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Fulminant Idiopathic Intracranial Hypertension

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Abstract

Purpose of Review The goal of this review is to describe the presenting features of fulminant idiopathic intracranial hypertension (IIH) and outline the multimodal approach to its treatment.

Recent Findings Venous sinus stenting may be an appropriate alternative to optic nerve sheath fenestration or cerebrospinal fluid shunting in select patients with fulminant IIH. Prompt surgical intervention maximizes the chance of visual recovery in patients with fulminant IIH.

Summary "Fulminant IIH" is defined as intracranial hypertension with no secondary cause, severe vision loss within 4 weeks of symptom onset, and progressive vision loss over days. Rapid recognition of the fulminant phenotype of IIH by emergency department physicians, neurologists, and ophthalmologists is critical. Without appropriate triage and rapid medical and surgical intervention, patients with fulminant IIH are at high risk for profound, permanent vision loss. Prompt surgical intervention with optic nerve sheath fenestration, cerebrospinal fluid shunting, or venous sinus stenting minimizes the chance of poor visual outcome. If a delay is anticipated, serial lumbar punctures or temporary cerebrospinal fluid drainage and medical therapy may forestall irreversible vision loss.

Keywords Idiopathic intracranial hypertension · Fulminant idiopathic intracranial hypertension · Malignant intracranial hypertension · Pseudotumor cerebri

Introduction

Elevated intracranial pressure without abnormal cerebrospinal fluid (CSF) constituents or a structural cause, commonly referred to as idiopathic intracranial hypertension (IIH), predominantly affects women of childbearing age who are overweight or have recently gained weight [1]. Severe visual field loss and sub-normal visual acuity are uncommon early in the course of IIH [1]. Weight loss and medical management typically are adequate to prevent visual loss in patients with IIH [2]. However, severe and rapidly progressive vision loss develops within 1 month of symptom onset in 2–3% of patients with IIH [3••]. These patients with fulminant IIH are at high risk for permanent legal blindness if diagnosis or definitive surgical treatment is delayed by a matter of days [3••]. The goal of this review is to describe the presenting features of fulminant IIH and to familiarize the reader with its medical and surgical management.

This article is part of the Topical Collection on *Neuro-Ophthalmology*

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Diagnosis of Fulminant IIH

Idiopathic intracranial hypertension (IIH) is defined by an abnormally high intracranial pressure (ICP) without iatrogenic cause in the setting of signs and symptoms of intracranial hypertension, a normal neurological examination (excepting 6th nerve palsy and papilledema), normal cerebrospinal fluid constituents and neuroimaging which excludes ventricular deformity or structural causes of intracranial hypertension (e.g., parenchymal mass or venous sinus obstruction) [2, 4•, 5]. Exact criteria for fulfilling the above premises may slightly differ between authors [2, 5]. Accepting that an isolated reading of ICP may reflect the peak or trough of a CSF wave and that variation in positioning and Valsalva may affect the recorded ICP, it is generally agreed that an ICP of ≥ 25 cm H_2O establishes the presence of intracranial hypertension. Wall

et al. consider an ICP of 20–25 cm H_2O adequate to establish the diagnosis in the setting of at least one of the following: pulse synchronous tinnitus, abducens palsy, Frisen grade II papilledema, absence of optic disc drusen on ultrasonography, transverse venous sinus stenosis/collapse, or a partially empty sella or enlarged optic nerve sheaths on neuroimaging [2]. Friedman et al. propose additional criteria to avoid overdiagnosis in patients without papilledema. These criteria are amalgamated in a simplified manner in Table 1. It should also be noted that some authors advocate for the term idiopathic intracranial hypertension whereas others employ *pseudotumor cerebri syndrome*, which allows for the inclusion of conditions wherein intracranial hypertension is not truly idiopathic, but rather associated with medications or general medical conditions [5].

Fulminant IIH is defined by its acute onset (less than 4 weeks between symptom onset and severe vision loss) and rapid worsening of vision over days (Table 2) [3••]. Few detailed descriptions of patients with fulminant IIH have been published [3••, 6–17]. The largest and most detailed series of patients with fulminant IIH was reported by Thambisetty et al. in 2007 [3••]. Among the 483 definite cases of IIH seen at Emory University between 1996 and 2006 and the 89 cases seen at Vanderbilt University between 2003 and 2006, a total of 14 (2.9%) and 2 patients (2.2%), respectively, met the diagnostic criteria for fulminant IIH. Similar to IIH patients who present with a non-fulminant course, the majority of patients with fulminant IIH are overweight women of childbearing age [2, 3••]. Features disproportionately represented in the fulminant subpopulation of IIH patients included nausea with recurrent vomiting in 9/16 (56%), subjective subacute vision loss in 13/16 (81%), and neck stiffness in 4/16 (25%). Similar to patients with a typical IIH presentation, headache was present in all 16 patients, pulsatile tinnitus in 9/16 (56%), and transient visual obscurations in 8/16 (50%). Smaller series and case reports of patients with fulminant IIH describe similar presenting features [6, 10, 13, 15]. Fulminant IIH also has been reported in atypical patient populations for IIH, including children and men [8, 9, 16].

The diagnosis of fulminant IIH may not be apparent at the first clinical encounter, particularly if a patient presents within days to weeks after onset of symptoms referable to intracranial hypertension and if formal visual field testing is unavailable. Bhandohal described a patient who presented to the emergency department with visual acuities of 20/30 in the right eye (OD) and 20/25 in the left eye (OS), progressing to NLP OD and 20/70 OS within a day [6]. Shaikh et al. reported the case of a patient who progressed from normal vision to no-light-perception (NLP) in both eyes (OU) over the course of 4 days [15]. These reports underscore the importance of maintaining an awareness of fulminant IIH among all physicians who treat headache or vision loss and the need to counsel patients to seek immediate attention should visual acuity or visual field markedly change following the initial clinical encounter [6].

Table 1 Diagnostic criteria for idiopathic intracranial hypertension

1. Symptoms and signs of intracranial hypertension[+]
2. Normal neurological examination (excepting 6th nerve palsy and papilledema) including mental status examination
3. Neuroimaging which excludes ventricular deformity or structural causes of intracranial hypertension (e.g., parenchymal mass or venous sinus obstruction)
4. Elevated ICP (ideally lateral decubitus with legs extended and abdomen relaxed)
   *≥ 25 cm H_2O measured reliably
   *20–24 cm H_2O is adequate to establish the diagnosis in the setting of at least one of the following:
   a. Pulse synchronous tinnitus
   b. Abducens palsy
   c. Frisen Grade II papilledema
   d. Exclusion of pseudopapilledema (e.g., optic disc drusen on ultrasound)
   e. Radiographic signs of intracranial hypertension (transverse venous sinus stenosis/collapse, partially empty sella, optic nerve tortuosity, or enlarged optic nerve sheaths)
5. No exposure to iatrogenic causes of elevated ICP
6. Normal CSF constituents

[+] IIH without papilledema may be diagnosed if the above criteria are fulfilled with an opening pressure of ≥ 25 cm H_2O measured reliably and in the presence of either a 6th nerve palsy or 3 or more of the following criteria (empty sella, flattening of the posterior globes, distention of the subarachnoid space around the optic nerves, or transverse sinus stenosis)

Table 2 Diagnostic criteria for fulminant idiopathic intracranial hypertension

1. Diagnostic criteria for IIH fulfilled (Table 1) including papilledema
2. Less than 4 weeks between symptom onset and severe loss of visual acuity or field
3. Rapid worsening of vision over days

Management of Fulminant IIH

The management of fulminant IIH differs fundamentally from the management of typical IIH. In most patients with non-fulminant IIH, papilledema improves over a period of months provided compliance with weight loss and medical management [2]. Surgical intervention is required in a minority of patients who remain refractory or intolerant to weight loss and medical treatment [1]. The relative rarity of profound, permanent vision loss in patients with IIH may lead to false reassurance when a diagnosis of IIH is established and the features of fulminant IIH are not recognized [13]. Because patients with fulminant IIH are at high risk for permanent, profound visual loss if surgical intervention is delayed [3••], patients encountered in the outpatient setting whose history and examination raise suspicion for fulminant IIH should be immediately admitted to a hospital for rapid confirmation of the diagnosis and prompt medical and surgical management

(Fig. 1). A neuro-ophthalmologist should be involved in the earliest stages of diagnosis and management. In patients with progressive visual loss, close monitoring of the visual status is essential with visual acuity and visual field assessment performed on a daily basis until medical and surgical treatments detailed below are found to be successful.

Effective treatment of fulminant IIH typically requires a combination of medical and surgical intervention [3••, 14]. All patients should be started on high-dose carbonic anhydrase inhibitors (typically acetazolamide given orally or intravenously at a dose of at least 2 g and up to 4 g per day) while awaiting surgical intervention. Intravenous methylprednisolone can also be considered as another temporizing measure. Surgical intervention, either in the form of CSF shunting, optic nerve sheath fenestration (ONSF), or venous sinus stenting (VSS), should be pursued as quickly as possible. If a delay in surgical intervention (24 h or more) is expected or if vision loss has been particularly rapid, temporizing CSF drainage should be considered via lumbar drain, extraventricular drain, or serial large-volume lumbar punctures.

Surgical Management

Surgical options for treating fulminant IIH include CSF diversion procedures (ventriculoperitoneal shunting [VPS] or lumboperitoneal shunting [LPS]), ONSF, and VSS. There are no prospective studies directly comparing the efficacy and complications of these interventions in patients with fulminant IIH. The choice between CSF diversion, ONSF, and VSS should be based upon the prominence of other symptoms of intracranial hypertension (shunting and VSS being favored over ONSF for relief of headache), the presence of venous sinus stenosis permitting VSS, and institutional familiarity with and availability of these procedures. CSF diversion procedures lower intracranial pressure and are favored if headache is prominent. Optic nerve sheath fenestration may be employed if headache and other symptoms of intracranial hypertension are absent or minimal but papilledema and vision loss are severe. Regardless of the specific choice of procedure, it is generally recommended that one of these procedures is performed promptly as fulminant IIH frequently results in poor visual outcomes if definitive surgery is delayed [3••]. In the largest series of patients with fulminant IIH, all patients who were not legally blind at follow-up underwent surgery (shunting or ONSF) within 4 days (median 2 days) of evaluation; all patients who remained legally blind at follow-up experienced delays in surgical intervention following diagnosis (3–37 days, median 6.5 days) [3••]. Definitive surgical management should not necessarily be abandoned if it cannot be performed within 4 days of diagnosis, though the chance of meaningful recovery of vision decreases with time [12].

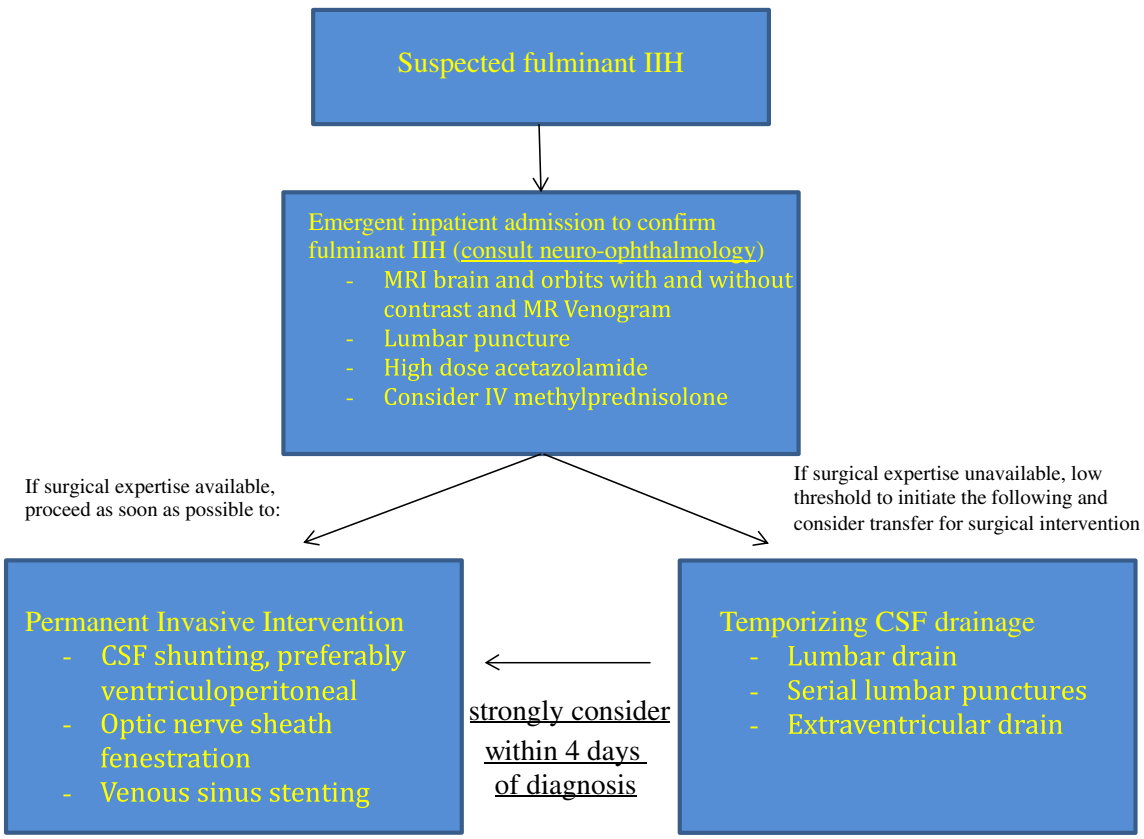

Fig. 1 Guide to assessment and treatment of fulminant idiopathic intracranial hypertension

Failure of one surgical intervention should lead to consideration of an urgent second procedure of different type [18•, 19].

A few reports exist of patients with fulminant IIH who experienced a good visual outcome with either medical therapy alone or temporary drainage while awaiting the effect of carbonic anhydrase inhibitors [8, 9, 14, 16]. However, these reports are rare, and non-surgical approaches to recovering lost vision in fulminant IIH are frequently unsuccessful [3••, 10, 17].

CSF Diversion

Ventriculoperitoneal shunting (VPS) involves diversion of CSF from the intraventricular compartment into the peritoneum via a subcutaneous shunt. Lumboperitoneal shunting (LPS) involves CSF diversion from the lumbar cistern into the peritoneum. Immediate success rates and short-term complications between the two procedures are comparable, though VPS is increasingly preferred due to lower rates of shunt revision and the ability to modify shunt settings.

The majority of patients who undergo shunting experience a near-immediate normalization of intracranial pressure resulting in reduction or elimination of headache and stabilization or improvement of vision [20]. Direct comparison of VPS and LPS in IIH patients indicates a similarly high success rate in normalization of ICP between the two procedures. One retrospective review of 25 shunted IIH patients, 8 of whom underwent shunting due to fulminant IIH, reported statistically equivalent rates of resolution of symptoms referable to elevated ICP between LPS and VPS (89% and 86%, respectively) [21•]. Of the subgroup shunted for fulminant IIH, 3/3 in the VPS group and 4/5 in the LPS group experienced immediate symptom relief without recurrence; the 5th patient in the LPS group required VPS due to malpositioning of the peritoneal catheter with subsequent resolution of symptoms. Another review of 24 IIH patients who underwent VPS or LPS reported rapid resolution of headache in 95% of patients [22]. Similar rates of success have been reported in studies employing exclusively VPS or LPS for IIH [23–26].

Though effective in normalizing intracranial pressure, shunting is only effective at recovering vision in fulminant IIH when employed promptly following diagnosis. In the series of fulminant IIH patients published by Thambisetty et al., 11 underwent shunting procedures (2 VPS and 9 LPS). Among this group, the only patients who were not legally blind at last follow-up were those who underwent shunting procedures within 4 days of diagnosis (2 VPS, 3 LPS), although intervention in 4 days or less did not necessarily insure success as 2 patients who underwent LPS 3 days after diagnosis (14 and 11 days after symptom onset) remained legally blind at follow-up.

One potential consideration in VPS is the 0.9% mortality rate reported by Curry et al. [27]. However, this figure is derived from a retrospective analysis of the Nationwide Inpatient Sample hospital discharge database, employing data collected between 1988 and 2002. Due to technical advances in shunting and the inability to review the specific circumstances of deaths in this study, the 0.9% mortality rate for VPS in the IIH population should be interpreted with caution.

Optic Nerve Sheath Fenestration

Optic nerve sheath fenestration (ONSF) entails the creation of a window in the optic nerve sheath, permitting egress of CSF from the perineural subarachnoid space and decreasing the pressure of CSF exerted on the optic nerve. There are a variety of surgical approaches described in the literature. ONSF is generally an effective and safe intervention. Recovery of visual function is more likely when ONSF is employed to treat acute rather than chronic papilledema [28]. Among the general IIH population, ONSF is frequently effective with stabilization or improvement of visual acuity in 95% of operated eyes [20]. Headache is less likely to improve following ONSF (44%) compared with shunting procedures (80%) [20]. Uncommon intraoperative complications include ischemic optic neuropathy and CRAO, which frequently cause severe, permanent vision loss [29].

Of the 16 patients included in the fulminant IIH series reported by Thambisetty et al., 5 underwent prompt ONSF and vision improved in all. However, specific caveats may apply to ONSF in patients with fulminant IIH. In one study of 40 patients with opening pressure ≥ 50 cm H_2O, 6 (15%) experienced progressive vision loss requiring a subsequent shunting procedure—a rate three times higher than in the 134 patients whose opening pressure was < 50 cm H_2O (4.5%). Progression of vision loss following ONSF requiring a shunting procedure was noted in 33.3% of patients whose presenting visual acuity was 20/200 or worse, in contrast to 2.4% for patients whose presenting acuity was 20/40 or better [18•]. Given the high proportion of patients with fulminant IIH who present with acuities of 20/200 or worse and an opening pressure of opening pressure ≥ 50 cm H_2O[3], patients who do undergo ONSF should not be discharged until it is clear that the procedure has successfully stabilized or improved vision.

In patients with fulminant IIH who do not have prominent symptoms of intracranial hypertension and who have highly asymmetric vision loss, unilateral ONSF may be considered. Although unilateral ONSF may be comparable with bilateral ONSF in patients with non-fulminant IIH, it is not clear whether that treatment strategy is applicable to patients with fulminant IIH and bilateral severe visual loss. A bilateral approach may be considered if ONSF is pursued in fulminant IIH patients with severe bilateral vision loss [30]. Surgeons performing ONSF may prefer to perform bilateral fenestration in serial procedures rather than simultaneously due to the small procedural risk of visual loss.

Venous Sinus Stenting

First described in 2003, venous sinus stenting (VSS) is a promising new treatment in select patients with fulminant IIH [31]. Given the relatively few patients with fulminant IIH who have been treated with VSS, this approach must be done cautiously with extremely close neuro-ophthalmic and neurosurgical surveillance.

Cerebrospinal fluid is reabsorbed by arachnoid granulations into the intracranial venous sinuses. Intracranial hypertension frequently causes stenosis of the venous sinuses, impairing the drainage of venous blood and resorbed CSF and thereby exacerbating intracranial hypertension and propagating further venous sinus stenosis. This vicious cycle may be interrupted by deploying one or more stents across hemodynamically significant stenotic intracranial venous sinuses to maximize venous (and thus CSF) drainage. In the setting of venous sinus stenosis where an appreciable trans-stenotic pressure gradient can be demonstrated (thus suggesting a hemodynamically significant stenosis), stenting has been shown to result in rapid normalization of intracranial pressure with improvement in afferent visual function and symptoms of intracranial hypertension [32•].

Dinkin and Patsalides reported a prospective trial of VSS in 13 patients with IIH, of whom 3 fulfilled the definition of fulminant IIH [32•]. Papilledema grade and visual field improved in all 6 eyes; no eyes developed optic atrophy. One patient was medication-free post-VSS and the acetazolamide dose was able to be reduced in a second. Elder et al. described 4 patients who underwent VSS for fulminant IIH following temporizing measures (3 with LPs, 1 with lumbar drain) while beginning antiplatelet therapy in advance of VSS [33]. All had a marked improvement in headache and a reduction in transverse sinus pressure gradient. Afferent visual function improved or stabilized in 3 patients, and the single patient whose vision worsened post-procedurally presented for VSS as a second intervention already having undergone ONSF.

Temporizing Invasive Management

If the surgical options above are not immediately available and the patient either cannot be transferred to a center where those interventions are available or refuses permanent surgical intervention, temporizing CSF diversionary measures should be considered while optimizing medical management of intracranial hypertension. These include serial lumbar punctures, lumbar drainage, and extraventricular drainage (EVD).

Serial Lumbar Punctures

Of the temporizing invasive interventions, serial lumbar puncture is likely the least morbid and has the advantage of being a procedure which can be done rapidly at the bedside by most neurologists, neurosurgeons, emergency medicine, and internal medicine physicians. The high body mass index typical to patients with fulminant IIH, however, may limit success of the procedure attempted without radiographic guidance. Though CSF can be withdrawn until the closing pressure normalizes, the beneficial effects of serial LP are short-lived due to the high rate of CSF production (approximately 15 mL/h) [8]. Depending on the interval between diagnosis of fulminant IIH and definitive surgical intervention, multiple LPs may be required [3••].

Lumbar Drainage

Lumbar drainage is an effective means of rapidly lowering ICP. CSF may be removed until the ICP is within the normal range, and subsequent drainage is typically recommended at 15 mL/h, which approximates the rate of CSF production although the rate may require individualized titration [8]. In select cases, lumbar drainage may be employed in patients with fulminant IIH as a bridge to adequate medical therapy rather than a definitive surgical intervention such as shunting, ONSF, or VSS [9]. Should this strategy be employed, the drain should be clamped after adequate medical therapy has been administered to determine whether medical therapy alone is adequate. Relapse after lumbar drain clamping despite adequate medical therapy indicates the need for shunting [7]. This strategy is appealing, but should lumbar drain placement fail, prompt definitive surgical intervention should be pursued.

Major complications of lumbar drainage include bacterial meningitis and symptomatic subdural or subarachnoid hemorrhage [34]. Prospective analysis of lumbar drain infections demonstrates a marked increase in incidence after 4 days of lumbar drainage, increasing from a cumulative incidence of ~ 5% by day 4 to ~ 10% by day 5 and ~ 20% by day 12 [35]. These data are supported by larger, retrospective analyses of lumbar drain infection [36]. In cases where lumbar drainage is employed as a bridge to adequate medical therapy, a clamping trial approximately 36–48 h following drain placement is suggested. This time frame permits acetazolamide levels to reach steady state concentration in the serum and if the clamping trial fails, definitive surgical intervention can be pursued and the lumbar drain discontinued prior to day 4 of lumbar drainage. During lumbar drainage, daily cerebrospinal fluid analysis is recommended to identify infections promptly. Patients with lumbar drains must also be monitored closely for intracranial hypotension, which may result in cranial neuropathies and posterior cerebral artery ischemia [37].

Extraventricular Drainage

In the event that serial lumbar puncture or lumbar drainage is unsuccessful, extraventricular drainage (EVD) could be considered as a bridge to ventriculoperitoneal shunting. An EVD

can be placed rapidly, in some cases while the patient is in the emergency department, and can be later converted into a VP shunt. The main complications of EVD placement are hemorrhage and meningitis, but the rate of significant hemorrhage related to placement is ~1% [38], and infectious risk is relatively low when used for a brief period as would typically be encountered in patients with fulminant IIH requiring a temporizing measure pending definitive surgical treatment [39]. Small ventricular size may increase the chances of EVD malpositioning and complication; this should be taken into account when selecting a temporizing CSF diversionary method.

Medical Management

Medical treatment is an important part of the management in fulminant IIH, but does not replace surgical intervention. Patients with fulminant idiopathic intracranial hypertension should be started on high-dose carbonic anhydrase inhibitors immediately following confirmation of the diagnosis and prior to definitive diagnosis if the index of suspicion for fulminant IIH is high. Intravenous methylprednisolone can also be considered as a temporizing measure.

Carbonic Anhydrase Inhibitors

Carbonic anhydrase inhibitors reduce the production of CSF in the choroid plexus [40]. Acetazolamide plays a central role in the care of most patients with IIH [2], and for fulminant IIH maximally tolerated doses (at least 2 g per day and up to 4 g per day) should be used. Should patients develop a clinically significant acidemia, buffering agents such as bicarbonate may be considered. The prompt initiation of high-dose acetazolamide in patients with newly diagnosed fulminant IIH is an important temporizing measure and should be continued after definitive surgical therapy to insure stabilization of vision [3••]. Intravenous acetazolamide may have a faster onset and has the additional benefit of guaranteed bioavailability in patients with emesis. Data supporting the use of topiramate are less robust, but it can replace acetazolamide in the setting of renal failure [41]. The use of topiramate for fulminant IIH is limited by the inability to initiate therapy at high doses due to the risk of Stevens Johnson syndrome.

Corticosteroids

Corticosteroids may be an effective treatment for IIH [42]. Intravenous methylprednisolone may be employed in patients with fulminant IIH awaiting surgical treatment [3••, 11]. Intravenous methylprednisolone at a dose of 250mg every 6 h is recommended with an oral or IV taper after definitive surgical intervention has normalized ICP.

Post-Acute Care of the Patient with Fulminant IIH

The immediate success of surgical procedures should not be assumed. Patients whose vision loss progresses despite VPS, ONSF, or VSS should undergo a second intervention of a different type promptly. Confirmation of surgical efficacy is critical prior to down-titration or withdrawal of medical therapy. Depending on the choice of surgical intervention, continued medical management may be required post-operatively. Shunting may obviate the need for carbonic anhydrase inhibitors. Down-titration or withdrawal of medical therapy should be done cautiously in patients who have undergone optic nerve sheath fenestration or venous sinus stenting.

Following stabilization or initial improvement in vision following surgical intervention, patients with fulminant IIH followed closely for recurrent intracranial hypertension or surgical complications. Eventual failure of the surgical interventions discussed above is common and relapses may be fulminant [43]. Weight loss remains crucial to prevent recurrent intracranial hypertension should VPS, VSS, or ONSF eventually fail. Even modest weight loss (6–10% of body weight at diagnosis) has the potential to improve or resolve IIH [44, 45]. Conversely, relapse is frequent and typically associated with an increase in weight [46•]. If weight loss cannot be achieved through lifestyle modification, surgical approaches to weight loss may be considered [47].

Shunt Failure

The risk of over-drainage, distal catheter migration, and infection is similar between VPS and LPS [22]. However, rates of shunt revision are 2–2.5 times higher in LPS than VPS, largely owing to obstruction and proximal catheter displacement from the lumbar cistern [21•, 22]. Among 18 patients who received LPS in one study, 10 (56%) required shunt revisions in contrast to 2 of 7 patients (29%) who underwent VPS [21•]. In patients who undergo either LPS or VPS and do require revision, multiple revisions are frequently required [21•, 25]. Causes of shunt revision include dislodgement at the proximal or distal end, shunt block, and shunt infection [21•, 25]. Patients with LPS or VPS who require revision typically undergo their first revision within the first 1–2 years following shunting [21•, 25], though later shunt failures have also been reported [42]. One Kaplan-Meier analysis of continued VPS function in 17 patients revealed 80% functioning at 12 months, 65% by 24 months, and 48% by 36 months [48]. Higher rates of VPS failure have also been reported, up to 60% by 24 months [26, 29]. Ventriculoatrial and ventriculopleural

shunting appear to have high rates of shunt failure compared with shunting procedures employing alternate distal shunt sites [26].

ONSF Failure

Fulminant IIH patients who undergo ONSF require particularly close outpatient follow-up. Despite high rates of initial success in stabilizing or improving vision, the likelihood of short-term failure of ONSF may be higher in patients with fulminant IIH than in the IIH population as a whole. The 3-month post-operative risk of ONSF failure correlates with ICP and inversely correlates with visual acuity at presentation [18•]. In one study, the 3-month failure rate of ONSF in patients with an opening pressure ≥ 50 cm H_2O and acuity of 20/200 or worse was 50%. While this should not prevent ONSF from being employed in the appropriate clinical setting, the particularly elevated opening pressure (mean 54.1 cm H_2O) and frequency of visual acuities of 20/200 or worse in at least one eye (11/16) reported by Thambisetty et al. should be considered prior to ONSF in patients with fulminant IIH. ONSF may still be an emergent, successful, sight-saving procedure in patients with fulminant IIH but a high index of suspicion should be maintained for ONSF failure with any post-operative worsening of optic nerve appearance or afferent visual function.

The majority of ONSF complications are apparent post-operatively and transient. However, serious and permanent complications including CRAO have been reported in up to 6% of patients, with repeat ONSF conferring a higher risk of retinal vascular occlusion [49, 50].

VSS Failure

Patients may occasionally require alternative surgical treatments after VSS. Goodwin et al. reviewed 18 IIH patients treated with VSS, 3 of whom underwent subsequent shunting [51]. Lumbar puncture opening pressure prior to VSS was higher in the group that required shunting after the stenting procedure (50 cm H_2O compared with 37 cm H_2O, $p = 0.012$). Restenosis adjacent to the stent was noted in 2 of 3 patients. However, in the series reported by Dinkin et al., no patients required further surgical treatment during the follow-up period (mean 499 days for all patients enrolled), VSS was well-tolerated in all 13 patients with no serious complications, and 7 patients were titrated off of medical therapy altogether [32•].

Conclusions

Fulminant vision loss occurs in 2–3% of patients with IIH. Patients with fulminant IIH are demographically typical to patients with non-fulminant IIH, but more frequently report emesis and neck stiffness. Rapid recognition of fulminant IIH is of critical importance as delay in definitive surgical intervention frequently results in permanent, profound visual disability. Acetazolamide should be initiated at a dose of between 2 and 4 g/day while awaiting surgical intervention, and IV methylprednisolone may be considered. Shunting, ONSF, or VSS within the first few days following diagnosis maximizes the chance of meaningful visual recovery. The rapidity with which one of these surgical interventions is employed likely is more impactful than the type of intervention chosen. If a delay in surgery is unavoidable, temporizing procedures such as serial lumbar punctures or lumbar drains should be considered. Down-titration of medical therapy following surgical intervention should be done cautiously. Post-acute care of patients with fulminant IIH involves close surveillance for failure of surgical intervention and relapse.

Compliance with Ethical Standards

Conflict of Interest The author declares that he has no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- •• Of major importance

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