Part 2:
The secondary headaches

5. Headache attributed to head and/or neck trauma
6. Headache attributed to cranial 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 cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures
12. Headache attributed to psychiatric disorder

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Primary or secondary headache?

Primary:

- no other causative disorder

Secondary

(*ie*, caused by another disorder):

- new headache occurring in close temporal relation to another disorder that is a known cause of headache
- coded as *attributed to* that disorder

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Primary or secondary headache?

A *pre-existing* primary headache made worse in close temporal relation to another disorder:

- judgement required to code
  - *either* as the primary headache only
  - *or* as both the primary headache and a secondary headache (attributed to the other disorder)

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Primary or secondary headache?

Diagnosis:	**Primary headache only**	**Primary + secondary**
Temporal relation of other disorder to headache exacerbation	Loose	Close
Degree of exacerbation	Slight	Marked
Other evidence that other disorder can cause secondary headache	Weak	Strong
Other disorder eliminated	Headache unchanged	Headache returns to previous pattern

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Diagnostic criteria for secondary headaches

A. Headache with one (or more) of the following [listed] characteristics and fulfilling criteria C and D

B. Another disorder known to be able to cause headache has been demonstrated

C. Headache occurs in close temporal relation to the other disorder and/or there is other evidence of a causal relationship

D. Headache is greatly reduced or resolves within 3 mo (shorter for some disorders) after successful treatment or spontaneous remission of the causative disorder

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Important general rules

8. The last criterion for most secondary headaches

D. Headache is greatly reduced or resolves within [specified time] after successful treatment or spontaneous remission of the causative disorder

is part of the evidence of causation

Before treatment or spontaneous resolution, criterion D is not fulfilled; code as *Headache probably attributed to [the disorder]*

5. Headache attributed to head and/or neck trauma

5.1 Acute post-traumatic headache
5.2 Chronic post-traumatic headache
5.3 Acute headache attributed to whiplash injury
5.4 Chronic headache attributed to whiplash injury
5.5 Headache attributed to traumatic intracranial haematoma
5.6 Headache attributed to other head and/or neck trauma
5.7 Post-craniotomy headache

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5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury

A. Headache, no typical characteristics known, fulfilling criteria C and D
B. Head trauma with at least one of the following:
   1. loss of consciousness for >30 min
   2. Glasgow Coma Scale (GCS) <13
   3. post-traumatic amnesia for >48 h
   4. imaging demonstration of a traumatic brain lesion
C. Headache develops within 7 d after head trauma or after regaining consciousness following head trauma
D. One or other of the following:
   1. headache resolves within 3 mo after head trauma
   2. headache persists but 3 mo have not yet passed

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5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury

Notes

- Criterion D does not relate to evidence of causation
- Causation is established by onset in close temporal relation to trauma, whilst it is well recognised that headache after trauma often persists
- When this occurs, 5.2.1 *Chronic post-traumatic headache attributed to moderate or severe head injury* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist

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5.2.1 Chronic post-traumatic headache attributed to moderate or severe head injury

As 5.1.1 *except*:

D. Headache persists for >3 mo after head trauma

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5.3 Acute headache attributed to whiplash injury

A. Headache, no typical characteristics known, fulfilling criteria C and D
B. History of whiplash (sudden and significant acceleration/deceleration movement of the neck) associated at the time with neck pain
C. Headache develops within 7 d after whiplash injury
D. One or other of the following:
   1. headache resolves within 3 mo after whiplash injury
   2. headache persists but 3 mo have not yet passed since whiplash injury

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5.3 Acute headache attributed to whiplash injury

Notes

- Criterion D does not relate to evidence of causation
- Causation is established by onset in close temporal relation to whiplash, whilst it is well recognised that headache after whiplash injury may persist
- When this occurs, 5.4 *Chronic headache attributed to whiplash injury* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist

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5.4 Chronic headache attributed to whiplash injury

As 5.3 *except*:

D. Headache persists for >3 mo after whiplash injury

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6. Headache attributed to cranial or cervical vascular disorder

6.1 Headache attributed to ischaemic stroke or transient ischaemic attack
6.2 Headache attributed to non-traumatic intracranial haemorrhage
6.3 Headache attributed to unruptured vascular malformation
6.4 Headache attributed to arteritis
6.5 Carotid or vertebral artery pain
6.6 Headache attributed to cerebral venous thrombosis
6.7 Headache attributed to other intracranial vascular disorder

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6.2 Headache attributed to non-traumatic intracranial haemorrhage

6.2.1 Headache attributed to intracerebral haemorrhage

6.2.2 Headache attributed to subarachnoid haemorrhage (SAH)

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6.2.2 Headache attributed to subarachnoid haemorrhage

A. Severe headache of sudden onset fulfilling criteria C and D
B. Neuroimaging (CT or MRI T2 or flair) or CSF evidence of non-traumatic subarachnoid haemorrhage with or without other clinical signs
C. Headache develops simultaneously with haemorrhage
D. Headache resolves within 1 mo

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6.3 Headache attributed to unruptured vascular malformation

6.3.1 Headache attributed to saccular aneurysm

6.3.2 Headache attributed to arteriovenous malformation (AVM)

6.3.3 Headache attributed to dural arteriovenous fistula

6.3.4 Headache attributed to cavernous angioma

6.3.5 Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)

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6.4 Headache attributed to arteritis

6.4.1 Headache attributed to giant cell arteritis (GCA)

6.4.2 Headache attributed to primary central nervous system (CNS) angiitis

6.4.3 Headache attributed to secondary central nervous system (CNS) angiitis

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6.4.1 Headache attributed to giant cell arteritis

A. Any new persisting headache fulfilling criteria C and D

B. At least one of the following:
   1. swollen tender scalp artery with elevated erythrocyte sedimentation rate (ESR) and/or C reactive protein (CRP)
   2. temporal artery biopsy demonstrating giant cell arteritis

C. Headache develops in close temporal relation to other symptoms and signs of giant cell arteritis

D. Headache resolves or greatly improves within 3 d of high-dose steroid treatment

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6.7 Headache attributed to other intracranial vascular disorder

6.7.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)

6.7.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)

6.7.3 Headache attributed to benign angiopathy of the central nervous system

6.7.4 Headache attributed to pituitary apoplexy

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6.7.1 CADASIL

A. Attacks of migraine with aura, with or without other neurological signs
B. Typical white matter changes on MRI T2WI
C. Diagnostic confirmation from skin biopsy evidence or genetic testing (Notch 3 mutations)

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7. Headache attributed to non-vascular intracranial disorder

7.1 Headache attributed to high cerebrospinal fluid pressure
7.2 Headache attributed to low cerebrospinal fluid pressure
7.3 Headache attributed to non-infectious inflammatory disease
7.4 Headache attributed to intracranial neoplasm
7.5 Headache attributed to intrathecal injection
7.6 Headache attributed to epileptic seizure
7.7 Headache attributed to Chiari malformation type I
7.8 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)
7.9 Headache attributed to other non-vascular intracranial disorder

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7.1 Headache attributed to high cerebrospinal fluid pressure

7.1.1 Headache attributed to idiopathic intracranial hypertension (IIH)

7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes

7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus

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7.1.1 Headache attributed to IIH

A. Progressive headache with ≥1 of the following characteristics and fulfilling criteria C and D:
   1. daily occurrence
   2. diffuse and/or constant (non-pulsating) pain
   3. aggravated by coughing or straining

B. Intracranial hypertension (*criteria on next slide*)

C. Headache develops in close temporal relation to increased intracranial pressure

D. Headache improves after withdrawal of CSF to reduce pressure to 120-170 mm H_2O and resolves within 72 h of persistent normalisation of intracranial pressure

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7.1.1 Headache attributed to IIH

B. Intracranial hypertension fulfilling the following criteria:

1. alert patient with neurological examination that either is normal or demonstrates any of the following abnormalities:
   a) papilloedema
   b) enlarged blind spot
   c) visual field defect (progressive if untreated)
   d) sixth nerve palsy
2. increased CSF pressure (>200 mm H_2O [non-obese], >250 mm H_2O [obese]) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
3. normal CSF chemistry (low CSF protein acceptable) and cellularity
4. intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
5. no metabolic, toxic or hormonal cause of intracranial hypertension

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7.2 Headache attributed to low cerebrospinal fluid pressure

7.2.1 Post-dural puncture headache

7.2.2 CSF fistula headache

7.2.3 Headache attributed to spontaneous (or idiopathic) low CSF pressure

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7.2.1 Post-dural (post-lumbar) puncture headache

A. Headache that worsens within 15 min after sitting or standing and improves within 15 min after lying, with ≥1 of the following and fulfilling criteria C and D:
   1. neck stiffness; 2. tinnitus; 3. hypacusia;
   4. photophobia; 5. nausea
B. Dural puncture has been performed
C. Headache develops within 5 d after dural puncture
D. Headache resolves either:
   1. spontaneously within 1 wk
   2. within 48 h after effective treatment of the spinal fluid leak

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7.3 Headache attributed to non-infectious inflammatory disease

7.3.1 Headache attributed to neurosarcoidosis

7.3.2 Headache attributed to aseptic (non-infectious) meningitis

7.3.3 Headache attributed to other non-infectious inflammatory disease

7.3.4 Headache attributed to lymphocytic hypophysitis

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7.4 Headache attributed to intracranial neoplasm

7.4.1 Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm

7.4.2 Headache attributed directly to neoplasm

7.4.3 Headache attributed to carcinomatous meningitis

7.4.4 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion

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7.4.2 Headache attributed directly to neoplasm

A. Headache with ≥1 of the following characteristics and fulfilling criteria C and D:
   1. progressive
   2. localised
   3. worse in the morning
   4. aggravated by coughing or bending forward

B. Intracranial neoplasm shown by imaging

C. Headache develops in temporal (and usually spatial) relation to the neoplasm

D. Headache resolves within 7 d after surgical removal or volume-reduction of neoplasm or treatment with corticosteroids

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7.6 Headache attributed to epileptic seizure

7.6.1 Hemicrania epileptica

7.6.2 Post-seizure (post-ictal) headache

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7.6.2 Post-seizure (post-ictal) headache

A. Headache with features of tension-type headache or, in a patient with migraine, of migraine headache and fulfilling criteria C and D
B. The patient has had a partial or generalised epileptic seizure
C. Headache develops within 3 h following the seizure
D. Headache resolves within 72 h after the seizure

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8. Headache attributed to a substance or its withdrawal

This Chapter Is Currently Under Review

9. Headache attributed to infection

9.1 Headache attributed to intracranial infection

9.2 Headache attributed to systemic infection

9.3 Headache attributed to HIV/AIDS

9.4 Chronic post-infection headache

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9.1 Headache attributed to intracranial infection

9.1.1 Headache attributed to bacterial meningitis

9.1.2 Headache attributed to lymphocytic meningitis

9.1.3 Headache attributed to encephalitis

9.1.4 Headache attributed to brain abscess

9.1.5 Headache attributed to subdural empyema

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9.1.1 Headache attributed to bacterial meningitis

A. Headache with ≥1 of the following characteristics and fulfilling criteria C and D:
   1. diffuse pain
   2. intensity increasing to severe
   3. associated with nausea, photophobia and/or phonophobia

B. Evidence of bacterial meningitis from examination of CSF

C. Headache develops during the meningitis

D. One or other of the following:
   1. headache resolves within 3 mo after relief from meningitis
   2. headache persists but 3 mo have not yet passed since relief from meningitis

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9.1.1 Headache attributed to bacterial meningitis

Notes

- Criterion D does not relate to evidence of causation
- Causation is established by onset during diagnosed bacterial meningitis, whilst it is well recognised that this headache often persists
- When this occurs, 9.4.1 *Chronic post-bacterial meningitis headache* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist

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9.4.1 Chronic post-bacterial meningitis headache

A. Headache with ≥1 of the following characteristics and fulfilling criteria C and D:
   1. diffuse continuous pain
   2. associated with dizziness
   3. associated with difficulty in concentrating and/or loss of memory

B. Evidence of previous intracranial bacterial infection from CSF examination or neuroimaging

C. Headache is a direct continuation of 9.1.1 *Headache attributed to bacterial meningitis*

D. Headache persists for >3 mo after resolution of infection

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9.2 Headache attributed to systemic infection

A. Headache with ≥1 of the following characteristics and fulfilling criteria C and D:
   1. diffuse pain
   2. intensity increasing to moderate or severe
   3. associated with fever, general malaise or other symptoms of systemic infection

B. Evidence of systemic infection

C. Headache develops during the systemic infection

D. Headache resolves within 72 h after effective treatment of the infection

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9.2 Headache attributed to systemic infection

9.2.1 Headache attributed to systemic bacterial infection

9.2.2 Headache attributed to systemic viral infection

9.2.3 Headache attributed to other systemic infection

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9.3 Headache attributed to HIV/AIDS

A. Headache with variable mode of onset, site and intensity fulfilling criteria C and D
B. Confirmation of HIV infection and/or of the diagnosis of AIDS, and of the presence of HIV/AIDS-related pathophysiology likely to cause headache, by neuroimaging, CSF examination, EEG and/or laboratory investigations
C. Headache develops in close temporal relation to the HIV/AIDS-related pathophysiology
D. Headache resolves within 3 mo after the infection subsides

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10. Headache attributed to disorder of homoeostasis

10.1 Headache attributed to hypoxia and/or hypercapnia
10.2 Dialysis headache
10.3 Headache attributed to arterial hypertension
10.4 Headache attributed to hypothyroidism
10.5 Headache attributed to fasting
10.6 Cardiac cephalalgia
10.7 Headache attributed to other disorder of homoeostasis

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10. Headache attributed to disorder of homoeostasis

Terminology change 1988-2004

- This section was previously
  10. *Headache associated with metabolic disorder*
- The new term captures more accurately their true nature
- Headaches caused by significant disturbances in arterial pressure and by myocardial ischaemia are now included in this section

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10.1 Headache attributed to hypoxia and/or hypercapnia

10.1.1 High-altitude headache

10.1.2 Diving headache

10.1.3 Sleep apnoea headache

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10.3 Headache attributed to arterial hypertension

10.3.1 Headache attributed to phaeochromocytoma

10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy

10.3.3 Headache attributed to hypertensive encephalopathy

10.3.4 Headache attributed to pre-eclampsia

10.3.5 Headache attributed to eclampsia

10.3.6 Headache attributed to acute pressor response to an exogenous agent

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11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures

11.1 Headache attributed to disorder of cranial bone
11.2 Headache attributed to disorder of neck
11.3 Headache attributed to disorder of eyes
11.4 Headache attributed to disorder of ears
11.5 Headache attributed to rhinosinusitis
11.6 Headache attributed to disorder of teeth, jaws or related structures
11.7 Headache or facial pain attributed to temporomandibular joint (TMJ) disorder
11.8 Headache attributed to other disorder of the above

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11.2.1 Cervicogenic headache

A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D

B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache

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11.2.1 Cervicogenic headache

C. Evidence that the pain can be attributed to the neck disorder or lesion based on ≥1 of the following:
   1. demonstration of clinical signs that implicate a source of pain in the neck
   2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls

D. Pain resolves within 3 mo after successful treatment of the causative disorder or lesion

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11.2.1 Cervicogenic headache

Notes

- Cervical spondylosis and osteochondritis are NOT accepted as valid causes fulfilling criterion B
- When myofascial tender spots are the cause, the headache should be coded under 2. *Tension-type headache* (subform *associated with pericranial tenderness*)

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11.3 Headache attributed to disorder of eyes

- 11.3.1 Headache attributed to acute glaucoma
- 11.3.2 Headache attributed to refractive errors
- 11.3.3 Headache attributed to heterophoria or heterotropia (latent or manifest squint)
- 11.3.4 Headache attributed to ocular inflammatory disorder

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11.3.1 Headache attributed to acute glaucoma

A. Pain in the eye and behind or above it, fulfilling criteria C and D
B. Raised intraocular pressure, with at least one of the following:
   1. conjunctival injection
   2. clouding of cornea
   3. visual disturbances
C. Pain develops simultaneously with glaucoma
D. Pain resolves within 72 h of effective treatment of glaucoma

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11.5 Headache attributed to rhinosinusitis

A. Frontal headache accompanied by pain in one or more regions of the face, ears or teeth and fulfilling criteria C and D
B. Clinical, nasal endoscopic, CT and/or MRI imaging and/or laboratory evidence of acute or acute-on-chronic rhinosinusitis
C. Headache and facial pain develop simultaneously with onset or acute exacerbation of rhinosinusitis
D. Headache and/or facial pain resolve within 7 d after remission or successful treatment of acute or acute-on-chronic rhinosinusitis

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11.5 Headache attributed to rhinosinusitis

Notes

- 11.5 *Headache attributed to rhinosinusitis* is differentiated from "sinus headaches", a commonly-made but non-specific diagnosis. Most such cases fulfil the criteria for 1.1 *Migraine without aura*, with headache either accompanied by prominent autonomic symptoms in the nose or triggered by nasal changes
- *Chronic sinusitis* is not a cause of headache or facial pain unless relapsing into an acute stage

11.7 Headache or facial pain attributed to temporomandibular joint disorder

A. Recurrent pain in ≥1 regions of the head and/or face fulfilling criteria C and D
B. X-ray, MRI and/or bone scintigraphy demonstrate TMJ disorder
C. Evidence that pain can be attributed to the TMJ disorder, based on ≥1 of the following:
   1. pain is precipitated by jaw movements and/or chewing of hard or tough food
   2. reduced range of or irregular jaw opening
   3. noise from one or both TMJs during jaw movements
   4. tenderness of the joint capsule(s) of one or both TMJs
D. Headache resolves within 3 mo, and does not recur, after successful treatment of the TMJ disorder

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12. Headache attributed to psychiatric disorder

New section in classification

12.1 Headache attributed to somatisation disorder

12.2 Headache attributed to psychotic disorder

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12. Headache attributed to psychiatric disorder

Notes

- There is very limited evidence supporting psychiatric causes of headache
- The only diagnoses included are those of headache attributed to psychiatric conditions known to be symptomatically manifested by headache
- Such cases are rare
- The vast majority of headaches occurring in association with psychiatric disorders are not causally related to them but instead represent comorbidity

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12.1 Headache attributed to somatisation disorder

A. Headache, no typical characteristics known, fulfilling criterion C
B. Presence of somatisation disorder fulfilling DSM-IV criteria
C. Headache is not attributed to another cause

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12.2 Headache attributed to psychotic disorder

A. Headache, no typical characteristics known, fulfilling criteria C-E
B. Delusional belief about the presence and/or aetiology of headache occurring in the context of delusional disorder, schizophrenia, major depressive episode with psychotic features, manic episode with psychotic features or other psychotic disorder fulfilling DSM-IV criteria
C. Headache occurs only when delusional
D. Headache resolves when delusions remit
E. Headache is not attributed to another cause

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