

The Clinical Features of Migraine With and Without Aura

Updates on the clinical features of migraine, especially migraine with aura.

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Although we see migraineurs almost daily and over 50 percent of us are migraineurs,¹ a review of the clinical features may be of interest.

Migraine has a one-year period prevalence of 12 percent (17.1 percent in women and 5.6 percent in men) and² some 35 million people annually have migraine in the United States. The cumulative incidence of migraine by age 85 is 18.5 percent in males and 44.³ percent in females with onset before the age of 25 in 50 percent of cases, in 75 percent before the age of 35 years, and only two percent over the age of 50.³ The median age of onset is 25 years. About 8 percent of boys and 11 percent of girls have migraine.⁴ Chronic migraine, with attacks occurring on 15 or more days per month for at least three months, occurs in about 3.2 million people per year in the United States, 80 percent women.⁵

MIGRAINE FEATURES

Migraine pain is unilateral in 60 percent of cases and bilateral in 40 percent. About 15 percent of migraineurs report so-called “side-locked” headaches, with migraine always occurring on the same side. The pain is often more intense in the frontotemporal and ocular regions before spreading to the parietal and occipital areas. Any region of the head or face may be affected, including the parietal region, the upper or lower jaw or teeth, the malar eminence, and the upper anterior neck. Throbbing pain is present in 85 percent of episodes of migraine, although up to 50 percent of patients describe non-throbbing pain dur-

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ing some attacks. Up to 75 percent of migraineurs report, along with the head pain, unilateral or bilateral tightness, stiffness, or throbbing pain in the posterior neck. The neck pain can occur during the migraine prodrome, the attack itself, or the postdrome.⁶

Migraines last four to 72 hours if left untreated or if unsuccessfully treated although pediatric migraine may have a duration of two to 72 hours and is more often bilateral and frontotemporal. Migraine which persists for more than 72 hours, termed status migrainosus by Taverner in 1978,⁷ has an unknown prevalence. Without treatment, 80 percent of patients have moderate to severe pain and 20 percent have mild pain. The pain, which is usually increased by physical activity or movement, is associated with nausea in about 80 percent of episodes, vomiting in about 30 percent, photophobia in about 90 percent, and phonophobia in about 80 percent.⁸

When patients deny a history of light and noise sensitivity, the questions should be asked, “During a headache,

would you prefer to be in bright sunlight or in a dark room?" and "During a headache, would you prefer to be in a room with loud music or in a quiet room?" About 20 percent additional patients become aware of associated light and noise sensitivity by use of the follow-up questions.⁹

The number of attacks per month for migraineurs is about as follows: 25 percent, four or more severe attacks a month; 35 percent, one to four severe attacks per month; 38 percent, one or less severe attacks per month; and 37 percent, five or more headache days per month.¹ During migraine attacks in one study, most migraineurs reported severe impairment or the need for bed rest (53.7 percent) with only 7.2 percent who reported no attack-related impairment. Over a three-month period, 35.1 percent of the migraineurs had at least one day of activity restriction related to headache.

Migraine triggers are present in 76 percent of migraineurs with the following endorsed by migraineurs with triggers in one study: stress, 89 percent; female hormones, 65 percent; not eating, 57 percent; weather, 53 percent; physical exhaustion or traveling, 53 percent; sleep disturbance, 50 percent; perfume or odor, 44 percent; bright lights, 38 percent; neck pain, 38 percent; alcohol, 38 percent; smoke, 36 percent; sleeping late, 32 percent; heat, 30 percent, food, 27 percent; and exercise, 22 percent.¹⁰

Cranial autonomic symptoms are caused by parasympathetic activation of the sphenopalatine ganglion during an attack which innervates the tear ducts and sinuses. At least one symptom is present in 56 percent of migraineurs, usually bilateral and not present with each attack, most commonly forehead/facial sweating, conjunctival injection and/or lacrimation, and nasal congestion and/or rhinorrhea.¹¹ So migraineurs and some physicians misdiagnose the headaches as "sinus" because the pain is referred to the face or forehead which they think is "sinus," change of weather is a common trigger, and because of the presence of cranial autonomic symptoms which also seem like "sinus" symptoms. Migraines are confused with stress or tension type headaches because they commonly occur in the neck and are often triggered by psychological stress.

BENIGN EPISODIC MYDRIASIS

Benign episodic mydriasis is transient isolated mydriasis with normal vision and pupillary reactivity to light which may occasionally accompany migraine headaches (although cases have been described without accompanying headache), typically in young adults or children.¹² The duration of episodes is 15 minutes to 24 hours often associated with blurred vision. Episodes average two to three per month. Eye and motility abnormalities are absent. The dilated pupil is due either to parasympathetic insufficiency

TABLE 1. HEADACHE CLASSIFICATION COMMITTEE OF THE INTERNATIONAL HEADACHE, 3RD EDITION (BETA VERSION) CRITERIA FOR MIGRAINE WITH AURA

- At least two attacks of aura with migraine headache
- The migraine aura fulfills criteria for one of the subforms of aura with migraine headache.
- The symptoms are not attributed to another disorder

Subforms of aura — The IHS criteria recognize six subforms of aura with migraine headache:

- Typical aura with migraine headache
- Typical aura with non-migraine headache
- Typical aura without headache
- Familial hemiplegic migraine (FHM)
- Sporadic hemiplegic migraine
- Basilar-type migraine

Typical aura — A "typical" aura fulfills the following IHS criteria.

First, the aura has at least one of the following characteristics without motor weakness:

- Fully reversible visual symptoms including positive features (e.g., flickering lights, spots, or lines) and/or negative features (eg, loss of vision)
- Fully reversible sensory symptoms including positive features (e.g., pins and needles) and/or negative features (e.g., numbness)
- Fully reversible dysphasic speech disturbance

Second, the aura has at least two of the following characteristics:

- Homonymous visual symptoms and/or unilateral sensory symptoms
- At least one aura symptom develops gradually over ≥ 5 minutes and/or different aura symptoms occur in succession over ≥ 5 minutes
- Each symptom lasts ≥ 5 and ≤ 60 minutes.

of the iris sphincter or sympathetic hyperactivity of the iris dilator. Angle closure glaucoma should be excluded.

MIGRAINE WITH AURA

Epidemiology

In the United States, the one year period prevalence of migraine with aura is 5.3 percent in females (30.8 percent of female migraineurs) and 1.9 percent in males (32 percent of male migraineurs).⁸ Up to 81 percent of those with migraine with aura also have attacks of migraine without aura.¹³

The reported age of onset ranges from a mean of 11.9 years (range 4–17)¹⁴ to a mean of 21 years old (range 5–77).¹⁵ In one study, 54.9 percent of patients suffered from less than one attack per month and 9.7 percent from more than three attacks per month.¹⁶

In another study, the mean of migraine with aura episodes/year/patient was reported to be 29 (ranging from less than one to 156).¹⁷

Clinical features

Table 1 provides the ICHD, 3rd edition criteria for migraine with aura. In a study of 362 patients with migraine with aura with a mean age of 46 years (range 12–90), at least in some attacks, 99 percent of patients had a visual aura, 54 percent had a sensory aura, and 32 percent had an aphasic aura.¹⁸ Most patients had a combination of aura symptoms as follows: 28 percent had a visual and sensory aura; 25 percent had a visual, sensory, and an aphasic aura; 6 percent had a visual and aphasic aura; and 39 percent had a visual aura exclusively. When more than one aura symptom occurred, they occurred in succession in 96 percent and simultaneously in four percent of patients. Just over 90 percent had a gradual onset of visual aura symptoms.

The “classic” visual aura is the fortification (looks like a fortified town as viewed from above) spectra or teichopsia (“seeing fortifications”) which is a jagged figure with fortification lines arranged at right angles to one another beginning from a paracentral area which usually spreads outwards leaving visual loss behind. There are often scintillations which may be white, gray, or have colors similar to a kaleidoscope in a semicircle or C shape surrounding the scotoma or area of visual loss. Scintillating scotomas are typically in one hemifield with visual field defects beginning around fixation and spreading outward. The symptoms reported by our patients may be quite variable, however.

Approximately 50 percent of patients report that the visual auras begin in the periphery and 50 percent in or adjacent to the center of the visual field. In a series of

122 clinic patients,¹⁹ the laterality of the visual auras was reported as follows: always on the same side of vision (right or left), 22.1 percent; one sided, but not always on the same side, 23.8 percent; always on both sides of vision, 23.8 percent; and sometimes on one side, sometimes on both sides, 29.5 percent.¹⁸ The color of the visual auras

TABLE 2. HEADACHE CLASSIFICATION COMMITTEE OF THE INTERNATIONAL HEADACHE, 3RD EDITION (BETA VERSION) CRITERIA FOR MIGRAINE WITH BRAINSTEM AURA

- A. At least two attacks fulfilling criteria B-D
- B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor or retinal symptoms
- C. At least two of the following brainstem symptoms:
 1. dysarthria
 2. vertigo
 3. tinnitus
 4. hypacusis
 5. diplopia
 6. ataxia
 7. decreased level of consciousness
- D. At least two of the following four characteristics:
 1. at least one aura symptom spreads gradually over 5 minutes, and/or two or more symptoms occur in succession
 2. each individual aura symptom lasts 5-60 minutes²
 3. at least one aura symptom is unilateral³
 4. the aura is accompanied, or followed within 60 minutes, by headache
- E. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

Notes:

1. When motor symptoms are present, code as 1.2.3 Hemiplegic migraine.
2. When for example three symptoms occur during an aura, the acceptable maximal duration is 3x60 minutes.
3. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

were reported as follows: always black-and-white, 30.3 percent; always black-and-silver, 20.5 percent; always colorful, 18 percent; both black-and-white and colorful, 22.2 percent; and have no color, nine percent. The visual phenomena were described with the following characteristics and percentages of patients: blurred vision, 54.1 percent; small bright dots, 47.5 percent; zigzag lines, 41.8 percent; flashes of bright light, 38.5 percent; "blind spots," 33.6 percent; flickering light, 30.3 percent; "like looking through heat waves or water," 24.6 percent; blindness in half of a visual field, 23.8 percent; white spots, 22.1 percent; colored dots/spots of light, 19.7 percent; curved or circular lines, 18.9 percent; small black dots, 17.2 percent; bean-like forms, like a crescent or C-shaped, 16.4 percent; black spots, 14.8 percent; and tunnel vision, 9.8 percent. Less common and rare visual auras include corona phenomena, palinopsia,²⁰ metamorphopsia, macropsia, micropsia, telescopic vision, teleopsia, mosaic vision, and multiple images.

According to the ICHD, 3rd edition criteria, migraine aura is considered to be of typical duration when lasting between five and sixty minutes. The criteria label aura lasting longer than an hour and less than a week as probable migraine with aura. Visual aura has been reported as lasting more than one hour in 6-10 percent of patients.²¹ Other aura symptoms can also last more than one hour as

follows: somatosensory aura in 14-27 percent and aphasic aura in 17-60 percent of patients.

A sensory aura consists of numbness, tingling, or a pins-and-needles sensation. The aura, which is usually unilateral, commonly affects the hand and then the face, or it may affect either one alone.²² Paresthesia of one side of the tongue is typical. Less often, the leg and trunk may be involved. A true motor aura is rare, but sensory ataxia or a heavy feeling is often misinterpreted as weakness. Patients often report a speech disturbance when the spreading paresthesias reach the face or tongue. Slurred speech may be present. With involvement of the dominant hemisphere, paraphasic errors and other types of impaired language production and comprehension may occur. Rarely, other aura symptoms may be described, including *deja vu* and olfactory and gustatory hallucinations.

Migraine aura is considered by many to be a distinct phase of the migraine attack preceding the headache. However, in a prospective study of 861 attacks of migraine with aura from 201 patients, during the aura phase, 73 percent of attacks were associated with headaches with 54 percent of the headaches fulfilling migraine criteria during the first 15 minutes within the onset of aura.²³ Aura follows headache in about 3-8 percent of cases.¹⁷ The headache may be contralateral to the side of the visual aura, ipsilateral in up to 62 percent of patients for some attacks,¹⁸ or bilateral.

TABLE 3. HEADACHE CLASSIFICATION COMMITTEE OF THE INTERNATIONAL HEADACHE, 3RD EDITION (BETA VERSION) CRITERIA FOR RETINAL MIGRAINE

- A. At least two attacks fulfilling criteria B and C
- B. Aura consisting of fully reversible monocular positive and/or negative visual phenomena (e.g. scintillations, scotomata or blindness) confirmed during an attack by either or both of the following:
 1. clinical visual field examination
 2. the patient's drawing (made after clear instruction) of a monocular field defect
- C. At least two of the following three characteristics
 1. the aura spreads gradually over 5 minutes
 2. aura symptoms last 5-60 minutes
 3. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and other causes of amaurosis fugax have been excluded.

Migraine aura as compared to cerebral ischemia and seizures

So unlike symptoms due to cerebral ischemia, migraine visual or sensory auras typically have a slow spreading quality where symptoms slowly spread across the visual field or body part followed by a gradual return to normal function in the areas first affected after 20 to 60 minutes.²⁴ Cerebral ischemic events typically have a sudden onset with an equal distribution in the relevant vascular territory although the affected area can expand step-wise if blood flow drops in additional vessels.²⁵ The progression in partial seizures is typically much more rapid. The return of function in areas first affected while symptoms are occurring in newly affected areas occurs in migraine aura but not in ischemia or seizures. As noted, when more than one aura type occurs, the auras almost always are reported as occurring one after the other, in contrast to cerebral ischemia where multiple neurological symptoms typically occur at the same time. Finally, migraine aura often begins with positive phenomena such as shimmering lights, zigzags in the vision, or tingling and then followed in minutes by negative symptoms such as scotoma, numbness, or loss of sensation which can also occur during seizures

but with a typically faster progression of symptoms. This progression from positive to negative symptoms is not typical of cerebral ischemia.

Late-life migraine accompaniments and migraine aura without headache

Fisher reported new onset “late life migrainous accompaniments” in 85 patients ages 40 to 73 years with episodes resembling transient ischemic attacks²⁶ similar to the 120 patients he had previously described.²⁷ Most had visual symptoms occurring alone (44/205) or combined with other aura symptoms. Headache was associated with the episodes in 40 percent of cases. In the Framingham study, visual migrainous symptoms were reported by 1.23 percent of subjects (1.33 percent of women and 1.08 percent of men) with onset after age 50 years in 77 percent with the following characteristics: stereotyped in 65 percent; never accompanied by headaches in 58 percent; the number of episodes ranging from 1 to 500 with 10 or more in 69 percent of subjects; and lasting 15-60 minutes in 50 percent.²⁸ In a study of 1000 patients presenting for a comprehensive eye examination in Alabama,²⁹ 6.5 percent reported visual symptoms consistent with migraine aura without headache, 8.6 percent females and 2.9 percent males with risk factors including female gender, a history of migraine headaches, and a history of childhood motion sickness.

A retrospective study of 100 aura patients³⁰ compared those with onset at ages of 45 years or older with those with onset before age 45 and found no differences in gender distribution, family or personal history of migraine without aura, type of aura symptoms or imaging findings. Aura symptoms were mostly visual. The aura duration was similar in both groups with duration in those with late onset aura as follows: <20 minutes, 47.8 percent; 20-60 minutes, 39.1 percent; >60 minutes, 13 percent. Headache were associated with auras less often in those with older onset. The patients with onset age 65 or older were similar to those with onset age 45 or older.

Alice in Wonderland Syndrome

Alice in Wonderland Syndrome, a term coined by Todd in 1955,³¹ is a rare migraine aura usually where patients experience distortion in body image characterized by enlargement, diminution, or distortion of part of or the whole body, which they know is not real. The syndrome can occur at any age but is more common in children. The cause may be migrainous ischemia of the nondominant posterior parietal lobule. Other causes include medications (topiramate,³² cough syrup with dihydrocodein phosphate and dl-methylephedrine hydrochloride),

Epstein-Barr virus and other infections,³³ depression, seizures and a right medial temporal lobe stroke.

Persistent visual aura

Rarely, migraineurs may have persistent visual aura with some 35 cases reported.^{34,35} This aura usually consists of simple, unformed hallucinations in the entire visual field of both eyes, including innumerable dots, television static, clouds, heat waves, flashing or flickering lights, lines of ants, a rainlike or snowlike pattern, squiggles, bubbles, and grainy vision. Occasionally, palinopsia (the persistence of visual images), micropsia, or formed hallucinations occur. ICHD, 3rd edition describes persistent aura without infarction as aura symptoms persisting for one week or more without evidence of infarction on neuroimaging.

Visual snow

In a just published article, Schankin and colleagues performed a study of “visual snow” (similar to the noise on an analog television set) or tiny dynamic flickering dots in the entire visual field in three groups: a chart review of 22 patients; an internet survey of self-reports with 235 respondents completing data sets; and a prospective semi-structured telephone interview of 78 people recruited from social media sites who had seen an ophthalmologist with normal fundoscopic exams and visual fields.³⁶ The prospective group of 78 people had the following: mean age of 30 years; a 1:1 female to male ratio; 59 percent with a history of migraine; 27 percent with a history of migraine with aura; 24 percent with a history of visual snow since childhood; 92 percent with a continuous history of visual snow from the beginning; 86 percent with palinopsia or after images; 81 percent with floaters; 79 percent with blue field entoptic phenomenon (“little cells that travel on a wiggly path”); 63 percent with spontaneous photopsia; 53 percent with swirls, clouds or waves with eyes closed; 74 percent with photophobia; and 68 percent with nyctalopia or impaired night vision.

In all groups, almost all of the people with visual snow had a variety of additional visual symptoms (palinopsia, enhanced entopic phenomena, photophobia, and impaired night vision). The authors suggest that visual snow is a unique visual disturbance clinically distinct from migraine aura without an effective treatment.

Migraine with brainstem aura

Table 2 provides the ICHD, 3rd edition, criteria for migraine with brainstem aura which was formerly termed basilar-type migraine which is now not used because involvement of the basilar artery is unlikely. Migraine with brainstem aura is a rare disorder that usually occurs from

ages 7-20 years and rarely presents in patients older than 50 years.³⁷ In one study, the following aura symptoms were reported: vertigo, 61 percent; dysarthria, 53 percent; tinnitus, 45 percent; diplopia, 45 percent; bilateral visual symptoms, 40 percent; bilateral paresthesias, 24 percent; decreased level of consciousness, 24 percent, and hypacusis, 21 percent. Visual symptoms, which usually take the form of blurred vision, shimmering colored lights accompanied by blank spots in the visual field, scintillating scotoma, and graying of vision, may start in one visual field and then spread to become bilateral. The median duration of aura was 60 minutes (range of 2 minutes to 72 hours) with two or more aura symptoms always occurring.

Retinal migraine

Retinal migraine is rare with a mean age at onset of 25 years presenting with fully reversible monocular positive and/or negative visual phenomena lasting less than one hour (Table 3 provides the criteria).³⁸ Typically, patients report flashing rays of light and zigzag lightning and less often, bright colored streaks, halos, or diagonal lines. Negative phenomena may be blurring, “gray-outs,” and “black-outs” causing partial or complete blindness. Elementary forms of scotoma are perceived as blank areas, black dots, or spots in the field of vision. Visual field defects can be altitudinal, quadrantic, central, or arcuate. The headache is usually ipsilateral to the visual loss. Almost 50 percent have a history of migraine with visual aura. Some patients who report monocular visual disturbance have hemianopsia which they are not aware of since they do not do a cover/uncover test. This is a diagnosis of exclusion of other causes of transient monocular blindness. Retinal migraine can lead to permanent monocular visual loss.

Migraine trait symptoms

In a study by mail of 219 migraineurs (32 percent with aura and 68 percent without) who had been seen in a tertiary headache clinic as compared to controls, the presence of both migraine and aura was associated with significantly higher frequencies of autokinesis, metamorphopsia, dyschromatopsia, cinematographic vision, illusionary visual spread, and synesthesia.³⁹ Double vision, inverted 2- and 3-dimensional vision, and altered perception of body weight and size were found more often in patients with migraine without aura than in those with aura. In contrast, aura was associated with the occurrence of visual splitting and corona phenomenon. The authors propose that the sensory and neuropsychological symptoms which occurred more often in migraineurs be termed “migraine trait symptoms” and suggest a prospective study to confirm these findings. ■

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