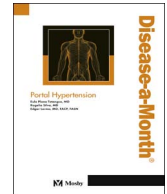


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## Primary Headache Disorders Part I- Migraine and the Trigeminal Autonomic Cephalalgias

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### A B S T R A C T

In Primary Headache Disorders, Part 1, we discuss three of the primary headache disorders using the headache definitions from ICHD-III (Beta): Migraine, with and without aura; its pathophysiology and treatment are discussed. We then discuss the Trigeminal Autonomic Cephalalgias (TACs), including Cluster Headache and Hemicrania Continua, two more primary headache disorders, as well as the other TAC Headaches. We discuss pathophysiology as well as diagnosis, treatment, and pharmacotherapeutic management of these headache diatheses.

### Introduction

Headache is a ubiquitous disorder, affecting almost half of the adult population globally who have experienced a headache at least a single time over the last year.

In adults aged 18–65 years of age, throughout the world, half to three quarters of adults in this group had a headache in the last year, with about 30% reporting migraine headache. Headache occurrence 15 days or more a month affect 1.7% to 4.0% of the world's population.<sup>1</sup>

In this two part article, we will detail the Primary Headache Disorders. Which ones? Migraine, Tension-type Headache and the Trigeminal Autonomic Cephalalgias (TACs) including Cluster Headache and Hemicrania Continua. We will also discuss the interaction between the Primary Headache Disorders and Medication Overuse Headache (MOH).

We will discuss the clinical description, the pathophysiology, and the pharmacotherapies and additional treatments of these headaches.

It is important to note that we will be discussing these headaches using the International Classification of Headache Disorders, 3<sup>rd</sup> Edition (ICHD III) Beta, published in Cephalalgia in 2013.<sup>2</sup> The final ICHD III should be approved in 2017 or 2018.

### Migraine Epidemiology

In the United States, it is estimated that more than 30 million people have one or more migraine headaches a year. Furthermore, migraine is encountered in 18% of females and 6% of males.<sup>3</sup>

Migraine accounts for 64% of severe headaches in women and 43% in males. About 75% of all people who experience migraine are women. One in six American women has migraine.

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The incidence of migraine with aura peaks in boys at about 5 years of age, and in girls at about 12–13 years of age. The incidence of migraine without aura peaks in boys at age 10–11, and in girls at age 14–17.<sup>4</sup> Prior to puberty, both prevalence and incidence of migraine are higher in boys than girls. After 12 years of age, the prevalence increases in both males and females, peaking at 30–40 years of age (YOA). The female to male ratio increases from 2.5:1 at puberty to 3.5:1 at 40 YOA.<sup>5</sup>

It should be noted that it was not so long ago that it was felt that there was no difference between migraine incidence and prevalence in young males and females prior to puberty.

The incidence of migraine does change, with decreased attack severity and frequency after menopause in about 2/3 of women. Most women with migraine have the attacks stop during pregnancy, as might be expected, as there are no significant peaks and troughs of estrogen. On the other hand, some women may manifest migraine at this time. The onset of migraine post 50 YOA is rare, but it can also begin after 60 YOA.

## Migraine Diagnoses- Nosology

Table 1 shows the Migraine Classification.<sup>6</sup> Note that “probably migraine without aura” 1.5 (or other types of migraine) is, by some headache specialists, questioned. “Migrainous disorder not fulfilling above criteria” also comes in for questioning, as if the situation doesn’t fulfill the noted criteria, how could it be a migraine? Indeed, if a patient with migraine didn’t have “C” in Table 2, how can one diagnose it as a migraine?

It is important to note here that in a Brainstem Aura, and Hemiplegic Migraine there must also be a “Typical Aura” that is visual, sensory or a speech problem.

Some things don’t change: a patient may have several aura’s which last between 5 min and 60 min and which may follow one after another, with, typically 1–3 occurring prior to the migrainous headache.

The new Appendix Criteria for Chronic Migraine by the IHS<sup>7</sup> is interesting in that it makes a diagnosis that the definition “changes”. This was, however, codified by the ICHD III Beta.

**Table 1**

The international classification of headache disorders 3rd Ed (Beta) (ICHD III)—outline<sup>6</sup>.

- 
1. Migraine
    - 1.1 Migraine without aura
    - 1.2 Migraine with aura
      - 1.2.1 Migraine with typical aura
      - 1.2.2 Migraine with brainstem aura
      - 1.2.3 Hemiplegic migraine
      - 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
    - 1.6 Episodic syndromes that may be associated with migraine
  2. Tension-type headache (TTH)
    - 2.1 Infrequent episodic tension-type headache
    - 2.2 Frequent episodic tension-type headache
    - 2.3 Chronic tension-type headache
  3. Trigeminal autonomic cephalalgias (TACs)
    - 3.1 Cluster headache
    - 3.2 Paroxysmal hemicrania
    - 3.3 Short-lasting unilateral neuralgiform headache attacks
      - 3.3.1 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
      - 3.3.2 Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)
    - 3.4 Hemicrania continua
    - 3.5 Probable trigeminal autonomic cephalalgia
  4. Other primary headache disorders
    - 4.1 Primary cough headache
    - 4.2 Primary exercise headache
    - 4.3 Primary headache associated with sexual activity
    - 4.4 Primary thunderclap headache
    - 4.5 Cold-stimulus headache
    - 4.6 External-pressure headache
    - 4.7 Primary stabbing headache
    - 4.8 Nummular headache
    - 4.9 Hypnic headache
    - 4.10 New daily persistent headache (NDPH)
- 

The International Headache Society (IHS) criteria for migraine without aura<sup>6</sup> notes: • Migraine without aura (about 75–80% of migraine) is formally diagnosed by the ICHD III (Beta)<sup>2</sup>

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