

Original Article**A Simplified Policy for Management of Idiopathic Intracranial Hypertension***Mohammed Adawi*, Islam Aboulfetouh and Ahmed Saleh*

Department of Neurosurgery, Benha University, Egypt

ABSTRACT

Background: Idiopathic intracranial hypertension (IIH) is a disorder of elevated intracranial pressure of unknown cause. It is characterized by increased intracranial pressure with its attendant signs and symptoms in an alert and oriented patient, but without localizing neurologic findings. **Objectives:** To evaluate management outcome of idiopathic intracranial hypertension (IIH) by depending on visual field changes as an indication for surgery. **Patients & Methods:** The study included twenty three IIH patients fulfilling Dandy criteria with free MRI brain. All patients underwent lumbar puncture (LP) with measurement of CSF opening pressure and analysis of the released volume CSF. All patients underwent complete ophthalmological assessment, and decision making was based on the results of visual field examination. **Results:** Seven patients presented with insignificant visual field changes passed smooth follow-up for 9-18 months without deterioration of visual field or recurrence of symptoms after the initial LP and the maintained medical treatment. Four patients presented with marked visual field affection and lumbo-peritoneal shunt (LPS) was inserted urgently. Twelve patients presented initially with mild visual field changes developed deterioration of visual field on follow-up visits and underwent LPS. **Conclusion:** Visual field testing is a reliable indication for surgery in patients with IIH as visual acuity and color perception are generally affected late in papilledema..

Received:

10 April 2016

Accepted:

20 August 2016

Key words:Idiopathic intracranial hypertension,
Lumbo-peritoneal shunt,
Visual field

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INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a pathological state defined as an isolated rise of intracranial pressure (ICP) that is not related to an intracranial process, cerebral venous thrombosis, or a meningeal process. Although not always literal, the term “idiopathic intracranial hypertension” is currently the preferred designation for this disorder rather than “pseudotumor cerebri” which often includes patients with other causes of raised ICP and than “benign intracranial hypertension” which is erroneously reassuring, considering that a substantial proportion of IIH patients irreversibly lose vision^{1,2}.

Typically IIH presents with symptoms and signs of raised ICP; headache is the most common symptom at presentation and is less likely to be reported by men than by women. Majority of patients showed the criteria for headache attributed with raised ICP including progressive, daily, diffuse, non-pulsatile headache with aggravation by coughing or straining.^{3,4}

Papilledema is the most common sign of IIH and can result in insidious and slowly progressive visual loss, which is usually reversible with appropriate treatment. However, up to 25% of IIH patients develop secondary optic atrophy and associated permanent visual loss. Visual loss is usually relatively mild at presentation but progresses insidiously. In fact, most

patients have visual field defects on automated perimetry at presentation, but are unaware of their visual dysfunction and visual field abnormalities that typically progress from enlargement of the physiologic blind spot, to nasal and arcuate defects, and ultimately to severe visual field constriction^{5,6}.

Treatment approach for IIH depends on the severity and time course of symptoms and visual loss. The main goals of treatment are alleviation of symptoms, including headache, and preservation of vision. All overweight IIH patients should be encouraged to weight loss, along with a low-salt diet. When there is mild visual loss, medical treatment with acetazolamide should be initiated. When visual loss is more severe or rapidly progressive, surgical interventions, such as optic nerve sheath fenestration or cerebrospinal fluid shunting, may be required to prevent further irreversible visual loss. The choice of intervention depends on the relative severity of symptoms and visual loss, as well as local expertise^{1,7,8}.

PATIENTS AND METHODS

The current study was conducted at Neurosurgery Departments at Benha University Hospitals, and Central Hospitals, KSA since Jan 2011 till March 2015 so as to allow a minimum follow-up period of 6 months for the last case enrolled in the study. Enrolment criteria included fulfillment of the Modified Dandy's Criteria for the diagnosis of IIH as shown in (Table 1)⁹. Exclusion criteria included intracranial pathology on MRI brain, patients with elevated intraocular pressure, and patients with retinopathy.

***Corresponding Author:**

Mohamed M Adawi, MD

Department of Neurosurgery, Benha University, Egypt

Email: mohamed.adawy@fmed.bu.edu.eg; Tel.: 01006299057

Table 1: Modified Dandy's Criteria for the diagnosis of IIH⁹

1.	Signs and symptoms of increased intracranial pressure (headache, nausea, vomiting, transient obscuration of vision, papilledema)
2.	No localizing, focal neurologic signs on neurological examination, except unilateral or bilateral VI nerve paresis.
3.	CSF opening pressure in the lateral decubitus position of > 200 mmH ₂ O in non-obese patients and >250 mmH ₂ O in obese patients; CSF is free of cytological or chemical abnormalities.
4.	Absence of deformity, obstruction, and displacement of the ventricular system as confirmed by radiological workup. Otherwise normal neuro-diagnostic studies except for increased ICP.
5.	Awake and alert patient
6.	No definite cause for increased ICP

All enrolled patients underwent detailed history taking including demographic data for age and gender, date of presentation, presenting symptoms of headache, nausea, vomiting, neurological symptoms, visual symptoms, and medications received for these symptoms and response to it. Also, history included inquiries about the presence and duration of any systemic disease, as hypertension, diabetes mellitus or relevant drug history as systemic corticosteroids, oral contraceptive pills, and vitamins.

Physical examination included determination of body weight, height and calculation of body mass index (BMI). BMI was defined as weight in kilograms divided by the square of the height in meters. Patients were graded according to the international classification of BMI into: underweight (BMI<18.5 kg/m²); normal weight range (BMI=18.5-24.99 kg/m²); overweight (BMI=25-29.99 kg/m²); Obese (BMI>30 kg/m²)¹⁰. The severity of headache was evaluated using the 11-point Numeric Rating Scale (NRS)¹¹ for assessment of pain intensity with numbers from 0 to 10 where 0 indicates no pain and 10 indicates worst pain imaginable.

A detailed ophthalmic examination was performed, including best corrected snellen visual acuity, refraction, ocular motility examination, pupil evaluation, color vision, intraocular pressure measurement, fundus examination with evaluation of degree of papilledema according to Frisén Scale of Papilledema¹², and visual field testing, which is considered the cornerstone in our decision making, is performed and graded according to classification system for the Idiopathic Intracranial Hypertension Treatment Trial (IIHTT).¹³

Then, all patients underwent complete neurological examination for cranial nerve deficits or associated neurological diseases. All patients underwent MRI brain for assurance of being free of pathology and diagnosing signs of increased intracranial pressure and underwent MRV to exclude sinus thrombosis.

All patients underwent lumbar puncture, after confirming free MRI brain and papilledema, to fulfill the diagnostic criteria of IIH. The opening pressure was measured using a manometer held at the level of the left atrium with the patient relaxed in the lateral decubitus position, prior to the removal of CSF. Generally, an opening pressure ≤200 mmH₂O is considered normal, a pressure value of 201-249 mmH₂O is inconclusive and a

pressure >250 mmH₂O indicated high intracranial pressure. CSF sample was obtained for chemical, cytological and bacteriological examination and then an appropriate volume of CSF was drained.

Headache was scored prior to and at completion of drainage and closure headache score was considered as baseline score for follow-up. Patients were maintained on medical treatment including acetazolamide with dose not less than 1 g/day, topiramate 100 mg/day was added when headache is the most distressing symptom. Patients were followed-up for their headache scores, Snellen's visual acuity, grading of papilledema and most important visual field once weekly for one month and monthly for 3 months and then every six months, even after shunt insertion or stopping medical treatment. Visual field deterioration was defined when follow-up Visual field mean deviation (MD) was worsened by ≥2 to 3 dB from the average baseline MD range (-2 to -7 dB). Any significant visual field deterioration, was considered an indication for insertion of lumbo-peritoneal shunt. Suggested treatment algorithm is shown in (Figure 1).

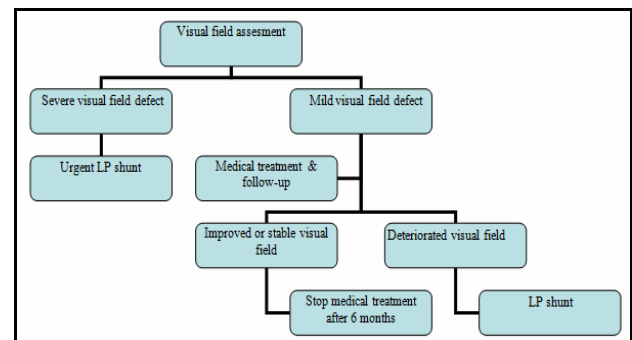


Fig. 1: Summary of management protocol after confirming diagnosis of idiopathic intracranial hypertension. LP shunt: Lumbo-peritoneal shunt.

RESULTS

The study included twenty three patients; seventeen females and six males with mean age of 37.2±6.4; range: 25-47 years. Mean BMI of enrolled patients was 32.5±6; range: 23.8-44.6 kg/m² and only four patients were of average weight and six patients were overweight, while thirteen patients were obese of varying extent (Table 2).

Table 2: Patients demographic data

			<i>Frequency</i>	<i>Mean (±SD)</i>
Age (years)	Strata	<30	4 (17.4%)	27±1.4
		30-40	8 (34.8%)	34.5±2.6
		>40	11 (47.8%)	42.7±2.1
Total			23 (100%)	37.2±6.4
Weight (kg)			23 (100%)	94.5±17.6
Height (cm)			23 (100%)	170.6±3.2
BMI (kg/m ²)	Strata	Average (<25)	4 (17.4%)	24.4±0.5
		Overweight (25-30)	6 (26.1%)	28.3±1
		Obese (>30-35)	6 (26.1%)	33.9±0.4
		Very obese (>35-40)	5 (21.7%)	37.5±0.4
		Morbid obese (>40)	2 (8.7%)	44.2±0.6
Total			23 (100%)	32.5±6

All patients presented by headache with a mean total NRS of 7.8±0.9; range: 6-9. Six patients had nausea and vomiting. Eighteen patients had reduced visual acuity and only one of them had counting fingers. Four patients had transient visual obscuration and nine patients had diplopia. Twenty-two patients had bilateral

disc edema, and one patient had unilateral edema. Twelve patients had papilledema grade 1, eight patients had papilledema grade 3 and three patients had papilledema grade 4. In all patients mean visual field deviation was -7.2; range: -12.8 to 7.8, (**Table 3**).

Table 3: Patients' clinical data

<i>Data</i>		<i>Findings</i>	
Headache		23 (100%)	
Reduced visual acuity		13 (56.5%)	
Transient visual obscuration		4 (17.3%)	
Nausea & vomiting		6 (26%)	
Diplopia		9 (39.1%)	
NRS scoring of headache severity	6	1 (6.3%)	
	7	8 (34.8%)	
	8	8 (34.8%)	
	9	6 (26.1%)	
	Total	7.8±0.9	
Ophthalmic findings	Snellen's visual acuity	6/6	5 (21.7%)
		6/9-6/18	7 (30.4%)
		6/24-6/60	10 (43.5%)
		CF	1 (4.34%)
	Disc edema score	Grading	1
3			8 (34.78%)
4			3 (13.04%)
Laterality	Bilateral	22 (95.6%)	
	Unilateral	1 (4.4%)	
Mean visual field deviation (dB)		-7.2	

Data are presented as numbers & mean±SD; percentages are in parenthesis; NRS: Numeric Rating Scale; CF: Counting Fingers

Out of the studied patients, only four patients presented with severe visual field defect and were sent for application of lumbo-peritoneal shunt on urgent basis. Two patients showed improved visual field defect (**Figure 2**), one patient showed persistence of visual field defect without significant change than preoperative test, but in the fourth patient visual field defect could not be determined precisely either pre or postoperatively due to markedly decreased visual acuity down to

counting fingers grade and showed no postoperative improvement.

Seven patients passed smooth follow-up without deterioration of visual field or recurrence of symptoms after the preliminary lumbar puncture and were well-maintained on medical treatment for at least 6 months. Twelve patients developed deterioration of visual field and underwent shunting after a mean duration of 6.5±1.8; range: 3-12 weeks.

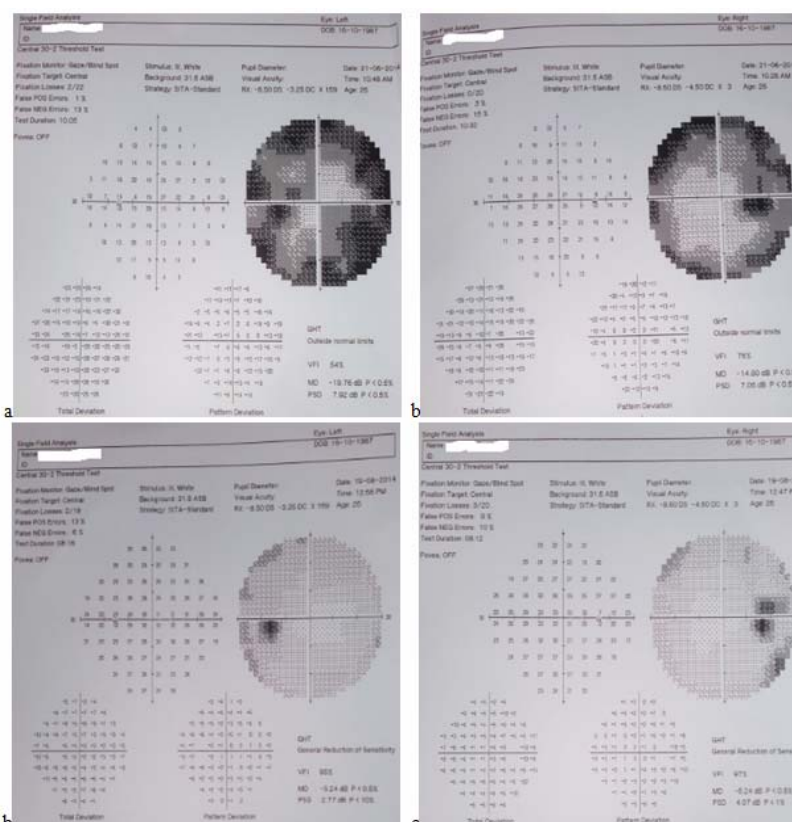


Fig.2 a-d: Visual field performed by Humphery automated perimetry with acceptable reliability indices. a: Pre-operative visual field of the left eye showing severe generalized reduction of retinal sensitivity, and central scotoma together with advanced annular defect giving rise to tubular vision. b: Pre-operative visual field of the right eye showing severe generalized reduction of retinal sensitivity, and paracentral scotoma together with double arcuate scotoma. c: Post-operative visual field of the left eye showing only mild generalized reduction of retinal sensitivity, with marked improvement of the pre-operative defect. d: Post-operative visual field of the right eye showing only mild generalized reduction of retinal sensitivity, with marked improvement of the pre-operative defect.

Procedure for lumbo-peritoneal shunt (LPS)

Shunting was performed under general anesthesia with the patient in the lateral decubitus position with the left side up. Lumbo-peritoneal shunt tube was inserted into the lumbar subarachnoid space using Touhy needle; then tunneled subcutaneously from the spine through 1-cm transverse incision situated at the midline on lumbar spine and subcutaneous tunnel was dissected blindly using a passer to reach another skin incision made over the rectus sheath at the level of umbilicus and the tube was inserted into the peritoneal cavity. Laparoscopic assistance was considered in obese patients and in revision cases.

Data collected including operative time, frequency of the need for fluoroscopic guidance for localization of the lumbar subarachnoid space, the need for laparoscopic assistance for peritoneal insertion of the shunt tubing, duration of hospital stay, and frequency of

postoperative complications. Patients were followed-up as usual with follow-up program described earlier.

Postoperative complications

One patient developed temporary over drainage in the form of headache on sitting and walking only, while there was no headache in resting stage; received intravenous fluid therapy and maintained in recumbant position; symptoms were improved gradually.

An obese female patient had persistent preoperative visual symptoms with aggravated headache after insertion lumbo-peritoneal shunt, abdominal ultrasonographic examination was inconclusive (seems to be due to the barrier effect of fat and inadequate experience of radiologist secondary low case number. CT examination detected extraperitoneal insertion of the distal end of the tube (**Figure 3**). Under general anesthesia, general surgeon was consulted and using single-port approach laparoscopy the distal end of the tube could be inserted in the peritoneal cavity.

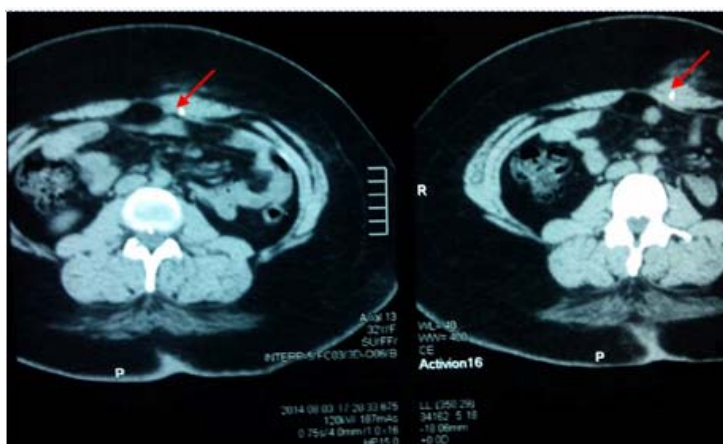


Fig. 3: Showing a case with extraperitoneal distal tube insertion (arrow)

Another patient presented with CSF leak from the back wound and recurrence of symptoms, 3D-CT imaging showed slipped peritoneal end of the LPS with coiling of the tube (**Figure 3**). Surgical exploration was

conducted and a subcutaneous stitch was detected hanging the tube (**Figure 4**), the stitch was removed and the peritoneal end of the tube was re-inserted after confirming its integrity.

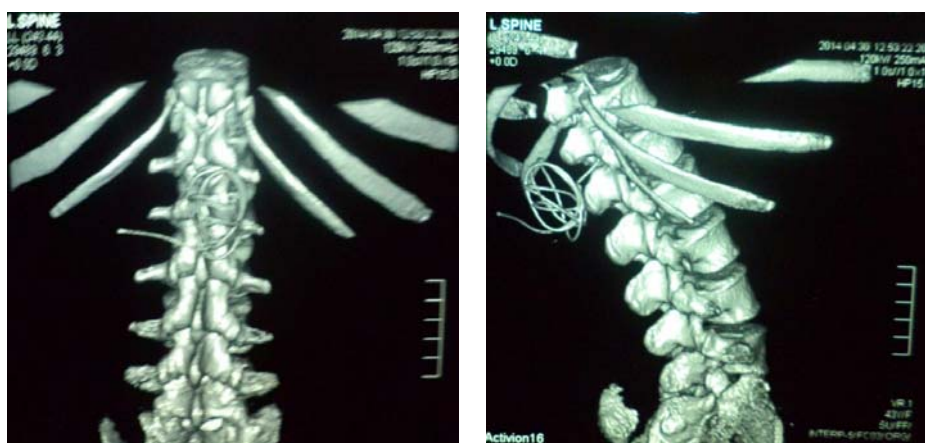


Fig. 4: 3D-CT imaging showing a case had slipped peritoneal end of the LPS with coiling of the tube



Fig. 5: Showing tube blockage by a stitch (arrow) taken wrongly around the tube during closure of subcutaneous space

DISCUSSION

The current study included twenty three patients fulfilling the diagnostic criteria of IIH; thirteen patients were obese of varying extent, six patients were overweight and only four were of average weight with BMI range of 23.8-44.6 kg/m². The high percentage of obese patients (56.5%) indicated the relationship between obesity and possibility of developing IIH and supported that previously reported in literature^{14,15,16}.

All patients underwent lumbar puncture, the mean opening pressure was 369.5±50; range: 283-386 mmH₂O, which coincided with that reported by Ambika et al.¹⁷ who in their series of 50 IIH patients, CSF opening pressure was 250-350 mmH₂O in 39 patients and was >350 mmH₂O in 11 patients. Also, El-Saadany et al.¹⁸ reported that among 22 patients who underwent lumbo-peritoneal shunt (LPS) placement for IIH, 16 patients had severe and fulminant opening CSF pressures with values of more than 400 mmH₂O.

Seven patients received and were maintained on medical treatment and passed smooth follow-up period for a mean duration of 14±2.9 (range 9-18) months. Five patients showed improvement of visual field, but two did not show any improvement of their mild defect. These findings indicated the possibility of control of intracranial pressure using medical treatment after pressure normalization by lumbar puncture and go in hand with Ambika et al.¹⁷ who after initial lumbar puncture started medical treatment for all patients and 70% of patients responded, while 30% patients had to undergo LPS.

Painhas et al.¹⁹ described a case developed headache, nausea and occasional vomiting since two months with recent transitory visual obscurations; examination revealed vision was 20/20 in both eyes, bilateral disc swelling, but normal neurologic examination and MRI, diagnostic lumbar puncture demonstrated an elevated opening pressure with normal compounds; oral acetazolamide was started and ocular and systemic symptoms totally disappeared in three weeks and disc swelling gradually improved in the following months.

In this study, four patients presented with severe visual field defect and were sent for application of lumbo-peritoneal shunt on urgent basis, two of them showed post-operative improvement in their visual field defect.

In line with shunting procedure as the 1st choice for management of IIH, Kumar et al.²⁰ presented a case of 16-year-old morbidly obese girl with a diagnosis of pseudotumor cerebri and decreasing visual acuity and contraction of her visual fields underwent LPS placement with microlaparoscopic-assisted approach for placement of the peritoneal end of the LPS and reported resolution of symptoms before hospital discharge with

normalization of her visual acuity and fields at 3-month follow-up.

Twelve patients (presented initially with mild defect and were maintained on medical treatment and follow-up) developed deterioration of visual field and underwent shunting after a mean duration of 6.5±1.8; range: 3-12 weeks. Post-operative follow-up visual field examination revealed variable degrees of improvement in all cases. This goes in hand with Rizzo et al.²¹ who retrospectively reviewed cases of IIH with CSF shunting and found CSF shunting resulted in significant improvement in the perimetric results with significant increase in the MD and significant decrease of average retinal nerve fiber layer thickness measured by optical coherence tomography and Frisen papilledema grade.

Dogan et al.²² presented a patient with Behçet's disease who developed bilateral papilledema and left abducens nerve palsy, but despite acetazolamide, prednisone, azathioprine and repeated lumbar punctures, his signs and symptoms of intracranial hypertension gradually worsened, so LPS was planned and rapid resolution of intracranial hypertension was observed.

In support of the favorable outcome of LPS procedure for management of IIH, Feldon²³ compared surgical techniques for management of visual loss in IIH unresponsive to medical treatment and reported improved or resolved vision deficit in 38.7% of patients after ventriculo-peritoneal shunt (VPS) placement, 47% of patients after stent placement, 44.6% of patients after LPS placement, and 80% of eyes after optic nerve sheath fenestration (ONSD) and documented that visual worsening was rare for all procedures evaluated. Toma et al.²⁴ retrospectively reviewed the clinical records of 18 patients who underwent LPS insertion and found that 14 patients with preoperative headache did not complain of headache postoperatively, and 4 had headache that was found not to be related to shunt function, two of the patients with preoperative visual complaints had ongoing visual problems postoperatively and thirteen patients (65%) did not require shunt revision and concluded that LPS is effective in treating pseudotumor cerebri.

Abubaker et al.²⁵ compared the outcome of IIH treatment by VPS to LPS and found that both shunts are effective in controlling all the clinical manifestations of IIH in the immediate postoperative period, with non-significantly higher failure rate with VPS and revision rate with LPS. El-Saadany et al.¹⁸ studied 22 patients showed failure or noncompliance to medical treatment and underwent LPS for IIH and reported recovery of headache in 19(86.4%) patients and complete resolution of papilledema in 16(72.7%) patients and concluded that LPS is easy and effective for treating intractable headaches and visual impairment associated with IIH.

Lai et al.²⁶ conducted a systematic analysis of case series to compare therapeutic efficacies, concerning

visual outcome and headache of surgical interventions for IIH management including optic nerve sheath fenestration, lumbo-peritoneal shunt, ventriculoperitoneal shunt and dural venous sinus stenting and documented that the pooled analysis indicated an overall similar improvement in visual outcomes and headache across treatment modalities with insufficient evidence to recommend or reject any treatment modalities for IIH.

In this series, we didn't consider headache indication for surgery and agree with Friedman & Rausch²⁷ who stated that: If the vision is good, treat headaches medically as LPS just creates a bigger headache for the patient, and the neurosurgeon.

In this series, two cases required revision and one case developed temporary overdrainage, that is not far from the results of McGirt et al²⁸ who found that LPS were 2.5 times more likely to become obstructed than VP and VA shunts. Their reported complications are catheter migration, overdrainage and shunt infection.

CONCLUSION

It could be concluded that this therapeutic policy of IIH management with depending on visual field changes as indication for surgery provided acceptable visual outcome. Maintaining follow-up of these patients together with inclusion of more cases is warranted for more precise evaluation.

Declaration

The author(s) declare no conflict of interest or any financial support and confirm the approval of the submitted article by the concerned ethical committee.

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