





TWO HUNDRED FACES OF HEADACHES

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Neurology Clinic Clinical Center of Serbia

Pain - Terrifying lord of life, October 22nd 2018

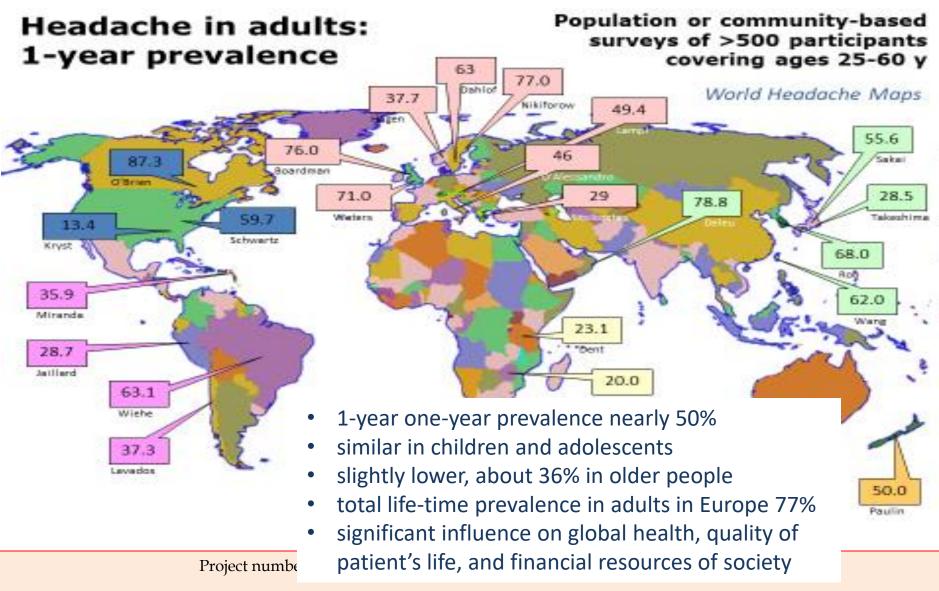
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There are more than 200 different headache types.

Classification

- helps to evaluate and treat the disorder
- provide higher precision to epidemiological studies and clinical research

Implementation of new scientific results in Classification established headaches from being one of the worst-classified neurological diseases to being the best.



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History of headache classification

The first headache classification

Cephalea

chronic, frequent, severe and long-lasting headache

Cephalalgia less frequent and mild



Aretaeus from Cappadocia 81- 138 year AD.

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History of headache classification

Thomas Willis 1672. De Cephalalgia "... out or in skull, universal or partial; short-lasting, continual or intermittent; inconstant or undetermined; forward, backward or lateralized and occasional or permanent...".



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History of headache classification

National Institute of Health

1962. Ad Hoc Committee on Classification of Headache Arnold Friedman, Knox Finley, John Graham, Charles Kunkle, Adrian Ostfeld i Harold Wolff

- was used in North America and some parts of Europe, was never internationally accepted and was not implemented in clinical practice.
- Imprecise headache definitions enabled different individual interpretations.

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History of headache classification

1. Vascular headache

Classic migraine, common migraine, cluster, hemiplegic, ophtalmoplegic migraine, lower-half headache

- 2. Muscle contraction headache
- 3. Combined headache: vascular and muscle contraction
- 4. Headache of nasal vasomotor reaction
- 5. Headache of delusional, conversion or hypochondriacal states
- 6. Nonmigrenous vascular headache
- 7. Traction headache
- 8. Headache due to overt cranial inflammation
- 9-13. Headache due to diseases of ear, nose, sinus, teeth
- 14. Cranial neuritides
- 15. Cranial neuralgias

Ad Hoc Committee on Classification of Headache. JAMA 1962; 179:717-718.

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History of headache classification

Definition of migraine

"...recurrent attacks of headache with broad range of intensity, frequency and duration. Attacks are usually lateralized at onset, often associated with anorexia, and sometimes with nausea and vomiting, some of them are preceded or associated with sensitive, motor or mood disorders and usually is familiar..."



Ad Hoc Committee on Classification of Headache. JAMA 1962; 179:717-718.

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History of headache classification

- 1982 International Headache Society IHS
- 1985 Headache Classification Committee 12 members, Jes Olesen



- 1988 International Classification of Headache Disorders ICHD I, San Diego
- 96 pages, 165 headache diagnosis
- operational diagnostic criteria for all headaches
- accepted by all national headache societies, WHO, WFNS
- translated into 22 languages and very soon entered everyday clinical practice and research.

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Division of migraine on scientific ground

 Aura rCBF with Xenon¹⁵³ and 254 channels

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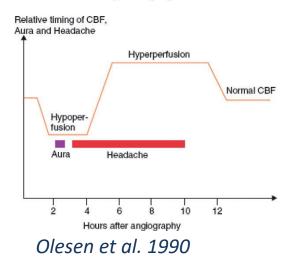
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Lauritzen and Olesen 1981-4



Before aura – focal hyperemia **Aura and the beginning of headache** – decreased flow (oligemia) spreading from occipital region forward during 15-45 min **Recovery of headache and flow**

Blood flow changes were not noted during migraine without aura.

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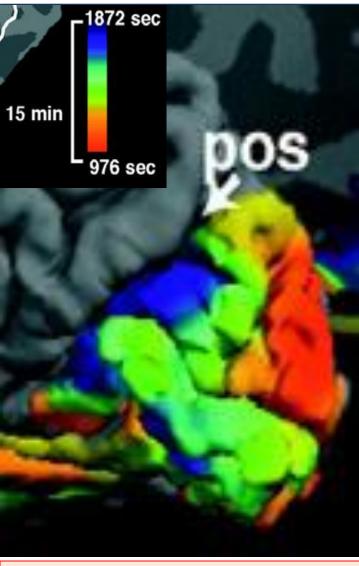
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CBF changes during migraine with aura









Aura – BOLD fMR

- aura provoked by exercises
- spreading from occipital region
- 3.5 ± 1.5 mm/min
- gradual onset and gradual disappearance
- prove that CSD generates aura in human cortex.

Hadjikhani N. et al. 2000

Bilateral occipital hypoperfusion during **migraine without aura** was demonstrated.

Denuelle et al. 2007

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History of headache classification

2003. The second revision of the Classification – ICHD-II

15 working groups with 3-10 experts, Jes Olesen

- 160 pages, 194 headache diagnosis
- hierarchically organized on 3(4) levels
- codes of ICD-10 were added to headache codes.

Volume 24 Supplement 1 2004

<u>Cephalalgia</u>

An International Journal of Headache

The International Classification of Headache Disorders

2nd Edition

Headache Classification Subcommittee of the International Headache Society





Cephalalgia 2004; 24 (suppl. 4): 9-160.

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ICHD-II

1.2	G43.1	Migraine with aura	
1.2.1	G43.10	Typical aura with migraine	
		headache	
1.2.2	G43.10	Typical aura with non-migraine	
		headache	
1.2.3	G43.104	Typical aura without headache	

1.5	G43.3	Complications of migraine
1.5.1	G43.3	Chronic migraine
1.5.2	G43.2	Status migrainosus
1.5.3	G43.3	Persistent aura without infarction
1.5.4	G43.3	Migrainous infarction
1.5.5	G43.3	Migraine-triggered seizures

Diagnostic criteria:

- A. Headache (tension-type-like and/or migraine-like) on \geq 15 days per month for >3 months² and fulfilling criteria B and C
- B. Occurring in a patient who has had at least five attacks fulfilling criteria B-D for 1.1 *Migraine without aura* and/or criteria B and C for 1.2 *Migraine with aura*
- C. On ≥ 8 days per month for >3 months, fulfilling any of the following³:

1. criteria C and D for 1.1 Migraine without aura

- 2. criteria B and C for 1.2 Migraine with aura
- believed by the patient to be migraine at onset and relieved by a triptan or ergot derivative
- D. Not better accounted for by another ICHD-3 diagnosis.

Headache Classification Committee, 2006.

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Cephalalgia 2004; 24 (suppl. 4): 9-160.





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ICHD-II

Hypnic headache

Diagnostic criteria:

- A. Recurrent headache attacks fulfilling criteria B-E
- B. Developing only during sleep, and causing wakening
- C. Occurring on ≥ 10 days per month for >3 months
- D. Lasting ≥15 minutes and for up to 4 hours after waking
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-3 diagnosis.

Primary thunderclap headache

Diagnostic criteria:

- A. Severe head pain fulfilling criteria B and C
- B. Abrupt onset, reaching maximum intensity in <1 minute
- C. Lasting for ≥ 5 minutes
- D. Not better accounted for by another ICHD-3 diagnosis.

Hemicrania continua

Diagnostic criteria:

- A. Unilateral headache fulfilling criteria B-D
- B. Present for >3 months, with exacerbations of moderate or greater intensity
- C. Either or both of the following:
 - 1. at least one of the following symptoms or signs, ipsilateral to the headache:
 - a) conjunctival injection and/or lacrimation
 - b) nasal congestion and/or rhinorrhoea
 - c) eyelid oedema
 - d) forehead and facial sweating
 - e) forehead and facial flushing
 - f) sensation of fullness in the ear
 - g) miosis and/or ptosis
 - 2. a sense of restlessness or agitation, or aggravation of the pain by movement
- D. Responds absolutely to therapeutic doses of indomethacin¹
- E. Not better accounted for by another ICHD-3 diagnosis.

Cephalalgia 2004; 24 (suppl. 4): 9-160.

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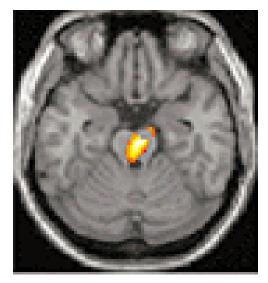
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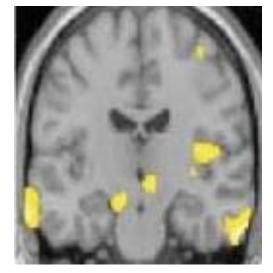




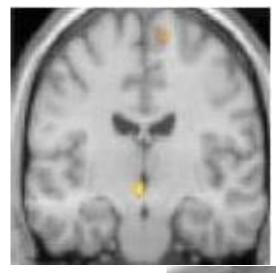
3.4 Hemicrania continua - fMRI



Weiller et al. 1995



Matharu et al. 2004



May et al. 1998







ICHD-II



Medication-overuse headache (MOH)

Diagnostic criteria:

- A. Headache occurring on ≥15 days per month in a patient with a pre-existing headache disorder
- B. Regular overuse for >3 months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache¹
- C. Not better accounted for by another ICHD-3 diagnosis.

- headache attributed to disorders of homeostasis
- headache attributed to psychiatric disorders.
- stronger connection between symptomatic headache and causative disorder was enabled by results of research.
 Cephalalgia 2004; 24 (suppl. 4): 9-160.

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History of headache classification

2013 The third revision – ICHD-III beta

18 working groups with 7-12 experts, Jes Olesen

- 180 pages, > 200 headache diagnosis
- test some entities in clinical practice
- harmonize with ICD-11



Cephalalgia 2013; 33: 629-808.

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History of headache classification

2014 Classification was translated into Serbian

National Headache Society of Serbia Nacionalno udruženje za glavobolje Srbije - **NUGS**





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ICHD-III beta

1.2 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
 - 2. sensory
 - 3. speech and/or language
 - 4. motor
 - 5. brainstem
 - 6. retinal
- C. At least two of the following four characteristics:
 - 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 minutes¹
 - 3. at least one aura symptom is unilateral²
 - the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

A. At least two attacks fulfilling criteria B and C

- B. One or more of the following fully reversible aura symptoms:
 - 1. visual
 - 2. sensory
 - speech and/or language
 - 4. motor
 - 5. brainstem
 - 6. retinal

С.

- At least three of the following six characteristics:
- at least one aura symptom spreads gradually over >5 minutes
- 2. two or more aura symptoms occur in succession
- 3. each individual aura symptom lasts 5-60
- minutes¹
- 4. at least one aura symptom is unilateral²
- 5. at least one aura symptom is positive³
- 6. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis.

3.1 Cluster headache

- C. Either or both of the following:
 - 1. at least one of the following symptoms or signs, ipsilateral to the headache:
 - a) conjunctival injection and/or lacrimation
 - b) nasal congestion and/or rhinorrhoea
 - c) eyelid oedema
 - d) forehead and facial sweating
 - e) forehead and facial flushing
 - f) sensation of fullness in the ear
 - g) miosis and/or ptosis
 - 2. a sense of restlessness or agitation

de Coo IF. Et al. 2016; 36:547-51.

Cephalalgia 2013; 33: 629-808.

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Li D, Christensen AF, Olesen J. 2015; 35: 748-756.







2018 Actual Classification – ICHD-3

- New diagnostic criteria for rare headache disorders such as primary cough, exercise and sexual headache, thunderclap headache, stabbing, nummular, hypnic and new daily persistent headache.
- New entity is introduced headache attributed to reversible cerebral vasoconstriction syndrome, aeroplane headache.
- New forms: persistent headaches attributed to arterial dissection or ischemic stroke.
- Revised criteria for headaches attributed to disturbances of cerebrospinal fluid.



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Principles of ICHD

- Diagnosis are hierarchically organized
- Level of classification according to diagnostic purpose

Code	Headache	Level of classification
1	Migraine	first
1.1 1.2	Migraine without aura Migraine with aura	second
1.2.1 1.2.2 1.2.3	Migraine with typical aura Migraine with brainstem aura Hemiplegic migraine	third
1.2.3.1 1.2.3.2	Familial hemiplegic migraine Sporadic hemiplegic migraine	fourth
1.2.3.1.1 1.2.3.1.2	Familial hemiplegic migraine type 1 Familial hemiplegic migraine type 2 	fifth







Principles of ICHD

The phenotype of headache is always classified

headache that is currently present, or that has been presented within the last year. For genetic studies or some other purposes all headache types that occurred during the lifetime of the patient could be diagnosed.

Each headache type is separately diagnosed and coded

The patient could receive several headache diagnosis at the same time, for example:

- 1.1 Migraine without aura
- 2.2 Frequent episodic tension-type headache and
- 8.2 Medication overuse headache.

Diagnosis are listed according to the importance for patient.

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Structure of ICHD

Part One: The Primary Headaches

- 1. Migraine
- 2. Tension-type headache
- 3. Trigeminal autonomic cephalalgias
- 4. Other primary headache disorders

Part Two: The Secondary Headaches

Introduction

- 5. Headache attributed to trauma or injury to the head and/or neck
- 6. Headache attributed to cranial and/or cervical vascular disorder
- 7. Headache attributed to non-vascular intracranial disorder
- 8. Headache attributed to a substance or its withdrawal
- 9. Headache attributed to infection
- 10. Headache attributed to disorder of homoeostasis
- 11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- 12. Headache attributed to psychiatric disorder

Part Three: Painful Cranial Neuropathies, Other Facial Pain and Other Headach

- 13. Painful lesions of the cranial nerves and other facial pain
- 14. Other headache disorders

Appendix

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Structure of ICHD

For every headache entity:

- Previously used terms
- Description of entity
- Diagnostic criteria (A-E) with notes
- Comments
- Bibliography

Due to precise inclusion and exclusion criteria, specificity > sensitivity. For particular diagnosis, the patient has to fulfill ALL criteria.

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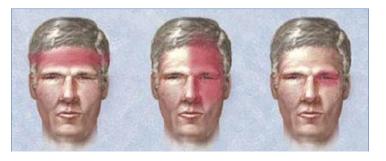




Structure of ICHD

Part One: The Primary Headaches

- 1. Migraine
- 2. Tension-type headache
- 3. Trigeminal autonomic cephalalgias
- 4. Other primary headache disorders



Diagnosis is based on symptoms – headache features

- monthly frequency
- intensity
- quality
- duration
- localization
- associated phenomena

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Structure of ICHD

Part Two: The Secondary Headaches

Introduction

- 5. Headache attributed to trauma or injury to the head and/or neck
- 6. Headache attributed to cranial and/or cervical vascular disorder
- 7. Headache attributed to non-vascular intracranial disorder
- 8. Headache attributed to a substance or its withdrawal
- 9. Headache attributed to infection
- 10. Headache attributed to disorder of homoeostasis
- Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- 12. Headache attributed to psychiatric disorder

Diagnosis of secondary headaches relay on causative disorder.

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Structure of ICHD

Part Three: Painful Cranial Neuropathies, Other Facial Pain and Other Headaches

13. Painful lesions of the cranial nerves and other facial pain 14. Other headache disorders

Consensus between the IHS and IASP - syndromes of painful lesions

- Neuralgia and neuropathy differences in clinical presentation and treatment approaches
- Central and peripheral neuropathic pain
- Presumed etiology: *classic*, idiopathic and secondary.

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Structure of ICHD

Appendix

- Headache entities with insufficient criteria to be extracted as separate and recognized entities.
- Alternative sets of diagnostic criteria for some entities with accepted criteria that are placed in the main body of the Classification.
- The first step to discard from the Classification those disorders that are traditionally considered as headache entities but without sufficient evidence until now.

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A10.8 Headache attributed to other disorder of homoeostasis

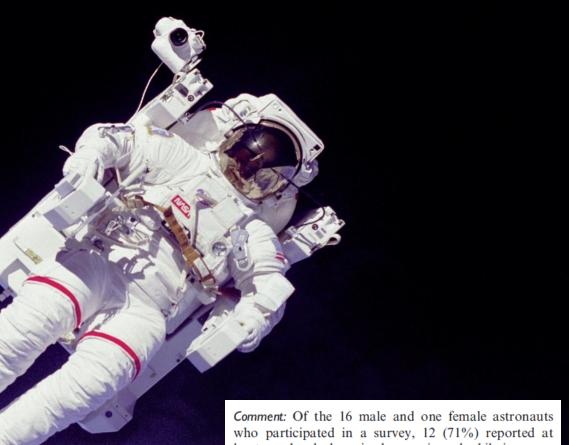
A10.8.1 Headache attributed to travel in space

Description: Non-specific headache caused by travel in space. The majority of headache episodes are not associated with symptoms of space motion sickness.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. The subject has travelled through space
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has occurred exclusively during space travel
 - 2. headache has spontaneously improved on return to earth
- D. Not better accounted for by another ICHD-3 diagnosis.

Vein et al. 2009.



Comment: Of the 16 male and one female astronauts who participated in a survey, 12 (71%) reported at least one headache episode experienced while in space, whereas they had not suffered from headache when on earth.

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