Diagnosing Headache: Clinical Clues and Clinical Rules

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ABSTRACT

Headache is classified as primary, including migraine, tension-type headache, or cluster headache, or secondary — due to trauma, another systemic disorder, or another medical cause. Because the prognosis and treatment are so different for primary and secondary causes of headache, it is crucial to distinguish them at presentation. The International Headache Society classification and diagnostic guidelines are invaluable tools for evaluating the headache patient. Failing to correctly diagnose primary headache disorders has significant treatment implications. Prevalence rates, operational diagnostic criteria, and clinical pearls or clues can be used to facilitate accurate diagnosis.


Prevalence and Burden

The vast majority of people worldwide experience headache at least once during a lifetime. Several international studies have shown that the lifetime prevalence of headache ranges from 60% to 93% in men and from 82% to 99% among women. One-year prevalence rates by gender vary greatly by country, ranging from 40% to 91% for women and 19% to 84% for men. These dramatic variations in headache prevalence may reflect racial differences. For example, one study found the prevalence of migraine in the United States was highest in Caucasians, intermediate in African Americans, and lowest in Asian Americans.

Although headache and migraine are more common among adults, children are not immune. Investigators have reported that headache occurs in 4% of 3-year-olds. By 5 years of age, as many as 20% of children experience a headache, and the incidence increases to 47% among children 6 to 7 years of age. Between 7 and 15 years of age, 57% of children have had a headache and, in the same age group, the percentage of migraine sufferers approaches 10%, which is comparable to the percentage of adult migraine sufferers. In addition, the incidence of migraine among children has markedly increased during the past 3 decades.

The burden of headache is significant, especially among those with primary headache disorders such as migraine. Studies using the Health-Related Quality of Life and the Short Form-12 assessments reveal a substantially reduced quality of life among migraine patients compared to controls. Pooled data from studies conducted in the United States and United Kingdom have documented that the median pain
intensity reported by migraine patients is 8 on a 10-point scale. In comparison, median pain intensity scores range from 4 to 6 among patients with arthritis, inflammatory bowel disease, and other disorders accompanied by chronic pain. More than 90% of migraine patients report some degree of impairment: 53% describe severe impairment or require bed rest and 39% report a lesser degree of impairment. More than 90% of patients with primary headache disorders are also significant. For example, health insurance data from Germany confirm that the average cost per inpatient with a chronic pain disorder was highest among patients with tension-type headache, followed by those with basilar migraine. For every 1000 employees with migraine, approximately 270 working days are lost per year due to an inability to work. For tension-type headache, the morbidity burden is even higher: 920 working days lost per year for every 1000 employees with this diagnosis.

**Prevalence by Headache Type**

Overall, more than 90% of headache patients suffer from the primary headache disorders (migraine, tension-type, or cluster headache), while the remaining patients will have secondary headache caused by tumor, meningitis, giant cell arteritis, sinusitis, or other medical disorders. A population-based study of the lifetime prevalence of headache found that tension-type headache was the most common primary headache, affecting 78% of patients, while migraine affected 16%.

Patients presenting with severe headache pain are most likely to have migraine or another primary headache disorder. Intracranial pathology is extremely uncommon among patients with primary headache disorders. One large meta-analysis revealed that only 0.18% of patients diagnosed with migraine, who have a normal neurological examination, will be diagnosed with a significant intracranial abnormality.

An analysis of 3799 patients seen at a 24-hour emergency headache clinic demonstrated that 86% had primary headache and of those, 61% were diagnosed with migraine. Only 6.4% of patients presented with secondary headache; sinusitis was the most common cause (1.7%), followed by post-traumatic headache (1.5%), cerebral spinal fluid leak (0.5%), and vascular disorders (0.5%). There are more than 300 causes of secondary headache and the International Headache Society (IHS) criteria for diagnosis and classification unfortunately do not address all of them.

**Diagnostic and Classification Guidelines**

In 1988, the IHS published the first headache classification system that was accepted worldwide. These diagnostic and classification guidelines allow for a precise diagnosis of headache disorders. Among patients with primary headache, 70% will meet IHS diagnostic criteria for migraine; 23% for tension-type headache, and 7% for migrainous headache. However, misdiagnoses are common. In one analysis, 48% of those diagnosed with sinus-related headache actually had migraine, as did 33% of those diagnosed with tension-type headache.

The original guidelines are currently undergoing revision. The new classification and diagnostic guidelines, when approved, are expected to provide an even more specific description of the various primary headache disorders including migraine with and without aura, chronic migraine, tension-type and chronic tension-type headache, and cluster headache. An expanded discussion of the secondary causes of headache also will be included. The IHS guidelines referred to below are based on the currently proposed revisions to the original document.

**Diagnosis: Secondary Headache**

Because secondary causes of headache can be life-threatening, the first step in diagnosis is to exclude a secondary cause. A consultation for headache should include a history and physical examination, evaluation of clinical clues, and a determination of whether features are present that suggest an investigation of secondary headache, often referred to as the “SN O O P” features (Table 1).
When any of these features is present, appropriate laboratory and imaging investigations and referral to the appropriate specialist should be aggressively pursued. For example, for the patient who presents with a headache in the setting of progressive weight loss, fever, and jaw claudication, the sedimentation rate should be determined. If the results are positive, the patient should be referred to the appropriate specialists for a temporal artery biopsy and long-term management of giant cell arteritis. In some regions of the country, a referral will be made to a headache specialist and in others, to a neurologist.

To exclude a structural cause, electroencephalography (EEG) is not useful for routine evaluation of patients with headache. However, EEG may be a useful tool in patients with headache and alteration of consciousness, encephalopathy or focal neurological deficits, or atypical aura symptoms.22

The role of computed tomography (CT) or magnetic resonance imaging (MRI) is unclear in patients with primary headache. In those with recurrent migraine headache, neither CT nor MRI is warranted unless there has been a recent change in headache pattern, new onset seizures, or focal neurological signs or symptoms.23,24

For patients with secondary headache, some causes may not be evident with CT.

If cerebrovascular processes including arterial dissection, cerebral venous sinus thrombosis, or central nervous system (CNS) vasculitis are suspected, a gadolinium-enhanced MRI is indicated. Other indications for gadolinium-enhanced MRI include suspicion of herpes encephalitis, high and low intracranial pressure syndromes, and tumors of the posterior fossa, pituitary, and leptomeninges. Suspected arterial dissection should be investigated with magnetic resonance angiography, and cerebral venous sinus thrombosis should be investigated with magnetic resonance venography.

If cranial imaging is normal, lumbar puncture is indicated in cases of high or low intracranial pressure; thunderclap headache; subacute progressive headache; and headache associated with fever, meningismus, confusion, or seizure.

**Diagnosis: Primary Headache**

Migraine. As mentioned, the vast majority of patients will have a primary headache syndrome. If worrisome or atypical features are not present and a secondary cause can be excluded, the IHS diagnostic criteria should apply (Table 2). When interpreting the IHS diagnostic criteria, it is important to note that no single criterion is necessary or sufficient for diagnosis because any single criterion will be absent in up to 40% of patients. The IHS criteria do not require gastrointestinal symptoms for a migraine diagnosis; vomiting occurs in less than one third of patients with migraine. While most migraine attacks are unilateral, 40% of patients report bilateral pain. Approximately 50% of patients describe the migraine as nonpulsating, and only 15% of patients will have an associated neurological aura.

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### Table 2. Migraine Without Aura

| A. At least 2 attacks fulfilling conditions B - E |
| B. Fully reversible visual and/or sensory and/or speech symptoms but no motor weakness |
| C. At least 2 of the following: |
| - Unilateral symptoms including positive and/or negative features |
| - At least 1 symptom that develops gradually over >5 minutes and/or different symptoms that occur in succession |
| - Each symptom lasts >5 minutes and <60 minutes |
| D. Headache meets criteria for migraine without aura and begins during the aura or follows aura within 60 minutes |
| E. Not attributable to another disorder |

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### Table 3. Migraine Aura (Typical, Hemiplegic, Basilar)

| A. At least 2 attacks fulfilling conditions B - E |
| B. Fully reversible visual and/or sensory and/or speech symptoms but no motor weakness |
| C. At least 2 of the following: |
| - Unilateral symptoms including positive and/or negative features |
| - At least 1 symptom that develops gradually over >5 minutes and/or different symptoms that occur in succession |
| - Each symptom lasts >5 minutes and <60 minutes |
| D. Headache meets criteria for migraine without aura and begins during the aura or follows aura within 60 minutes |
| E. Not attributable to another disorder |

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Headache Classification Committee of the IHS.20
In practice, certain clinical features not necessarily included in the IHS criteria can be used to help solidify the migraine diagnosis. These include a temporal association with the menstrual cycle, which is frequent among migraine patients. Characteristic triggers such as thirst, or a food craving may be present before the attack. Family history of migraine may be present, and the patient may have premonitory symptoms. There may also be a paradoxical relationship with sleep; migraine frequently occurs during sleep or upon awakening but may also be abated by sleep.25

Migraine Aura. The IHS diagnostic criteria for migraine aura are shown in Table 3. It is important to note that more than one symptom can occur in succession so that a patient may have a visual aura followed sequentially by speech or sensory symptoms such as paraesthesia. Certain medical conditions can produce symptoms that mimic migraine aura. These include stroke and transient ischemic attack (TIA), seizure disorders, tumors, venous thrombosis, arteriovenous malformation, and carotid artery dissection. Particularly common may be the need to differentiate migraine aura from TIA (Table 4).26 The symptoms of TIA and other migraine aura mimics will usually violate several of the clinical rules for migraine diagnosis.

Tension-Type Headache. Tension-type headache can be differentiated from migraine using IHS criteria. Tension-type headaches may have a duration of between 30 minutes and 7 days and are not associated with nausea, photophobia, or phonophobia. In addition, at least 3 of the following 4 characteristics must be present for a tension-type headache diagnosis:

- A pressing/tightening, nonpulsating quality
- Mild to moderate intensity that does not prohibit activity
- Bilateral location
- Not aggravated by routine physical activity

Cluster Headache

According to current IHS criteria, cluster headache can be diagnosed when there have been at least 5 attacks of severe unilateral, orbital, supraorbital, or temporal pain lasting between 15 and 180 minutes without treatment. Frequency of cluster headache can range from 1 attack every other day to 8 attacks per day. There should be no suggestion of another headache disorder or a secondary cause. If another headache disorder is present concomitantly, the first presentation of cluster headache should not have occurred in close temporal association to the other headache disorder. If a secondary cause had been suspected, it must have been ruled out with the appropriate investigations. In addition, cluster headache must be associated with the patient experiencing at least 1 of the following symptoms on the side of the head where the pain occurs:

- Conjunctival injection
- Lacrimation
- Nasal congestion
- Rhinorrhea
- Forehead and/or facial swelling
- Miosis
- Ptosis
- Eyelid edema

Chronic Daily Headache

Any discussion of headache diagnosis and characterization must address “chronic daily headache.” While the term is commonly used in clinical practice, “chronic daily headache” is not a diagnosis. Patients either have a primary chronic headache or a secondary chronic headache that may be post-traumatic, postinfectious, cervicogenic, or due to vascular disorders or intracranial hypertension.27 A similar approach using clinical clues can help differentiate primary and secondary forms of chronic headache. It is estimated that between 4% and 5% of the U.S., European, and Asian population have primary chronic headache defined as occurring on more than 15 days of each month whether or not the patient is using medication. Primary chronic headaches cannot be related to another cause such as structural or systemic disease.28,29

<table>
<thead>
<tr>
<th>Table 4. Migraine Aura vs TIA</th>
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<tbody>
<tr>
<td><strong>Migraine</strong></td>
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<tr>
<td>Positive visual symptoms</td>
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<tr>
<td>Gradual onset/evolution</td>
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<tr>
<td>Sequential progression</td>
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<tr>
<td>Repetitive attacks of identical nature</td>
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<tr>
<td>Flurry of attacks mid-life</td>
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<tr>
<td>Duration up to 60 minutes</td>
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<td>Headache follows ~50%</td>
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</table>

A chronic headache of more than 4 hours' duration is most often primary headache, typically chronic migraine (Table 5) or tension-type headache. In clinical practice, approximately 78% of patients with chronic headache will have chronic migraine, and 15% will have chronic tension-type headache. Only 7% will have chronic headache due to another cause.28-30

Chronic headache of less than 4 hours' duration is often a secondary headache and may be due to paroxysmal hemicrania (spasmodic pain or aching on one side of the head), or hypnic headache, which occurs during sleep and causes frequent awakening.27 These patients typically present with an idiopathic stabbing headache. However, chronic daily headache of less than 4 hours also may be cluster headache.27

WHEN TO REFER THE PATIENT WITH HEADACHE

Certainly, whenever any of the warning symptoms or signs listed in Table 1 is present, and routine laboratory or imaging studies do not reveal an underlying cause, referral to a neurologist or headache specialist is appropriate. In addition, patients with a primary headache disorder should be referred when:

• The diagnosis remains unclear.
• Treatment with conventional acute or prophylactic management is not effective, tolerated, or contraindicated.
• Frequency of attacks exceeds 1 per week or attacks are prolonged and disabling despite the use of specific acute and preventive medications.

CASE STUDY

This case helps illustrate the use of the diagnostic criteria and clinical clues to accurately characterize a particular headache.

A 46-year-old woman with a history of migraine aura without headache pain reported 3 such episodes of 20-minutes duration each during the past 6 years. She presented to the emergency department with an abrupt aura consisting of a right, homonomous scintillating scotoma followed by left-sided headache with nausea and photophobia that had lasted 2 days.

The patient was postmenopausal and taking estrogen replacement therapy. Routine examination was normal and CT revealed no brain abnormalities. She received an initial diagnosis of migraine with aura. However, during a subsequent office visit, review of clinical clues and of “SNOOP” features (Table 1) raised the suspicion that a secondary etiology might be present. The patient had neurological symptoms manifesting as aura and while previous episodes of aura had lasted less than 20 minutes, this episode of aura was persistent, representing a change in pattern.

In addition, the characteristics of this patient’s current headache violated several of the clinical rules for migraine aura (Table 3). The neurological symptoms were not reversible, onset was abrupt rather than gradual, aura symptoms persisted for longer than 60 minutes, and there was a delay of more than 60 minutes from the time aura began to the time of the onset of headache pain.

Investigations were conducted to rule out conditions that may mimic migraine aura. However, a 4-vessel angiography revealed an irregularity in the left posterior cerebral artery. MRI revealed an increased diffusion-weighted signal in the medial left occipital cortex that was consistent with a subacute cerebral infarction. Thus, a secondary cause of this particular headache was confirmed.

CONCLUSION

In summary, the majority of headache cases seen in primary care will be due to a primary headache disorder such as migraine or tension-type headache. Primary headache syndromes are diagnosed by defining the clinical features of an individual’s attacks and viewing them in light of the definitions of headache type, and the clinical rules for diagnosis established by the IHS.

If care is taken to identify any worrisome features present during a particular headache attack, the chances of missing the diagnosis of a secondary headache caused by a serious disorder can be greatly diminished.

Table 5. Chronic Migraine

| A. Average migraine frequency ≥15 days per month for ≥3 months fulfilling B - D |
| B. Attacks fulfill criteria for migraine without aura |
| C. No overuse of symptomatic drugs |
| D. Not attributable to another disorder |

Headache Classification Committee of the IHS.24
REFERENCES