HEADACHE

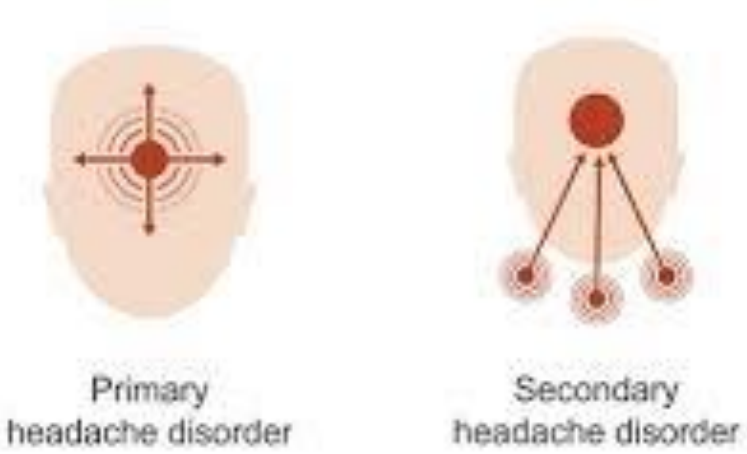
Headaches are one of the most common neurological problems presented to GPs and neurologists. They are painful and debilitating for individuals, an important cause of absence from work or school and a substantial burden on society.

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

This high prevalence results in significant disability and lost productivity: headache disorders are the 3rd highest cause of **years lost to disability** worldwide

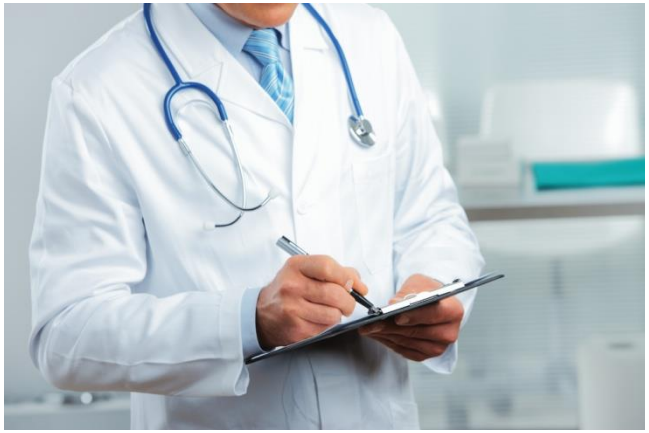
TWO MAJOR CATEGORIES

Primary headache disorders (those caused by the headache disorder itself, not due to other causes) and **secondary headache disorders**, those caused by (or "symptomatic of") another underlying medical problem



The pain can be due to the involvement of pain-sensitive structures in the head , 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 multifactorial headaches, so a detailed history and examination are necessary to identify the contributing factors



Key points from the history

When obtaining the history the following information 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

Additional aspects of the history 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)

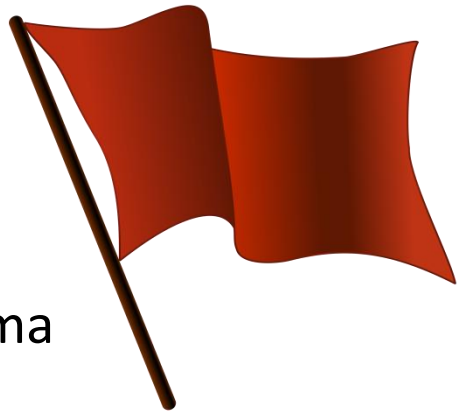
Social history, including detailed screening for illicit drugs

Family history

Sleep, including a history of insomnia and snoring, symptoms suggestive of obstructive sleep apnea

Evaluate people who present with headache and any of the following features, and consider the need for further investigations and/or referral:

- Worsening headache with fever
- Sudden-onset headache reaching maximum intensity within 5 minutes
- New-onset neurological deficit
- New-onset cognitive dysfunction
- Change in personality
- Impaired level of consciousness
- Recent (typically within the past 3 months) head trauma
- Headache triggered by cough, valsalva (trying to breathe out with nose and mouth blocked) or sneeze
- Headache triggered by exercise
- Orthostatic headache (headache that changes with posture)
- Symptoms suggestive of giant cell arteritis
- Symptoms and signs of acute narrow angle glaucoma
- A substantial change in the characteristics of their headache.



Consider further investigations and/or referral for people who present with new-onset headache and any of the following:

- Compromised immunity, caused, for example, by HIV or immunosuppressive drugs
- Age under 20 years and a history of malignancy
- A history of malignancy known to metastasise to the brain
- Vomiting without other obvious cause.

In the era of the smartphone, patients may bring pictures of themselves to a clinician for review if they have paroxysmal symptoms and signs(such as ptosis or lacrimation), which can aid in the diagnosis

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

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

Fever may suggest an underlying infection, including a CNS infection

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

A detailed head and neck exam includes evaluating for nuchal rigidity, cervical myofascial pain, occipital Tinel sign(evaluated by eliciting tenderness or tingling when palpating near the occipital protuberance along the occipital nerve), and palpation of the TMJ, assessment of dental wearing or chipping to suggest bruxism, and observing the oropharynx for narrowing that could suggest obstructive sleep apnea

A full neurological exam should also be performed, with emphasis on the funduscopic exam to assess for papilledema

The cortical sensory exam can suggest cortical dysfunction that may occur with venous sinus thrombosis

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

Condition	Notes
Migraine	Recurrent, severe headache which is usually unilateral and throbbing in nature May be be associated with aura, nausea and photosensitivity Aggravated by, or causes avoidance of, routine activities of daily living. Patients often describe 'going to bed'. In women may be associated with menstruation
Tension headache	Recurrent, non-disabling, bilateral headache, often described as a 'tight-band' Not aggravated by routine activities of daily living
Cluster headache*	Pain typical occurs once or twice a day, each episode lasting 15 mins - 2 hours with clusters typically lasting 4-12 weeks Intense pain around one eye (recurrent attacks 'always' affect same side) Patient is restless during an attack Accompanied by redness, lacrimation, lid swelling More common in men and smokers

*some neurologists use the term trigeminal autonomic cephalgia to group a number of conditions including cluster headache, paroxysmal hemicrania and short-lived unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). It is recommended such patients are referred for specialist assessment as specific treatment may be required, for example it is known paroxysmal hemicrania responds very well to indomethacin

TABLE 10-1. Key Features of Primary Headache Disorders

	Episodic Migraine	Episodic Tension	Episodic Cluster	Paroxysmal Hemicrania	SUNCT and SUNA
Sex	Female > male	Female > male	Male > female	Female > male	Male > female
Location	Unilateral > bilateral	Bilateral (band around the head)	Unilateral (behind or around the eye)	Unilateral (behind or around the eye)	Unilateral (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 features	No	No	Yes	Yes	Yes

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

Temporal arteritis	Typically patient > 60 years old Usually rapid onset (e.g. < 1 month) of unilateral headache Jaw claudication (65%) Tender, palpable temporal artery Raised ESR
Medication overuse headache	Present for 15 days or more per month Developed or worsened whilst taking regular symptomatic medication Patients using opioids and triptans are at most risk May be psychiatric co-morbidity

Other causes of headache

Acute single episode

Meningitis
Encephalitis
Subarachnoid haemorrhage
Head injury
Sinusitis
Glaucoma (acute closed-angle)
Tropical illness e.g. Malaria

Chronic headache

Chronically raised ICP
Paget's disease
Psychological

TENSION-TYPE HEADACHE

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

Pain is usually bilateral and described as pressure or tightness

It is usually mild to moderate and lasts for under an hour to several days

Unlike migraine, it is not associated with photophobia, phonophobia, nausea, or vomiting

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

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

Patients with frequent or chronic tension-type headaches benefit from treatment

Tension-type headache treatment

1. Abortive treatments

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

For those with moderate to severe pain, NSAIDs are the mainstay of treatment

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

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

2. Preventive treatments

Antidepressants are the first-line preventive therapy for chronic tension headache

The tricyclic amitryptiline is the most studied to date and has good evidence for efficacy

Other antidepressants , including mirtazapine and venlafaxine are second-line therapies

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

3. Adjuvant treatments

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

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

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

Migraine

The International Headache Society has produced the following diagnostic criteria for migraine without aura:

Point	Criteria
A	At least 5 attacks fulfilling criteria B-D
B	Headache attacks lasting 4-72 hours* (untreated or unsuccessfully treated)
C	Headache has at least two of the following characteristics:• 1. unilateral location* 2. pulsating quality (i.e., varying with the heartbeat)• 3. moderate or severe pain intensity• 4. aggravation by or causing avoidance of routine physical activity (e.g., walking or climbing stairs)•
D	During headache at least one of the following:• 1. nausea and/or vomiting* 2. photophobia and phonophobia•
E	Not attributed to another disorder (history and examination do not suggest a secondary headache disorder or, if they do, it is ruled out by appropriate investigations or headache attacks do not occur for the first time in close temporal relation to the other disorder)

Migraine

Migraine with aura (seen in around 25% of migraine patients) tends to be easier to diagnose with a typical aura being progressive in nature and may occur hours prior to the headache. Typical aura include a transient hemianopic disturbance or a spreading scintillating scotoma ('jagged crescent'). Sensory symptoms may also occur

Auras may occur with or without headache and:

- Are fully reversible
- Develop over at least 5 minutes
- Last 5-60 minutes

The following aura symptoms are atypical and may prompt further investigation/referral;

motor weakness

double vision

visual symptoms affecting only one eye

poor balance

decreased level of consciousness.

Management

Acute treatment

First-line: offer combination therapy with

an oral triptan and an NSAID, or

an oral triptan and paracetamol

For young people aged 12-17 years consider a nasal triptan in preference to an oral triptan

If the above measures are not effective or not tolerated offer a non-oral preparation of metoclopramide or prochlorperazine and consider adding a non-oral NSAID or triptan

Caution should be exercised when prescribing metoclopramide to young patients as acute dystonic reactions may develop

Prophylaxis

Prophylaxis should generally be given if '*Migraine attacks are having a significant impact on quality of life and daily function, for example they occur frequently (more than once a week on average) or are prolonged and severe despite optimal acute treatment*'

Options:

Propranolol

Topiramate: should be avoided in women of childbearing age as it may be teratogenic and it can reduce the effectiveness of hormonal contraceptives

Amitriptyline

For women with predictable menstrual migraine treatment NICE recommend either frovatriptan (2.5 mg twice a day) or zolmitriptan (2.5 mg twice or three times a day) as a type of 'mini-prophylaxis‘

Monoclonal antibodies directed against the calcitonin gene-related peptide (CGRP) receptor: examples include erenumab

Cluster headache

Cluster headaches are known to be one of the most painful conditions that patients can have the misfortune to suffer. The name relates to the pattern of the headaches - they typically occur in clusters lasting several weeks, with the clusters themselves typically once a year.

Cluster headaches are more common in men (3:1) and smokers. Alcohol may trigger an attack and there also appears to be a relation to nocturnal sleep.

Features

Intense sharp, stabbing pain around one eye. Pain typical occurs once or twice a day, each episode lasting 15 mins - 2 hours

The patient is restless and agitated during an attack due to the severity

Clusters typically last 4-12 weeks accompanied by redness, lacrimation, lid swelling nasal stuffiness miosis and ptosis in a minority

Investigations

Most patients will have neuroimaging - underlying brain lesions are sometimes found even if the clinical symptoms are typical for cluster headache

MRI with gadolinium contrast is the investigation of choice

Management

NICE recommend seeking specialist advice from a neurologist if a patient develops cluster headaches

Acute

100% oxygen (80% response rate within 15 minutes)

Subcutaneous triptan (75% response rate within 15 minutes)

Prophylaxis

Verapamil is the drug of choice

There is also some evidence to support a tapering dose of prednisolone

Some neurologists use the term trigeminal autonomic cephalgia to group a number of conditions including cluster headache, paroxysmal hemicrania and short-lived unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT). It is recommended such patients are referred for specialist assessment as specific treatment may be required, for example it is known paroxysmal hemicrania responds very well to indomethacin

SECONDARY HEADACHE DISORDERS

Secondary headache disorders are headaches caused by a medical condition or medication

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

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

Treatment is based on addressing the underlying disorder

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

1. **Vascular causes**

There are numerous vascular causes of headache

All cerebral hemorrhages can cause headache

This includes subarachnoid hemorrhage(SAH) , intraparenchymal hemorrhage, and subdural and epidural hematomas

These hemorrhages may be spontaneous (associated with stroke or hypertension) or traumatic

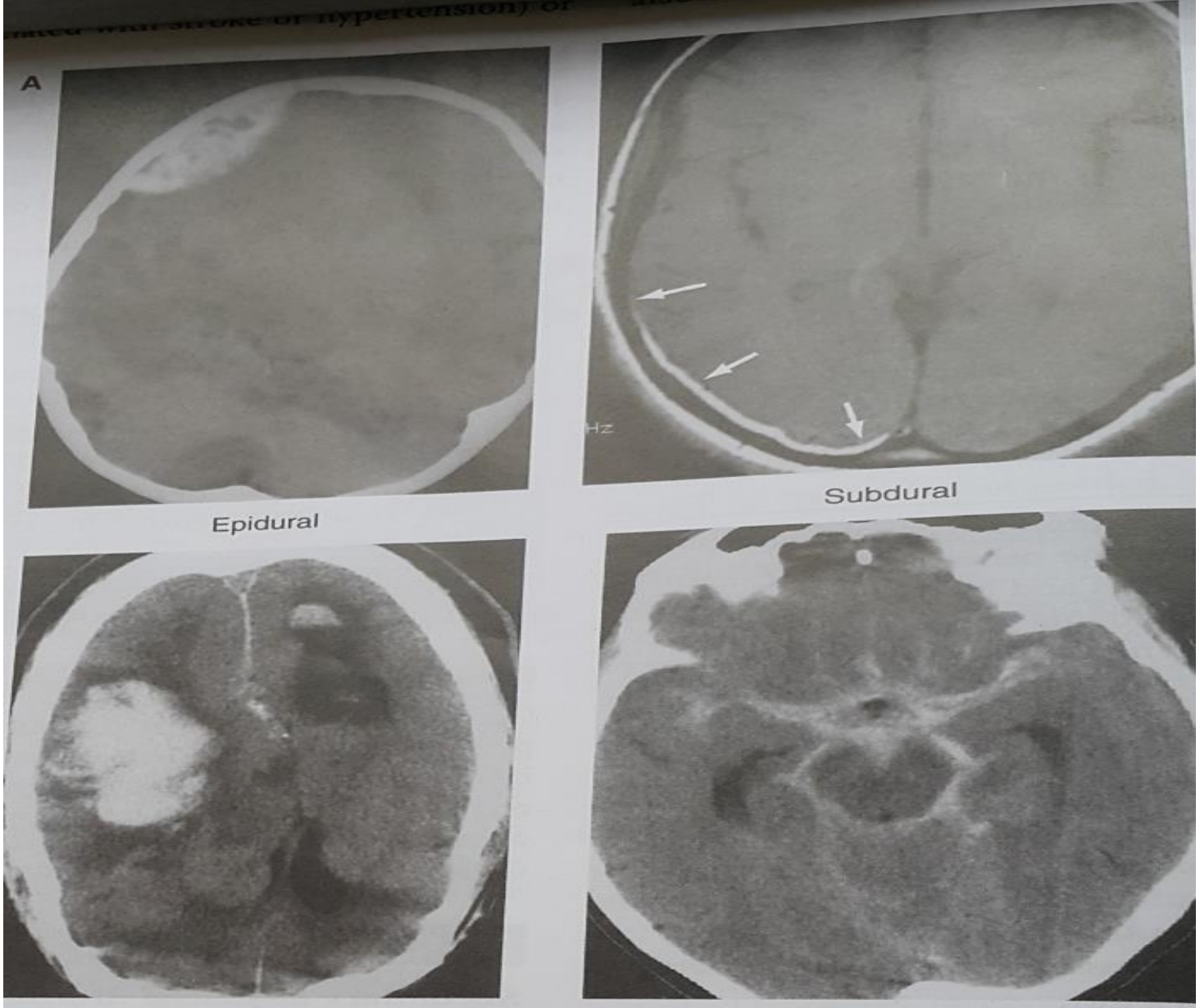
Patients **with intracerebral hemorrhages** typically present with what is referred to as thunderclap headache; the onset is abrupt and severe

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

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

Their semiology is nonspecific, but typically abrupt in onset

Patients may have focal neurologic deficits which aid in the diagnosis



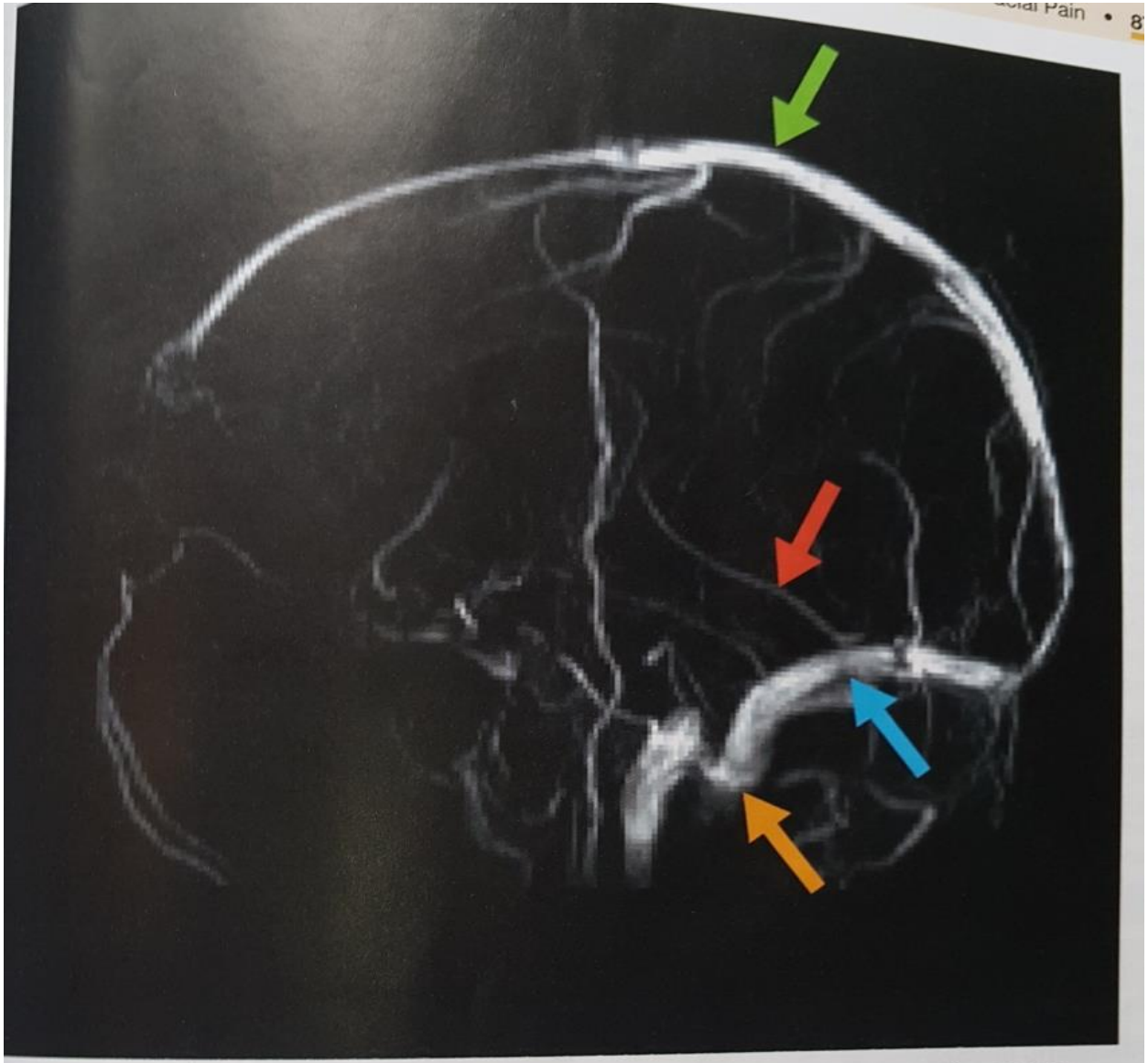
s of intracerebral hemorrhages on CT scans. **(A)** Epidural hemorrhage. **(B)** Su
hemorrhage. **(D)** Subarachnoid hemorrhage. Arrows point to the subdural he

Cerebral thrombosis, either arterial or venous, can also cause headache

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

Thrombosis should be considered particularly in patients with hypercoagulability states, including pregnancy

The diagnosis is made on imaging, including that of cerebral vessels (figure)



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

Giant cell arteritis (GCA) ,

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

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

Involvement of the branches of the external carotid artery, and occasionally the ophthalmic artery, can result in blindness if not readily identified and treated promptly

Patients usually have elevated inflammatory markers(erythrocyte sedimentation rate and C-reactive protein)

Empiric steroids should be started in any patient with a high clinical concern

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

2. Infectious or inflammatory:

Intracranial infections , such as encephalitis and meningitis, usually present with headache and often fever

They may also have nuchal rigidity and altered mental status

As the infection progresses , seizures and focal neurological deficits may occur

Infections can be bacterial, viral , fungal, or parasitic; the headache semiology does not help to differentiate the underlying cause

LP is the crucial diagnostic test and necessary in any patient for whom there is concern for a CNS infection

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

CNS inflammatory and autoimmune conditions such as sarcoidosis and lupus frequently present with headache

3. Neoplastic causes

Intracranial neoplasms may present with headaches, especially when there is significant mass effect

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

The headache may occur early or late with neoplasms and with any type of primary cancer

4. Traumatic causes

Head and neck injuries often result in headache

To be attributed to trauma, the headache must develop with a temporal association with the injury

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

There is no specific headache semiology that helps with the diagnosis

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

5. Intracerebral pressure disorders

Intracranial hypertension and hypotension can cause headaches, but with markedly different semiologies

A) Intracranial hypertension

Intracranial hypertension may be “ idiopathic”(most common in obese young woman) or due to medications or systemic disorders

The headache is often described as worse when *supine* or sleeping

(awakening the patient from sleep) , or with Valsalva maneuver

Patients often have associated features , including papilledema, pulsatile tinnitus, or visual symptoms

Patients should have imaging to exclude a mass lesion or venous sinus thrombosis

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

Intracerebral pressure (ICP) is elevated above 200 mm CSF

Acetazolamide is the first-line treatment for idiopathic intracranial hypertension (IIH)

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

B) Intracranial hypotension

Intracranial hypotension may be spontaneous or traumatic

The headache improves when the patient is supine but worsens with standing

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

It is frequently traumatic- occurring after an attempted epidural puncture or LP

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

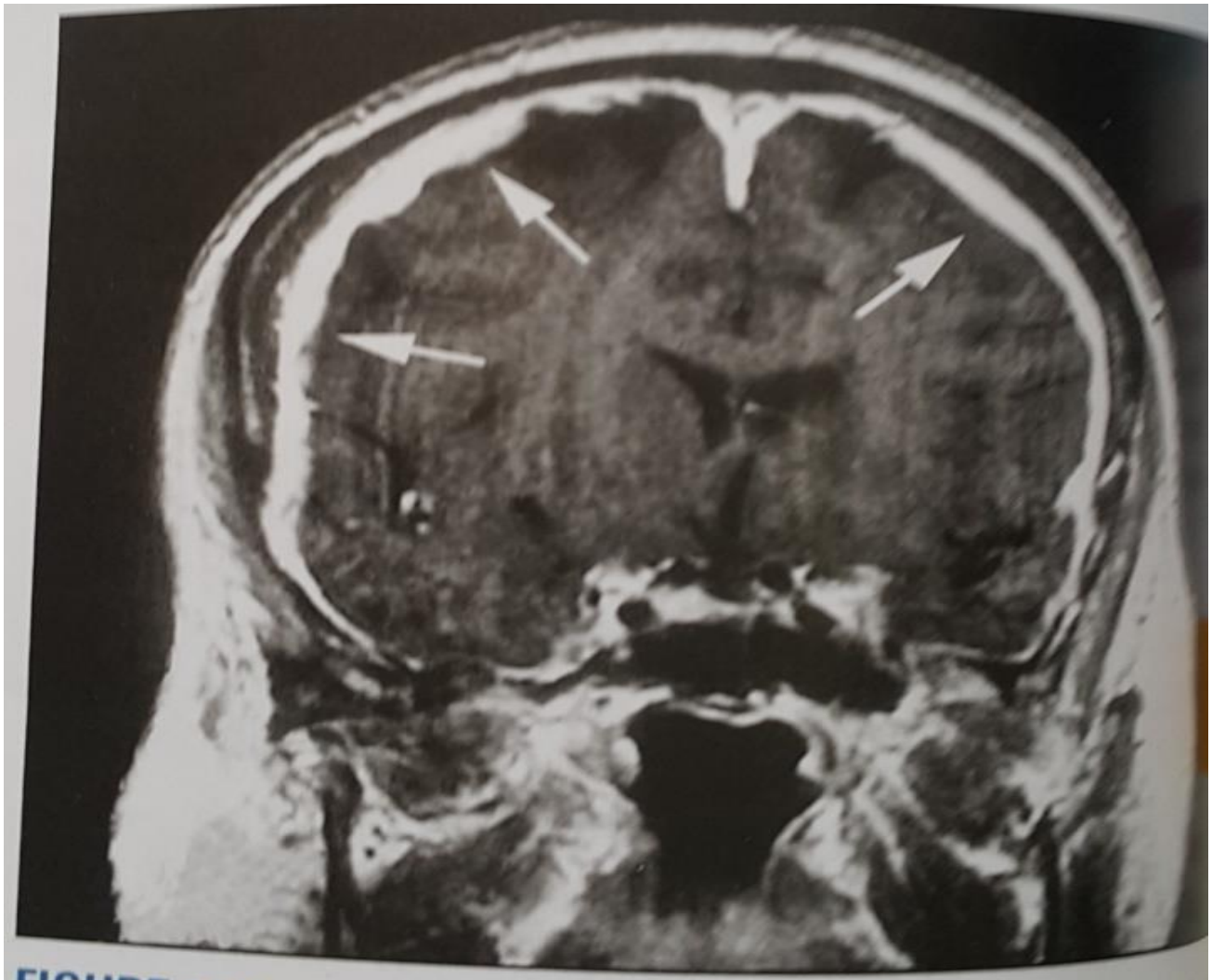


FIGURE 10-5. Intracranial hypotension. Gadolinium-enhanced MRI scan of a patient with intracranial hypotension. There is widespread, symmetric meningeal enhancement (*arrows*).

The definitive diagnosis is made with an LP showing an opening pressure below 60 mm CSF

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

6. Medication causes

Medications associated with headaches are numerous

They range from hormonal therapies, including contraceptives, to nitric oxide

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

Headache semiology is nonspecific, but the temporal association to medication change helps establish the diagnosis

Head and neck disorders

There are many different structural disorders that can cause headache

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

Sinusitis is commonly associated with a headache

It may be bifrontal or unilateral

Acute sinusitis is often associated with other symptoms of a respiratory tract infection

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

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

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

Cervical range of motion is often reduced

Trigeminal neuralgia

The pain of trigeminal neuralgia is shock-like, severe, occurring in one or all branches of the trigeminal nerve

The pain is usually paroxysmal and recurrent

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

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

The following ar red flags suggesting serious underlying cause:

1. Sensory changes
2. Deafness or ear problems
3. Pain only in the ophthalmic devision
4. Optic neuritis
5. Family history of MS
6. Age of onset before 40

For these changes do brain MRI with contrast

Carbamazepine is a first-line treatment

SUMMARY

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

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

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