Journal of Neurology & Experimental Neuroscience

https://doi.org/10.17756/jnen.2020-071

Research Article

Open Access

Idiopathic Intracranial Hypertension: A Review of Nomenclature, Diagnostic Criteria and Management Strategies

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Received: May 13, 2020
Accepted: June 22, 2020
Published: July 01, 2020

Citation: Gupta D, Pradeep R, Mehta A, Javali M, Acharya PT, et al. 2020. Idiopathic Intracranial Hypertension: A Review of Nomenclature, Diagnostic Criteriaand Management Strategies. *J Neurol Exp Neurosci* 6(2): 31-39.

Published by United Scientific Group

Abstract

Background: Idiopathic Intracranial Hypertension (IIH) is a disorder characterized by elevated intracranial pressure without an identifiable underlying aetiology. Cerebrospinal fluid (CSF) analysis and neuro-imaging may be normal, though certain MRI signs have been described which can help in establishing the diagnosis of IIH.

Objective: The prevalence of IIH is on the rise secondary to the global obesity epidemic. The diagnostic criteria and treatment strategies are subject to constant modification and upgradation.

Materials and method: We reviewed literature pertaining to the development of concept and history of IIH, it's varied nomenclature, the newer classification criteria and the treatment strategies.

Results and conclusion: Despite being a century old disease, the nomenclature and diagnostic criteria have undergone constant modifications. The most common symptoms are headache, visual disturbances and tinnitus. Treatment strategies include weight reduction, lifestyle modification, Acetazolamide, analgesics for headache, topiramate and surgical procedures like shunt diversion or optic nerve sheath fenestration.

Keywords

Benign intracranial hypertension, Headache with vision loss, Idiopathic intracranial hypertension, Pseudotumor cerebri syndrome, Papilledema

Key message

The terminology 'benign intracranial hypertension' is best avoided. Clinical course is variable and Acetazolamide is the first line therapy, followed by surgical management.

Introduction

As the name suggests, Idiopathic Intracranial Hypertension (IIH), is a disorder of unknown aetiology, presenting with signs and symptoms of raised intracranial pressure (ICP). The clinical course of the disease is variable and, if untreated, may lead to permanent loss of vision [1]. Recent studies including the Idiopathic Intracranial Hypertension treatment trial (IIHTT) have helped in better understanding of etio-pathogenesis and the treatment protocols [2].

Evolution of Concept of IIH

Perhaps the first detailed report of IIH were published by German internist, Heinrich Quincke, who invented the eponymous procedure, lumbar puncture. He was a pioneer in measuring intracranial pressures via 'Quincke's procedure' and extensively studied cerebrospinal fluid (CSF) dynamics. Through his report in 1896, Quincke proposed an association between headache, visual disturbances and raised intracranial pressure [3].

Another German neurologist, Max Nonne, described 18 cases presenting with a constellation of symptoms indubitable of tumours of either cerebral hemisphere or posterior cranial fossa [4]. However, the patients had an atypical clinical course and evaluation for tumours was negative. In some cases, the CSF pressures were elevated and unlike brain tumours, the patients responded to conservative management. Nonne advocated the term 'pseudotumour cerebri' for such patients.

In 1930, Charles P. Symonds, working in National Hospital, London, published his observations regarding raised ICP in patients with middle ear infection and the therapeutic effects of CSF drainage. He thought that the ear infection was the cause of high ICP and labelled this condition as 'Otitic hydrocephalus', for which the treatment was CSF drainage [5].

Subsequently a number of scientists reported similar cases, with Dyke and Davidoff using the term "hypertensive meningeal hydrops" for a series of patients with clinical syndrome similar to pseudo-tumour cerebri [6].

Walter Dandy, in his eloquent review, described 22 patients with clinical syndrome suggestive of a brain tumour, yet the same being ruled out by ventriculography. 16 of these were females and most of them presented with headache followed by blurring of vision, giddiness, tinnitus, drowsiness and vomiting in various combinations. The outstanding feature in all was symmetrical, bilateral papilledema with or without hemorrhages, enlarged blind spots and scotomas [7]. Neurological examination was largely normal. Lumbar puncture showed an opening pressure ranging from 250 to 550 mm of water with a normal CSF analysis. This formed the basis of first diagnostic criteria for the condition Dandy called as 'Intracranial pressure without Brain Tumour'. With the advent of modern imaging and diagnostic facilities, these criteria have undergone a number of modifications [8].

Foley re-defined the salient features, underlying pathogenesis and prognosis of this syndrome. He opined that prognosis is 'invariably good with the condition subsiding within few weeks or months.' He thus used the term 'Benign intracranial hypertension' for this syndrome [9]. It is now known that the prognosis is not always favourable and untreated cases may lead to permanent blindness. Hence the term 'Benign intracranial hypertension' hast lost significance in recent times. The terminology to define this syndrome is one of the most controversial nomenclature in modern medicine with Deborah Freidman re-popularizing the term 'pseudo-tumour cerebri syndrome (PTCS),' a century after this was introduced by Nonne [10].

Wall et al recommended 'Idiopathic intracranial hypertension' as the most appropriate terminology. In patients in whom a definite cause is identified, terminology like 'tetracycline-induced intracranial hypertension,' 'vitamin A-induced intracranial hypertension', 'steroid withdrawal related intracranial hypertension' can be used otherwise, the term 'intracranial hypertension of unknown cause' is best suited [11].

Epidemiology

Population based studies in the United States of America revealed an annual incidence of 0.9 per 100,000 people in general population [12]. Females were 8 times more commonly affected as males and the average body weight of patients was 38% higher. When corrected for weight, the incidence increased to 13-14.85 per 100,000 for reproductive age females with body weight 10% more than ideal and, to 19.3 per 100,000 in females with body weight 20% more than the ideal weight [12, 13]. Various other studies have reported incidence ranging from 0.03 per 100,000 in Japan to 2.2 per 100,000 per year in Libya [14-17]. A recent Scottish study noted that the incidence of IIH is higher in areas of social deprivation, which is likely due to increased prevalence of obesity in these areas. The incidence of IIH in obese reproductive age females in this study was reported to be 37.9 per 100,000 [18].

Most of the studies on IIH focus on women and thus there is a paucity of data on IIH in male population and children. Bursztyn reported an annual incidence of 0.6 per 100,000 children [19]. A large Canadian retrospective study reported an incidence of 0.9 per 100,000 children, with predominance in females and adolescents (12-15 years of age) [20]. There is a glaring lack of epidemiological studies on IIH in the south-east Asian region, including the Indian subcontinent. Most small-scale studies done in tertiary care referral centres have reported a female preponderance of disease, with mean age of presentation between 25.6 years to 32.89 years [21-24].

Etiology

Although the term Idiopathic intracranial hypertension points to an unknown underlying cause of the disease, a number of secondary causes have been identified (Table 1) [25]. Friedman et al have advocated the umbrella term Pseudo-tumour cerebri syndrome (PTCs) for all such cases with sub-division to primary and secondary PTCs. IIH is then considered a subset within the primary PTCS [10].

Relation of Obesity with IIH

Obesity is traditionally considered to be the strongest risk factor for IIH. Obesity leads to high intra-abdominal and intra-thoracic pressures, which causes functional obstruction to cerebral venous outflow via the jugular venous system. Racial differences have been reported in the association of obesity with IIH. The lower obesity rates in Asian countries have been postulated to account for a lower incidence of IIH,

Table 1: Risk factors and reported associations with IIH.

Established risk factors	a) Female sex
	b) Obesity/ Overweight
	c) Excess Vitamin- A
	1. All trans retinoic acid/ Isotretinoin (ATRA)
	d) Growth hormone
	e) Steroid withdrawal
	f) Endocrine disorders
	1. Addison's disease
	2. Hypoparathyroidism
Probable associations	a) Excess tetracyclines
	b) Thyroid hormone therapy
	c) Indomethacin and ketoprofen in Barter's syndrome
Possible associations	a) Anemia- iron deficiency
	b) Hypovitaminosis A
	c) Drugs/ Antibiotics
	1. Lithium
	2. Amiodarone
	3. Sulpha antibiotics
	4. Nalidixic acid
	d) Sarcoidosis
	e) Systemic lupus erythematosus
	f) Obstructive sleep apnoea (OSA)
Unproven associations	a) Human immunodeficiency virus infection
	b) Systemic hypertension
	c) Menarche
	d) Pregnancy
	e) Menstrual irregularities
	f) Oral contraceptive usage
	g) Long term Corticosteroids
	h) Chronic muti-vitamin tablet use
	i) Chronic kidney disease
Secondary causes of Intracranial hypertension	a) Intracranial smass lesions
	b) Impaired cerebral venous drainage due to
	1. Venous sinus thrombosis
	2. Inherited thrombophilia/ hypercoagulable states
	3. Face and ear, nose, throat infections
	4. Mastoiditis, middle ear infections
	5. Bilateral jugular vein thrombosis/ ligation
	6. Superior venacava syndrome
	7. Increased right heart pressure
	8. Glomus jugulare
	9. Head and neck surgery
	10. Radical neck dissections
	c) Post-inflammatory sequalae (SAH, meningitis)
	d) Elevated CSF proteins levels (GBS, spinal tumours, obstruction)
	e) A-V malformations

as compared to the western world. It is speculated that obesity plays a minor role in Asian IIH patients and they have lower mean BMI compared to west [25, 26]. We propose that much like the South-Asian or the Indian- phenotype for diabetes, similar mechanisms may be playing a role in the non-obese Asian patients with IIH. More studies using the Asian and Indian cut-offs for BMI are needed to better understand this paradox [27]. Association between obesity and IIH is much weaker in pediatric IIH when compared to adults. Balcer et al reported that 43% children aged 3-11 years with IIH were obese, while this proportion increased to 81% in 12-14 years and 91% in 15-17-year-old children, thus postulating that younger children with IIH are less likely to be obese than older ones [28].

Clinical Presentation

The mean age at diagnosis is between 24 to 39 years, with a clear female preponderance (Table 2) [16, 29-34]. The most common symptom is headache, which can mimic any of the primary headache disorders (Table 3).

Table 2: Clinical presentation of IIH in previously reported studies.

	Incidence	Mean age at presentation	Most common symptom
Wall, Iowa [29]		31 years	Headache
Carta, Parma, Italy [30]	0.28/ 100,000	36 years	
Idiculla, Oman [31]	2.18/ 100,000	25 years	Headache
D'Amico, Italy [32]	-	39 years	Headache
Kesler, Israel [33]	2.02/ 100,000	-	Headache
Wall, IIHTT [34]	-	29 years	Headache
Pal, India [35]	-	24.3 years	Dull aching, holocranial pain

Table 3: Relative Frequency of symptomatology of IIH.

1	Headache	75-94%
2	Transient Visual Obscuration	68-72%
3	Pulsatile tinnitus	52-60%
4	Dizziness	53%
5	Photophobia	42-73%
6	Neck pain	42%
7	Visual loss	32%
8	Nocturia	30%
9	Cognitive dysfunction	20%
10	Radicular pain	19%
11	Diplopia	18%

1. Most patients report chronic daily pulsatile bifrontal or holocranial pain of moderate to severe intensity which may be associated with nausea but usually no vomiting.

2. Lateralized throbbing pain, associated with nausea and vomiting.

3. The pain is usually more at night and may awaken the patient from sleep.

4. Changes in posture (bending over or lying down) aggravate pain.

5. Violent coughing or straining may aggravate pain.

6. Associated symptoms of retro-orbital pain, photophobia, phonophobia, neck pain, back ache, radiating or radicular pains may be present.

The other common and troubling symptom complex includes visual disturbances, usually in form of sudden, episodic, brief loss of vision, lasting for few seconds and spontaneously recovering within a minute. These transient visual obscurations (TOV) may be multiple and may involve one or both the eyes [33-36]. It becomes imperative to differentiate TOVs of IIH from amaurosis fugax or visual transient ischemic attacks. Similar to headache, postural aggravations are seen in TOVs. The exact pathogenesis of TOVs is controversial with Sadun et. al. postulating transient optic nerve ischemia, consequent to axonal swelling, intraneural transport and increased influx of interstitial fluid into the optic nerve head [37]. As the underlying pathogenesis is optic nerve dysfunction and edema, TOVs are not pathognomonic of IIH or papilledema.

Papilledema and secondary optic atrophy might ultimately lead to permanent blindness in untreated patients. Abducens nerve palsies, though uncommon, may present with diplopia in 20% of the patients. In a small proportion of patients, anatomic compartmentalization of subarachnoid space around the optic nerve may stop the high CSF pressure gradient from reaching the retrolaminar part of nerve, thus preventing the development of papilledema in IIH.

Nearly half of the patients may present with dizziness and tinnitus, which may be intermittent or continuous, unilateral or bilateral [38]. Unilateral transverse sinus thrombosis causes increased turbulent blood flow in the opposite site, leading to contralateral pulsatile (pulse-synchronous) tinnitus [39]. Hence any IIH patient presenting with tinnitus should raise a suspicion of venous sinus thrombosis and warrants further neuro-imaging. This can further be ascertained if the tinnitus resolves on compression of the internal jugular vein.

Other less common symptoms include neck pain, back pain, radicular pains and mild cognitive decline. It is worth remembering that patients with IIH, have well preserved mentation and symptoms such as altered consciousness, seizures, behavioural changes, gait disturbances and lateralizing neurological deficits preclude the diagnosis and warrant search for another aetiology like intracranial space occupying lesions.

Examination

The patients may be overweight to obese with occasional systemic hypertension. Neurological examination is unremarkable except for unilateral or bilateral abducens nerve palsy [40]. Ophthalmological examination reveals optic disk edema with blurring of disk margins, elevated disk and obscuration of blood vessels. Historically, the presence of papilledema has been considered the sine-que-non of IIH, though recent observations of IIH without papilledema (IIHWOP) have necessitated a revision of this diagnostic criterion. Conversely congenitally anomalous disk or optic nerve head drusen may masquerade as papilledema in normal population. Hence, it is very important to distinguish true from pseudo-papilledema [41, 42]. Loss of visual acuity is seen in advanced disease, but visual field anomalies develop early in the course of disease. The most common field defects are enlarged blind spot, loss of nasal field of vision and generalized constriction of visual field [43]. Physical examination is incomplete without a thorough search for underlying causes like anaemia, ENT infections, obstructive sleep apnoea (OSA), connective tissue disorders like lupus, endocrine abnormalities like Addison's disease and features of steroid withdrawal. Blood pressure monitoring is essential to rule out malignant hypertension.

Investigations

Newer advances in the field of neuro-ophthalmology have enabled objective assessment of optic nerve structure and function using trans-orbital sonography [44], optical coherence tomography [45, 46] and fundus fluorescein angiography [47]. Coloured Fundus photography is an excellent tool to record the fundus findings and use as objective evidence in long term monitoring of the disease. Optical Coherence Tomography is a non-invasive, quick tool and is superior to fundus photography. Ultrasonographic B-scan and fundus fluorescein angiography can also be used to assess the optic disc height and papilledema.

Role of Brain Imaging

IIH is usually a disease of reproductive age females and Computed tomography (CT) is best avoided due to radiation risk. Magnetic Resonance Imaging (MRI) with contrast is the favoured modality and should include orbital images and an accompanying venogram of the brain. Fat suppression images help in better visualization of the intra-orbital part of the optic nerve [42]. Exclusion of structural lesions, hydrocephalous and venous sinus thrombosis is a prerequisite for diagnosis of IIH. Recent advances in neuroimaging have identified some common radiological signs (Figure 1 and Table 4), though none of them is considered pathognomonic for IIH [48-50]. Whether all suspected IIH patients should have neuro-imaging is debatable, but paediatric patients, men with suspected IIH, elderly patients, thin females and patients with an atypical symptomatology or tinnitus at presentation should definitely undergo imaging to exclude secondary causes [51, 52].

Lumbar Puncture

A lateral decubitus LP, with measurement of CSF opening pressure is mandatory for diagnosis of IIH. Diurnal fluctuations and postural variations of CSF pressure are known, in addition to transient elevation of CSF pressure

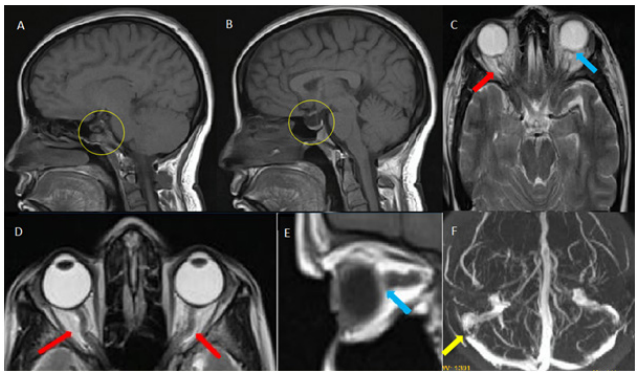

Figure 1: Neuro-Imaging in IIH.A-B: MRI Sagittal FLAIR images showing empty sella; C-D: MRI axial cuts showing tortuous and distended optic nerves (arrows); E: MRI orbit sagittal cuts showing flattening of posterior part of globe and tortuous optic nerve; F: MRI venogram showing right transverse sinus stenosis (arrow).

Table 4: Neuro-imaging signs of IIH.

Empty sella or flattening of the pituitary
Tight subarachnoid spaces,
Flattening of the posterior globe
Protrusion of the optic nerve head
Enhancement of the prelaminar portion of the optic nerve head
Distension of the optic nerve sheath
Vertical tortuosity of the optic nerve

during crying, shouting and Valsalva manoeuvre [53]. As such repeated lumbar punctures may be required for an accurate pressure reading. A useful clue is asking the patient for temporary symptomatic improvement in headache and visual complaints following CSF withdrawal [42]. The 95% reference interval for CSF opening pressure is shown to be 10-25 cm of water [54]. Hence, the diagnosis of IIH requires demonstration of an opening pressure more than 25 cm of water in lateral decubitus position with legs relaxed and extended [55].

Diagnosis of IIH

The original criteria proposed by Dandy [7] in 1937 were further modified by Smith in 1955 and were the preferred criteria in the late 20th century [8, 9, 34]. With an improvement in understanding of the nature and pathophysiology of IIH, diagnostic criteria have been updated from time to time and the most recently used criteria in the Idiopathic Intracranial hypertension trial are given in table 5.

Relation of IIH with CSF pressure

Traditionally IIH is diagnosed by demonstrating elevated CSF pressures of > 25 cm of water. However, normal fluctuations of CSF opening pressure are well known. In case of CSF opening pressure between 20-25 cm of water, it is better to repeat the lumbar puncture at a different time or do a 24-hour ICP monitoring. In case the patients do not consent for the same, additional fundus or MRI criteria are required to make a diagnosis of IIH [5, 11]. The cause of 'lower' CSF pressures in this small subsection of IIH patients may be related to inherent susceptibility of their optic discs to even slight changes in ICP. Whether genetic or environmental factors play a role is yet to be elucidated. Presence of a CSF leak (rhinorrhoea or otorrhoea may also decrease the CSF pressure. Recognising these patients early is of utmost importance, as untreated papilledema and IIH can have devastating outcomes.

Inherent susceptibility of optic disc to CSF pressure may also explain the sub group of patients on the opposite spectrum, that is high CSF pressure without papilledema. Though advancements in neuro-imaging have led to criterion for diagnosing papilledema-negative IIH, its existence is rare and contentious. The etio-pathogenesis and diagnostic criterion of childhood IIH is still debated and pre-pubertal IIH is different from IIH in older children (Table 5).

Treatment of IIH

The main goal of IIH treatment is prevention of visual loss along with symptomatic relief of headache. Management is largely based on clinical experience and expert opinions as there is a paucity of literature to define evidence-based management guidelines. The recent IIH-treatment trial (IIHTT) has provided a better understanding of the treatment options [2, 56].

Lifestyle modification and weight reduction strategies

IIH is typically a disease of obese, and weight reduction becomes an essential management strategy. In a small prospective study in United Kingdom, low calorie diet (425 Kcal/day) with 2L fluids per day with or without acetazolamide has been shown to reduce weight dramatically, leading to significant reduction in intracranial pressure with improvement in headache and papilledema [57]. Similar results were obtained in a Danish study, where weight loss of more than 3.5% BMI correlated with significant decrease in disease activity, CSF pressures and a favourable outcome [58]. Behavioural and dietary weight loss is often challenging and ill-sustained and most patients regain some proportion of their original weight within 1-5 years [59]. Sustainable weight loss approaches are the need of the hour and in this respect, the role of bariatric surgery is being evaluated in the IIH-weight trial (IIH-WT), with the results expected around 2022 [60].

Pharmacological management

The carbonic anhydrase inhibitor, Acetazolamide, forms the cornerstone of IIH management. Its efficacy has been well established, however side effects like altered taste, dizziness, parasthesias, nausea, vomiting and diarrhoea preclude its use in clinical setting. Literature on the dosage requirement and the duration of therapy in IIH are lacking. IIH-treatment trial has shown improvement in visual field function after six-month therapy with acetazolamide in patients with mild visual loss [2, 56]. The treatment effect was greater in patients with higher grade papilledema at baseline. With a mean dose of 2.5 gm, gastro-intestinal side effects, fatigability, and parasthesias were significantly more frequent.

Table 5: Diagnostic criteria for IIH.

Modified Dandy criteria	a) Signs and symptoms of increased ICP- headache, nausea, vomiting, transient visual obscuration, papilledema
	b) No localizing signs with exception of unilateral or bilateral abducens nerve palsy
	c) Elevated CSF pressure > 25 cm of water with a normal CSF composition
	d) Normal to small (slit) symmetrical ventricles on imaging with no intracranial mass.
IIHT trial Modified Dandy criteria [34]	a) Signs and symptoms of increased ICP
	b) Absence of localizing findings on neurological examination. (except those known to occur with increased ICP)
	c) Absence of deformity, displacement, or obstruction of the ventricular system and otherwise normal neuro-diagnostic studies, except for evidence of increased cerebrospinal fluid pressure (> 20 cm water). Abnormal neuroimaging except for empty sella turcica, optic nerve sheath with filled out CSF spaces, and smooth-walled non-flow-related venous sinus stenosis or collapse should lead to another diagnosis
	d) Awake and alert patient
	e) No other cause of increased intracranial pressure present
For CSF pressure 20-25 cm of water	One additional criterion required
	a) Abducens palsy
	b) Pulse synchronous tinnitus
	c) Frisen grade II papilledema
	d) Echography for drusen-negative and no other disk anomalies mimicking disk edema present
	e) MR venography with lateral sinus collapse/ stenosis preferably using auto-triggered eliiptic centric-ordered technique
	f) Partially empty sella on coronal or sagittal views and optic nerve sheaths with filled out CSF spaces next to globe on T2-weighted axial scans.
IIH without Papilledema [11, 71]	The following should be satisfied, along with unilateral or bilateral abducens palsy.
	a) Normal neurological examination except for cranial nerve abnormalities
	b) Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
	c) Normal CSF composition.
	d) Elevated lumbar puncture opening pressure [> 25 cm CSF in adults and > 28 cm CSF in children (25 cm CSF if the child is not sedated and not obese)] in a properly performed lumbar puncture
	In absence of abducens palsy, 3 of the following neuroimaging findings in addition to above may SUGGEST a diagnosis of IIH
	a) Empty sella
	b) Flattening of posterior aspect of globe
	c) Distention of peri optic subarachnoid space with or without a tortuous optic nerve
	d) Transverse venous sinus thrombosis
Pre-pubertal IIH [40, 72]	a) If symptoms or signs present, they may only reflect those of generalized intracranial hypertension or papilledema; normal mental status
	b) Documented elevated intracranial pressure (age appropriate) measured in lateral decubitus position: neonates > 76 mm of water, 1-8 years > 280 mm of water
	c) Normal CSF composition except for neonates who may have upto 19 WBC/mm3 (if 0-28 days) and upto 9 WBC/mm^3 (if 29-56 days). Protein may be as high as 150mg/dL
	d) No evidence of hydrocephalus, mass, structural or vascular lesion on MRI, with and without contrast, and MR venography. Narrowing of transverse sinuses is allowed.
	e) Cranial nerve palsies are allowed if there is no other identifiable aetiology and improve with reduction in CSF pressure or resolution of other signs and symptoms of intracranial hypertension.
	f) No other identified cause of intracranial hypertension.

In our clinical experience, dosages starting at 500 mg/day and slowly titrating to 1.5-2 gm/day, in 3-4 divided doses have been adequate. Many patients complain of diarrhoea which can be minimized by smaller, more frequent doses.

Headache is the primary reason for seeking a medical consultation, therefore, pain relief assumes a priority in such patients. A number of pathogenic mechanisms, including raised intracranial pressures, venous sinus thrombosis, co-existing migraine and medication overuse contribute to the genesis of headache. In addition, we have encountered patients with post-LP low pressure headaches, which complicates the management further. Treatment with acetazolamide plus non-steroidal anti-inflammatory drugs usually suffices, though occasional patients need anti-migraine prophylactic therapy. A combination of cyproheptadine with propranolol works well for patients with post LP low pressure headaches. The medications are generally continued till the patient gets symptomatic benefit and papilledema resolves.

Topiramate has been tested due to pleiotropic effects including weight loss, CSF pressure reduction and migraine prophylaxis. The potential advantages are offset by disabling side effects, not the least of which include cognitive dysfunction and adverse effect on foetus [61]. In patients refractory to medical therapy, Greater Occipital nerve block may be considered for pain relief.

Surgical management

It is vital to identify high risk patients- those with impending visual loss- for urgent surgical intervention for prevention of permanent visual loss. Patients showing partial or incomplete response to medical therapy after 6 months may also be considered for surgical management. A number of procedures for reducing CSF pressure are available, though head to head trials of one over the other are limited [62]. Hence the choice of procedure usually depends on the surgeon's preference, experience and expertise. These include CSF diversion by lumbo-peritoneal (LPS) or ventriculo-peritoneal (VPS) shunts. Ventriculo-jugular, Ventriculo-atrial and Ventriculo-pleural shunts are now out of favour, but deserve a mention for their historical significance. CSF shunting is a temporary measure in patients with fulminant presentation as it can improve visual function [63]. Post shunting, up to three-fourth patients have persistent or recurrent headache with nearly 28% of them reporting a change of phenotype to low pressure headache [64]. Complications like abdominal pain, backache, infections and shunt block necessitate shunt revision in more than half of the patients. Though newer shunts with valve system and CSF reservoir have improved outcomes, it is recommended to use shunts as the last resort after failure of first line conservative management options.

De Wecker described Optic nerve sheath fenestration (ONSF) in 1872 as an incision in the meninges surrounding the optic nerve, leading to reduction in intracranial pressure. A unilateral procedure might lead to resolution of headache as well as improvement in contralateral disk edema, through a filtration effect propagated throughout the CSF circulatory system. Consequently, a bilateral ONSF may not always be required in patients with bilateral papilledema [65, 66]. Exploration and incision around the optic nerve needs technical skill and can lead to devastating complications like traumatic optic neuropathy, retinal vascular occlusion, diplopia and pupillary dilation in amateur and novice hands [67].

Endovascular venous sinus stenting is gaining prominence as a modality for patients with proven stenosis, classically of the transverse sinus. Stenting increases CSF drainage leading to lowering of CSF pressure and symptomatic relief [68]. Similar to CSF shunts, stent thrombosis, migration, venous sinus perforation and recurrent stenosis proximal to the stent location may need revision [69]. Our experience with surgical management is limited and none of the patients have required a revision of procedure in the first 6-12 months.

Management of the underlying aetiology may also help in reversing the symptoms of IIH as evidence by remission of IIH following counter-clockwise maxilla-mandibular advancement (CC-MMA), done for a 47-year-old female with obstructive sleep apnoea [70].

IIH- The road ahead

Despite being a century old disease, newer insights into pathogenesis and evaluation of IIH add to the existing body of literature. Some authors have put forward the concept of IIH without papilledema (IIHWOP) which requires further investigation [71, 72]. Clinical application of newer modalities like ophthalmic ultrasonography, optical coherence tomography (OCT), fundus fluorescein angiography and Transcranial doppler need further investigation [73]. Till date, there are no universal guidelines for the management of IIH, and there's an unmet need for large scale prospective clinical trials evaluating the same.

Conflicts of Interest

Nil

Source of Funding

Nil

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