Part I

Principles of Headache: Primary and Secondary Headache Disorders

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The Basics of Headache Classification and Diagnosis

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Introduction

The causes of headache range from the short-lived and trivial (e.g., a hangover headache) to the intermittent and quality-of-life threatening (migraine), to the unremitting and life-threatening (subarachnoid hemorrhage headache). This broad range of etiologies is matched by the very high prevalence of headache in the general population. In combination, the diversity of causes and high prevalence mandate a systematic approach to classification and diagnosis. Classification refers to a set of categories with diagnostic rules that provide the framework for a clinical approach. Diagnosis is the process of applying the rules to individual patients, defining their place in the classification.

In this chapter, we present an approach to both headache classification and diagnosis. We begin by describing the classification for headache disorders (see Tables 1.1, 1.2, 1.3, and 1.4 in the relevant sections). We recommend a three-step diagnostic process. First, we emphasize the identification or exclusion of secondary headache disorders by history, physical examination, and judicious use of diagnostic tests (see Table 1.5 and Figure 1.1). Second, we consider four groups of primary headache disorders that are defined based on headache frequency and duration (see Table 1.6) and refer to these as primary headache syndromes. Finally, we emphasize the identification of specific disorders within syndromic groups.

Approach to classification

The classification system for headache disorders has evolved over the past 50 years. Good classification systems should be valid, reliable, generalizable, and complete. A valid system provides categories that correspond, as much as possible, to biologic reality. As a practical matter, a valid system should usefully predict prognosis, response to treatment, and pathobiology. In a reliable system, two clinicians seeing the same patient should assign the same diagnoses. A generalizable system should work in a variety of settings including population studies, primary care, specialty care, and clinical trials. Finally, a complete system has an appropriate category for every headache type.

The first modern attempt at headache classification, sponsored by the National Institute of Health in 1962, was undertaken by the Ad Hoc Committee on Headache Classification of Headache [1]. While the Ad Hoc Committee's

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descriptions of headache types, for the most part, stand to this day, disorders were described in terms of their typical features without explicit operational rules regarding the characteristics required to diagnose or exclude particular disorders. In the absence of such rules, diagnostic reliability is not possible.

For the past 25 years, headache classification efforts have been led by the International Headache Society, which has published two editions of its International Classification of Headache Disorders (known as ICHD-1 and -2) [2, 3]. A third edition (ICHD-3) is in development. Learning the lessons taught by the Diagnostic and Statistical Manual of Mental Disorders system in psychiatry, the ICHD system provides clear boundaries among the primary headache disorders (Table 1.1). Although it is based on both evidence and expert opinion, the effort to be explicit inevitably leads to rules that are somewhat arbitrary. For example, to diagnose migraine without aura, at least five headache attacks are required. A patient

Table 1.1. The classification system (modified) (numbers refer to the ICHD-2 code)

A. Primary headache disorders

1. Migraine
   - 1.1. Migraine without aura
   - 1.2. Migraine with aura
   - 1.3. Childhood periodic syndromes that are commonly precursors with migraine
   - 1.4. Retinal migraine
   - 1.5. Complications of migraine
   - 1.6. Probable migraine
2. Tension-type headache
   - 2.1. Infrequent episodic tension-type headache
   - 2.2. Frequent tension-type headache
   - 2.3. Chronic tension-type headache
   - 2.4. Probable tension-type headache
3. Cluster headache and other trigeminal autonomic cephalalgias (TAC)
   - 3.1. Cluster headache
     - 3.1.1. Episodic cluster headache
     - 3.1.2. Chronic cluster headache
   - 3.2. Paroxysmal hemicrania
     - 3.2.1. Episodic paroxysmal hemicrania
     - 3.2.2. Chronic paroxysmal hemicrania
   - 3.3. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
   - 3.4. Probable TAC
4. Other primary headaches
   - 4.1. Primary stabbing headache
   - 4.2. Primary cough headache
   - 4.3. Primary exertional headache
   - 4.4. Primary headache associated with sexual activity
   - 4.5. Hypnic Headache
   - 4.6. Primary thunderclap headache
   - 4.7. Hemicrania continua
   - 4.8. New daily persistent headache (NDPH)

B. Secondary headache disorders

5. Headache attributed to head and/or trauma
   - 5.1. Acute post-traumatic headache
   - 5.2. Chronic post-traumatic headache

Table 1.1. (*Continued*)

6. Headache attributed to cranial or cervical vascular disorder
   - 6.1. Headache attributed to ischemic stroke or transient ischemic attack
   - 6.2. Headache attributed to non-traumatic intracranial hemorrhage
   - 6.3. Headache attributed to unruptured vascular malformation
   - 6.4. Headache attributed to arteritis
   - 6.5. Carotid or vertebral artery pain
   - 6.6. Headache attributed to cerebral venous thrombosis
   - 6.7. Headache attributed to other intracranial vascular disorder
7. Headache attributed to non-vascular intracranial disorder
   - 7.1. Headache attributed to high cerebrospinal fluid pressure
   - 7.2. Headache attributed to low cerebrospinal fluid pressure
   - 7.3. Headache attributed to non-infectious inflammatory disease
   - 7.4. Headache attributed to intracranial neoplasm
   - 7.5. Headache attributed to intrathecal injection
   - 7.6. Headache attributed to epileptic seizure
   - 7.7. Headache attributed to Chiari malformation type I
   - 7.8. Syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis
   - 7.9. Headache attributed to other non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
   - 8.1. Headache induced by acute substance use or exposure
   - 8.2. Medication-overuse headache (MOH)
   - 8.3. Headache as an adverse event attributed to chronic medication
   - 8.4. Headache attributed to substance withdrawal
9. Headache attributed to infection
   - 9.1. Headache attributed to intracranial infection
   - 9.2. Headache attributed to systemic infection
   - 9.3. Headache attributed to HIV/AIDS
   - 9.4. Chronic post-infection headache
10. Headache attributed to disorder of homeostasis
    - 10.1. Headache attributed to hypoxia and/or hypercapnia
    - 10.2. Dialysis Headache
    - 10.3. Headache attributed to hypertension
    - 10.4. Headache attributed to hypothyroidism
    - 10.5. Headache attributed to fasting
    - 10.6. Cardiac cephalalgia
    - 10.7. Headache attributed to other disorder of homeostasis

C. Headache or facial pain attributed to disorder of cranial structures, psychiatric disorders, cranial neuralgias

11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures
    - 11.1. Headache attributed to disorder of cranial bone
    - 11.2. Headache attributed to disorder of neck
    - 11.3. Headache attributed to disorder of eyes
    - 11.4. Headache attributed to disorder of ears
    - 11.5. Headache attributed to rhinosinusitis
    - 11.6. Headache attributed to disorder of teeth, jaws or related structures
    - 11.7. Headache or facial pain attributed to temporomandibular joint disorder
    - 11.8. Headache attributed to other disorder of cranium, neck, eyes, ears, nose sinuses, teeth, mouth or other facial or cervical structures

(*Continued*)

Table 1.1. (*Continued*)

12. Headache attributed to psychiatric disorder
    - 12.1. Headache attributed to somatisation disorder
    - 12.2. Headache attributed to psychotic disorder
13. Cranial neuralgias and central causes of facial pain
    - 13.1. Trigeminal neuralgia
    - 13.2. Glossopharyngeal neuralgia
    - 13.3. Nervus intermedius neuralgia
    - 13.4. Superior laryngeal neuralgia
    - 13.5. Nasociliary neuralgia
    - 13.6. Supraorbital neuralgia
    - 13.7. Other terminal branch neuralgias
    - 13.8. Occipital neuralgia
    - 13.9. Neck-tongue syndrome
    - 13.10. External compression headache
    - 13.11. Cold-stimulus headache
    - 13.12. Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions
    - 13.13. Optic neuritis
    - 13.14. Ocular diabetic neuropathy
    - 13.15. Head or facial pain attributed to herpes zoster
    - 13.16. Tolosa-Hunt syndrome
    - 13.17. Ophthalmoplegic "migraine"
    - 13.18. Central causes of facial pain
    - 13.19. Other cranial neuralgia or other centrally mediated facial pain
14. Other headache, cranial neuralgia, central or primary facial pain
    - 14.1. Headache not elsewhere classified
    - 14.2. Headache unspecified

with four typical attacks of migraine and a family history of migraine almost certainly has migraine, although they may not meet full criteria for the disorder.

Since ICHD-1, the criteria have evolved, based on formal field testing and clinical experience [3]. Major improvements have been made in the area of completeness as important disorders not included in ICHD-1, such as chronic migraine, new daily persistent headache, and hypnic headache were added to ICHD-2, published in 2004 [3]. In the years that followed, as field testing revealed limitations to ICHD-2, proposed revisions have been published, which will culminate in ICHD-3. This chapter is based on ICHD-2 and the published proposals for revision that will inform the shape of ICHD-3.

The ICHD approach to classification defines three overarching categories: primary headache disorders, secondary headache disorders, and cranial neuralgias and facial pains. For the primary disorders, the headache disorder is the problem; there is no underlying disease. For secondary headaches, the headache is attributed to an underlying cause. In this chapter, the discussion of secondary headaches will deal mainly with the features that differentiate them from the primary headaches, i.e., the "red flags" that indicate the need for further diagnostic evaluation. The primary headaches are divided into four broad groups: migraine, tension-type headache (TTH), trigeminal autonomic cephalalgias (TACs; including cluster headache), and other uncommon primary headaches.

Approach to diagnosis

Headache history

Because most patients seeing their doctor for headache have normal medical and neurologic examinations, the history is essential for accurate diagnosis. We recommend beginning with an open-ended statement such as, "Tell me about your headaches and how they affect your life." Although many clinicians fear that such an open-ended statement will generate a very long and unhelpful narrative, most patients speak for less than 90 seconds and usually provide useful information about both the headache's features and the impact on their life. Studies show that the use of open-ended questions makes headache visits shorter and increases both clinician and patient satisfaction with the visit.

After the patient's narrative, directed questions are used to fill in the details. The age of onset of the headache and the accompanying circumstances are important. Ask about the features of the head pain, including the location (unilateral, bilateral, or focal), quality (throbbing, stabbing, or steady ache), and intensity of the pain (on a scale of 1 to 10). The duration and frequency of headaches are important. The timing of attacks may assist both diagnosis and therapy. What time in the 24-hour day do they occur; are they associated with menstruation; do they cluster in time?

☆ TIPS AND TRICKS

Patients often first describe their worst or most recent headache. They may rate the intensity as 10 on a scale of 1 to 10, and the duration as constant since last Tuesday. Ask about the range of intensity and the typical duration. Similarly, they may state that the headache lasts for 3 days and occurs three times per week. It then is useful to determine the number of headache-free days per week or per month.

Ask about possible warning or premonitory symptoms that typically precede the onset of the headache by hours or days. Typical premonitory features include changes in mood such as irritability, food cravings, and neck stiffness among others. Auras are focal neurologic symptoms that usually begin 15–60 minutes before the headache. Premonitory features and auras are most typical of migraine although not entirely specific. Symptoms accompanying the headache may include nausea, vomiting, photophobia, and phonophobia as well as tearing of the eye, conjunctival injection, and nasal discharge among others.

It is also helpful to understand factors associated with an increased chance of headache onset (triggers). Avoidance or management of triggers becomes a therapeutic strategy. Aggravating factors increase the severity of a preexisting headache but are not associated with an increased rate of headache initiation. Ameliorating factors provide diagnostic clues. For example, hibernation in a dark, quiet room suggests a diagnosis of migraine. Understanding the effects of treatment and the course of the headache over time is also helpful.

☆ TIPS AND TRICKS

Patients often list their prescription medication, but fail to mention the over-the-counter (OTC) medications they use. OTC medications and dietary caffeine are important causes of medication overuse headache. After the history of the present illness has been obtained, other historical factors should be elicited. Is there a past history of illness, surgery, trauma, depression, or anxiety? Family history should include questions on other members with headache. Social history factors relating to marital status, education, occupation, interests, habits (alcohol, nicotine, and illicit drugs), sleep patterns, childhood abuse, and past and present stressors may all be relevant.

Diagnosing headache is as easy as 1, 2, 3

The first step in evaluating a patient with headache is to differentiate primary from secondary headaches. Secondary headaches are attributable to an underlying neurologic, medical, or psychiatric illness (Figure 1.1). Secondary headache is suspected if red flags on the history or examination suggest it. Primary headache disorders are likely if red flags are absent or if diagnostic testing excludes secondary headache. In addition, if a

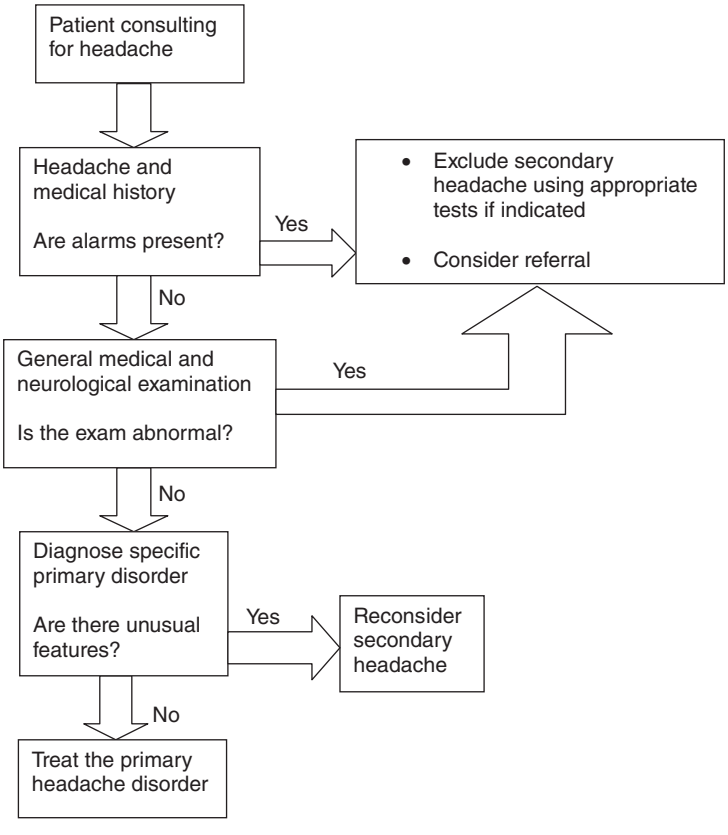

Figure 1.1. Headache diagnosis. (Reproduced from *Wolff's Headache and Other Pain*, 8th edition by Lipton et al. (2008) [15]. By permission of Oxford University Press, USA.)

secondary cause of headache is present but temporally remote from a pre-existing headache disorder, both primary and secondary headaches are likely. Consider a woman with typical attacks of migraine with aura from age 10 who develops a new form of daily headache at age 70. The MRI reveals a meningioma. Most likely, the patient has migraine with aura and a secondary headache attributed to her meningioma.

☆ TIPS AND TRICKS

When a headache changes in intensity or frequency, or in another feature, it may be a worsening of the primary headache, but one must always consider the superimposition of a new secondary headache. Searching for a new potential cause is warranted.

In making the diagnosis of a primary headache, the history and physical and neurologic examinations do not suggest secondary headache, or the latter can be ruled out either by investigations or by a distant temporal relationship (see Table 1.5). Symptoms that point to a specific primary headache but may not include all of the diagnostic criteria listed in ICHD-2 are preceded by the title "probable," for example "probable migraine." If the patient has two or more types of headache contemporaneously, all should be classified. The presence of a primary headache does not, of course, exclude the development of a secondary headache. Inattention to that obvious fact is a common and sometimes fatal error.

After excluding a secondary headache disorder, the next step is to identify a primary headache syndrome. Common headache syndromes and the specific disorders to consider within each syndromic group are summarized in Table 1.6.

These syndromes are defined based on the number of headache days per month and on the average duration of attacks. Based on headache days, two examples with long and short frequencies are chronic TTH, with 15 or more days per month, and episodic TTH with fewer than 15 headache days per month. Based on duration, we consider short-duration attacks as lasting less than 4 hours and long-duration attacks as lasting for 4 hours or more, for example cluster headache and chronic migraine, respectively. Some ICHD-2 defined disorders are included in more than one syndromic group. Most people with migraine have episodic headache of a long duration, but a clinically important subgroup have chronic migraine, the most important cause of chronic daily headache with a long attack duration.

Having identified the syndrome, the next task is to diagnose the specific disorder within the syndromic group. In the sections that follow, we discuss a number of specific primary headache disorders.

Migraine (ICHD-2 1.0)

Migraine is divided into two categories based on the characteristics of the attack: migraine without aura and migraine with aura. Attacks typically occur from less than once to several times a month. Migraine is also categorized based on headache days per month. When migraine occurs on 15 or more days per month for at least 3 months, the term "chronic migraine" is applied. The term "chronic" has several possible meanings in ordinary English and in medicine. As applied to migraine and TTH, it has the very specific meaning of headaches occurring on more days than not over at least 3 months. There are rare variants of migraine: retinal migraine, familial or sporadic hemiplegic migraine, and basilar-type migraine. Migraine is a very common phenomenon, affecting 12% of the population—18% of women and 6% of men in any given year. In contrast to past concepts, epidemiologic studies have shown that the prevalence of migraine varies inversely with socioeconomic status [4].

Migraine without aura

Migraine without aura (ICHD-2 1.1) represents about 75%–80% of all migraineurs. The criteria for migraine without aura are listed in Table 1.2. The typical features of an attack are a headache that is one-sided, pulsating, moderate or severe in pain intensity, lasting for hours, and associated with nausea and/or vomiting, photophobia, and phonophobia. In children, the duration is often shorter, 1 or 2 hours, and the associated features of nausea or vomiting or both are sometimes more prominent than the headache. Migraine attacks may not be typical but still fulfill the diagnostic criteria. For example, the location is often bilateral, and the quality may be a steady ache rather than pulsating. In the absence of these two features, the diagnosis is still appropriate if the two other features of the headache are present (pain of moderate or severe intensity and aggravation by physical activity) and other criteria are fulfilled. Similarly, only one of the two associated features—nausea and sensitively to light and sound—need be present. The frequency of attacks may vary from one per week to one per year. When the frequency increases to more than 15 days per month, the additional classification of chronic migraine is applied.

Migraine with aura

Migraine with aura (ICHD-2 1.2) (Table 1.2) occurs in about 20% of people with migraine. The most common aura of migraine is a positive homonymous visual phenomenon, a scintillating arc of zigzag lines that occurs for minutes before the onset of the headache and then completely disappears. Any neurologic symptom or sign may be an aura of migraine, but unusual conditions are more specifically subclassified as hemiplegic migraine, basilar-type (brainstem dysfunction) migraine, or retinal migraine (see "Subtypes of migraine"). The symptoms are usually positive, such as flickering lights or tingling sensations, in contrast to negative symptoms such as loss of vision or impaired sensation. The visual symptoms are typically homonymous, and the sensory symptoms usually affect the hand and face. These focal symptoms are most often on the side opposite the headache. Dysphasic speech is the least common aura manifestation. The aura usually evolves gradually and lasts from 5 to 60 minutes. Headache with features of migraine without aura usually begins as soon as the aura stops but sometimes begins at the same time as the aura and may begin as long as 1 hour after the aura. The aura may occur without headache, especially during middle or late age. Typical auras

Table 1.2. Diagnostic criteria for migraine without and with aura

ICHD-2 diagnostic criteria for 1.1 migraine without aura

A. At least 5 attacks fulfilling criteria B–D
B. Headache attacks last 4–72 hours (untreated or unsuccessfully treated)
C. Headache has at least 2 of the following characteristics:
   1. Unilateral location
   2. Pulsating quality
   3. Moderate or severe pain intensity
   4. Aggravation by, or causing avoidance of, routine physical activity (e.g., walking or climbing stairs)
D. During the headache attack, at least 1 of the following:
   1. Nausea and/or vomiting
   2. Photophobia and phonophobia
E. Symptoms not attributed to another disorder

ICHD-2 diagnostic criteria for 1.2 migraine with aura (modified)

A. At least 2 attacks fulfilling criteria B–D
B. Aura consists of at least 1 of the following:
   1. Fully reversible visual symptoms
   2. Fully reversible sensory symptoms
   3. Fully reversible dysphasic speech disturbance (not motor weakness)
C. At least 2 of the following:
   1. Homonymous visual symptoms and/or unilateral sensory symptoms
   2. At least one aura symptom develops gradually over more than 5 minutes and/or different aura symptoms occur in succession over 25 minutes
D. Headache fulfills criteria B–D for 1.1 migraine without aura
E. Symptoms not attributed to another disorder

sometimes occur with nonmigraine headaches and have been reported with cluster headache, chronic paroxysmal hemicranias, and hemicrania continua.

Cerebrovascular disease, especially transient ischemic attacks, may closely resemble the aura of migraine. Features that differentiate the two entities are the aura's slow evolution and the positive nature of the visual and sensory symptoms (e.g., scintillating scotomas rather than blindness). Investigations to rule out cerebrovascular disease are warranted if the symptoms are of rapid onset (less than 5 minutes), the visual or sensory symptoms are negative (blindness or sensory loss), the aura is prolonged (more than 1 hour), or the patient has risk factors for vascular disease including onset after the age of 50 [5].

Subtypes of migraine

There are several unusual forms of migraine named for the area of the affected nervous system. In all cases, the manifestation of these unusual auras is reversible.

Familial hemiplegic migraine (FHM) (ICHD-2 1.2.4), as the name implies, is manifested by hemiparesis sometimes with hemisensory impairment. Ataxia or other features of basilar-type migraine may occur. Unlike a typical aura, the hemiparesis may last for hours. This is the first headache syndrome linked to genetic polymorphisms. The loci for the condition are on chromosomes 1, 2, or 19 [6]. One or more first-degree relatives have similar attacks. People without a family history but otherwise meeting the criteria are classified as having sporadic hemiplegic migraine. The genetics of FHM are discussed in detail in Chapter 6.

In patients with *basilar-type migraine* (ICHD-2 1.2.6), the symptoms associated with the headache implicate the posterior fossa. The ICHD-2 classification requires at least two of the following aura symptoms: vertigo, tinnitus, decreased hearing, double vision, visual defects in both the temporal and nasal fields of both eyes, ataxia, dysarthria, bilateral paresthesias, or decreased consciousness.

Retinal migraine (ICHD-2 1.4) is manifested by scintillation, other positive visual symptoms, or more likely scotomas or partial or complete blindness limited to one eye (in contrast to common homonymous symptoms). Many of these patients experience a permanent partial defect representing retinal infarction [7]. Other causes of monocular visual loss (transient ischemic attacks, optic neuropathy, and retinal detachment) must be considered.

Childhood periodic syndromes are often precursors of migraine. *Cyclic vomiting* (ICHD-2 1.3.1) is manifested by intractable recurrent vomiting lasting for hours or days. Pallor and lethargy are usually associated. There is no evidence of gastrointestinal disease. Patients with *abdominal migraine* (ICHD-2 1.3.2) have attacks of abdominal pain associated with anorexia or nausea, with or without vomiting, and pallor. The ICHD-2 criteria require at least two of the three associated symptoms. Again, examination and studies rule out gastrointestinal disease. In patients with *benign paroxysmal vertigo of childhood* (ICHD-2 1.3.3), there are recurrent attacks (at least five) of severe vertigo that may last for minutes to hours; the attacks resolve spontaneously. Neurologic and vestibular functions are normal between attacks, as are imaging studies and electroencephalography. When this condition occurs after childhood, the patients often have bouts of migraine during or independent of vertigo. "Migrainous vertigo" or "vertiginous migraine" are not accepted entities in the ICHD-2 classification. Benign recurrent vertigo may occur at any age and often coexists with migraine. It is considered a comorbid condition.

Complications of migraine

Chronic migraine (ICHD-2 1.5.1) is defined as migraine headaches (usually without aura) or TTHs recurring on more than 15 days per month for more than 3 months [8]. At least 50% of the headaches should be migraine based on the presence of migraine features or response to migraine treatment. Chronic migraine evolves from episodic migraine. Many risk factors contribute to the evolution of chronic migraine from episodic migraine (see Chapter 5). Risk factors include frequent episodic headaches, high levels of disability, allodynia, depression, traumatic head injury, and overuse of drugs, most importantly opioid- and barbiturate-containing analgesics. The diagnosis of chronic migraine in the setting of medication overuse is complex (see Chapter 10). In practice, we treat medication overuse as a modifier, I.e., chronic migraine with medication overuse.

Status migrainosus (ICHD-2 1.5.2) is a term used for a severe and disabling migraine that lasts for more than 3 days. *Prolonged aura* is an extension of the patient's typical aura for more than 1 hour but less than 1 week. *Persistent aura without infarction* (ICHD-2 1.5.3) refers to persistence of an aura for more than 1 week. The aura is otherwise typical of past attacks. Evaluations do not reveal cerebral infarction. *Migrainous infarction* (ICHD-2 1.54) rarely occurs. The typical aura symptom or symptoms persist beyond 1 hour, and neurologic examination or neuroimaging or both confirms cerebral infarction. Other causes of stroke must be excluded.

Many patients exhibit all but one of the criteria for migraine. The term "probable migraine" (ICHD-2 1.6) is then appropriate.

TTH (ICHD-2 2.0)

This term replaces the former names of "tension headache" (implying emotional stress) and "muscle contraction headache" (implying a muscular origin of the headache). Although TTH is the most common primary headache, its mechanism is the least well understood. Many believe that TTHs are fragments of migraine. The diagnostic criteria are listed in Table 1.3. The headache characteristics are purposely nonmigrainous. The location is bilateral, the quality is nonpulsating, the intensity is mild to moderate, and the headache is not aggravated by physical activity. Similarly, there is a paucity of associated symptoms: no nausea or vomiting, although either photophobia or phonophobia may be present, but not both. There may or may not be associated pericranial tenderness.

TTHs are classified into episodic (fewer than 15 attacks per month) and chronic (more than 15 attacks per month). Episodic TTH is further subdivided into *infrequent episodic TTH* (ICHD-2 2.1) (less than 1 day per month) and *frequent episodic TTH* (ICHD-2 2.2) (1–14 days per month). *Chronic TTH* (ICHD-2 2.3) usually evolves from the episodic form and in its purest form should not be diagnosed in patients overusing acute medication. In practice, patients with medication overuse headache (a secondary headache)

Table 1.3. ICHD-2 Diagnostic criteria for 2.0 tension-type headache (TTH) (modified)

A. Episodes occuring in frequencies below (see 2.1, 2.2 and 2.3A)
B. Headache lasting from 30 minutes to 7 days
C. Headache has at least 2 of the following characteristics:
   1. Bilateral location
   2. Pressing/tightening (non-pulsating) quality
   3. Mild or moderate intensity
   4. Not aggravated by routine physical activity, such as walking or climbing upstairs
D. Both of the following:
   1. No nausea or vomiting (anorexia may occur)
   2. No more than one of the following: photophobia or phonophobia
E. Symptoms not attributed to another disorder

2.1 Infrequent episodic TTH
At least 10 episodes occurring on <1 day per month on average (<12 days per year) and fulfilling criteria B–D

2.2 Frequent episodic TTH
At least 10 episodes occurring on ≥1 but <15 days per month for at least 3 months (≥12 and <180 days per year) and fulfilling criteria B–D

2.3 Chronic TTH
A. Headache occurring on >15 days per month on average for >3 months (>180 days per year) and fulfilling criteria C–E (above)
B. Headache lasts hours or may be continuous

usually manifest the features of chronic TTH. As with the classification of migraine, the diagnosis of *probable TTH* (ICHD-2 2.4) is warranted if all but one of the ICHD-2 criteria are met. Another headache that phenotypically may resemble chronic TTH is new daily persistent headache (see "Other primary headaches").

TACs (ICHD-2 2.3.0)

The group of primary headaches known as TACs are characterized by trigeminal activation with unilateral pain typically affecting one orbit and ipsilateral autonomic activation (predominantly parasympathetic) [9]. The pain may spread to adjacent areas of one side and is usually locked to the same side on repeated attacks.

Cluster headache (ICHD-2 3.1) is the best known of the TACs, and the criteria for diagnosing it are noted in Table 1.4. The pain, usually in or around one orbit, is excruciating and is typically boring, sharp, or stabbing. It begins quickly and lasts from 15 minutes to 3 hours. During the attack, one or more of the following associated features are present: ipsilateral redness and tearing of the eye, clogging or drainage from the nostril, and ptosis and miosis. The attacks may occur between one and eight times per day, often

Table 1.4. ICHD-2 diagnostic criteria for cluster headache

3.0 Cluster headache (modified)
A. At least 5 attacks fulfilling criteria B–D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15–180 minutes if untreated
C. Headache is accompanied by at least 1 of the following:
   1. Ipsilateral conjunctival injection and/or lacrimation
   2. Ipsilateral nasal congestion and/or rhinorrhea
   3. Ipsilateral eyelid edema
   4. Ipsilateral forehead and facial sweating
   5. Ipsilateral miosis and/or ptosis
   6. A sense of restlessness or agitation
D. Attacks have a frequency from one every other day to 8 per day
E. Not attributed to another disorder

3.1 Episodic cluster headache occurs in periods lasting 7 days to 1 year

3.2 Chronic cluster headache occurs for more than 1 year without remission

awakening the patient from sleep. During the headache, in contrast to the behavior during migraine, the patient cannot lie still but is impelled to move about or sit and rock. Cluster headaches affect men more than women. These people are often heavy smokers.

The most common subtype is *episodic cluster headache* (ICHD-2 3.1.1), recurring every day for weeks or months (in clusters), and then remitting for one or more months annually, often recurring during the same season or seasons every year. During these cluster periods, alcoholic beverages and other vasodilating agents may trigger an attack. The unfortunate individuals who do not experience a remission within a year or have only a brief remission of less than 1 month are classified as having *chronic cluster headache* (ICHD-2 3.1.2).

Paroxysmal hemicranias, as the name implies, are unilateral headaches that are of short duration (2–30 minutes) and occur more than five times per day [9]. Associated with these are some or all of the autonomic features of cluster headache. The headaches invariably respond to prophylactic doses of indomethacin. The headaches usually occur without remission: *chronic paroxysmal hemicrania* (ICHD-2 3.2.2). Less frequent are remissions of 1 month or more: *episodic paroxysmal hemicrania* (ICHD-2 3.2.1).

Short-lived unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome (ICHD-2 3.3)—the name of this syndrome tells it all [10]. The headache and autonomic features are again similar to those of cluster headaches and paroxysmal hemicranias, but the duration is shorter (5 seconds to 4 minutes) and the frequency is higher (3–200 attacks per day).

Other primary headaches

Hemicrania continua (ICHD-2 4.7) is a daily and continuous unilateral headache that always responds to therapeutic doses of indomethacin [11]. The daily pain is of moderate intensity, but superimposed exacerbations often occur and may mimic migraine or cluster headache. Patients sometimes complain only of the migraine or cluster headache symptoms. A sensation of conjunctival irritation is often noted. Without inquiring about lesser daily headaches, the diagnosis of hemicrania continua may be missed.

☆ TIPS AND TRICKS

Patients often describe the headache that is most intense or disabling, such as migraine. Ask whether or not some headache or discomfort is present between the severe attacks. This may lead to the diagnosis of hemicrania continua with exacerbations that resemble a migraine-like headache.

Primary stabbing headaches (ICHD-2 4.1) are manifested by episodes of localized stabs of pain [12]. Formerly called "jabs and jolts," the pains last only one or a few seconds and occur at random without a consistent location. The frequency may range from a few per day to a few per month. Associated symptoms are lacking. These headaches are not uncommon in people with migraine.

Other primary headaches may be triggered by *cough*, *exertion*, or *sexual activity* (ICHD-2 4.2, ICHD-2 4.3, ICHD-2 4.4) and are labeled as such. Organic disease may cause headache evoked by these activities. Neuroimaging with special attention to the posterior fossa is mandatory to rule out cerebral aneurysm, subarachnoid hemorrhage, arterial dissection, and Arnold–Chiari malformation. Headache associated with sexual activity may be preorgasmic, causing a dull ache in the head and neck, or orgasmic, which often has an explosive quality.

Primary thunderclap headache (ICHD-2 4.6) is a severe headache of sudden onset [13]. The attack mimics the pain of subarachnoid hemorrhage, which must be excluded. The headache lasts from 1 to 10 days and may recur at random over weeks or months.

Hypnic headaches (ICHD-2 4.7) awaken patients from sleep, often at the same time each night. The pain is not specific and is without associated symptoms. Usually mild to moderate in intensity, the headache lasts about 30 minutes. It occurs mainly in late middle age or beyond.

New daily persistent headache (ICHD-2 4.8) was initially thought to mainly resemble chronic TTH. Recent evaluation, however, has revealed that most people with this condition have many features of migraine, and a modified classification has been proposed [14]. Patients can often

identify the exact day or days of onset. The headaches begin within 3 days of the noted day and are constant for more than 3 months. Acute medication overuse must be first considered, and chronic migraine should also be ruled out before the diagnosis is made.

Headache diagnosis

In beginning to formulate a diagnosis, the few steps in the algorithm of Figure 1.1 should be helpful. As noted before, headaches secondary to organic disease or dysfunction must be ruled out before a diagnosis of primary headache disorder is established. Neurologic workup is not necessary with every new patient, but if the headache is atypical or has recently changed or there is a history of red flags, a search for underlying disease is warranted (Table 1.5).

In formulating the differential diagnoses of the primary headaches, it is useful to follow a sequence of steps (Table 1.6). First, classify the headaches into low or high frequency (less or more than 15 headache days per month). Second, divide the headaches into those of short or long

Table 1.5. Alarms warranting further consideration

Red flag	Consider	Possible investigation(s)
Sudden-onset headache	Subarachnoid hemorrhage, bleed into mass or AVM, mass lesion (especially posterior fossa), pituitary apoplexy	Neuroimaging Lumbar puncture (after neuroimaging evaluation)
Worsening-pattern of headache (change in frequency or character)	Subdural hematoma, other mass lesion, medication overuse	Neuroimaging
Headache with systemic illness (fever, neck stiffness, rash)	Meningitis, encephalitis, Lyme disease, systemic infection, collagen vascular disease, arteritis	Neuroimaging Lumbar puncture Biopsy Blood tests
Focal neurologic signs, or symptoms other than typical visual or sensory aura	Mass lesion, AVM, collagen vascular disease	Neuroimaging Lumbar puncture (after neuroimaging evaluation) Blood tests
Papilledema	Mass lesion, idiopathic intracranial hypertension, encephalitis, meningitis	Neuroimaging Lumbar puncture (after neuroimaging evaluation)
Triggered by cough, exertion or sexual activity	Subarachnoid hemorrhage, mass lesion	Neuroimaging Consider lumbar puncture
Headache during pregnancy or post-partum	Cortical vein/cranial sinus thrombosis, carotid dissection, pituitary apoplexy	Neuroimaging
New headache type in a patient with cancer	Metastasis	Neuroimaging Lumbar puncture
Lyme disease	Meningoencephalitis	Neuroimaging Lumbar puncture
HIV	Opportunistic infection, tumor	Neuroimaging Lumbar puncture
Age of onset over 50	Organic disease: e.g., giant cell arteritis, mass lesion	Neuroimaging Erythrocyte sedimentation rate

AVM, arteriovenous malformation.

Table 1.6. Primary headache diagnosis by syndromic group

Frequency	Short duration (<4 hours)	Long duration (4 hours or more)
Low to moderate (<15 days per month)	Primary stabbing headache Thunderclap headache SUNCT syndrome Hypnic headache	Migraine Episodic TTH
Chronic (15 or more days per month)	Cluster headache and other TACs Paroxysmal hemicrania Primary stabbing headache Thunderclap headache SUNCT syndrome Hypnic headache	Chronic migraine Chronic TTH Hemicrania continua New daily persistent headache

duration (less than or more than 4 hours). Finally, consider headaches of short duration of low or high frequency and the presence or not of triggering factors. This approach should lead to the correct diagnosis for almost all patients.

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