http://jmscr.igmpublication.org/home/ ISSN (e)-2347-176x ISSN (p) 2455-0450 crossref DOI: https://dx.doi.org/10.18535/jmscr/v8i3.79



Journal Of Medical Science And Clinical Research An Official Publication Of IGM Publication

<u>Research Article</u> Study of Ophthalmic Profile and Prognosis in patients of Idiopathic Intracranial Hypertension

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Abstract

Aim: To study the ophthalmic profile of Idiopathic intracranial hypertension and correlation between ophthalmic profile with demographic, clinical and investigation profile as available from department of Medicine /Neurology.

Material and Methods: This study was a Prospective and Retrospective observational study conducted in 30 cases in whom diagnosis of IIH has been made according to Dandys criteria in Upgraded Department of Ophthalmology in MGMMC & MY Hospital during March 2018 to March 2019.

Results: Most (24/30, 80%) of the patients were females and the mean age of presentation was 31years. Chief complaints were headache in all 30 (100%) patients, 17 (56.6%) patients had blurring of vision, 1 (3.3%) patient had diplopia. There is no relation between high BMI and occurrence of IIH as in this study 90% patients has normal BMI. Papilloedema was seen in all 30 (100%) cases, 13(43.5%) patients had enlarged blind spot as the common visual field defect. 93.3% of eyes improved vision between 6/12-6/6, 3.33% of eyes has vision between 6/24-6/12 and visual acuity of rest 3.33% of eyes were remain <6/60 even after treatment. Majority of patients (43.3%) presented with GRADE 1 papilloedema followed by GRADE 2 in 33.3%, GRADE 3 in 16.6% and GRADE 4 in 6.6% patients. Papilloedema resolved (GRADE 0) in 90% of patients following treatment. 1 Patient had grade 1 papilloedema and 2 patients had deterioration with GRADE 4 Papilloedema after treatment. 7 patients (23%) became asymptomatic, including normalized vision, at 7th day of follow-up and after 3 months 28 patient (93%) improved, except 2 patient which showed visual deterioration even after 3 month of complete treatment.

Conclusions: *IIH* represents an increasing burden to public health in our country. In this study it is found that medical treatment in early stage (relatively good visual acuity, early disc edema, and normal / minimal field defect) has prevented them from ending up with irreversible vision loss and optic atrophy. A strong suspicion of idiopathic intracranial hypertension in chronic severe headache and immediate investigation followed by proper treatment can salvage vision of patient even in case of established papilloedema. The goal of this study is to delineate the clinical course of this disorder with an emphasis on visual prognosis. **Keywords:** *Idiopathic intracranial hypertension, Papilloedema, pseudotumorcerebri.*

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Introduction

Idiopathic intracranial hypertension, also known pseudotumorcerebri (PTC) or benign as Intracranial Hypertension (BIH) is a neuroophthalmic disease of unknown etiology¹. With an annual incidence of 0.9/100000, mainly in females of age group 15-44 years, IIH has been a long standing challenge for clinicians because of unknown pathophysiology. It is characterized by elevated intracranial pressure in the absence of underlying conditions, typically resulting in bilateral optic nerve head edema. If left untreated, IIH can result in irreversible loss of vision. The majority of patients presenting with IIH have symptoms that include a headache. Other signs and symptoms may include transient visual obscurations, visual blurring, horizontal diplopia, nausea, vomiting, vision loss, impaired visual fields. papilloedema. Investigation and management depends on symptoms and signs and requires an interdisciplinary team approach. A strong suspicion of idiopathic intracranial hypertension in chronic severe headache and immediate investigation following proper treatment can salvage vision of patient even in case of established papilloedema. The goal of this study is to delineate the clinical course of this disorder with an emphasis on visual prognosis.

Material and Methods

This study was a Prospective and Retrospective observational study conducted in Upgraded Department of Ophthalmology in MGMMC & MY Hospital, Indore during March 2018 to March 2019. The study was approved by Institutional Review board and written informed consent was obtained from all subjects prior to participation. Some of the consents were in local language to ensure validity. Patients were free to withdraw from study at any time and were assured that the study would not compromise the quality of their eye care.

• **Sample Size:** 30 cases in whom diagnosis of IIH has been made according to Dandys criteria.

Dandys Criteria

- 1) Symptoms of raised intracranial pressure (headache, nausea, vomiting, transient visual obscurations) or Papilledema.
- 2) No localizing signs with the exception of abducens (sixth) nerve palsy.
- 3) The patient is awake and alert.
- 4) Normal CT/MRI findings without evidence of thrombosis.
- 5) LP opening pressure of >25 cm of H2O and normal biochemical and cytological composition of CSF.
- 6) No other explanation for the raised intracranial pressure.

Inclusion Criteria

• All patients between 20- 40 years of age presenting with headache to Ophthalmology OPD and Neurology OPD as clinically suspected cases of IIH.

Exclusion Criteria

- Other causes of Intracranial hypertension
- Abnormal MRI brain as per imaging protocol of department of Radiology.
- Any contraindication for MRI/ CSF Tap.

All the patients who present to the Department of ophthalmology and neurology, who are suspected to have IIH will be evaluated as follows: A detailed history was taken, including demographic features of age and gender, date of presentation, symptoms of headache, presenting nausea, vomiting, neurological symptoms, and ocular symptoms like transient visual loss, loss of central vision, visual field defects, and double vision. A detailed ophthalmic examination was performed, including snellen's visual acuity, refraction, ocular motility examination, pupil evaluation, color vision, anterior segment examination, intraocular pressure measurement, optic disc examination Dbiomicroscopy, with and fundus +90examination with binocular indirect ophthalmoscopy and +20 D lens. Humphrey's 30-2 visual field analysis, neuroimaging (MRI scan with MR venography), and lumbar puncture: cerebrospinal fluid (CSF) analysis and pressure recording was done for all patients.

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After all the investigations all the patients managed according to their symptoms and vision status. Patient with normal vision and minimal papilloedema treated with medical management and monitored frequently for the development of symptoms or vision loss. All patients were treated medically with oral acetazolamide at a dose of 250mg QID/day. The dosage was adjusted depending on the response of patients. Visual acuity, optic disc changes, and visual field defects were checked in all the patients during follow-up, which was done at 7 th day and at 3 months. All these data was recorded meticulously in proforma and in master chart for further systematic tabulation for observations and analysis. Summary and conclusions was drawn after discussion with review of literature.

Observations & Results

The present study "Ophthalmic profile and prognosis in patients of Idiopathic Intracranial Hypertension" was carried out in 30 cases who presented to the Department of ophthalmology and neurology at MGM Medical College and MY hospital, Indore during March 2018 to March 2019. All the data was recorded meticulously in proforma and in master chart for further systematic tabulation for observations and analysis. Summary and conclusions was drawn after discussion with review of literature.

The study sample consisted of patients with the minimum age of 20 years and maximum age of 40 years. 56% patients fall within the range of 30-35 years with the mean age of 31.5 years (Table-1). The number of female patients was more than male patients that is 80% of sample size. There is no relation between high BMI and occurrence of IIH as in this study 90% patients has normal BMI (Table-2). Headache and papilloedema are the most common presenting features present in all the 30(100 %) patients, blurring of vision present in 17(56.6%) patients and, diplopia in only 1(3.3%) patient (Table-3).

93.3% of eyes improved vision between 6/12-6/6, 3.33% of eyes has vision between 6/24-6/12 and

visual acuity of rest 3.33% of eyes remain<6/60 even after treatment (Table-4,5).Most common visual field defect in IIH was Enlarged Blind Spot seen in 13 (43.3%) patient (20 out of 60 eyes).

Majority of patients (43.3%) presented with GRADE 1 papilloedema followed by GRADE 2 in 33.3%, GRADE 3 in 16.6% and GRADE 4 in 6.6% patients. Papilloedema resolved (GRADE 0) in 90% of patients following treatment. 1 Patient had grade 1 papilloedema and 2 patients had deterioration with GRADE 4 Papilloedema after treatment (Table-6,7).In 90% of patients CSF opening pressure was mild within the range of 25-30 cm of $H_2O(Table-8)$. Visual acuity, optic disc changes, and visual field defects were checked in all the patients during follow-up, which was done at 7th day and at 3 months. 7 patients (23%) became asymptomatic, including normalized vision, at 7thday of follow-up and after 3 months 28 patients (93%) improved, except 2 patient which showed visual deterioration even after 3 month of complete treatment (Table-9).

 Table 1 Age Distribution

S.No.	AGE (In years)	NO.OF PATIENTS	PERCENTAGE (%)
1.	20-25	3	10
2.	25-30	9	30
3.	30-35	17	56.6
4.	35-40	1	3.3

Table 2 Body mass index (WHO InternationalClassification) distribution

S.No	BMI	No .Of Patients	PERCENTAGE (%)
1	NORMAL [<25]	27	90
2	OVERWEIGHT	2	6.6
	[25-29.9]		
3	OBESE [>30]	1	3.3

Table 3 Signs and Symptoms

S.No	SYMPTOMS	NO.OF	PERCENTAGE
		PATIENTS	[%]
1.	Headache	30	100
2.	Blurring of vision	17	56.6
3.	Diplopia	1	3.3
4.	Lateral rectus	0	0
	palsy		
5.	Papilloedema	30	100
6.	Enlarged blind	13	43.3
	spot		

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S.No.	VISION	NO.OF EYES	PERCENTAGE (%)
1	<6/60	2	3.33
2	6/60-6/24	3	5
3	6/24-6/12	15	25
4	6/12-6/6	40	66.6

Table 4 Pretreatment visual aquity

 Table 5 Post treatment visual acuity

S.No.	VISION	NO.OF EYES	PERCENTAGE (%)
1	<6/60	2	3.33
2	6/60-6/24	2	3.33
3	6/24-6/12	0	0
4	6/12-6/6	56	93.3

Table 6 Papilloedema at the time of Presentation

S.No	GRADING	NO.OF PATIENTS	PERCENTAGE (%)
1	GRADE 1	13	43.3
2	GRADE 2	10	33.3
3	GRADE 3	05	16.6
4	GRADE 4	02	6.6

 Table 7 Post treatment papilloedema

S.No.	GRADING	NO.OF PATIENTS	PERCENTAGE (%)
1	GRADE 0	27	90
2	GRADE 1	1	3.33
3	GRADE 2	0	0
4	GRADE 3	0	0
5	GRADE 4	2	6.66

Table 8 CSF Opening Pressure

The CSF pressure classification was based on El-Saadany et al study. The pressure was categorized into mild (25-30), moderate (30-40), and severe (>40)

S.No.	CSF OPENING PRESSURE(cm H ₂ O)	NO.OF PATIENTS	PERCENTAGE (%)
1	25-30	27	90
2	30-40	1	3
3	>40	2	6.66

Table 9 Follow Up

Followu P	Number of patients with improve d vision	Percentag e (%) of patients with improved vision	Number of patients with no improveme nt	Percentage (%)of patients with no improveme nt
7 th day	7	23.3%	23	76%
3 month	28	93%	2	6.66 %

Discussion

The present study "Ophthalmic profile and prognosis in patients of Idiopathic Intracranial Hypertension" was carried out in 30 cases who presented to the Department of ophthalmology and neurology at MY hospital and MGM Medical College Indore during March 2018 to March 2019. The study included total 30 cases with the minimum age of 20 years and maximum age of 40 years. 56% patients fall within the range of 30-35 years with the mean age of 31.5 years. A study by **Daniels** *et al*² (2007) reported the mean age of presentation as 32 years .Similar study done by Ambika S et al.³(2010) reported the mean age of presention 31.3 year. Fraser C et al⁴ in (2011) reported the age of presentation in IIH was between 29 and 30 years. In this study, the number of female patients (24) was more than male patients(6) that is 80% of sample size(30), hence shows a female preponderance. Radhakrishnan K et al⁵ (1993) conducted a study and reported 93.8% females and 6.17% male study sample. Raoof N, Sharrack B et al⁶ (2011) reported 93.7% females and 6.25% males in his study sample. A study done by John chen and Michael Wall⁷ (2014) had quoted that female gender is a risk factor for IIH since almost 90% of the affected population were females. Above studies shows female preponderance in IIH .A possible explanation for this predilection of IIH for women in their childbearing years supports a potential role of hormonal influences in the development of this disorder. This study shows that there is no relation between high BMI and occurrence of IIH as in this study 90% patients has normal BMI. **B.B.Bruce** et al⁸(2010) identified 4% IIH patients with normal BMI.

Headache is the most frequently reported symptom in IIH and was the presenting complaint of all the subjects in our study followed by reduced visual acuity, blurring of vision and diplopia. In Wall M study⁸⁶ (1990) 92.0% patient has headache as a chief complaint in IIH. George D et al⁹ (1991) reported headache in 92% IIH patients. **Binder** et al¹⁰.(2004) reported 90%

headache patient in IIH. **Friedman DI et al**¹¹ (2017) also reported 84% headache patient in IIH population. In present study headache is improved in all the patient with medical management. Vision loss is the main long term complication of IIH.patient with mild to moderate vision loss tend to recover vision following medical therapy.

In this study totally, 44 out of 60 eyes had a visual acuity in the range of 6/12-6/6 (66.6%), 15(25%) patients had visual acuity in the range of 6/24-6/12, 3(5%) had vision between 6/60-6/24 and only 2 (3.33%) had visual acuity <6/60. All patients are improved during the course of treatment except for 2 patients ,1 whose vision decreased from 2/60 to 1/60 right eye and 1 whose right eye vision showed only 1 line improvement which was associated with worsening of edema. This patients was initially started on oral acetazolamide 250 mg QID/ day. However, the showed visual deterioration despite patients medical therapy and were advised for ventriculoperitoneal (VP) shunt by neurologist but the patient was not willing for surgery and was lost to follow-up. Rowe FJ, Sarkies NJ. et al ¹²1998 noted Visual loss at presentation and during follow-up in up to 87% of patient. Corbett JJ, et al.¹³ (1982) reported 24.6% patients with vision loss in IIH.

The most common visual field defect was an enlarged blind spot which was noted in 43.3% patients or 33.3% eyes. With treatment there is perimetric improvement in all the pateints except 2 patients which undergo progressive deterioration. Raju K V et al¹⁴ 2009. reported 56% of the patient in their study showed field defect with enlargement of blind spot. Thurtell MJ et al Wall M^{15} (2013) reported enlarged blind spots as early visual losses. In this study all of the cases had papilloedema and of these (43.3%) presented with GRADE 1 papilloedema followed by GRADE 2 in 33.3%, GRADE 3 in 16.6% and GRADE 4 in 6.6% patients.

Untreated papilloedema can lead to progressive irreversible visual loss. Patients have more visual loss in their eyes with high-grade papilloedema than in those eyes with low-grade papilloedema range may well have visual loss; however, their values have not yet reached the abnormal range.

The CSF pressure classification was based on El-Saadany et al¹⁶ study. The pressure was categorized into mild (25-30) cm of H₂O, moderate (30-40) cm of H_2O , and severe (>40) cm of H₂O It is has shown in our study that in 90% of patients CSF opening pressure was mild within the range of 25-30 as majority of cases presented with mild to moderate disc edema. Visual acuity, optic disc changes, and visual field defects were checked in all the patients during follow-up, which was done at 7th day and at 3 months. 7 patients (23%) became asymptomatic, including normalized vision, at 7thday of follow-up and after 3 months 28 patients (93%) improved, except 2 patient which showed visual deterioration even after 3 month of complete treatment.

Conclusions

IIH represents an increasing burden to public health in our country. It is a disease primarily affecting women of child bearing age. High BMI may not be a dominant risk factor for the development of IIH. The most common presenting symptom is headache followed by blurring of vision, diplopia, Neurodeficits. MRI can be absolutely normal in these cases or may show findings such as empty sella. CSF pressure recording is mandatory in these patients and only early treatment can help salvaging the vision. Increased opening pressure on lumbar puncture is proportional to the grades of papilloedema seen on fundus examination and thus to worse visual prognosis. In this study it is found that medical treatment in early stage (relatively good visual acuity, early disc edema, and normal/minimal field defect) has prevented them from ending up with irreversible vision loss and optic atrophy. Further studies with larger sample size are required to determine the role of other factors responsible for its pathogenesis. These patients tend to have a good response to medical

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management alone, without lasting visual loss, needing surgical intervention.

A strong suspicion of idiopathic intracranial hypertension in chronic severe headache and immediate investigation following proper treatment can salvage vision of patient even in case of established papilloedema. The goal of this study is to delineate the clinical course of this disorder with an emphasis on visual prognosis.

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