

7.1 Headache attributed to high cerebrospinal fluid pressure

Coded elsewhere:

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

7.1.1 Headache attributed to idiopathic intracranial hypertension (IIH)

Previously used terms:

Benign intracranial hypertension (BIH), pseudotumor cerebri, meningeal hydrops, serous meningitis

Diagnostic criteria:

- A. Progressive headache with at least one 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 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 mmH₂O in the non-obese, >250 mmH₂O in the obese) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
 3. normal CSF chemistry (low CSF protein is 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
- C. Headache develops in close temporal relation to increased intracranial pressure
- D. Headache improves after withdrawal of CSF to reduce pressure to 120-170 mmH₂O and resolves within 72 hours of persistent normalisation of intracranial pressure

Comments:

IIH most commonly occurs in young obese women.

Although the majority of patients with IIH have papilloedema, IIH without papilloedema is observed. Other symptoms or signs of IIH include intracranial noises, tinnitus, transient visual obscurations and diplopia.

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

Coded elsewhere:

Headache attributed to increased intracranial pressure due to head trauma, vascular disorder or intracranial infection is coded to whichever one of those disorders is present. Headache attributed to raised intracranial pressure occurring as a side-effect of medication is coded as 8.3 *Headache as an adverse event attributed to chronic medication.*

Diagnostic criteria:

- A. Headache with at least one 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 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 mmH₂O in the non-obese, >250 mmH₂O in the obese) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
 - 3. normal CSF chemistry (low CSF protein is acceptable) and cellularity
 - 4. intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
- C. Headache develops after weeks or months of endocrine disorder, hypervitaminosis A or intake of substances (other than medications) that can elevate CSF pressure
- D. Headache resolves within 3 months after removal of the cause

Comment:

Normal pressure hydrocephalus does not cause headache.

7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus

Diagnostic criteria:

- A. Headache with at least two of the following characteristics and fulfilling criteria C and D:
 - 1. diffuse pain
 - 2. worse in the morning
 - 3. worse with Valsalva-like manoeuvres
 - 4. accompanied by vomiting
 - 5. associated with papilloedema, sixth nerve palsy, altered level of consciousness, gait instability and/or increased head circumference (in children <5 years old)
- B. High-pressure hydrocephalus fulfilling the following criteria:
 - 1. ventricular enlargement on neuroimaging
 - 2. intracranial pressure >200 mmH₂O in the non-obese or >250 mmH₂O in the obese
 - 3. no other intracranial disorder causing increased CSF pressure
- C. Headache develops in close temporal relation to increased CSF pressure
- D. Headache resolves within 72 hours of normalisation of CSF pressure