Idiopathic intracranial hypertension diagnosis

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The syndrome of increased intracranial pressure without hydrocephalus or mass lesion and with normal CSF composition is a diagnosis of exclusion now termed idiopathic intracranial hypertension (IIH). Diagnostic criteria of this disorder have not been updated since the Modified Dandy Criteria were articulated in 1985. Since then, new developments, including advances in neuroimaging technology and recognition of additional secondary causes of intracranial hypertension, have further enhanced the ability to diagnose conditions that may mimic IIH. These factors are not addressed in the Modified Dandy Criteria [1)].

Horev et al. retrospectively evaluated cerebral venous sinus stenosis using the conduit Farb score (CFS) and other radiological findings suggestive of IIH by computed tomography venography and magnetic resonance venography in females ≥ 18 years-old with chronic headaches, suspected IIH, and LPOP < 250 mm. Eighty-eight women (56 with LPOP < 200 mm H2O and 32 with LPOP ranging between 200 and 250mmH2O) were included. Among patients with LPOP 200-250mmH2O, 40% (12) exhibited three or more radiological findings supporting IIH, compared to 17% (8) in the LPOP < 200 mmH2O group ($p = 0.048$). Cerebral venous stenosis (CFS ≤ 5) was observed in 80% (24) of those with LPOP 200-250 mmH2O, contrasting with 40% (19) of those with LPOP < 200 mmH2O ($p < 0.001$). Cerebral venous stenosis was significantly more common in patients with LPOP 200-250 mmH2O than < 200 mmH2O, suggesting that they may benefit from IIH treatment [2)]

The idiopathic intracranial hypertension diagnosis criteria (IIH) were last revised and updated in 2013 by Friedman et al. [3] As stated by Friedman et al., one of the most important criteria is a lumbar puncture opening pressure (LPOP) > 250 mm H20. When the LPOP is less than 250 and the other criteria are met (evidence of papilledema, normal cerebrospinal fluid (CSF) profile, normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion, and no abnormal meningeal enhancement on MRI), a diagnosis of IIH is probable but not definite.

Modified Dandy Criteria

1. Signs and symptoms of intracranial hypertension.

2. Absence of localizing findings on neurologic examination

3. Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neurodiagnostic studies, except for evidence of increased cerebrospinal fluid pressure (greater than 200 mm water). * Abnormal neuroimaging except for empty sella, optic nerve sheath with filled-out CSF spaces, and smooth-walled non-flow-related venous sinus stenosis or collapse should lead to another diagnosis

4. Awake and alert

5. No other cause of increased intracranial pressure is present *For CSF opening pressure of 200 to 250 mm water required at least one of the following:

-Pulse synchronous tinnitus

-VI palsy

-Frisen Grade II papilledema see Papilledema and idiopathic intracranial hypertension

-Echography for drusen negative and no other disc anomalies mimicking disc edema present

-MRV (Magnetic Resonance Venography) with lateral sinus collapse/stenosis preferably using the ATECO technique

-Partially empty sella on coronal or sagittal views and optic nerve sheaths with filled-out CSF spaces next to the globe on T2 weighted axial scans.

Increased cerebrospinal fluid (CSF) around the bilateral optic nerve sheath complexes with flattening of the optic discs and partially empty sella can be associated with idiopathic intracranial hypertension

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It was found that obesity and papilledema were less common in Asian IIH patients when compared with Caucasian patients. Although patients with CSF pressures of 200-250 mm CSF had a less severe

phenotype, the risks of having headache or visual loss were comparable to those in the > 250 mm CSF group. It is possible that a diagnostic cutoff of > 200 mm CSF could be more suitable for Asians, although further studies are still needed [4)]

Idiopathic intracranial hypertension (IIH) is still a grey area in the knowledge of the aetiology, diagnosis and management of neurosurgical diseases.

The diagnosis requires a multidisciplinary approach.

The diagnosis may be suspected on the basis of the history and examination. To confirm the diagnosis, as well as excluding alternative causes, several investigations are required; more investigations may be performed if the history is not typical or the patient is more likely to have an alternative problem: children, men, the elderly, or women who are not overweight.

Idiopathic intracranial hypertension (IIH) is characterized by increased cerebrospinal fluid (CSF) pressure and normal or slit ventricles.

Physical examination

Physical examination of the nervous system is typically normal.

Ophthalmic examination

Idiopathic intracranial hypertension Ophthalmic examination.

Magnetic resonance imaging

Magnetic resonance imaging for Idiopathic intracranial hypertension diagnosis.

Optical coherence tomography

Optical coherence tomography may be used as a supplementary method to aid in the reliable detection of papilledema in evaluating a child for idiopathic intracranial hypertension [5)].

Lumbar puncture

Lumbar puncture for idiopathic intracranial hypertension diagnosis.

Transcranial color coded duplex sonography for idiopathic intracranial hypertension

Transcranial color coded duplex sonography for idiopathic intracranial hypertension.

Intracranial pressure monitoring for idiopathic intracranial hypertension

see Intracranial pressure monitoring for idiopathic intracranial hypertension.

1)
Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology. 2002 Nov 26;59(10):1492-5. Review. PubMed PMID: 12455560.
2)
Horev A, Eliav T, Sherer I, Biederko R, Ben-Arie G, Shelef I, Zlotnik Y, Regev T, Tsumi E, Honig A, Givaty G. Radiological signs supporting idiopathic intracranial hypertension in symptomatic patients with lumbar puncture opening pressure < 250 mm. Sci Rep. 2024 Aug 21;14(1):19450. doi: 10.1038/s41598-024-70588-z. PMID: 39169176; PMCID: PMC11339333.
3)
Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology. 2002 Nov 26;59(10):1492-5. doi: 10.1212/01.wnl.0000029570.69134.1b. PMID: 12455560.
4)
Hsu HT, Cheng HC, Hou TW, Tzeng YS, Fuh JL, Chen SP, Chen WT, Lee WJ, Pai YW, Lee YC, Lirng JF, Wang SJ, Wang YF. Idiopathic intracranial hypertension in Asians: a retrospective dual-center study. J Headache Pain. 2024 Sep 4;25(1):144. doi: 10.1186/s10194-024-01852-w. PMID: 39232671; PMCID: PMC11373263.
5)
Lee YA, Tomsak RL, Sadikovic Z, Bahl R, Sivaswamy L. Use of Ocular Coherence Tomography in Children With Idiopathic Intracranial Hypertension-A Single-Center Experience. Pediatr Neurol. 2015 Dec 10. pii: S0887-8994(15)30039-4. doi: 10.1016/j.pediatrneurol.2015.10.022. [Epub ahead of print] PubMed PMID: 26971313.