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ORIGINAL ARTICLE

Short-term Prognosis of Vision in Idiopathic Intracranial Hypertension

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ABSTRACT

Background: Idiopathic intracranial hypertension (IIH) is a condition where intracranial pressure is raised in the absence of both abnormal neuroimaging and abnormal cerebrospinal fluid composition. We evaluate the clinical and ophthalmological parameters that could influence short term poor outcome of vision in IIH patients. **Methods**:A total of 18 IIH patients admitted to the department of Neurology, Zagazig University were included in this prospective cohort study. Complete clinical evaluation and ophthalmological assessment were done and included visual acuity, papilledema grading, field of vision, contrast sensitivity and color vision. Short term visual outcome was assessed after 3 months guided by visual field results and was correlated with clinical and ophthalmological parameters. **Results:** Visual acuity impairment from the onset was significant among patients with higher degrees of headache (p = 0.008), patients with impaired contrast sensitivity (p = 0.027 and 0.003 for RTand LT eye respectively) and patients with higher levels of serum cholesterol (p = 0.006). Short term visual outcome was assessed after 3 months and good prognosis was significantly correlated with young age (p = 0.004) and shunt operation (p = 0.038).

Conclusion: Young age and early intervention by lumboperitoneal shunt were significant factors for good outcome of vision in IIH. Doctors should recognize that in order to preserve vision and lower morbidity.

Keywords: Idiopathic intracranial hypertension, Visual acuity, Papilledema, Contrast sensitivity, Shunt.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a disease of high intracranial pressure in absence of a space occupying lesion, ventriculomegaly or abnormal cerebrospinal fluid (CSF) analysis **[1].** It is more prevalent among overweight females in their childbearing period with female: male ratio of 5:1, in whom the incidence reaches up to 3.5 per 100,000 compare to 0.9 only per 100,000 general populations **[2].**

The IIH patients mainly complain of pain in head that is worsened on the mornings hours and is characteristic of raised intracranial tension **[3].** Edema of the optic disc, known as papilledema, is an important sign on examination **[4].** Patients with high grade papilledema combined with diminished visual acuity at presentation are more likely to suffer failure of treatment **[5].** The main goals of management of patients with IIH are to protect vision in addition to minimizing the headache morbidity **[6].** Objective clinical and neuro-ophthalmologic factors may help early detection of patients requiring aggressive medical or surgical intervention **[5].** This study attempts to determine the short visual outcome of IIH with respect to various clinical and neuro-ophthalmological parameters.

METHODS

This prospective cohort study was conducted in neurology department and outpatient clinics of Zagazig University Hospitals during

the period March 2022 to February 2023. Eighteen patients ranging from 19 to 42 years old were included. The patients were diagnosed with IIH according to the revised criteria described by Friedman and colleagues, 2013 **[7].** A definite diagnosis of IIH was given if the patients had papilledema, normal neurologic examination except for occasional isolated sixth nerve palsy, normal brain structure with no evidence of hydrocephalus, space occupying lesion on magnetic resonance imaging and venography (MRI and MRV) along with normal CSF composition and high lumbar puncture opening pressure of more than 250 mm with a properly performed lumbar puncture **[7].** Exclusion criteria including patients with abnormal neuroimaging, patients with endocrine disturbance, patients on steroid therapy or other drugs known to cause elevated intracranial pressure.

Full history was taken from all subjects with focus on demographics (gender, age, body weight) and clinical data including visual symptoms (e.g., diplopia and blurring of vision). The severity of headache was graded using a self-administered questionnaire into mild, moderate and severe **[8].** Drug history of hormonal contraception was obtained. All patients underwent routine lab work-up including complete lipid profile. The presence and grade of papilledema were assessed through fundus examination using modified Frisén scale which scores the signs on optic disc and surrounding retina to grade the edema on a scale from 0 to 5. The staging is zero (normal fundus). Grade 1 means excess blurred nasal border in addition to subtle gray halo with temporal gap at the disc, grade 2 means blurring of all temporal margin, halo surrounds disc, grade 3 consistent with increase diameter of optic nerve head, obfuscating one or more major vessels, grade 4 is elevation of whole nerve head, grade 5 means dome-like protrusion of optic nerve head **[9].**

Visual acuity was assessed with Snellen chart **[10].** Snellen's test measures only perception of sharp and clear outlines of very small objects, not the changes in illumination **[11].** The affection of vision cannot be evaluated properly using only visual acuity **[12].** Visuospatial ability would be determined using contrast sensitivity (CS) which assess the changes in lighting perception between a target and its background **[13].**

Contrast sensitivity was assessed using Pelli Robson contrast sensitivity sheet**.** A score of 2.0 is considered normal, score of less than 1.5 represents mild visual impairment and less than 1.0 represents indicates severe visual impairment or disability **[14].** Visual field testing was done by the Humphrey (Humphrey Instruments, San Leandro, CA) **[15].** All our patients received medical therapy in the form of acetazolamide and did lumbar puncture to assess CSF opening pressure. At the same set, spinal tapping of CSF was done as a part of treatment to decrease intracranial pressure.

Patients were assessed after 3 months with regards to improvement of headache severity along with visual parameters of visual acuity, field of vision, contrast sensitivity and papilledema grading on fundus examination. Final visual outcome was defined according to results obtained from the worst eye on follow up after 3 months and was compared with the clinical and ophthalmological parameters in those patients. The degree of visual field loss represented the visual outcome, graded on a scale from zero to 2 point. Zero is normal or enlarged spot; 1 represents scattered central or paracentral scotoma and 2 denotes generalized field constriction.

Because blind spot enlargement is widely common, and additional plus lenses can improve this defect, we did not consider it a visual loss unless it interfered with fixation **[16].** Severe visual field contraction and scotomas are other common changes **[17].** Based on this, we considered Zero grade as good visual outcome, while 1,2 grades represented poor visual outcome.

Ethics approval and constant to participate

The study was approved from the Institutional Ethics of the faculty of medicine. Zagazig University (ZU-IRB #9475- 6/4-2022). Written informed consent was obtained from all the participants after explaining the details and benefits as well as risks to them.

Statistical analysis: Categorical variables were tabled as frequencies and percentages and statistically tested with Chi-squared or Fisher's exact test. Means and standard

deviations expressed continuous variables and tested using independent sample T test or Mann-Whitney test. P value less than 0.05 was the cut off for statistical significance.

RESULTS

We included 18 patients of IIH. The median age of our patients was 28.5 (IQR 19–42 years). 83.3 % were females.

Our study showed that 10 (55.6%) of our patients had impaired vision from the onset. Asymmetrical papilledema was detected in 5 (27.8%). All patients had intact color vision but contrast sensitivity was affected on testing both eyes. 9 (50%) of participants had visual field abnormalities **(table 1).** The results showed that visual acuity impairment from the onset of disease was significant among patients with higher degrees of headache (p = 0.008) and patients with impaired contrast sensitivity (p = 0.027 and 0.003 for right and left eye respectively). Higher cholesterol level was significant among patients with poor vision (p = 0.006). Although mean CSF opening pressure was higher in patients with poor visual acuity (40.8±13.8), the difference was not statistically significant, p = 0.09 **(table 2).**

Good visual outcome was determined after 3 months of onset guided by results of visual field examination. Good outcome was significant among younger age group (p = 0.004). Patients treated by shunt operation showed statistically significant good outcome (p = 0.038). However, other clinical and ophthalmological parameters were not significantly correlated with outcome **(table 3, 4).**

Table (1): Demographic, clinical and ophthalmological data of the studied patients

Patients (n=18)	Median (IQR) or n (%)
Age (years):	28.5 (19-42)
Females	15 (83.3%)
BMI (kg/m^2), (25-29.9)	8 (44.4%)
BMI (kg/m^2), (30 or above)	10 (55.6%)
History of contraceptive pills in females	9 (60%)
CSF opening pressure (cm H2O)	31 (25-60)
Headache	16 (89.9%)
Blurring of vision	15 (83.3%)
Diplopia	2 (11.1%)
Visual acuity impairment worst eye	10 (55.6)
Asymmetrical papilledema	5 (27.8)
Visual field affection	9 (50%)
Contrast sensitivity affection worst eye	8 (44.4)

CSF: Cerebrospinal fluid ,BMI: Body mass index

Table (2): Comparison between degree of visual acuity at onset of disease (determined by visual acuity in worst eye) and demographic, clinical, laboratory and ophthalmological data of the patients.

Variables		Initial visual acuity		Test of significance	P Value
		Normal n.8	Impaired n.10		
Age per years	Mean ±SD	33.6±5.9	28.1±5.5	1.8	0.09
Sex	Female	5 (62.5%)	10 (100.0%)	F	0.069
	Male	3 (37.5%)	0 (0.0%)		
BMI	Mean ±SD	28.4±3.6	34.9±4.9	2	0.058
Headache severity	Severe	0 (0.0%)	5 (50.0%)	11.9	**0.008***
	Mild	6 (75.0%)	2 (20.0%)		
	Moderate	0 (0.0%)	3 (30.0%)		
	No	2 (25.0%)	0 (0.0%)		
Papilledema	I	1 (12.5%)	0 (0.0%)	7.5	0.058
	II	3 (37.5%)	0 (0.0%)		
	III	2 (25.0%)	2 (20.0%)		
	IV	2 (25.0%)	8 (80.0%)		
Right contrast	Normal	8 (100%)	4 (40%)	7.2	**0.027***
	Impaired	0 (0%)	2 (20%)		
	Disable	0 (0%)	4 (40%)		
Left contrast	Normal	8 (100%)	2 (20%)	11.5	**0.003***
	Impaired	0 (0%)	5 (50%)		
	Disable	0 (0%)	3 (30%)		
CSF open pressure	Mean ±SD	31.5±4.3	40.8±13.8	1.8	0.09
Total cholesterol	Mean ±SD	169±.31.6	182±27.2	3.1	**0.006***
Triglyceride	Mean ±SD	101.2±25.8	114.9±54.9	0.65	0.53
HDL	Mean ±SD	46.6±7.6	48.2±6.7	0.46	0.65
LDL	Mean ±SD	109.4±28.9	103.5±7.8	0.53	0.61

f=Fisher Exact test ,* Significant ,CSF: Cerebrospinal fluid , BMI: Body mass index ,HDL: High density lipoprotein ,LDL: Low density lipoprotein

Table (3): Relation between visual field outcome and demographic, clinical data, shunt operation in idiopathic intracranial hypertension.

Variables		Visual field at follow up		Test of significance	P Value
		Good outcome n.11	Poor outcome n.7		
Age per years	Mean ±SD	27.4±4.2	35.4±5.9	3.5	**0.004***
Sex	Female	9 (81.8%)	6 (85.7%)	F	0.99
	Male	2 (18.2%)	1 (14.3%)		
Body mass index	Overweight	5 (45.5%)	3 (42.9%)	F	0.99
	Obese	6 (54.5%)	4 (57.1%)		
CSF open pressure	Mean ±SD	36.3±11.2	37.3±12.2	0.18	0.86
Cholesterol	Mean ±SD	176.5±31.9	175.9±26.6	0.04	0.97
Triglyceride	Median (range)	100(56.9-140.6)	102(65-259)	0.74	0.47

HDL	Mean ±SD	47.2±7.04	47.9±7.4	0.92	0.83
LDL	Mean ±SD	114.5±21.5	93±22.8	1.94	0.053
Medical treatment+ lumbar puncture	Yes	11 (100.0)	7 (100.0)	-	-
Shunt	Yes	6 (54.5%)	0 (0.0%)	F	**0.038***
	No	5 (45.5%)	7 (100.0%)		

f=Fisher Exact test ,* Significant ,CSF: Cerebrospinal fluid ,HDL: High density lipoprotein ,LDL: Low density lipoprotein

Table (4): The association between headache severity, visual parameters and short term outcome in IIH patients.

Variables		**Visual field at follow up**		**Test of significance**	**P value**
		Good outcome n.11	**Poor outcome n.7**		
Headache severity	Severe	5 (45.5%)	0 (0.0%)	5.2	.16
	Mild	3 (27.3%)	5 (71.4%)		
	Moderate	2 (18.2%)	1 (14.3%)		
	No	1 (9.1%)	1 (14.3%)		
Right contrast	Normal	6 (54.5%)	6 (85.7)	2.2	0.33
	Impaired	2 (18.2%)	0 (0.0%)		
	Disable	3 (27.3%)	1 (14.3%)		
Left contrast	Normal	6 (54.5%)	4 (57.1%)	0.047	0.98
	Impaired	3 (27.3%)	2 (28.6%)		
	Disable	2 (18.2%)	1 (14.3%)		
Visual acuity worst eye	6/6	4 (36.4%)	4 (57.1%)	4.7	0.36
	6/9	2 (18.2%)	0 (0.0%)		
	6/12	3 (27.3%)	0 (0.0%)		
	6/18	1 (9.1%)	2 (28.6%)		
Papilledema grade	I	1 (9.1%)	0 (0.0%)	0.88	0.83
	II	2 (18.2%)	1 (14.3%)		
	III	2 (18.2%)	2 (28.6%)		
	IV	6 (54.5%)	4 (57.1%)		

f: Fisher exact test

DISCUSSION

We aimed to assess short-term visual outcome in patients with IIH. Most patients were females (83.3%), young with age between 19 to 42 years and mean of 30.6±6.2 years, which agrees with previous studies **[18-20].** Although IIH has a higher predilection in women, yet the link between IIH and female sex remains uncertain with many suggested explanations. For instance,

gender-specific hormones were supposed to have a key role in the pathophysiology and development of IIH **[21].** Men are not likely to have IIH; a recent large series found that the occurrence of IIH in men was only about 10% **[22].** It was also observed that IIH is also strongly associated with obesity. In our study, all of our patients had high body mass index (BMI) with 55.6% were obese (BMI 30 or over).

A multicenter, case-control research comparing recently diagnosed IIH patients to those with other neuro-ophthalmologic illnesses established that a higher BMI is linked to a higher chance of developing IIH **[23].** A recent moderate increase in weight was linked to an increased risk of IIH **[24].**

In our study, we reported that 60% of our female group of patients were on oral contraceptive pills (CPs). On the other hand, a large retrospective study which recruited patients over 16-year duration found that CPs were not correlated with risk of IIH, refuting the need for women with IIH to abort them **[25].** In a Saudi study, 3 (6.6%) of the females were on oral CPs which is a low number **[26].**

Symptoms we recorded were comparable to other studies. And like which, headache was the most common encountered presentation in our patients (84%) **[5,27]**.

In the current study, severe visual impairment was found in 11.2% of our patients. Symmetrical papilledema was found in 72.2%. Our study showed that patients with visual acuity impairment from the onset of disease was significant among patients with higher degrees of headache and patients with impaired contrast sensitivity. Higher cholesterol level was significant among patients with poor vision at $p = 0.006$. This observation might be explained by the fact that obesity and dyslipidemia contributes to the occurrence of metabolic syndrome **[28].** Future work to investigate the role of lipids and its metabolites on elevated intracerebral pressure would be of interest.

The optic neuropathy and optic disc edema are likely the cause of vision involvement in IIH patients. The latter's pattern is typically bilateral and symmetrical, while it occasionally exhibits asymmetrical or unilateral presentation **[29].** In a study of 559 patients with IIH, 20 (3.6%) had very asymmetric papilledema at initial evaluation which was defined as a ≥2 grade difference in modified Frisén between the 2 eyes **[30].**

The translaminar pressure gradient at the head of the optic nerve is critical to the pathophysiology of papilledema. Axonal flow stasis and microscopic alterations are identical when peri-optic CSF pressure is high or low **[31].**

Assessing only visual acuity by Snellen chart provides incomplete view of visual abilities **[12].** The visual targets in space is evaluated using contrast sensitivity (CS); which is known by the ability to distinguish the degree of whiteness to blackness of a specific target in addition to perception of sharp demarcation of extremely small objects. CS plays an important role as an early detector of impaired visual function in cataract, elevated intraocular pressure and many retinal problems **[32].**

The Pelli Robson chart was used for CS evaluation and in our study, it was significantly associated with visual acuity impairment in our patients of .027 and .003 significances in right and left eye respectively. The initial assessment of CS in IIH was done by Wall M in 1986, who used six Arden grating plates to examine 12 patients. In 9/12 patients (75%) and 13/24 eyes (54%), CS loss was found. It was observed that when the papilledema subsided, CS got better. The authors came to the conclusion that CS was helpful for serial follow-up of IIH patients and for detecting visual loss **[33].** Similar to our results, Argwal and Tidakereported a similar link between deterioration in visual acuity and decrease of contrast sensitivity **[34].** CS was the only visual parameter that was significantly associated with the symptom of persistent visual loss in another study **[12].**

We use the visual field as a parameter to assess outcome of vision since such defects typically develop in a steady pattern as a result of the initial impairment of peripheral vision, while the central field might be spared for longer duration and late stages leaving the patient oblivious of his condition and resulting progressive complications **[35].**

Enlargement of physiological blind spot is the

earliest visual field defect to appear, which is a refractive scotoma brought on by the elevation of the peripapillary retina **[36]** followed by infernasal scotomas and late field constriction. Neurosensory detachments and choroidal folds are two retinal causes of vision loss **[17].** However, affection of vision in IIH is due to elevated pressure of CSF causing flow stagnation and subsequently intra-axonal swelling; and compression, resulting in ischemic damage **[37].**

Visual affection with field loss is the most dreaded consequence, mainly determining the therapy and outcome of the syndrome. Whether those parameters could predict final visual outcome in IIH patients is controversial. Surprisingly, in our study, these parameters did not show significant relation with final visual outcome. This lack of significance may be attributed to different definition of vision outcome in each study. Also, presence of confounding factor which is early surgical intervention in patients showing impending visual loss along with short follow up period. In addition, it is essential to note that there is inter-individual variation of papilledema grading based on the ophthalmologist assessment **[٣8].** The optic disc appearance is a key parameter of disease status in IIH. The Frisén classification grades optic disc swelling (grades 0-5). It is the international known classification used in both clinical settings and research purpose **[9].**

The good visual outcome in our work was statistically correlated with younger age and those underwent shunt operation. This finding is passing with that of Argwal and Tidake**[34]** where pediatric IIH had a favorable visual outcome in terms of visual field and visual acuity, while one study had founded puberty to be a risk factor for an unfavorable outcome **[٣9].** On the other hand, some studies suggested that the visual outcome may be better in pediatrics than adults **[40, ٤1],** with spontaneous remission after diagnostic lumbar puncture being more common **[٤1].**

Bynke and colleagues **[٤2]** evaluated 17 IIH patients treated by shunt operation, and reported 18% improvement in visual acuity and a 65% improvement in visual field defects. However, in disagreement, another study **[٤3]** revealed no relation between CSF shunting and visual field outcome.

This study shows several obstacles; small sample size, the subjective grading of papilledema severity with a high degree of interobserver variability, the battery of neuro-ophthalmological tests used in the study may need additional exams like visual evoked potential, optical coherence topography (OCT) to assess retinal nerve fiber layer thickness (RNFL). Contrast sensitivity in IIH patients was evaluated using a chart and not an automated advanced machines due to unavailability of the machine in our hospital.

Recommendations: Further multicenter study with a larger sample and long-term follow up is recommended. Assessment of RNFL thickness using OCT is recommended.

CONCLUSION

In our study we determined that good vision outcome after 3 months of IIH diagnosis, was related to young age and early surgical intervention in patients.

Conflict of interest:

The authors declared that they have no conflicts of interest with respect to the authorship and/ or publication of this article.

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