

# Idiopathic Intracranial Hypertension

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**Abstract:** The syndrome of intracranial hypertension without structural brain or cerebrospinal fluid abnormalities and without identifiable cause, now most appropriately termed idiopathic intracranial hypertension, was described over a century ago. Although the pathogenesis of this condition remains unknown, diagnostic and therapeutic developments during the past two decades have substantially advanced patient management.

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## DEFINITION

The syndrome of increased intracranial pressure (ICP) without ventriculomegaly or mass lesion, and with normal cerebrospinal fluid (CSF) composition, was first described more than a century ago, yet we still know little about its pathogenesis (1). Often referred to as “pseudotumor cerebri” but more appropriately called “idiopathic intracranial hypertension” (IIH), it is a surprisingly common disorder. In young overweight women, the annual incidence is as high as 20 per 100,000 persons (2).

The definition of IIH has evolved with clinical experience and advances in imaging technology. Currently, IIH can be diagnosed only if the following criteria are met (Table 1): 1) symptoms and signs attributable to increased ICP or papilledema; 2) elevated ICP recorded during lumbar puncture in the lateral decubitus position; 3) normal CSF composition; 4) no imaging evidence of ventriculomegaly or a structural cause for increased ICP, such as a brain parenchymal, ventricular, meningeal, or venous sinus abnormality; and 5) no other cause of intracranial hypertension identified, such as use of certain medications.

The diagnosis and management of IIH remains based largely on anecdotal evidence. However, substantial developments during the past two decades have provided clinicians with more tools for excluding disorders that mimic IIH and for facilitating its diagnosis and management.

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## NOMENCLATURE

The nomenclature for IIH remains controversial. “Benign intracranial hypertension” is no longer accepted, as significant visual morbidity may occur with this disorder (3). The term “pseudotumor cerebri,” a historically popular and all-encompassing term, leaves the impression that IIH is not a real disease. IIH is currently the favored term for the primary (idiopathic) disorder. For those patients with an identified cause of intracranial hypertension without structural brain imaging or CSF constituent abnormalities, the appropriate diagnostic term would be “intracranial hypertension secondary to (...).” The typical patient with IIH is an obese woman of childbearing age (4). Atypical patients include men, slim women, prepubescent children, and patients older than age 44 years.

## NATURAL HISTORY AND VISUAL PROGNOSIS

There are few prospective data in the era of modern imaging (computed tomography [CT], magnetic resonance imaging [MRI], and magnetic resonance venography [MRV]) to document the natural history of the disorder. Clinical experience, however, suggests that it is common for patients to have a protracted course lasting months to years, during which they may be asymptomatic but have chronic papilledema, or have symptoms that require medical agents to lower their ICP. This suggests that intracranial hypertension, symptomatic or not, persists in many patients with IIH. Indeed, one series found that 10 (83%) of 12 patients in long-term follow-up who underwent repeated lumbar punctures showed persistently elevated ICPs ranging from 220 to 550 mm H<sub>2</sub>O (5). Recurrent symptoms and papilledema have been reported in 8% to 37% of patients often years after the initial diagnosis (5,6).

The principal morbidity of IIH is papilledema-associated visual loss. A prospective study of 50 patients with IIH found visual field defects at initial presentation in at least one eye in 96% of patients assessed with the Goldmann perimeter, and in 92% of patients assessed with the Humphrey perimeter (4). After treatment, 60% of patients improved, 30% remained stable, and 10% worsened, as assessed with Goldmann perimetry (4). Assessed with the Humphrey perimeter, 50% of patients improved, 28% remained stable, and 22% worsened (4).

**TABLE 1. Clinical criteria for diagnosing idiopathic intracranial hypertension\***

Symptoms, if present, represent increased intracranial pressure or papilledema
Signs represent increased intracranial pressure or papilledema
Documented elevated intracranial pressure during lumbar puncture measured in the lateral decubitus position
Normal cerebrospinal fluid composition
No evidence of ventriculomegaly, mass, structural, or vascular lesion on magnetic resonance imaging or contrast-enhanced computed tomography for typical patients, and magnetic resonance imaging and magnetic resonance venography for all others
No other cause (including medication) of intracranial hypertension identified

\* Adapted from reference 7.

## DIAGNOSIS

### Imaging

With advances in neuroimaging techniques and a growing understanding of the pathophysiology of IIH, the diagnostic criteria for this condition have recently been revised (7) (Table 1). A noncontrast CT was previously considered an adequate imaging study because it can exclude ventriculomegaly or a mass lesion. However, conditions that increase ICP without producing ventriculomegaly or mass lesions, such as gliomatosis cerebri, meningitis, and cerebral venous thrombosis, may mimic IIH yet not have associated CT abnormalities to provide a clue to the true underlying condition. Accordingly, this imaging technique may be suboptimal, particularly for atypical patients. Unless there are external constraints (weight limitations, availability), MRI with MRV is currently the study of choice. Using a special technique, three-dimensional gadolinium-enhanced MRV appears to be more sensitive than conventional MRV for detecting areas of subtle cerebral venous stenosis (8). The clinical relevance of these changes is uncertain, and likely reflects a compensatory response to increased ICP.

### CSF Opening Pressure

The medical literature contains various, and sometimes conflicting, recommendations regarding the minimum CSF opening pressure required for diagnosing IIH. In general, however, a CSF pressure greater than 250 mm H<sub>2</sub>O is consistent with the diagnosis, less than 200 mm H<sub>2</sub>O is normal, and 201 to 249 mm H<sub>2</sub>O is nondiagnostic (9). Contrary to popular belief, there is no evidence that body weight influences these cutoff values. The upper limit of normal

CSF pressure in children is generally considered to be 180 to 200 mm H<sub>2</sub>O; the effect of obesity in children has not been studied.

### Visual Fields

Although visual acuity and color perception are generally preserved in papilledema unless it enters a chronic and atrophic stage (10–12), visual fields and contrast sensitivity may be abnormal earlier. Visual field testing is far more sensitive for detecting optic nerve damage producing visual loss, particularly in the early stages of the disorder. Quantitative perimetry with static or kinetic methods is the current standard for assessing visual fields in IIH. The sensitivity to the detection of visual field defects is similar using either technique, assuming an experienced perimetrist performs the kinetic test (11,13).

Newer perimetric techniques, such as frequency-doubling technology perimetry, short-wavelength automated perimetry, tendency-oriented perimetry, and high-pass resolution (ring) perimetry have been examined in patients with glaucoma, but, with the exception of high-pass resolution perimetry (14), not well-studied in other optic neuropathies or in IIH. Motion perimetry, in which computer graphics generate small circular regions of coherent motion perception targets throughout the central visual field, identified the visual field defects in patients with IIH detected with conventional automated perimetry, as well as some defects that were not identified using automated perimetry (15). These results, and those elicited with other newer perimetric techniques, must be confirmed and validated before the newer tools replace the current visual field testing methods.

### Monitoring the Optic Nerve Head

Whereas the results of visual field testing provide functional information concerning the degree of optic nerve damage, assessment of the degree of papilledema change over time often provides a useful structural measure of the clinical course and effect of treatment. In some patients, however, papilledema never resolves completely despite resolution of symptoms and stabilization of visual function. It is important to document the appearance of the optic disc with photographs, ideally at the first evaluation and whenever there is a change.

Confocal scanning tomography is a new tool that can quantify the degree of papilledema and measure changes over time (16). Tomographic parameters of the optic nerve head seem to correlate with visual field sensitivity loss (17). For routine patient care, however, confocal scanning tomography is not a practical tool and may not provide more useful information than carefully performed and interpreted visual fields. Future studies are needed before deciding if this technique will find a place in routine patient management.

**Visual Acuity**

Loss of visual acuity generally does not occur in acute papilledema unless there is macular edema. As untreated papilledema becomes more chronic, however, progressive impairment of visual acuity can be expected from a variety of causes (Table 2).

**Contrast Sensitivity**

Loss of contrast sensitivity is frequently identified in patients with IIIH, regardless of the technique used (10–12). For that reason, some investigators recommend contrast sensitivity testing as an adjunctive measure to assess optic nerve function. Whereas this tool may detect a global abnormality of optic nerve function when other standard measures are normal (11), its specificity for optic nerve dysfunction is low.

**Visual Evoked Potentials**

Assessment of visual evoked potentials (VEP) is often performed to screen for injury to the optic nerve. However, this technique probes only the central 10 degrees of visual field, a region that is insensitive to visual loss in papilledema (11). Thus, there is no role for VEP in evaluating patients with IIIH. The future role of multifocal VEP, which is capable of assessing nonfoveal neurotransmission, remains to be determined.

**RISK FACTORS FOR VISUAL LOSS**

Several clinical series have identified factors that may influence visual outcome in patients with IIIH (Table 3) (2,18–22). The reliability of these variables in clinical practice may be limited on an individual case basis because they were determined from retrospective studies. Some reports provide conflicting results.

**MIMICKERS OF IIIH**

As long as the diagnostic guidelines outlined in Table 1 are followed, there is little chance of failing to diagnose an ominous mimicker of IIIH. Still, some cases of cerebral venous sinus thrombosis, gliomatosis cerebri, and leptomeningeal infiltration by a chronic neoplastic or infectious process may escape detection with brain imaging and CSF

**TABLE 2. Causes of loss of visual acuity in idiopathic intracranial hypertension**

Chronic (atrophic) papilledema
Chorioretinal folds
Macular edema or exudates
Infarction of the optic disc
Subretinal peripapillary hemorrhage extending through the fovea
Subretinal peripapillary neovascular membrane

**TABLE 3. Predictors and nonpredictors of visual loss in patients with idiopathic intracranial hypertension\***

Factors predictive of visual loss
Recent weight gain
High-grade papilledema
Atrophic papilledema
Subretinal hemorrhage
Significant visual field loss at presentation
Hypertension
Factors not predictive of visual loss
Duration of symptoms
Transient visual obscurations
Double vision
Pulsatile intracranial noises
Degree of headache
Opening pressure during lumbar puncture
Pregnancy

\* Adapted from references 2, 17–21.

analysis until late in their course. Red flags that should signal the possible presence of a mimicker are outlined in Table 4 (23–26).

One should be particularly cautious about falsely diagnosing IIIH in patients with abnormal visual fields that are psychogenic in nature, in those with anomalous optic discs, and when the opening pressure during lumbar puncture was improperly measured. Because concentric visual field constriction is a finding common to both psychogenic visual loss and IIIH, misinterpretation of this visual field defect is a particularly frequent problem (5).

**TREATMENT**

**Indications for Treatment**

Not all patients with IIIH require treatment. After establishing the diagnosis, asymptomatic individuals with

**TABLE 4. “Red flags” that suggest the presence of a mimicker of idiopathic intracranial hypertension**

Atypical demographic profile
Cranial nerve palsies other than sixth nerve palsy
Alteration in level of consciousness
Focal neurologic signs apart from sixth nerve palsy
Abnormal CSF profile (low glucose or high protein concentrations)
Explosive onset of symptoms
Rapid development of visual loss and progression of symptoms
Global ophthalmoparesis
Internuclear ophthalmoplegia
Vertical gaze disorder

normal vision and minimal papilledema can be monitored frequently for the development of symptoms or visual decline. A small percentage of patients improve after their diagnostic lumbar puncture (LP). The reason for the apparent cure is uncertain and may relate to re-establishing normal CSF homeostasis or cerebral venous pressure when normal CSF pressure is temporarily restored (27). Patients experiencing transient visual obscurations with normal visual function may be observed unless they have moderately severe papilledema. Some patients with headaches and minimal visual signs (visual field loss limited to a slightly enlarged blind spot) may also be managed conservatively.

Therapy is initiated in the presence of visual acuity or visual field loss (apart from mild enlargement of the blind spot), moderate to severe (Frisén grade 3–5) papilledema or persistent headaches (28). Visual signs and symptoms often co-exist with headache, but the two manifestations are approached independently. Treatment is always indicated when patients are aware of their visual deficit.

### Dietary Management

Dietary management and weight loss are time-honored treatments, supported by several observational studies (29–36). The earliest report described rapid resolution of papilledema in nine obese patients treated with a very low-calorie (400–1,000 calories daily) rice diet (29). One retrospective study correlated weight loss to papilledema in 15 women with IIH who were treated with acetazolamide at the time of diagnosis (30). Within the 24-week study period, 11 patients had improvement or resolution of papilledema. The six patients who had complete resolution of marked papilledema underwent a mean weight loss of 6.2% total body weight. Patients who were unable to achieve the same degree of weight loss had lesser degrees of improvement in their papilledema grade. The four patients with unchanged optic disc swelling had no weight loss during the study period.

Another retrospective series evaluated the effect of weight loss on visual function and papilledema grade in 58 patients (31). There was more rapid improvement in papilledema and visual fields in overweight women with IIH who lost weight (mean weight loss  $13.3 \pm \text{SD } 9.9$  lb) than in those who did not lose weight (mean weight loss  $0.2 \pm \text{SD } 0.6$  lb). Surgically induced weight loss (mean loss  $57 \pm 5$  kg) was associated with decreased CSF pressure and resolution of papilledema in eight patients examined  $34 \pm 8$  months postoperatively (32). Various procedures used over the 11-year study period included horizontal gastropasty, vertical banded gastropasty, proximal Roux-en-Y gastric bypass, and distal gastric bypass.

There is little scientifically robust information regarding specific dietary measures for IIH. Limiting vitamin A consumption and a low tyramine diet may be beneficial

(33–36). Dietary sources rich in vitamin A include fish, eggs, carrots, sweet potatoes, leafy greens, broccoli, red bell peppers, tomatoes, apricots, and cantaloupe. Supplemental vitamin A preparations are available over-the-counter. Tyramine naturally accumulates in food during the aging process. Foods and beverage that have high tyramine content include aged cheese and meat, pickled foods, overripe or dried fruit, beer, and wine. As many of the high-tyramine foods are also migraine triggers, patients may be instructed to use resources that are available to migraineurs regarding diet.

### Repeated Lumbar Punctures

Repeated LPs are sometimes used in patients with occasional symptom relapses, in pregnant women, or in the setting of rapidly declining vision to temporarily lower the CSF pressure while planning a more aggressive treatment. However, the procedure may be painful, technically difficult to perform, and cause a low-pressure headache (37). Other complications of LP, such as infection, tonsillar herniation, radiculopathy, and arachnoiditis, are rare. Considering that 50 mL of CSF are produced in a day in humans at a rate of approximately 0.35 mL/min, 20 mL of CSF removed by LP is replenished in one hour, provided there is no persistent CSF loss through the dural puncture site or alteration in CSF production caused by the LP (38).

### Carbonic Anhydrase Inhibitors

Acetazolamide is generally accepted as a first-line medication for lowering the intracranial hypertension in patients with IIH. Its carbonic anhydrase inhibition decreases the secretion of CSF by the choroid plexus. Doses of 1 to 2 g are generally used and some advocate increasing to the maximum tolerated dose if necessary. Side effects are common but are better accepted when patients are aware of their potential occurrence and medication doses are built up gradually. Alternatively, methazolamide may be used but it has no particular therapeutic advantage over acetazolamide. Topiramate, an anti-epileptic medication with carbonic anhydrase inhibitory properties, may prove to be useful for IIH, particularly because it is also useful for headache prophylaxis and often produces weight loss (39). Currently, treatment of IIH with topiramate has not been studied and is considered off-label usage.

### Other Diuretics

Furosemide also has beneficial effects on CSF secretion and may be used (39). Other diuretics are used but no consistent therapeutic trend has been reported. Most diuretics contain a sulfa moiety that may be problematic in patients who are allergic to sulfa. Triamterene and spironolactone are useful in this circumstance, although they have no proven effect on CSF production.

**Corticosteroids**

Corticosteroids are not advocated for routine or long-term management of IIH. They are useful as an adjunctive treatment in patients with rapid deterioration while arranging a surgical procedure (see “malignant” IIH) (40). Withdrawal of corticosteroids may lead to a rebound increase in ICP (41,42). Moreover, their side effects (weight gain, fluid retention, hyperglycemia) are problematic in IIH patients.

**Management of Headaches**

The chronic headaches of IIH are best treated with conventional headache prophylaxis, although in some cases lowering the ICP with medical methods is effective (37). Because of the potential dangers, we do not advocate CSF shunting procedures for headache alone. At the same time, many of the agents used for headache prophylaxis in IIH (tricyclic antidepressants, selective serotonin reuptake inhibitors, sodium valproate, calcium channel blockers) may produce weight gain or edema that is undesirable in this population. Many patients with IIH also have migraine, tension-type headaches, or analgesic overuse headaches (43). Headache prevention is recommended in IIH as long as the patient is monitored for medication-induced weight gain (43).

Medical treatment of IIH is seldom life-long. When the patient’s visual status and optic nerve appearance have stabilized, or when the disease has been in remission for at least six months, ICP-lowering agents may be tapered and discontinued. Patients should still be periodically monitored at this point because recurrences are not rare. Weight gain is associated with recurrence in some patients (44). Recurrence of symptoms may warrant reinstitution of medications but headaches are typically managed without diuretics or carbonic anhydrase inhibitors unless there is evidence of elevated ICP.

**SURGICAL TREATMENT**

Surgery is considered under the following circumstances: 1) progressive loss of vision despite maximal medical therapy; 2) severe or rapid visual loss at onset (see “malignant IIH”), including the development of an afferent pupillary defect or signs of advancing optic nerve dysfunction; and 3) severe papilledema causing macular edema or exudates (37,45).

Surgical procedures used for the treatment of visual loss include optic nerve sheath decompression (ONSD) and CSF diversion procedures. Whether one procedure is superior to the other is controversial and the decision often depends on available resources and expertise. The success rate is comparable between ONSD and lumboperitoneal shunt (46). Advantages of ONSD are shorter anesthesia and hospitalization times. ONSD avoids the complications of

shunting (Table 6). Approximately 50% of patients experience improvement in the nonoperated eye after a single ONSD (47).

**Optic Nerve Sheath Decompression**

The mechanism by which ONSD benefits IIH is uncertain. One possibility is a filtering effect with local CSF pressure reduction improving the peripapillary circulation (47). A second possibility is a generalized decrease in ICP, which has been demonstrated after ONSD (48,49). A third possibility is that scarring of the arachnoid after the procedure may protect the nerve head from the elevated CSF pressure. ONSD in monkeys produces connective tissue proliferation and obliteration of the subarachnoid space near the area of operation (50); similar changes were found in one human postmortem study (51).

The complication rate of ONSD ranges broadly from 4.8% to 45%, with a mean of 12.9% (52–62). Meta-analysis is difficult because there are so many variables, including the surgical approach (medial versus lateral), surgical experience, duration of follow-up, criteria for an event to be considered a complication, and primary procedure versus reoperation. Complications of ONSD are listed in Table 5. In a review of 317 published cases by Anthony Arnold, MD (presented at the International Neuro-Ophthalmology Society Meeting, 2002), failure (progressive visual loss postoperatively or need for reoperation) occurred in 42 cases (13%). The most commonly reported complications are extraocular motility dysfunction (often transient) and pupillary abnormalities. Extraocular movement dysfunction, usually caused by lateral rectus palsy, generally resolves. Visually threatening complications are rare. Transient and protracted postoperative blindness have been reported (59,60) and are attributed to ischemic injury to the optic nerve. The complication risks after reoperation are approximately the same as those for the first procedure (61,62).

**TABLE 5. Complications of optic nerve sheath decompression**

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Extraocular movement dysfunction
Pupillary dysfunction
Vascular occlusion
Central retinal artery occlusion
Branch retinal artery occlusion
Choroidal ischemia/infarction
New visual field defect
Orbital hemorrhage
Transient or protracted blindness
Globe perforation

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**TABLE 6. Complications of lumboperitoneal shunting**

Shunt valve or tubing obstruction
Over-shunting (low pressure headache)
Catheter migration/abdominal pain
Lumbar radiculopathy
Infection
CSF leak
Cerebellar tonsillar herniation (acquired Chiari malformation)
Shunt dependency

### CSF Diversion Procedures

A CSF diversion procedure treats IIH by lowering ICP but requires insertion of a foreign body. Lumboperitoneal shunting is more commonly performed than ventriculoperitoneal (VP) shunting because insertion and maintenance of patency may be more difficult in the latter procedure. However, VP and cisterna magna shunting may be successfully used (63,64,67). Complications of shunts are summarized in Table 6. The revision rate for lumboperitoneal shunts ranges from 38% to 64%, with an overall revision rate of 52% (78 of 150 cases) (46,61–66). The number of revisions per patient is 2.3% to 6.6% (mean 3.9%), but this value may be skewed because of the small number of reported patients. The reported interval between shunt placement to first revision is 9 to 27 months (46,66,67). Major causes of shunt failure include catheter obstruction, low ICP, catheter migration, and lumbar radiculopathy (66,68). A programmable shunt valve usually prevents low-pressure headaches, obviating the need for reoperation; this complication is less frequent in VP than in lumboperitoneal shunting. Visual loss may herald shunt malfunction, but may also occur with a functioning shunt (66–75). Uncommonly, patients may become “shunt dependent” after being in remission for years, with worsening of signs and symptoms when the shunt is obstructed or removed. Severe, acute ICP elevation upon insertion or removal of a shunt has been reported (70).

Over-shunting may lead to an acquired Chiari malformation or chronic intracranial hypotension. The symptoms and signs of low ICP are often similar to those of elevated ICP. Most patients will experience a postural headache that worsens with sitting or standing. Neck pain, vomiting, photosensitivity, blurred vision, transient visual obscurations, peripheral visual field loss, and sixth nerve paresis may occur (71). MRI changes of intracranial hypotension include leptomeningeal enhancement, tonsillar herniation, and subdural effusions (72,73).

### Bariatric Surgery

Bariatric surgery may be considered in morbidly obese patients in whom medical and surgical treatments are

ineffective (31). Although the procedure has risk, it offers the additional health benefits of reduced cardiovascular risk, type II diabetes, and lumbar disc degeneration that occur with significant weight loss. Bariatric surgery may be considered for long-term management but is not an appropriate treatment of patients with actively worsening vision.

## SPECIAL CONSIDERATIONS

### “Malignant” IIH

Aggressive treatment is required for patients in whom rapid visual decline (“malignant” IIH) develops. Significant visual field loss and marked papilledema are evident at presentation, often with decreased visual acuity. Visual loss may occur rapidly over days to weeks. Temporizing management includes serial lumbar punctures or insertion of a lumbar drain, and the administration of intravenous acetazolamide and corticosteroids (76). Prompt surgical treatment is indicated with ONSD, shunting, or both procedures. Cerebral venous sinus thrombosis is an important diagnostic exclusion as it is managed with heparinization and, in some cases, thrombolytic treatment. Occasionally, the thrombosis may not be apparent on the initial MRI study, and re-imaging or catheter angiography may be fruitful.

### IIH in Pregnancy

Although pregnancy is not considered an independent risk factor for IIH, the disease may start or worsen during pregnancy (21,77). IIH during pregnancy is managed in cooperation with the obstetrician. Often the condition is successfully controlled with headache management and serial LPs. Patients are advised to avoid excessive weight gain with guidance from their obstetrician. Low-calorie diets and weight reduction are not recommended. Acetazolamide is a category C medication in pregnancy (risk cannot be ruled out because data are lacking). However, large clinical experience among neuro-ophthalmologists indicates overall safety without known teratogenic effects on the fetus, especially if the medication is used after the first trimester. Another option is chlorthalidone, a diuretic with a category B rating (no evidence of risk in humans). Corticosteroids may be administered without undue risk if needed for visual loss. Surgery is rarely required. If it is, ONSD is preferred over shunting because of potential shunt obstruction by the enlarging uterus (77). No special measures are required for delivery, and vaginal delivery is not contraindicated.

### IIH in Children

Treatment of children with IIH is similar to that of adults, with the caveat that a secondary cause is often found in children (78–81). IIH in children may follow a febrile

illness, and common secondary causes are tetracycline (including minocycline), hypervitaminosis A (including retinoid use), and cerebral venous sinus thrombosis (81).

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