Chapter 1
Diagnosis of Migraine and Tension-Type Headaches

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Introduction to Diagnosis

Headache diagnosis in the office is predicated on deciding if the patient's headache is primary or secondary. With this determination, the clinician will know how to proceed. Aiding diagnosis is the use of the *International Classification of Headache Disorders,* third edition, beta version (ICHD-3), published in 2013. Parts I and II of this clinical manual cover the diagnosis of primary headaches of adults in the office, Part III covers diagnosis of secondary headaches, and Part IV covers diagnosis of pediatric headaches. The remaining parts of the book cover treatment. A knowledge of basic headache epidemiology, some familiarity with ICHD-3, some shortcuts and clinical pearls, and recognition of when to be worried for the possibility of sinister headaches will equip the clinician with a route to satisfactory diagnosis (Table 1.1).

Epidemiology of Primary Headaches

Primary headaches are very common, and the headache usually encountered in the office is migraine. Outside the doctor's office, tension-type headache (TTH) is by far the most common diagnosis in the general population. But in clinical practice, when a patient complains of episodic headache, the diagnosis is usually migraine.

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Table 1.1 Steps to quick, correct diagnosis of headaches

1. Know basic epidemiology of primary and secondary headaches
2. SNOOP: a mnemonic for secondary workup (see Chaps. 5 and 6 in Part III of this book)
3. ICHD-3 criteria
4. Pattern recognition
5. Brief screeners
6. Impact/disability-based diagnosis

Table 1.2 Basics of epidemiology of migraine

1. Migraine occurs in 12% of the general population, 18% female, 6% male
2. When a patient complains of a stable pattern of episodic, disabling headache in the office, the likelihood of migraine or probable migraine is greater than 90%

Migraine occurs in about 12% of the US population, 18% of females, 6% of males, numbers established in three large US population-based studies from 1989 to 2007. Thus, unless there are red flags present, migraine is the likely diagnosis of an office patient with a stable pattern of episodic, disabling headache.

One study in 14 countries of primary care offices and nonheadache specialists established that when a patient complained of episodic headache, either as a chief complaint, secondary complaint, or checked "headache" off on the review of systems, the diagnosis was migraine or probable migraine (PM) in 94% of the patients. The remaining 6% were evenly divided between TTH and other types of headache. It is a pretty good bet that in the absence of concerns for secondary headache discussed in Part III, migraine should be the default diagnosis in patients complaining of a stable pattern of episodic, disabling headache in clinical practice (see Table 1.2).

Reminder on the Red Flags of Headache Diagnosis

As noted, diagnosis of secondary headaches will be covered in Part III of this book. However, it is worth stating at the beginning that a workup of patients with red flags is necessary before diagnosing primary headaches. When in doubt, investigate the atypical.

Dr. David Dodick, Professor of Neurology at the Mayo Clinic, first published the use of a mnemonic for red flags suggesting sinister or secondary headaches. His mnemonic, which will be repeated in Chap. 4, tells the clinician when to "snoop" for secondary headache and is adapted in Table 1.3.

If the red flags are not noted, it is time to decide which primary headache is presenting.

Table 1.3 The SNOOP mnemonic for red flags for secondary headache. (Adapted from Dodick 2003)

*S*ystemic symptoms (fever, weight loss) or
*S*econdary risk factors: underlying disease (HIV, cancer, autoimmune disease)
*N*eurologic symptoms or abnormal signs (confusion, impaired alertness or consciousness, focal exam)
*O*nset: sudden, abrupt, or split-second (first, worst)
*O*lder age onset: new-onset and progressive headache, especially at age > 50 (giant cell arteritis, cancer)
*P*attern change: first headache or different, change from
*P*revious headache history: attack frequency, severity, or clinical features
SSNOOPP

Table 1.4 Migraine without aura, ICHD-3 criteria

1. Having had more than five attacks, the patient should meet the following criteria
2. Headaches last from 4 to 72 h
3. Any two of the following four
(a) Moderate to severe intensity
(b) Throbbing quality
(c) Worsened by physical activity
(d) Unilateral location
4. The headaches need to have any one of the following:
(a) Nausea
(b) Photophobia and phonophobia
5. Secondary causes eliminated (normal exam, imaging, etc.)

Diagnosis Using the ICHD-3

The ICHD-3 provides validated international criteria for diagnosing headaches. Previously, the ICHD-2 was adopted by the National Institutes of Health (NIH), the Food and Drug Administration (FDA), the World Health Organization (WHO), and all major clinical professional organizations in the USA, including the American Academy of Neurology (AAN). The plan is for the ICHD-3 to be connected to future versions of the International Classification of Diseases (ICD) billing codes. Using the ICHD-3 can be very helpful; when a patient does not fit the ICHD-3 criteria for a given primary headache disorder, it is time to contemplate the possibility that a secondary headache exists.

The ICHD-3 of the International Headache Society (IHS) is an extensive, detailed document, 400 pages long, and many doctors use pattern recognition or other short cuts to diagnosis instead. Nonetheless, careful scrutiny of IHS criteria can be very useful, especially in more atypical headache disorder presentations. The ICHD-3 criteria for migraine without aura are summarized in Table 1.4.

A few clinical pearls help with using the ICHD-3 criteria. Although migraine is often suggested by the company it keeps (menstrual, stress, red wine, or weather triggers, history of motion sickness, family history), triggers are not included in the strict criteria for diagnosis.

Table 1.5 Clinical pearls on diagnosing migraine without aura

1. Migraine can be suggested by the company it keeps
   (a) Menstrual trigger
   (b) Red wine trigger
   (c) Weather trigger
   (d) Stress trigger
2. Location is not included in the diagnostic criteria
   (a) Neck pain in migraine is very common
   (b) Bilateral location occurs in at least 40 % of migraine
3. Migraine attacks vary between patients and in the same patient across time
4. Response to triptans and ergots is not diagnostic of migraine
5. Migraine has negative impact on patients in their daily activities. Tension-type headaches do not generally result in disability
6. Many migraine patients have either a family history of "headaches," personal histories of motion sickness, especially in childhood, or both
7. Ninety-four percent of patients complaining in the office to primary care doctors of stable, episodic headaches had migraine or probable migraine. Only 3 % had tension-type headache as the primary diagnosis

Location is not included in the diagnostic criteria. For example, neck pain, often thought to suggest TTH, is present in at least 75 % of migraine patients. Forty percent of migraine is bilateral. Bilateral maxillary pain, often thought to suggest "sinus headache," is a nonspecific symptom. In other words, do not make a diagnosis by location of pain alone.

Migraine is variable both inter- and intrapatient across time. Severity of migraine can be moderate, location can be bilateral, quality can be nonthrobbing, and nausea and aura can both be absent. Many migraine patients have either a family history of "headaches," personal histories of motion sickness, especially in childhood, or both.

Response to medication does not prove diagnosis. Meningitis and subarachnoid hemorrhage pain can transiently respond to migraine-specific treatments, such as triptans and ergots, so response to triptans is not conclusive for migraine diagnosis or even a primary headache disorder. Cluster headaches also respond to both triptans and dihydroergotamine.

Migraine adversely affects patients in their daily activities, while TTH does not generally result in disability. The ICHD-3 checklist can assure that the default primary episodic headache diagnosis is accurate. Clinical pearls on diagnosing migraine without aura are summarized in Table 1.5.

Pattern Recognition Diagnosis of Migraine

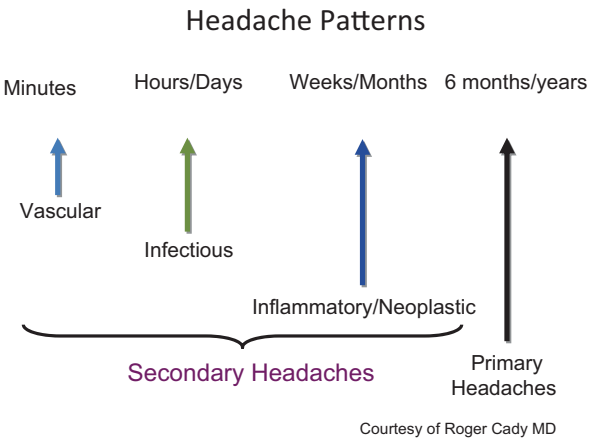
The duration of headache history can add to pattern recognition. Recent onset of headache should be of more concern (see Tables 1.6 and 1.7).

New and sudden headaches, often described as such as having thunderclap onset, raise the question of bleed. A presenting new headache of days' duration without a

Table 1.6 The aphorism of pattern recognition of migraine

• A patient presenting with a stable pattern of at least 6 months duration of episodic disabling headache has migraine until proven otherwise

Table 1.7 Headache patterns. (Courtesy of Roger Cady MD)



previous history of headaches raises the question of meningitis or encephalitis. New headaches of slow, progressive onset suggest neoplasm or vasculitis. And finally, the comfort of stable, episodic headaches of at least 6 months duration is the pattern of migraine.

Brief Screeners for Migraine Diagnosis

ID Migraine

Because some care providers find the ICHD-3 criteria too cumbersome, several brief screeners have been validated. The most important of these is ID Migraine, which consists of three questions: presence or absence of photophobia, presence or absence of nausea, and presence or absence of impact on activities. If the patient has the presence of 2/3 symptoms, ID Migraine has a sensitivity of 0.81 and a specificity of 0.75 (see Table 1.8).

Table 1.8 ID Migraine. (adapted from Lipton et al. 2003)

Yes or no answers
With your headaches
1. Do you have dislike of light?
2. Do you have nausea?
3. Do your headaches have impact on work, school, or recreational activities?
2/3 "yes" answers suggest migraine

Table 1.9 Migraine Disability Assessment Scale (MIDAS). (Adapted from Stewart et al. 1999)

MIDAS is a five-item questionnaire on headache disability which can be summarized as:

- "How many days in the last 3 months were you at least 50% disabled at work, home, school, or recreational activities?"
- Scores greater than 11 days suggest at least moderate disability and also suggest a diagnosis of migraine

Single Screener for Migraine: Nausea

Dr. Vincent Martin from the University of Cincinnati found that nausea alone, when associated with episodic headache, yields a sensitivity of 0.81 and a specificity of 0.83. So if your patient with a stable pattern of episodic disabling headache has nausea, that patient meets both 2/3 of the ID Migraine criteria and the single criterion. Brief screeners can be very useful at short-cutting to the diagnosis.

Impact-Based Diagnosis of Migraine

Impact is the third criterion of ID Migraine. Migraine is the recurring, episodic primary headache which causes disability and has impact. The impact of migraine is why the aphorism is for a stable pattern of at least 6 months of episodic, *disabling* migraine. TTH rarely has any impact at all.

Two screeners of disability or impact in episodic primary headache can indirectly suggest migraine. These are the Migraine Disability Assessment Scale (MIDAS) and the Headache Impact Test (HIT-6).

MIDAS uses a five-item questionnaire to ask the question, "How many days in the last 3 months were you at least 50% disabled by your headaches at work, home, school, or recreational activities?" (see Table 1.9). If the answer is greater than 11 days, migraine diagnosis is suggested.

HIT-6 uses questions in six domains to evaluate headache impact. If the HIT-6 score is greater than 60, migraine diagnosis is suggested.

Diagnosis of Tension-Type Headache

TTH was described by the late Dr. Fred Sheftell as the featureless headache. The diagnosis of TTH is made predicated on the fact that it is not migraine.

The ICHD-3 criteria for episodic TTH (ETTH) are summarized in Table 1.10.

The criteria for ETTH posit that it is not migraine: not unilateral, not throbbing, not severe, not worse with activity, no nausea, and generally no photophobia and no phonophobia. ETTH rarely causes any lasting impact. Patients rarely complain of it, and it is almost never seen in the office. As noted above, it is featureless. Also,

Table 1.10 Infrequent episodic tension-type headache, ICHD-3 criteria

1. At least ten episodes occurring less than once a month or less than 12 times per year average and fulfilling the following criteria
2. Headaches last from 30 min to 7 days
3. Headache has at least two of the following:
   (a) Not unilateral
   (b) Not throbbing
   (c) Mild or moderate intensity, not severe
   (d) Not aggravated by routine physical activity
4. Both of the following:
   (a) No nausea
   (b) No more than one of photophobia or phonophobia or neither
5. Not secondary

Table 1.11 Migraine without aura/(*episodic tension-type headache, ETTH*)

1. At least five (*ten*) attacks lasting 4–72 h (*30 min to 7 days*) with
2. At least two of the following four
   (a) Unilateral (*bilateral*)
   (b) Pulsating (*not pulsating*)
   (c) Moderate to severe intensity, inhibits or prohibits activities (*mild to moderate*). *Note, migraine has impact, not TTH!*
   (d) Physical activity aggravates (*does not aggravate*). *Note, migraine, not TTH has impact!*
3. At least one of the following
   (a) Nausea and/or vomiting (*no nausea or vomiting*)
   (b) Photophobia and phonophobia (*one or neither*)
4. Both have normal history, exam, or imaging test

note that the ICHD-3 criteria do not mention location or triggers, so ETTH is not diagnosed by a neck location or a stress trigger.

The ICHD-3 classification differentiates infrequent ETTH (episodes occurring on < 1 day per month on average, < 12 days per year) and frequent ETTH (occurring on ≥ 1 but < 15 days per month for at least 3 months, ≥ 12 and < 180 days per year). Some European headache clinicians think of patients with chronic daily headache (CDH) as having a background of ETTH punctuated by episodes of migraine, while American headache specialists believe that CDH is usually chronic migraine (see Chap. 4).

The ICHD-3 also differentiates ETTH with and without pericranial tenderness. It is not clear that this distinction has any clinical importance at all.

Migraine vs. ETTH

Migraine can be distinguished from ETTH using Table 1.11, which lists the features of migraine without aura, followed by the characteristics of ETTH in parentheses and italics.

Table 1.12 Clinical pearls on the diagnosis of probable migraine

- A "probable" diagnosis in the ICHD-3 means that a primary headache disorder is missing one criterion
- Diagnosis of "probable" should trigger the clinician to consider the possibility that the patient has secondary headache, rather than a primary headache disorder, because the patient does not meet all of the IHS criteria for a given primary headache

Diagnosis of Probable Migraine

Probable migraine (PM) is the term used by the ICHD-3 for migraine missing one criterion. For example, a patient has more than five headaches lasting 24 h, which are bilateral, nonthrobbing, and of moderate intensity, worse with activity, with photophobia but no phonophobia, thus missing one of the "D" criteria.

The ICHD-3 instructs clinicians to diagnose based on the highest *complete* set of criteria, so that patients who meet criteria for both ETTH and PM should be diagnosed as having ETTH. However, there is a large group of clinicians who disagree and feel that the diagnosis should be based on the worst headache, namely migraine.

The diagnosis of "probable" should trigger the clinician to consider the possibility that the patient has secondary headache, rather than a primary headache disorder, because the patient does not meet all of the IHS criteria for a given primary headache. Think again about whether the patient merits a workup before assuming it is primary and treating (Table 1.12).

The Spectrum of Migraine

There is evidence that patients with migraine have a spectrum of episodic headaches across time. That is, some of their attacks will meet criteria for ETTH, some for PM, and some for migraine.

There is also evidence that the lower-level headaches of *migraineurs* respond to migraine-specific medications, such as triptans. However, ETTH attacks in people who *never get* migrainous headaches, the so-called pure ETTH, do not respond to triptans any better than placebo. The lower-level headaches of *migraineurs* behave as lower-level migraines, and their TTH are, in essence, phenotypically tension-type, but genotypically, and clinically, migraines.

Migraineurs thus have a spectrum of attacks, with clinical variability, but all of their attacks are likely manifestations of their migrainous disorder. People with "pure" TTH have no migrainous attacks and rarely complain of headaches in the doctor's office. Although TTH is more common than migraine, it is seldom the reason an individual seeks medical care.

Table 1.13 Organization of ICHD-3 migraine aura

1. Typical aura (≥1 aura spreads gradually over ≥5 min and/or ≥2 symptoms occur in succession and/or each aura lasts 5–60 min and/or ≥1 aura is unilateral)
   (a) Visual
   (b) Sensory
   (c) Speech and/or language
2. Brainstem aura
   (a) Typical aura meeting above criteria *plus*
   (b) At least two of the following brainstem symptoms: dysarthria, vertigo, tinnitus, hyperacusis, diplopia, ataxia, decreased level of consciousness, with each brainstem aura symptom lasting 5–60 min
3. Hemiplegic aura
   (a) Typical aura meeting above criteria *plus*
   (b) Fully reversible motor weakness lasting <72 h
4. Retinal aura: Fully reversible confirmed monocular visual phenomena meeting other criteria for typical aura

Chronic Migraine and Chronic TTH

Episodic migraine and TTH can transform into daily or nearly daily headache, and diagnosis of these chronic disorders will be described in Chap. 4.

Migraine with Aura

The ICHD-3 changed the organization of aura to make the entire system more logical. The criteria begin with a definition of aura as "recurrent attacks, lasting minutes, of unilateral fully reversible visual, sensory, or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms." The auras are listed as visual, sensory, speech and/or language, motor, brainstem, or retinal. The first three types of aura are gathered under the term "typical," motor aura is "hemiplegic," basilar-type migraine is now "brainstem aura," and migrainous monocular visual change is "retinal" (Table 1.13).

Migraine with Typical Aura

Typical aura is defined as a reversible neurologic event, lasting from 5 to 60 min, in which headache occurs with the event or follows the aura within an hour. Aura only occurs in about 20% of migraineurs and often does not occur with each attack. In addition, the headache which accompanies aura does not always meet ICHD-3 criteria for migraine, and sometimes headache does not occur with aura at all.

Table 1.14 First pearl on precise diagnosis of aura

• Do not use the outdated, imprecise, and unacceptable terms "complicated migraine" or "complex migraine." They are not included in the ICHD-3

Table 1.15 Migraine with typical aura, ICHD-3 criteria

1. At least two attacks with the following
2. Aura must be a completely reversible visual, sensory and/or speech/language symptom or dysfunction with no motor symptoms such as weakness, no brainstem or posterior fossa symptoms and no retinal symptoms, suggested by retinal vasoconstriction
3. At least two of the following four
   (a) At least one aura symptom develops gradually over ≥5 min and/or different aura symptoms occur in succession over ≥5 min
   (b) Each aura symptom lasts 5–60 min. Duration can be 180 min with successive aura symptoms
   (c) ≥1 aura symptom is unilateral. Aphasia is considered unilateral, as language usually comes from a dominant hemisphere. The ICHD-3 states that dysarthria may or may not be considered unilateral
   (d) Headache occurs during the aura or follows the aura within 60 min
4. Not secondary

Table 1.16 Second pearl on precise diagnosis of aura

• If the visual aura is 5 -60 min long and not monocular, the diagnosis is typical aura without headache

The ICHD-3 criteria for typical aura are quite specific and include types of neurologic migrainous aura events that were previously called "complicated migraine" or "complex migraine" before the first ICHD. Neither term was included in the ICHD, and neither should be used in diagnosis of primary headaches (Table 1.14).

A diagnosis of typical aura requires at least two events as described in Table 1.15.

Typical aura can occur with migraine, with a headache that does not meet criteria for migraine, such as a TTH, and can occur with no headache following at all. The ICHD-3 now divides typical aura into two categories, that occurring with headache and that occurring without headache. Frequently, both aura without and with headache present over the course of a patient's lifetime. Rarely do patients have auras with every migraine headache.

Typical aura without headache has in the past been named "ocular migraine," "acephalic migraine," "migraine equivalents," and "late-life migraine accompaniments." These terms are no longer used. Late-life migraine accompaniments was a term coined by Dr. C. Miller Fisher for typical aura without headache, which commonly occurs in older patients (Table 1.16).

Typical aura symptoms can occur sequentially, with each one lasting up to an hour. This overall duration would have suggested a prolonged aura using old terminology, but now is simply described as "typical aura." There is no "prolonged aura" term in the ICHD-3. There is, however, a diagnosis of persistent aura without

Table 1.17 Persistent aura without infarction

1.	The present attack in a patient with migraine with aura is typical of previous aura except that one or more aura symptoms persist for ≥ 1 week
2.	Not secondary, no neuroimaging confirmation of stroke

infarction, defined as an aura lasting ≥ 1 week without neuroimaging confirmation of stroke in a person with a previous history of typical aura and with the persistent aura the same as the previous typical aura (see Table 1.17).

Migraine with Brainstem and Hemiplegic Auras

As noted above, the old terms "complicated migraine," "complex migraine," and "prolonged migraine" are no longer in use. Migraine with unusual aura is diagnosed either as typical aura with successive symptoms or as migraine with brainstem aura or hemiplegic migraine. The term migraine with brainstem aura replaced the terms basilar migraine and basilar-type migraine in ICHD-3.

Brainstem Aura

Note that typical aura and brainstem aura can never include weakness. Aura associated with weakness is either hemiplegic migraine or may not be migraine at all.

The diagnosis of migraine with brainstem aura requires the following: (1) the attacks must include a typical aura, (2) the attack also includes an aura with at least two brainstem or posterior fossa symptoms, (3) there can be no motor weakness, (4) the duration of each aura symptom is 5–60 min, and (5) headache occurrs during the aura or following the aura within an hour (see Tables 1.18 and 1.19).

Hemiplegic Migraine

Hemiplegic migraine diagnosis is contingent on the presence of at least two attacks with typical aura *and* motor weakness as well. The duration of the typical aura remains 5–60 min, while the motor weakness lasts < 72 h, without residual.

Many spells are erroneously attributed to migraine when they are unexplained and weakness is present. Remaining true to the ICHD-3 criteria, requiring typical aura with reversible weakness avoids that diagnostic error (see Tables 1.20 and 1.21).

Diagnosis of familial hemiplegic migraine (FHM) requires at least one first- or second-degree relative to be diagnosed with FHM, otherwise the diagnosis becomes sporadic hemiplegic migraine. Multiple genes have been cloned to explain the

Table 1.18 Migraine with brainstem aura, ICHD-3 criteria

1. At least two attacks with the following
2. A typical aura occurs with completely reversible visual, sensory and/or speech/language symptoms or dysfunction with no motor symptoms, such as weakness, and no retinal symptoms suggested by retinal vasoconstriction
3. The patient also has an aura consisting of at least two of the following brainstem or posterior fossa reversible symptoms, but *no motor weakness*
   (a) Dysarthria
   (b) Vertigo
   (c) Tinnitus
   (d) Hypacusis or phonophobia
   (e) Double vision
   (f) Ataxia
   (g) Decreased level of consciousness
4. Each aura symptom lasts ≥5 min up to 60 min, but they can be sequential
5. Headache begins during the aura or follows aura within 60 min
6. Not secondary

Table 1.19 Key pearls on diagnosing brainstem aura

- The patient must have a history of migraine with aura, meaning a history of at least two attacks including typical aura
- Each attack must include a typical aura
- The attack also includes an aura with at least two brainstem or posterior fossa symptoms
- There can be no motor weakness
- The duration of each aura symptom is 5–60 min
- Headache occurs during the aura or following the aura within an hour

Table 1.20 Hemiplegic migraine, ICHD-3 criteria

1. At least two attacks with the following
2. Typical aura and completely reversible motor weakness
3. At least 2/4 of the following
   (a) Duration of typical aura onset is at least 5 min, and the typical aura symptoms can occur sequentially
   (b) Duration of typical aura is 5–60 min although the typical auras can be additive when sequential, so up to 3 h. Motor weakness lasts less than < 72 h
   (c) At least one aura symptom is unilateral
   (d) Headache occurs during the aura or follows the aura within an hour
4. Not secondary

pathogenesis of FHM. The ICHD-3 lists three validated gene mutations triggering hemiplegic migraine, CACNA-1A (FHM-1), ATP1A (FHM-2), and SCN1A (FHM-3), and diagnosis of these causative mutations can be obtained by blood screening. The three gene mutations all result in excess glutamate in the synapse, increasing neuronal excitability postsynaptically, probably by activating the *N*-methyl-D-aspartate (NMDA)–glutamate receptor. There is also a classification in the IHCD-3 for FHM from other loci, anticipated for future discoveries.

Table 1.21 Key pearls on diagnosing hemiplegic migraine

- The patient must have a history of migraine with aura, meaning a history of at least two attacks including typical aura
- Each attack must include a typical aura lasting 5–60 min
- The attack also includes an aura with reversible motor weakness lasting 5 min to < 72 h
- Headache occurs during the aura or following the aura within an hour

Table 1.22 Retinal migraine, ICHD-3 criteria

1. At least two attacks with
2. Fully reversible monocular
   a. Positive visual changes (e.g., scintillations)
   b. Negative visual changes (e.g., blindness)
3. Confirmation by ≥ 1 of
   a. Clinical visual field exam
   b. Patent drawing of monocular visual change after careful instruction
4. ≥ 1 of
   a. Aura spreads gradually over ≥ 5 min
   b. Aura lasts 5–60 min
   c. Headache occurs with aura or within 60 min of its end
5. Not secondary, so other causes of transient monocular blindness have been eliminated

Retinal Migraine

Retinal migraine is defined by the ICHD-3 as "repeated attacks of monocular visual disturbance, including scintillations, scotomata, or blindness, associated with migraine headache." This could be due to neuronal activation in the retina or to vascular change, but is monocular. The criteria require *monocular* positive or negative visual changes otherwise meeting criteria for a migraine aura with clinical confirmation, listed in Table 1.22. For clinical pearls on diagnosing migraine with aura, see Table 1.23, and for a review of the clinical pearls on migraine with brainstem aura and hemiplegic migraine see Table 1.24.

Episodic Syndromes that may be Associated with Migraine (Previously Called Childhood Periodic Syndromes that are Commonly Precursors of Migraine)

These syndromes (recurrent gastrointestinal disturbance, cyclical vomiting syndrome, abdominal migraine, and benign paroxysmal vertigo) will be covered in Chap. 8.

Table 1.23 Clinical pearls on migraine with aura

- Typical aura can be visual, hemisensory, and/or an alteration of speech or language. Events lasting 5–60 min can follow each other successively, that is, up to 3 h, and still be diagnosed as typical aura
- Typical aura now requires at least one of

1. At least one aura symptom spreads gradually over at least 5 min, and/or at least two symptoms occur in succession
2. Duration of the aura is at least 5–60 min
3. At least one aura symptom is unilateral. Aphasia is considered unilateral
4. Headache occurs with the aura or within 1 h

- Aura can occur with migraine headache, with nonmigraine headache, and without headache. ICHD-3 divides typical aura into that with headache and that without
- Primary reversible migrainous aura should be diagnosed as typical, brainstem, hemiplegic, retinal, or persistent without infarction
- Help the patient distinguish between monocular symptoms (retinal aura) or homonomous symptoms (typical aura). Positive cortical phenomena, such as seeing zigzag fortification spectra seen in typical aura, take the symptom out of the eye to the visual cortex. Typical aura is far more common!

Table 1.24 Review of the clinical pearls on migraine with brainstem aura and hemiplegic migraine

- There is no ICHD-3 term of: complex migraine, complicated migraine, acephalic migraine, ocular migraine, or migraine equivalent. Avoid these terms as they are meaningless
- Brainstem aura is diagnosed by having at least two episodes with a *typical aura* and at least two symptoms of brainstem or posterior fossa dysfunction lasting 5–60 min, with headache or followed by headache within an hour
- There is never motor weakness in migraine with brainstem aura
- Neither migraine with brainstem aura nor hemiplegic migraine can be diagnosed without the typical aura component
- Hemiplegic migraine diagnosis requires at least two episodes of *typical aura* lasting 5–60 min, with motor weakness lasting < 72 h with headache or followed by headache within an hour
- Hemiplegic migraine cannot be diagnosed without the typical aura component
- *Note well*: Neither migraine with brainstem aura nor hemiplegic migraine can be diagnosed by ICHD-3 criteria without at least two identical attacks of a typical aura meeting ICHD-3 criteria, lasting 5–60 min, in addition to the target aura of brainstem symptoms of 5–60 min, or motor weakness lasting < 72 h, and a headache with the aura or within 60 min of the aura

Complications of Migraine

The ICHD-3 lists the following as complications of migraine: status migrainosus, persistent aura without infarction (already covered above), migrainous infarction, and migraine aura-triggered seizure.

Table 1.25 Status migrainosus, ICHD-3 criteria

1. The present attack in a patient with migraine without aura is typical of previous attacks except for its duration
2. Headache has both of the following features
(a) Unremitting for > 72 h
(b) Severe intensity
3. Not secondary

Table 1.26 Migrainous infarction, ICHD-3 criteria

1. The present attack in a patient with migraine with aura is *typical of previous attacks except that one or more aura symptoms persists for > 60 min*
2. Neuroimaging demonstrates ischemic infarction in the relevant area
3. Not secondary (no other stroke etiologies)

Status Migrainosus

This is simply a migraine that will not quit and commonly occurs in migraineurs at some point in their life. The ICHD-3 criteria are a migraine duration that exceeds 72 h, with pain or associated features that are "debilitating." Many menstrually related migraines (MRM) go longer than 3 days, so conventionally typical long menstrual migraines are excluded from this diagnosis (see Table 1.25).

Migrainous Infarction

Migraine and stroke, both being common, often occur together. Stroke risk is increased in migraine with aura patients, with the risk greatest in women under the age of 45. However, it is very rare that migraine actually appears to cause stroke, an event in which the stroke evolves out of the migraine.

This type of event, a true migrainous infarction, is so infrequent that the ICHD-3 criteria are very strict. The stroke must occur in a patient with previously established aura, and in the same distribution as the aura. Further, the ICHD-3 requires imaging confirmation of the stroke (see Table 1.26), although clinically objective evidence of the stroke, for example reflex asymmetry, a Babinski, or a pronator drift, should suffice.

Secondary causes for stroke occurring in a migraine need to be scrupulously eliminated. The rarity of true migrainous infarction cannot be overstressed (Table 1.27).

Migraine Aura-Triggered Seizure (Migralepsy)

As with migrainous infarction, migraine aura-triggered seizures can occur, but are very unusual. The seizure, when triggered by migraine with aura, must occur during

Table 1.27 Key pearls on migrainous infarction

- Migrainous infarction requires that the stroke occurs in the territory of *previously established aura in a patient with migraine with aura*
- In addition, by ICHD-3 criteria, the stroke must be imaged *in this previously established territory*
- In addition, secondary causes must be scrupulously ruled out
- So beware of this diagnosis: it is very, very rare!

Table 1.28 Migraine aura-triggered seizure (migralepsy), ICHD-3 criteria

1. Patient must have migraine with aura
2. The seizure occurs during or within 1 h after a migraine aura

Table 1.29 Menstrual Migraine, ICHD-3 beta appendix definitions

1. Both forms of menstrual migraine occur only as migraine without aura
2. *Pure menstrual migraine:* Attacks occur exclusively on days −2 to +3 of menstruation in at least 2/3 menstrual cycles *and at no other times of the cycle*
3. *Menstrually related migraine without aura:* Attacks occur on days −2 to +3 of menstruation in at least 2/3 menstrual cycles and *additionally at other times of the cycle*

the migraine or within an hour of the migrainous aura, and once again, secondary causes must be excluded (see Table 1.28).

A critical part of the diagnosis of migraine aura-triggered seizure is that by criteria, the seizure can only be triggered in a patient with migraine with aura, not in migraine without aura. This makes the diagnosis even more rare.

Menstrual Migraine

Menstrual migraine is only defined in the appendix of the ICHD-3 beta, but the current definitions are widely accepted and adopted. Menstrual migraine occurs in about 2/3 of women, and is just a migraine with a menstrual trigger. Hormonal issues and headache will be covered more extensively in Chap. 21.

There are two forms of menstrual migraine, but it is not clear that the differences are clinically meaningful. The usual form is referred to as menstrually-related migraine (MRM), in which attacks occur both during the menses and outside the menses. Pure menstrual migraine (PMM) attacks occur only during menses and not outside the menses. Both forms require that the menstrual attacks be migraine without aura.

For the purpose of diagnosis, the first day of flow is numbered +1, and to be a menstrual migraine the attack must begin between day −2 and +3. Menstrual migraines must occur in 2/3 of periods (see Table 1.29).

Clinically, menstrual migraines are often longer and more severe than non-menstrual migraines, so identifying them can help with planning a treatment regimen. There is a validated test for menstrual migraine, the Menstrual Migraine

Table 1.30 Menstrual Migraine Assessment Tool (MMAT), a quick screener for menstrual migraine. (Tepper et al. 2008)

Q1: Do you get headaches during your period?
Q2: Do your menstrual headaches get severe?
Q3: Do you get dislike of light during your menstrual headaches?
2/3 yes answers strongly suggest menstrual migraine

Assessment Test (MMAT), which consists of just three questions, presence or absence of attacks during menses, presence or absence of severe attacks, and presence or absence of photophobia. With 2/3 of these present, the sensitivity of MMAT for menstrual migraine is 0.94 and the specificity is 0.74, so it is well worth using in clinical practice for identifying menstrual migraine (see Table 1.30).

Conclusions on Diagnosis of Migraine and Tension-Type Headache

- A patient complaining of a stable pattern of at least 6 months of episodic, disabling headache likely has migraine
- The presence of nausea with long-established episodic headaches strongly suggests migraine
- Location does not determine diagnosis; migraine usually is accompanied by neck pain
- Triggers do not determine diagnosis; the most common trigger for migraine is stress
- TTH is featureless and without impact; patients almost never complain of tension-type headaches in the office
- Most patients with migraine have a spectrum of attacks, from attacks that appear like tension-type headaches to attacks that appear like PM (missing one migraine criterion) to ICHD-3 migraine, and all three levels of headache are likely forms of migraine responding to triptans
- Typical aura can be visual, sensory, or as an alteration of speech or language, or all three sequentially
- Both brainstem aura and hemiplegic aura attacks also require the presence of typical aura
- Migrainous infarction and migraine aura-triggered seizures are both rare and occur only in patients with established migraine with aura
- Menstrual migraine occurs from day −2 to +3, often with severe intensity and photophobia

Suggested Reading

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