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"A study on the clinical profile of Idiopathic Intracranial Hypertension (IIH) in a tertiary care centre in India"

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Idiopathic Intracranial Hypertension (IIH) or Pseudo tumour is a syndrome of increased intracranial pressure without any clear etiology and pathogenesis. If not recognised early and no treated, it may lead to permanent visual loss. There are few studies from India describing the clinical profile of this condition. Objectives : To study the clinical profile of idiopathic intracranial hypertension with emphasis on symptoms, signs, and radiological features. Study design : This is a retrospective study using the copies of discharge cards of patients with idiopathic intracranial hypertension admitted in neurology ward with IIH from August 2019 (01/08/2019 to July 2020(31/07/2020). They are enrolled based on definite Inclusion criteria: Modified dandy's criteria. Total sample size was 35 patients. ***Results :*** *Out of the 35 patients, 34 were females, majority of whom were obese. Median age of the patients was 33. Headache the most common symptom, present in all patients. It was followed by diminution of vision which was present in 15 patients, Two of them showed progressive visual loss and had to be referred for surgical procedures. Visual field defects included enlargement of the blind spot in 11 patients and peripheral constriction of field in 27 patients. Out of the 35 patients, 31 had bilateral symmetric Papilledema. CSF pressure was above 25 cm of water and patients with increased CSF of pressure had high grade of Papilledema.*

Key words: *IIH (Idiopathic Intracranial Hypertension), Pseudotumor cerebri, Papilledema, Headache, LP (Lumbar Puncture), CSF (Ceribro Spinal Fluid), ICP (Intracranial Pressure), IOP (Intraoccular Pressure), ONS (Optic Nerve Sheath)*

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I. Introduction

The old name for Idiopathic Intracranial Hypertension (IIH) was Pseudotumour cerebri or BIH (Benign Intracranial Hypertension). Idiopathic Intracranial Hypertension (IIH), also known as Pseudotumor, is a phenomenon characterised by elevated intracranial pressure with no known cause or pathophysiology. Over the decades the "not so benign nature" of the condition has been discovered. If not recognized early and left untreated, it may lead to permanent visual loss. Although global prevalence of IIH is increasing, there are few studies from India describing the clinical profile of this condition.[1]

A brief review

It has been a big challenge to Neurologists and Physicians all around the world to explain the etiology and pathophysiology of IIH and it is still unfortunate that our current knowledge about of this subject is limited. Previously IIH was known by names like Serus Meningitis, Pseudotumour cerebrai or benign intracranial hypertension. But with the recent advances we have become aware that it is not a benign condition and there are risks of visual loss and the chronic headache is disabling.[2]

The diagnostic criteria for IIH was first forwarded in1937 by Dandi and later modified by Smith in 1985.In 2013 Friedman et al proposed the term of pseudo tumour Cerebri Syndrome **(PTCS).** It was further classified into primary or secondary (PTCS) depending on the presence or absence of identifiable clauses. As a result IIH falls within the primary PTCS category.IIH is a disease defined by elevated CSF pressure normal CSF fluid content, a normal brain with normal or small ventricles in imaging studies, and normal results of neurological examinations except for abducent nerve palsy. Documentation of an elevated CSF pressure (>250mm in adults and >280mm in children) is mandatory for the diagnosis of definite PTCS. The diagnosis of probable PTCS may be given to patients with strongly suggestive clinical history, bilateral papal edema, supportive neuro imaging and abnormal CSF opening pressure.[3]

The incidence rate of IIH is about 0.5-2/1 lakh/year. IIH is becoming increasingly prevalent globally along with the epidemic of obesity. It is more present among obese young females. Although women are about 8 times more commonly affected than men, it is rarely found in men and children also. It is more common among

overweight people.

PTCS clinically presents with headache in 90 of patients and these are characterized by pressure like throbbing headaches. Visual loss is the most feared sequel of PTC and it may be transient in nature. Vision loss may take the form of impairment in visual field presenting us tunnel vision. Visual field defects are also common. Other symptoms include Pulsatile tinnitus photopia, eye pain and diplopia.

Raised intracranial pressure maybe due to known secondary causes like mass lesions, meningitis, venous sinusthrombosis.IIH is a syndrome of raised ICP without any identifiable brain pathology and with normal cerebrospinal fluid CSF composition. Hence it is a diagnosis of exclusion. Recognition and exclusion of this condition has crucial therapeutic and prognostic implication.[4] MRI has definite role in the diagnosis of IIH. Many imaging signs have been described in IIH. They include perioptic nerve sheath distension, vertical buckling of optic nerve, globe flattening, optic nerve head protrusion. MR venogram is very essential in excluding cortical venous thrombosis an important cause of secondary PTCS.[5,6]

Management of IIH includes administration of drugs like Mannitol, Acetazolamide, Lasix and repeated CSF drainage with lumbar puncture. If the vision is seriously compromised procedures like fenestration of optic nerve and surgical shunt procedures like ventriculo- peretonnial shunt, ventriculo-cavalshunt etc. may have to be performed.

IIH is one of the common neurological causes of visual loss which can be prevented if recognized earlier. Most often this entity is overlooked due to the uncommon presentation. So awareness of the typical signs and symptoms and high index of suspicion are important for salvaging the vision.

Objectives

1. To study the clinical profile of idiopathic intracranial hypertension with emphasison symptoms, signs, and radiological features.

II. Material Sand Methods

Study design - Cross sectional observational Record based
Study setting - Tertiary care centre in Kerala, India
Study participants

This is a retrospective study using the discharge cards in Neurology department of patients with IIH admitted in neurology ward from August 2018 (01/08/2018 to 31st July 2021(31/07/2021).

Inclusion criteria

1. Unilateral or bilateral papilledema
2. Normal neurological examination except for isolated abducens nerve palsy.
3. A wake and alert patients.
4. Normal brain MRI imaging/MRV/CT scan.
5. CSF pressure more than 250 mm of water with normal CSF fluid content.

Exclusion criteria

1. Other causes of intracranial hypertension.
2. Abnormal MRI brain or MRV/CT.
3. All cases of pseudo papilledema
4. All patients below 12 years
5. Study period - 1month

Methodology - Data of Patients with IIH like clinical presentation, age of onset, gender, progression and clinical tests performed and diagnostic test employed were collected using a structured proforma.
Variables Included **Age of onset,** Gender Presentation (clinical presentation)
Radiological features etc..

Data Analysis

Data was entered in MS excel & analysed using appropriate software like Epiinfo. Qualitative data will be analysed using proportions. Were tabulated & discussed.

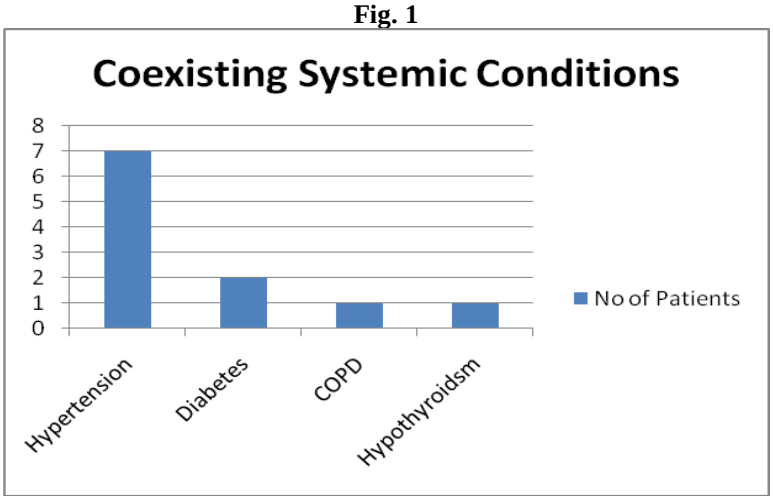
Ethical consideration

This is a record based study. It was started after getting the approval from there search and ethical committee. Confidentiality of data will be maintained.

III. Results:

Sex/Age: Out of the 35 patients, 34 for were females. The average age of onset was 31 (range 19-42 years). A detailed history of the patients were taken including history of drug intake. and association with other systemic disease. Important points that were noted in the drug history included intake of oral contraceptive pills, steroids, tetracycline etc. 5 patients were taking oral contraceptives and 2 steroids. Also history of systemic disorders like hypertension, COPD, sarcoidosis, SLE and thrombocytopenia, Addison's disease, history of weight gain or weight loss, previous history of head injury or intracranial surgeries were also noted. In our study 11cases had systemic risk factors. 7 cases had systemic hypertension 2 had diabetes mellitus, 1 had COPD and 1 had hyperthyroidism. All these patients were getting medications for the above ailments.

Fig. 1



Body mass index of all the patients were calculated. Applying the WHO criterian for obesity, patients were subdivided into 4 categories. No patient belonged to the group with BMI< 18.5 (underweight), 10 patients belonged to the group with BMI 18.5-24.9 (normal weight), 20patients belonged to the group with BMI 25-29.9[over weight and 5patients had a BMI over 30. When this criteria was applied to our study group. 30 out of the 35 (85%) patients belonged to the overweight / obese group

Fig. 2A

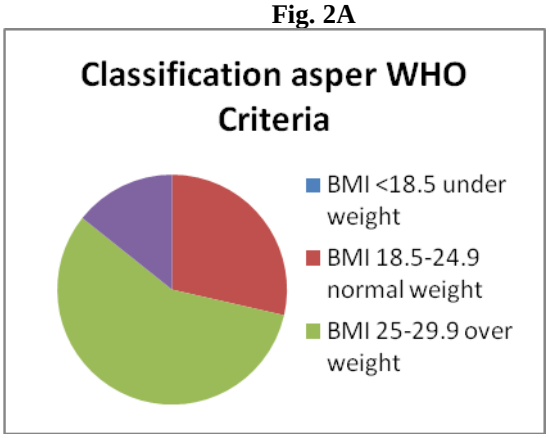
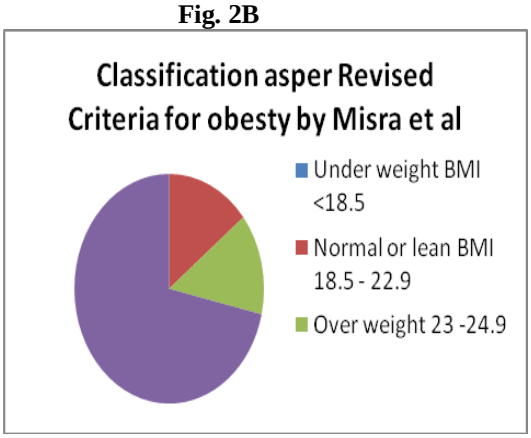


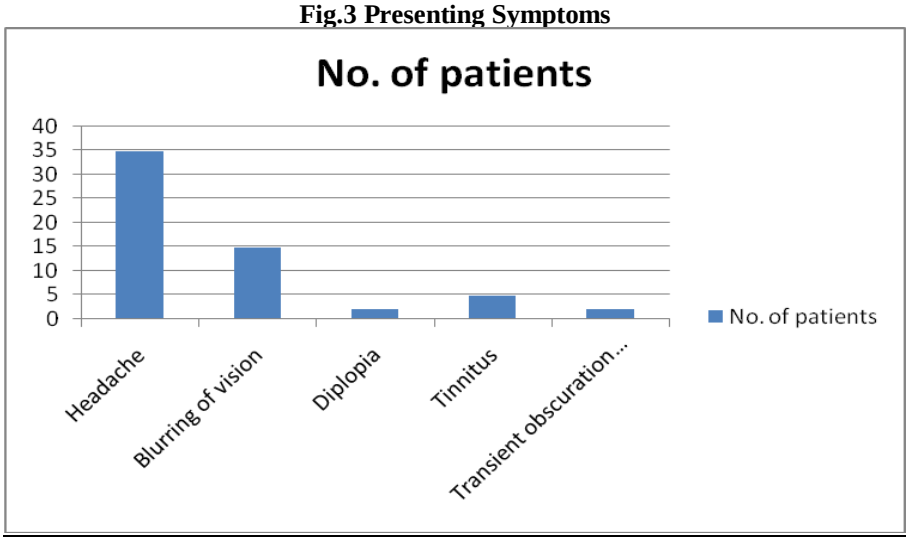
Fig. 2B



Headache:

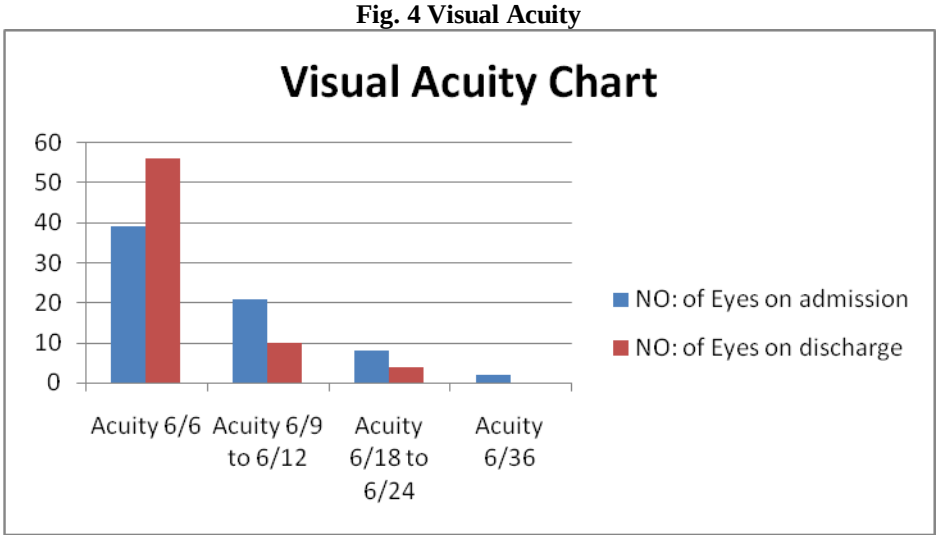
Headache is the most common symptoms and it was present in all cases in our study. Headache is usually very severe throbbing type of pain. Blurring of vision was the second common complaint in our study and it was seen in 15patients (45%) Transient visual obscurations were seen in 2 patients (5.7%). Nausea and vomiting was observed in 12 patients (35%), pulsatile tinnitus in 5 patients (14%), and double vision in 2 patients (5.7%)

Fig.3 Presenting Symptoms



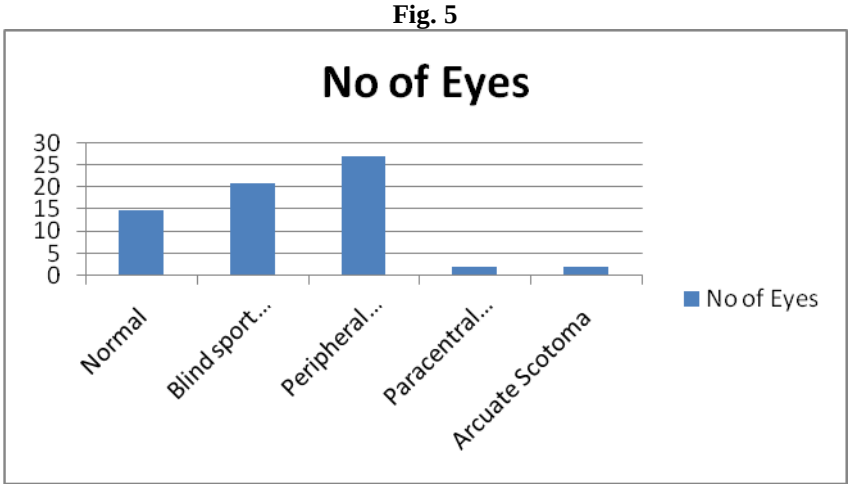
Visual acuity of patients was in the range from 6/6 to 6/36 at the on admission. Out of the 70 eyes 39 had visual acuity of 6/6, 21 eyes had a visual acuity ranging from 6/9 to 6/12, 8 patients visual acuity ranging from 6/18 to 6/24 and 2 patients a visual acuity of 6/36. All patients had treatment with Acetazolamide, Mannitol and Lasix and lumbar puncture was done for diagnostic and therapeutic purposes. Lumbar puncture had to done repeatedly in some patients. Except for 2 patients, all others showed an improvement in visual acuity with treatment. One patient had to be referred for ventriculoperitonial shunt and other for optic nerve fenestration.

Fig. 4 Visual Acuity



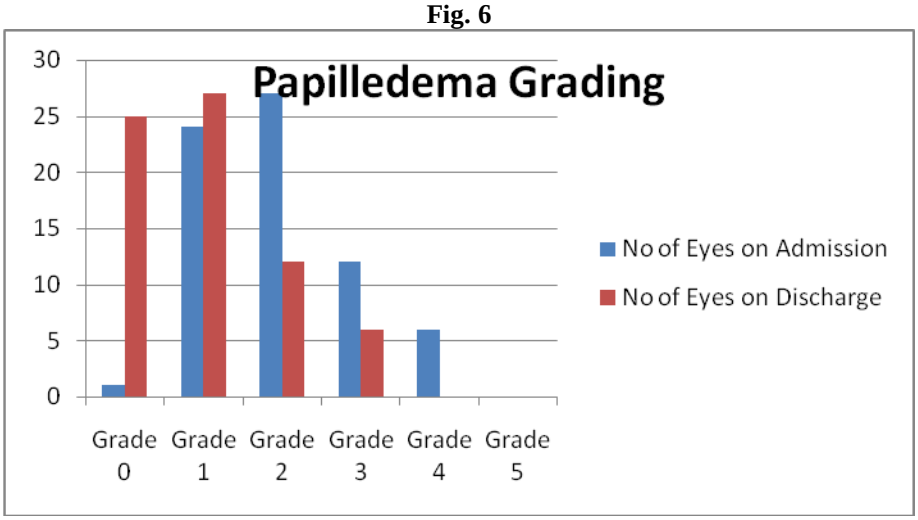
15 patients (44%) showed no visual field defects. Abnormal visual field test included an enlarged blind spotin 21 eyes (30%), peripheral field constriction iJn 27 eyes out of the 70 (39%), paracentral scotoma in 2 out of 70 eyes (3%) and arcuate scotoma in another 2 patients (2%).

Fig. 5



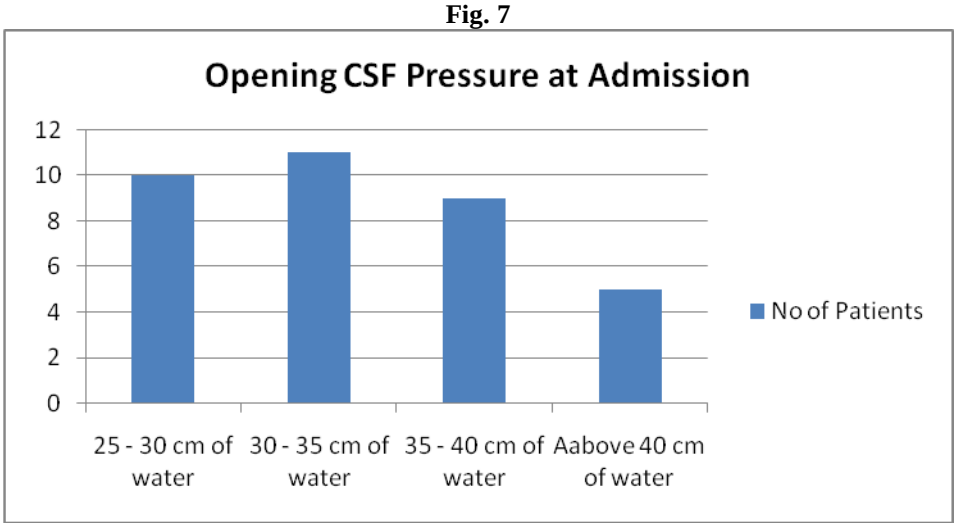
Bilateral and symmetrical papiIledema was observed in 31 patients and asymmetric papiIledema in 4 cases. Papilledema is a cardinal sign of IIH. Other causes of papiIledema a like brain tumors, hydrocephalus , and cortical venous thrombosis were excluded by an MRI brain and MR venogram. These two imaging procedures were normal in all patients. It was graded as per the modified Frisen scale from 0 to5 depending on the severity. The severity of papiIledema at time of presentation and at discharge are given in Fig6

Fig. 6



CSF pressure: Lumbar puncture was done in all patients and serial measurements of CSF pressure was done. An opening pressure of 25 – 30 cm was recorded in 5 patients , 30 -35 cm was present in 10 patients , 35 – 40 cm was present in 10 patients and above 40cm was present in 5 patients. The median CSF pressure was 36.5cm in our study. CSF composition was normal in all patients in our study.

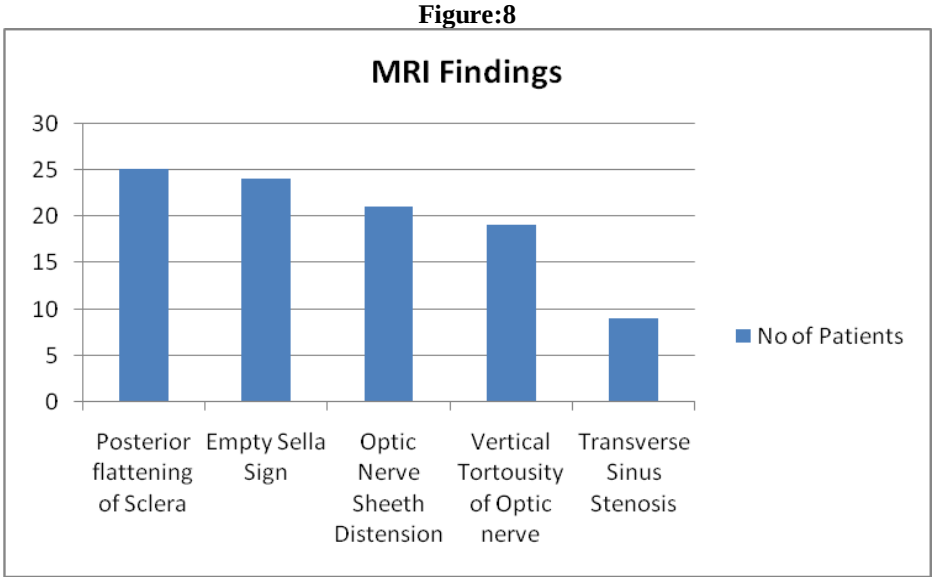
Fig. 7



Radiological signs:

Neuroimaging procedures like MRI brain and MR venogram were done in all patients. None of the patients had an abnormal ventricular size. The following signs were noted in the MRI and MRV of the patients. Posterior flattening of the globe was seen in 25[71%] of our patients. Empty sella syndrome was seen in 24 [68%] of our patients. Optic nerve sheath distension was seen in 21 [60%]% of our patients. Vertical tourtousity of optic nerve was seen in 19[55%] of our patients. Stenosis of transverse sinuswas seen in 9 [25%]of our patients.

Figure:8



IV. Discussion:

The incidence rate of IIH is about 0.5-2/1 lakh/year.[7] It is usually seen in obese females between 15 and 45 years of age. In our study of the 35 patients, 34 were females 97%. The female propendarance is reported in most the previously conducted studies [8.] The average age of onset was 31 years (range 9-45 years). A similar mean age of onset has been reported by Daniels et al.[9]

Important points that should be noted in the history of the patients include intake of oral contraceptive pills, steroids, tetracycline etc. also history of systemic disorders like hypertension, COPD, sarcoidosis, SLE and thrombocytopenia, addison's disease. History of weight gain or weight loss, previous history of head injury or intracranial surgeries. In our study **8** cases had systemic risk factors and the most common one was hypertension. A similar finding has been reported by Bruce et al[10].

Body mass index: was calculated for all patients and patients were grouped as per WHO Criteria of obesity. 25 patients were either over weight or obesce.(71%) Another system of categorization of BMI for Asian Indians has been proposed by Misra et al.[11] This system was to account for the high prevalence of diabetes and

cardiac disease and to accommodate differing associations of BMI with body fat in this population when compared with the Westerners. As per this criteria patients are grouped as under weight (<18.5kg/m2), normal or lean (18.5-22.9kg/m2), over weight (23 -24.9kg/m2) and obese (> 25kg/m2). When this criteria was applied to our study group. 30 out of the 35 (85%) patients belonged to the overweight / obese group Abdul Gafoor et al as reported and incidents of 82% obesity in his study [12].

Headache is the most common symptoms and it was present in all cases in our study. It was seen in 90% of cases by Binder et al[13]**.** Headache is usually very severe throbbin type of pain. Diminution of vision was the second common complaint in our study and it was seen in 45% of patients. Transient visual obscurations were seen in 5.7%. This phenomenon usually occurs due to deficiency of the perfusion of swollen optic nerve head with mild changes in blood pressure. This was observed in higher numbers in other studies by Kim et al [14] Diplopia was seen in 1 patient in our study while its incidence was reported to be 8% and 60% by other studies[15] Pulsatile tinnitus was described by 5 patients in our study. This was described as hearing a swishing heartbeat like sound in one or both ears. Cause of the tinnitus is attributed flow disturbances in the cerebral venous system.

A careful evaluation and monitoring of visual field are necessary in IIH patients.[15] In our study, 44% patients showed no visual field defects. Other field defects included an enlarged blind spot in 21 eyes (30%), peripheral field constriction in 27 eyes out of the 70 (39%), paracentral scotoma in 2 out of 70 eyes (3%) and arcuate scotoma in another 2 patients (2%). Visual field defects have been described by Suh and Kim et al.[16]

Bilateral and symmetrical papal edema was observed in 31 (88%) patients and asymmetric papilledema in 4 cases. Asymmetric papilledema in IIH has been described by Sammual Bidot, et al [17] He has described that this asymmetry is due to the different in size of the bony optic cannel. 30 patients in our study had an opening pressure of 30 cm of H2O.The median CSF pressure was 36.5cm in our study. Riggeal et al has reported a median CSF opening pressure 35cm of H_2O. In his study[18] . CSF composition was normal in all patients in our study. In our study it was found that patients who presented with high CSF opening pressure at the time of admission had a higher grading of Papilledema. When other variables were compared it was found that a high CSF pressure was directly proportional to grade of Papilledema with a P value of < 0.001. Similar finding has been reported by Ayush Dubey et al[19]

Structural MRI is an important tool in the work up of IIH to rule out secondary causes of elevated ICP and to identify certain neuro imaging signs observed in IIH. The following signs were seen in the MRI images of our patient.

(a) Posterior flattening of the globe-This is one of the commonest sign in IIH .There is a direct correlation between elevated ICP and IOP through the transmission of elevated CSF pressure in the subarachnoid space extending through the ONS to the posterior globe . An increase in ICP increases IOP and results in flattening of the posterior sclera[20]. (b)Empty sella sign :It is described as the reduction of mid sagittal height of the pituitary[21]. This is thought to be due to the herniation of arachenocele seen through the diaphragm sellae. (c)Optic nerve sheath distension – ONS enlargement appears as a widened ring of CSF around optic nerve. This will appear as a compressed signal on coronal images and as widened CSF signal intensity on axial images[22]. (d)Vertical tourtousity of optic nerve. This sign is also associated with increased ICP. The optic nerve's distal and proximal points of fixation allow it to bend freely in its path to the globe when intracranial contents protrude under pressure[23]. (e) Stenosis of transverse sinus- MR venography is mainly done to rule out cortical venous thrombosis which is an important cause of raised ICP. But transverse sinus narrowing is a common finding in many cases of IIH. The bony groove in the occiput may be small or absent and this factor along with the compressible nature of the transverse sinus makes it vulnerable to stenosis with increased ICP[24]. Frequency of the signs in our study were 71% , 68% , 60% , and 55% respectively Hingwala et al found the frequency of the signs as 71% , 76% , 95%, and 61% respectively in his study[25]**.** Transverse Sinus stenosis was observed in 25% of our patients. Farb et al has reported a higher incidence of this findings 93% in his study [26]

V. Conclusion

IIH is common in young over weight and obese females and is easily overlooked as other type of benign headache . The most common symptom is headache , followed by blurring of vision and field defects . Documentation of Papilledema is an important step in the diagnosis. A high CSF opening pressure during lumbar puncture is diagnostic. A normal MRI will help in excluding other causes of raised ICP and in clinching the diagnosis. Those patients who presented with high opening CSF pressure presented with a higher grade of Papilledema. Serial assessment of visual acuity ,visual field charting, fundus examination etc are needed in all cases , as some of them may deteriorate even with treatment. Such patients may have to be referred for surgical procedures. So high index of suspicious , meticulous neurological examination and use of MR imaging techniques will help in detecting the cases early. Early and prompt treatment will prevent complications like visual loss.

Conflicts of Interest
There are no conflicts of interest

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