

Idiopathic intracranial hypertension

J.Sales-Llopis

Neurosurgery Department, *General University Hospital Alicante, Spain*

- Association of ganglion cell-inner plexiform layer thinning with visual function in pediatric papilledema
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Definition

Idiopathic [intracranial hypertension](#) without an identifiable cause. Absence of vascular or space-occupying lesions, and without enlargement of the cerebral ventricles, for which no identifiable causative factor can be determined. [Idiopathic intracranial hypertension](#) is the preferred term for this condition, replacing [pseudotumor cerebri](#), which often includes cerebral [venous sinus thrombosis](#) and other etiologies of [intracranial hypertension](#), and [benign intracranial hypertension](#), which does not take into account that some IIH patients do not have a “benign” course and go on to irreversibly [vision loss](#) ¹⁾.

The definition of [Idiopathic intracranial hypertension](#) (IIH) has been reviewed over time and many hypotheses have been expressed as the cause of the disease.

It is characterized by increased [cerebrospinal fluid pressure](#) and normal or [slit ventricles](#).

[Heinrich Quincke](#) in 1897 reported the first cases of IIH shortly after he introduced the [lumbar puncture](#) into medicine. It was named [pseudotumor cerebri](#) in 1904 but was not well delineated clinically until the 1940s when [cerebral angiography](#) was added to [pneumoencephalography](#) to identify cases of cerebral mass lesions. Foley coined the term [benign intracranial hypertension](#) in 1955 but reports from the 1980s demonstrated a high incidence of [visual loss](#) ^{2) 3)} and the term

“benign” is no longer appropriate.

Epidemiology

[Idiopathic intracranial hypertension epidemiology](#).

Etiology

see [Idiopathic intracranial hypertension etiology](#).

Clinical Features

[Idiopathic intracranial hypertension clinical features](#).

Diagnosis

see [Idiopathic intracranial hypertension diagnosis](#).

Differential diagnosis

[Idiopathic intracranial hypertension differential diagnosis](#).

Treatment

see [Idiopathic intracranial hypertension treatment](#).

Prognosis

[Idiopathic intracranial hypertension prognosis](#).

Systematic reviews

A systematic review of the surgical treatment of IIH was carried out. Cochrane Library, MEDLINE and EMBASE databases were systematically searched from 1985 to 2014 to identify all relevant manuscripts written in English. Additional studies were identified by searching the references of

retrieved papers and related narrative reviews.

Forty-one (41) studies were included (36 case series and 5 case reports), totalling 728 patients. Three hundred forty-one patients were treated with optic nerve sheath fenestration (ONSF), 128 patients with lumboperitoneal shunting (LPS), 72 patients with ventriculoperitoneal shunting (VPS), 155 patients with venous sinus stenting and 32 patients with bariatric surgery. ONSF showed considerable efficacy in vision improvement, while CSF shunting had a superior headache response. Venous sinus stenting demonstrated satisfactory results in both vision and headache improvement along with the best complication profile and low relapse rate, but longer follow-up periods are needed. The complication rate of bariatric surgery was high when compared to other interventions and visual outcomes have not been reported adequately. ONSF had the lowest cost.

No surgical modality proved to be clearly superior to any other in IIH management. However, in certain contexts, a given approach appears more justified. Therefore, a treatment algorithm has been formulated, based on the extracted evidence of this review. The traditional treatment paradigm may need to be re-examined with sinus stenting as a first-line treatment modality ⁴⁾.

Trials

The Idiopathic Intracranial Hypertension Treatment Trial represents the largest prospectively analyzed cohort of untreated patients with IIH. The data show that IIH is almost exclusively a disease of obese young women. Patients with IIH with mild visual loss have typical symptoms, may have mild acuity loss, and have visual field defects, with predominantly arcuate loss and enlarged blind spots that require formal perimetry for detection ⁵⁾.

Retrospective observational studies

Brain [Magnetic Resonance Venography](#) features were enumerated and assessed for correlation with [body mass index](#) (BMI) and [lumbar puncture opening pressure](#) (LPOP). Sensitivity, specificity, positive predictive value (PPV), and likelihood ratios (LRs) were calculated for each MRI sign. Significance was set at $P < 0.05$.

One hundred one patients diagnosed with IIH, and 119 control patients had complete files and were included. Patients with IIH were predominantly female (92.8% vs 59.7%; $P = <0.001$), younger (30.6 years vs 46.4 years; $P < 0.001$), and more obese (mean BMI = 35.2 vs 29.3; $P < 0.001$) than controls. Mean (SD) number of MRI findings was 2.21 (1.8) in IIH and 0.6 (1.2) in controls; ($P < 0.001$). Vertical nerve tortuosity (44.1%; $P < 0.001$), TVSS (37.8%; $P < 0.001$), sheath expansion (36.0%; $P < 0.001$), globe flattening (22.5%; $P < 0.001$), slit ventricles (18.9%; $P < 0.001$), optic disc protrusion (9.9%; $P = 0.007$), and complete empty sella (12.6%; $P < 0.042$) were observed more in patients with IIH than control patients. In the IIH group, mean (SD) LPOP was 33.6 (11.11) cmH₂O and weakly correlated with number of MRI findings ($\rho = 0.182$, $P = 0.057$). TVSS (sensitivity 37.84%; confidence interval [CI] 29.3%-47.2%, specificity 98.32%; CI 93.5%-99.6%) had the highest PPV (95.45%) and positive LR (22.51) for IIH diagnosis.

These results are consistent with IIH predominance in young, obese females. In patients with IIH, the number of MRI findings exceeded controls and positively correlated with LPOP. TVSS was most predictive of IIH ⁶⁾

Case series

[Idiopathic intracranial hypertension case series.](#)

Case reports

[Idiopathic intracranial hypertension case reports.](#)

References

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