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Headaches that Make Your Head Ache

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- Optometrists are in many cases the first line of care for patients with headaches
- It is essential to have a basic working foundation of the common primary and secondary headache syndromes to:
  1. Recognize emergent situations that could be life-threatening
  2. Match your treatment plan to the type of headache and severity of illness

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Headache Patterns

- The amount of time a headache has been present can give direction on which etiologies to consider
- Certain chronic headache conditions are characterized by the recurrence of headache with relatively long headache-free intervals (e.g. migraine or cluster)
- Headaches that are significantly different from prior headaches can have a different etiology

Spierings ELH. Headache: The Most Common Complaints. Butterworth-Heinemann Ltd; 1998:92.

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Initial evaluation-Important Questions to Ask

1. Is this your first or worst headache? How bad is your pain on a scale of 1 to 10 (1 means not too bad, and 10 means very bad)? Do you have headaches on a regular basis? Is this headache like the ones you usually have?
2. What symptoms do you have before the headache starts? What symptoms do you have during the headache? What symptoms do you have right now?
3. When did this headache begin? How did it start (gradually, suddenly, other)?
4. Where is your pain? Does the pain seem to spread to any other area? If so, where?
5. What kind of pain do you have (throbbing, stabbing, dull, other)?
6. Do you have other medical problems? If so, what?
7. Do you take any medicines? If so, what?
8. Have you recently hurt your head or had a medical or dental procedure?

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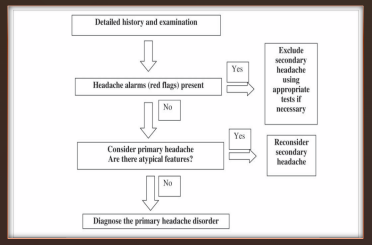
Headache evaluation

- **Step 1:** Distinguishing primary from secondary headache
  - Search for "red flags" that suggest the possibility of a secondary headache
  - Conduct the appropriate work-up to exclude the conditions suggested by the red flag
  - Once secondary headache has been excluded by history, physical and neurological evaluation, the next step is to diagnose the primary headache disorder
- **Step 2:** Classifying the primary headache based on duration and frequency



Bigal ME, Lipton RB. The differential diagnosis of chronic daily headaches: an algorithm-based approach. J Headache Pain January 2007;8:263-72.

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Headache Evaluation

- **Work-up:**
  - MOST IMPORTANT: Take a thorough history
  - Cranial nerve evaluation
  - Vitals
  - Ophthalmic examination
  - Blood work
  - Neuroimaging
  - Lumbar puncture
  - Referral for consultation (neurology/neuro-ophthalmology/internal medicine)

Ravishankar K. The art of history-taking in a headache patient. Ann Indian Acad Neurol January 2012;15:7.

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Recognizing red flag headaches

Worrisome Headache Red Flags—"SNOOP"
• **S**YSTEMIC SYMPTOMS (fever, weight loss) or SECONDARY RISK FACTORS (HIV, systemic cancer)
• **N**EUROLOGIC SYMPTOMS or abnormal signs (confusion, impaired alertness or consciousness)
• **O**NSET: sudden, abrupt, or split-second
• **O**LDER: new onset and progressive headache, especially in middle age >50 yr (giant cell arteritis)
• **P**REVIOUS HEADACHE HISTORY: first headache or different (change in attack frequency, severity, or clinical features)

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Prevalence of Headache in General Population

Headache	Prevalence
Primary	
Tension-type	78%
Migraine	16%
Secondary	
Fasting	19%
Sinus-related	15%
Head trauma	4%
Non-vascular intracranial Disease (including tumor)	0.5%

Rasmussen BK, Jensen R, Schroll M, Olesen J. Epidemiology of headache in a general population — a prevalence study. J Clin Epidemiol 1991;44:1147-1157.

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Primary Headache Disorders

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Tension-Type Headache (TTH)

- Criteria according to International Headache Society:
  - A. At least 10 episodes fulfilling criteria B-D, below
  - B. Episodes last from 30 mins to 7 days
  - C. Headache has at least two of the following characteristics:
    1. Pressing or tightening quality
    2. Mild or moderate intensity
    3. Bilateral location
    4. Not aggravated by routine physical
  - D. During headache, both of the following:
    1. No nausea or vomiting
    2. Photophobia or phonophobia

Spierings ELH. Headache: The Most Common Complaints. Butterworth-Heinemann Ltd; 1998:92.

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Tension-Type Headache, cont'd

- **Episodic:**
  - Commonly triggered by stress, tension, fatigue, lack of sleep, and not eating on time
  - Headaches typically begin during the day, typically in the late afternoon, and may be relieved by the evening
  - These headaches tend to have a diffuse location (i.e. across the forehead, on top of, or in the back of the head)
  - Tend to respond well to OTC analgesics

Spierings ELH. Headache: The Most Common Complaints. Butterworth-Heinemann Ltd; 1998:92.

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Tension-Type Headache, cont'd

- **Chronic TTH**
  - Typically occur daily or almost daily
  - Similar to episodic tension-type headaches, but are present upon waking or shortly after rising
  - Can be caused by chronic anxiety, but are often related to chronic fatigue due to lack of sleep
  - These headaches are often secondary and develop out of episodic TTH

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Prevalence of migraine and severe headache

- Affects 1 out of 6 Americans
- 1 in 5 women over a 3 month period
- 15.3% overall
  - Males: 9.7%
  - Females: 20.7%
- Race
  - Highest in American Indian or Alaskan Natives (18.4%) compared with Caucasians, African Americans, or Hispanics
  - Lowest prevalence in Asians (11.3%)

Burch R, Rizzoli P, Loder E. The prevalence and impact of migraine and severe headache in the united states: Figures and trends from government health studies. *Headache: The Journal of Head and Face Pain*. 2018;58(4):496-505.

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Migraine Headache (without aura)

- Criteria according to International Headache Society:
  - A. At least five attacks fulfilling conditions B-D, below
  - B. Attacks lasting 4-72 hours
  - C. Headache has at least two of the following characteristics:
    1. Unilateral location
    2. Pulsating quality
    3. Moderate or severe intensity
    4. Aggravated by routine physical activity
  - D. During headache, at least one of the following:
    1. Nausea or vomiting
    2. Photophobia or phonophobia

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Migraine Triggers

- Menstrual cycle
  - Associated with estrogen withdrawal
  - Oral contraceptive use
- Stress
  - Headache typically occurs after the stress when relaxation occurs
- Vasoactive agents
  - Vasodilators
    - Alcohol, sodium nitrite
  - Vasoconstrictors
    - Caffeine, sympathomimetic amines, tyramine (cheese, red wine), phenylethylamine (dark chocolate)

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Migraine with Aura ICHD-3,1.2

- In classic migraines, or migraine with aura, the headaches are preceded by transient focal neurologic symptoms
  - Sub-classifications:
    - Typical aura with headache
    - Typical aura without headache
- Per the International Headache Society (ICHD-3) classification:
  - Migraine with aura in which aura consists of visual and/or sensory and/or speech/language symptoms, but no motor weakness, and is characterized by gradual development, duration of each symptom no longer than one hour, a mix of positive and negative features and complete reversibility.
  - Diagnostic criteria:
  - A. Attacks fulfilling criteria for migraine listed previously
  - B. Aura with both of the following:
    - fully reversible visual, sensory and/or speech/language symptoms
    - no motor, brainstem or retinal symptoms.

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- **Visual aura-typical description**
  - Scintillating scotoma, also called *teichopsia* or *fortification spectra*
  - Vision often obscured by zigzag lines and band of dimness that lies against it on the inside of the crescent
  - Typically lasts 10-30 minutes (average ~20 minutes)

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Migraine Aura, cont'd

- **Somatosensory disturbance**
  - Also lasts for approximately 10-30 minutes
  - Typically presents in the form of digitolingual paresthesias (*cheiro-oral syndrome*)
    - Feeling of tingling or pins-and-needles that start in the fingers of one hand with gradual extension upward into the arm and face on the same side
  - Often follows the visual aura symptoms
  - Headache follows the visual or somatosensory disturbances either right away or after a certain interval of time (e.g. 1 hour)

Spierings ELH. Headache: The Most Common Complaints. Butterworth-Heinemann Ltd; 1998.

Hershey AD, Kabbouche MA, O'Brien HL, Kacperski J. Headaches. 4th ed.(Wolters Kluwer):2017.

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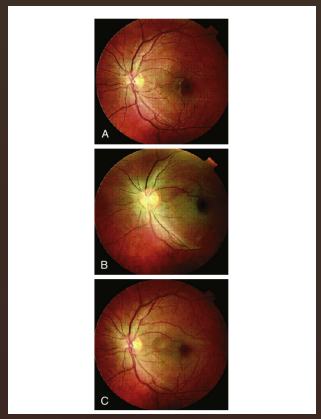
Retinal (ocular) migraine

- Characterized by monocular aura without headache
- Monocular aura typically implies dysfunction anterior to the optic chiasm
- Retinal artery vasospasm has been observed during episodes



Doyle E, Vote BJ, Casswell AG. Retinal migraine: caught in the act. *Br J Ophthalmol.* 2004;88(2):301–302. doi:10.1136/bjo.2003.021808

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Ophthalmoplegic migraine

- Very rare condition characterized by recurrent attacks of migrainous headache associated with reversible paresis of one or more ocular cranial nerves
- CNIII involved in more than 95% of cases
  - Pupil usually spared during attack
- More common in children than in adults
- Ophthalmoplegia follows the headache, typically within 1 to 4 days

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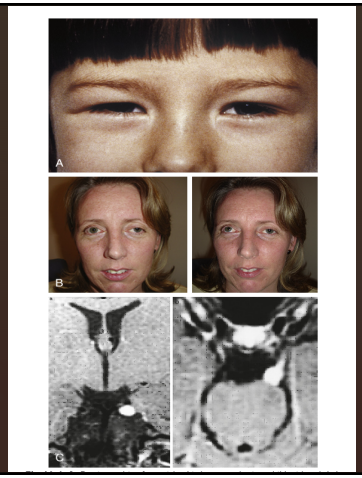

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Migraine complications

- Serious complications are rare, but there is extensive literature relating migraines to the following:
  - **Status migrainosus**
  - **Migrainous infarction:**
    - mostly in posterior circulation and in young women
  - **Seizures**
  - **Persistent aura without infarction**

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Cluster Headaches-Presentation

Characteristics	Finding
Age of onset	20-40 years old
Sex distribution	90% **male**
Episodic	85% of patients
Chronic	
Primary	10%
Secondary	5%
Attack duration	30 minutes to 2 hours
Attack frequency	1-2 per day
Duration of episodes	2 weeks to 2 months
Duration of remissions	6 months to one year
Laterality	
Right	50% of patients
Left	40% of patients
Either	10% of patients

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Cluster headache symptoms

- Pain is always unilateral and located in or behind the eye, in the forehead, or in the temple
- Autonomic symptoms often occur, including:
  - Reddening and tearing of the eye
  - Edematous swelling and drooping of the upper eyelid
  - Narrowing of the pupil
  - Increased sweating over the forehead
  - Stuffiness/running of the nose
- Systemic symptoms such as nausea and vomiting that are common in migraine headaches are rare with cluster headaches

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Cluster headache, cont'd

Common triggers

- Most consistently related to alcohol and daytime napping
- When triggered by alcohol, it typically occurs 30-45 minutes after ingestion of even a small quantity
- Occurrence of episodes has been related to the lengthening and shortening of days in spring and fall, respectively

Patient profile

- Described as the *leonine mouse syndrome* (John R. Graham)
  - Refers to the husky appearance of many of these patients and their leonine facial features, including:
    - Ruddy complexion, deep furrows, and prominent eyebrows
  - They also tend to have a very timid personality, with increased dependency needs
  - Cluster headache patients tend to smoke and drink excessively

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Medication Overuse Headache

- **International Headache Society definition:**
  - **Description:**
    - Headache occurring on 15 or more days/month in a patient with a pre-existing primary headache and developing as a consequence of regular overuse of acute or symptomatic headache medication (on 10 or more or 15 or more days/month, depending on the medication) for more than 3 months. It usually, but not invariably, resolves after the overuse is stopped.
  - **Diagnostic criteria:**
    - Headache occurring on ≥15 days/month in a patient with a pre-existing headache disorder
    - Regular overuse for >3 months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache[1,2,3]
    - Not better accounted for by another ICHD-3 diagnosis.

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Secondary Headache Disorders

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Headache or Facial Pain attributed to Eye

- Angle closure
- Uveitis
- Scleritis or orbital inflammation
- Optic neuritis
- Refractive disorders and muscle imbalance
- Ophthalmic herpes zoster → Post-herpetic neuralgia (PHN)

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Headache associated with infection

- **Meningitis**
  - Incidence and etiology: 10-20 cases per 100,000 population per year. Children <5 YOA account for 70% of the cases
  - Presentation and Diagnosis:
    - Develops acutely over hours or days.
    - May be preceded by respiratory or gastrointestinal illness
    - Severe, bilateral headache associated with photophobia, nausea, and vomiting
    - Fever and signs of meningeal irritation observed in 80% of cases
    - Dx made through lumbar puncture and analysis of spinal fluid

Clinch CR. Evaluation of acute headaches in adults. Am Fam Physician February 2001;63:685-92.
Spierings ELH. Headache: The Most Common Complaints. Butterworth-Heinemann Ltd; 1998:92.

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Meningitis, cont'd

Bacterial

- Usually more severe symptoms
- Cell count >1000/ul
- Mostly polymorphonuclear leukocytes
- Protein level elevated
- Glucose decreased

Viral

- Cell count <1000/ul
- Mostly lymphocytes
- Protein and glucose levels normal

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Headache Attributed to Non-traumatic Intracranial Hemorrhage

- **Subarachnoid hemorrhage**
  - Incidence and etiology: annual incidence of subarachnoid hemorrhage in the U.S. is estimated at 10-15 cases per 100,000 population
    - In 75% of patients, the hemorrhage occurs from a ruptured aneurysm 25% by arteriovenous malformation or bleeding disorder
  - Presentation
    - 2/3 of patients are between 40-60 YOA
    - Women>men
    - Precipitated by activities such as lifting, straining, intercourse, or emotional excitement
    - Can occur during sleep, probably related to the increase in blood pressure during REM sleep
  - Diagnosis
    - Characterized by headache of hyperacute onset, which is KEY to diagnosis
    - Develops in a matter of seconds, like a blow to the head or neck
      - Warrants diagnostic imaging, then lumbar puncture if imaging negative

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Headaches associated with disorders of homeostasis

- **Hypertensive crisis without encephalopathy ICHD-3, 10.3.2**
- A hypertensive crisis is defined as a paroxysmal rise in systolic (to ≥180 mm Hg) and/or diastolic (to ≥120 mm Hg) blood pressure.
  - Headache fulfilling criterion C
  - Both of the following:
    - a hypertensive crisis1 is occurring
    - no clinical features or other evidence of hypertensive encephalopathy
  - Evidence of causation demonstrated by at least two of the following:
    - headache has developed during the hypertensive crisis
    - either or both of the following:
      - a) headache has significantly worsened in parallel with increasing hypertension
      - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the hypertensive crisis
  - headache has at least one of the following three characteristics:
  - a) bilateral location
  - b) pulsating quality
  - c) precipitated by physical activity

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Headache Attributed to Cranial or Cervical Vascular Disorder

- **Giant cell arteritis**
  - Inflammation of medium and large-sized arteries in the body (vasculitis)
  - Presentation:
    - Occurs almost exclusively after age 50 and women are afflicted with the disease 2 to 3 times more commonly than men
    - Most common systemic vasculitis among adults
      - Incidence: 18.9 per 100,000
      - Prevalence: 228 per 100,000

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Systemic symptoms

- General malaise
- Generalized weakness
- Easy fatigability
- Lack of appetite
- Weight loss
- Low grade fever

Specific symptoms

- Headache
- Jaw claudication
- Blindness
- Stroke

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- Examination:
  - Specific abnormalities are not often observed on physical examination
  - Routine lab testing typically reveals an elevated ESR and CRP
    - The sedimentation rate is generally increased 50-100mm per hour, but may also be normal
  - With strong clinical suspicion, even with a normal ESR, a biopsy should be performed
- Treatment:
  - Corticosteroids

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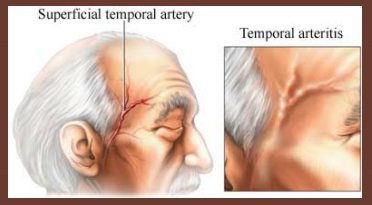

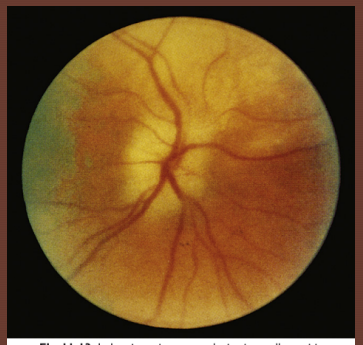

Fig. 11-13. Ischemic optic neuropathy in giant cell arteritis.

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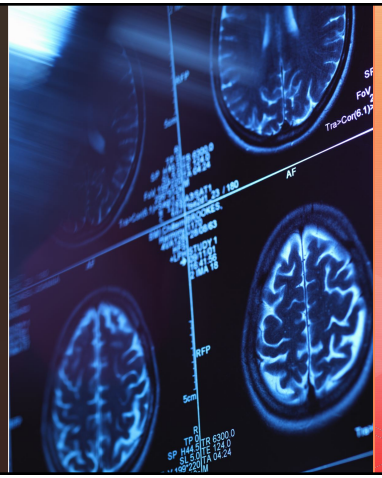
- Other causes:
  - Hemorrhagic stroke
  - Pituitary apoplexy
  - Unruptured arteriovenous malformation



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Headaches Attributed to Non-Vascular Intracranial Disorder

- **Cerebral tumor:**
  - Etiology:
    - Divided into primary and secondary (metastatic, 30%)
    - Reach the brain from hematogenic seeding, most often from breast or lung cancer
  - Incidence:
    - 5-10 cases per 100,000 population per year
    - Age: Between 50 and 70 years of age

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- **Types of cerebral tumors:**
  - Glioma:
    - 50% of cases, tumor originates from glial cells of brain
    - 60% of gliomas are malignant, often resulting in death within 6 months to one year
    - Other gliomas such as astrocytoma and oligodendroglioma are much less disastrous, but are still malignant
    - Initial manifestation of symptoms is generally not headache, but neurological symptoms or seizure

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- Meningioma
  - Typically benign tumors that arise from arachnoid cap cells of the middle meningeal layer – can be malignant and highly aggressive
  - Women>men
  - Can be found in brain, spinal cord, or optic nerve
  - Headache is localized to the side of the tumor (not to exact location)
    - May be intermittent or continuous, but often progresses in intensity
    - Easily mistaken for tension-type or migraine headaches

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Headaches associated with nonvascular intracranial disorders

- **Idiopathic intracranial hypertension**
  - Symptoms and signs
    - Pseudotumor cerebri is a condition of increased intracranial pressure caused by impaired absorption of CSF or edematous swelling of the brain, or both.
    - The condition is often seen in overweight women of reproductive age
    - Headache is present in more than 80% of patients and generalized in location
    - Other symptoms: Transient vision loss, double vision, pulsatile tinnitus

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- Diagnosis:
  - Typically made through lumbar puncture, which often reveals a markedly elevated pressure in the range of 300-400mm H2O
- Management
  - Vigorous weight loss management is key to treatment
  - Lumbar punctures may help decrease the ICP
  - Lumboperitoneal shunt can also be considered if lumbar puncture not effective
  - Acetazolamide and furosemide are often used to decrease the formation of CSF
  - In cases involving severe papilledema, optic nerve sheath fenestration may be indicated to spare vision

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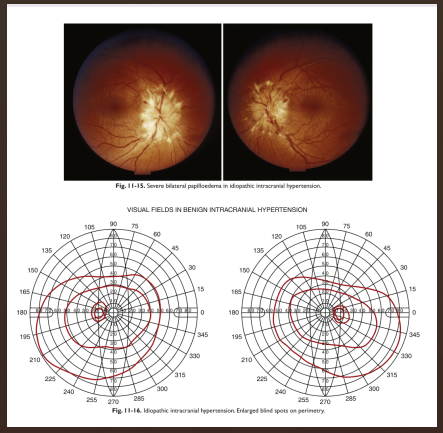

Fig. 11-15. Severe bilateral papilledema in idiopathic intracranial hypertension.

Fig. 11-16. Idiopathic intracranial hypertension. Enlarged blind spots on perimetry.

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Headache associated with trauma

- **Subdural hematoma:**
  - Etiology:
    - Most often the result of a head injury
    - Hematoma of venous origin located over the cerebral hemisphere and can be unilateral or bilateral
      - Caused by a rupture of a bridging vein resulting from trauma to the head
  - Presentation/Symptoms/Signs:
    - Age > 50 years, incidence increases with age
    - More common in men than women
    - Headache is frequently the presenting symptom, but it may be absent
    - Confusion, disorientation, and drowsiness are also common presenting symptoms

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Headache associated with trauma

- **Diagnosis:**
  - Neurodiagnostic imaging (CT or MRI)
  - CT shows it as hyperdense, isodense, or hypodense
- **Treatment:**
  - Medical: Corticosteroids
  - Surgical: Evacuation of the hematoma and is associated with the general risks of surgery

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Summary

- It's impossible to know about every single type of headache and all of the associated clinical features, but we have a big role in the differential diagnosis and treatment/management of these patients

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