

1 Overview of Common and Important Headache Disorders

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Abstract: The purpose of this chapter is to provide a summary of headache disorders that are common or important in primary care. Later chapters cover all aspects in greater detail.

The key headache disorders are primary – migraine, tension-type headache, and cluster headache. A relatively small number of secondary headache disorders may be seen in primary care and are important because they are serious and need to be recognized.

Collectively, headache disorders are common and ubiquitous. They have a neurological basis, but headache rarely signals serious underlying illness. The huge public-health importance of headache arises from its causal association with personal and societal burdens of pain, disability, damaged quality of life, and financial cost.

Headache disorders have many types and subtypes, but a very small number of them impose almost all of these burdens. Most of these can be effectively treated. They are diagnosed clinically, requiring no special investigations. Their management belongs in primary care.

Mismanagement and overuse of medications to treat acute headache are major risk factors for disease aggravation.

Headache

Headache is a painful feature of a relatively small number of primary headache disorders that in many cases are lifelong conditions. Headache also occurs as a characteristic symptom of many other conditions; these are termed secondary headache disorders. Although the International Headache Society classifies almost 200 distinct headache disorders – under 14 headings, the first four of which cover the primary headache disorders (▶ [Table 1.1](#)) – most are not of significance in primary care. A few are, and must be recognized, because they are serious.

There are regional variations, but headache disorders are highly prevalent throughout the world. Collectively, they are among the most common disorders of the nervous system. They affect people of all ages, races, income levels, and geographical areas. Furthermore, some of them cause substantial disability, which is imposed on all populations along with a very considerable socioeconomic burden.

Despite this, headache is underestimated in scope and scale, and headache disorders remain under-recognized and undertreated everywhere.

Migraine

Migraine is a primary headache disorder, probably with a genetic basis. Activation of a mechanism deep in the brain causes release of pain-producing inflammatory substances around the nerves and blood vessels of the head. Why this happens periodically and what brings the process to an end in spontaneous resolution of attacks are uncertain.

Usually starting at puberty, migraine is recurrent throughout life in many cases. Adults with migraine describe episodic attacks with specific features (▶ [Table 1.2](#)), of which headache and nausea are the most characteristic. In children, attacks tend to be shorter-lasting and abdominal symptoms more prominent. Attack frequency is typically once or twice a month but can be anywhere between once a year and once a week, often subject to lifestyle and environmental factors.

■ Table 1.1

The international classification of headache disorders (International Headache Society Classification Subcommittee 2004)

Primary headaches	1. Migraine, <i>including</i> : 1.1 Migraine without aura 1.2 Migraine with aura	
	2. Tension-type headache, <i>including</i> : 2.1 Infrequent episodic tension-type headache 2.2 Frequent episodic tension-type headache 2.3 Chronic tension-type headache	
	3. Cluster headache and other trigeminal autonomic cephalalgias, <i>including</i> : 3.1 Cluster headache	
	4. Other primary headaches	
Secondary headaches	5. Headache attributed to head and/or neck trauma, <i>including</i> : 5.2 Chronic posttraumatic headache	
	6. Headache attributed to cranial or cervical vascular disorder, <i>including</i> : 6.2.2 Headache attributed to subarachnoid hemorrhage 6.4.1 Headache attributed to giant cell arteritis	
	7. Headache attributed to nonvascular intracranial disorder, <i>including</i> : 7.1.1 Headache attributed to idiopathic intracranial hypertension 7.4 Headache attributed to intracranial neoplasm	
	8. Headache attributed to a substance or its withdrawal, <i>including</i> : 8.1.3 Carbon monoxide-induced headache 8.1.4 Alcohol-induced headache 8.2 Medication-overuse headache 8.2.1 Ergotamine-overuse headache 8.2.2 Triptan-overuse headache 8.2.3 Analgesic-overuse headache	
	9. Headache attributed to infection, <i>including</i> : 9.1 Headache attributed to intracranial infection	
	10. Headache attributed to disorder of homeostasis	
	11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures, <i>including</i> : 11.2.1 Cervicogenic headache 11.3.1 Headache attributed to acute glaucoma	
	12. Headache attributed to psychiatric disorder	
	Neuralgias and other headaches	13. Cranial neuralgias, central and primary facial pain and other headaches, <i>including</i> : 13.1 Trigeminal neuralgia
		14. Other headache, cranial neuralgia, central or primary facial pain

■ **Table 1.2**

Typical features of adult migraine headache

Headache	Moderate or severe in intensity One-sided and/or pulsating Aggravated by routine physical activity
Duration	Hours to 2–3 days
Accompanying symptoms	Nausea and sometimes vomiting and/or dislike or intolerance of normal levels of light and sound

Migraine is most disabling to those aged 35–45 years, but it can trouble much younger people, including children. In Europe and the USA, migraine affects 6–8% of men and 15–18% of women. The higher rates in women are hormonally driven. Similar patterns probably exist in Central and South America, with prevalences only slightly lower. In India, migraine is likely to be common: high temperatures and light levels for more than 8 months of the year, the heavy noise pollution and the Indian habits of omitting breakfast, frequent fasting, and eating rich, spicy, and fermented food are thought to be common triggers. Migraine appears less prevalent, but still common, elsewhere in Asia (around 8–10%) and in Africa (3–7%). In these areas, population-based studies are now being performed.

Tension-Type Headache

The mechanism of tension-type headache is poorly understood although it has long been regarded as a headache with muscular origins. It may be stress-related or associated with musculoskeletal problems in the neck.

Tension-type headache has distinct subtypes. Episodic tension-type headache, like migraine, occurs in attack-like episodes. These usually last no more than a few hours but can persist for several days. Chronic tension-type headache, one of the chronic daily headache syndromes, is present most of the time and can be unremitting over long periods. It is less common, but much more disabling to those affected.

Headache in either case is usually mild or moderate and generalized, though it can be one-sided. It is described as pressure or tightness, like a band around the head, sometimes spreading into or from the neck. It lacks the specific features and associated symptoms of migraine.

Tension-type headache pursues a highly variable course, often beginning during the teenage years and reaching peak levels in the 30s. It affects three women to every two men. Episodic tension-type headache is the most common headache disorder, reported by over 70% of some populations although its prevalence appears to vary greatly worldwide. In Japan, for example, 22% of the population report this disorder, while a prevalence of only 3% has been recorded in a rural population of Saudi Arabia. Lack of reporting and underdiagnosis are likely factors here, and cultural attitudes to reporting a relatively minor complaint may explain at least part of the variation elsewhere. Chronic tension-type headache affects 1–3% of adults.

Cluster Headache

Cluster headache is one of a group of primary headache disorders (trigeminal autonomic cephalalgias) of uncertain mechanism that are characterized by frequently recurring, short-lasting but extremely severe headache.

Cluster headache also has episodic and chronic forms. Episodic cluster headache occurs in bouts (clusters), typically of 6–12 weeks' duration once a year or 2 years and at the same time of the year. Strictly one-sided intense pain develops around the eye once or more times daily, mostly at night. Unable to stay in bed, the affected person agitatedly paces the room, even going outdoors, until the pain diminishes after 30–60 min. The eye is red and waters, the nose runs or is blocked on the affected side, and the eyelid may droop. In the less common chronic cluster headache, there are no remissions between clusters. The episodic form can become chronic, and vice versa.

Though relatively uncommon, affecting no more than 3 per 1,000 adults, cluster headache is highly recognizable. It is unusual among primary headache disorders in affecting six men to each woman. Most people developing cluster headache are in their 20s or older. Once present, the condition may persist intermittently for 40 years or more.

Medication-Overuse Headache

Chronic excessive use of medication to treat headache is the cause of medication-overuse headache, another of the chronic daily headache syndromes.

Medication-overuse headache is oppressive, persistent, and often at its worst on awakening in the morning. A typical history begins with episodic headache – migraine or tension-type headache – treated with an analgesic or other medication for each attack. Over time, headache episodes become more frequent, as does medication intake. In the end stage, which not all patients reach, headache persists all day, fluctuating with medication use repeated every few hours. This evolution occurs over a few weeks or much, much longer. A common and probably key factor at some stage in the development of medication-overuse headache is a switch to preemptive use of medication, in anticipation of the headache.

All medications for the acute or symptomatic treatment of headache, in overuse, are associated with this problem, but what constitutes overuse is not clear in individual cases. Suggested limits are the regular intake of simple analgesics on 15 or more days per month or of codeine- or barbiturate-containing combination analgesics, ergotamine or triptans, or any combination of these, on more than 10 days a month. Frequency of use is important: even when the total quantities are similar, low daily doses carry greater risk than larger weekly doses.

In terms of prevalence, medication-overuse headache far outweighs all other secondary headaches. It affects more than 1% of some populations, women more than men, and children also.

Serious Secondary Headaches (Headaches to Worry About)

Some headaches signal serious underlying disorders. These may demand immediate intervention. Although relatively uncommon, they worry nonspecialists because they are in the

differential diagnosis of primary headache disorders. The reality is that intracranial lesions give rise to histories and physical signs that should bring them to mind.

A history indicative of raised intracranial pressure should first suggest *intracranial neoplasm*. Intracranial tumors rarely produce headache until quite large, when raised intracranial pressure is apparent in the history and, in all likelihood, focal neurological signs are present. Because of their infrequency, brain scanning is not justified as a routine investigation in patients with headache.

Meningitis and its associated headache occur in an obviously ill patient. The signs of fever and neck stiffness, later accompanied by nausea and disturbed consciousness, reveal the cause.

Subarachnoid hemorrhage is by far the most common cause of incapacitating headache of abrupt onset, often described as the worst headache ever. It is usually unilateral at onset and accompanied by nausea, vomiting, and impaired consciousness, but may be less severe and without associated signs. Neck stiffness may take some hours to develop. Subarachnoid hemorrhage is very serious (50% of patients die, often before arriving at hospital, and 50% of survivors are left disabled). Unless there is a clear history of similar uncomplicated episodes, headache with these characteristics demands urgent investigation.

New headache in any patient over 50 years of age should raise the suspicion of *giant cell (temporal) arteritis*. This condition is conspicuously associated with headache, which can be severe. The patient, who does not feel entirely well, may also complain of marked scalp tenderness. Jaw claudication is highly suggestive. This disorder must be recognized: there is major risk of blindness, preventable by immediate steroid treatment.

Primary angle-closure glaucoma, rare before middle age, may present dramatically with acute ocular hypertension, a painful red eye with the pupil mid-dilated and fixed, and, essentially, impaired vision. In other cases, headache or eye pain may be episodic and mild.

Idiopathic intracranial hypertension is a rare cause of headache not readily diagnosed on the history alone. Papilledema indicates the diagnosis in adults, but is not seen invariably in children with the condition.

More commonly encountered in the tropics are the acute infections, *viral encephalitis*, *malaria*, and *Dengue fever*, all of which can present with sudden severe headache with or without a neurological deficit. These conditions should be kept in mind wherever they are likely to occur.

Other disorders seen more in the tropics that may present with subacute or chronic headache are *tuberculosis*, *neurocysticercosis*, *neurosarcoidosis*, and *HIV-related infections*. These are often diagnosed only on imaging.

Other Headaches Common in Primary Care

Only a small number of other headache disorders are likely to be seen in primary care. Guidance on their diagnosis is given elsewhere in this Handbook.

Chronic posttraumatic headache, usually secondary to moderate or severe head injury, has no specific features but often occurs as part of the posttraumatic syndrome. This includes symptoms such as equilibrium disturbance, poor concentration, decreased work ability, irritability, depressive mood, and sleep disturbances.

Headache attributed to low cerebrospinal fluid (CSF) pressure has three subtypes, presenting similarly but distinguished by etiology. This headache may develop up to 5 days after lumbar

puncture (often resolving spontaneously within a week). Persistent CSF leakage may be caused by another clinical procedure, or by trauma. Low CSF pressure may also develop spontaneously, often with a history of trivial increase in intracranial pressure, such as occurs on vigorous coughing, or after a sudden drop in atmospheric pressure.

Classical trigeminal neuralgia is characterized by unilateral brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve (usually the second or third). These occur spontaneously, or are evoked by trivial stimuli such as washing, shaving, brushing the teeth, smoking, or talking. They commonly cause facial muscle spasm on the affected side (*tic douloureux*). Between paroxysms there may be no symptoms, or a dull background pain may persist.

Persistent idiopathic facial pain (formerly known as atypical facial pain) is deep and poorly localized, confined at onset to a limited area on one side of the face, and present daily for all or most of the day. It lacks the characteristics of a cranial neuralgia and is not attributable to another disorder (pain may be initiated by surgery or injury to the face, teeth or gums, but persists without any demonstrable local cause).

Over-Diagnosed Headaches

Headache should not be attributed to sinus disease in the absence of other symptoms indicative of it. Errors of refraction are overestimated as a cause of headache. Dental problems may cause jaw or facial pain but rarely headache.

Management and Prevention of Headache Disorders

Health-Care Policy

The volume of headache referrals to neurologists seen in better-resourced countries is difficult to justify (see [▶ Chap. 5](#)), and should not be repeated in countries where headache-related health services are being developed. The common headache disorders require no special investigation: they are diagnosed and managed with skills generally available to physicians. Management of headache therefore belongs in primary care for all but a very small minority of patients. Models of health care vary but, in most countries, primary care has an acknowledged and important role. It is a role founded on recognition that decisions in primary care take account of important patient-related factors such as family medical history and patients' individual expectations and values. The continuity and long-term relationships of primary care generate awareness of these, while promoting trust and satisfaction among patients.

Even in primary care, however, the needs of the headache patient are not met in the time usually allocated to a consultation in many health systems. Nurses and pharmacists can complement the delivery of health care by primary care physicians.

Successful management of headache disorders follows five essential steps:

- The affected person must seek medical treatment.
- A correct diagnosis should be made.

- The treatment offered must be appropriate to the diagnosis.
- The treatment should be taken as directed.
- The patient should be followed up to assess the outcome of treatment, which should be changed if necessary.

Headache Diagnosis

The key to getting the diagnosis right is sufficient time committed to a systematic history. This must highlight or elicit description of the characteristic features of the important headache disorders described above (► [Table 1.3](#)). Different headache types are not mutually exclusive: patients are often aware of more than one headache type, and a separate history should be taken for each.

The correct diagnosis is not always evident initially, especially when more than one headache disorder is present, but the history should awaken suspicion of the important secondary headaches. Once it is established that there is no serious underlying disorder, a diary kept for a few weeks to record the pattern of attacks, symptoms, and medication use will usually clarify the diagnosis.

Physical examination rarely reveals unexpected signs after an adequately taken history, but should include blood pressure measurement and a brief but comprehensive neurological examination including the optic fundi: more is not required unless the history

■ **Table 1.3**

An approach to the headache history

1. <i>How many different headache types does the patient experience?</i> Separate histories are necessary for each. It is reasonable to concentrate on the most bothersome to the patient but others should always attract some enquiry in case they are clinically important.	
2. <i>Time questions</i>	(a) Why consulting now? (b) How recent in onset? (c) How frequent, and what temporal pattern (especially distinguishing between episodic and daily or unremitting)? (d) How long lasting?
3. <i>Character questions</i>	(a) Intensity of pain (b) Nature and quality of pain (c) Site and spread of pain (d) Associated symptoms
4. <i>Cause questions</i>	(a) Predisposing and/or trigger factors (b) Aggravating and/or relieving factors (c) Family history of similar headache
5. <i>Response questions</i>	(a) What does the patient do during the headache? (b) How much is activity (function) limited or prevented? (c) What medication has been and is used, and in what manner?
6. <i>State of health between attacks</i>	(a) Completely well, or residual or persisting symptoms? (b) Concerns, anxieties, fears about recurrent attacks, and/or their cause

is suggestive. Examination of the head and neck may find muscle tenderness, limited range of movement or crepitation, which suggest a need for physical forms of treatment but not necessarily headache causation.

Investigations, including neuroimaging, rarely contribute to the diagnosis of headache when the history and examination have not suggested an underlying cause.

Realistic Objectives

There are few patients troubled by headache whose lives cannot be improved by the right management, with the objective of minimizing impairment of life and lifestyle (▶ [Table 1.4](#)). Cure is rarely a realistic aim in primary headache disorders, but people disabled by headache should not have unduly low expectations of what is achievable through optimum management. Medication-overuse headache and other secondary headaches are, at least in theory, resolved through treatment of the underlying cause.

Predisposing and Trigger Factors

Migraine in particular is subject to certain physiological and external environmental factors. While predisposing factors increase susceptibility to attacks, trigger factors may initiate them. The two may combine.

Attempts to control migraine by managing these factors are often disappointing. A few predisposing factors (stress, depression, anxiety, menopause, head or neck trauma) are well recognized but not always avoidable or treatable. Trigger factors are important and their influence real in some patients, although dietary sensitivities affect, at most, 20% of people with migraine. Other lifestyle and environmental trigger factors suggest people with migraine react adversely to change in routine. Many attacks have no obvious triggers and, again, those that are identified are not always avoidable. Diaries may be useful in detecting triggers but the process is complicated as triggers appear to be cumulative, jointly overflowing the “threshold” above which attacks are initiated. Too much effort in seeking triggers causes introspection and

■ **Table 1.4**

Seven elements of good headache management

1. Evident interest and investment of time to inform, explain, reassure, and educate
2. Correct and timely diagnosis
3. Agreed high but realistic objectives
4. Identification of predisposing and/or trigger factors and their avoidance through appropriate lifestyle modifications
5. Intervention (optimal management of most primary headaches combines adequate but not excessive use of effective and cost-effective pharmaceutical remedies with non-pharmacological approaches; secondary headaches generally require treatment of the underlying cause)
6. Follow-up to ensure optimum treatment has been established
7. Referral to specialist care when these measures fail

can be counterproductive. Enforced lifestyle change to avoid triggers can itself adversely affect quality of life.

In tension-type headache, stress may be obvious and likely to be etiologically implicated. Musculoskeletal involvement may be evident in the history or on examination. Sometimes, neither of these is apparent. In the Muslim world, a marked rise in tension-type headache incidence on the first day of fasting is observed in people ordinarily susceptible to headache.

Cluster headache is usually but not always a disease of smokers, many of them heavy. However, patients with cluster headache who still smoke cannot be promised that giving up will end or even improve their headaches. Alcohol potently triggers cluster headache and most patients have learnt to avoid it during clusters.

Patient Information

Patients who are informed about and understand their disorder, and the purpose, nature, and expected effects of treatments for it, can be expected generally to adhere better to recommendations and have better outcomes. In this Handbook, each of the sections on the major headache disorders concludes with suggested information to patients.

Therapeutic Intervention

The purpose of pharmacotherapy of primary headache, once nondrug measures have been fully exploited, is to control symptoms so that the impact of the disorder on each individual patient's life and lifestyle is minimized. This requires a therapeutic plan tailored for each patient, and patients with two or more coexisting headache disorders are likely to require separate plans for each disorder.

Use of drugs for headache should, where possible, follow local guidelines that take account of local resources. The following are general guidelines.

No details of treatment are included in this overview: later chapters cover these for each headache type. The following observations are worth making here.

Migraine

Most people with migraine require drugs for the acute attack. Large numbers manage themselves, with no more than symptomatic over-the-counter remedies, and for many this appears adequate. Advice on correct usage (formulation, timing, dosage) may improve outcomes.

For the remainder, the great majority should be perfectly well manageable in primary care. The goal of acute therapy – resolution of symptoms and full return of function within 2 h – is not attainable by everyone with drugs currently available. When symptom control with best acute therapy is inadequate, it can be supplemented with prophylactic medication aiming to reduce the number of attacks. Drugs in a range of pharmacological classes have limited but often useful prophylactic efficacy against migraine. The choice of agent is guided by local availability, but otherwise by comorbidities and contraindications. Because poor compliance is a major factor impairing effectiveness, drugs given once daily are preferable, all else being equal.

Tension-Type Headache

Reassurance and over-the-counter analgesics are usually sufficient for infrequent episodic tension-type headache. Most people with this condition manage themselves: it is self-limiting and, although it may be temporarily disabling, it rarely raises anxieties.

People consult doctors because of episodic tension-type headache when it is becoming frequent and, in all likelihood, no longer responding to painkillers. Symptomatic medication is contraindicated for tension-type headache occurring on more than 2 days/week: when it is already being taken at high frequency, a diagnosis of chronic tension-type headache rather than medication-overuse headache cannot be made with confidence.

Cluster Headache

Because of its relative rarity, cluster headache has a tendency to be misdiagnosed, sometimes for years. It is the one primary headache that may not be best managed in primary care, but the primary care physician has an important role not only in recognizing it at once but also in discouraging inappropriate “treatments” (tooth extraction is not infrequent).

Medication-Overuse Headache

Prevention is the ideal management of medication-overuse headache, with education the key factor: many patients with medication-overuse headache are otherwise unaware of it as a medical condition. Once this disorder has developed, early intervention is important since the long-term prognosis depends on the duration of medication overuse. Treatment is withdrawal of the suspected medication(s). Despite initial worsening, within 2 weeks usually, the headache shows signs of improvement, which continues for weeks to months; 50–75% of patients revert to their original episodic headache type.

Other Headaches

All of the serious secondary headaches described above require specialist referral. In most cases, this should be immediate or urgent.

Headache attributed to low CSF pressure is likely to require specialist intervention whatever its etiology if it persists beyond a week.

Chronic posttraumatic headache and persistent idiopathic facial pain may be difficult to manage, and generally require specialist care. This is true also of classical trigeminal neuralgia. Many, possibly most, patients with this condition have compression of the trigeminal root by tortuous or aberrant vessels for which surgery may be appropriate. Rarely this disorder occurs bilaterally, in which case a central cause such as multiple sclerosis must be considered.

Follow-Up, and Referral

Neither the first diagnosis, nor the first-proposed treatment plan, may be correct. Follow-up is essential.

For migraine and episodic tension-type headache, the interval to follow-up is usually determined by attack frequency. Acute treatment may need several trials before its effect can be judged. Prophylaxis generally achieves observable benefit only after 3–4 weeks (although adverse effects may occur sooner).

For chronic tension-type headache, follow-up provides often-needed psychological support while recovery is slow.

In medication-overuse headache, early review is essential once withdrawal from medication has begun in order to check that it is being achieved: Nothing is less helpful than discovering, 3 months later, that the patient ran into difficulties and gave up the attempt. During later follow-up, the underlying primary headache condition is likely to reemerge and require reevaluation and a new therapeutic plan. Most patients with medication-overuse headache require extended support: the relapse rate is around 40% within 5 years.

Urgent referral for specialist management is recommended at each onset of cluster headache. Weekly review is unlikely to be too frequent, and allows dosage incrementation of potentially toxic drugs to be as rapid as possible.

In all other cases, specialist referral is appropriate when the diagnosis remains (or becomes) unclear or these standard management options fail.

Acknowledgment

The text of this chapter is closely based on educational materials developed for primary-care physicians by *Lifting The Burden: the Global Campaign Against Headache*.

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