Approaching The Patient With Headache

By Jonathan P. Gladstone, BSc, MD, and John G. Edmeads, MD, FRCPC

In this article:

1. What are the crucial parts of taking the patient's history?
2. What are the key aspects of the physical exam?
3. What is the differential diagnosis?
4. What are the warning signs?

Headache is a common presenting complaint to family practitioners, with the lifetime prevalence of headache in the general population being over 90%.[1] The vast majority of headaches are attributable to a benign primary headache disorder and are not due to a sinister cause. Nonetheless, physicians need to be vigilant for secondary, possibly ominous, causes of headache. Familiarity with common and rare benign headache syndromes will help in the appropriate diagnosis and management of them.

A thorough history is the most critical aspect of the evaluation of headache (Table 1). The physical examination findings should be anticipated based upon the history, but may occasionally yield unexpected findings. A focused headache examination should include fundoscopy; visual fields; extraocular eye movements; assessment of strength and reflexes; coordination testing; and head and neck palpation (Table 2).

What is the differential diagnosis?

A useful approach is to divide headache types according to a temporal pattern, as follows: episodic and self-limited; abrupt and fulminant; and subacute or chronic. Each temporal pattern generates a unique differential diagnosis, and a different approach to investigation and treatment.

Episodic and self-limited headache entities.

Episodic tension-type headache (ETTH). This is recognised by recurrent attacks of mild-to-moderate headaches, lasting from 30 minutes to seven days untreated.[2] ETTH is bilateral or holocranial, and often presents as a dull pressure or squeezing discomfort that is responsive to over-the-counter analgesics. It is the most common headache type, but due to its mild-to-moderate nature, it is infrequently the sole cause for visiting the family doctor; notwithstanding, it is frequently mentioned (amongst other issues) during office visits.

Migraine with or without aura. Migraine is a common, often incapacitating, headache disorder. It is characterised by episodic attacks of moderate-to-severe headache and various combinations of neurologic (*i.e.,* sensory, motor or speech auras) and gas-

Headache

Table 1
Getting the patient's history

Age at headache onset: most primary headache disorders begin during childhood, adolescence or early adulthood.
Precise headache location: hemicrania continua almost invariably remains on the same side.
Duration of pain: idiopathic stabbing headache lasts only seconds at a time.
Frequency and timing of attacks: *i.e.,* cluster attacks occur one to eight times per day, typically at predictable times.
Pain severity and quality: *i.e.,* tension-type headaches are rarely severe.
Associated systemic features: *i.e.,* arthralgia, myalgia and malaise in an elderly patient suggest temporal arteritis.
Precipitating and/or aggravating factors: *i.e.,* standing up worsens spontaneous intracranial hypotension and coughing or straining may precipitate a headache associated with an Arnold-Chiari malformation, upper cervical or other posterior fossa lesions.
Ameliorating factors: *i.e.,* sleep typically relieves migraine attacks.
Associated neurologic symptoms: *i.e.,* transient visual obscurations may be the first sign of idiopathic intracranial hypertension.
Family history: strong family history in migraineurs of migraine, motion sickness or episodic dizziness.
Recent travel or exposure to tropical disease: tuberculosis, lyme or cocciodiomycosis can cause indolent low-grade meningitis.
Past medical history: previously quiescent cancer can reappear with brain metastasis.
Previous abortive treatment trials: *i.e.,* cluster patients are typically refractory to narcotics.
Impact of the headache: an episodic, disabling headache occurring over many years is almost always migraine.

trointestinal (*i.e.,* nausea and vomiting) dysfunction.[3] Migraines occur in 18% of females and 6% of males, and are the most frequent headache entity prompting patients to consult their physicians.

Cluster headache. This type of headache is the most dramatic and excruciating of the primary headache disorders. Attacks are more common in males and typically begin in the third or fourth decades of life.[4] The pain is usually retro-orbital and spreads to the ipsilateral head and face. The pain is excruciating in intensity and boring in quality. Attacks last between 15 minutes and 180 minutes, and occur from once every other day to eight times per day. Headaches are associated with at least one of the following: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, ptosis or miosis. Headache attacks occur in clusters for days, weeks or even months, and then usually remit for extended periods.

Idiopathic stabbing headache. Commonly known as "ice-pick headache" or "jabs and jolts," idiopathic stabbing headache consists of ultrashort (*i.e.,* one to two seconds) stabbing pains, which often frighten patients. It is more common in females, with an average age of onset of 47 years. Location, frequency and pattern can vary, and there are no accompanying neurologic or autonomic symptoms.

Dr. Gladstone is neurology resident, University of Toronto, Toronto, Ontario.

Dr. Edmeads is professor, division of neurology, University of Toronto, and neurologist, Sunnybrook & Women's College Health Sciences Centre, Toronto, Ontario.

Hypnic headaches. An unusual headache group that wakes patients from sleep, hypnic headaches occur more commonly in females and individuals over age 50.[5] The headache usually lasts less than 60 minutes, is moderately intense and occurs without accompanying symptoms. Oftentimes, hypnic headaches occur several times per week at predictable times and then disappear. Caffeine before bed may be an effective treatment technique.

Paroxysmal hemicranias. This describes frequent, short-lasting headaches in the orbito-temporal region. Attacks last between two minutes and 45 minutes (average duration is 10 minutes) and occur five to 40 times per day (average occurrence is 10 times). Pain with paroxysmal hemicranias is typically unilateral and accompanied by autonomic features, such as lacrimation and nasal congestion. Like idiopathic stabbing headaches, these headaches are responsive to indomethacin.

Causes of acute onset, fulminant headaches.

Headaches in this category are abrupt in onset and reach maximal intensity over seconds to hours. These headaches are frightening and may prompt worried telephone calls to family doctors, unscheduled drop-in visits or emergency room visits. The etiologies of acute onset, fulminant headache are varied and include subarachnoid hemorrhage, infarction (*i.e.,* ischemic or hemorrhagic), subdural hematoma, arterial dissection, meningitis, venous sinus thrombosis and obstructive hydrocephalus. The workup is dictated by the rapidity of onset, presence or absence of associated neurologic symptoms, and fever or systemic symptoms.

Subarachnoid hemorrhage. This is an acute onset excruciating headache, reaching maximal intensity within seconds. It should be judged as being subarachnoid hemorrhage until proven otherwise. A computed tomography (CT) scan should be immediate and, if negative, a lumbar puncture (LP) shunt is mandatory. Cerebrospinal fluid (CSF) xanthochromia remains positive for 10 to 14 days. If the patient presents to you in a delayed fashion, urgent referral to a neurologist is warranted for consideration of magnetic resonance (MR) angiography or conventional

Practice Pointer

Cluster headache is the most dramatic and excruciating of the primary headache disorders. Attacks are more common in males and typically begin in the third or fourth decades of life.

Table 2
Key Aspects of the Physical Exam

Fundoscopy: optic disc blurring may indicate increased intracranial pressure due to pseudotumour cerebri, space-occupying lesions or venous sinus thrombosis.
Visual fields: a field cut may be the only clue to a tumour.
Extraocular eye movements: a sixth-nerve palsy may be a "false localizing sign" secondary to increased intracranial pressure.
Assessment of strength and reflexes: a subtle pronator drift, asymmetry of power or reflexes, or an upgoing toe are highly suspicious for intracranial pathology.
Co-ordination testing: dysmetria or ataxic gait are highly suggestive of a posterior fossa lesion.
Head and neck palpation: for blood vessel tenderness or muscle spasm. This will typically be normal, but patients expect their head to be touched.

angiography to rule out a sentinal leak from aneurysm or arteriovenous malformation.

Infarction. Up to 20% of ischemic strokes have accompanying headache. Occasionally, patients may complain of headache, but do not appreciate their visual field loss from an occipital stroke or sensory neglect from a parietal stroke. A thorough neurologic examination to screen for focality is warranted in all patients with a fulminant headache.

Subdural hematomas. They occur most commonly in the elderly and in alcoholics, in whom brain atrophy increases the potential subdural space and falls are frequent. Notably, subdurals may occur spontaneously, without trauma, secondary to anticoagulants/antiplatelet agents, a vascular malformation or idiopathic causes. Subdurals may present with headache alone or with accompanying focal or cognitive symptoms and should be easily identified on CT scan.

Arterial dissection. This may be idiopathic or secondary to neck trauma, or a hyperelastic connective tissue disorder. With carotid dissection, a painful Horner's syndrome may be the sole symptom (*i.e.,* retro-orbital pain with ptosis and miosis). Occipital or nuchal pain may be the sole manifestation of vertebral dissection, or it may be accompanied by evidence of a posterior circulation stroke. MR angiography or conventional angiography are the investigations of choice.

Hydrocephalus. Acute obstruction to CSF flow by a cyst or tumour can lead to acute hydrocephalus, presenting with headache or, more dramatically, with projectile vomiting or loss of consciousness. This will be obvious on CT scan, but the underlying cause may be better delineated by magnetic resonance imaging (MRI).

Practice Pointer

Up to 20% of ischemic strokes have accompanying headache. Occasionally, patients may complain of headache, but do not appreciate their visual field loss from an occipital stroke or sensory neglect from a parietal stroke. A thorough neurologic examination to screen for focality is warranted in all patients with a fulminant headache.

Causes of subacute or chronic headaches.

Subacute or chronic headache is a common and challenging presentation of headache. The differential diagnosis is considerable, and a detailed history and physical examination are paramount.[6]

Benign primary causes of chronic headache. Chronic tension-type headache (CTTH) is a diffuse, dull headache. It often involves the posterior head and neck or it may be non-localizable. CTTH arises in individuals with a history of ETTH and may be exacerbated by overuse of medication. Treatment with tricyclic antidepressants, in combination with non-pharmacologic approaches (*i.e.,* relaxation, biofeedback), is helpful.

Chronic (transformed) migraine usually begins with episodic migraine, beginning when the patient is in his/her teens or twenties. Headaches become more frequent and associated migrainous features (*i.e.,* nausea, phono-photophobia) become less prominent. It typically occurs in females, is frequently seen with medication overuse and may coexist with psychiatric comorbidity.

Hemicrania continua is a rare, unilateral, continuous headache that waxes and wanes without ever completely subsiding. Frequent severe exacerbations occur and are often associated with autonomic disturbances like ptosis, tearing and nasal congestion. It can be severely debilitating and typically patients see numerous physicians without relief. The headache is dramatically sensitive to indomethacin and patients previously on significant doses of narcotics can be headache-free for the first time in years after a three-day course of indomethacin. (A response to a trial of 75 mg on day one, 150 mg on day two and 225 mg on day three confirms or refutes the diagnosis.)

Secondary causes of subacute or chronic headaches.

There are a large number of disease processes that can present with chronic headache. The physician needs to be aware of the possibilities to detect the unfortunate patient, whose headache is not benign. A focused search for the red flags and an appropriate investigation will detect the vast majority of secondary headaches (Table 3).

Infectious causes. Bacterial (*i.e.,* lyme, tuberculosis), fungal (*i.e.,* coccidiomycosis), parasitic (*i.e.,* neurocysticercosis) and viral meningitis (*i.e.,* human immunodeficiency virus seroconversion) can cause subacute or chronic headache. Patients have a constant, moderate intensity headache with or without low-grade fever, neck stiffness or constitutional symptoms. CSF profile and cultures help differentiate. Viral meningitis typically occurs during the fall, with most cases due to entero or arbo viruses.

Sinusitis typically should be diagnosed only in patients with purulent discharge. In a patient without symptoms of nasal disease, however, sphenoid sinusitis may masquerade as an intractable

For a good move see page 100

Table 3
Headache Red Flags

Acute onset of unusually severe headache

Consider subarachnoid hemorrhage, infarct, bleed, dissection and hydrocephalus.

Change in headache pattern

Be wary of changes in quality, location, frequency or severity, and remember headache types can coexist.

Presence of fever or systemic symptoms, such as:

Meningitis/encephalitis: low-grade with viral, tuberculosis, fungal or parasitic infection.

Sinusitis: sphenoid sinusitis may occur without nasal symptoms.

Vasculitis: primary central nervous system or secondary to other inflammatory/rheumatologic conditions.

Temporal arteritis: weak, fatigued, vision loss, polymyalgia rheumatica.

Focal neurologic findings

Look for optic disc swelling, field cut, focal weakness and lack of co-ordination.

Onset age of older than 50 years

Consider temporal arteritis, brain tumour (*i.e.,* primary or secondary) or infarct.

Occurring de novo in the peri-partum period

Consider venous sinus thrombosis or idiopathic intracranial hypertension.

Precipitated or exacerbated by positional changes, valsalva, bending or coughing

If worse when standing, think spontaneous intracranial hypotension.

If worse when supine, or with valsalva, think increased intracranial pressure and/or posterior fossa abnormality.

Progressive headache or escalating medication requirements

Re-evaluate original diagnosis, and consider a secondary cause.

Be wary of caffeine or medication overuse.

History of cancer, immunocompromise or human immunodeficiency virus

Be wary of metastatic disease or cerebral infection.

headache, unresponsive to analgesics and interfering with sleep. All CT scans for chronic headache patients should include sinus views.

Disorders of intracranial pressure. Idiopathic intracranial hypertension (IIH, also known as benign intracranial hypertension or pseudotumour cerebri) presents around age 30, but can be seen in childhood and middle-age.[7] It is more common in females and the obese, but may occur in non-obese males. Headaches are often constant and moderately severe, without distinguishing features. Accompanying features of headaches may include transient visual obscurations (*i.e.,* blurring or blacking out of parts of the visual field) and intracranial noises (*i.e.,* tinnitus or whooshing sounds). These may be elicited only upon specific probing. Secondary causes include venous sinus thrombosis, tetracycline/minocycline and vitamin A derivatives.

Identifying possible IIH and initiating prompt referral is critical, as visual loss can be irreversible. Its workup includes: ophthalmologic consultation to look for papilledema and establish baseline visual acuity and fields (papilledema is rarely absent); neurologic consultation (to screen for secondary causes and facilitate investigations); MRI (looking for space-occupying lesions); magnetic resonance venography (MRV) (looking for sinus thrombosis); and LP (looking for elevated opening pressure).

Spontaneous intracranial hypotension has recently been described and is analogous to post-LP headaches.[8] It is caused by a spontaneous tear in the dura along the spinal axis that leads to a continuous, tiny CSF leak. Traction on the brain's pain-sensitive structures then creates the head pain. A moderately severe headache will develop abruptly or subacutely, and is relieved by lying down and exacerbated by standing or walking. It is vital to trace headaches back to their onset, as postural components may blur or disappear over time. LP may show low pressure and MRI with gadolinium can show diffuse meningeal enhancement and downward displacement of the cerebellar tonsils.

Miscellaneous others. Excessive or daily consumption of analgesics (especially with caffeine, codeine, opiates or butalbital) can foster or perpetuate headaches. Obstructive sleep apnea may lead to headaches upon awakening. Severe hypertension occasionally contributes to headaches. Cervical spine, temporomandibular joint (TMJ) syndrome or dental pathology can lead to chronic cranial, nucchal or facial pain.[9] CME

Take-home message

Headaches are an extremely common complaint, with the vast majority being benign. An awareness of the common and rare benign primary headache types, combined with a focused approach to the red flags for sinister secondary causes of headaches, will aid clinicians in their day-to-day approach to the patient presenting with headache.

References

1. Mannix LK: Epidemiology and impact of primary headache disorders. Med Clin North Am 2001; 85(4):887-95.
2. Schulman EA: Overview of tension-type headache. Curr Pain Headache Rep 2001; 5(5):454-62.
3. Goadsby PJ, Lipton RB, Ferrari MD: Migraine: Current understanding and treatment. N Engl J Med 2002; 346(4):157-70.4.
4. Dodick DW, Capobianco DJ: Treatment and management of cluster headache. Curr Pain Headache Rep 2001; 5(1):83-91.
5. Dodick DW, Mosek AC, Campbell JK: The hypnic ("alarm clock") headache syndrome. Cephalalgia 1998; 18:152-6.
6. Silberstein SD, Lipton RB: Chronic daily headache. Curr Opin Neurology 2000; 13:277-88.
7. Digre K: Idiopathic intracranial hypertension headache. Curr Pain Headache Rep 2001; 6:217-35.
8. Mokri B: Spontaneous intracranial hypotension. Curr Pain Headache Rep 2001; 5:284-90.
9. Edmeads J: The cervical spine and headache. Neurology 1988; 38:1874-8.