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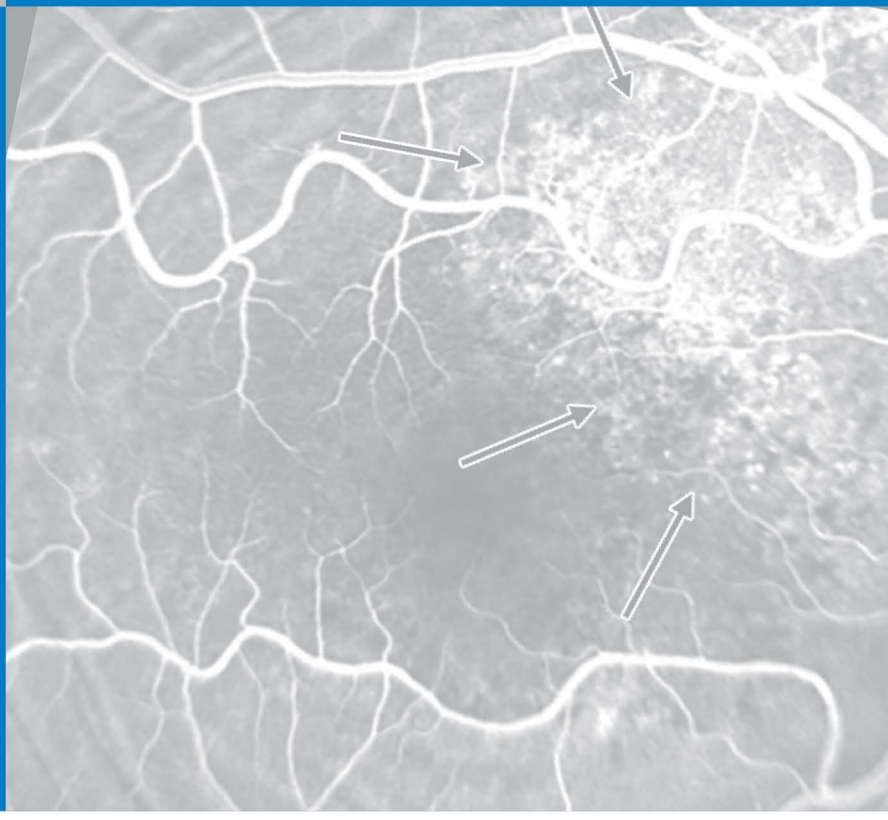
Modern Day Management of HEADACHE

Questions and Answers





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CHAPTER 2

The International Classification of Headache Disorders (ICHD-3 Beta Version)

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Q. What is the International Classification of Headache Disorders?

A. Disease definition is a fundamental prerequisite for professional communication both in clinical and research settings. Having structural criteria is especially mandatory for disorders, such as headache, where the diagnosis is often based on clinical symptoms. Researchers and clinicians as well as patients can use the same diagnostic system. The structural and operational diagnostic criteria of the International Classification of Headache Disorders (ICHD) were developed by the classification subcommittee of the International Headache Society (IHS). The first ICHD was published in 1988,[1] with a second edition in 2004.[2] The beta version of latest third edition ICHD-3 was published in 2013.[3] The chairman of these three editions is Professor Jes Olesen. Each edition represents a huge effort involving many experts worldwide and has taken at least 3 years to finish. Furthermore, some criteria of ICHD-3 are elaborated after comprehensive discussion with other related specialty societies or associations. The two cross-specialty criteria are vestibular migraine, according to a consensus with the Bárány Society;[4] and trigeminal neuralgia, a product by a joint committee of members of the IHS and members of the International Association for the Study of Pain (IASP).[5] For purposes of synchronizing ICHD-3 with the World Health Organization's next version of International Classification of Diseases (ICD-11), the ICHD-3 is a beta version now. The final version of ICHD-3 may be published in 2017 after field testing over 3-year period,[5] but is expected to be similar to the current beta version[3] that means clinicians and researchers should start using this classification now even though it is a beta version.

Q. How can one use the International Classification of Headache Disorders?

A. As suggested by Professor Jes Olesen, the classification is a reference that should be consulted over and over again rather than learnt by rote.[3] The ICHD adopted a hierarchical classification system. All headache disorders are classified into major groups and each group is then subdivided one, two, or three times into headache types, subtypes, and subforms. One must decide how detailed he or she wants to make the diagnosis, ranging from the first digit level to the fifth, e.g., migraine (code 1) and familial hemiplegic migraine type 1 (code 1.2.3.1.1). One gets a rough idea about which group the patient belongs to. Then one obtains information allowing a more detailed diagnosis.

The practicing family physician may only need to diagnose the headache disorder to the first or second levels. However, practicing neurologists or headache specialists would normally diagnose the precise subform to the third or more levels. In clinical practice, you do not need the classification system for obvious migraine or tension-type headache (TTH) patients, but it is useful for research purpose or when the diagnosis is uncertain.

Q. What is new for migraine in the International Classification of Headache Disorders-3 beta version?

A. All categories of migraine are rearranged more logically in the following order: migraine without and with aura, chronic migraine (CM), migraine complications, probable migraine, and other syndromes that maybe associated with migraine. Chronic migraine[6] in ICHD-3 beta is a distinct type of migraine with no ambiguity in the criteria rather than merely a complication of migraine. Unlike in ICHD-2, patients fulfilling criteria for both CM and for medication overuse headache (MOH) should be given both diagnoses according to the ICHD-3 beta. Episodic syndromes that maybe associated with migraine are a group of disorders occurring in patients who also have migraine or who have an increased likelihood to develop migraine. Although historically noted to occur in childhood, they may also occur in adults. Therefore, the word childhood is no longer used in ICHD-3 beta. Benign paroxysmal torticollis is now a formal diagnosis in the main body of the classification because a recent study has validated its relation to migraine.[7] Infantile colic[8] and vestibular migraine[4] are two new episodic migraine-associated syndromes included in the appendix of ICHD-3 beta.

Q. What is new for migraine aura in the International Classification of Headache Disorders-3 beta version?

A. The option "at least one aura symptom is unilateral" is added to aura criteria as one of four polythetic requirements in ICHD-3 beta. There are six aura categories that include visual, sensory, speech or language, brainstem, motor, and retinal. There are many patients with visual aura who occasionally have sensory or speech aura symptoms. Conversely, patients with sensory or speech aura always experience visual aura. In ICHD-3 beta, distinction between these three aura symptoms—visual, sensory, and speech—is not recognized. They are all included as typical aura. The term migraine with brainstem aura is now used to replace basilar-type migraine that was used in ICHD-2 because, in such patients, involvement of the basilar artery is not likely. Retinal aura was a new aura type which was included as a distinct type of migraine in ICHD-2. However, its existence has not been well-documented.[9] Thus, there are in total, four subtypes of migraine with aura: migraine with typical aura, migraine with brainstem aura, hemiplegic migraine, and retinal migraine (Table 1).

TABLE 1: Aura categories and subtypes of migraine with aura

Aura symptoms	Aura categories	Subtypes of migraine with aura
Visual	Typical aura	1.2.1 Migraine with typical aura
Sensory		
Speech/language		
Brainstem	Brainstem aura	1.2.2 Migraine with brainstem aura
Motor	Motor aura	1.2.3 Hemiplegic migraine
Retinal	Retinal aura	1.2.4 Retinal migraine

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Q. What is the difference between persistent aura without infarction and migraine aura status?

A. Persistent aura without infarction is the term used for patients with migraine with aura who have one or more aura symptoms similar to the previous aura but persisting for 1 week or more without evidence of cerebral infarction on neuroimaging. This is a rare condition, but has been well-documented. To make this diagnosis, underlying symptomatic conditions should be excluded, such as cerebral infarction. One study concluded that while belonging to the migraine spectrum, persistent visual aura may be considered a distinct disorder because magnetoencephalography data on the excitability changes in the visual cortex differentiates persistent visual aura from other migraine disorders.[10] On the other hand, migraine aura status, being in the appendix of the ICHD-3 beta, simply indicates migraine with frequent aura attacks that require at least two auras occurring per day for more than 3 days to meet the diagnostic criteria. Symptomatic conditions similar to persistent aura without infarction, such as reversible cerebral vasoconstriction syndrome, posterior reversible encephalopathy syndrome, or arterial dissection should be excluded before making the diagnosis.

Q. Patients with migraine, who frequently complain of vertigo or dizziness, may fit into one of the following three diagnoses: migraine with brainstem aura, benign paroxysmal vertigo, and vestibular migraine. How do you differentiate between these conditions?

A. Vertigo occurring in patients with migraine with brainstem aura is a type of migrainous aura, which means its duration should be within the time frame of 5–60 minutes as defined for an aura symptom. Its attack should have a temporal relationship to migrainous headache. Most vertigo episodes of vestibular migraine cannot meet the above criterion and therefore cannot be regarded as migraine aura.[11] Although vertigo is one of the most common symptoms of brainstem dysfunction reported by patients with migraine with brainstem aura,[12] at least two brainstem symptoms in addition to visual, sensory, or speech aura symptoms are necessary for this diagnosis. Most patients with vestibular migraine have only vertigo without other brainstem symptoms or typical aura and cannot fulfill these criteria. Therefore, migraine with brainstem aura and vestibular migraine are not synonymous. The diagnosis of benign paroxysmal vertigo requires at least five episodes of vertigo resolving spontaneously after minutes to hours. Being a precursor syndrome of migraine, a previous diagnosis of migraine is not necessary. On the contrary, the diagnosis of vestibular migraine requires an existing diagnosis of migraine and a limited time frame of 5 minutes to 72 hours. When the patients meet criteria of both disorders in separate attacks, both diagnoses can be given.

Q. What is new for tension-type headache in International Classification of Headache Disorders-3 beta version?

A. Diagnostic criteria for TTH have not changed much over three editions of the classification except that anorexia has been removed as a criterion from associated symptoms of TTH in the ICHD-3 beta. This headache is relatively featureless and has no biological marker to make the diagnosis. Current criteria are quite open that a headache without migraine characteristics and without any other known causes can be classified as TTH. Tension-type headache is divided into three types according to attack

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frequency: *infrequent episodic* (<1 day per month), *frequent episodic* (1–14 days per month), and *chronic* (≥15 days per month) TTH. All types of TTH are further divided into two subtypes: with and without pericranial muscle tenderness. Increased pericranial tenderness recorded by manual palpation is the most commonly mentioned finding in patients with TTH and it may add value and credibility to the explanations given to the patient. However, its validity and clinical significance are not known. Its link to the pathophysiology is not clear because, compared to migraine research, much less research has been conducted in TTH in recent years.

Q. What is new for trigeminal autonomic cephalalgias in International Classification of Headache Disorders-3 beta version?

A. Hemicrania continua (HC) was a diagnostic entity included under other primary headaches in ICHD-2 but is now included under trigeminal autonomic cephalalgias (TACs) as it has shared features of unilateral head pain and associated ipsilateral cranial autonomic symptoms. Furthermore, functional imaging also suggests that all syndromes in TACs including HC have activation in the region of posterior hypothalamic gray during the headache episode.[13] For consistency, new criteria unified all cranial autonomic symptoms in each type of TAC. There are in all seven autonomic symptoms. Two newly added symptoms are forehead and facial flushing, and sensation of fullness in the ear. Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) is considered a formal diagnostic entity. Whether short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is a subform of SUNA or not requires further study although a recent report has suggested that they are part of the same continuum.[14,15] Meanwhile, each is classified as a separate subtype.

Q. What is new for other primary headache disorders in International Classification of Headache Disorders-3 beta version?

A. This chapter includes primary headache disorders that are clinically heterogeneous and have poorly understood pathogenesis. They are classified into four categories, headaches associated with physical exertion; headaches attributed to direct physical stimuli; head pain over the scalp; and other miscellaneous primary headache disorders. Headaches with similar characteristics can also be symptomatic; therefore, this group demands careful evaluation by appropriate tests. Cold stimulus headache and external pressure headache are considered as primary headache in this chapter because they are induced by non damaging stimuli. Two subforms of headache associated with sexual activity, i.e., pre-orgasmic and orgasmic headache, were merged into one diagnostic code in the ICHD-3 beta because their clinical profiles are similar.[16] Nummular headache, an increasingly reported headache disorder,[17] has headache in a sharply contoured round or elliptical area of fixed size and shape, 1–6 cm in diameter in any part of scalp. It is often chronic and focal with mild-to-moderate pain intensity. New criteria for hypnic headache in the ICHD-3 beta has removed age limit[18] and requires attack frequency only more than 10 attacks per month rather than daily. Criteria for new daily persistent headache (NDPH), defined as TTH feature in the ICHD-2, has been simplified to a continuous headache that has a clearly remembered onset and persists for more than 3 months without any remission.[19] There is no restriction of headache features, i.e., either migraine-like or TTH-like features can both be diagnosed as NDPH.[20]

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Q. What is new for secondary headaches in International Classification of Headache Disorders-3 beta version?

A. A de novo headache occurring with another disorder recognized to be capable of causing it is diagnosed as secondary. Such patients shall be given only one headache diagnosis. When a patient has had a primary headache that worsens in close temporal relation to another disorder, the rules for attribution used in the ICHD-2 allowed one or two diagnoses in such circumstances, but relied on judgment. They have been modified in the ICHD-3 beta in order to be less open to interpretation. In the ICHD-3 beta, when a preexisting primary headache becomes chronic or worsens significantly in close temporal relation to a causative disorder, both the primary and the secondary headache diagnoses should be given, provided there is good evidence that the underlying disorder can cause headache. The word "chronic" was used in ICHD-2 to describe secondary headache persisting more than 3 months after the causative disease. It is replaced with the word "persistent" in ICHD-3 beta. In addition, the secondary headache group includes post-traumatic headache; homeostasis and cerebrospinal fluid (CSF) pressure related headaches; and headaches attributed to infection, substances, cranial vascular and nonvascular disorders, and psychiatric disorders.

Q. What is new for headache attributed to trauma or injury to the head and/or neck in International Classification of Headache Disorders-3 beta version?

A. There are no specific headache features to distinguish headaches in this chapter from other types.[21] Consequently, their diagnosis depends largely on the close temporal relation between the trauma and headache onset. Consistent with those of ICHD-2, the diagnostic criteria of ICHD-3 beta for all subtypes require that headache must be reported within 7 days after trauma or after regaining consciousness following injury to the head. Although this 7-day interval is somewhat arbitrary, there is not enough evidence to change this requirement. Those headaches that develop more than 7 days after trauma are named delayed onset acute or persistent headache attributed to traumatic injury to the head and are placed in the appendix of ICHD-3 beta waiting for further research to validate.[22] Post-traumatic amnesia lasting more than 48 hours that had been used to differentiate mild from moderate or severe head injury in the ICHD-2 is changed to more than 24 hours in the ICHD-3 beta.

Q. What is new for headache attributed to cranial or cervical vascular disorder in International Classification of Headache Disorders-3 beta version?

A. Reversible cerebral vasoconstriction syndrome (RCVS) is a new and important subtype of headache attributed to other acute intracranial arterial disorder. Reversible cerebral vasoconstriction syndrome is poorly understood, characterized clinically by severe diffuse, thunderclap-like headaches, mimicking aneurysmal subarachnoid hemorrhage, and accompanied by reversible multiple vasoconstrictions on imaging. Reversible cerebral vasoconstriction syndrome is the most frequent cause of thunderclap headache recurring over a few days or weeks.[23] If a headache attack presents with typical thunderclap features but with no vasoconstriction demonstrated on cerebral angiography, a diagnosis of probable RCVS could be given temporally. Three subtypes of genetic vasculopathy which were formerly listed in headache attributed to other intracranial vasculopathy are now placed as distinct types named headache attributed

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to genetic vasculopathy including cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy; mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS); and other genetic vasculopathy.

Q. What is new for headache attributed to nonvascular intracranial disorder in International Classification of Headache Disorders-3 beta version?

A. Both increased and decreased CSF pressure can lead to headache. For diagnosing headache attributed to idiopathic intracranial hypertension, ICHD-3 beta clearly defines CSF pressure more than 250 mmH_2O measured by lumbar puncture, epidural or intraventricular monitoring, with normal CSF chemistry and cellularity. A single measurement performed within minutes may not be as accurate as the average CSF pressure over 24 hours. Distinction of upper limit for CSF pressure between obese and nonobese has been mentioned in ICHD-2, but has been discarded in the ICHD-3 beta because body mass index is only weakly related to CSF pressure.[24] Headache attributed to low CSF pressure, i.e., intracranial hypotension, requires either CSF pressure less than 60 mmH_2O or evidence of CSF leakage on imaging as criteria to be fulfilled for the diagnosis. Description of headache characteristics and associated symptoms in criteria for CSF pressure related headaches are removed from ICHD-3 beta, as they cannot be relied on as a diagnostic criterion.[25] Headache attributed to colloid cyst of the third ventricle is a new subform under headache attributed to intracranial neoplasms and has headache characteristics different when compared to other intracranial neoplasm headaches. Most colloid cysts of the third ventricle are asymptomatic. But if their locations are immediately adjacent to the foramen of Monro, they can result in sudden obstructive hydrocephalus, causing headache with thunderclap onset and reduced level of consciousness. This highly characteristic presentation signals a life-threatening emergency.[26]

Q. What is new for headache attributed to a substance or its withdrawal in the International Classification of Headache Disorders-3 beta version?

A. Headache has long been thought to be an adverse effect of many drugs or some substances, maybe merely reflecting its high prevalence rather than causative relationship. Headache is regarded as a true side effect only when it occurs more often after an active drug than after placebo in double-blind controlled trials. Most headaches related to withdrawal of certain medications or substances are based on empirical experience. Of note, regular use or withdrawal of exogenous hormones can both be associated with an increase in frequency or new development of headache. In this chapter, the headache characteristics described as criteria for caffeine-, opioid-, and other substance-withdrawal headaches are removed as they are not specific. Caffeine-withdrawal headache describes headache developing within 24 hours after withdrawal of regular caffeine in excess of 200 mg/day for more than 2 weeks. Headache should resolve either within 1 hour after intake of caffeine 100 mg or within 7 days in the absence of further consumption.

Q. What is new for medication overuse headache in International Classification of Headache Disorders-3 beta version?

A. One of the essential criteria for MOH in ICHD-2 was that the headache resolved or reverted to its previous pattern within 2 months after discontinuation of the offended

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medication. This criterion is no longer present in the ICHD-3 beta because, in this context, MOH can only be diagnosed 2 months after discontinuation of medications. In addition, the criterion, headache has developed or markedly worsened during medication overuse in ICHD-2R,[27] is also removed from the ICHD-3 beta. The new criteria simply require that the patient has a primary headache and uses acute medications more frequently than allowed according to the ICHD-3 beta diagnostic criteria.[28] This makes the diagnosis of MOH more practical and may facilitate diagnosis of MOH. Being a secondary headache, MOH criteria also follows the general rule that patients with a preexisting primary headache who, in association with another causative disorder, e.g., MOH, develop a new type of headache or a marked worsening of their preexisting headache should be given diagnoses of both the causative disorder and the preexisting headache, i.e., both MOH and primary headache (such as chronic migraine) should be coded simultaneously.

Q. What is new for headache attributed to infection in International Classification of Headache Disorders-3 beta version?

A. In most cases of intracranial bacterial infection, it is difficult to distinguish involvement of pure meninges from pure encephalon. The distinction does not lead to different treatment and prognosis. Therefore, headache attributed to bacterial meningitis and encephalitis is included in the same subtype of headache attributed to bacterial meningitis or meningoencephalitis. However, viral meningitis does differ from encephalitis prognostically. For this reason, separate criteria are given for headache attributed to viral meningitis and headache attributed to viral encephalitis. Headache attributed to human immunodeficiency virus (HIV) infection is no longer a formal diagnosis in ICHD-3 beta because it is difficult to distinguish headache attributed purely to HIV infection from the primary-like headaches reported by most HIV patients.[29]

Q. What do acute, chronic, and persistent mean in headache attributed to infection?

A. Headache attributed to infection is usually the consequence of active infection. The headache would remit in parallel with resolution of the infection after treatment or immune activity. This headache is termed acute when the whole course is shorter than 3 months. In rare cases, the infection resolves but the headache does not remit; after 3 months, such headache is termed persistent (in accordance with other secondary headaches). In some other cases, the pathogen cannot be eradicated and the infection remains active. The headache in these cases may persist, as the cause remains present. It is referred to as chronic after 3 months.

Q. What is new for headache attributed to homeostatic disorder in International Classification of Headache Disorders-3 beta version?

A. Mild (140–159/90–99 mmHg) or moderate (160–179/100–109 mmHg) chronic arterial hypertension does not appear to cause headache. Ambulatory blood pressure monitoring in patients with mild and moderate hypertension has shown no convincing relationship between blood pressure fluctuations and headache.[30] Therefore, criteria of headache attributed to arterial hypertension defines blood pressure more than 180/120 mmHg as essential which was more than 160/100 mmHg

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in ICHD-2. Headache attributed to airplane travel is a new subtype of headache attributed to hypoxia and/or hypercarpnia to describe headache, often severe, usually unilateral and periocular and without autonomic symptoms, occurring during and caused by airplane travel.[31] It occurs during landing in more than 85% of patients and remits within 30 minutes after landing. Another new type is headache and/or neck pain attributed to orthostatic (postural) hypotension listed in the appendix of ICHD-3 beta. Here, the pain is mostly in the back of neck but can sometimes spread to the occipital region, accompanied by postural hypotension and the headache develops only in the upright posture.[32] The other interesting new subtype is headache attributed to travel in space, which is a nonspecific headache caused by travel in space. Out of the 17 astronauts, 12 reported at least one headache episode experienced while in space mostly without other symptoms of space motion sickness, whereas they had not suffered from headache when on earth.[33]

Q. What is new for headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cervical structure in International Classification of Headache Disorders-3 beta version?

A. Because of nociceptive field overlap and convergence in the nociceptive pathways of the head and neck, any eye, nose, or ear source of pain may lead to headache. However, most of the diagnostic criteria in this chapter lack scientific validations. Of note, if the causative disorder is unilateral, headache is likely to be localized ipsilateral to it. Headache attributed to disorder of the nose or paranasal sinuses is not "sinus headache"; the latter is a commonly-used but nonspecific diagnosis. Most sinus headaches in fact, are migraine or TTH but triggered by nasal disorder.[34] Headache attributed to temporomandibular disorder (TMD) is caused by a disorder involving the temporomandibular region. Diagnosis of TMD can be difficult. The use of diagnostic criteria evolved by the International Research Diagnostic Criteria or TMD Consortium Network and Orofacial Pain Special Interest Group is recommended.[35] There is some overlap between headache attributed to TMD and TTH. When the diagnosis of TMD is uncertain, the headache should be coded as TTH.

Q. What is new for cervicogenic headache in International Classification of Headache Disorders-3 beta version?

A. Cervicogenic headache[36] is often confused with TTH. Features that help to distinguish cervicogenic headache from TTH include side-locked pain, provocation of pain by digital pressure on neck muscles and by head movement, and posterior-to-anterior radiation of pain. However, they are not unique to it. "Headache is made significantly worse by provocative maneuvers" is added as one of four polythetic options in criterion C of cervicogenic headache which emphasizes maneuver provocation as a causative evidence in addition to temporal relation. Lesser degree of migrainous features such as nausea, vomiting, and photo or phonophobia may be a symptom of cervicogenic headache and may help to differentiate some cases. Tumors, fractures, infections, and rheumatoid arthritis of the upper cervical spine have not been validated formally as causes of headache, but maybe accepted depending on the individual case, just as with cervical spondylosis and osteochondritis.

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Q. Headache attributed to psychiatric disorders. Why are there only two disorders in International Classification of Headache Disorders-3 beta version?

A. Epidemiological data show that headache and psychiatric disorders occur together at frequencies higher than expected by chance.[37] True comorbidities are possible, indicating the likelihood of an underlying association. Possible casual associations include the headache causing the psychiatric disorder, the psychiatric condition causing the headache, bidirectional influence, and a common underlying factor leading to both headache and the psychiatric disorder. Only psychiatric disorders directly causing headache would be classified in this chapter. However, evidence supporting psychiatric causes of headache remains scarce. Thus, the diagnostic categories in chapter 12 are limited to those few cases in which a headache occurs in the context and as a direct consequence of a psychiatric condition known to be symptomatically manifested by headache. Because of uncertainties concerning the causal relationships and relative lack of evidence, criteria for headaches attributed to other psychiatric disorders have been included only in the appendix. Further study to clarify the mechanisms underlying these causal associations is necessary for study conclusions. When headache disorders occur coincidentally with other psychiatric disorders without any evidence of a causal relationship, both a primary headache diagnosis and a separate psychiatric diagnosis should be made.

Q. What is new for headache attributed to psychiatric disorder in International Classification of Headache Disorders-3 beta version?

A. Somatization disorder is characterized in the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) by a combination of multiple distressing symptoms and an excessive or maladaptive response to these symptoms.[38] Headache is one of the multiple symptoms. When there is evidence of causation such as headache worsens or remits in parallels with other somatic symptoms attributed to somatization disorders, diagnosis of headache attributed to somatization disorder could be given. Somatization disorder per se is not included in the DSM-5. It has been replaced by the category somatic symptom disorder which includes both individuals with headache who have disproportionate concerns about the headache as well as classic cases of somatization disorder with a lifelong pattern of multiple somatic symptoms including headache. Due to the enormous heterogeneity of this category, it would be possible to assert attribution only when headache was part of a larger pattern of multiple somatic complaints. Another psychiatric disorder attributing headache is delusional disorder, i.e. Headache attributed to psychotic disorder. Of note again, many psychiatric disorder related headaches, such as depression and anxiety are not formal diagnostic entities in the ICHD-3 and are put into the appendix because evidence is lacking for these headache entities.

Q. What is new for trigeminal neuralgia in International Classification of Headache Disorders-3 beta version?

A. Trigeminal neuralgia was classified into two subtypes, classical and symptomatic in ICHD-2. When a vascular loop was found to compress the nerve on imaging or at operation, the neuralgia should be regarded as secondary. However, it remains uncertain as to whether they are primary or secondary if no surgery or imaging was done. For this

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reason, the term classical rather than primary has been applied to those patients with a typical history even though a vascular compression may be discovered during its course. The term secondary can then be reserved for those patients with other clear etiologies. In ICHD-3 beta, trigeminal neuralgias of secondary causes are included in painful trigeminal neuropathy and are further classified according to their etiologies: attributed to acute herpes zoster, postherpetic, post-traumatic, attributed multiple sclerosis plaque, attributed to space-occupying lesion, and other disorders. Classical trigeminal neuralgia is also further classified into two subtypes depending on whether concomitant persistent facial pain is present or not. Classical trigeminal neuralgia with concomitant persistent facial pain has been called atypical trigeminal neuralgia. It responds poorly to treatment. Neurovascular compression is less likely to be demonstrated in such patients. It is thought to be a different subtype of trigeminal neuralgia.[39]

Q. What is new for painful cranial neuropathies and other facial pains in International Classification of Headache Disorders-3 beta version?

A. Tolosa-Hunt syndrome describes unilateral orbital pain associated with paresis of the third, fourth, and/or sixth cranial nerves. The new ICHD-3 beta diagnostic criteria not only specify locations of granulomatous lesions in the cavernous sinus, superior orbital fissure, or orbit but also discard the criterion of its response to corticosteroid. However, pain and paresis always resolve when treated adequately with corticosteroids. Syndrome of repeated attacks of paresis of one or more ocular cranial nerves with ipsilateral headache has been called ophthalmoplegic migraine in the ICHD-2. Recurrent painful ophthalmoplegic neuropathy is the term used in ICHD-3 beta because this syndrome is not considered a migraine subtype but rather a painful neuropathy because of the demonstration of cranial nerve inflammation by magnetic resonance imaging.[40] However, recent studies showed some patients might be migraine subtype because of the negative results of neuroimaging.[41] Burning mouth syndrome describes an intraoral burning sensation, recurring daily for more than 2 hours per day over more than 3 months, without causative lesions. Persistent idiopathic facial pain, that was earlier called atypical facial pain, requires recurrent poorly localized and dull, aching quality of facial pain for more than 2 hours per day for more than 3 months and where a dental cause has been excluded.

Q. What is the meaning of headache not elsewhere classified and headache unspecified?

A. Although probable subcategories have been applied to many headache disorders in order to include conditions fulfilling all criteria except one for specific headache disorder. There are still some headaches that cannot fit into any set of headache disorder described in ICHD-3 beta. Headache not elsewhere classified includes new headache entities that have never been described in ICHD-3. They would become distinct headache disorders in the next version of ICHD if clinical evidence is sufficient. Headache unspecified reserves for those headaches without enough information to be classified. Usually patients can hardly describe their headache in detail in the first visit. Headache diary and repeated clinical visits may help solve this problem. When information collected enough to classify, another optimal diagnosis should be given. Obviously this code must not be used as an excuse for not taking headache history in detail. Physicians should gather as much information as possible

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to make a diagnosis. This code could be used only when communication is not possible or the patient is not available.

CONCLUSION

It is important for any field of medicine to have a universally accepted disease classification. This is particularly true for a developing field like headache. Physicians and researchers should use these criteria to diagnose headache disorders. Of note, after the introduction of ICHD, scientific and therapeutic progress as well as public awareness of headache has vastly increased. The development of ICHD also inspired some medical specialty so much to develop their own disease criteria, e.g., International Classification of Vestibular Disorders from the Bárány Society.[42] The diagnostic system for headache disorders has proven utility at the different levels of health care systems all over the world. The IHS hopes to promote consistency in the way we classify, diagnose and treat headache patients throughout the world.

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