Idiopathic intracranial hypertension (pseudotumor cerebri): Clinical features and diagnosis

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INTRODUCTION — Idiopathic intracranial hypertension (IIH) is also commonly called pseudotumor cerebri. It is a disorder defined by clinical criteria that include symptoms and signs isolated to those produced by increased intracranial pressure (eg, headache, papilledema, vision loss), elevated intracranial pressure with normal cerebrospinal fluid composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations [1].

While once called benign intracranial hypertension, to distinguish it from secondary intracranial hypertension produced by a neoplastic malignancy, it is not a benign disorder. Many patients suffer from intractable, disabling headaches, and there is a risk of severe, permanent vision loss.

This topic will discuss the clinical features and diagnosis of IIH. The epidemiology and pathogenesis, as well as the prognosis and treatment of this disorder are discussed separately. (See "Idiopathic intracranial hypertension (pseudotumor cerebri): Epidemiology and pathogenesis" and see "Idiopathic intracranial hypertension (pseudotumor cerebri): Prognosis and treatment").
SYMPTOMS — In one case series, the most common symptoms of idiopathic intracranial hypertension (IIH) were [2]: Headache (92 percent) Transient visual obscurations (72 percent) Intracranial noises (pulsatile tinnitus) (60 percent) Photopsia (54 percent) Retrobulbar pain (44 percent) Diplopia (38 percent) Sustained visual loss (26 percent)

These symptoms, even as a cluster, are not specific for IIH. In one case-control study, these symptoms were also common in age and gender-matched controls who were recruited from hospital waiting areas; although the prevalence, severity, and frequency were less in this group [3].

Headache — Headache is the most common presenting symptom of IIH. However, the features of headaches in IIH patients are variable and are not specific to IIH. Many, but not all patients note that the pain is of unusual severity [2,4]. The headaches are often lateralized and throbbing or pulsatile in character. They may be intermittent or persistent, occur daily or less frequently. Associated nausea and vomiting are not infrequent. Some patients describe headache exacerbation with changes in posture and some may report that relief occurs with nonsteroidal anti-inflammatory medications and/or rest. Retrobulbar pain and pain with eye movement or globe compression are somewhat more specific features for IIH. In some patients, the pain follows a trigeminal or cervical nerve root distribution [4]. Neck stiffness is also commonly reported [2,5].

In most cases, the features of headache are consistent with other primary headache disorders including migraine and tension-type headache [6,7]. The often refractory nature of the headache may lead the patient to overuse analgesic medication, suggesting or even causing a superimposed rebound headache, further obscuring the diagnosis [8]. (See "Headache syndromes other than migraine", section on Medication overuse headache).

Rare patients present without headache [9,10]. Among younger children, headache is a less universal finding [5,11]; in one series, 29 percent of children with IIH did not have headache [12]. In one large case series, men were less likely to complain of headache than women [13].

Transient visual obscurations — Transient visual obscurations occur in about two-thirds of patients with papilledema. These last seconds at a time and can be bilateral or unilateral [2]. The frequency is variable, ranging from rare or isolated episodes to those occurring several times a day. Some patients note that these can be precipitated by changes in position (usually standing,
but sometimes lying down or bending over), Valsalva, bright light, or eye movement (ie, gaze-evoked) [2,3]. The occurrence of transient visual obscurations does not appear to correlate with the degree of intracranial pressure elevation or the extent of disc swelling, and is not predictive of future visual loss [3,14].

Photopsias, brief sparkles or flashes of light, can also occur in patients with IIH and, similar to visual obscurations, can be provoked by positional changes and Valsalva [3].

Intracranial noise — Pulsatile tinnitus is common in IIH and in the setting of headache is somewhat specific for the diagnosis [3,8,15]. Patients often describe hearing rushing water or wind. This symptom can be persistent or intermittent and is believed to represent vascular pulsations transmitted by cerebrospinal fluid under high pressure to the venous sinuses [16].

Diplopia — Patients with IIH may report intermittent or continuous horizontal diplopia. This is typically due to a unilateral or bilateral sixth cranial nerve palsy from increased intracranial pressure. Rarely, other causes of diplopia can occur in IIH (eg, other cranial neuropathies, decompensated phoria) [17]. (See "Other cranial nerve deficits" below).

EXAMINATION — The most common signs in IIH are Papilledema Visual field loss Sixth nerve palsy

Papilledema — Papilledema is the hallmark sign of IIH (show picture 1A-1B). Papilledema is described in detail separately. (See "Overview and differential diagnosis of papilledema").

While typically bilateral and symmetric, papilledema may be asymmetric or frankly unilateral [17-20]. In one series, 10 percent of 478 IIH patients had highly asymmetric papilledema with greater visual loss in the eye with higher grade of papilledema [18]. Such patients may also have a relative afferent pupillary defect.

Papilledema can be graded in severity. We use the Frisén scale for grading papilledema. Patients with more severe papilledema are at higher risk of permanent visual loss [18]. We take photographs of both optic discs and grade the severity of papilledema at the initial visit and each follow-up. (See "Idiopathic intracranial hypertension (pseudotumor cerebri): Prognosis and treatment", section on General treatment and follow-up considerations).
Other findings on funduscopic examination may include choroidal compression folds across the macula, choroidal neovascularization, and serious retinal elevation around the nerve head [21].

There are some reports of IIH without papilledema [7-9,22-24]. These patients typically present with intractable headaches and are diagnosed with IIH after an elevated opening pressure is documented on lumbar puncture. We consider the absence of papilledema to be a rare occurrence in IIH [1]. Such patients are typically not at risk for vision loss [7,25].

Visual loss — Loss of vision is the major morbidity in IIH and may be present on initial evaluation [2,14]. Vision loss is usually gradual but can be abrupt. Such patients have a more fulminant course and more significant permanent vision loss. (See "Idiopathic intracranial hypertension (pseudotumor cerebri): Prognosis and treatment", section on Fulminant IIH).

Visual acuity is less than 20/20 in 10 to 29 percent of patients on presentation [2,26-28]. However, visual acuity is an insensitive measure of vision loss in IIH. Visual field loss occurs before loss of acuity; confrontation visual fields are abnormal (nasal loss, temporal loss, visual blurring) in up to 32 percent at presentation [2,28]. Perimetry gives a more accurate and detailed assessment of visual field abnormalities and is an essential feature of the evaluation of a patient with IIH. (See "Visual field testing" below).

The visual field loss is typically peripheral with predominate nerve fiber bundle type defects. Central visual field can be involved late in the course or earlier if there is concomitant macular pathology (serous detachment, macular hemorrhage, or edema), choroidal folds, or choroidal neovascular membrane. In addition to chronic papilledema, other mechanisms, such as a serous retinal detachment, may produce visual loss.

Abducens palsy — A sixth cranial nerve (abducens) palsy may be unilateral or bilateral in patients with IIH [9]. This reflects a nonlocalizing effect of elevated intracranial pressure on the sixth nerve, which has a long intracranial course before exiting the skull.

Other cranial nerve deficits — Other cranial nerve deficits that resolve with IIH treatment are noted in case reports. These may be more common in prepubertal children than in older patients.
DIFFERENTIAL DIAGNOSIS — Because the headache features of IIH are nonspecific, a fundoscopic examination is critical to identify patients with IIH. When papilledema is present, this suggests elevated intracranial pressure, which can have many etiologies in addition to IIH. There are also many etiologies of unilateral or bilateral optic disc swelling, which can have a similar appearance to papilledema.

Secondary intracranial hypertension — Any entity that increases intracranial pressure may lead to papilledema. These include: Intracranial mass lesions (tumor, abscess) Increased cerebrospinal fluid (CSF) production, eg, choroid plexus papilloma Decreased CSF absorption, eg, arachnoid granulation adhesions after bacterial or other infectious meningitis, subarachnoid hemorrhage Obstructive hydrocephalus Obstruction of venous outflow, eg, venous sinus thrombosis, jugular vein compression, neck surgery Idiopathic intracranial hypertension (pseudotumor cerebri)

Secondary intracranial hypertension due to cerebral venous thrombosis can have a very similar clinical presentation as IIH [36-38]. Other unusual causes of obstructed venous outflow include transverse sinus septum causing sinus stenosis [39]; osteopetrosis of the jugular foramen [40-44]; depressed skull fracture and stenosis of the superior sagittal sinus [45]. Venous hypertension and secondary increase in intracranial hypertension can also be caused by cerebral arteriovenous malformations, dural arteriovenous malformations, and arteriovenous fistulas [27,46-55]. Some patients thought to have IIH have later been discovered to have one of these conditions [27,36,56,57].

Others — There are many causes of an elevated optic nerve head. While the term papilledema is sometimes used to describe the findings in these conditions, it should be reserved for patients who have elevated optic disc heads as a consequence of increased intracranial pressure. The Table lists the causes of optic disc swelling (show table 1). These are discussed in detail separately. (See "Overview and differential diagnosis of papilledema")

DIAGNOSIS — IIH is diagnosed according to the modified Dandy criteria [1]: Symptoms and signs of increased intracranial pressure (eg, headache, transient visual obscurations, pulse synchronous tinnitus, papilledema, visual loss) No other neurologic abnormalities or impaired level of consciousness Elevated intracranial pressure with normal cerebrospinal fluid (CSF) composition A
neuroimaging study that shows no etiology for intracranial hypertension. No other cause of intracranial hypertension apparent.

It follows that the clinical evaluation of IIH includes a complete history, including documentation of any conditions or medications associated with IIH. (See "Idiopathic intracranial hypertension (pseudotumor cerebri): Epidemiology and pathogenesis", section on Associated conditions).

A complete ocular exam should document formal visual field examination, dilated fundus examination, and optic nerve photographs. Neuroimaging is required to exclude secondary causes of intracranial hypertension, followed by a lumbar puncture to document opening pressure and exclude other conditions.

All patients with bilateral optic disc edema should have a measurement of the systemic blood pressure as optic neuropathy related to malignant hypertension can mimic papilledema and can also produce headache and other symptoms that might be mistaken for IIH. (See "Hypertensive emergencies: Malignant hypertension and hypertensive encephalopathy").

Some also suggest that all patients with IIH have a complete blood count to exclude an anemia that may be causative or contributory to the patient's condition [58].

Neuroimaging — In a patient with headache and papilledema, the purpose of neuroimaging is to exclude secondary causes of increased intracranial hypertension. (See "Secondary intracranial hypertension" above).

Magnetic resonance imaging (MRI) is the preferred test. A CT scan may be necessary for patients with contraindications to MR imaging (eg, pacemakers, metallic clips in head, metallic foreign bodies) and obese or claustrophobic patients. The use of contrast enhancement increases the sensitivity of the study particularly for subtle intracranial masses (eg, gliomatosis cerebri) and meningeal-based pathologies [59].

The MRI often shows abnormalities suggestive of IIH [60-63]: Flattening of the posterior sclera (80 percent) Distension of perioptic subarachnoid space (50 percent) Enhancement (with gadolinium)
of the prelaminar optic nerve (45 percent) Empty sella (70 percent) Intraocular protrusion of the prelaminar optic nerve (30 percent) Vertical tortuosity of the orbital optic nerve (40 percent)

These findings are not, individually or in the aggregate, diagnostic of IIH [60,64].

While many cases of cerebral venous thromboses (CVT) are visualized on MRI, MR venography (MRV), particularly with contrast administration, is more sensitive [56]. It is our practice to include a postcontrast MRV with MRI when evaluating patients with suspected IIH. If a patient comes to us with a normal MRI without MRV, and there are no other risk factors for CVT (including no oral contraceptive use), and the patient has a typical risk profile for IIH (an obese woman of childbearing age), we may defer MRV unless there is rapid clinical progression or poor response to treatment.

There are many reports of cerebral venous abnormalities on MRV in patients with IIH. While the presence of clear venous sinus thrombosis had definite clinical significance, the relationship between apparent venous sinus narrowing or stenosis is unclear. (See "Idiopathic intracranial hypertension (pseudotumor cerebri): Epidemiology and pathogenesis", section on Intracranial venous hypertension). Patients with equivocal findings on MRV may require further diagnostic evaluation to exclude CVT. (See "Etiology; clinical features; and diagnosis of cerebral venous thrombosis", section on Diagnosis).

Lumbar puncture — If the neuroimaging study reveals no structural etiology for intracranial hypertension, a lumbar puncture (LP) is performed. In addition to measuring the opening pressure, the cerebrospinal fluid (CSF) is analyzed for cell count and differential, glucose, and protein. Appropriate CSF studies for microbial agents, CSF cytology, and antigen testing (eg, CSF VDRL) may be indicated if the CSF content is abnormal or the clinical situation suggests additional testing. (See "Lumbar puncture: Technique; indications; contraindications; and complications in adults").

The upper limit of normal for opening pressure in adults is 200 mmH2O. Some believe that obese patients may have a higher upper limit of normal, with opening pressures that may normally approach 250 mmH2O [65]. However, others have not correlated obesity with elevated intracranial pressure in the absence of IIH [66,67]. We and others consider pressures less than 200 mmH2O to be normal, greater than 250 mmH2O to be abnormal, and 200-250 mmH2O to be equivocal [1,59,66]. In young children, a lower upper limit of normal, 180 mmH2O, may be more appropriate, but this is not defined [25,26].
For accurate recordings, the patient should be relaxed and lying in the lateral decubitus position with legs extended [59]. Other positions (prone, sitting) can give falsely elevated readings, as can anxiety and pain. Misleading low readings can be obtained after multiple LP attempts, or in the setting of hyperventilation and treatment with intracranial pressure lowering medications. CSF pressures can vary, and a normal reading in a patient with IIH may reflect an atypically low reading for that patient. Repeating the LP may be required in a patient if suspicion for IIH remains high after one normal CSF reading [1,25]. In rare patients, CSF pressure monitoring may be required to document elevated CSF pressure, but this is exceptional [68,69].

Visual field testing — Visual field testing is essential in IIH to assess the severity of optic nerve involvement and monitor response to treatment. Options include Goldmann kinetic perimetry and the computer-assisted static perimetry. While each has advantages and limitations, the latter is generally preferred, as it provides a more reliable measure for follow-up examinations [21].

The most common findings on perimetry are [21,70-72]: Enlarged blind spot Generalized constriction Inferonasal vision loss

Less commonly central, paracentral arcuate and altitudinal scotomas may occur [59].

The frequency of visual field loss and acuity loss with IIH is somewhat variable. In one case series, Goldmann perimetry was abnormal in at least one eye in 96 percent of patients on the initial evaluation [2]. Grade 2 loss or higher in at least one eye was present 62 percent. In other reports, visual field loss was noted in 71 to 100 percent of eyes using various forms of perimetry [11,27,70,73].

Other testing — Confocal scanning tomography is a technique that provides a quantitative measure of papilledema and has been shown to correlate with CSF opening pressure and with visual field sensitivity losses and recovery with treatment [74-76]. While this technique may prove clinically useful, it is not widely available, nor is it clear that it offers significant clinical information over perimetry.
SUMMARY AND RECOMMENDATIONS — The clinical features of idiopathic intracranial hypertension (IIH) are believed to result directly from increased intracranial pressure and in most patients include headache and papilledema. The characteristics of the headache in IIH are variable and nonspecific, but usually include daily occurrence, unusual severity, and a throbbing quality. (See "Headache" above). Papilledema is usually bilateral and symmetric; the severity of papilledema is associated with the risk of permanent visual loss. Grading the severity of papilledema and taking photographs of the optic discs is a useful mechanism for following the patient’s course and response to treatment. (See "Papilledema" above). Other common features of IIH that are unusual in other primary headache disorders are transient visual obscurations, pulsatile tinnitus, and diplopia. (See "Symptoms" above). Other common examination features include restricted visual fields and uni- or bilateral abducens palsy. (See "Examination" above). IIH must be distinguished from other causes of increased intracranial pressure and other etiologies of optic nerve head swelling. Cerebral venous thrombosis in particular may have a very similar clinical presentation to IIH. (See "Differential diagnosis" above). A neuroimaging study is required in patients suspected as having increased intracranial pressure to exclude other causes of elevated intracranial pressure. Magnetic resonance imaging (MRI) with and without contrast and including postcontrast MR venography is the imaging study of choice. (See "Neuroimaging" above). Lumbar puncture should follow MRI unless a source of elevated intracranial pressure is clearly delineated. An opening pressure greater than 250 mm water taken with the patient lying on his side with legs extended confirms elevated intracranial pressure. Pressures between 200 and 250 mm water are considered equivocal. Cerebrospinal fluid analysis should be normal in IIH. (See "Lumbar puncture" above). Visual field testing is an essential part of the evaluation of patients with IIH and provides a means for following the patient and directing treatment (See "Visual field testing" above).

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