Idiopathic Intracranial Hypertension (IIH)

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Abstract: Idiopathic intracranial hypertension (IIH) was previously known as pseudotumor cerebri or benign intracranial hypertension. Though rare, it is an important cause of vision loss, especially in women of child-bearing age. It presents clinical features consisting of raised intracranial pressure, including papilledema. Neuroimaging reveals no mass lesions, but a lumbar puncture shows high CSF pressure, while normal findings are revealed by CSF studies. IIH is usually managed using conservative treatment, but surgical intervention is needed for threatened vision and severe resistant headaches. In this chapter, clinical presentation, diagnostic workup, and management principles are summarized. Indications of surgical management along with surgical treatments including optic nerve sheath fenestration, CSF diversion (ventriculoperitoneal shunts and lumboperitoneal shunts), and venous stenting are mentioned in the later part of this chapter.

Abbreviations

BMI	body mass index	CSF	cerebrospinal fluid
CT	computed tomography	ICP	intracranial pressure
IIH	idiopathic intracranial hypertension	LP	lumboperitoneal shunt
MRI	magnetic resonance imaging	OCT	optical coherence tomography
ONSF	optic nerve sheath fenestration	OP	opening pressure
VP	ventriculo-peritoneal		

1. Introduction

Idiopathic intracranial hypertension (IIH) is a seldom-seen illness characterized by increased intracranial pressure (ICP) and papilledema in obese women of childbearing age. Patients with IIH experience chronic disabling headaches as a result of elevated ICP, which is considered to be generated by altered CSF fluid dynamics (Mollan et al. 2016). Considering the modified Dandy criteria (Mollan et al. 2018), IIH is largely a diagnosis of exclusion, in which a rising ICP due to other causes is ruled out. A lumbar puncture (diagnostic) must have an opening pressure (OP) greater than 25cm of CSF, indicating that the cerebrospinal fluid (CSF) is primarily acellular and meets normal biochemical criteria. There must also be papilledema, or swelling of the optic disk. With the exception of empty sellae, neuro-anatomical anomalies in suspected IIH patients are ruled out. With the exception of abducent nerve palsy, patients must also have usual cranial nerve function. Diagnostic criteria of IIH is shown in Table 1.

Table 1. IIH diagnostic criteria.

Diagnostic Criteria of IIH
Papilledema
Normal neurological findings with the exception of sixth nerve palsy
Proof of normal anatomy
Normal CSF constituents
Increased opening pressure (OP) on lumbar puncture (>250 mm CSF)

Source: Table adapted from Mollan et al. (2018), used with permission.

2. Epidemiology

In the Western world, the per annum incidence of IIH is around 0.9/100,000 people, and 3.5/100,000 in females aged 15 to 44. The prevalence of IIH is rising in tandem with the present obesity pandemic. Other demographic studies have found that obese women of reproductive age make up the vast majority of IIH cases. The average age of diagnosis is around 30 years. IIH can affect children, males, and the elderly; however, it is less common in these populations. Males account for less than 10% of IIH sufferers in adulthood. Men with IIH are often overweight, much like women. Males with IIH, on the other hand, may have worse visual acuity than women, according to research. IIH has no obvious racial predisposition. While the bulk of epidemiological studies demonstrate that the incidence of IIH is generally the same across countries, a few studies suggest that IIH is

less common among Asians. This is assumed to be due to the lower levels of obesity in various Asian countries (Chen and Wall 2014).

3. Pathogenesis

The pathophysiology of IIH is uncertain; potential theories include aberrant cerebral venous outflow, raised CSF outflow hindrance, obesity-related higher intraabdominal and intracranial venous pressure, abnormal sodium as well as water reabsorption processes, and vitamin A metabolism problems (Bari et al. 2005). The effects of excessive secretion of interstitial fluid as well as extracellular cerebral edema on the blood–brain barrier (BBB) are most likely caused by a pathogenic mechanism: the CSF is generated in a normal way; there are more transactions from the interstitial fluid to the CSF at the trans-ependyma and trans-cerebral pial levels; and there is an increase in resorption of the CSF, and there is a swift venous efflux (Figure 1) (Iencen et al. 2015).

The clinical sign of rapid and possibly turbulent venous flow is tingles in synchronization with an individual's pulse, which are frequent in idiopathic intracranial hypertension. One may consider that an uncertain cause generates impacts on the blood–brain barrier, with the gradual appearance of cerebral edema due to the excess interstitial fluid. The pressure is balanced by the trans-ependymal interchange of edema interstitial fluid from the cerebral parenchyma to the CSF, followed by enhanced resorption of the cerebrospinal fluid, which maintains cerebral blood flow and represents a compensation mechanism (Iencen et al. 2015).

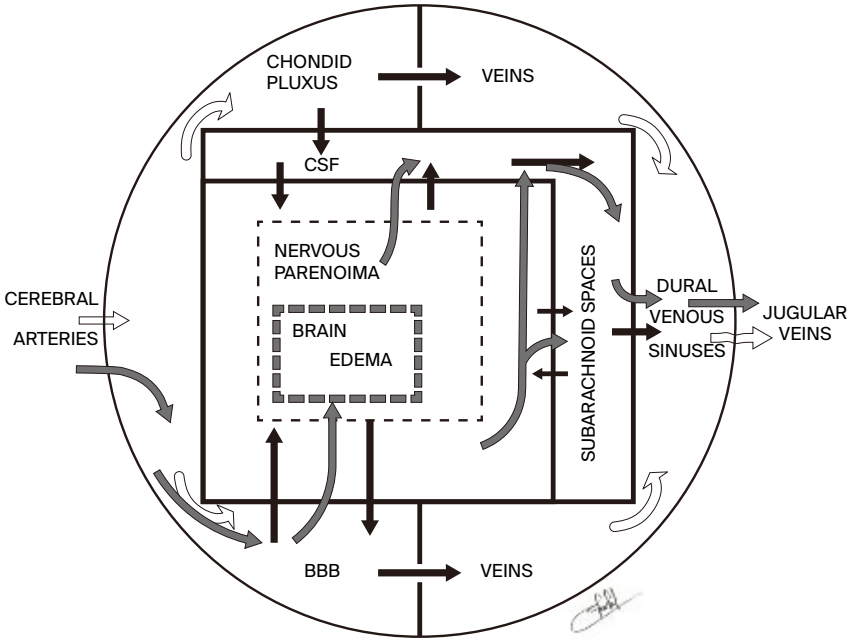

Figure 1. Hydrodynamic model of pathogeny in idiopathic intracranial hypertension. Source: Figure by authors.

When the BBB is gradually damaged, the amount of interstitial fluid grows gradually, and cerebral edema develops (Friedman 2005; Iencean 2003; Iencean 2004; Tabassi et al. 2005; Vorstman et al. 2002; Wraige et al. 2002). The basic ingredient for this balancing mechanism of pressure rise is the interstitial fluid's trans-ependymal and trans-pial route towards the cerebrospinal fluid (Figure 1).

The rise in intracranial pressure is exceedingly slow, and it has a long-term effect (Figure 2). This gradual rise in intracranial pressure permits proper pressure adjustment and near-normal cerebral sanguine flux maintenance. The pathogenic ICP range is extremely high, ranging from 60 to 80 mm Hg, and can remain stable for lengthy periods of time (Figure 2) (Iencen et al. 2015; Wraige et al. 2002).

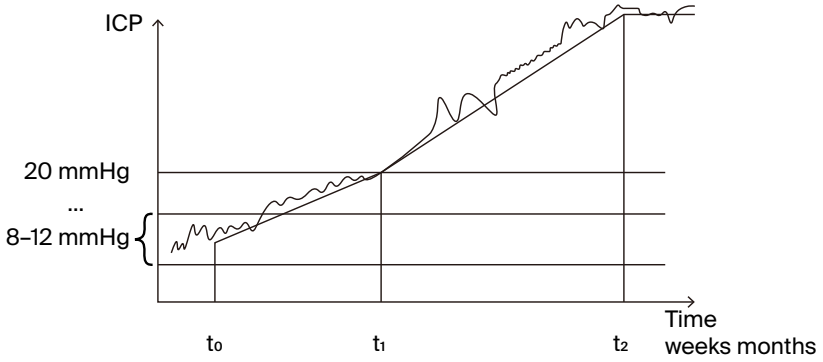

Figure 2. ICP increase in idiopathic intracranial hypertension, with a prolonged infraclinical period and an extremely long duration of pathological pressure value presentation. Source: Figure by authors.

4. Symptoms of IIH

The symptoms of IIH are headache (severe), transient visual obscuration (TVO), blurring of vision, double vision, pulsatile tinnitus, photophobia, retrobulbar pain, and dizziness. According to the International Classification of Headache Disorders, 3rd edition (ICHD-3) (Headache Classification Committee of IHS 2013), most patients with IIH have symptoms such as headaches, which become increasingly worse and more common (Mollan et al. 2018). Headaches caused by increased ICP, like any headaches, can worsen upon waking up and performing the Valsalva maneuver (KesKın et al. 2018).

5. Signs of IIH

Papilledema, visual field loss, and abducent nerve palsy are the most prevalent symptoms of IIH. Papilledema is frequently bilateral and similar; however, it can also be unilateral or asymmetric. Secondary intracranial hypertension caused by cerebral venous thrombosis might have symptoms that are quite similar to those of IIH. It is known that IIH is a long-term disease, which lasts months to years (Whelton et al. 2017). Symptoms of IIH generally worsen slowly; however, a subset has a more fulminant course. The risk factors of IIH are shown in Table 2.

Table 2. Risk factors of IIH.

Major	Others
Gender (>90% of patients afflicted with IIH are female); Obesity.	Addison's disease; Hypoparathyroidism; Steroid discontinuation; Growth hormone utilization in pediatric patients; Malnutrition; Hypervitaminosis A.

Source: Authors' compilation based on data from Chen and Wall (2014).

6. Diagnostic Approach

- The phenotype of headache is extremely variable, and it might be mistaken for various common headache syndromes (Table 1). Investigation and management in this regard are based on symptoms and indicators. For appropriate management, an interdisciplinary approach is essential.
- To exclude malignant hypertension, blood pressure (BP) readings must be as follows: a diastolic blood pressure more than or equivalent to 120 mm Hg and a systolic BP more than or equivalent to 180 mm Hg (Mollan et al. 2018; Whelton et al. 2017).
- Ophthalmological examination: every patient should have confirmed papilledema and a risk evaluation of their visual function. When papilledema is present, the following should be noted:
  - Visual acuity;
  - Pupil evaluation findings;
  - Intraocular pressure;
  - Confrontational visual field test (perimetry) results;
  - Dilated fundoscopy for grading the degree (severity) of the papilledema (Figure 3) and to exclude ocular etiologies for disc edema.

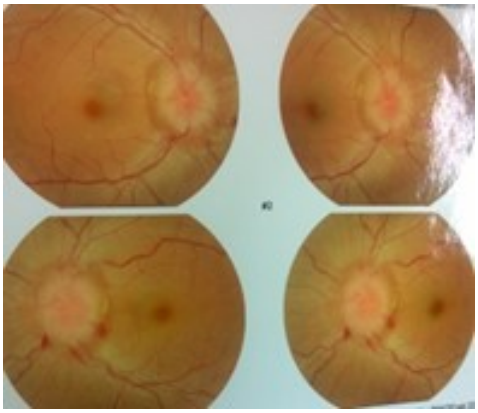

Figure 3. Preoperative fundal photograph of an IIH patient showing advanced papilledema. Source: Figure by authors.

Drawings of the fundus image and significant findings on the optic nerve head should be made and obtained, respectively, whenever possible (hyperemia, hemorrhages, exudate, cotton wool spots, arterio-venous nipping, blurring of the vessels, etc.). The use of photographs and/or optical coherence tomography (OCT) images is beneficial (Mollan et al. 2018). Regular ocular examination is required if visual function is in danger, as this will influence timely management.

- Neurological examination
  - Cranial nerve testing. There must be no cranial nerve dysfunction other than 6th cranial nerve palsy/palsies when IIH is suspected (Mollan et al. 2018).
  - Alternative diagnoses should be considered if other cranial nerves and/or abnormal signs are present.
- Neuroimaging
  - Urgent MRI of the brain and orbit within twenty-four hours; if not possible within twenty four hours, urgent CT scan of brain followed by MRI of the brain if no pathology is found (Mollan et al. 2018).
  - No signs of HCP, a mass, an anatomical or vascular lesion, or aberrant meningeal enhancement should be present.
  - A CT or MR venography must be performed within 24 h to rule out cerebral sinus thrombosis.
  - Features of increased ICP may be found in imaging results, though these are not diagnostic of IIH (Mollan et al. 2018; Degnan and Levy 2011; Hoffmann et al. 2013; Farb et al. 2003; Kelly et al. 2013).

6.1. Neuroimaging Characteristics of Raised ICP

- Empty sellae;
- Partially empty sellae/reduced pituitary height;
- Greater tortuosity and elongation of optic nerve;
- Expanded optic nerve sheath (peri-optic nerve subarachnoid space) (Figure 4);
- Flattened posterior sclera;
- Deflection of the cerebral venous sinuses, involving bilateral transverse sinus narrowing or stenosis of a prime transverse sinus;
- Intraocular projection of the optic nerve head. (Mollan et al. 2018)

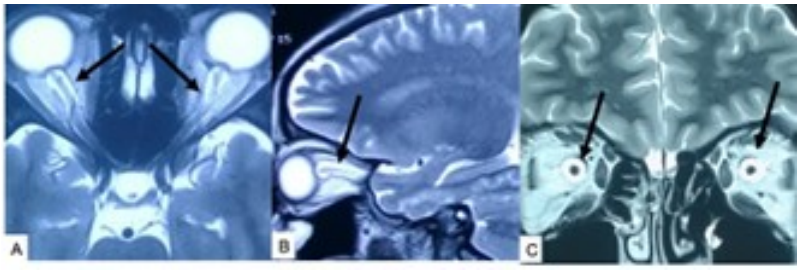

Figure 4. MRI of the brain and orbit: (**A**) axial, (**B**) sagittal, and (**C**) coronal T2W images showing elongated optic nerve with horizontal and vertical kinking, convex optic nerve head invaginating into globe, peri-optic nerve with increased CSF levels, partial empty sella, and normal brain with ventricles. Source: Figure by authors.

6.1.1. Lumbar Puncture

- Patients with papilledema must undergo a lumbar puncture (LP) after normal imaging has been conducted to confirm opening pressure (OP) and normal composition (Mollan et al. 2018).
- In the lateral decubitus posture, measure the LP opening pressure (Friedman et al. 2013). Pressure documentation should take place with the patient in a relaxed state and with their legs extended after the placement of the needle into the CSF space. Allow time for the CSF level to settle before documentation. CSF protein, glucose, and cell count analyses should be conducted.
- To diagnose IIH, a cut-off OP of >250 mm CSF is required per the diagnostic criteria (Friedman et al. 2013).

6.1.2. Exclusion of All Other Possible Causes of Increased ICP

- All patients' should have their medical history thoroughly collected to rule out any secondary reasons for elevated intracranial pressure that have previously been associated with increased ICP.
- To rule out anemia, every patient should be subjected to a complete blood count (Biousse et al. 2003; Mollan et al. 2009).
- If a patient is regarded as abnormal, additional blood testing to rule out secondary reasons may be considered.
- If a patient's condition is regarded as abnormal, further neuroimaging may be considered. More proximal neuroimaging of the neck vessels may be used to rule out internal jugular blockage.

7. Management Principles of IIH

For the optimal management of patients with IIH, there must be seamless interaction between clinician and neurosurgeon to allow smooth joint care between the different specialties. The main goals of treatment of IIH are as follows (Mollan et al. 2018):

1. Mitigate the underlying etiology;
2. Save visual function;
3. Reduce the morbidity due to headache.

IIH management principles: Modify the underlying disease by reducing weight.
Weight loss decreases ICP and is effective in alleviating papilledema and headaches (Sinclair et al. 2010).

- As soon as definitive IIH is established, every patient with a BMI greater than 30 kg/m^2 should be counseled about weight reduction. This should be handled with care.
- It is unknown how much weight reduction is needed to cause a patient to enter remission. It has been noticed that a diagnosis of IIH is related to 5–15% weight gain in the year preceding the diagnosis (Daniels et al. 2007) and that an up to 15% weight reduction is necessary for remission for IIH (Mollan et al. 2018; Sinclair et al. 2010).
- The IIH Treatment Trial (IIHTT) found that combining acetazolamide plus a low-sodium weight-loss diet led to a moderate elevation in visual field function in individuals with mild visual loss when compared to that facilitated by the diet alone. At 6 months, the IIHTT reported better quality-of-life results associated with acetazolamide (Mollan et al. 2014).

The algorithom of managment of IIH is shown in Figure 5.

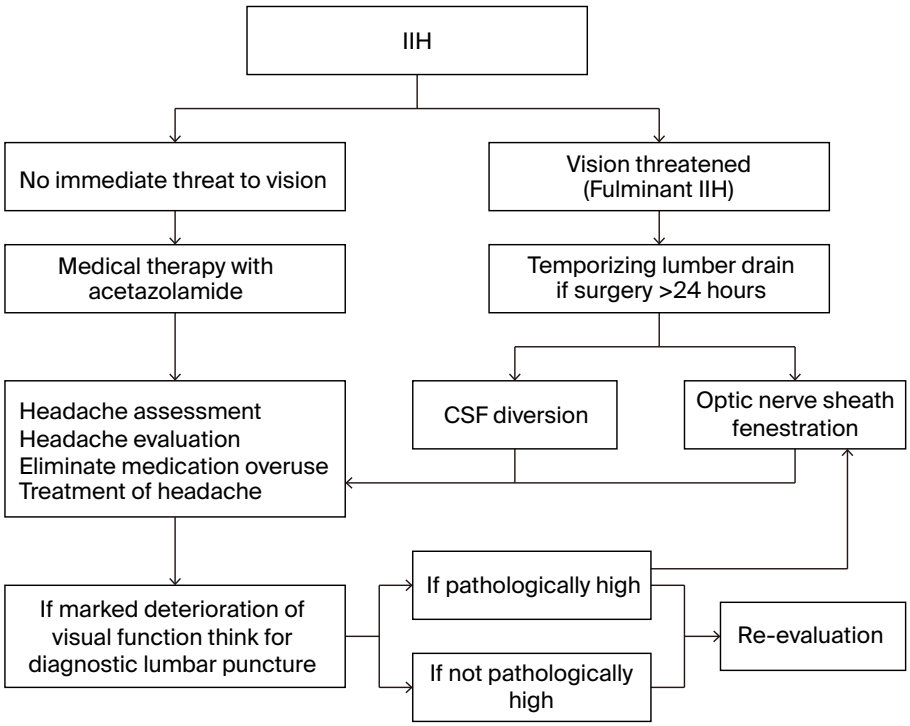

Figure 5. The algorithom of managment of IIH. Source: Authors' compilation based on data from Mollan et al. (2018).

7.1. To Protect Vision

Surgical procedures are indicated for less than 10% of IIH (Friedman et al. 2013) patients who present with a fast progressive reduction in visual function (called fulminant IIH) and require an immediate decrease in ICP to protect their eyesight (Lipton and Michelson 1972; Sodhi et al. 2017). A ventriculoperitoneal (VP) shunt is the most common CSF diversion. A lumboperitoneal (LP) shunt and optic nerve sheath fenestration (ONSF) are two such alternatives. VP shunts are recommended over lumbo-peritoneal shunts (Lipton and Michelson 1972; Sodhi et al. 2017) due to the former's lower reported revision rates (1.8 versus 4.3 revisions per patient, respectively) (Lipton and Michelson 1972; Sinclair et al. 2010).

Many surgical operations, for example, CSF diversion as well as optic nerve sheath fenestration (ONSF), have been shown to be effective in the short term (Mollan et al. 2018; Uretsky 2009). Weight reduction should be used to modify the underlying condition while the procedures are proving effective.

7.2. CSF Diversion

Neurosurgical CSF diversion is the chosen surgical treatment.

- Because of the lower reported number of revisions per patient, a VP shunt should be the recommended CSF diversion method for threatened vision in relation to IIH (Kalyvas et al. 2017).
- An LP shunt could be employed as well.
- When placing VP shunts, it is best to employ neuronavigation.
- To limit the possibility of low-pressure headaches, programable valves containing antigravity or antisiphon systems should be taken into consideration (Mollan et al. 2018).

7.3. Optic Nerve Sheath Fenestration

ONSF is said to have fewer difficulties than CSF diversion (Figure 6), but no cases of fatalities have been found in the literature. Diplopia, ansiocoria, and optic nerve head hemorrhages have all been recorded as transient side effects. More permanent complications, such as branch and central retinal artery occlusions, have been documented on a very infrequent basis. ONSF is sometimes recommended as the first step in the treatment of patients with malignant fulminant IIH as well as those with asymmetrical papilledema causing blindness in one eye (Spitze et al. 2013). If this surgery fails, CSF diversion, which is more intrusive, may be explored (Mollan et al. 2018). Another alternative is ONSF surgery, which is particularly useful for asymmetric papilledema or when

visual problems are the most prominent. It has the potential to cause serious consequences such as visual loss, diplopia, and pupillary dysfunction (Chowdhury et al. 2020; Mulla et al. 2015).

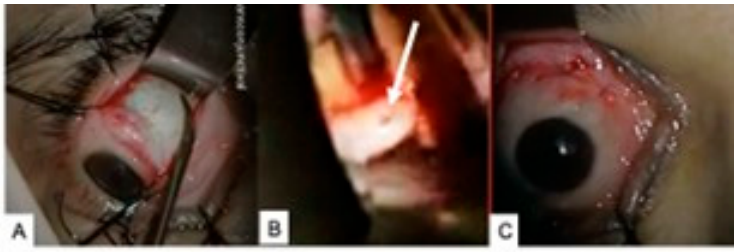

Figure 6. Perioperative pictures of the transconjunctival ONSF approach. (**A**) Right-eye conjunctival incision and approach to optic nerve, (**B**) fenestration on optic nerve sheath, and (**C**) conjunctival repair with sutures. Source: Figure by authors.

7.4. Venous Stenting

Many people with IIH exhibit anatomical anomalies of the cranial venous sinus system observable thanks to advances in venography imaging (Mollan et al. 2018). A narrowing of the prime or both transverse sinuses is one of these anomalies. Stenosis can be caused by the intrinsic dural sinus structure or external compression caused by raised ICP, and lowering ICP might result in the stenosis being resolved.

The degree of narrowing does not appear to be related to ICP or vision loss in a consistent way (Riggeal et al. 2013). In a number of studies, neurovascular stenting has been shown to relieve the symptoms of intracranial hypertension. Many people experience a brief ipsilateral headache after the procedure, especially in the case of stent-adjacent stenosis, which may need retreatment through a third surgery, and artery perforation resulting in acute subdural hematoma, stent migration, and thrombosis in rare cases are possible complications (Mollan et al. 2018).

- Neurovascular stenting's role in IIH has yet to be determined.
- After neurovascular stenting, a long-term antithrombotic drug therapy, is recommended for at least 6 months (Mollan et al. 2018).

7.5. Management of Headache

7.5.1. Lumbar Puncture

Because CSF is released at a rate of 25 mL/hour from the choroid plexus, headache relief after an LP is often short-lived (Wright 1978). As a result, the volume extracted in a so-called therapeutic lumbar tap is quickly replenished. Furthermore, repeated LPs (lumbar punctures) and lumbar–peritoneal shunting have been utilized successfully in the clinical setting. Via lumbar puncture, roughly 30 mL of CSF is drained, or the CSF is drained until the intracranial pressure (as measured via the lumbar technique) is reduced to roughly half of its initial value.

- In spite of the alleviation of headache in nearly 75% of patients, serial LPs are not advised for the treatment of IIH. Many patients experience severe anxiety as a result of LPs, which can lead to acute as well as chronic back pain (Duits et al. 2016).

7.5.2. Acetazolamide

- Acetazolamide, a carbonic anhydrase (CA) inhibitor, is commonly utilized to lower ICP. It is assumed to work by lowering CSF secretion at the choroid plexus.

Two studies showed moderate benefits for acetazolamide for a few outcomes. There is inadequate evidence to advise or reject the efficacy of this drug. Its function and utility are debatable (Biousse et al. 2003; Mollan et al. 2009; Ball et al. 2011).

- It can be used at a maximum dose of 4 g per day (ten Hove et al. 2016); however, Ball et al. (2011) found that 48% of people stopped taking it at 1.5 g because of negative effects.
- A total of 250–500 mg twice a day is a common starting dose of acetazolamide.

7.5.3. Topiramate

Topiramate inhibits hunger by inhibiting carbonic anhydrase. In an uncontrolled open-label study on IIH, it was compared to acetazolamide (Celebisoy et al. 2007). Topiramate has been shown to be effective in the treatment of migraines (Silberstein 2017). With weekly dose increments from 25 mg to 50 mg bd, topiramate may play a role in treating IIH (Mollan et al. 2018; Williams et al. 2017). Topiramate, an anti-epileptic and migraine preventative, was found to be non-inferior to acetazolamide in one trial (Mollan et al. 2014).

7.5.4. Analgesic

In the first few weeks after a diagnosis, short-term painkillers may be beneficial. Nonsteroidal anti-inflammatory medications (NSAIDs) or paracetamol are examples. Indomethacin may provide some benefit due to its ability to lower ICP (Mollan et al. 2018; Liu et al. 2017). Acetazolamide has not been proven to be effective in treating headaches on its own.

8. Definitions

- Idiopathic intracranial: Intracranial hypertension caused by idiopathic factors (IIH). Patients with a high ICP due to an unknown cause meet the criterion.
- Fulminant IIH: Patients with fulminant IIH who meet the criteria for a rapid loss in vision within four weeks of being diagnosed with IIH are said to have fulminant IIH.
- Typical IIH: Females of reproductive age with a body mass index (BMI) more than 30 kg/m^2 are typical IIH patients.
- Atypical IIH: Patients with atypical IIH are not women, are not of reproductive age, or have a BMI of less than 30 kg/m^2. These patients need a more thorough examination to rule out any other potential causes.

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