

Idiopathic Intracranial Hypertension and the Evaluation of Suspected Papilledema

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ABSTRACT

Papilledema is swelling of the optic disc secondary to an increase in intracranial pressure. This is diagnosed not only through funduscopic examination, but also in conjunction with lumbar puncture, ultrasonography, optical coherence tomography, and neuroimaging. There are several disorders that result in increased intracranial pressure, some being potentially life threatening and therefore warranting an expeditious evaluation. As papilledema may be the earliest presenting sign of a life-threatening condition, it is often the eye care provider's role to identify and evaluate patients with suspected papilledema.

Keywords: Papilledema; Idiopathic intracranial hypertension; Cerebrospinal fluid

INTRODUCTION

Papilledema is the swelling of the optic disc only in the setting of increased intracranial pressure (ICP) [1]. Swelling of the optic nerve in papilledema is most often bilateral and symmetric, though there have been documented cases of unilateral and asymmetric papilledema [2]. The Monroe-Kellie hypothesis states that the volume of the cranium, made up of brain parenchyma, vessels, blood, and cerebrospinal fluid, is fixed. As such, any increase in one of the constituents must be offset by the other in order to maintain balance. When an increase in one of the above components is not offset by an increase in volume, intracranial hypertension occurs and may manifest clinically as papilledema [2]. Because the optic nerve is enveloped by a sheath that is continuous with the meninges of the brain, an increase in intracranial pressure is transmitted to the perioptic meninges and impairs the metabolic activity of the nerve, leading to edema [3]. The exact pathogenesis is unknown, and two theories are currently under investigation. The first, the mechanical theory, states that the elevation in ICP directly compresses the axons of the optic nerve, expanding its prelaminar axons as their axoplasm accumulates at the level of the lamina cribrosa. The second theory is the ischemic theory, which suggests that the dilatation of the optic nerve sheath compresses neurovascular structures thus impeding vascular flow to the optic nerve [3]. As the optic disc distends as a result of these mechanisms, it displaces the retina and can cause Paton’s folds, a wrinkling of the retinal nerve fiber layer appearing as greyish-white lines on funduscopic exam [4], which may appear with the presence of disc hemorrhages (Figure 1). Optic disc swelling secondary to elevated ICP often begins inferiorly, progressing superiorly and nasally, with the temporal area last to swell [1]. An important sign that may aid in the accurate diagnosis of papilledema is the loss of a spontaneous venous pulse (SVP). With elevated ICP there is an associated increase in CSF pulse pressure. When the CSF pulse pressure exceeds the retinal venous pulse pressure, the SVP is eliminated [5]. This diagnostic sign allows the clinician to accurately distinguish papilledema from ocular conditions that may mimic it, referred to as pseudopapilledema (Table 1). However, the previous documentation of a SVP is necessary as it may be absent in 10-20% of the normal population [5].

Table 1: Causes of papilledema and those conditions that may mimic its appearance.

Causes of Papilledema	Causes of Pseudopapilledema
Space-occupying tumor or mass	Optic disc drusen
Hydrocephalus	Hypoplastic disc
Dural venous sinus thrombosis	Optic neuritis
Vascular causes: stroke, AVM, aneurysm	Non arteritis ischemic optic neuropathy
Spinal cord lesion	
Infectious agents	
idiopathic	

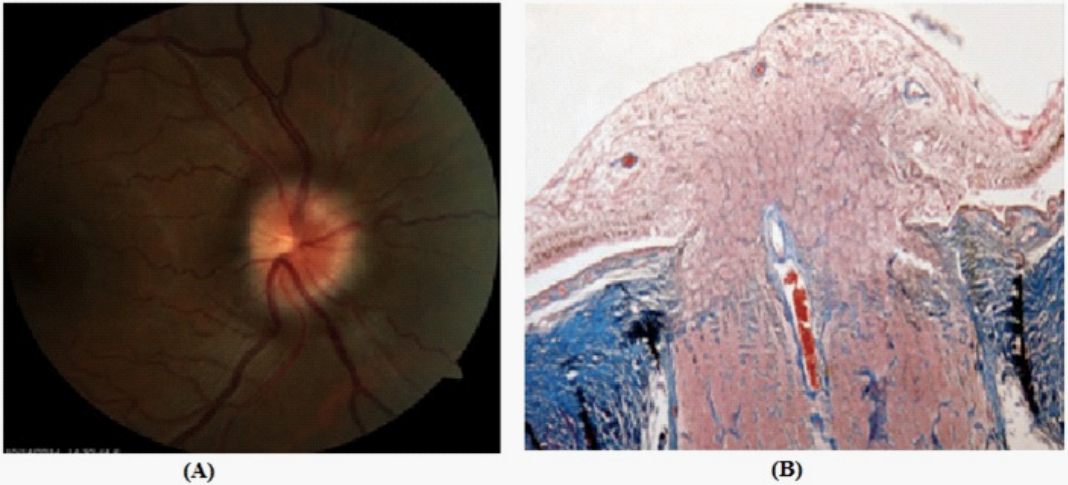


Figure 1: A (left): Papilledema on fundoscopic examination with blurred disc margins and Paton’s folds. B (right): Microscopic view of optic disc demonstrating papilledema Courtesy Dr. John Deck, Toronto, CA. Reprinted from Fig. 25-34 D p 586 in Carmody RF Chapter 25 [In Naidich TP et al. Imaging of the Brain 2013 Philadelphia, Elsevier. Reprinted with permission.

The diagnosis of papilledema, regardless of underlying cause, is most commonly confirmed by lumbar puncture demonstrating an elevated CSF opening pressure [1-3]. As it is not feasible to perform a lumbar puncture in an office setting, surrogate tests may be performed to aid in the diagnosis. In addition to the fundoscopic findings described above, B scan ultrasonography is often utilized in the differentiation of papilledema from pseudopapilledema. Patients with severe disc swelling secondary to increased ICP may present with an anechoic “crescent” sign on B scan. This appearance on imaging is due to an area of fluid accumulation within the optic nerve sheath. Optic nerve sheath diameter can also be measured through this technique, 3 mm behind the retina in the horizontal axial direction. A measurement greater than 4.5 mm is abnormal and indicative of papilledema [6]. One study reports 93% accuracy in the diagnosis of increased intracranial pressure through measurement of optic nerve sheath diameter on sonographic assessment [7].

Optical coherence tomography (OCT) is a useful, in office examination for the measurement of average peripapillary retinal nerve fiber layer and total retinal thickness, as well as optic nerve head volume [8]. This scan is often utilized in follow up evaluation of papilledema to document progression or regression of disease. However, due to the OCT’s ability to quantify retinal and optic nerve structures, it is also helpful in the initial diagnosis of optic disc swelling and in characterizing the pattern of swelling [1] (Figure 2). Visual field testing is also of great clinical value, especially in monitoring long term visual function in patients. Standard automated perimetry will most often reveal an enlarged blind spot due to the edematous nature of the optic nerve. Other common visual field defects include a generalized reduction in sensitivity, arcuate defects, nasal step, paracentral scotomata, generalized field constriction, and central scotomata [9].

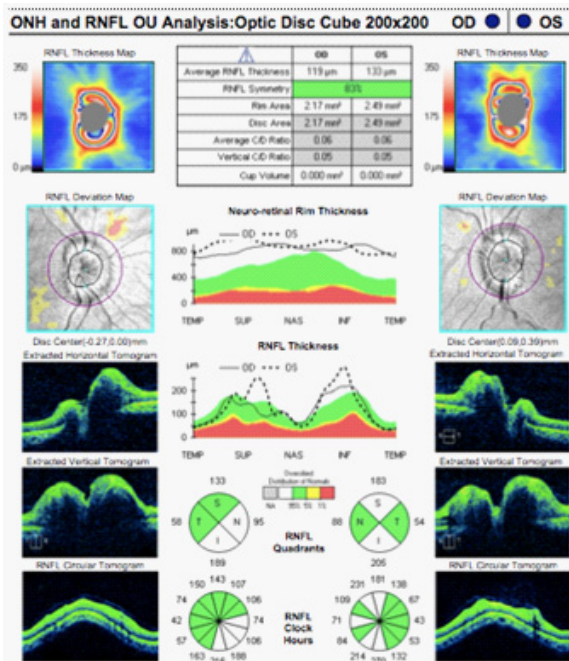


Figure 2: OCT printout in a patient with papilledema showing elevated retinal nerve fiber layer, greatest inferiorly with least swelling temporally.

Frisen L developed the numerical grading scale for papilledema based on level of severity. Stage 1 is the blurring of the nasal disc border and subtle, circumferential gray ring in the absence of temporal swelling. Stage 2 includes blurring of the temporal margin as well, with or without the presence of choroidal folds. Stage 3 is moderate papilledema, with total increase in optic nerve head diameter and the obscuration of one or more segments of the underlying blood vessels at the disc margin. Stage 4 is distinguished by complete absence of the cup and total obscuration of an underlying segment of central retinal artery or vein. Finally, stage 5 is the most severe, presenting as a dome-shaped protrusion of the nerve head and well-demarcated halo. This occurs with or without the presence of segmental obscuration of blood vessels [10].

Papilledema, if left untreated, may result in permanent vision loss. Serious unilateral or bilateral vision loss has been reported in up to 25% of patients [11]. Long-term papilledema results in optic atrophy and vision loss, which is irreversible [12].

NORMAL CEREBROSPINAL FLUID DYNAMICS

Cerebrospinal fluid (CSF) is a colorless fluid surrounding the brain and spinal cord. It provides a cushion for the brain and facilitates the spread of nutrients through the central nervous system. Microbiological studies have revealed that it also plays an integral role in maintaining homeostasis of the brain's interstitial fluid and in regulating the normal physiology of neuronal function. The production, absorption, and circulation of CSF are implicated in neurological disease processes, especially those affecting intracranial pressure [13,14].

The average volume of CSF is approximately 150 mL in adults, though there is significant individual variation. 80% of this volume is produced by the choroid plexus and in the lateral, third, and fourth ventricles and then located in the subarachnoid space, while the remaining volume is produced by the brain parenchyma and then located in the ventricles [15]. The choroid plexus secretes CSF at a rate of 20 mL per hour, with the entire volume replaced every six hours [13-16]. CSF secretion occurs in two steps. The first involves passive filtration from the choroidal capillaries to the interstitial space. This occurs across the single layer of choroidal epithelium and is driven by a pressure gradient. Active transport through membrane proteins then transports the fluid from the interstitial space to the ventricular lumen, thus constituting the second step in CSF secretion. These processes are facilitated by several proteins, including carbonic anhydrase (affected by acid and base balance), aquaporin water channels (high expression of aquaporin 1 and 4 in the brain), and membrane carrier proteins [17]. Secretion is mediated by cholinergic, adrenergic, and autonomic innervation. Sympathetic stimulation decreases CSF secretion and parasympathetic stimulation increases CSF secretion. There are circadian variations in the secretion of CSF, which are likely a result of autonomic stimulation [16].

Following secretion, CSF circulates from the lateral ventricles to the third ventricle (via the foramina of Monroe), then into the fourth ventricle (via the cerebral aqueduct), and finally from the fourth ventricle into the subarachnoid space (via the median foramen of Magendie and the lateral foramina of Luskha) [13-17]. This flow is pulsatile and synchronous with the systolic phase of the adjacent choroidal arteries [14]. Once within the subarachnoid space CSF travels caudally, towards the spinal cord, or cranially. The cranially directed CSF is absorbed by arachnoid granulations (or villi) into the dural venous sinuses. Once within the sinuses, the CSF flows with venous blood towards the skull base and ultimately into the systemic circulation via the internal jugular veins [13].

DIFFERENTIALS OF INCREASED ICP

A disruption in the aforementioned CSF pathway may cause an increase in intracranial pressure and resultant papilledema. Various etiologies include, but are not limited to, obstruction, infection, mass lesions, vascular, and idiopathic causes [18,19]. Determining the source of the pressure buildup through a thorough history and the appropriate diagnostic imaging is essential in proper treatment and management of the patient as some of these causes may be deadly.

Hydrocephalus

Hydrocephalus is enlargement of one or more components of the ventricular system that occurs from either an overproduction of CSF or impaired drainage [19]. Overproduction can be seen with tumors of the choroid plexus, namely choroid plexus papillomas and carcinomas. Impaired drainage can occur due to a multitude of causes that narrow or occlude the conduits of CSF flow. An intracranial mass, such as a colloid cyst, can occlude the foramen of Monroe. The cerebral aqueduct is a narrow conduit and flow through it may be severely impaired by a regional mass lesion or aqueductal stenosis (Figure 3), particularly in children [19,20].



Figure 3: Coronal non-contrast head CT in a 7-year old boy with hydrocephalus secondary to aqueductal stenosis. The white arrow denotes a ventriculoperitoneal shunt, which is only partly visualized.

Regardless of etiology, the excess CSF causes an elevation of ICP. Chronic headaches are a frequently reported symptom, which do not improve despite the use of pain relievers. Patients may also complain of subtle gait disturbances and cognitive dysfunction increasing with duration of the condition [20].

Diagnosis is made with either computerized tomography (CT) or magnetic resonance imaging (MRI). Imaging studies demonstrate enlargement of one or more ventricles [19,20]. If there is enlargement of only a portion of the ventricular system while the rest are not, this is referred to as non-communicating hydrocephalus, most commonly resulting from a mass lesion at the foramen of Monro or cerebral aqueduct. In communicating hydrocephalus, all of the ventricles are enlarged and this is likely due to reduced CSF resorption [19].

Space Occupying Lesion

In the presence of papilledema, it is imperative to exclude the presence of a space-occupying lesion. The skull, being a fixed space, is not subject to expansion with the development of a mass. As such, the area remains constant while the contents in the skull increase, resulting in an increase of pressure. Expansive intracranial processes may also cause alterations in intracranial volume and lead to the development of brain edema, which then causes intracranial hypertension. Edema is usually sectorial in the brain with differences in cerebrospinal fluid compartments [21]. Space occupying lesions include non-neoplastic processes, such as arachnoid cysts, and neoplastic processes, both benign and malignant. Common intracranial neoplasms in adults include

meningioma, glioblastoma, and metastasis, among others [22] (Figure 4 and Figure 5). There is either a very slow or rapid increase in intracranial pressure, dependent on type of mass lesion [21]. Brain tumors in children are more commonly located in the posterior fossa, and thus present more frequently with papilledema [2].

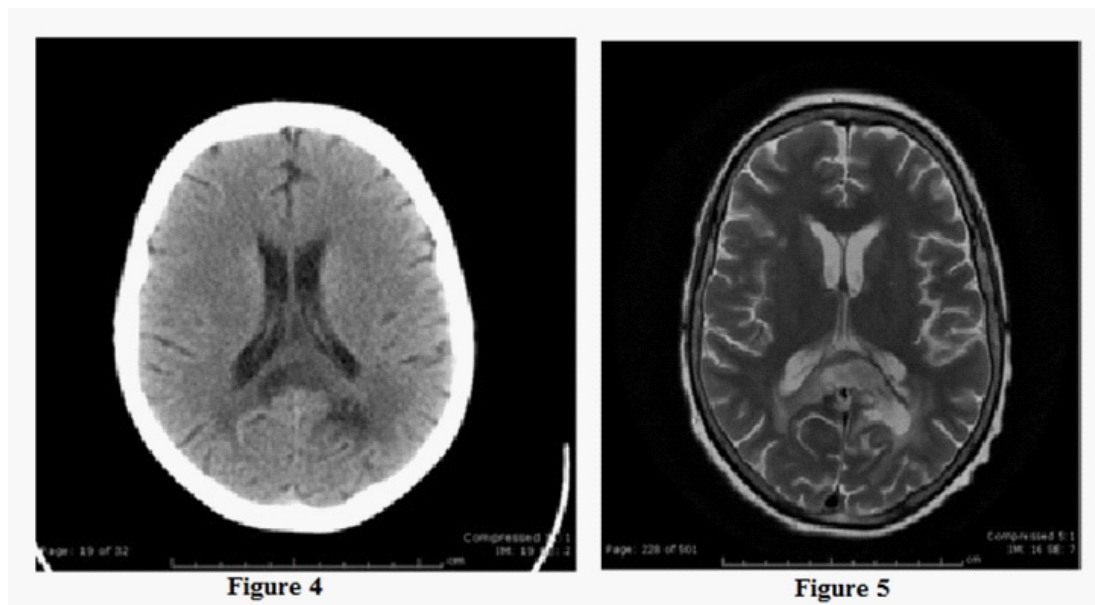


Figure 4 and Figure 5: Axial non-contrast head CT (left) and T2-weighted MRI of the brain (right) demonstrate a lesion at the splenium of the corpus callosum. Surgical pathology was compatible with glioblastoma.

Headaches are the most commonly reported symptom and are typically worse in the morning. Nausea, vomiting, and seizures have also been commonly reported [22,23]. Diagnosis is made with radiologic studies. In the evaluation of papilledema it is important to perform a baseline CT to exclude a mass prior to performing a lumbar puncture. Removing CSF in the presence of an intracranial mass may result in herniation with significant morbidity and mortality [23]. This depends largely on the size of the mass and proximity to the foramen magnum [24].

Spinal Cord Lesions

Spinal cord tumors are an important consideration in the evaluation of increased ICP in the absence of an intracranial mass. Among patients with papilledema secondary to a spinal cord lesion, 50% of cases are due to ependymomas and neurofibromas. They are often located in the thoracic and lumbar regions of the spine, leading to upward swelling and compression of the cerebellum, which in turn will impede CSF outflow through the foramen magnum [2].

Dural Venous Sinus Thrombosis (DVST)

Dural venous sinus thrombosis is a rare condition with three to four reported cases per million per year [25]. It presents most commonly in patients with hypercoagulable states due to pregnancy and the early postpartum period, as well as due to the use of oral contraceptive pills. Other causes include neoplastic conditions, intracranial infection, dehydration, or head trauma [26,27].

Dural venous sinus thrombosis, depending on size and location, may occlude one of the sinuses and reduce CSF outflow, causing buildup of ICP. Additionally, the thrombosis can lead to venous hypertension with resultant parenchymal infarcts and hemorrhage, thus contributing further to an increase in ICP [27]. Headache is the most commonly reported symptom of dural venous sinus thrombosis. Other less common symptoms include vomiting, gait ataxia, focal neurological deficits, convulsions, or altered consciousness [25,26]. In severe cases of venous thrombosis, cognitive dysfunction and seizures may also occur.

This condition is difficult to diagnose due to a varying clinical presentation. Neuro-imaging is utilized for diagnosis in the form of CT and MRI. The most sensitive diagnostic tools are CT angiography (CTA) or magnetic resonance venography (MRV) with and without gadolinium (Figure 6). These techniques will illustrate the area of absent venous flow and delineate the location of the thrombosis [28].



Figure 6: 3-D reconstruction from MRV with and without gadollium performed on a 30-year old female with headache. The dotted white arrow demonstrates a normal patent right transverse sinus. The solid white arrow denotes thrombosis of the left transverse sinus.

Vascular Causes

Increased ICP can result from a variety of vascular abnormalities in the brain including stroke, large aneurysm, hemorrhage, or arteriovenous malformation (AVM) [29]. The increase in ICP from vascular causes is due to mass effect or resulting edema. Edema is caused by congestion from the increase in cerebral volume or the decrease in cerebral blood flow. Ischemic strokes, which account for over 80% of all strokes, lead to increases in capillary permeability in the absence of an intact blood brain barrier. Consequently, edema and pressure inside the brain increases and leads to a severe increase in pressure [21] (Figure 7).

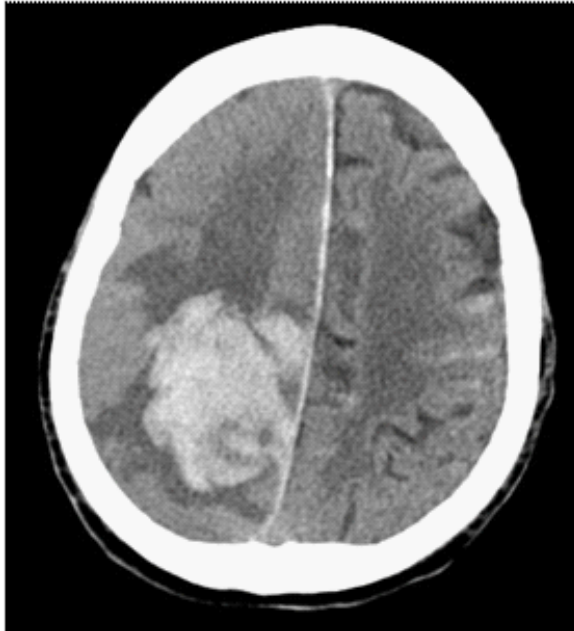


Figure 7: Axial non-contrast head CT demonstrates a parenchymal hematoma at the right frontoparietal lobe with associated vasogenic edema. Small subarachnoid hemorrhage is also seen along the contralateral cerebral hemisphere.

Diagnosis of any of the above conditions is made primarily with CT or MRI. For the diagnosis of aneurysm or AVM, angiography is utilized [24].

Infectious Causes

Infections such as an abscess, meningitis, tuberculosis, and Lyme disease have been linked to papilledema. Infectious agents may cause inflammation of the arachnoid granulations in the subarachnoid space thus impairing the proper resorption of CSF, which results in backup of fluid. Papilledema resulting from these infectious causes is rare, as low as 2.5% with most infections but up to 25% in tuberculosis associated meningitis [2]. In concurrence with neuroimaging (see figure 8), lumbar puncture is performed to evaluate the content of CSF for infection [30,31].

IDIOPATHIC INTRACRANIAL HYPERTENSION

Epidemiology

When no cause is identified, the diagnosis of idiopathic intracranial hypertension (IIH) is permissible. IIH is a neurological disorder characterized by an increase in intracranial pressure in the absence of a mass lesion or any clinical, laboratory, or radiological abnormality indicative of infection, vascular change, or hydrocephalus [32,33]. The incidence is reported as two cases per 100,000 per year, with a continued increase due to rising obesity, as it mainly affects overweight individuals. It also predominantly affects women between the ages of 15-44 years old, making it the number one non-reproductive disorder with the highest female sex predilection. The incidence of IIH in men is approximately 9% with an even lesser degree of acquisition in children [34].

Pathophysiology

Though there is no known cause for IIH, there are several presumed pathophysiologic theories under investigation. Since the majority of cases are in obese individuals, it has been hypothesized that an increase in abdominal mass raises intrathoracic and central venous pressure, thus deterring cerebral venous drainage. This reduction in flow leads to an intracranial pressure and manifestation of clinical papilledema [34-36]. Further studies have elucidated that there is a greater association between lower body adipose accumulation and the development of IIH as compared to upper body accumulation [37]. Adipose tissue acts as an endocrine organ, releasing hormones and leading to an increased level of estrogen [35], which may offer explanation as to why this condition primarily affects women of childbearing age. Another proposed mechanism is that obesity, being an inflammatory condition, contributes to a pro-thrombotic state and increased cytokine expression as demonstrated by CSF laboratory analysis [34]. This leads to the theory that microthrombi develop within the cerebral veins, either from inflammatory conditions or thrombophilia, and are responsible for interfering with arachnoid granulations and impairing CSF absorption [37,38].

Recent studies using magnetic resonance venography (MRV) revealed that 90% of patients with diagnosis of IIH also displayed bilateral transverse venous sinus stenosis. However, it is unclear whether the stenosis is caused by the increase in pressure or if the pressure is causing the compression of the sinuses, producing outflow obstruction and further worsening venous hypertension [36,37]. Few studies have even exhibited that serial lumbar punctures can relieve stenosis and that venous sinus stenting procedures have been effective in such cases [31]. This not only may contribute to possible pathogenesis of IIH, but also highlights the importance of MRV in the evaluation of papilledema.

Aquaporins are membrane proteins responsible for the bidirectional transport of water through an osmotic gradient and maintaining water balance [39,40]. When these dynamics are altered,

parenchymal edema develops and may elicit an increase in intracranial pressure. Aquaporin 1 (AQP1) is located in the apical membrane of the choroid plexus and normally secretes water into the subarachnoid space. AQP1 has been thought to play a role in drug-induced IIH, as medications, such as steroids, induce its expression and cause edema. AQP1 is also responsible for weight gain [35]. Aquaporin 4 (AQP4) is located in the plasma membrane of astrocytes and is also responsible for osmosis of water and cerebral edema [39]. While it was previously found that AQP4 does not play a causative role in the development of IIH [40], more recent studies demonstrate that a decrease in AQP4 levels in the CSF of patients with IIH, increasing susceptibility to vasogenic edema [41].

Symptoms and Mechanisms

The most commonly reported symptom in IIH is headache, present in over 90% of patients, which is the most significant symptom interfering with quality of life in these patients [42]. These headaches are daily, holocranial, and often worse upon awakening. Often, headaches are worsened with postural change and valsalva maneuvers [11,33,43]. They are associated with neck pain, nausea and vomiting [37]. The cause of headaches is due to the diffuse compression of the cranial nerves and intracranial vasculature in the setting of an elevated ICP, manifesting symptomatically as cephalgia, a feeling of fullness in the head [35]. Allodynia in the cranial nerve V1 distribution accompanies headaches in half of patients [34]. The International Headache Classification states that headaches secondary to IIH should only be diagnosed if there is a clear temporal relationship with the increase in ICP, which disappear by way of measures to normalize ICP [44]. They typically improve within the first month of diagnosis, though some cases persist past one year [45].

Transient visual obscurations are the second most frequently reported symptom, described as a momentary “gray out” or “fogginess” of vision and usually lasting seconds. They may either be unilateral or bilateral, and have also been shown to be exacerbated by postural change [36]. It is thought that these phenomena are due to transient ischemia to the optic nerve from pressure on its microcirculation [33,36,46].

Pulsatile tinnitus, described as a swishing heartbeat sound out of one or both ears [33], is present in approximately 65% of patients with IIH [47]. This is most likely a result of the transmission of CSF pulsations from compression and narrowing of the dural venous sinuses and subsequent flow turbulence [35,36,48]. The disruptions in laminar blood flow and resulting turbulence are audible [47]. While this is not the most common symptom, it is the most highly suggestive of IIH, with IIH being the leading cause of pulsatile tinnitus [48]. Studies have also suggested the possibility of dural venous sinus wall anomalies as a cause of pulsatile tinnitus in patients with IIH [49].

Diplopia is not an uncommon symptom in IIH (approximately 30% of cases), which is typically horizontal in nature and resulting from a unilateral or bilateral abducens nerve palsy [11,33].

Elevation in ICP may cause a downward displacement of the brainstem, stretching the abducens nerve or compressing it against the clivus [50]. Rarely, skew deviations, fourth, and third nerve palsies have also been reported [11].

IIH Without Papilledema

It is important to note that some cases of IIH occur in the absence of papilledema but in the presence of chronic headaches. Patients who have had poorly controlled and chronically raised ICP will sometimes present with CSF leaks, typically CSF rhinorrhea, and years later [51]. The correlation between spontaneous CSF leaks in patients with IIH is clearly documented in the literature in recent years, and is established as a variant of IIH [52]. IIH is becoming increasingly recognized as a cause of spontaneous CSF rhinorrhea, with studies documenting up to 72% of patients also meeting diagnostic criteria for IIH [53]. The initial relationship was investigated due to the strong overlap in the demographics of both IIH patients and those with spontaneous CSF rhinorrhea, namely obese females [12,52,54]. Persistent pressure from raised ICP may eventually lead to erosions and remodeling of the skull base, which leads to areas of structural weakness that allow for CSF to pass through [51,53]. The most common area of skull weakness causing CSF leak is within the ethmoid bone and lateral wall of the sphenoid bone [53]. Enlargement of the foramen magnum and development of the skull base with arachnoid pits are also present in these patients [54]. CSF is diverted because of these leaks, allowing the elevated ICP to decompress and avoiding the manifestation of papilledema [51,54]. However signs of IIH remain apparent on neuroimaging, such as a partially empty sella turcica [53]. Often, the repair of CSF rhinorrhea through endoscopic procedures will raise ICP and elicit the signs and symptoms of IIH, especially papilledema [53]. These findings suggest that patients with CSF rhinorrhea should be screened for papilledema weeks following repair to prevent vision loss, and that patients who do have IIH should also be screened for the development of CSF rhinorrhea [53].

Patients with IIH can also develop spontaneous meningoceles and encephaloceles, protrusions of meninges and brain parenchyma, respectively, through bony defects. This association is statistically significant and can be seen in 11-50% of these patients [55]. This process is likely related to the remodeling and erosion of the skull base in IIH. Once formed, there may be herniation of intracranial contents through these osseous defects. The most common location of spontaneous meningoceles is the floor of the middle cranial fossa, which is made up of the temporal bone and sphenoid bone. It is possible that the meningoceles also act to partially normalize ICP by either increasing the volume of the subarachnoid space or through small leaks from microruptures in the meninges. Additional imaging in patients with IIH should be considered due to this correlation [55].

Diagnosis

The current clinical diagnosis of IIH is made through the Modified Dandy's criteria, which requires five conditions to be met. These conditions include the following: (1) the patient

demonstrates symptoms of increased intracranial pressure or papilledema, (2) the patient demonstrates signs of increased intracranial pressure or papilledema, (3) there are no localizing findings on neurological examination and neuroimaging demonstrates no evidence of hydrocephalus, mass, structural, or vascular lesion on MRI or CT, (4) there is an elevated opening pressure (>25 cm H₂O) on lumbar puncture with normal composition, (5) and that the patient is awake and alert [54]. To make the diagnosis of IIH without papilledema, the rest of the diagnostic criteria must be satisfied, with the additional finding of a unilateral or bilateral abducens nerve palsy. If not present, the diagnosis of IIH without papilledema may only be suggesting through findings on radiological imaging, discussed below [56].

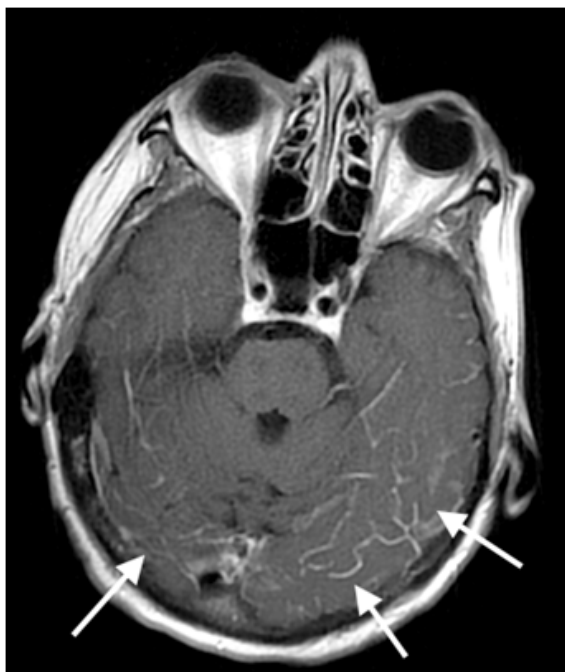


Figure 8: T1-weighted post contrast MRI of the brain in a 67-year old female with confusion. Abnormal enhancement of the leptomeninges (pia and arachnoid) was identified, as denoted by the white arrows. These findings were compatible with the diagnosis of meningitis and was confirmed histologically.

The most common neuroimaging finding in IIH is an empty or partially empty sella turcica (Figure 9), present in 65%-80% of cases and best seen on mid-sagittal MRI. The increase in ICP flattens the pituitary gland, allowing CSF to fill in and give the appearance of an empty sella [56]. When this sign is present in IIH, it often indicates chronic disease [57]. Complete resolution of empty sella appearance has been noted following treatment of IIH. However, this finding is not specific to increased ICP secondary to IIH, but is also present in patients with other causes such as intracranial space occupying lesions, as well as the normal population [56]. Slit-like ventricles are also a sign of IIH, though rarely seen and making it of poor clinical importance [57].

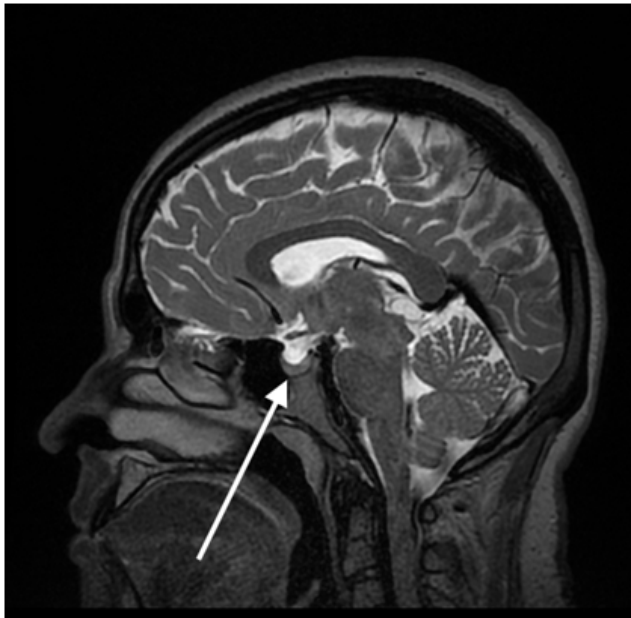


Figure 9: Sagittal T2-weighted MRI in a 37-year old female patient with IIH. There is loss of pituitary volume and the normal upwards convexity of the gland with an increased volume of CSF within the sella turcica (white arrow). These findings represent the “empty sella” sign.

Narrowing of the transverse venous sinuses are also seen much less commonly in patients with IIH from the increase in ICP and compression of the sinuses. This is best viewed using MRV techniques, but may also be present on sagittal or axial MRI [57]. While MRV was once thought to rule out the presence of venous sinus thrombosis in elevated ICP, it is now utilized to evaluate the presence of venous sinus stenosis, a potential cause or complication of elevated ICP. It has been proposed that bilateral transverse sinus stenosis, or the stenosis of a dominant transverse venous sinus, leads to decreased outflow with subsequent venous hypertension, thus impeding CSF resorption and further elevating ICP. This elevated ICP then further compresses the venous sinuses and continues the cycle of events [56]. This MRV finding, while present in about 90% of patients with IIH, is not specific to IIH and is also found in other conditions [56].

Flattening of the posterior globe on MRI and, in more severe cases, protrusion of the optic nerve into the globe is indicative of papilledema and often found in patients with IIH [56]. This occurs because of the transmission of elevated ICP through the subarachnoid space and optic nerve sheath, overcoming the intraocular pressure and pushing against the posterior sclera [57] (Figure 10). This finding is most commonly seen in IIH, though it is rarely seen in ocular hypotony [56]. Other orbital findings include dilation of the optic nerve sheaths and tortuosity of the nerve. Optic nerve sheath distention is a manifestation of the elevated ICP in the perioptic subarachnoid space, and also results in the optic nerve's tortuous appearance between the two fixed areas of its course, which is the insertion at the sclera and the orbital apex [56].



Figure 10: Axial T2-weighted MRI of the brain in a 21-year old female with IIH. This is bilateral dilation of the optic nerve sheaths (white arrow), flattening of the posterior sclera, and papilledema (black arrows).

Treatment and Management

Treatment is targeted at lowering intracranial pressure. Weight loss is an effective first line treatment to this end. Studies report a significant decrease in CSF opening pressure in patients who lost 15% of their body weight and even a marked decrease in patients with only 3.5% reduction in weight when compared to no weight loss [36]. Drug treatment is most effective with acetazolamide, a carbonic anhydrase inhibitor, which inhibits the movement of water across the choroid plexus and decreased secretion of CSF [36,34]. Topiramate and furosemide may also be used, as they appear to work through similar mechanisms [11].

If these treatment modalities fail, more invasive approaches are considered. Lumbar punctures, although typically diagnostic, may be used as treatment to remove fluid and relieve pressure [58]. In patients positive for venous sinus stenosis on MRV, stenting procedures may be utilized to equalize the pressure gradient over the stenosed sinus [11]. Diversion of CSF is made possible through shunting procedures, mainly ventriculoperitoneal and lumboperitoneal shunts [36]. Optic nerve sheath fenestration is another treatment option, which allows excess CSF to leak into periorbital fat with subsequent absorption through the veins in the fat [11]. Finally, bariatric surgery is effective in achieving long term weight loss and relieving signs and symptoms of IIH [36].

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