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SURGERY

Updates on Recent Developments in Idiopathic Intracranial Hypertension

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Abstract

Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri syndrome (PTCS) is a condition in which the intracranial pressure is increased without radiographic or laboratory evidence of intracranial pathology or localizing neurological signs, with the exception of papilledema and/or a cranial nerve VI palsy. As the pathophysiology of IIH is not well understood, current treatment strategies center on managing symptoms and preventing morbidity. This review aims to provide an updated overview on typical and atypical presentations, diagnostic criteria, work up, treatment options, and current theories on the pathophysiology of IIH.

Keywords Idiopathic intracranial hypertension · Pseudotumor cerebri · Papilledema · Headache · Obesity

Introduction

Primary idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is defined as elevated intracranial pressure (ICP) in the absence of identifiable intracranial pathology and etiology. In comparison, secondary IIH encompasses known secondary causes. Herein, only primary IIH shall be discussed. Although the precise pathophysiology remains unknown, researchers and clinicians continue to evolve methods to diagnose and treat IIH. Current strategies center on reducing risk factors, managing symptoms, and preventing morbidity, such as chronic headache and blindness. This review aims to provide an updated overview of the spectrum of presentation, diagnostic criteria, work up, treatment options, and current theories on the pathophysiology of primary IIH.

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Diagnostic Criteria

As of 2021, the diagnostic criteria most commonly reflected in the literature are those established by Friedman et al. (Table 1) [1].

Clinical Presentation

A significant challenge to diagnosing IIH is the variability in clinical presentation. The most common and disabling presenting symptom is a non-specific headache, reported in over 84% of patients [2–5]. Stereotypical headaches associated with increased intracranial pressure are positional headaches that are worse upon wakening or during Valsalva maneuvers. Recent surveys by the International Headache Society now type IIH-related headaches as episodic migraines, chronic migraines, or tension-type headaches [4].

Additional symptoms of IIH include pulse synchronous tinnitus, horizontal diplopia, neck and back pain, vertigo, nausea, and vomiting. Patients with papilledema may also experience bilateral transient visual obscurations (TVO) lasting seconds [1, 5–8].

Of note, some patients with IIH are asymptomatic and diagnosed after optic disc edema was incidentally found on optometric exam [9, 10]. Although this has led to suspicions of an underdiagnosed population of asymptomatic obese patients, studies in bariatric populations found that only 0.4–

Table 1 2013 Friedman diagnostic criteria for pseudotumor cerebri syndrome [1] A diagnosis of IIH is considered definite if A-E are satisfied and probable if A-D are met

1. Required for diagnosis of pseudotumor cerebri syndrome
A. Papilledema
B. Normal neurologic examination except for unilateral or bilateral cranial nerve VI palsies
C. Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass or structural lesion, and no abnormal meningeal enhancements on MRI, with and without gadolinium, and magnetic resonance venography; if MRI is unavailable or contraindicated, contrast-enhanced CT and CT Venogram may be used
D. Normal CSF composition
E. Elevated lumbar puncture opening pressure (>25 cm H2O in adults and >28 cm H2O in children [25 cm CSF if the child is not sedated and not obese]) when performed in the lateral decubitus position
2. Diagnosis of pseudotumor cerebri syndrome without papilledema
In the absence of papilledema, a diagnosis of pseudotumor cerebri syndrome can be made if B-E from above are satisfied, and in addition the patient has a unilateral or bilateral abducens (CN VI) nerve palsy
In the absence of papilledema or sixth nerve palsy, a diagnosis of pseudotumor cerebri syndrome can be suggested but not made if B-E from above are satisfied, and in addition at least 3 of the following neuroimaging criteria are satisfied: (i) empty sella, (ii) flattening of the posterior aspect of the globe, (iii) distention of the perioptic subarachnoid space with or without a tortuous optic nerve, and (iv) transverse sinus stenosis

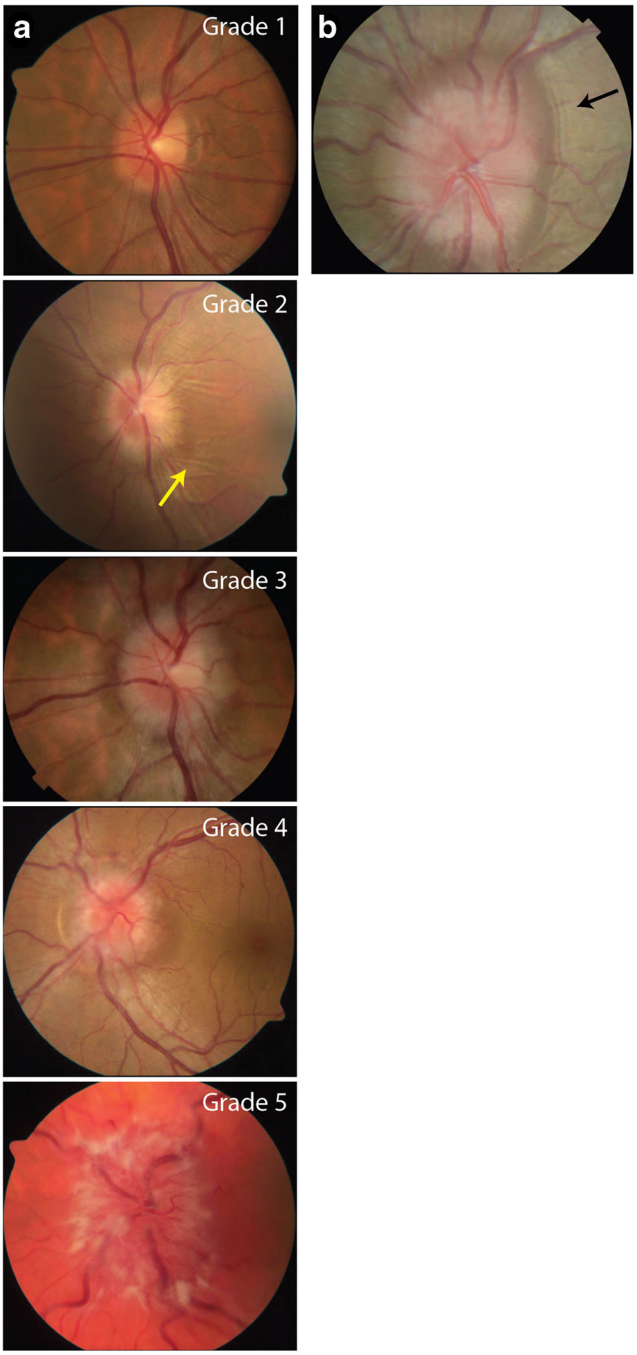

Fig. 1 Fundoscopic examples of the Frisén papilledema grading scale. **a** Grade 1, optic disc margin blurring with temporal sparing; grade 2, circumferential optic disc blurring with radial choroidal folds (yellow arrow); grade 3, obscuration of one or more minor retinal vessels at disc margin with fringed circumpapillary halos; grade 4, obliteration of the major blood vessels at the disc margin; grade 5, predominantly anterior expansion of nerve head with obliteration of optic cup. **b** Paton's folds, fringed circumpapillary retinal folds (black arrow)

0.5% of patients had asymptomatic mild optic disc edema resulting in a diagnosis of IIH [11, 12].

Common clinical signs of IIH include papilledema, visual field defects, and ophthalmoplegia secondary to CN VI palsy. [6, 7] Papilledema or swelling of the optic nerve head secondary to increased ICP results from direct pressure on the nerve at the base of the globe from the CSF and can be visualized directly with fundus ophthalmoscopy. In 1982, Lars Frisén proposed the staging system we currently use for grading papilledema (Fig. 1) [13]. More recently, practitioners use optical coherence tomography [14], a non-invasive retinal imaging technique, to measure the thickness of the retinal nerve fiber layer (RNFL) at the optic nerve head [15]. While OCT should not supplant Frisén grading during the funduscopic exam, OCT is useful for tracking changes during treatment (Fig. 2) [16]. Indeed, studies such as the IIHTT have shown a strong correlation between OCT measurements and Frisén grade [17].

Under the Frisén scale (Fig. 1a), stage 0 signifies a normal optic disc. Stage 1 represents blurring of the optic disc or development of a subtle gray halo with temporal margin sparing. Stage 2 involves circumferential blurring of the optic disc margin. Radial choroidal folds that emanate from the optic nerve, produced by posterior CSF pressure, may also become apparent at stage 2 (Fig. 1a). Stage 3 papilledema describes more pronounced swelling that obscures major retinal blood vessels at the disc margin. Fringed circumpapillary retinal folds, known as Patton's folds, may also appear (Fig. 1b). Swelling of the nerve head is even more pronounced in stage 4, resulting in obscuration of the major blood vessels that

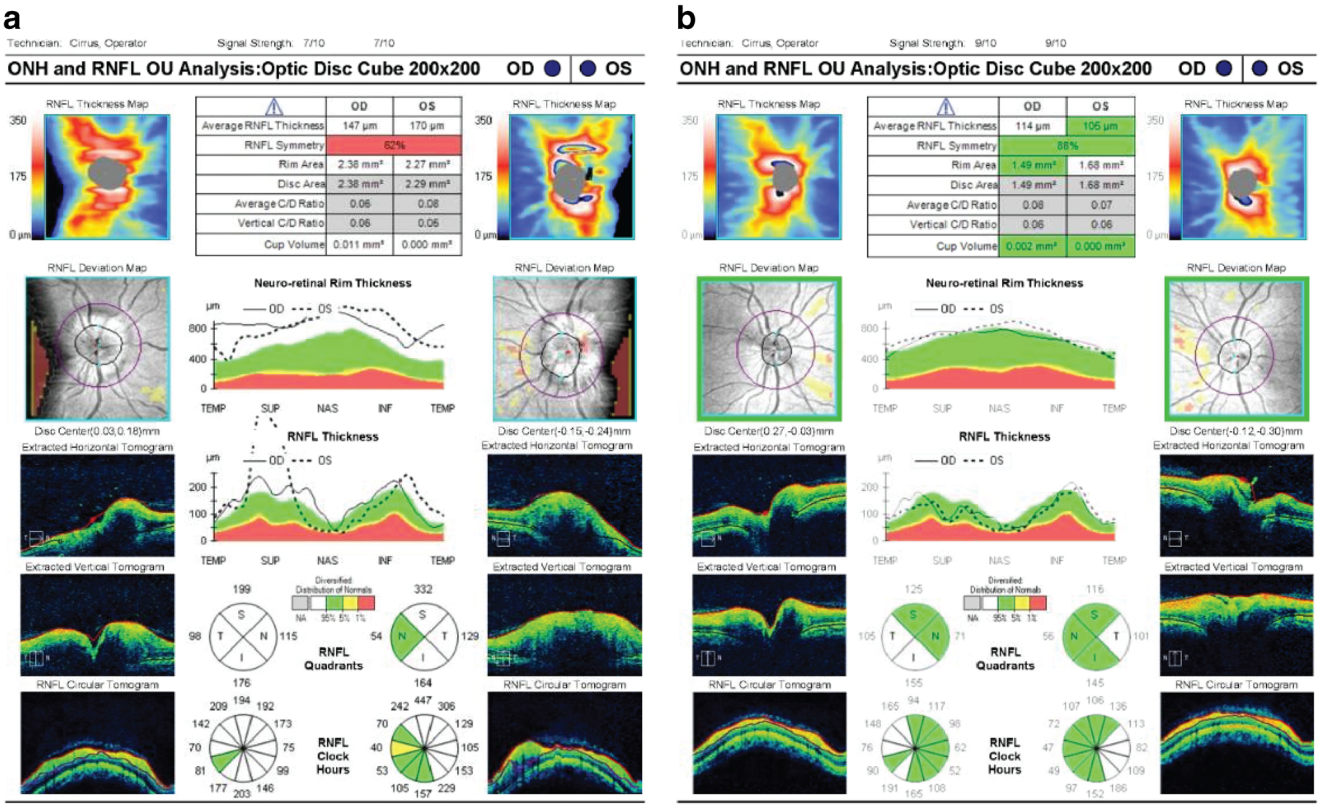

Fig. 2 Improvement in papilledema detected by spectral domain-ocular coherence tomography. An example of a patient with IIH who demonstrated improvement in retinal nerve fiber layer hyperemia as detected by spectral domain-ocular coherence tomography after treatment with oral acetazolamide

transverse through the disc. In stage 5, the most severe stage, the nerve head and optic cup are completely obliterated from swelling. The optic nerve head may also be surrounded by a thin, well-demarcated halo [13]. The aforementioned radial choroidal folds and fringed circumpapillary halos also known as Patton's folds represent signs of acquired disc swelling (i.e., not found in pseudopapilledema). Nerve fiber layer hemorrhages and venous dilation and tortuosity can also occur with papilledema and are a reliable sign of active elevated ICP, but are not part of the Frisén grading scale [13]. Funduscopic examination should also include attempts to identify spontaneous venous pulsations (SVPs), best seen at the optic nerve margin. As SVPs are estimated to exist in 87.6% of the general adult population, their isolated absence does not equate to elevated ICP [18]. Rather the disappearance of previously documented SVPs suggests elevated ICP. Finally, optic nerve head shunt vessels can develop from the chronic papilledema and their presence indicates active ICP elevation.

The swelling of the RNFL in papilledema impedes light transmission to underlying photoreceptors. This can be detected as an enlarged blind spot on formal visual field testing in early stages (Fig. 3). As the grade of optic nerve swelling continues to worsen, visual field defects progress to arcuate scotomas or loss of peripheral vision. Of note, loss of central visual acuity occurs as a late, end-stage consequence of severe papilledema, and thus, normal visual acuity alone should never be considered to be a reassuring sign without formal visual field testing.

Demographic at Risk

The typical IIH patient is an overweight young adult female who has undergone recent rapid weight gain [19]. The annual incidence of IIH in the USA has been reported to be 0.9–1.15 per 100,000 with an 8:1 female:male ratio and mean age at diagnosis of 27.8 years [20–22]. When specifically evaluating females age 15 to 44, Mayo Clinic reported an incidence of 3.3 per 100,000 per year, which increased to 7.9 per 100,000 when only those with a body mass index >26 were included. Incidences as high as 14.85 per 100,000 were reported in Iowa for women between 20 to 44 years of age. Given this observed link between obesity and IIH, it is unsurprising that one study found a correlation between the increasing incidences of IIH with the rise of obesity [23]. In terms of income and race, the highest incidence of IIH was reported in low income (1.56 compared to 1.21 per 100,000 of middle/high-income) and Black individuals (2.05 compared to next highest incidence of 1.04 per 100,000 in Whites) in the USA [24]. Although the incidence of IIH in males is lower than their female

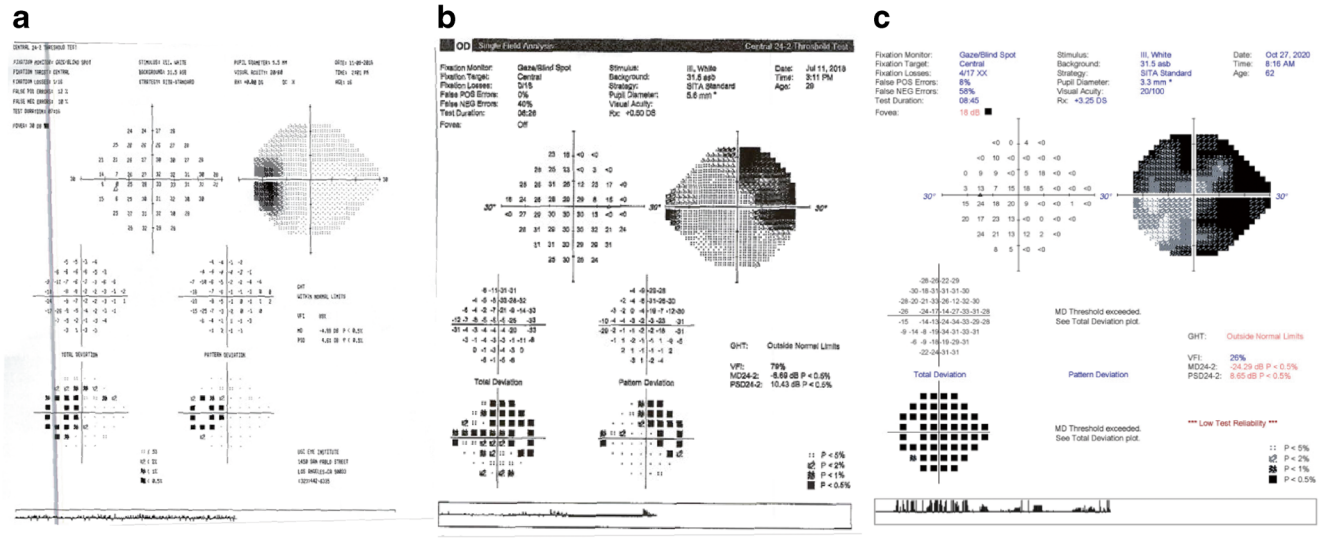

Fig. 3 Visual field changes seen in IIH. Examples of different peripheral field changes seen in patients with IIH as detected by Humphery perimetry 24–2. **a** Enlarged blind spot. **b** Superior arcuate scotoma with early nasal step. **c** Severe peripheral field constriction

counterparts, men with IIH are more likely to have transient visual obscurations and develop severe vision loss, but less likely to report headache [25].

Work Up

When a patient presents with signs and symptoms suggestive of increased ICP pressure, their physician should perform a comprehensive and appropriate workup for underlying causes. The initial exam should assess visual function, including visual acuity, intraocular pressure, pupil examination, and formal visual field testing. A fundoscopic examination is necessary for grading the severity of papilledema on the Frisén scale. If papilledema is present, the patient's blood pressure should be measured to rule out malignant hypertension (diastolic pressure greater than or equal to 120 mmHg or systolic pressure greater than or equal to 180 mmHg). The presence of hyperemia, hemorrhage, or obscuration of vessels on the optic nerve head should be documented with fundus photography. Documentation and monitoring of retinal nerve fiber layer hyperemia can also be performed with ocular coherence tomography [14, 26].

The neurologic workup should include examination of the cranial nerves. With the exception of a CN VI palsy, and rarely a CNVII palsy, [27] involvement of other cranial nerves suggests an alternative explanation [26]. Neuroimaging studies are an essential component of the work up and MRI is the gold standard for identifying secondary causes of intracranial hypertension such as hydrocephalus, tumor, meningitis, or mass-occupying lesion, which would exclude a diagnosis of IIH [26, 28]. In patients with IIH, an empty or enlarged sella and a flattened posterior optic globe may be seen [29]. CT or MR venography should always be used to exclude cerebral venous sinus thrombosis [30], which can be a secondary cause of elevated ICP even when minute [30, 31]. Distinguishing between the two is important as therapeutic management and prognosis of CVT differs greatly from IIH [30]. Although CVT is rare, unilateral or bilateral transverse sinus stenosis is a common finding in IIH patients on venography (>90%) [32].

Lumbar puncture (LP) is an essential tool in the work up of IIH. LPs should be performed with the patient in the lateral decubitus position with legs and neck extended [28] as incorrect positioning may produce false pressure readings. The lumbar opening pressure should be measured and recorded along with CSF contents. If there is high clinical suspicion for IIH but the opening pressure is normal, repeat lumbar punctures can be performed as CSF pressure can fluctuate [6, 31]. ICP is elevated if the opening pressure is ≥25 cm H_2O in adults and ≥ 28 cm H_2O in children although the pressure at which point patients become symptomatic may vary. Incorrect patient positioning, use of sedation, and improper technique will affect the validity of the measurement. As such, lumbar opening pressures should be interpreted with caution to prevent over diagnosis and unnecessary testing [33]. IIH should only be considered when the CSF contents are normal; otherwise, investigation into secondary causes should be initiated. Abnormal CSF composition such as pleocytosis could suggest an underlying infection/meningitis, or autoimmune disorder such as systemic lupus erythematosus or neurosarcoidosis.

In the event of a negative work up, causes of secondary PTCS should be elicited including recent use of certain antibiotics such as tetracycline and minocycline, excess vitamin A and retinoids, anemia, respiratory dysfunction such as sleep apnea, abrupt corticosteroid withdrawal following prolonged administration, chromosomal disorders, metabolic disorders, renal failure, anabolic steroid use, growth hormones, lithium,

nalidixic acid, chlordecone, hormone replacement medications, and Norplant(r), a levonorgestrel implant system [34–36]. Presence of these factors raises suspicion for secondary PTCS.

Differential Diagnosis for Optic Disc Edema

The first step in the workup of optic disc edema is to distinguish between true disc swelling and pseudopapilledema. Pseudopapilledema is characterized by a full appearing optic disc in the absence of elevated ICP. Upon close examination, pseudopapilledema lacks retinal nerve fiber layer hyperemia, venous dilation and tortuosity, Paton's and choroidal folds, and disc hemorrhages—clinical signs of true papilledema. Common causes of pseudopapilledema include congenitally crowded discs, seen more often in hyperopes, tilted discs, more commonly found in myopes, optic disc drusen, myelinated nerve fiber layer, and glial tufts. Differentiating between true and pseudopapilledema can be facilitated by spectral-domain OCT: Lee et al. found that the thickness of the nasal quadrant of the optic nerve disc was consistently lower than 78um in patients with optic disc drusen compared to patients with optic disc edema [37]. Optic disc autofluorescence imaging and B-scan ultrasonography can be used to facilitate detection of buried optic disc drusen. Although the exact nature of the relationship is still under active investigation, the presence of spontaneous venous pulsations (SVPs) in the setting of normal intraocular pressures is another reliable indication that the ICP is within normal limits (<18–19 cm H2O) [18]. Upon establishing that the disc swelling is real and acquired, disc edema from optic neuropathies including malignant hypertension, optic neuritis, infiltrative optic neuropathy, and ischemic optic neuropathy should be excluded. Subtle disc swelling can also be seen in toxic, metabolic, and inherited optic neuropathies.

Pathophysiology

In healthy individuals, approximately 500 mL of CSF is produced daily, primarily by the choroid plexus lining the lateral, third, and fourth ventricles, yet the volume of CSF within the subarachnoid space [38] and ventricular system ranges between 135 and 150 mL in adults. Thus, the rate of CSF production and reabsorption must be tightly balanced and regulated to maintain an appropriate ICP. Although the exact pathophysiology of IIH continues to be debated, the clear association with obesity and rapid weight gain (5–15% within the year prior) has led many to suspect neuro-metabolic dysregulation as an underlying cause of deranged CSF dynamics [19]. Ko et al. observed that patients without recurrence of IIH had stable BMI, compared to patients with recurrence (average 6% weight gain) [39]. Reinforcing this phenomenon, there were findings from Sinclair et al. showing an average loss of 15% body weight resulted in remission when weight loss was maintained [40]. Some possible mediators of this effect include glucose-like peptide 1 (GLP-1), androgens, and 11B-hydroxysteroid dehydrogenase 1 (11B-HSD1).

GLP-1 is an incretin secreted by the distal small intestines upon ingestion of food [41]. Although GLP-1 receptors (GLP-1R) have been long known to be expressed in the kidney and to play a role in volume homeostasis, recent studies found a subset of these receptors to also be expressed in the choroid plexus. In fact, extendin-4, a GLP-1 agonist originally designed to treat diabetes, was discovered to also signal through GLP-1R in the choroid plexus, leading to a 65% reduction in ICP in rodent studies. GLP-1R activation decreases Na+/K+ ATPase activity in the choroid plexus needed for CSF production [42]. However, studies examining the relationship between gut peptide and IIH are limited and their role in IIH pathogenesis is unclear. Regardless, GLP-1 agonists may represent a potential treatment for ICP management in IIH.

Hormonal dysregulation is a common feature of patients with IIH and is also believed to play a role in disease development as androgen receptors are expressed in the choroid plexus [43]. O'Reilly et al. found that women with IIH have significantly elevated levels of both precursor androstenedione and active androgen testosterone in their CSF compared to PCOS, and simple obesity controls and rodent models show that testosterone increases CSF secretion. The androgen excess theory, however, is unable to account for the gender disparity in the IIH population. Some support can be drawn from the overlapping metabolic consequences of androgen excess in obese women and deficiency in men [44, 45]. Given the prevalence of obesity and androgen excess in IIH patients, it is not surprising that women with IIH have 2-fold increased risk for developing cardiovascular disease (heart failure, ischemic heart disease, and stroke/transient ischemic attack) compared to age and BMI matched controls [46].

Given the high prevalence of obesity in IIH patients, other groups have suggested that increased truncal fat leading to increased pressure on the inferior vena cava, transferring resistance to venous outflow via the internal jugular veins, a major drainage pathway of the venous sinuses, as a cause of IIH [47]. Alperin et al. found that patients with IIH had larger extraventricular CSF volume and significantly smaller total venous outflow through their jugular veins compared to healthy controls [47]. In fact, in IIH patients, truncal fat mass, not BMI, was found to correlate with LP opening pressure [48] and truncal weight loss in patients with IIH was significantly correlated with a reduction in disease activity [49]. Given the higher prevalence of truncal obesity in men compared to women, however, this theory also fails to explain the gender disparity in IIH [50].

Another possible mechanism linking obesity to IIH proposes that increased adiposity leads to remodeling of the arachnoid granulations, impairing CSF absorption. Adipose tissue increases production of sex hormones, including estrogen [51]. Animal studies have shown that estradiol and progesterone reduce choroid plexus uptake and ion transport [52]. From this, some propose that female sex hormones, particularly estrogen, induce remodeling of arachnoid granulations and subsequent development of IIH.

Treatment Options

Treatment of IIH requires a multidisciplinary team of physicians, including a neurologist, ophthalmologist, neurosurgeon, and the primary care physician. The necessity and type of treatment will vary depending on the severity of symptoms and the availability of surgical interventions at a given medical institution. The mainstay of IIH management begins with methods for weight loss, including diet, exercise, and bariatric surgery in some cases. Medications are often used in conjunction to reduce ICP and treat headaches. The majority (93%) of patients do well with conservative treatment, [26] with surgical intervention reserved for cases with rapidly progressing visual loss or intractable headaches. Surgical options include shunting, venous stenting, and optic nerve sheath decompression.

Weight Loss

Given that 79–88% patients with IIH are obese (BMI > 30), weight loss is the mainstay of treatment in patients with primary IIH [22, 53]. Studies have demonstrated that a weight loss of approximately 6–10% of body weight or > 3.5% of total body mass index is strongly correlated with reduction in ICP and IIH symptoms [54, 55]. Weight loss interventions are more effective when guided by a licensed dietician or nutritionist and incorporate modifications to both diet and exercise. In some patients, conservative weight management efforts will fail, particularly long-term. Bariatric surgery can be considered in the morbidly obese and has been shown to produce sustained weight loss subsequently leading to a reduction in IIH-associated intracranial pressure, headache, and papilledema [56–58].

Medical Management

Acetazolamide, a carbonic anhydrase inhibitor, is often prescribed in conjunction with weight management to treat IIH. The physiological role of the carbonic anhydrase enzyme is to convert water and carbon dioxide to hydrogen ions and bicarbonate, thus exerting an effect on the rate of ion transportation and associated water diffusion. When carbonic anhydrase is inhibited, this reduces ion transportation and changes the electrochemical gradient. Subsequently, less water crosses the epithelium of the choroid plexus via facilitated diffusion, effectively decreasing the rate of CSF secretion [59]. The NORDIC (Neuro-Ophthalmology Research Disease Investigator Consortium) Idiopathic Intracranial Hypertension Treatment Trial was the first multi-center, randomized, double-blind study to assess the efficacy and safety of acetazolamide in conjunction with a low-sodium weight-loss diet for treating mild peripheral vision loss in IIH. Acetazolamide in conjunction with weight loss was shown to improve visual field function and papilledema in the patients who initially presented with mild visual loss, compared to weight interventions alone. Importantly, this effect was still significant even when weight loss due to acetazolamide was accounted for. While acetazolamide improved visual outcomes, there was no significant correlation with the reduction of headache or transient visual obscurations (TVOs) compared to the control group [16]. Prescriptions of acetazolamide have been shown to be safe and well-tolerated in doses up to 4 g/day for non-pregnant patients [60]. Although no teratogenic effects were reported in an observational study, caution should be exerted when prescribing acetazolamide to pregnant women as these studies had a small sample size [61].

Topiramate is another weak carbonic anhydrase inhibitor that can be used to treat IIH. Although there are no head to head comparisons of topiramate versus acetazolamide, one open-label study found topiramate to be as effective as acetazolamide for treatment of IIH through reduction in weight and CSF production [62]. Topiramate is contraindicated in pregnant patients due to the increased risk of teratogenic conditions [63].

Loop diuretics, such as furosemide (Lasix), serve as an alternative or adjunct to treating IIH when acetazolamide is contraindicated or insufficient [64, 65]. Furosemide appears to reduce intracranial pressure via diuresis and decreased CSF production secondary to carbonic anhydrase inhibition [64]. This slight difference in action between acetazolamide and furosemide indicates a potentially additive effect on ICP reduction when administered concurrently [66].

Headache Management

Headaches are a common and significant driver for reduced quality of life in IIH [67]. ICP management alone, however, has only shown to improve headache in 43% of patients [68]. Furthermore, in the IIHTT trial, there was no association found between mean lumbar opening pressure and headache [2]. This suggests that the etiology of headaches in IIH is likely multifactorial and targeted treatment should be provided to reduce morbidity and increase quality of life for patients.

The European Headache Federation has put forth recommendations for treatment of headaches in IIH [26]. For patients with headaches and persistently elevated ICP, weight

loss can dramatically improve symptoms. A small study (n = 20) found that an average weight loss of 15% body weight led to significant improvement in headache severity (HIT-6), frequency, and weekly analgesic use. These improvements were still seen 3 months after the study when weight loss was maintained [40]. In patients with suspected medication-overuse headaches (comorbidity of 37% in IIH), a period of medication withdrawal may be beneficial. [2] Finally, for patients with IIH in ocular remission but with persistent migraine headaches, acute (e.g., triptans and non-steroidal anti-inflammatory drugs) and preventive migraine treatments may be effective [28]. Prophylactic medications for headache that do not induce weight gain are preferred. Although there is only one clinical study evaluating the efficacy of topiramate for ICP reduction, many practitioners prescribe topiramate for its secondary benefits as migraine prophylaxis without weight gain. Other potential prevention options include candesartan and onabotulinum toxin A [28].

Shunt

In cases where conservative management has failed, medication intolerance or non-compliance, or the patient is at risk for rapid visual decline, surgical intervention such as CSF shunting may become necessary. Shunting treats IIH by diverting CSF flow from the ventricles (VP shunt) or lumbar spine (LP shunt) into the peritoneal cavity. Shunting is effective at improving common IIH-associated symptoms such as cranial nerve six palsies, TVOs, and vision loss from papilledema. Its ability to improve headaches, however, is variable with groups reporting between 72.8 and 80% efficacy [69, 70]. For this reason, it is only considered in patients with severe headaches that are not controlled with medications and weight loss but who find symptomatic relief with large volume lumbar punctures. Complications associated with shunting include subsequent revisions due to obstruction (as high as 86% [71, 72]), migration or, in rare cases, infection. Some patients can develop low-pressure headache as a result of excessive CSF diversion [72, 73]. Furthermore, neither procedure guarantees the improvement of all symptoms nor the duration of symptom remission.

Optic Nerve Sheath Fenestration

The optic nerve is ensheathed in meninges (dura mater, arachnoid mater, pia mater) that are continuous with the dura of the brain. Thus, the same CSF found in the subarachnoid space of the brain surrounds the optic nerve. In fact, it is through this connection that the force from elevated intracranial pressure is transmitted onto the optic nerve, compressing it behind the globe, leading to swelling of the optic nerve head. Optic nerve sheath fenestration (ONSF) is a surgical procedure in which a slit window is cut into the meninges of the optic nerve to reduce the pressure within the subarachnoid space surrounding the optic nerve, thereby diverting CSF into the retro-orbital space [74]. Others propose that ONSFs decrease pressure around the optic nerve by inducing scarring that impedes transmittance of ICP from the cranium to the optic nerve [74, 75]. ONSF is an effective surgical option for treating severe IIH when vision loss is the primary symptom. ONSF has the advantage of being able to improve papilledema, visual acuity, and other visual parameters faster than shunting procedures (Fig. 4). Its ability to reduce headache, however, has only been reported in less than 15% of cases [74, 76, 77]. In a retrospective review, Starks et al. found the retinal nerve fiber layer to decrease an average of 10.93 um after ONSF [78]. Although papilledema is usually found to occur bilaterally in IIH, bilateral ONSF may not be required as unilateral ONSF has been shown to produce improvement in both the ipsilateral and contralateral eye [77, 79]. In less than 1% of eyes, repeat ONSF or another CSF diversion procedure was required later [74]. Common complications of ONSF are usually short-lived and benign, including diplopia in about 26% of patients, anisocoria in around 10%, and less than 10% experience corneal delle formation and orbital or conjunctival abscess. Rare dreaded complications from ONSFs include optic nerve hematoma and traumatic optic neuropathy, both leading to severe permanent vision loss [74]. In one of the largest case series studying ONSF for progressive vision loss, Moreau et al. reported that 94.4% and 95.9% of 578 eyes of 331 patients had improvement or stability in vision and visual fields, respectively, after ONSF, demonstrating the safety and efficacy of ONSF [80]. When qualified surgeons are available, ONSF should be not be delayed for cases of IIH dominated by progressive peripheral and central visual decline [81].

Venous Stenting

Recently, dural venous sinus stenting [82] has emerged as a surgical option for IIH although its safety and utility remain controversial. Studies have shown a high prevalence of unilateral or bilateral sigmoid/transverse sinus stenosis that is associated with a high-pressure gradient along the vein in IIH patients [83]. Although it is unclear whether venous stenosis is the cause of elevated ICP in patients with IIH or develops as a consequence of elevated ICP, [84, 85] its high prevalence in the IIH population (>90% compared to 40% in the general population) makes venous stenting an attractive option.

Venous sinus stenting [86] is a radiologically guided procedure that places a catheter along the stenotic sinus,

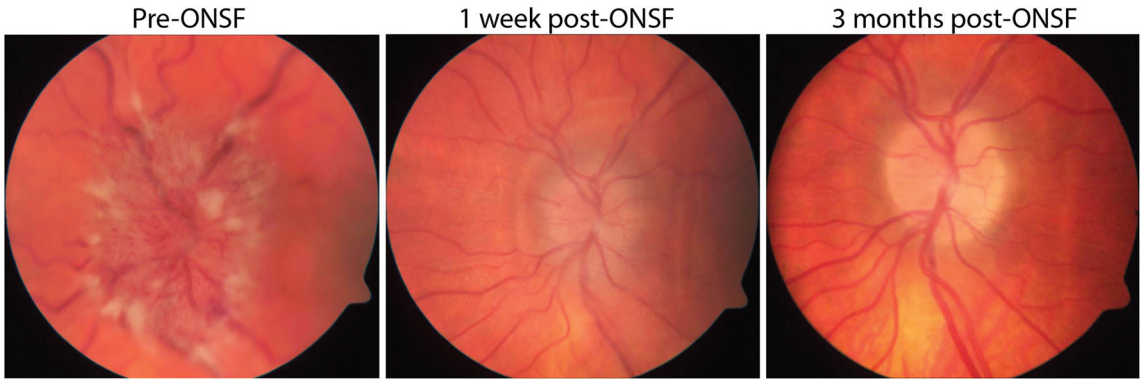

Fig. 4 Fundoscopic changes after optic nerve sheath fenestration. Example of how rapidly papilledema resolves after optic nerve sheath fenestration

decreasing the venous pressure gradient and allowing for improved venous flow and CSF absorption. A systematic review found VSS improved visual fields and visual acuity by 72.7% and 64.6%, respectively [82]. Headache improvement with VSS was comparable to CSF shunting, 72.1 to 69.8% of patients respectively. Papilledema normalized in 87.1% of VSS cases, 78.9% of shunt placements, and 90.5% of ONSF cases. Notably, treatment failure occurred in 12.3% of patients due to restenosis. While results from some studies have been promising, VSS carries rare but major complications such as subdural/intracerebral hematoma, stent migration, obstructive hydrocephalus, thrombosis, and death [82, 87]. In experienced hands, VSS may be a viable option for medically refractory IIH; however, patients should be counseled on the potential unique complications.

In summary, the goals of management are to prevent vision loss, treat diplopia, and minimize headache morbidity using the most appropriate combination and sequence of medical and surgical interventions [26]. Accordingly, patients in whom papilledema was found incidentally with no evidence of peripheral field loss should be monitored for progression or self-resolution. Patients with a BMI >30 kg/m2 and IIH should be counseled on weight management. If vision function is affected or if there is an increased risk of vision loss, medical management versus surgical intervention should be considered depending on the severity and rapidity of vision loss. CSF diversion through an LP or VP shunt is an acute surgical option that is effective for preserving and improving visual acuity when implemented in a timely fashion [88]. Another option is optic nerve sheath fenestration, which has been shown to improve or stabilize vision and visual fields in IIH patients with progressive vision loss [80]. Recently, the NORDIC group designed a multi-center clinical trial (Surgical IIH Treatment Trial) to compare the efficacy of medical plus surgical therapy (shunt vs ONSF) to treat IIH patients with moderate to severe vision loss. However, due to low enrollment, this trial is currently on hold. Neurovascular stenting is an emerging approach for cases of IIH with venography proven cerebral venous sinus abnormalities [26, 89].

Disease Progression

The majority of patients with IIH are able to recover from mild vision loss and decrease their dependence on pressure lowering medications within months of initiating treatment, especially if they are able to maintain weight loss. Despite reduction in ICP, a good portion of patients experience residual symptoms including chronic headache and pulsatile tinnitus [90]. Recurrence rates are reported to be between 8 and 40% of patients and generally correlated with recurrent weight gain [91, 92].

Pregnancy and IIH

As a condition most prevalent in women of child-bearing age, it is important to address the effect of pregnancy on IIH. While there have been case reports of visual impairment in pregnant patients with known IIH, [93, 94] controlled studies have found no difference in the frequency of visual loss between pregnant and non-pregnant IIH patients [95, 96]. IIH onset can occur in any trimester of pregnancy; however, a majority of diagnoses are made in the first half of pregnancy [95]. This is perhaps attributable to the rapid weight gain associated with gestation. Management of IIH in pregnant patients should not differ from that of non-pregnant patients aside from restriction of teratogenic medications and modification of dietary and weight loss goals so as to avoid ketosis injury to the fetus [96].

Conclusion

As we continue to advance our understanding of the pathophysiology underlying idiopathic intracranial hypertension along with technological advances, we hope to more accurately diagnose IIH, with minimally invasive mechanisms, and differentiate it from similarly presenting conditions. The Idiopathic Intracranial Hypertension Treatment Trial has demonstrated the efficacy of a conservative treatment plan consisting of weight

loss and acetazolamide for cases associated with mild vision loss. Results of large-scale studies comparing surgical approaches to conservative treatment for moderate vision loss are needed. Ultimately, treatment decisions for medically refractory cases are limited by the availability of a procedure in a given institution. A better understanding of the mechanisms underlying IIH will allow for the development of novel treatment strategies and prevention of associated morbidities, such as permanent vision loss.

Author Contributions KG conceptualized and designed the manuscript. MP and KG performed the literature review and interpretation. MP and KG drafted and reviewed the manuscript. All authors read and approved the final manuscript.

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