021. | This topic last updated: Apr 06, 2020.

INTRODUCTION

Headache is among the most common medical complaints. An overview of the approach to the patient with a chief complaint of headache is presented here. The approach to adults presenting with headache in the emergency department is reviewed elsewhere. (See "Evaluation of the adult with nontraumatic headache in the emergency department".)

The clinical features and diagnosis of specific primary headache syndromes are discussed separately:

· Migraine:

Pathophysiology, clinical manifestations, and diagnosis of migraine in adults

Chronic migraine

Vestibular migraine

Hemiplegic migraine

Migraine with brainstem aura (basilar-type migraine)

• Tension-type headache:

Tension-type headache in adults: Pathophysiology, clinical features, and diagnosis

· Trigeminal autonomic cephalalgias:

Cluster headache: Epidemiology, clinical features, and diagnosis

Paroxysmal hemicrania: Clinical features and diagnosis

Short-lasting unilateral neuralgiform headache attacks: Clinical features and diagnosis

Hemicrania continua

• Other primary headache disorders:

Primary cough headache

Exercise (exertional) headache

Primary headache associated with sexual activity

Cold stimulus headache

Primary stabbing headache

Nummular headache

Hypnic headache

New daily persistent headache

CLASSIFICATION

As many as 90 percent of all primary headaches fall under a few categories, including migraine, tension-type, and cluster headache. While episodic tension-type headache (TTH) is the most frequent headache type in population-based studies, migraine is the most common diagnosis in patients presenting to primary care physicians with headache.

Cluster headache typically leads to significant disability and most of these patients will come to medical attention. However, cluster headache remains an uncommon diagnosis in primary care settings because of overall low prevalence in the general population (<1 percent).

The epidemiology of these headaches is reviewed in detail separately. (See "Tension-type headache in adults: Pathophysiology, clinical features, and diagnosis", section on 'Epidemiology' and "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults", section on 'Epidemiology' and "Cluster headache: Epidemiology, clinical features, and diagnosis", section on 'Epidemiology'.)

Clinicians can easily become familiar with the most common primary headache disorders and how to distinguish them, as summarized in the table (<u>table 1</u>).

Migraine — Migraine is a disorder of recurrent attacks. The headache of migraine is often but not always unilateral and tends to have a throbbing or pulsatile quality. Accompanying features may include nausea, vomiting, photophobia, phonophobia, or osmophobia during attacks (<u>table 2</u>). (See <u>"Pathophysiology, clinical manifestations, and diagnosis of migraine in adults"</u>.)

Migraine trigger factors (table 3) may include stress, menstruation, visual stimuli, weather changes, nitrates, fasting, wine, sleep disturbances, and aspartame, among others. (See "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults", section on 'Precipitating and exacerbating factors'.)

Tension-type headache — The typical presentation of a TTH attack is that of a mild to moderate intensity, bilateral, nonthrobbing headache without other associated features (<u>table 4</u>). Pure TTH is a rather featureless headache. (See <u>"Tension-type headache in adults: Pathophysiology, clinical features, and diagnosis"</u>.)

Cluster headache — Cluster headache belongs to a group of idiopathic headache entities, the trigeminal autonomic cephalalgias (table 5), all of which involve unilateral, often severe headache attacks and typical accompanying autonomic symptoms. Cluster headache is characterized by attacks of severe unilateral orbital, supraorbital, or temporal pain accompanied by autonomic phenomena (table 6). Unilateral autonomic symptoms are ipsilateral to the pain and may include ptosis, miosis, lacrimation, conjunctival injection, rhinorrhea, periorbital edema, facial sweating, and nasal congestion.

Restlessness can also be a typical feature of a cluster headache attack. Attacks usually last 15 to 180 minutes. (See "Cluster headache: Epidemiology, clinical features, and diagnosis".)

Cluster headache may sometimes be confused with a life-threatening headache, since the pain from a cluster headache can reach full intensity within minutes. However, cluster headache is transient, usually lasting less than one to two hours.

Secondary headache — A headache caused by an underlying condition is termed a secondary headache [1]. Clinicians who evaluate patients with headache should be alert to signs that suggest a serious underlying disorder [2]. (See 'Danger signs' below.)

In the Brazilian primary care study, 39 percent of patients presenting with headache had a headache that was due to a systemic disorder (most commonly fever, acute hypertension, and sinusitis), and 5 percent had a headache that was due to a neurologic disorder (most commonly posttraumatic headache, headaches secondary to cervical spine disease, and expansive intracranial processes) [3].

EVALUATION

The appropriate evaluation of headache complaints includes the following:

- Rule out serious underlying pathology and look for other secondary causes of headache (algorithm 1).
- Determine the type of primary headache using the patient history as the primary diagnostic tool (table 1). There may be overlap in symptoms, particularly between migraine and tension-type headache (TTH) and between migraine and some secondary causes of headache such as sinus disease. A headache diary can be helpful in further clarifying the headache diagnosis, the frequency of headache, potential triggers, and the disability from the headache [4].

A systematic case history is the single most important factor in establishing a headache diagnosis and determining the future work-up and treatment plan. Imaging is not necessary in the vast majority of patients presenting with headache. Nevertheless, brain imaging is warranted in the patients with danger signs suggesting a secondary cause of headache. (See 'Indications for imaging' below.)

History and examination — A thorough history can focus the physical examination and determine the need for further investigations and imaging exams. A systematic history should include the following:

- Age at onset
- Presence or absence of aura and prodrome
- · Frequency, intensity, and duration of attack
- Number of headache days per month
- Time and mode of onset
- Quality, site, and radiation of pain
- · Associated symptoms and abnormalities

- · Family history of migraine
- · Precipitating and relieving factors
- · Exacerbation or relief with change in position (eg, lying flat versus upright)
- · Effect of activity on pain
- · Relationship with food/alcohol
- · Response to any previous treatment
- · Review of current medications
- · Any recent change in vision
- · Association with recent trauma
- · Any recent changes in sleep, exercise, weight, or diet
- · State of general health
- · Change in work or lifestyle (disability)
- · Change in method of birth control (women)
- · Possible association with environmental factors
- Effects of menstrual cycle and exogenous hormones (women)

The examination of an adult with headache complaints should cover the following areas:

- · Obtain blood pressure and pulse
- · Listen for bruit at neck, eyes, and head for clinical signs of arteriovenous malformation
- · Palpate the head, neck, and shoulder regions
- · Check temporal and neck arteries
- · Examine the spine and neck muscles

The neurologic examination should cover mental status testing, cranial nerve examination, funduscopy and otoscopy, and symmetry on motor, reflex, cerebellar (coordination), and sensory tests. Gait examination should include getting up from a seated position without any support and walking on tiptoes and heels, tandem gait, and Romberg test.

Low-risk features — The following features can serve as indicators of patients who are unlikely to have serious underlying cause for headache [5,6]:

- Age ≤50 years
- Features typical of primary headaches (table 1)
- · History of similar headache
- · No abnormal neurologic findings
- · No concerning change in usual headache pattern
- No high-risk comorbid conditions
- No new or concerning findings on history or examination

Patients with headache who meet these criteria do not require imaging.

The majority of patients with headache complaints have a completely normal physical and neurologic examination. However, some types of primary headache may be associated with specific abnormalities:

- With TTH, there may be pericranial muscle tenderness.
- With migraine, there may be manifestations related to sensitization of primary nociceptors and central trigeminovascular neurons, such as hyperalgesia and allodynia.
- With hemicrania continua or one of the other trigeminal autonomic cephalalgias (cluster headache, paroxysmal hemicrania, and short-lasting unilateral neuralgiform headache attacks), there may be evidence of autonomic activation.

Other abnormalities on examination should raise suspicion for a secondary headache disorder. Likewise, danger signs (ie, red flags) should prompt further evaluation, as discussed in the sections below. (See '<u>Danger signs'</u> below.)

Features suggesting migraine — The most common headache syndromes frequently present with characteristic symptoms (table 1). However, there may be considerable symptom overlap; one population-based survey found that less than one-half of patients who complained of headaches that met criteria for migraine were properly diagnosed [7]. Migraine symptoms may also overlap with other causes of headache. As an example, a significant number of patients with migraine may have nasal symptoms that suggest sinus disease [8]; in addition, a study of primary care patients with recurrent sinus headache found that 90 percent experienced attacks that met the International Headache Society criteria for migraine [9]. (See 'Sinus symptoms' below.)

Given these pitfalls, a number of diagnostic instruments have been proposed, mainly to assist with the diagnosis of migraine, the most common primary headache syndrome in patients presenting to primary care physicians. One such instrument (ID Migraine) preselects eligible subjects as those who had two or more headaches in the previous three months and indicated either that they might want to speak with a health care professional about their headaches or that they experienced a headache that limited their ability to work, study, or enjoy life [10]. The screen employs three questions:

During the last three months, did you have the following with your headaches?

- Photophobia Did light bother you (a lot more than when you do not have headaches)?
- Incapacity Did your headaches limit your ability to work, study, or do what you needed to do for at least one day?
- Nausea Did you feel nauseated or sick to your stomach?

The mnemonic **PIN** is a reminder of the questions used in the ID Migraine screen that can help identify migraine. The ID Migraine screen is positive if the patient answers "yes" to two of the three items. In a systematic review of 13 studies that involved over 5800 patients, the pooled sensitivity and specificity of ID Migraine was 0.84 and 0.76, respectively [11]. A positive ID Migraine increased the pretest probability of migraine from 59 to 84 percent, whereas a negative ID Migraine score reduced the probability of migraine from 59 to 23 percent.

Another simple and validated instrument, the brief headache screen, consists of three to six questions [12]. One version includes the following four questions:

- How often do you get severe headaches (ie, without treatment it is difficult to function)?
- How often do you get other (milder) headaches?
- How often do you take headache relievers or pain pills?
- Has there been any recent change in your headaches?

In one study, the presence of episodic disabling headache correctly identified migraine in 136 of 146 patients (93 percent) with episodic migraine, and 154 of 197 patients (78 percent) with chronic headache with migraine, with a specificity of 63 percent [12]. Only 6 of 343 patients (2 percent) with migraine were not identified by disabling headache. Thus, virtually any patient with severe episodic headaches can be considered to have migraine.

Among the questions above, the second on frequency of headache and the third on the need for pain pills may be helpful for identifying patients with medication overuse (eg, patients who use symptomatic medications more than three days per week and/or who have daily headaches). The last question about recent changes in the headache is particularly helpful for identifying patients who may have an important secondary cause of headache; a patient with a stable pattern of headache for six months is unlikely to have a serious underlying cause.

Danger signs — Paying attention to danger signs is important since headaches may be the presenting symptom of a space-occupying mass or vascular lesion, infection, metabolic disturbance, or a systemic problem. The following features in the history can serve as warning signs of possible serious underlying disease [13-15]. (See "Evaluation of the adult with nontraumatic headache in the emergency department".)

The mnemonic **SNNOOP10** is a reminder of the danger signs ("red flags") for the presence of serious underlying disorders that can cause acute or subacute headache [16]:

- Systemic symptoms including fever
- Neoplasm history
- Neurologic deficit (including decreased consciousness)
- Onset is sudden or abrupt
- Older age (onset after age 50 years)
- Pattern change or recent onset of new headache
- Positional headache
- Precipitated by sneezing, coughing, or exercise
- Papilledema
- Progressive headache and atypical presentations
- Pregnancy or puerperium
- Painful eye with autonomic features
- Post-traumatic onset of headache
- · Pathology of the immune system such as HIV
- Painkiller (analgesic) overuse (eg, medication overuse headache) or new drug at onset of headache

Any of these findings should prompt further investigation, including brain imaging with magnetic resonance imaging (MRI) or computed tomography (CT).

Specific features suggesting a secondary headache source — Other features that suggest a specific source of headache pain include the following:

- Strictly unilateral pain that does not switch sides (ie, side-locked pain) is associated with an increased likelihood of secondary headache disorders (especially cervicogenic headache and post-traumatic headache), although only a minority may be related to a serious underlying disease (eg, intracranial neoplasm, cervical arterial dissection, giant cell arteritis, cerebral venous sinus thrombosis) [17]. Thus, further evaluation should be pursued in patients presenting with side-locked headache.
- Impaired vision or seeing halos around light suggests the presence of glaucoma. Suspicion for subacute angle closure glaucoma should be raised by
 relatively short duration (often less than one hour) unilateral headaches that do not meet criteria for migraine arising after age 50 [18]. Acute myopia
 and secondary angle closure glaucoma are rare adverse effects of topiramate (often used to treat migraine), typically within one month of starting
 treatment.
- · Visual field defects suggest the presence of a lesion of the optic pathway (eg, due to a pituitary mass).
- Sudden, severe, unilateral vision loss suggests the presence of optic neuritis. Optic neuritis typically presents with painful, monocular visual loss that evolves over several hours to a few days. One-third of patients have visible optic nerve inflammation (papillitis) on funduscopic examination. (See "Optic neuritis: Pathophysiology, clinical features, and diagnosis".)
- Blurring of vision on forward bending of the head, headaches upon waking early in the morning that improve with sitting up, and double vision or loss
 of coordination and balance should raise the suspicion of raised intracranial pressure (ICP); this should also be considered in patients with chronic,
 daily, progressively worsening headaches associated with chronic nausea. Idiopathic intracranial hypertension (pseudotumor cerebri) typically affects
 obese women of childbearing age. Characteristic features are headache, papilledema, vision loss or diplopia, elevated lumbar puncture (LP) opening
 pressure with normal cerebrospinal fluid (CSF) composition. (See "Evaluation and management of elevated intracranial pressure in adults" and
 "Idiopathic intracranial hypertension (pseudotumor cerebri): Clinical features and diagnosis".)
- In patients who present with headache that is relieved with recumbency and exacerbated with upright posture, the diagnosis of headache attributed to spontaneous intracranial hypotension, or to spontaneous spinal cerebrospinal fluid (CSF) leak with normal CSF pressure [19], should be considered. An additional major feature of this headache syndrome is diffuse, pachymeningeal enhancement on brain MRI. The accepted etiology is CSF leakage, which may occur in the context of disruption of the meninges. (See "Spontaneous intracranial hypotension: Pathophysiology, clinical features, and diagnosis".)
- The presence of nausea, vomiting, worsening of headache with changes in body position (particularly bending over), a focal neurologic deficit, papilledema, new-onset seizure, and/or a significant change in prior headache pattern suggests a brain tumor as a possible cause. The features of brain tumor headache are generally nonspecific and vary widely with tumor location, size, and rate of growth. Brain tumor headache may resemble tension-type headache, migraine, or a variety of other headache types. (See "Brain tumor headache" and "Overview of the clinical features and diagnosis of brain tumors in adults".)
- Intermittent headache with generalized sweating, tachycardia, and/or sustained or paroxysmal hypertension is suggestive of pheochromocytoma. (See "Clinical presentation and diagnosis of pheochromocytoma".)
- Morning headache is nonspecific and can occur as part of a primary headache syndrome or may be secondary to a number of disorders including
 sleep apnea, sleep-related bruxism, chronic obstructive pulmonary disease, caffeine withdrawal, medication overuse headache, and the obesityhypoventilation syndrome. (See "Clinical presentation and diagnosis of obstructive sleep apnea in adults" and "Chronic obstructive pulmonary
 disease: Definition, clinical manifestations, diagnosis, and staging" and "Clinical manifestations and diagnosis of obesity hypoventilation syndrome".)

Need for emergency evaluation — A small proportion of patients present with serious or life-threatening headaches that require referral for emergency diagnosis and treatment. These include:

- Sudden onset "thunderclap" headache Severe headache of sudden onset (ie, that reaches maximal intensity within a few seconds or less than one minute after the onset of pain) is known as thunderclap headache because its explosive and unexpected nature is likened to a "clap of thunder." Thunderclap headache requires urgent evaluation as such headaches may be harbingers of subarachnoid hemorrhage and other potentially ominous etiologies (<u>table 7</u>). (See "Overview of thunderclap headache".)
- Acute or subacute neck pain or headache with Horner syndrome and/or neurologic deficit Cervical artery dissection is usually associated
 with local symptoms including neck pain or headache, and often results in ischemic stroke or transient ischemic attack. Horner syndrome is seen in
 approximately 39 percent of those with carotid and 13 percent of those with vertebral artery dissection [20,21].
- Headache with suspected meningitis or encephalitis Fever, altered mental status, with or without nuchal rigidity may indicate central nervous system infection.
- Headache with global or focal neurologic deficit or papilledema Headache is the primary symptom of increased ICP, which should be suspected when accompanied by bilateral papilledema, focal neurologic deficit, or repeated episodes of nausea and vomiting.

- Headache with orbital or periorbital symptoms Headache with visual impairment, periorbital pain, or ophthalmoplegia could indicate acute angle closure glaucoma, infection, inflammation, vascular congestion from a cavernous sinus thrombosis or draining arteriovenous malformation, or tumor involving the orbits.
- Headache and possible carbon monoxide exposure Headache is a nonspecific symptom of carbon monoxide exposure; the intensity varies with the carbon monoxide level [1]. The headache tends to be bilateral and mild at low levels of carbon monoxide, pulsating at levels of 20 to 30 percent, and severe with nausea, vomiting, and blurred vision at levels of 30 to 40 percent.

The evaluation of the adult with headache in the emergency department is described elsewhere. Laboratory tests, imaging, and LP for CSF analysis may be included in the evaluation. (See "Evaluation of the adult with nontraumatic headache in the emergency department".)

Imaging — CT or MRI are the common modalities used to diagnose many causes of secondary headache. Choice of exact body part (eg, head, neck, face) and use of contrast varies with clinical scenario.

Indications for imaging — Patients with the danger signs or other features suggesting a secondary headache source will require imaging. (See 'Danger signs' above and 'Specific features suggesting a secondary headache source' above.)

Imaging is usually not warranted for patients with a stable migraine pattern and a normal neurologic examination, although a lower threshold for imaging is reasonable for patients with atypical migraine features or in patients who do not fulfill the strict definition of migraine [22]. As an example, imaging is indicated for patients presenting with recent-onset headache that is featureless (ie, bilateral, non-throbbing, without nausea and without sensitivity to light, sound, or smell) [23,24]. However, imaging for no other reason than reassurance is sometimes performed in clinical practice. It is important that the clinician provide the patient with a clear explanation of both the diagnosis and the reason for imaging, especially if it is being performed in someone suspected of having primary headache [23]. The patient should also be informed that incidental findings (eg, vascular lesion, small neoplasm) likely unrelated to the headache can be seen in 1 to 2 percent of MRI exams and that there are few data providing guidance as to how they should be managed [25,26]. In a population-based imaging study of 864 adults, major intracranial abnormalities were not more likely among subjects with headache compared with headache-free individuals [27].

The vast majority of patients without danger signs do not have a secondary cause of headache [28,29]. As an example, in a study of 373 patients with chronic headache at a tertiary referral center, all had one or more of the following characteristics that prompted referral for head CT scan: increased severity of symptoms or resistance to appropriate drug therapy, change in characteristics or pattern of headache, or family history of an intracranial structural lesion [30]. Only two exams (less than 1 percent) showed potentially significant lesions (one low-grade glioma and one aneurysm); only the aneurysm was treated.

Choice of imaging exam — The choice of imaging modality and need for intravenous (IV) contrast depends upon the clinical setting and indications [31].

- Emergency settings In emergency settings, CT has several advantages compared with MRI:
 - CT is widely available in most hospitals and takes only a few minutes to perform, a practical advantage in busy emergency departments.
 - Most life-threatening causes of headache, such as intracranial hemorrhage and tumor, are easily detected on CT as an initial screening examination in emergency settings. CT is highly sensitive for hemorrhage, the signs of which are usually evident to the general radiologist and clinician, whereas imaging signs of subtle hemorrhage may be difficult to appreciate on MRI.
 - CT is safer than MRI for unstable patients who require monitoring and/or life-support while in the radiology department.

With newer techniques, the radiation exposure from CT has been minimized for most adult and pediatric patients.

MRI as an initial examination is usually reserved for new headache with optic disc edema or trigeminal pain, chronic headache with new features, or headache in the context of red flags (eg, known or suspected cancer, subacute head trauma, neurologic deficit, immunosuppressed state, pregnancy), keeping in mind that CT remains an alternative when MRI is unavailable.

• Non-emergency settings – Given that most headaches are benign, MRI is usually reserved as a non-emergent elective study. MRI of the head is more sensitive than CT for most secondary causes of headache and does not result in radiation exposure [32]. One of the expert consensus recommendations from the "Choosing Wisely" initiative is that clinicians should not order CT imaging for headache when MRI is available, except in emergency settings [33].

The <u>ACR Appropriateness Criteria</u> provides general guidance for many common clinical scenarios of headache [34]. When the decision is not obvious, consultation with the radiologist is helpful to facilitate patient referral. For imaging of the vessels, cerebral and cervical angiography using computed tomography (CTA) or magnetic resonance angiography (MRA) is performed as an added exam to MRI (or CT) and usually requires IV contrast administration. MRA and CTA image the arteries, veins, or both, depending on the indication. Exams tailored for imaging the orbits and ear (encompassing the skull base and pituitary), face, and maxilla (encompassing the paranasal sinuses), or the temporomandibular joint are sometimes

added to the head imaging if an underlying diagnosis that localizes anatomically is suspected. The approximate effective radiation dose for a head CT is 2 millisievert (mSv).

Lumbar puncture — LP for CSF analysis is urgently indicated in patients with headache when there is clinical suspicion of subarachnoid hemorrhage in the setting of a negative or normal head CT. In addition, LP is indicated when there is clinical suspicion of an infectious, inflammatory, or neoplastic etiology of headache. An LP is also necessary in cases of suspected idiopathic intracranial hypertension (pseudotumor cerebri), but is generally not necessary in cases where MRI is consistent with the diagnosis of spontaneous spinal cerebrospinal fluid (CSF) leak with normal CSF pressure or spontaneous intracranial hypotension due to a CSF leak. These issues are discussed in detail elsewhere. (See "Aneurysmal subarachnoid hemorrhage: Clinical manifestations and diagnosis", section on 'Evaluation and diagnosis' and "Lumbar puncture: Technique, indications, contraindications, and complications in adults", section on 'Indications' and "Idiopathic intracranial hypertension (pseudotumor cerebri): Clinical features and diagnosis", section on 'Lumbar puncture'.)

Common clinical scenarios — Patients with a chief complaint of headache accompanied by factors that suggest a serious but **not** immediately life-threatening diagnosis should be evaluated promptly in the outpatient or inpatient setting. Differences in patient demographics, comorbidities, and headache features can guide the evaluation to help ensure appropriate diagnosis and management. (See <u>'Danger signs'</u> above and <u>'Specific features suggesting a secondary headache source'</u> above.)

New or recent onset headache — The absence of similar headaches in the past, when combined with high-risk features, suggests a possible serious disorder. Head MRI without and with contrast should be obtained to evaluate for an intracranial mass lesion (eg, primary or metastatic neoplasm, abscess, hematoma), communicating or obstructive hydrocephalus, or cerebral edema from ischemia or infarction (ie, stroke). If MRI is not available or contraindicated, head CT without and with contrast should be performed instead.

In patients with a new or recent onset of headache, high-risk features include:

- Older age New headache in patients older than 50 years may suggest underlying pathology.
- Cancer New headache type in a patient with cancer suggests metastasis. (See "Evaluation of the adult with nontraumatic headache in the emergency department", section on 'New headache in a cancer patient'.)
- Febrile or with Lyme disease New headache associated with fever and altered mental status with or without nuchal rigidity can indicate meningitis. New headache in a patient with Lyme disease suggests meningoencephalitis. (See "Evaluation of the adult with nontraumatic headache in the emergency department", section on 'New headache with suspected meningitis or encephalitis' and "Nervous system Lyme disease", section on 'Lyme encephalomyelitis'.)
- Immunosuppression New headache type in a patient with immunosuppression suggests an opportunistic infection or tumor.

Brain tumor is a rare cause of headache but should be considered in patients presenting with a change in headache pattern, focal neurologic signs, papilledema, or seizures, particularly when new-onset headaches occur in adults older than 50 years. (See "Brain tumor headache".)

In the absence of danger signs, patients who present with a new or recent onset headache and a normal neurologic examination are most likely to have primary headache, such as migraine or TTH (table 1).

Older patients — Older patients are at increased risk for secondary types of headache (eg, giant cell arteritis, trigeminal neuralgia, subdural hematoma, acute herpes zoster and postherpetic neuralgia, and brain tumors) and some types of primary headache (eg, hypnic headache, cough headache, and migraine accompaniments) [35]. Need for imaging depends on the suspected diagnosis. Diagnostic consideration include:

• Giant cell (temporal) arteritis (GCA) is a chronic vasculitis of large and medium sized vessels. The disease seldom occurs before age 50 years, and its incidence rises steadily thereafter. A new type of headache occurs in two-thirds of affected individuals. The head pain tends to be located over the temporal areas but can be frontal or occipital in location. The headaches may be mild or severe. Other common symptoms can include fever, fatigue, weight loss, jaw claudication, visual symptoms, particularly transient monocular visual loss and diplopia, and symptoms of polymyalgia rheumatica. Laboratory testing may reveal an elevated erythrocyte sedimentation rate and/or serum C-reactive protein, or thrombocytosis, but these are not specific.

The diagnosis of GCA is based on histopathology or imaging exams. Histopathologic evidence of GCA is most often acquired by temporal artery biopsy. Color Doppler ultrasound (CDUS) of the head, as performed by experienced operators, is an alternative diagnostic procedure. CDUS can visualize temporal artery abnormalities (eg, mural edema as shown by the "halo sign" and "compression sign") characteristic of GCA. When the diagnosis of GCA is still suspected in a patient who has had a negative temporal artery biopsy and/or CDUS, the possibility of large vessel involvement can be evaluated by imaging the torso with CT/CTA, MRI/MRA, or positron emission tomography (PET). (See "Clinical manifestations of giant cell arteritis" and "Diagnosis of giant cell arteritis".)

- Trigeminal neuralgia is defined by sudden, usually unilateral, severe, brief, stabbing or lancinating, recurrent episodes of pain in the distribution of one or more branches of the fifth cranial (trigeminal) nerve. The incidence increases gradually with age; most idiopathic cases begin after age 50 years. Once the diagnosis is suspected on clinical grounds, it is important to search for secondary causes. Patients with suspected trigeminal neuralgia or those with recurrent attacks of pain limited to one or more divisions of the trigeminal nerve and no obvious cause (eg, herpes zoster or trigeminal nerve trauma) should undergo imaging to help distinguish classic trigeminal neuralgia from secondary causes. MRI and MRA of the head without and with contrast tailored to evaluate the trigeminal nerve is the preferred imaging exam to evaluate for compression of the nerve by adjacent vessels or other structures. (See "Trigeminal neuralgia".)
- Chronic subdural hematoma may present with the insidious onset of headaches, light-headedness, cognitive impairment, apathy, somnolence, and occasionally seizures. Imaging with noncontrast CT or MRI is essential to confirm the diagnosis. (See "Subdural hematoma in adults: Etiology, clinical features, and diagnosis".)
- Acute herpes zoster and postherpetic neuralgia often involve cervical and trigeminal nerves. Pain is the most common symptom of zoster and approximately 75 percent of patients have prodromal pain in the dermatome where the rash subsequently appears. The major risk factors for postherpetic neuralgia are older age, greater acute pain, and greater rash severity. Acute herpes zoster is usually a clinical diagnosis based upon the characteristic vesicular lesions in a restricted dermatomal pattern. The diagnosis of postherpetic neuralgia is made when pain persists beyond four months in the same distribution as a preceding documented episode of acute herpes zoster. (See "Epidemiology, clinical manifestations, and diagnosis of herpes zoster" and "Postherpetic neuralgia".)
- Brain tumor should be considered as a possible cause of new-onset headaches in adults over age 50 years, as discussed above. (See 'New or recent onset headache' above and "Brain tumor headache".)
- Hypnic headache, also known as "alarm clock headache," occurs almost exclusively after the age of 50 years and is characterized by episodes of dull head pain, often bilateral, that awaken the sufferer from sleep. The diagnosis requires the exclusion of nocturnal attacks caused by other primary and secondary headaches. Therefore, imaging of the brain, preferably by MRI without and with contrast, should be obtained to look for a structural cause. (See "Hypnic headache".)
- **Primary cough headache** most often affects people older than age 40 years and is provoked by coughing or straining in the absence of any intracranial disorder. Patients presenting with de novo headache precipitated by coughing should have imaging, preferably brain MRI without and with contrast, to exclude a structural lesion. (See <u>"Primary cough headache"</u>.)

Pregnancy — New headache or change in headache during pregnancy may be due to migraine or another primary headache, but many other conditions can present with headache at this time, particularly pre-eclampsia, postdural puncture headache, and cerebral venous thrombosis. Pre-eclampsia must be ruled in or out in every pregnant woman over 20 weeks of gestation with headache. (See <u>"Preeclampsia: Clinical features and diagnosis"</u>.)

MRI without contrast is recommended when there is concern for a secondary headache, and MR venography without contrast should be included if cerebral venous sinus thrombosis is a concern. If MRI is not immediately available or contraindicated, head CT without and with contrast can be used to evaluate for hemorrhage, mass effect or hydrocephalus. (See "Headache in pregnant and postpartum women".)

Fever — Fever associated with headache may be caused by intracranial, systemic, or local infection, as well as other etiologies (table 8).

Emergency evaluation is indicated if fever is accompanied by symptoms suggestive of meningitis or encephalitis (eg, altered mental status, with or without nuchal rigidity). (See "Evaluation of the adult with nontraumatic headache in the emergency department", section on 'New headache with suspected meningitis or encephalitis'.)

Oromaxillofacial symptoms — TMJ disorders, trigeminal neuralgia and odontogenic conditions (eg, tooth impaction, dental abscess) may present as headache with facial pain. Imaging tailored for the anatomic site of concern is usually indicated. (See "Overview of craniofacial pain" and "Temporomandibular disorders in adults" and "Trigeminal neuralgia" and "Complications, diagnosis, and treatment of odontogenic infections".)

Head injury — Approach to diagnostic evaluation and imaging of headache following trauma and is discussed elsewhere. (See <u>"Postconcussion syndrome"</u>, <u>section on 'Headaches'</u> and <u>"Acute mild traumatic brain injury (concussion) in adults"</u>, <u>section on 'Imaging'</u>.)

Headache is variably estimated as occurring in 25 to 78 percent of persons following mild traumatic brain injury. Paradoxically, headache prevalence, duration, and severity is greater in those with mild head injury compared with those with more severe trauma. Most often, the headache following head trauma can be clinically classified similarly to nontraumatic headaches; migraine and TTH predominate.

Sinus symptoms — Although sinus headache is commonly diagnosed by clinicians and self-diagnosed by patients, acute or chronic sinusitis appears to be an uncommon cause of recurrent headaches [36-38].

Headache of sinonasal origin usually does not require imaging for diagnosis as it is best evaluated with nasal endoscopy. If intracranial complications of sinus disease is suspected, head MRI without and with contrast is indicated. (See "Acute sinusitis and rhinosinusitis in adults: Clinical manifestations and

diagnosis".)

Autonomic features characteristically occur in trigeminal autonomic cephalalgias such as cluster headaches and are also common with migraine headache. These symptoms may include nasal congestion, rhinorrhea, tearing, color and temperature change, and changes in pupil size. (See "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults".)

The prominence of sinus symptoms often leads to the misdiagnosis of "sinus headache" in patients who meet diagnostic criteria for migraine or, less often, TTH. This point is illustrated by an observational study that enrolled 2991 patients with a history of clinician- or self-diagnosed sinus headache and no previous history of migraine; 88 percent of these patients fulfilled criteria for migraine or migrainous headache, and 8 percent fulfilled criteria for TTH [39]. In the patients with migraine or migrainous headache, sinus pain, pressure, and congestion commonly occurred in association with typical migraine features such as pulsing head pain and sensitivity to activity, light, and sound (as depicted in Figure 2 of the observational study).

Pain related purely to sinus conditions may have some features that aid in distinguishing it from migraine [40,41]. Sinus-related pain or headache is typically described as a pressure-like or dull sensation that is usually bilateral and periorbital. However, it can be unilateral with deviated septum, middle or inferior turbinate hypertrophy, or unilateral sinus disease. In addition, sinus-related pain is typically associated with nasal obstruction or congestion, lasts for days at a time, and is usually not associated with nausea, vomiting, photophobia, or phonophobia. (See "Acute sinusitis and rhinosinusitis in adults: Clinical manifestations and diagnosis".)

The severity, extent, and location of sinus-related pain do not correlate with the extent or location of mucosal disease as revealed by imaging [41].

In general, the following principles apply to the relationship of rhinosinusitis and headache [40,42,43]:

- A stable pattern of recurrent headaches that interfere with daily function is most likely migraine.
- Recurrent self-limited headaches associated with rhinogenic symptoms are most likely migraine.
- Prominent rhinogenic symptoms with headache as one of several symptoms should be evaluated carefully for otolaryngologic conditions.
- · Headache associated with fever and purulent nasal discharge is likely rhinogenic in origin.

Chronic headache — Chronic daily headache is not a specific headache type, but a syndrome that encompasses several primary and secondary headaches. The term "chronic" refers either to the frequency of headaches or to the duration of the disease, depending upon the specific headache type. (See "Overview of chronic daily headache".)

In adults with chronic recurrent headaches, including those with migraine aura, with no recent change in headache pattern, no history of seizures, and no other focal neurologic signs or symptoms, the routine use of imaging is not warranted. The yield of head CT or MRI in identifying potentially treatable lesions is <1 percent [44]. However, imaging to exclude a secondary cause of headache is indicated in the initial evaluation of patients presenting with hemicrania continua, new daily persistent headache, cluster headache, paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks, and hypnic headache, described below.

With headache subtypes of long duration (ie, four hours or more), "chronic" indicates a headache frequency of 15 or more days a month for longer than three months in the absence of organic pathology. These headache subtypes are:

- Chronic migraine headache (see "Chronic migraine")
- Chronic TTH (see "Tension-type headache in adults: Pathophysiology, clinical features, and diagnosis")
- Medication overuse headache, which is typically preceded by an episodic headache disorder (usually migraine or TTH) that has been treated with
 frequent and excessive amounts of acute symptomatic medications (see "Medication overuse headache: Etiology, clinical features, and diagnosis")
- Hemicrania continua, a strictly unilateral, continuous headache with superimposed exacerbations of moderate to severe intensity accompanied by autonomic features and sometimes by migrainous symptoms (see "Hemicrania continua")
- New daily persistent headache, characterized by headache that begins rather abruptly and is daily and unremitting from onset or within three days of onset at most, typically in individuals without a prior headache history (see "New daily persistent headache")

With headache subtypes of shorter duration (ie, less than four hours), "chronic" refers to a prolonged duration of the condition itself without remission. The headache subtypes in this category are the following:

- Chronic cluster headache (see "Cluster headache: Epidemiology, clinical features, and diagnosis")
- Chronic paroxysmal hemicrania, characterized by unilateral, brief, severe attacks of pain associated with cranial autonomic features that recur several times per day with individual headache attacks that usually last 2 to 30 minutes (see "Paroxysmal hemicrania: Clinical features and diagnosis")
- Short-lasting unilateral neuralgiform headache attacks, characterized by sudden brief attacks of severe unilateral head pain in orbital, peri-orbital, or temporal regions, accompanied by ipsilateral cranial autonomic symptoms (see <u>"Short-lasting unilateral neuralgiform headache attacks: Clinical features and diagnosis"</u>)
- Hypnic headache, also known as "alarm clock headache," which occurs almost exclusively after the age of 50 years and is characterized by episodes
 of dull head pain, often bilateral, that awaken the sufferer from sleep (see "Hypnic headache")

• Primary stabbing headache, characterized by sudden brief attacks of sharp, jabbing head pain in orbital, peri-orbital, or temporal regions (see "Primary stabbing headache")

Misconceptions — Several misconceptions may hinder headache evaluation and diagnosis.

- Although sinus headache is commonly diagnosed by clinicians and self-diagnosed by patients, acute or chronic sinusitis appears to be an uncommon cause of recurrent headaches, and many patients presenting with sinus headache turn out to have migraine [36-38]. (See 'Sinus symptoms' above.)
- Patients frequently attribute headaches to eye strain. However, an observational study suggested that headaches are only rarely due to refractive error alone [45]. Nevertheless, correcting vision may improve headache symptoms in some of patients with headache and refractive error.
- Some patients believe that hypertension is the cause of their headaches. While this can be true in the case of a hypertensive emergency, it is probably not true for typical migraine or TTH. As an example, a report from the Physicians' Health Study of 22,701 American male physicians ages 40 to 84 years analyzed various risk factors for cerebrovascular disease and found no difference in the percentage of men with a history of hypertension in the migraine and nonmigraine groups [46]. Furthermore, a prospective study of 22,685 adults in Norway found that high systolic and diastolic blood pressures were actually associated with a reduced risk of nonmigrainous headache [47].

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Migraine and other primary headache disorders".)

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

- Basics topic (see "Patient education: Headaches in adults (The Basics)")
- Beyond the Basics topics (see "Patient education: Headache causes and diagnosis in adults (Beyond the Basics)" and "Patient education: Headache treatment in adults (Beyond the Basics)")

SUMMARY AND RECOMMENDATIONS

• While episodic tension-type headache (TTH) is the most frequent headache type in population-based studies, migraine is the most common diagnosis in patients presenting to medical attention with headache. Clinicians can easily become familiar with the most common primary headache disorders and how to distinguish them (table 1). (See 'Classification' above.)

Using the patient history as the primary diagnostic tool, the initial headache evaluation (<u>algorithm 1</u>) should determine whether the headache is primary or secondary and, if the latter, whether the underlying cause is serious. (See <u>'Evaluation'</u> above.)

- · The following features can serve as indicators of patients who are unlikely to have serious underlying cause for headache:
 - Age ≤50 years
 - · Features typical of primary headaches
 - · History of similar headache
 - · No abnormal neurologic findings
 - · No concerning change in usual headache pattern
 - · No high-risk comorbid conditions
 - · No new or concerning findings on history or examination

Patients with headache who meet these criteria do not require imaging. (See 'Low-risk features' above.)

- The mnemonic **SNNOOP10** is a reminder of the danger signs ("red flags") for the presence of serious underlying disorders that can cause acute or subacute headache:
 - · Systemic symptoms including fever
 - · Neoplasm history
 - · Neurologic deficit (including decreased consciousness)
 - · Onset is sudden abrupt onset
 - Older age (onset after age 50 years)
 - Pattern change or recent onset of new headache
 - · Positional headache
 - Precipitated by sneezing, coughing, or exercise
 - Papilledema
 - · Progressive headache and atypical presentations
 - Pregnancy or puerperium
 - Painful eye with autonomic features
 - Post-traumatic onset of headache
 - · Pathology of the immune system such as HIV
 - Painkiller (analgesic) overuse (eg, medication overuse headache

Patients with danger signs or other features of a secondary headache source require prompt evaluation and imaging (<u>algorithm 1</u>). (See <u>'Danger signs'</u> above and <u>'Specific features suggesting a secondary headache source'</u> above and <u>'Common clinical scenarios'</u> above.)

Emergency diagnosis and treatment is indicated for sudden onset of severe headache (ie, thunderclap headache) and for new headache with suspected meningitis or encephalitis, with neck pain with Horner syndrome suggesting cervical artery dissection, with focal neurologic deficit or papilledema suggesting increased intracranial pressure (ICP), or with orbital or periorbital symptoms (<u>algorithm 1</u>). (See <u>'Need for emergency evaluation'</u> above.)

• Computed tomography (CT) or magnetic resonance imaging (MRI) of the head is the preferred imaging exam for headache. Choice of modality and need for intravenous (IV) contrast depends on the clinical indications (see 'Common clinical scenarios' above). For imaging of the vessels, cerebral and cervical angiography using computed tomography (CTA) or magnetic resonance angiography (MRA) is performed as an added exam to head CT or MRI and usually requires IV contrast administration. (See 'Imaging' above.)

ACKNOWLEDGMENT

The editorial staff at UpToDate would like to acknowledge Zahid H Bajwa, MD, who contributed to an earlier version of this topic review.

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Topic 3349 Version 33.0

GRAPHICS

Characteristics of migraine, tension-type, and cluster headache syndromes

Symptom	Migraine	Tension-type	Cluster
Location	Adults: Unilateral in 60 to 70%, bifrontal or global in 30% Children and adolescents: Bilateral in majority	Bilateral	Always unilateral, usually begins around the eye or temple
Characteristics	Gradual in onset, crescendo pattern; pulsating; moderate or severe intensity; aggravated by routine physical activity	Pressure or tightness which waxes and wanes	Pain begins quickly, reaches a crescendo within minutes; pain is deep, continuous, excruciating, and explosive in quality
Patient appearance	Patient prefers to rest in a dark, quiet room	Patient may remain active or may need to rest	Patient remains active
Duration	4 to 72 hours	30 minutes to 7 days	15 minutes to 3 hours
Associated symptoms	Nausea, vomiting, photophobia, phonophobia; may have aura (usually visual, but can involve other senses or cause speech or motor deficits)	None	Ipsilateral lacrimation and redness of the eye; stuffy nose; rhinorrhea; pallor; sweating; Horner syndrome; restlessness or agitation; focal neurologic symptoms rare; sensitivity to alcohol

Graphic 68064 Version 7.0

Diagnostic criteria for migraine

Migraine without aura A. At least five attacks fulfilling criteria B through D B. Headache attacks lasting 4 to 72 hours (untreated or unsuccessfully treated) C. Headache has at least two of the following characteristics: Unilateral location Pulsating quality Moderate or severe pain intensity $\label{prop:continuous} \mbox{Aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)}$ D. During headache at least one of the following: Nausea, vomiting, or both Photophobia and phonophobia E. Not better accounted for by another ICHD-3 diagnosis Migraine with aura A. At least two attacks fulfilling criteria B and C B. One or more of the following fully reversible aura symptoms: Speech and/or language Motor Brainstem C. At least three of the following six characteristics: At least one aura symptom spreads gradually over ≥ 5 minutes Two or more symptoms occur in succession Each individual aura symptom lasts 5 to 60 minutes At least one aura symptom is unilateral At least one aura symptom is positive* The aura is accompanied or followed within 60 minutes by headache D. Not better accounted for by another ICHD-3 diagnosis Features of migraine in children and adolescents Attacks may last 2 to 72 hours ¶ Headache is more often bilateral than in adults; an adult pattern of unilateral pain usually emerges in late adolescence or early adulthood Photophobia and phonophobia may be inferred by behavior in young children

ICHD-3: International Classification of Headache Disorders, 3rd edition.

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Graphic 50876 Version 15.0

^{*} Scintillations and pins and needles are examples of positive symptoms.

[¶] The evidence for untreated durations of less than 2 hours in children has not been substantiated.

Headache triggers

Diet	Stress		
Alcohol	Let-down periods		
Chocolate	Times of intense activity		
Aged cheeses	Loss or change (death, separation, divorce, job change)		
Monosodium glutamate	Moving		
Aspartame	Crisis		
Caffeine	Changes of environment or habits		
Nuts	Weather		
Nitrites, nitrates	Travel (crossing time zones)		
Hormones	Seasons		
Menses	Altitude		
Ovulation	Schedule changes		
Hormone replacement (progesterone)	Sleeping patterns		
Sensory stimuli	Dieting		
Strong light	Skipping meals		
Flickering lights	Irregular physical activity		
Odors			
Sounds, noise			

Graphic 57424 Version 4.0

Episodic tension-type headache diagnostic criteria

No more than one of photophobia or phonophobia. **E.** Not better accounted for by another ICHD-3 diagnosis.

Description: 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. Increased pericranial tenderness may be present on manual palpation.

A. At least 10 episodes of headache fulfilling criteria B through D. Infrequent and frequent episodic subforms of TTH are distinguished as follows:

Infrequent episodic TTH: Headache occurring on <1 day per month on average (<12 days per year).

Frequent episodic TTH: Headache occurring on 1 to 14 days per month on average for >3 months (≥12 and <180 days per year).

B. Headache lasting from 30 minutes to seven days.

C. At least two of the following four characteristics:

Bilateral location.

Pressing or tightening (nonpulsating) quality.

Mild or moderate intensity.

Not aggravated by routine physical activity such as walking or climbing stairs.

D. Both of the following:

No nausea or vomiting.

TTH: tension-type headache; ICHD-3: The International Classification of Headache Disorders, 3rd edition.

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Graphic 79672 Version 9.0

Clinical features of the trigeminal autonomic cephalalgias

	Cluster headache	Paroxysmal hemicrania	SUNCT and SUNA	Hemicrania continua
Sex (female:male)	1:3 to 1:7	1:1 to 2.7:1	1:1.5	2:1
Pain				
Туре	Stabbing, boring	Sharp, stabbing, throbbing	Burning, stabbing, sharp	Throbbing, sharp, pressure, dull, burning, aching, or stabbing
Severity	Excruciating	Excruciating	Severe to excruciating	Mild to severe
Site	Orbit, temple	Orbit, temple	Periorbital	Orbital, frontal, temporal; less often occipital
Attack frequency	1 every other day to 8 per day	1 to 40 a day (>5 per day for more than half the time)	1 to 200 per day	Continuous pain with exacerbations
Duration of attack	15 to 180 minutes	2 to 30 minutes	1 to 600 seconds	Months to years
Autonomic features	Yes	Yes	Yes (prominent conjunctival injection and lacrimation with SUNCT)	Yes
Restlessness and/or agitation	Yes	Yes	Frequent	Yes
Migrainous features (nausea, photophobia, or phonophobia)	Yes	Yes	Rare	Frequent
Alcohol trigger	Yes	Occasional	No	Occasional
Cutaneous triggers	No	Rare	Yes	No
Indomethacin effect	None	Absolute	None	Absolute
Abortive treatment	Sumatriptan injection or nasal spray Oxygen	Nil	Lidocaine intravenous infusion	Nil
Prophylactic treatment	Verapamil Methysergide Lithium Galcanezumab	Indomethacin	Lamotrigine Topiramate Gabapentin	Indomethacin

SUNCT: short-lasting unilateral neuralgiform pain with conjunctival injection and tearing; SUNA: short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms.

Graphic 65541 Version 11.0

Diagnostic criteria for cluster headache

Cluster headache: Diagnostic criteria for cluster headache require the following:

- A. At least five attacks fulfilling criteria B through D
- B. Severe or very severe unilateral orbital, supraorbital, and/or temporal pain lasting 15 to 180 minutes when untreated; during part (but less than half) of the active time course of cluster headache, attacks may be less severe and/or of shorter or longer duration
- C. Either or both of the following:
 - 1. At least one of the following symptoms or signs ipsilateral to the headache:
 - a) Conjunctival injection and/or lacrimation
 - b) Nasal congestion and/or rhinorrhea
 - c) Evelid edema
 - d) Forehead and facial sweating
 - e) Miosis and/or ptosis
 - 2. A sense of restlessness or agitation
- D. Attacks have a frequency between one every other day and eight per day; during part (but less than half) of the active time-course of cluster headache, attacks may be less frequent
- E. Not better accounted for by another ICHD-3 diagnosis

Episodic cluster headache: Diagnostic criteria for episodic cluster headache require the following:

- A. Attacks fulfilling criteria for cluster headache and occurring in bouts (cluster periods)
- B. At least two cluster periods lasting from seven days to one year (when untreated) and separated by pain-free remission periods of three months or more

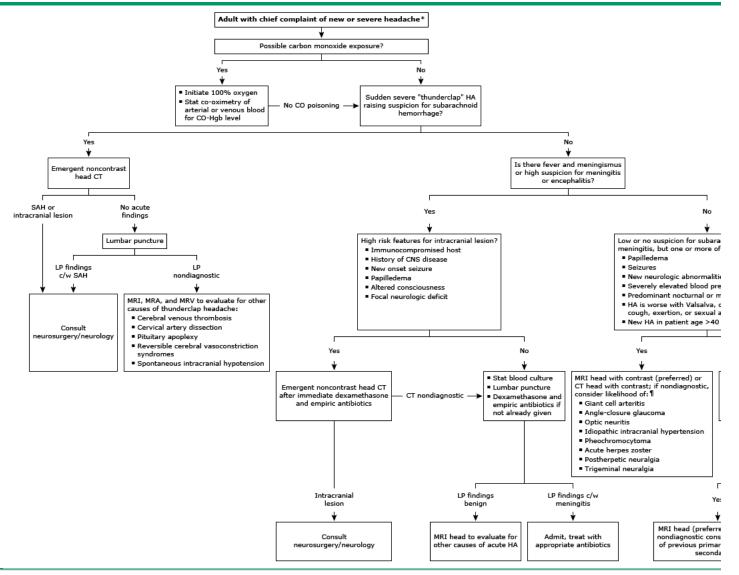
Chronic cluster headache: Diagnostic criteria for chronic cluster headache require the following:

- A. Attacks fulfilling criteria for cluster headache
- B. Attacks occurring without a remission period, or with remissions lasting less than three months, for at least one year

ICHD-3: International Classification of Headache Disorders, 3rd edition.

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Graphic 80843 Version 8.0



This is an overview of our approach to the urgent evaluation of headache in an adult. It should be used in conjunction with other UpToDate content on headache.

CO-Hgb: carboxyhemoglobin; CO: carbon monoxide; CT: computed tomography; HA: headache; CNS: central nervous system; MRA: magnetic resonance angiography; MRI: magnetic resonance imag LP: lumbar puncture; SAH: subarachnoid hemorrhage; c/w: consistent with.

* All patients with acute or subacute head trauma should have head CT scan; pregnant women > 20 weeks gestation or women in early postpartum should be evaluated for preeclampsia.

¶ The main clinical features of these conditions are listed below:

- Giant cell (temporal) arteritis typically affects people age >50 years and may be associated with systemic manifestations fever, fatigue, weight loss, jaw claudication, visual symptoms, partic diplopia, or symptoms of polymyalgia rheumatic.
- Acute angle-closure glaucoma may present with vision loss, headache, severe eye pain, light halos, nausea, and vomiting. Exam reveals a red eye, corneal cloudiness or edema, a shallow an dilated pupil.
- Optic neuritis typically presents with painful, monocular visual loss that evolves over several hours to a few days. One-third of patients have visible optic nerve inflammation (papillitis) on fundiorbits with gadolinium contrast confirms the diagnosis in most cases.
- Idiopathic intracranial hypertension (pseudotumor cerebri) typically affects obese women of child-bearing age. Characteristic features are headache, papilledema, vision loss or diplopia, elev with normal cerebrospinal fluid composition. MRI with MR venography is the preferred study to exclude secondary causes of elevated intracranial pressure, particularly cerebral venous thrombosi
- Pheochromocytoma is a rare condition that may present with episodic headache, sweating, and tachycardia. Sustained or paroxysmal hypertension is the most common sign, but 5 to 15% of I
 Acute herpes zoster usually presents with a vesicular rash and acute painful neuritis. Prodromal pain may precede the rash by days to weeks. In unusual cases, pain may be related to herpes:
- **Postherpetic neuralgia** usually affects the thoracic, cervical, and trigeminal nerves. In most cases, the diagnosis is made when pain persists beyond four months in the same distribution as a pherpes zoster.
- Trigeminal neuralgia is defined by sudden, usually unilateral, severe, brief, stabbing or lancinating, recurrent episodes of pain in the distribution of one or more branches of the fifth cranial (tri

Graphic 107496 Version 2.0

Etiologies of thunderclap headache

Most common causes of thunderclap headache:	
Subarachnoid hemorrhage	
Reversible cerebral vasoconstriction syndromes (RCVS)	
Conditions that less commonly cause thunderclap	headache:
Cerebral infection (eg, meningitis, acute complicated sinusitis)	
Cerebral venous thrombosis	
Cervical artery dissection	
Spontaneous intracranial hypotension	
Acute hypertensive crisis	
Posterior reversible leukoencephalopathy syndrome (PRES)	
Intracerebral hemorrhage	
Ischemic stroke	
Conditions that uncommonly or rarely cause thun	derclap headache:
Pituitary apoplexy	
Colloid cyst of the third ventricle	
Aortic arch dissection	
Aqueductal stenosis	
Brain tumor	
Giant cell arteritis	
Pheochromocytoma	
Pneumocephalus	
Retroclival hematoma	
Spinal epidural hematoma	
Varicella zoster virus vasculopathy	
Vogt-Koyanagi-Harada syndrome	
Disputed causes of thunderclap headache:	
Sentinel headache (unruptured intracranial aneurysm)*	
Primary thunderclap headache ¶	

Graphic 81710 Version 8.0

^{*} Sentinel headache due to an unruptured intracranial aneurysm is a possible cause of thunderclap headache, but supporting data are weak.

¶ There is controversy as to whether thunderclap headache can occur as a benign and potentially recurrent headache disorder in the absence of underlying organic intracranial pathology.

Differential diagnosis of headache with fever

Intracranial infection
Meningitis
Bacterial
Fungal
Viral
Lymphocytic
Encephalitis
Brain abscess
Subdural empyema
Systemic infection
Bacterial infection
Viral infection
HIV/AIDS
Other systemic infection
Other causes
Familial hemiplegic migraine
Pituitary apoplexy
Rhinosinusitis
Subarachnoid hemorrhage
Malignancy of central nervous system

HIV/AIDS: human immunodeficiency virus/acquired immunodeficiency syndrome.

Graphic 80966 Version 5.0