Migraine Variants

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- Migraine is a disorder affecting more than 13% of the general population
- 80% of migraineurs have migraine without aura
- 15–20% of cases have migraine with typical aura
- Isolated migraine aura without headache (acephalalgic migraine) may be encountered in 5% of patients

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Migraine variant (migraine equivalent)

exhibits itself in a form other than head pain

usually affect children and young adults

characterized by:

- Atypical sensory, motor, or visual aura
- Paroxysmal episodes of prolonged visual auras
- Confusion
- Dysarthria
- Focal neurologic deficits
- Gastrointestinal (GI) manifestations
- Other constitutional symptoms, with or without a headache

20% of cases may experience prodromal symptoms without subsequent headaches

- 1.2.1 Migraine with typical aura
  - 1.2.1.1 Typical aura with headache
  - 1.2.1.2 Typical aura without headache
- 1.2.2 Migraine with brainstem aura
- 1.2.3 Hemiplegic migraine
  - 1.2.3.1 Familial hemiplegic migraine (FHM)
    - 1.2.3.1.1 Familial hemiplegic migraine type 1 (FHM1)
    - 1.2.3.1.2 Familial hemiplegic migraine type 2 (FHM2)
    - 1.2.3.1.3 Familial hemiplegic migraine type 3 (FHM3)
    - 1.2.3.1.4 Familial hemiplegic migraine, other loci
  - 1.2.3.2 Sporadic hemiplegic migraine (SHM)
- 1.2.4 Retinal migraine

1.3 Chronic migraine

1.4 Complications of migraine

- 1.4.1 Status migrainosus
- 1.4.2 Persistent aura without infarction
- 1.4.3 Migrainous infarction
- 1.4.4 Migraine aura-triggered seizure

1.5 Probable migraine

- 1.5.1 Probable migraine without aura
- 1.5.2 Probable migraine with aura

1.6 Episodic syndromes that may be associated with migraine

- 1.6.1 Recurrent gastrointestinal disturbance
  - 1.6.1.1 Cyclical vomiting syndrome
  - 1.6.1.2 Abdominal migraine
- 1.6.2 Benign paroxysmal vertigo
- 1.6.3 Benign paroxysmal torticollis

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Migraine variants defined by Third Edition of the International Classification of Headache Disorders (ICHD-3):

- Hemiplegic migraine
- Migraine with brainstem aura
- Retinal migraine
- Episodic syndromes that may be associated with migraine
- Complications of migraine

Hemiplegic Migraine

- recurrent headaches associated with temporary unilateral hemiparesis or hemiplegia
- hemiparesis may resolve before the headache in minutes to hours or may persist for days to weeks(less than 72 hours)
- The prevalence is 0.03%
- Two forms are known
  - Familial hemiplegic migraine (FHM)
    - FHM type I
    - FHM type II
    - FHM type III
  - Sporadic hemiplegic migraine(SHM)

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Diagnostic criteria

- A. Attacks fulfilling criteria for *Migraine with aura* and criterion B below
- B. Aura consisting of both of the following:
- C. fully reversible motor weakness(Motor symptoms generally last less than 72 hours)
- D. fully reversible visual, sensory and/or speech/language symptoms.

Diagnostic criteria(migraine with aura)

- **A. At least two attacks fulfilling criteria B and C**
  B. One or more of the following fully reversible aura symptoms:
  - visual
  - sensory
  - speech and/or language
  - motor
  - brainstem
  - retinal
- **C. At least three of the following six characteristics:**
  - at least one aura symptom spreads gradually over ≥5 minutes
  - two or more aura symptoms occur in succession
  - each individual aura symptom lasts 5-60 minutes[1]
  - at least one aura symptom is unilateral[2]
  - at least one aura symptom is positive[3]
  - the aura is accompanied, or followed within 60 minutes, by headache
- **D. Not better accounted for by another ICHD-3 diagnosis.**

FHM

- autosomal dominant disorder and a channelopathy
- at least one first- or second-degree relative has migraine aura including motor weakness.
- in **FHM1** there are mutations in the **CACNA1A gene** (coding for a calcium channel) on chromosome 19
- in **FHM2** there are mutations in the **ATP1A2 gene** (coding for a K/Na-ATPase) on chromosome 1
- in **FHM3** there are mutations in the **SCN1A gene** (coding for a sodium channel) on chromosome 2

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TREATMENT

- beta blockers
- low-dose tricyclic antidepressants
- anticonvulsants
- and calcium channel blockers
- **Acetazolamide** but its benefit is questionable
- **Triptans and ergotamine preparations are contraindicated**

Basilar Migraine(Bickerstaff syndrome)

- most frequently in adolescent girls and young women
- symptoms of vertebrobasilar insufficiency

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Diagnostic criteria:

- **A. Attacks fulfilling criteria for Migraine with aura and criterion B below**
  B. Aura with both of the following:
    1. at least two of the following fully reversible brainstem symptoms:
       a) dysarthria[1]
       b) vertigo[2]
       c) tinnitus
       d) hypacusis[3]
       e) diplopia[4]
       f) ataxia not attributable to sensory deficit
       g) decreased level of consciousness (GCS ≤13)[5]
    2. no motor[6] or retinal symptoms.

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Retinal Migraine

- monocular visual disturbance, including scintillations, scotomata or blindness
- lasting from minutes to 1 hour
- associated with minimal or no headache
- Postural changes, exercise, and oral contraceptive agents may precipitate attacks
- optic pallor or narrowing of the retinal vessels can be seen
- Ruling out eye disease or vascular causes is important
- **Vasoconstrictive agents such as triptans and ergots should be avoided**

Diagnostic criteria:

- **A. Attacks fulfilling criteria for 1.2 Migraine with aura and criterion B below**
- **B. Aura characterized by both of the following:**
  - 1. fully reversible, monocular, positive and/or negative visual phenomena (eg, scintillations, scotomata or blindness) confirmed during an attack by either or both of the following:
    - – clinical visual field examination
    - – the patient's drawing of a monocular field defect (made after clear instruction)
  - 2. at least two of the following:
    - – spreading gradually over ≥5 minutes
    - – symptoms last 5-60 minutes
    - – accompanied, or followed within 60 minutes, by headache
- **C. Not better accounted for by another ICHD-3 diagnosis, and other causes of amaurosis fugax have been excluded.**

Episodic Syndromes That May Be Associated with Migraine

- Recurrent gastrointestinal disturbance
  - Cyclic vomiting syndrome
  - Abdominal migraine
- Benign paroxysmal vertigo
- Benign paroxysmal torticollis

- multiple cyclic attacks of pain or vomiting, with our without migraine headaches
- common in children and adolescents
- commonly precursors of migraine
- High-resolution MRI and (MRA) are indicated in suspicious cases in the **absence of a supportive family history**
- precipitated by infection, menstruation, or physical or emotional stress

Diagnostic criteria of Cyclical vomiting syndrome

- **A. At least five attacks of intense nausea and vomiting, fulfilling criteria B and C**
- **B. Stereotypical in the individual patient and recurring with predictable periodicity**
- **C. All of the following:**
  - nausea and vomiting occur at least four times per hour
  - attacks last for ≥1 hour, up to 10 days
  - attacks occur ≥1 week apart
- **D. Complete freedom from symptoms between attacks**
- **E. Not attributed to another disorder1.**

Diagnostic criteria of abdominal pain

- **A. At least five attacks of abdominal pain, fulfilling criteria B–D**
- **B. Pain has at least two of the following three characteristics:**
  - midline location, periumbilical or poorly localized
  - dull or "just sore" quality
  - moderate or severe intensity
- **C. At least two of the following four associated symptoms or signs:**
  - anorexia
  - nausea
  - vomiting
  - pallor
- **D. Attacks last 2-72 hours when untreated or unsuccessfully treated**
- **E. Complete freedom from symptoms between attacks**
- **F. Not attributed to another disorder[1].**

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***Diagnostic criteria of* Benign paroxysmal vertigo**

- **A. At least five attacks fulfilling criteria B and C**
- **B. Vertigo[1] occurring without warning, maximal at onset and resolving spontaneously after minutes to hours without loss of consciousness**
- **C. At least one of the following five associated symptoms or signs:**
  - nystagmus
  - ataxia
  - vomiting
  - pallor
  - fearfulness
- **D. Normal neurological examination and audiometric and vestibular functions between attacks**
- **E. Not attributed to another disorder**

Diagnostic criteria of Benign paroxysmal torticollis

- A. Recurrent attacks[1] in a young child, fulfilling criteria B and C
- B. Tilt of the head to either side, with or without slight rotation, remitting spontaneously after minutes to days
- C. At least one of the following five associated symptoms or signs:
  - pallor
  - irritability
  - malaise
  - vomiting
  - ataxia[2]
- D. Normal neurological examination between attacks
- E. Not attributed to another disorder

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Complicated Migraine

- Persistent aura without infarction
- Migrainous infarctions
- Migraine aura-triggered seizure(Migralepsy)
- Status migrainosus

Diagnostic criteria of Persistent aura

- A. Aura fulfilling criterion B
- B. Occurring in a patient with 1.2 *Migraine with aura* and typical of previous auras except that one or more aura symptoms persists for ≥1 week
- C. Neuroimaging shows no evidence of infarction
- D. Not better accounted for by another ICHD-3 diagnosis.

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Diagnostic criteria of Migrainous infarction

- A. A migraine attack fulfilling criteria B and C
- B. Occurring in a patient with 1.2 *Migraine with aura* and typical of previous attacks except that one or more aura symptoms persists for >60 minutes
- C. Neuroimaging demonstrates ischaemic infarction in a relevant area
- D. Not better accounted for by another ICHD-3 diagnosis.

- mostly occurs in the posterior circulation and in younger women
- Triptans, ergots, and dihydroergotamine are contraindicated
- Long-term antiplatelet therapy is indicated

Diagnostic criteria of Migralepsy

- A. A seizure fulfilling diagnostic criteria for one type of epileptic attack, and criterion B below
- B. Occurring in a patient with 1.2 *Migraine with aura*, and during or within 1 hour after an attack of migraine with aura
- C. Not better accounted for by another ICHD-3 diagnosis.

Less Common Migraine Variants

- **Acute confusional migraine**
  - almost exclusively seen in young children
  - manifested by episodes of confusion, disorientation, and vomiting, with or without headaches
  - Relieved by sleep
- **Vertiginous migraine**
- **Nocturnal migraine**
  - is not a true migraine variant
  - during the middle of the night or the early morning hours
  - related to circadian activation of certain neurotransmitters during sleep

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- migraine variants respond to typical migraine preventive medications
- **Treatment is divided into 3 components:**

  A. **Elimination of specific triggers**

  such triggers include monosodium glutamate (MSG), nitrates-containing processed meat, aged or smoked cheese, onions, pickled products, avocados, dairy products, nuts, chocolate, caffeine, and alcoholic beverages (in particular, red wine)

  B. **Acute management of attacks**

  C. **Long-term prevention**

- avoid the use of hormonal replacement therapy , smoking cessation

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