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# Mini Review

# **Understanding Headaches Classification**

Erin K. Purdy-Payne, MD1\*; Larry B. Mellick, MS, MD, FAAP, FACEP2

<sup>1</sup>Resident Physician, Department of Emergency Medicine, Medical College of Georgia, Augusta University, Augusta, Georgia, USA <sup>2</sup>Professor, Department of Emergency Medicine and Pediatrics, Medical College of Georgia, Augusta University, Augusta, Georgia, USA

# \*Corresponding author

Erin K. Purdy-Payne, MD

Resident Physician, Department of Emergency Medicine, Medical College of Georgia, Augusta University, Augusta, Georgia, USA; E-mail: EPAYNE@augusta.edu

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#### ABSTRACT |

In this mini-review, the authors introduce and discuss the 3<sup>rd</sup> edition of the International Classification of Headache Disorders (ICHD-3), an important tool in the diagnosis and management of headaches and other causes of orofacial pain, such as the cranial neuropathies.

#### Keywords

International Classification of Headache Disorders (ICHD); ICHD-3; Headaches; Headaches classification; Emergency department; Emergency medicine; Differential diagnosis of headache; Headache management; Primary headaches; Secondary headaches; Cranial neuropathies.

# INTRODUCTION |

This year saw the publication of the third edition of the International Classification of Headache Disorders (ICHD-3), a comprehensive diagnostic guide which categorizes and clinically defines all types of headaches as they are presently understood. This follows the publication of the beta edition in 2013, which was available online for the past several years both as a tool for diagnosis and classification, and to promote field testing and evidence- and clinical-based feedback before finalization of the new edition. <sup>1-2</sup> In either case, the ICHD-3 is a very helpful guide for any clinicians who manage patients presenting with headache. <sup>1-2</sup>

Despite its utility, and the availability of the beta version in a user-friendly online format since 2013, many clinicians are unaware of this helpful and important resource. Headache is a prevalent disorder worldwide across many settings, and a particularly common chief complaint in the emergency department.<sup>3-7</sup> Because of the extensive array of various headache diagnoses, which can have significant overlap in presentation and sometimes subtle points of diagnostic differentiation, the ICHD-3 is a resource meant to be easily accessible and frequently referred back to whenever it is needed to assist in diagnosing specific headache disorders.<sup>1-2</sup> While this would be less important if all headaches were best managed in a similar fashion, therapeutic management in fact

varies significantly depending on the specific type of headache. Therefore, accurate diagnosis is an important factor in therapeutic outcome, and such a resource can help clinicians effect a positive impact on the lives of untold numbers of patients.<sup>3</sup>

Detailed delineations and diagnostic descriptions of specific headache types are provided in the International Classification of Headache Disorders (ICHD-3), which, while the full document published in *Cephalalgia* is over 200 pages, is also presented in an easily-accessible, outline-based format online at www.ichd-3.org.<sup>2</sup> The goal of this mini-review, therefore, is not to restate these diagnostic criteria; but to increase awareness of this useful clinical resource, and present some of its higher yield information *via* simplified tables and an algorithm.

#### ICHD-3 CLASSIFICATIONS

The ICHD-3 classification reference has three main sections. Part one covers primary headaches, part two covers secondary headaches, and part three covers painful cranial neuropathies, as well as other facial pains and headaches. 1-2

The differentiation between primary and secondary headaches are relatively simple and easy to understand; a primary headache is due to the actual headache condition itself and cannot

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be attributed to another etiology, while a secondary headache is a symptom of another underlying condition. In cases where a headache otherwise meets diagnostic criteria for a primary headache type, such as migraine, but is due to an underlying cause, it only receives one diagnosis—that of the secondary headache disorder.<sup>1,2</sup>

The third section, cranial neuropathies and other facial pains, describes an array of disorders which are attributed to dysfunction or afferent activation of isolated craniofacial nerves, rather than true headaches. While the craniofacial pains are indistinct categories from the headaches, the clinical differentiation is not always so distinct—there is quite a bit of overlap in some of the presentations, and some headache types have even changed categories between the prior edition and the current one. For example, what is now known as recurrent painful ophthalmoplegic neuropathy was previously categorized an ophthalmoplegic migraine. 1.2

#### PRIMARY HEADACHES CLASSIFICATION

The first section of the ICHD-3, primary headaches, include four subtypes—migraine, tension-type headache, trigeminal autonomic cephalalgias, and other primary headache disorders. Each of these four headache classifications is further sub-classified as well.<sup>1,2</sup>

Migraine headaches with and without aura are subject to specific diagnostic criteria which specify not only what types of headaches can be diagnosed as migraines, but also when accompanying neurologic symptoms can be defined as an aura. In the simplest of terms, they are unilateral, pulsating, moderate-to-severe, and accompanied by either nausea and vomiting, or photophobia and phonophobia. In the emergency department setting, antidopaminergic medications such as have been shown to be the most efficacious treatment, and are often administered alongside other analgesics and adjunct treatments, sometimes including diphenhydramine to reduce the incidence of akathisia. 5

Tension-type headaches also have specific diagnostic criteria, including mild-to-moderate pressure-type pain in a bilateral distribution, and may occur with or without pericranial tenderness. Most guidelines recommend treating these headaches with overthe-counter analgesics, including combination formulations, some of which contain caffeine. That being said, while the differentiation between a migraine and tension-type headaches is sharply demarcated in the ICHD-3, many clinicians and researchers have pointed out the subtlety of patients' presentations in real-life practice, and that in many cases, the presentation may be more blurred or a combination of two or more headache types. In addition, tension-type headaches have shown favorable responses to treatments primarily used to treat migraines. This data has led some to believe that they may, in fact, lie on the same spectrum and should therefore be managed similarly.

The third section of the primary headaches classification covers the trigeminal autonomic cephalalgias, which include cluster headaches, paroxysmal hemicrania, short-lasting unilateral neuralgiform headaches, hemicrania continua, and probable trigeminal autonomic cephalalgia; each of which is characterized by sharp pain occurring in a unilateral distribution, associated with ipsilater-

al autonomic symptoms. Despite these similarities, their timing and other diagnostic criteria are quite distinct from each other, as are their management recommendations. The ICHD-3 should be always consulted for differentiation of these disorders, as their similar features yet important distinctions would be almost impossible to memorize other than by clinicians specialized in this particular area.

The fourth category of primary headaches is for other primary headache disorders, and includes a number of less common headache conditions, including a primary cough headache, primary exercise headache, primary headache associated with sexual activity, primary thunderclap headache, cold-stimulus headache, an external-pressure headache, primary stabbing headache, numular headache, hypnic headache and new daily persistent headache. While many of these headaches occur secondary to other stimuli, they are still classified as primary headaches because they do are not the result of any other disorder. A helpful diagnostic algorithm for primary headaches (Algorithm 1), and a table including the primary headache classifications, current management, prophylaxis recommendations, and mimics (Table 1), are provided in this mini-review.

#### SECONDARY HEADACHES CLASSIFICATION

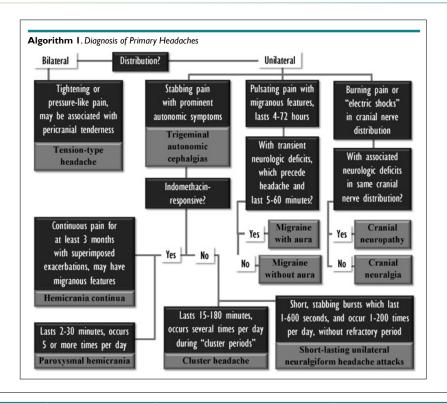
The classification of secondary headaches is much more extensive, due to the vast array of underlying conditions which can produce a headache. Subcategories include headaches attributed to trauma or injury to the head and/or neck, cranial or cervical vascular disorders, non-vascular intracranial disorder, a substance or its withdrawal, infection, a disorder of homeostasis, a disorder of various structures of the head and neck, or psychiatric disorder. These categories, too, are further divided into sub-categories, which are generally, the specific medical diagnoses which have produced a headache.<sup>1,2</sup>

A secondary headache is a headache which is present because of another medical condition, and generally, carry a higher-level of concern for clinicians because the underlying etiology is sometimes a life-threatening condition such as bacterial meningitis or intracranial hemorrhage. The biggest challenge associated with secondary headaches is in differentiating them from the primary headache etiologies, which they frequently mimic and which are almost universally less worrisome. Failure to recognize and act upon a secondary headache will frequently have serious consequences and complications, depending on the etiology. Less Table 2 for a selection of these secondary headaches which may mimic more common primary headaches, as well as their emergent management.

# PAINFUL CRANIAL NEUROPATHIES, OTHER FACIAL PAINS, AND OTHER HEADACHES

The thirdsection in the International Classification of Headache Disorders (ICHD-3) is dedicated to the cranial neuropathies, other facial pains, and other headaches. Many, but not all of these conditions, are characterized by neuropathic-type pain in a specific





	Chara	cteristics	Emergency Department Management	Prophylaxis	Mimics	
Migraine headache	Unilateral, pulsating, severe, 4-72 hours, migrainous features, with or without aura		Neuroleptics or other anti-dopaminergics, combined with an over-the-counter analgesic, and an antiemetic	Antiepileptics, botulinum toxin injections	Ischemic stroke or TIA intracranial hemorrhage giant cell arteritis, centr venous sinus thrombosi	
	Management ac	Management alternatives to consider: sub-dissociative ketamine infusions, triptans ment adjuncts to consider: diphenhydramine (for prevention or treatment of akathisia), corticosteroids (for prevention of recurrence)				
Tension-type headache	Bilateral, tightening or pressure-like, with or with- out pericranial tenderness		Over-the-counter analgesics, may consider formulation with caffeine	Antidepressants, physical therapy, biofeedback	Many types of seconda headaches	
Paroxysmal hemicrania	Severe, stabbing, with ipsilateral parasympathetic autonomic symptoms	Lasts 2-30 minutes, occurs several times per day	Indomethacin	Indomethacin	Cluster headaches	
Hemicrania continua		Continuous pain with waxing and waning fluctuations			Migraine	
Cluster headache		15-180 minutes, several per day during 'cluster periods'	100% oxygen, subcutaneous/nasal triptan	Suboccipital corticosteroid injections	Migraine, paraoxysma hemicrania	
Short-lasting unilateral neuralgiform headache attacks		Short stabbing bursts which last a few seconds and occur many times per day	IV lidocaine or corticosteroids	Anti-epileptics, gabapentin	Primary stabbing head aches, trigeminal neura	
Primary stabbing headaches	Short stabbing burts, no autonomic symptoms		Indomethacin, COX-2 inhibitors	Indomethacin, tricyclic anti-depressants, gabapentin	Short-lasting unilaters neuralgiform headache tacks, cranial neuralgia	
Primary thunderclap Headache	Severe, sudden onset, maximal at onset		Rule out emergent causes		Acute intracranial hem rhage or othervascula problems	
Nummular headache	Small, well-circumscribed area of mild pain		Over-the-counter analgesics	Only for severe or refractory cases	Underlying structura lesion	
Hypnic headache	Onsets exclusively during sleep (2-4 am), usually moderate bilateral pain		Analgesics formulation containing caffeine	Lithium	Variable, depends or features	
New daily persistent headaches	Occur daily for at least 3 months in patients with limited prior headache history, variable features		Similar to migraine headaches		Underlying structral les	



	May mimic	Differentiating features	Emergency Department Management	
Giant cell arteritis	Migraine, certain cranial neuropathies	Ipsilateral monocular vision changes (very rare with migraine); may also have scalp tenderness, jaw claudication, fevers, polymyalgia	High dose corticosteroids, arrange for temporal arter biopsy	
Central venous sinus thrombosis	Migraine, encephalopathy, intracranial hemorrhage, ischemic stroke, or other intracranial vascular problems	Highly variable, may have neurologic find- ings including seizures, vision changes, focal deficits, alterations of mental status or level of consciousness	Anticoagulation, antiepileptic prophylaxis, monitoring and management of intracranial pressure	
Medication overuse headache	Migraine or tension-type headaches	Worsening or new type of headache in the setting of chronic overuse of medication intended for acute headache management	Tapered or abrupt withdrawal of acute medications, optimize preventative management of chronic headaches	
Cervicogenic headache	Tension-type headaches, occipital neuralgia	Headache onsets in neck before spreading to head, may have bony tenderness and/or signs of cervical nerve root compression	Consider imaging of cervical spine to evaluate for underlying problem if acute	

cranial nerve distribution, including trigeminal neuralgia, occipital neuralgia, optic neuritis and other, lesser-known neuropathies. This section also includes a category dedicated to headaches without a clear-cut etiology or not fitting specific diagnostic criteria for any other headache diagnosis. Again, the most current diagnostic criteria for each of these conditions, as determined by the ICHD-3 a panel of experts, are provided for the clinician. The treatments for these conditions range from local anesthetics, to antiepileptics and tricyclic antidepressants, to surgical interventions, depending on the nerve involved and the underlying cause.

### CONCLUSION |-

In summary, the 3<sup>rd</sup> edition of the International Classification of Headache Disorders (ICHD-3) is an extremely valuable tool in the diagnosis and therapeutic management of the complex spectrum of head and face pain syndromes.<sup>1</sup> Because therapeutic success is most likely to occur when the type or cause of a headache or face pain is correctly identified, a precise diagnosis is critical.<sup>2</sup>

## CONFLICTS OF INTEREST |

The authors have no conflicts of interest to disclose.

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