

PART – I

INTRODUCTION

Idiopathic Intracranial Hypertension (IIH) is defined as a clinical entity characterized by raised intracranial pressure without an identifiable pathology.

It is characterized by the presence of symptoms like head ache, vomiting, pulsatile tinnitus, double vision, transient visual obscurations and signs of raised intracranial pressure (ICP) like VI th sixth nerve palsy.

This study tries to analyse the various clinical presentation, investigations of IIH, with the outcome of illness and to study various management options in these patients.

EPIDEMIOLOGY

The annual incidence of IIH in general population is 0.9/1,00,000 and 3.5/1,00,000 among females. In obese women between the age group of 25 to 40 years, the incidence is 19/1,00,000. The female to male ratio is 2:1. Around ninety percent of IIH patients are obese, women of child-bearing age group.

HISTORICAL REVIEW

This disease has been referred by various terminologies since Quincke first described it as “meningitis serosa” in 1893¹. Various other terms used includes ‘otitic hydrocephalus’², ‘Pseudotumour cerebri’, ‘benign intracranial hypertension’.

The term ‘Benign intracranial hypertension’ was widely in use for very long time, since Foley coined it in 1955. Benign nature of IIH was questioned by the presence of loss of vision in some patients and hence it was renamed as ‘Idiopathic Intracranial Hypertension’.

REVIEW OF LITERATURE

Ambika S et al described “the clinical profile, evaluation and management of IIH in a prospective study of 50 patients”.³

Wall M et al described the risk factors for visual outcome in patients with idiopathic intracranial hypertension.⁴

B.B. Bruce studied the Idiopathic intracranial hypertension in men.⁵

F J Rowe et al studies the relationship between obesity and idiopathic intracranial hypertension.⁶

Sandeep Randhawa and Gregory P.Van Stavern studied current treatment of idiopathic intracranial hypertension and concluded “that newer treatment modalities are being experimented for IIH refractory to standard medical therapy, but then their efficacy and safety must be evaluated in large studies before adopting”⁷

Martin G.Radvany et al described “ visual and neurological outcomes following surgical intervention - Endovascular Stenting for Pseudotumor Cerebri associated with Transverse sinus stenosis”⁸

Supuran CT studied and concluded that “Carbonic anhydrase inhibitor Acetazolamide is effective in treating Idiopathic intracranial hypertension.”⁹

Hannerz J and Ericson K studied on “The relationship between idiopathic intracranial hypertension and obesity¹⁰.

Obi EE et al concluded that Optic nerve sheath fenestration is a safe procedure and stabilises visual function in majority of refractory patients.¹¹

John Chen described that “IIH is a disease of child-bearing age group women and its prevalence is growing due to the worldwide obesity epidemic¹²

Bryan D.Riggeal et al described that transverse sinus stenosis is common in Idiopathic intracranial hypertension.¹³

Susan P Mollan et al concluded that “ IIH necessitates a multidisciplinary approach management with ophthalmologists, neurologists and neurosurgeons¹⁴

Fridley J et al studied the role and efficacy of bariatric surgery in treatment of IIH in obese patients¹⁵

Hingwala DR et al studied the imaging results and concluded that Optic nerve head protrusion and posterior globe flattening are associated with IIH.¹⁶

Mulla Y et al described that “ Quality of life in IIH patients is significantly improved by effective headache management.¹⁷

ANATOMY OF OPTIC NERVE

Optic nerve extends from the optic disc to the optic chiasma. It is the continuation of retinal nerve fibre layer which compromise the ganglion cell axons. The optic nerve is about 47-50 mm in length.

PARTS OF THE OPTIC NERVE

Based on the course it can be divided into 4 parts:

1. Intraocular part
2. Intraorbital part
3. Intracanalicular part
4. Intracranial part

INTRAOCULAR PART

Also referred as optic nerve head, optic papilla or optic disc. ONH is the distal part of the optic nerve of 1mm in length.

Nerve fibres converge upon the ONH. Connective tissue support is delivered by the glial cells and astrocytes. The axons at the superior and inferior poles have less structural support. The axons exit through a fenestrated sclera, the lamina cribrosa. Optic nerve diameter expands to about 3mm as it become myelinated in the retrolaminar part.

The normal optic disc is vertically oval and pink in colour.

Zones of optic nerve head

1. Surface nerve fibre layer
2. Prelaminar region
3. Lamina cribrosa
4. Retrolaminar region

INTRAORBITAL PART

Intraorbital part is extending from behind the globe to optic foramen. It is 30mm in length. Here, the optic nerve is covered by three layers of meninges.

Near the exit at optic foramen, the optic nerve is closely related to the annulus of Zinn. The long and short ciliary nerves and vessels surround the optic nerve before entering the eyeball.

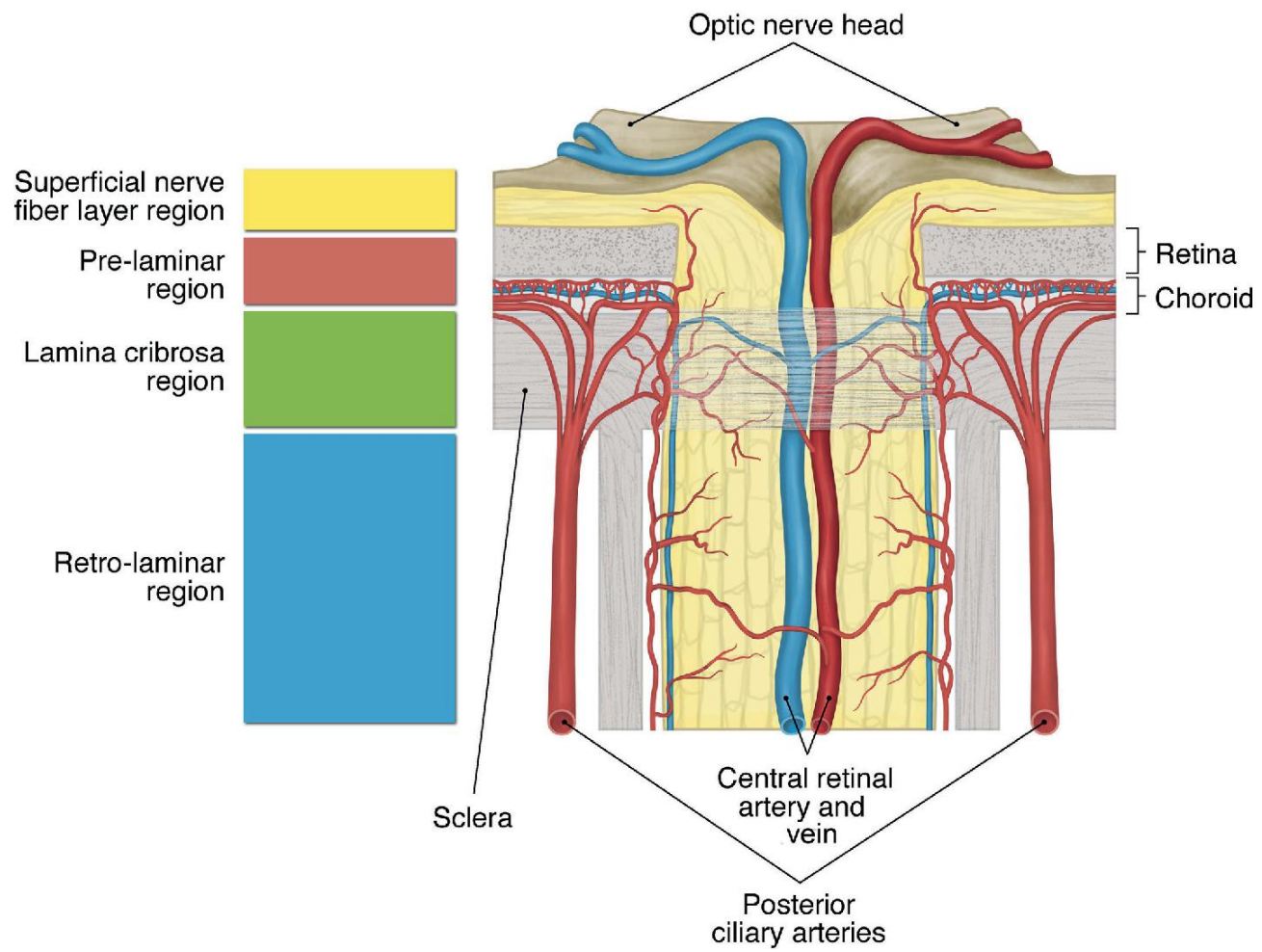


Figure 1: Anatomy of Optic nerve head

INTRACANALICULAR PART

Optic nerve is closely related to the ophthalmic artery in the intracanalicular part. It is 6-9 mm in length.

INTRACRANIAL PART

This part of the optic nerve is about 10 mm in length. It is ensheathed only by pia mater in this portion.

AXOPLASMIC TRANSPORT

Axoplasmic transport is responsible for the supply of the nutrients within the axons.

- Antegrade flow
- Retrograde flow
- Fast & slow components

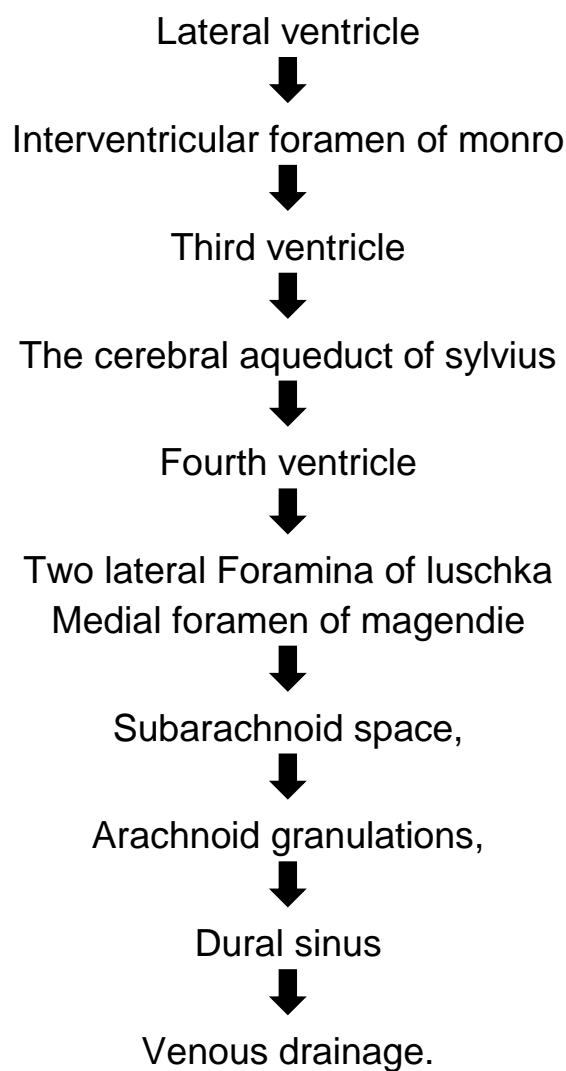
FACTORS AFFECTING THE NORMAL AXOPLASMIC FLOW

- Intracranial pressure
- Intra axonal pressure
- Intra ocular pressure
- Optic Nerve Tissue pressure (ONTP)

CSF DRAINAGE PATHWAY:

CSF is produced by ependymal cells in the choroid plexus of lateral ventricles. The rate of CSF production is 500ml per day with a 140ml of CSF in circulation at any point of time renewed every 6-8 hrs.

Choroid plexus



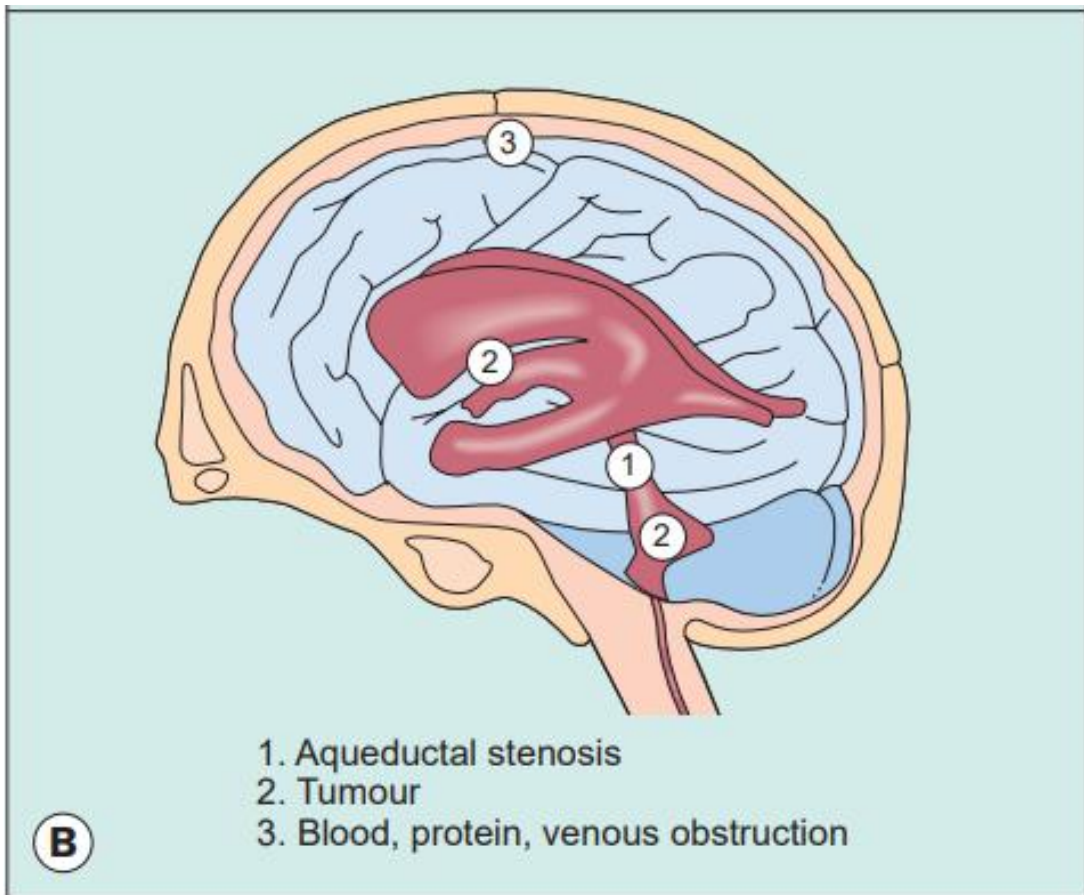


Figure 2: CSF drainage pathway and causes for obstruction

PAPILLEDEMA:

The term Disc edema refers to any type of optic disc swelling regardless of the etiology. Whereas the term Papilledema specifically denotes optic disc swelling that are due to increased intracranial pressure.

PATHOPHYSIOLOGY OF PAPILLEDEMA²⁰

The subarachnoid space around the optic nerve is continuous with the subarachnoid space around the brain.

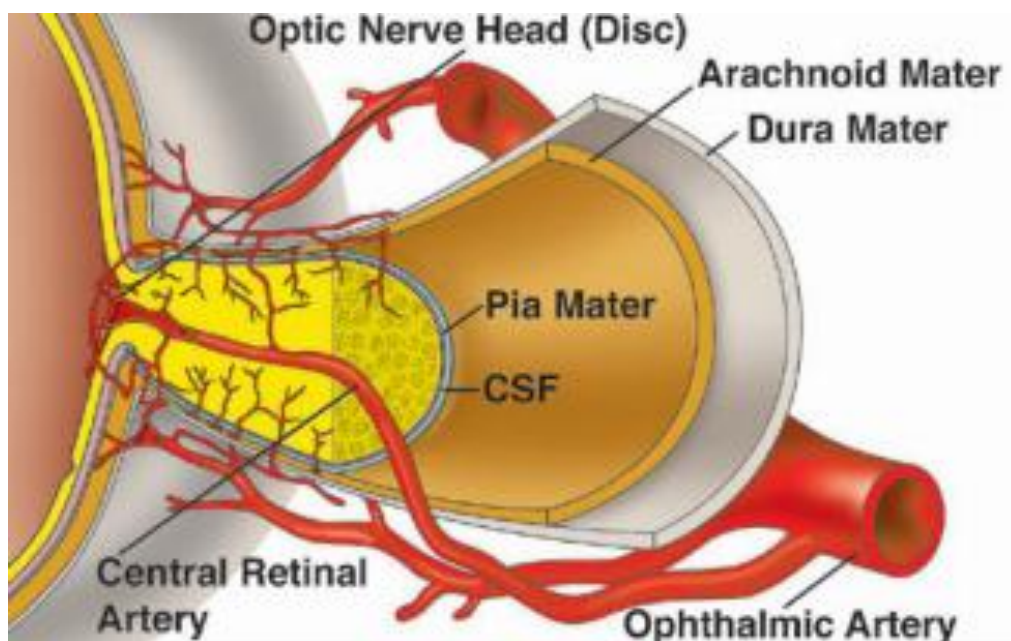


Figure 3 : Coverings of Optic Nerve

Increased ICP causes axoplasmic stasis and also stagnation of venous return. This is the most accepted theory of Axoplasmic stasis proposed by Hayreh.²¹

CAUSES OF RAISED INTRACRANIAL PRESSURE:

- Space occupying lesions
- Cerebral edema
- Blockage of CSF flow
- Reduction in CSF resorption
- Increased CSF production
- Idiopathic intracranial hypertension

MODIFIED DANDYS CRITERIA FOR DIAGNOSIS OF IIH^{18,19}

1. Symptoms or signs of raised intracranial pressure.
2. Elevated CSF opening pressure more than 250mm of water.
3. Normal CSF composition.
4. Normal neuroimaging except for small ventricles and empty sella.
5. No other cause of increased ICP found.

EXOGENOUS SUBSTANCES COMMONLY ASSOCIATED WITH IIH

Hormonal

- Corticosteroid withdrawal
- Levonorgestral
- Danazol
- Tamoxifen
- Growth hormone
- Anabolic steroids

Antibiotics

- Tetracycline
- Doxycycline
- Minocycline
- Nalidixic acid
- Nitro furantoin

NSAID'S

- Indomethacin
- Rofecoxib

Vitamin A

- Retinol
- Retinoids

SYSTEMIC ILLNESS ASSOCIATED WITH IHH

- Obesity
- Anemia
- Systemic hypertension
- Sleep apnoea syndrome
- Polycystic ovarian disease
- Systemic lupus erythematosus
- Hyperthyroidism
- Sarcoidosis
- Renal failure

SECONDARY CAUSES OF ITH

OBSTRUCTION OF SUPERIOR SAGITTAL AND TRANSVERSE SINUS

- Primary hematologic
- Activated protein C deficiency
- Essential thrombocythemia
- Factor V Leiden mutation
- Idiopathic thrombocytopenic purpura
- Paroxysmal nocturnal hemoglobinuria

OBSTRUCTION OF TRANSVERSE SINUS

- Dural arteriovenous fistula
- Infection (mastoiditis)
- Haematologic causes
- Tumours (extra vascular)

OBSTRUCTION OF INTERNAL JUGULAR VEIN

- Iatrogenic
- Indwelling catheter
- Surgery/traumatic/tumour

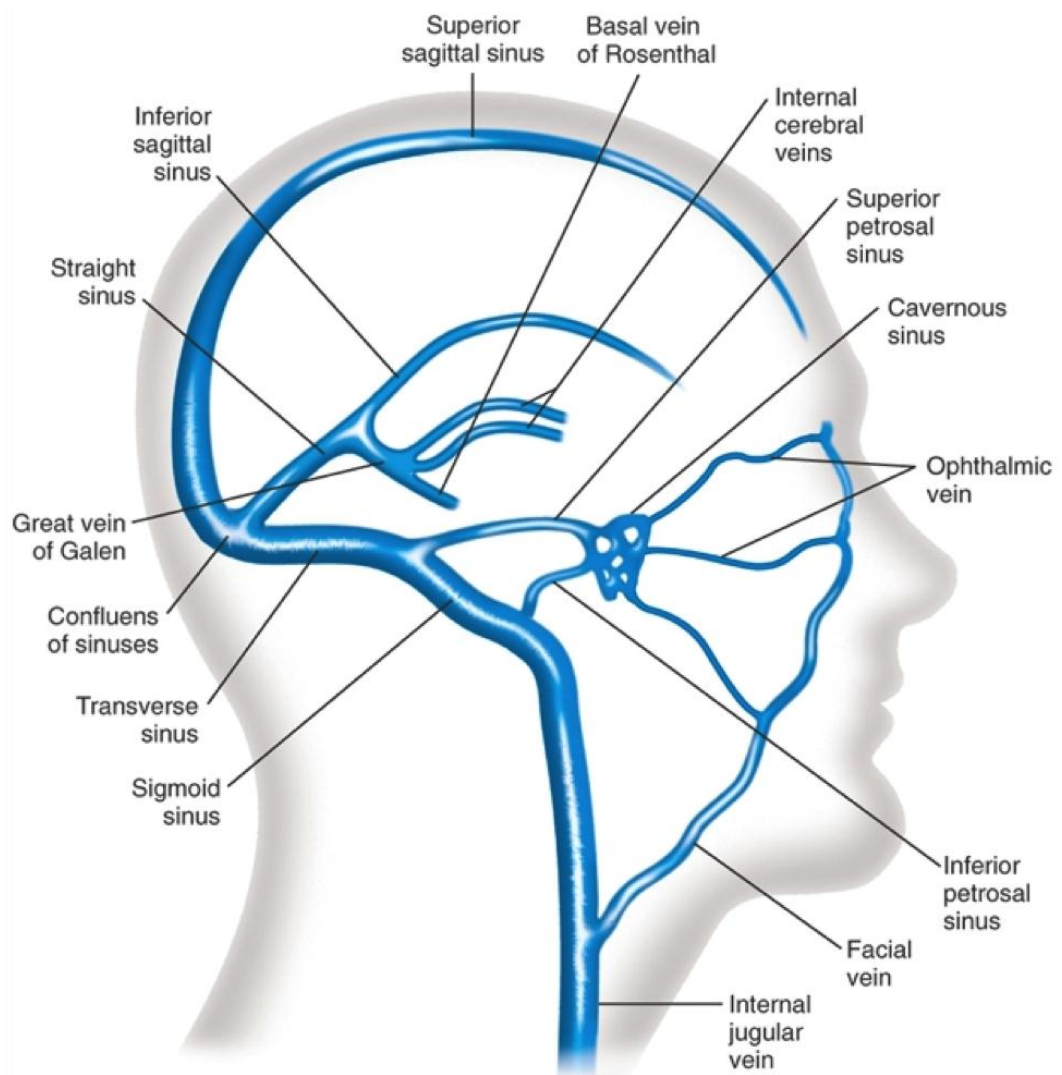


Figure 4: Cerebral venous sinuses

OBESITY AND IIH

Body mass index of more than 30 kg/m² is classified as Obesity.

Mechanisms of IIH association in obesity:

- 1) Obstructive sleep apnoea which is commonly associated with obesity, leads to nocturnal hypercapnia which in turns increases the intrathoracic pressure thereby raising the ICP.
- 2) Increased abdominal pressure subsequently elevates the intra-thoracic pressure and rise in cerebral venous pressure affecting ICP.²²
- 3) Pro inflammatory cytokines released by the adipose tissue like Leptin, Adiponectin play a role in IIH.

CLINICAL FEATURES

NON VISUAL SYMPTOMS

- Headache
 - Most common presenting symptom
 - Generalized headache
 - Mild to moderate intensity
 - Worse in the morning, wakes the patient from sleep
 - Associated with vomiting and photophobia
 - Severity is not in relation with the intracranial pressure
 - Increased on straining / valsalva manoever
 - Headache is due to stretching of meninges because of increased ICP

- Nausea / vomiting
 - Often associated with increased ICP
 - Projectile vomiting can also occur
 - Vomiting with bradycardia suggests herniation of intracranial contents
- Pulsatile tinnitus
 - Unilateral or bilateral
 - Intracranial noises often referred whooshing sound
- Loss of consciousness
 - Occurs due to compression of cerebral cortex
- Motor rigidity
 - Due to tentorial herniation and compression of crura cerebri

VISUAL SYMPTOMS

- Visually asymptomatic
 - Routine snellens visual acuity testing is not capable of detecting the visual loss in IIH
 - Field defects can be detected only in perimetry
 - Few patients may complaint of a dark spot which is due to enlargement of blind spot
 - Visual loss occurs in chronic cases because of optic atrophy
- Transient visual obscurations
 - Lasting from 5 seconds to 30 seconds

- May present as minimal blurring to complete gray out of vision
- Upto 20 episodes per day
- Some patients even present with photopsias
- Obscurations are due to transient ischemia of optic nerve
- Horizontal diplopia
 - It is a false localizing sign
 - Due to sixth nerve palsy caused by stretching of one or both abducens nerves over the petrous tip
- Pupillary dilatation
 - Caused by compression of oculomotor nerve

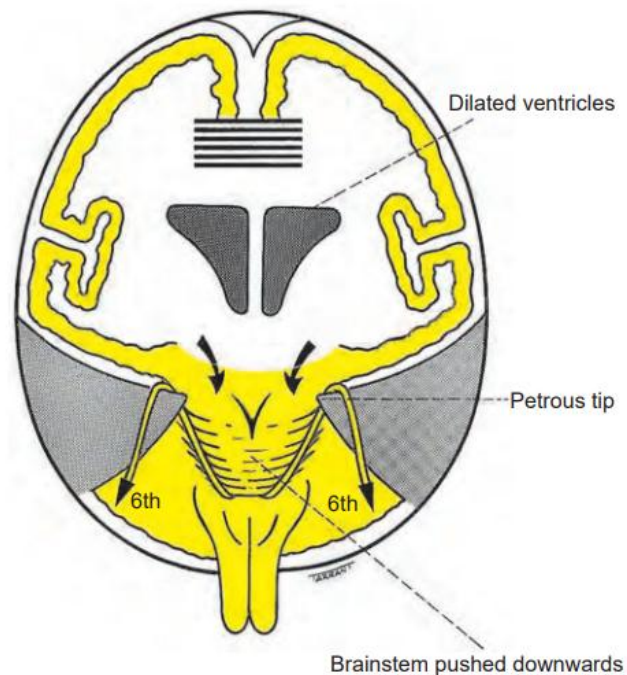


Figure 5: Compression of Abucens nerve

SIGNS OF PAPILLEDEMA

- Elevation of the optic disc
- Blurring of the optic disc margins
- Filling in of the physiological cup
- Edema of the peripapillary nerve fiber layer
- Retinal or choroidal folds



Figure 6: Papilledema

VASCULAR SIGNS OF PAPILLEDEMA

- Hyperemia of the optic disc due to increased vascularity of the optic disc and opening up of the smaller blood vessels over the disc surface.
- Venous dilatation and tortuosity because of reduction in the venous outflow and increased pressure within venules.
- Peripapillary hemorrhages caused by rupture of small blood vessels.
- Exudates in the disc or peripapillary area by leakage of proteins and lipids from congested blood vessels.
- Nerve fiber layer infarcts are caused by decreased flow in the vessels of posterior ciliary circulation.
- Tortuous vessels on or surrounding the disc
- Peripapillary splinter hemorrhages
- Cotton wool spots and hard exudates over the disc and the macula as incomplete star

PATON LINES :

- The edema may extend beyond the optic nerve head in the form of circumferential retinal folds and parallel choroidal folds over the posterior pole.



Figure 7 Patons line

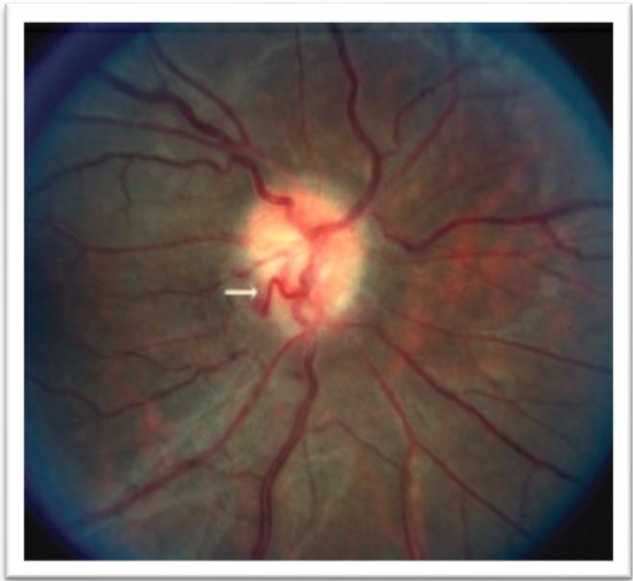


Figure 8: Optociliary shunt

OPTOCILIARY SHUNTS :

- Pre-existing veins connecting the retinal and choroidal vasculature
- Increased ICP compresses Retinal vein and opens this shunt vessels
- May disappear once ICP is reduced

CLASSIFICATION:

Papilledema can be classified based on the duration and the severity.

Based on duration, it can be

- Early
- Fully developed
- Chronic
- Atropic

FRISENS' STAGING OF PAPILLEDEMA

- Frisens have graded the papilledema according to the severity,

Normal optic disc (Frisen stage 0)

- Mild elevation of the nasal nerve fiber layer.
- Rarely a major vessel may be obscured in the upper pole

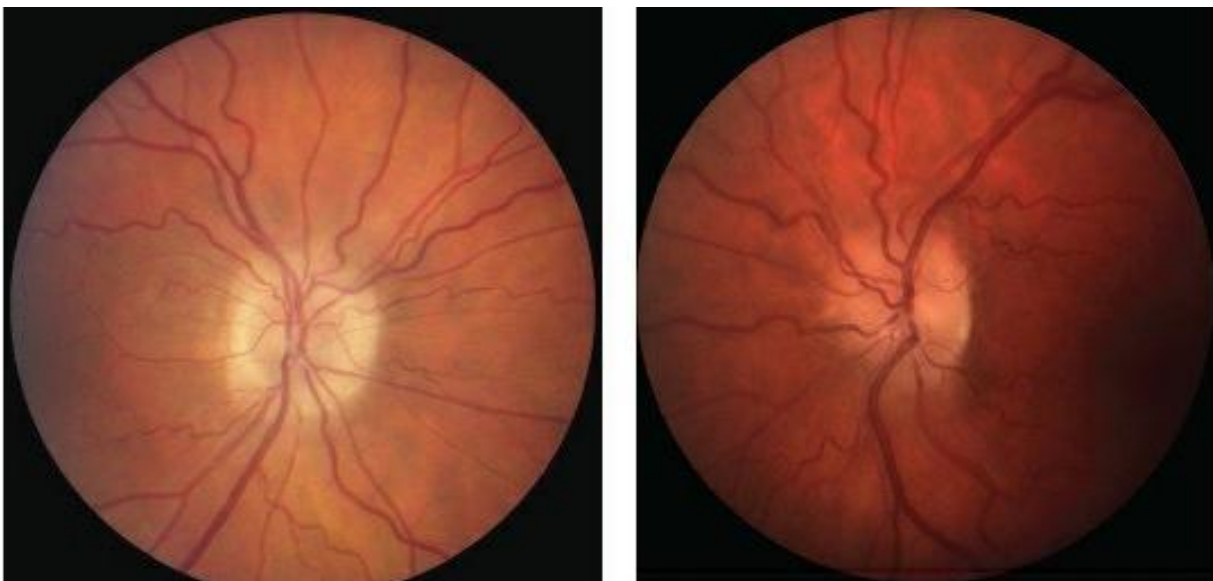


Figure 9: Stage 0 & Stage 1 Papilledema

Very early papilledema (Frisen stage 1)

Nasal disc margin obscuration

No elevation of disc margins

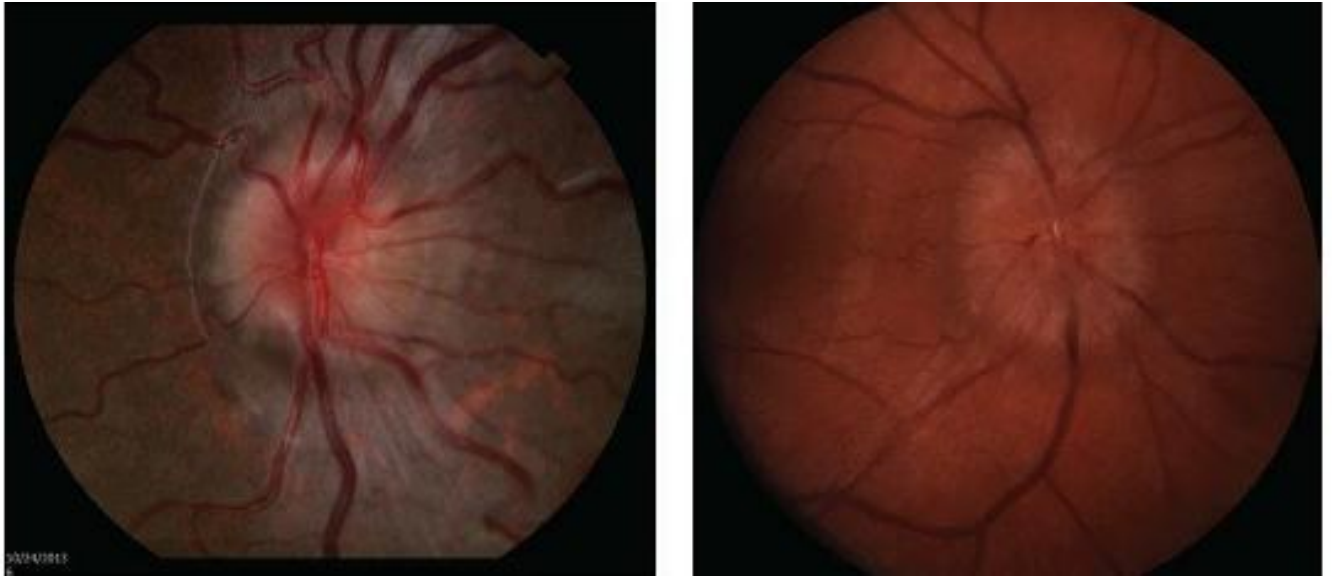


Figure 10: Stage 2 & Stage 3 Papilledema

Early papilledema (Frisen stage 2)

- Obscuration of all the optic disc borders,
- Nasal border elevation
- Complete peripapillary halo

Moderate papilledema (Frisen stage 3)

- All the margins of the optic disc are elevated.
- Obscuration of one or more segments of major blood vessels leaving the disc
- Peripapillary halo

Marked papilledema (Frisen stage 4)

- Elevation of entire optic nerve head. Obscuration of all the borders
- Prominent peripapillary halo.
- Total obscuration of a segment of a major blood vessel.

Severe papilledema (Frisén stage 5)

- Dome-shaped configuration of optic nerve head
- Obliteration of the optic cup

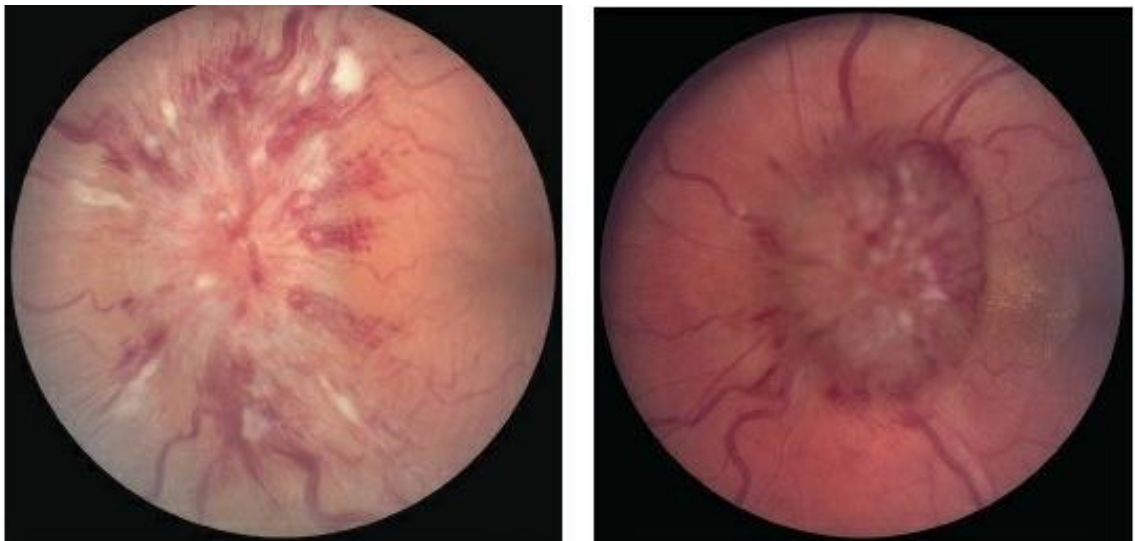


Figure 11: Stage 4 & Stage 5 Papilledema

PSEUDO PAPILLEDEMA

Blurring of margins resembling Disc edema with absence of

- Microvascular abnormalities on the ONH surface
- Opacification of the peripapillary retinal RNFL is referred to as Pseudopapilledema.

DIFFERENTIAL DIAGNOSIS FOR PSEUDOPAPILLEDEMA:

1. Optic disc drusen
2. Tilted optic disc
3. Peripapillary myelinated nerve fibers
4. Crowded disc in hypermetropia



Figure 12: Optic Disc Drusen



Figure 13: Myelinated nerve fibre

INVESTIGATIONS

1. PERIMETRY

- Visual field defects are more common in IHH which are detected by static perimetry. Static perimetry uses a stationary target. Luminance is adjusted to vary its visibility.
- Commonly used automated perimeter are
 - Humphrey Field Analyzer
 - Octopus perimeter.
- The amount of time required for static perimetry varies with the strategy used.
- In Humphrey Field Analyzer,
 - Full-threshold test usually - 10 to 12 minutes per eye, whereas use of the Swedish interactive threshold algorithm (SITA)
Standard strategy - about 4 to 6 minutes
SITA - Fast strategy - 3 to 4minutes
 - Depending upon the size of the field to be tested, most neuro-ophthalmologists use a
24-2 test - central 24 degrees is tested
30-2 test - entire field is tested out to 30 degrees.

For patients with central or paracentral defects, 10-2 - 10 degree field test is used.

RELIABILITY INDICES:

- Reliability indices are used to analyse the significance of visual field test done.
- Fixation loss – Indicates the gaze fixation during the test
- False-positive errors – Responses are recorded even when no stimulus is presented
- False-negative errors – Not responding to a stimulus which is previously recorded
- If the Fixation losses are more than 20% and False positive, False negative more than 33%, the test is considered not reliable.

FIELD DEFECTS

- Enlargement of blind spot - Most common field defect found in IHH
 - Nasal step defect
 - Arcuate scotoma
 - Peripheral constriction of fields
 - Occurs in cases of longstanding IHH

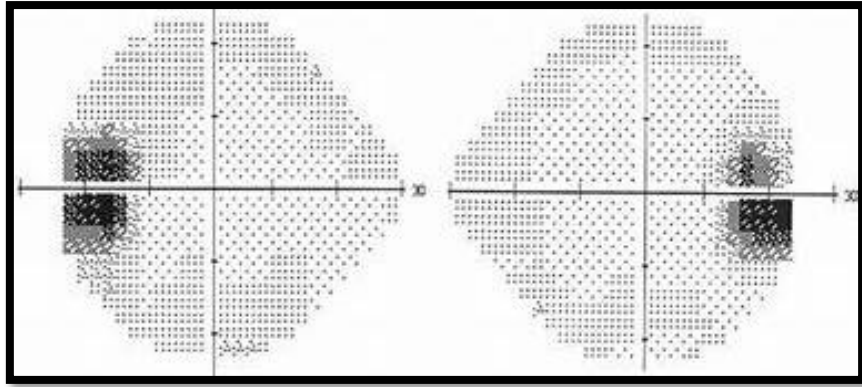


Figure 14: Enlarged blind spot

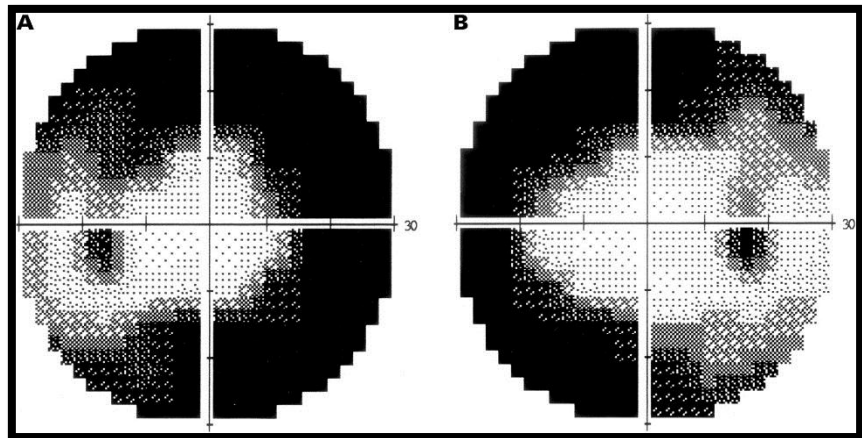


Figure 15: Peripheral constriction of fields

2. ULTRASONOGRAPHY - B SCAN

- Measures the external diameter of the optic nerve sheath (ONSD)
- Optic nerve sheath is said to be distended if the measurement is 5.0–5.7 mm or greater at 3.0 mm behind the globe.

CRESCENT SIGN:

- An echolucent area in the anterior intraorbital nerve is referred as crescent sign in ultrasonography
- It represent increased separation of the nerve from its sheath

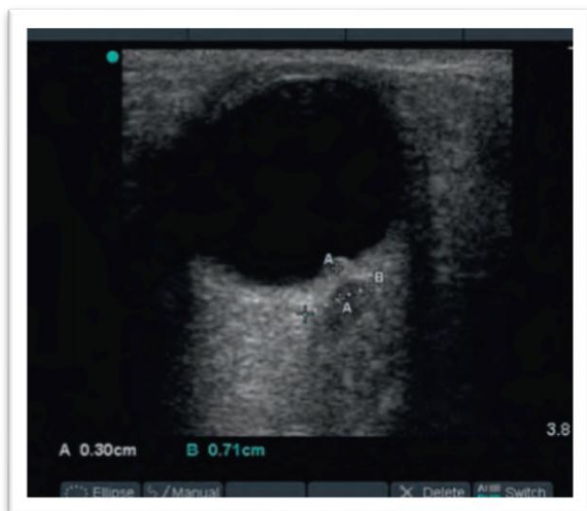


Figure 16: Optic Nerve Sheath Diameter

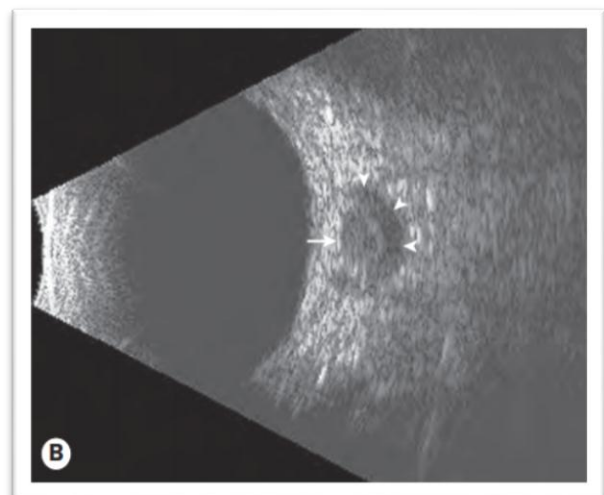


Figure 17: Crescent sign

LATERAL GAZE / THIRTY DEGREE TEST:

- Commonly leads to a 10% reduction in diameter on A-scan measurement in the presence of excess fluid

3. NEUROIMAGING

- COMPUTED TOMOGRAPHY

- To rule out intracranial and space occupying lesions
- Helps in diagnosing cerebral edema
- Acute vascular causes like hemorrhages and infarcts can be made out

- MAGNETIC RESONANCE IMAGING – MRI ANGIOGRAPHY / VENOGRAPHY

- Empty sella
- Protrusion of optic papilla into posterior globe resulting in flattening of sclera
- Dilated optic nerve sheath
- Tortuosity of optic nerve
- Prominent perioptic space
- Slit like ventricles
- Superior sagittal sinus thrombosis
- Lateral transverse sinus thrombosis

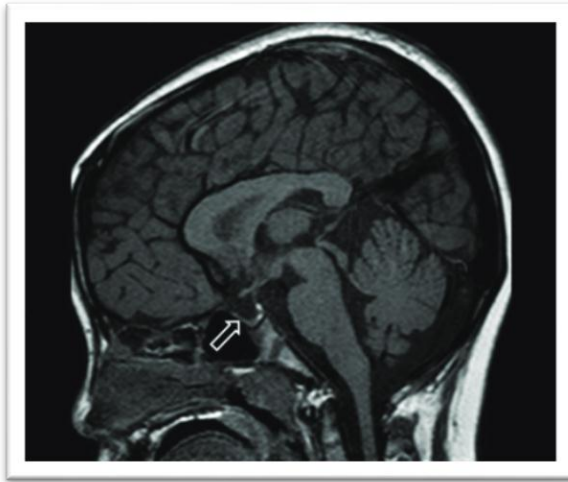


Figure 18: Empty sella

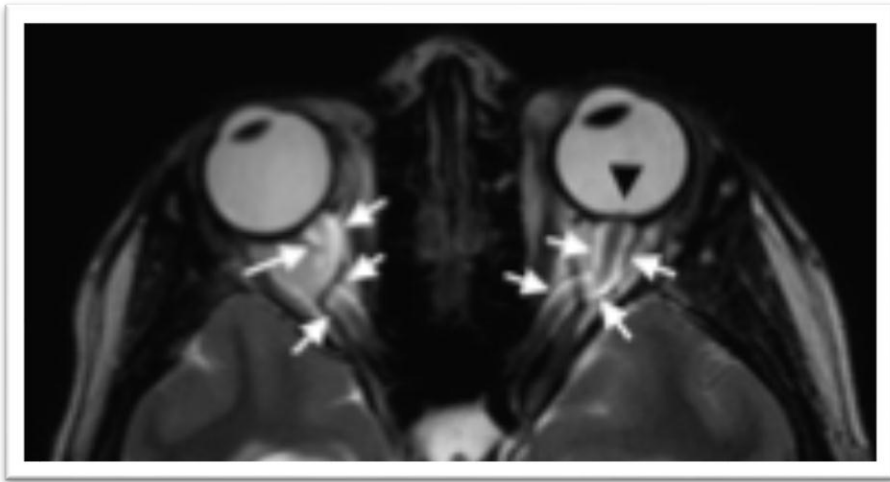


Figure 19: Dilated optic nerve sheath

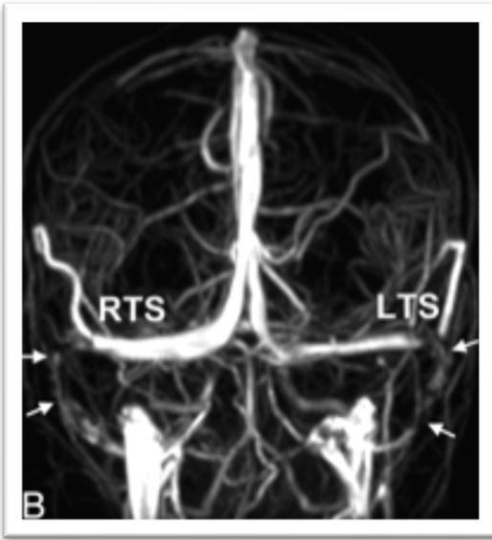


Figure 20: Transverse sinus Thrombosis

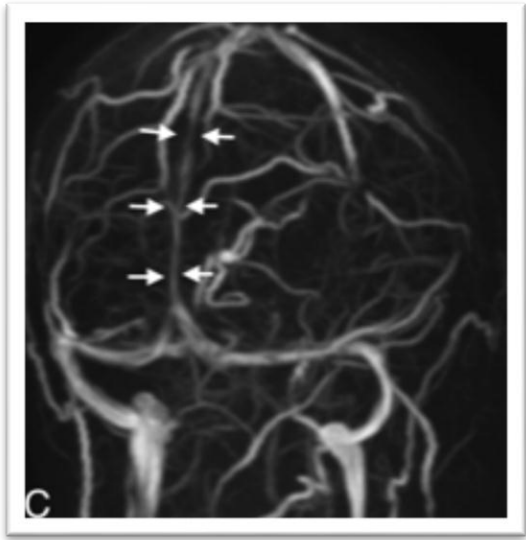


Figure 21: Sigmoid sinus Thrombosis

4. LUMBAR PUNCTURE

- Lumbar puncture is relatively safe procedure done with 18-20 gauge needle in left lateral decubitus position

PREQUISITE

- Must not be carried out until imaging has excluded a intracranial space-occupying lesion that may cause downwards herniation of the intracranial contents through foramen magna leading to fatal results
- To rule out any clotting abnormality, including therapeutic anticoagulation

DIAGNOSTIC USE

- To check the opening pressure in cases of Idiopathic intracranial hypertension
- Opening pressure of more than 250mm of water in adults and more than 280mm of water in children is suggestive of IIH
- To exclude other causes of raised intracranial tension by CSF analysis
- CSF analysis include Total / Differential leucocyte count, glucose, protein and cytology

THERAPEUTIC USE

- Therapeutic in cases of IIH which relieve the symptoms of headache

COMPLICATIONS

- Painful
- Difficult to perform in cases of obese patients
- Post procedural headache may occur in 40% of patients
- Infection
- Acquired Arnold chiari malformation
- Herniation of intracranial contents if space occupying lesions are not ruled out

5. MONITORING OF CHANGES IN THE OPTIC NERVE HEAD :

FUNDUS PHOTOGRAPHY

- Fundus photography is useful in analyzing the progression and documentation of disc edema in subsequent visits.
- Response to treatment can be evident in follow up visits.

CONFOCAL SCANNING TOMOGRAPHY

- Newer method to quantify the edema of the optic nerve head.
- It demonstrates the gradually increasing retinal surface elevation.

OPTICAL COHERENCE TOMOGRAPHY

- Noninvasive imaging technique useful for evaluation and monitoring of IIH.
- It quantifies RNFL thickening in eyes with optic disc edema.

TREATMENT

The primary goals of treatment in IIH are

- 1) To preserve vision
- 2) Alleviate headache and other symptoms of raised intracranial tension

The treatment of IIH is based on Idiopathic Intracranial Hypertension Treatment Trial.

Therapeutic decisions are mainly based on individual patient's characteristics.

Factors to consider

- (1) Presence and severity of headaches;
- (2) Severity of visual loss at presentation;
- (2) Presence of associated aggravating factor.

Treatment can be medical management or by surgical management.

MEDICAL MANAGEMENT:

WEIGHT LOSS:

Weight loss is advised for IIH in obese patients based on earlier analyses. Modest weight loss of 3% to 6% may help in patients' symptomatic improvement.

Caloric restriction to 400 kcal/day lead to IIH resolution in majority of patients.

In obese patients with intracranial hypertension secondary to hypoxia and hypercapnia due to obstructive sleep apnea may respond not only need weight loss, but treatment of obstructive sleep apnea.

CARBONIC ANHYDRASE INHIBITOR:

Acetazolamide is the most commonly used drug for IIH. Mechanism of action is by reducing the production of CSF from the choroid plexus along with its diuretic effect.

Dosage - IIHTT showed that Acetazolamide in a dose of 1gm to 4gm per day along with weight loss is significant in reducing the symptoms and papilledema in cases of IIH.

Side effects of acetazolamide:

- Light headedness,
- Dizziness,
- Hypersensitivity reaction,
- Renal stone,
- Metabolic acidosis,
- Hypokalemia.

DIURETICS:

Furosemide is a loop diuretic used in the management of IHH. It acts as second line of management next to acetazolamide.

Dosage – 20mg to 40mg Bd

Side effects:

- Ototoxicity,
- Hypokalemia,
- Hyperuricemia,
- Hyperglycemia,
- Metabolic alkalosis.

CORTICOSTEROIDS:

Although beneficial in the treatment of raised ICP associated with systemic inflammatory disorders like sarcoidosis and systemic lupus erythematosus, they have their own side effects.

Side effects:

- Rebound increase in ICP
- Weight gain which is undesirable in obese IHH patients
- Corticosteroids should be reserved for emergent treatment of patients with severe progressive visual loss going for a definitive surgical procedure.

TOPIRAMATE:

Topiramate is an anticonvulsant used to treat the headache which also provides weak carbonic anhydrase inhibition.

Side effects:

- Drowsiness, Fatigue
- Cognitive dysfunction
- Loss of appetite and weight loss which are beneficial in treating IHH.

OCTREOTIDE:

Octreotide is a somatostatin analogue found to be useful for migraine and cluster headache associated with IHH

REPEATED LUMBAR PUNCTURE:

Repeated LP has been helpful in patients

- Who were refractory to medical treatment
- Not ideally fit for surgical management
- Who are planning surgery for symptomatic relief
- It provides symptomatic relief to patients with headache. But the effect is temporary and needs repeated lumbar punctures.

SURGICAL MANAGEMENT:

Surgery is indicated in patients with severe optic neuropathy at initial presentation or when medical treatment is not successful in preventing progressive visual loss.

The decision of whether to go with a CSF shunting procedure or an Optic nerve sheath decompression (ONSD) depends on individual patients. ONSD may be preferred in patients with severe visual loss and disc edema but minimal headache, whereas CSF shunting procedure are for patients with visual loss, disc edema, and severe headaches.

OPTIC NERVE SHEATH FENESTRATION:

In this surgery, Optic nerve sheath pressure is reduced by creating windows or multiple slits in the dura sheath and arachnoid that surrounds the optic nerve just behind the globe. This allows CSF to leak leading to decrease of CSF pressure and thereby improving the severity of papilledema and visual function immediately. But it is not effective in reducing the intracranial pressure.

INDICATION:

- IIH patients who present with severe or progressive visual loss.
- Patient's refractory to medical management.
- Severe papilledema progressing to macular edema and exudates.

APPROACHES:

1. Medial transconjunctival orbitotomy
2. Lateral orbitotomy
3. Superior eyelid approach

MECHANISM:

1. Filtering effect reduces the local CSF pressure which improves the peripapillary circulation.
2. Generalized decrease in intracranial pressure.
3. Fibrous scarring occurs between dura and arachnoid site of surgery protects the optic nerve head from raised CSF pressure.

COMPLICATIONS:

- Optic nerve injury
- Extraocular movements restriction
- Diplopia
- Pupillary dysfunction
- Vascular occlusion causing permanent or transient loss of vision
- New onset Visual field defects
- Orbital hemorrhage
- Globe perforation
- Infection

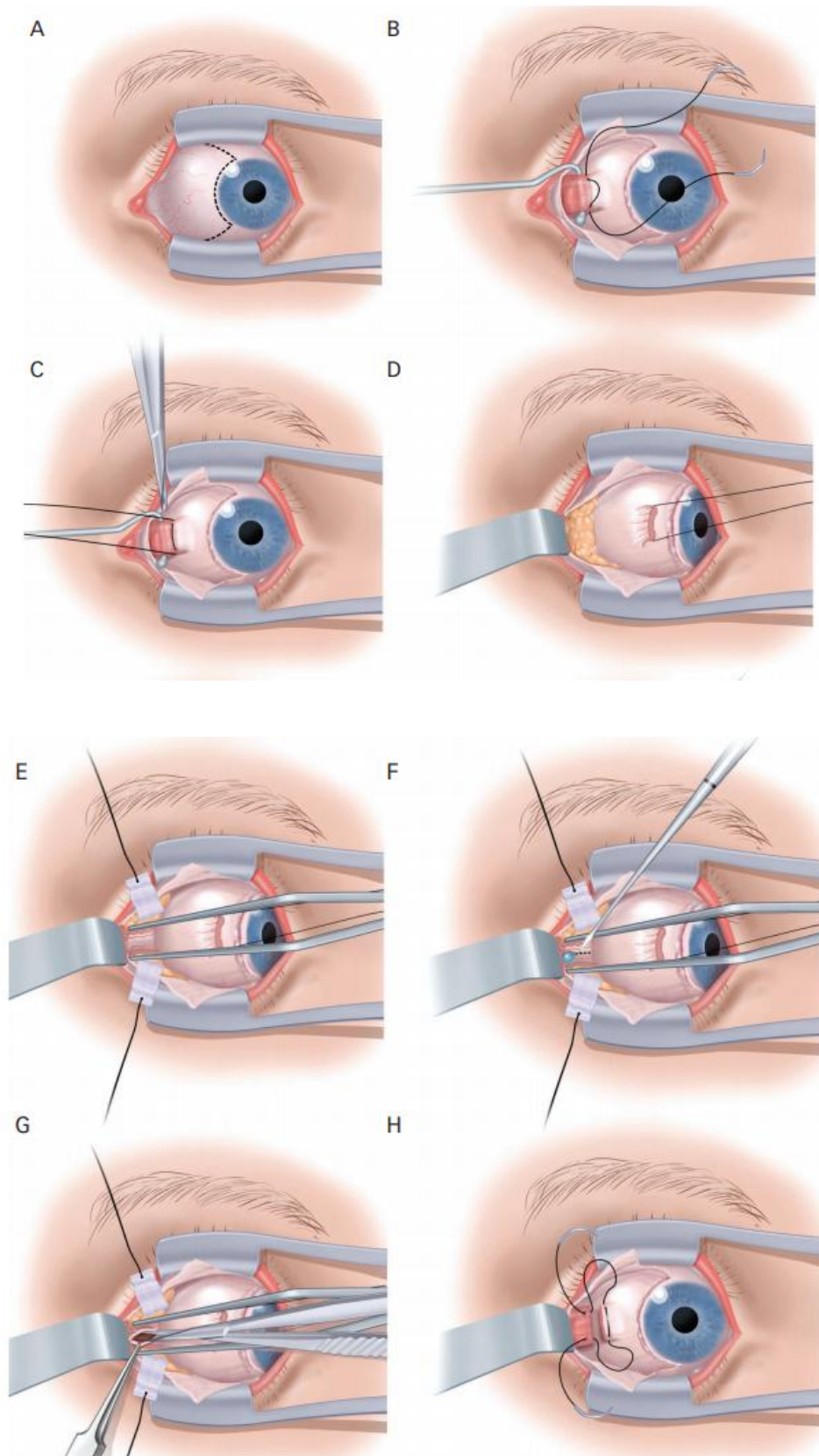


Figure 22: Steps of Optic Nerve Fenestration Surgery

CSF SHUNTING PROCEDURES:

CSF shunting procedures are very effective in lowering the intracranial pressure in IIH patients. Immediate decrease of ICP, resolution of papilledema along with visual recovery is seen in patients treated with shunting procedures. Risk for optic nerve damage which is threatening in ONSD is absent in CSF shunting procedures.

TYPES OF CSF SHUNTING PROCEDURES:

1. Lumbo peritoneal shunting
2. Ventriculo peritoneal shunting

Lumbo peritoneal shunting procedures are more preferred over the other procedure.

INDICATIONS:

- Cases refractory to medical treatment
- Medically intractable headache

COMPLICATIONS:

- Mechanical shunt dysfunction due to obstruction most commonly at peritoneal end
- Infections
- Migration of shunt
- Radiculopathy

- Arnold chiari malformation
- CSF leak
- Abdominal pain
- Need for repeated revisions
- High failure rate

BARIATRIC SURGERY:

Vertical band gastroplasty is a gastric exclusion procedure. It helps essentially in treating the comorbid conditions of obesity such as diabetes mellitus and obstructive sleep apnoea.

Bariatric surgeries are useful in long term management of IIH rather than relieving immediate symptoms.

VENOUS SINUS STENTING:

MRV shows more number of cases with narrowing or stenosis of transverse venous sinuses which obstructs venous return thereby increasing the intracranial pressure . This stenting of obstructed transverse or sigmoid sinus procedure reduces cerebral venous pressure and intracranial pressure.

COMPLICATIONS:

- Epidural and sub dural hematoma
- Anaphylaxis and shock
- Hearing loss

Because of dreaded complications, venous sinus stenting is reserved for cases of Fulminant IHH

SPECIAL CIRCUMSTANCES

PREGNANCY

Women who present with IHH during pregnancy are diagnosed and treated as similar to nonpregnant women. Most women do well, with little or no permanent visual loss. There is no contraindication to pregnancy or special protocols unless other medical complications are present.

Acetazolamide may be given after 20 weeks of gestation. If vision worsens, corticosteroids may be used. ONSF or CSF shunting may be performed if warranted. Suspicion of cerebral venous sinus thrombosis is needed in cases of raised Intracranial pressure.

FULMINANT IHH

These are patients with IHH who experience a

- Rapid onset of visual symptoms
- Significant visual deterioration
- Marked visual field loss
- Loss of central visual acuity loss
- Established papilledema at presentation
- Macular edema
- Ophthalmoparesis

Aggressive treatment that is needed which includes Intravenous corticosteroids and insertion of a lumbar drain in cases of Fulminant IHH

PART II

PART – II

AIM OF THE STUDY:

- To study the clinical profile of patients presenting with idiopathic intracranial hypertension.

OBJECTIVES:

- To assess the type of presentation and visual function in cases of idiopathic intracranial hypertension.
- To study the response to medical management of idiopathic intracranial hypertension.

MATERIALS AND METHODS:

Subject selection:

- All patients presenting with idiopathic intracranial hypertension attending RIOGOH services are taken up for study after consent.

Inclusion criteria:

- All patients with features of raised intracranial pressure fulfilling Modified Dandy criteria will be included in the study after obtaining a written informed consent.
1. Symptoms or signs of raised intracranial pressure.
 2. Elevated CSF opening pressure more than 250mm of water.
 3. Normal CSF composition.

4. Normal neuroimaging except for small ventricles and empty sella.
5. No other cause of increased ICP found.

Exclusion criteria:

Intracranial hypertension due to cerebral venous sinus thrombosis, space occupying lesion, metabolic causes, meningitis/encephalitis and trauma will be excluded from the study.

Methods:

All patient's reliable history including onset of headache, blurring of vision, pulsatile tinnitus and history of any other neurological symptom are enquired.

All patients are subjected to snellen's visual acuity, slit lamp biomicroscopy, fundus examination, colour vision, fields by automated perimetry, neuroimaging.

Follow up:

The patients are to be followed up at 1 week, 1 month and 3 months of treatment. At each follow up visits' patient examination should include snellen's visual acuity, pupillary reaction, colour vision, fields by automated perimetry, fundus examination.

ANALYSIS AND RESULTS

Analysis of collected data was done based on the following

1. Age distribution

- Male – 1
- Female - 2

2. Sex distribution

- 11- 20 years – 1
- 21 – 30 years – 2
- 31 – 40 years – 3
- More than 40 years - 4

3. Presence or absence of various symptoms

- Yes – 1
- No - 0

4. Best corrected visual acuity

- 6/6 to 6/18 - 1
- 6/24 to 6/60 - 2
- < 6/60 - 3

5. Colour vision by Ishihara's chart

- Normal - 1
- Defective - 2

6. Fields by Automated perimetry

- Normal - 1
- Defective - 2

7. Lateral rectus palsy

- Present – 1
- Absent - 2

8. Presence of systemic illness

9. Past treatment history

10. Body mass index

- a. Healthy 18.5 to 25 - 1
- b. Overweight 25 to 30 - 2
- c. Obese 30 to 35 - 3

11. Fundus examination

- a. Early papilledema - EP
- b. Established papilledema - ESP
- c. Chronic papilledema - CP

d. Resolving papilledema - RP

e. Normal – N

12. Presence of Radiological findings

○ Yes - 1

○ No - 0

13. Fundus photography

14. Treatment – Weight reduction / Medical

15. Visual acuity at follow up

16. Fundus status at follow up

Statistical analysis:

Statistical analysis done by Mean SD, Frequency and Percentage .

RESULTS

1. AGE DISTRIBUTION

AGE	FREQUENCY	PERCENTAGE
21 – 30	9	30%
31 – 40	14	47%
41 – 50	7	23%

TABLE 1 SHOWING AGE DISTRIBUTION IN IIH PATIENTS

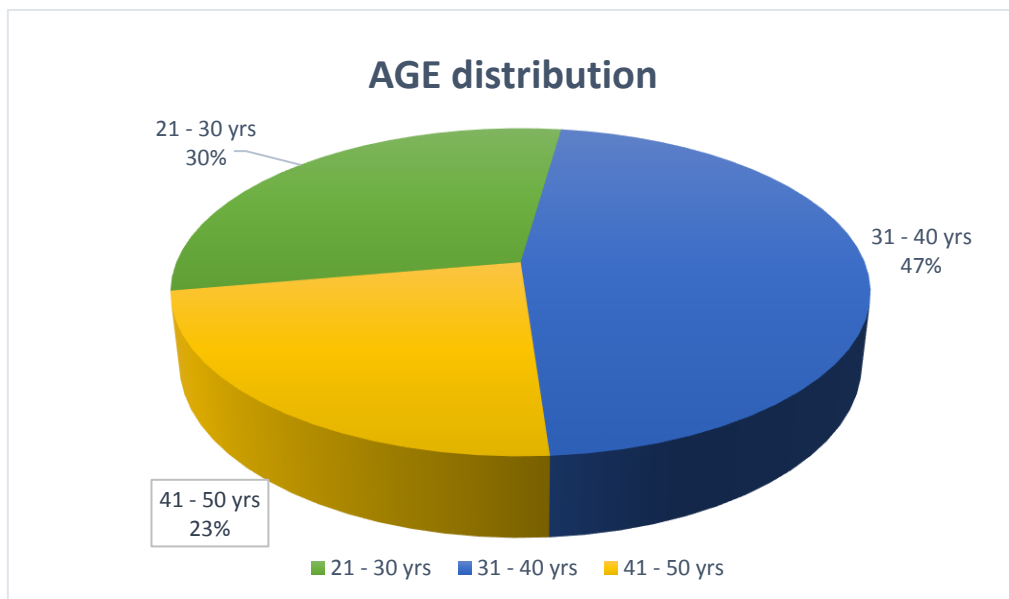


CHART 1 SHOWING AGE DISTRIBUTION IN IIH PATIENTS

INTERPRETATION

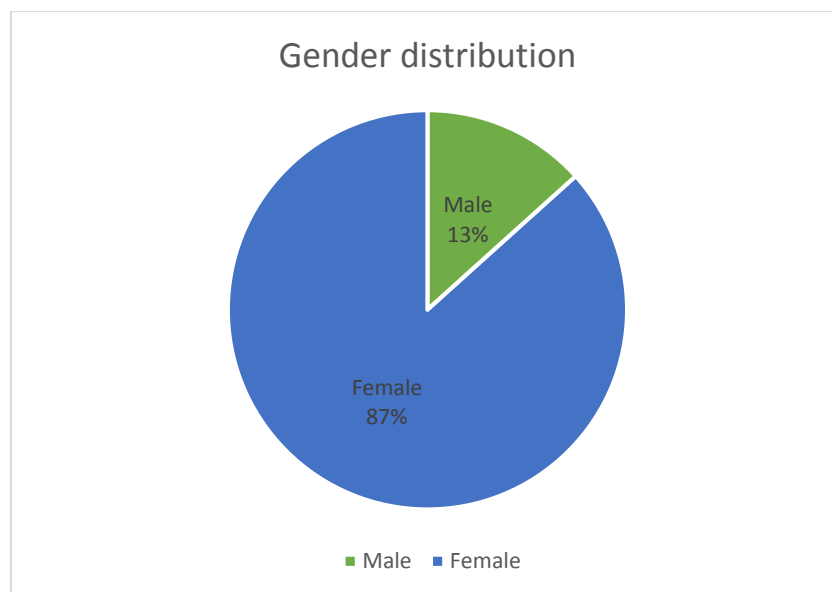
In our study, majority of the patients were between the age group of 31 to 40 years. The mean age is 34.2 years.

2. GENDER DISTRIBUTION

TABLE 2 SHOWING GENDER DISTRIBUTION IN IIH PATIENTS

GENDER	FREQUENCY	PERCENTAGE
MALE	4	17%
FEMALE	26	83%

CHART 2 SHOWING GENDER DISTRIBUTION IN IIH PATIENTS



INTERPRETATION

Among the patients 26 were female, comprising 83% of total sample. Though being rare, we had 4 male patients presented with signs and symptoms of IIH.

3. COMPLAINTS

TABLE 3 SHOWING COMPLAINTS IN IHH PATIENTS

COMPLAINTS	FREQUENCY	PERCENTAGE
Headache	28	93%
Transient obscuration of vision	19	63%
Defective vision	8	26%
Diplopia	11	37%
Nausea / vomiting	13	43%
Tinnitus	10	33%

INTERPRETATION

Symptoms like headache, nausea, vomiting, diplopia, tinnitus were analyzed among 30 patients. Headache was present in 93% of patients and being the presenting symptoms in majority of them.

The headache was pulsatile and associated with nausea, vomiting in 43% of the patients.

Transient visual obscurations was the second common symptom presenting in 63% of the patients.

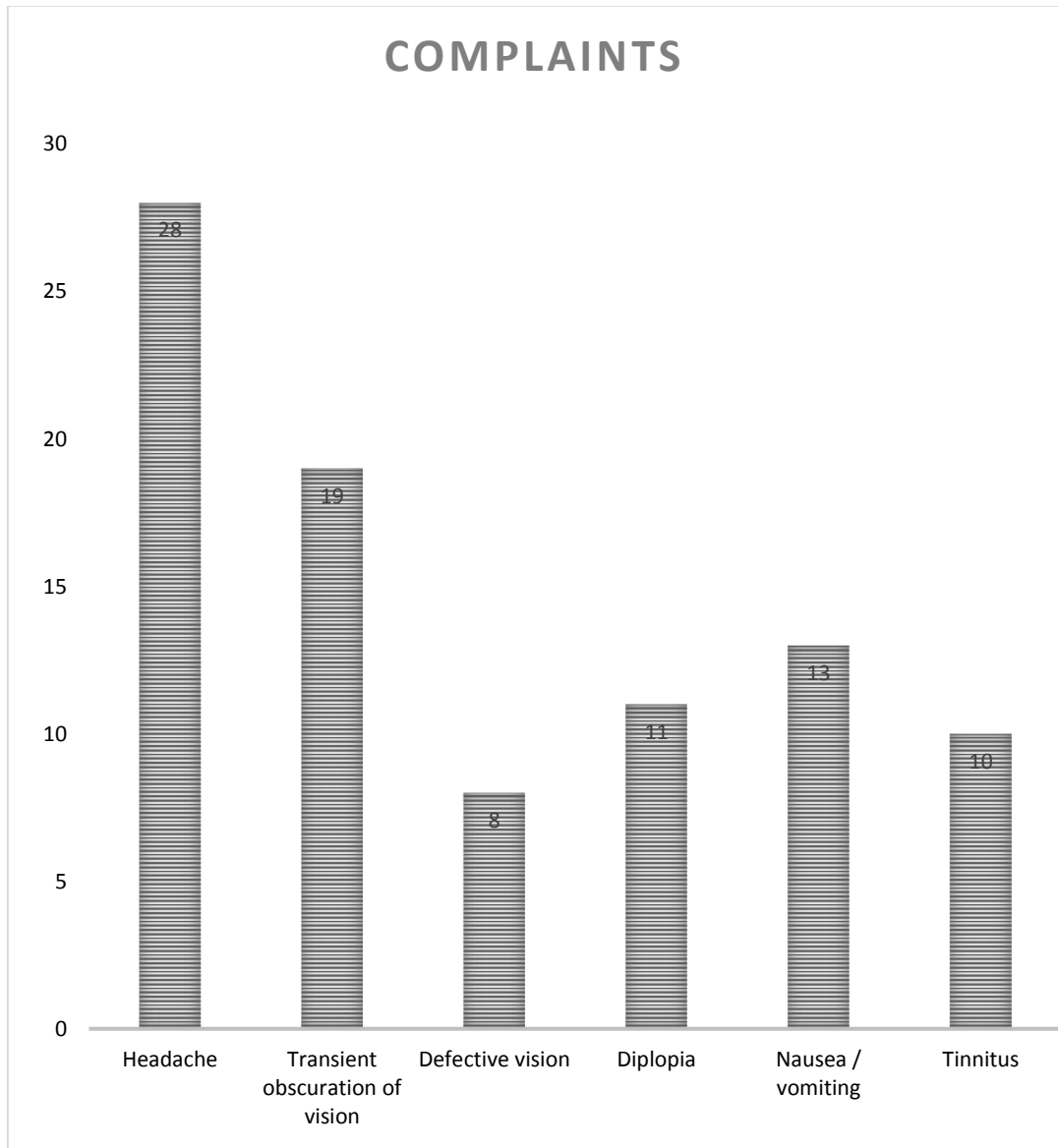


CHART 3 SHOWING COMPLAINTS IN IHH PATIENTS

4. SYSTEMIC DISEASE

TABLE 4 SHOWING SYSTEMIC DISEASES ASSOCIATED WITH IHH

SYSTEMIC DISEASE	FREQUENCY	PERCENTAGE
Hypertension	3	10%
Diabetes mellitus	1	3%
PCOD	3	10%
Hypothyroid	2	6%
HIV	1	3%
Rheumatoid arthritis	1	3%

INTERPRETATION

Hypertension and polycystic ovarian disease with intake of oral contraceptive pills intake together account for 20% of the cases.

Obstructive sleep apnea was found to a risk factor in 2 cases.

One patient presented with a history of fever and intake of Tab Doxycycline for the same prior to the onset of IHH.

5. WEIGHT

TABLE 5 SHOWING MEAN WEIGHT IN IHH PATIENTS

WEIGHT	MEAN SD
Visit 1	68.43 kg
Visit 2	66.37 kg
Visit 3	65.18 kg

6. BODY MASS INDEX

Body mass index is calculated from the formula:

Body mass index = Weight in Kg / Height in meter square

TABLE 6 SHOWING CLASSIFICATION OF OBESITY

CLASSIFICATION	BMI
NORMAL	18.5 - 24.9
OVERWEIGHT	25.0 – 29.9
OBESE	➤ 30

TABLE 7 SHOWING RELATIONSHIP OF BMI WITH IHH

BMI	FREQUENCY	PERCENTAGE
Healthy	5	18%
Overweight	7	26%
Obese	18	56%

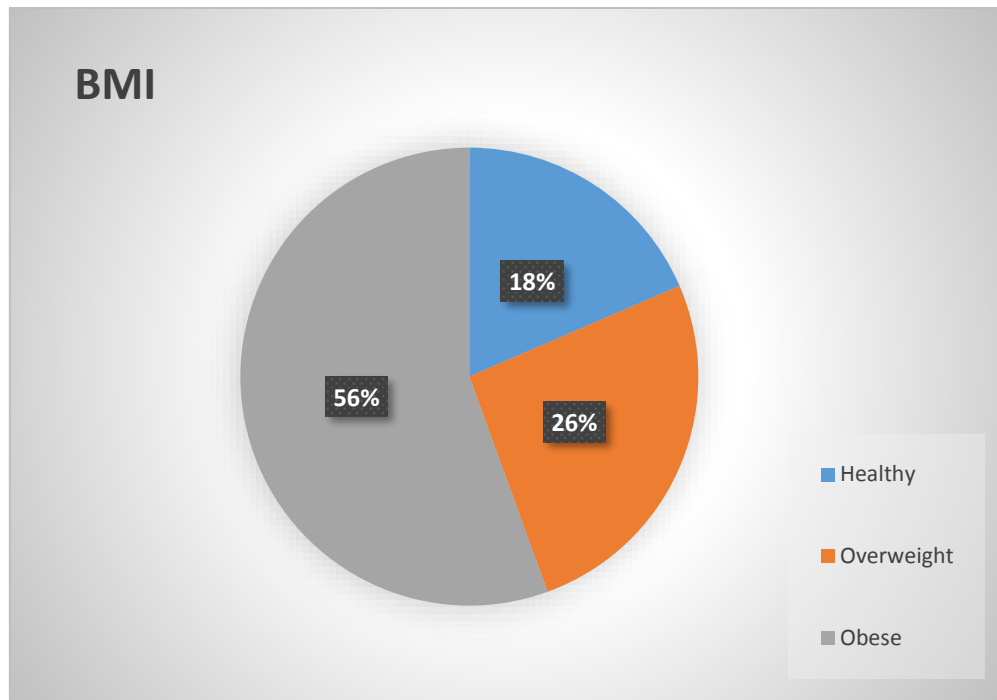


CHART 4 SHOWING RELATIONSHIP OF BMI WITH IHD

INTERPRETATION

Majority of the patients (72%) presented were obese and overweight.

OCULAR EXAMINATION DATA

7. BEST CORRECTED VISUAL ACUITY

TABLE 8 SHOWING VISUAL ACUITY IN IIH PATIENTS

VISUAL ACUITY	Visit 1	Visit 2	Visit 3
6/6 – 6/18	46 [77%]	45 [83%]	40 [87%]
6/24 – 6/60	13 [22%]	9 [17%]	6 [13%]
Less than 6/60	1 [0.02%]	-	-
Total eyes tested	60	54	46

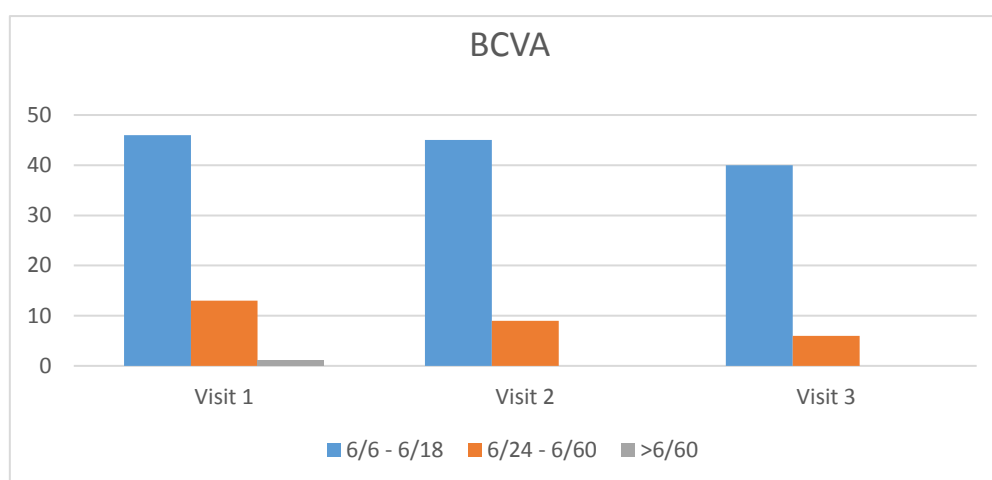


CHART 5 SHOWING VISUAL ACUITY IN IIH PATIENTS

INTERPRETATION

Vision was grossly unaffected in most of the patients. Only 22% of patients had vision less than 6/18.

Paired Samples Statistics

	Mean	N	Std. Deviation	Std. Error Mean
Pair 1 PRE.VISION	.4180	46	.18959	.02795
POST.VISION	.6526	46	.24263	.03577

Paired Samples Correlations

	N	Correlation	Sig.
Pair 1 PRE.VISION & POST.VISION	46	.721	.001

8. EOM

TABLE 9 SHOWING EOM IN IIIH PATIENTS

EOM	FREQUENCY	PERCENTAGE
Full	22	71%
Abduction restriction	8	29%

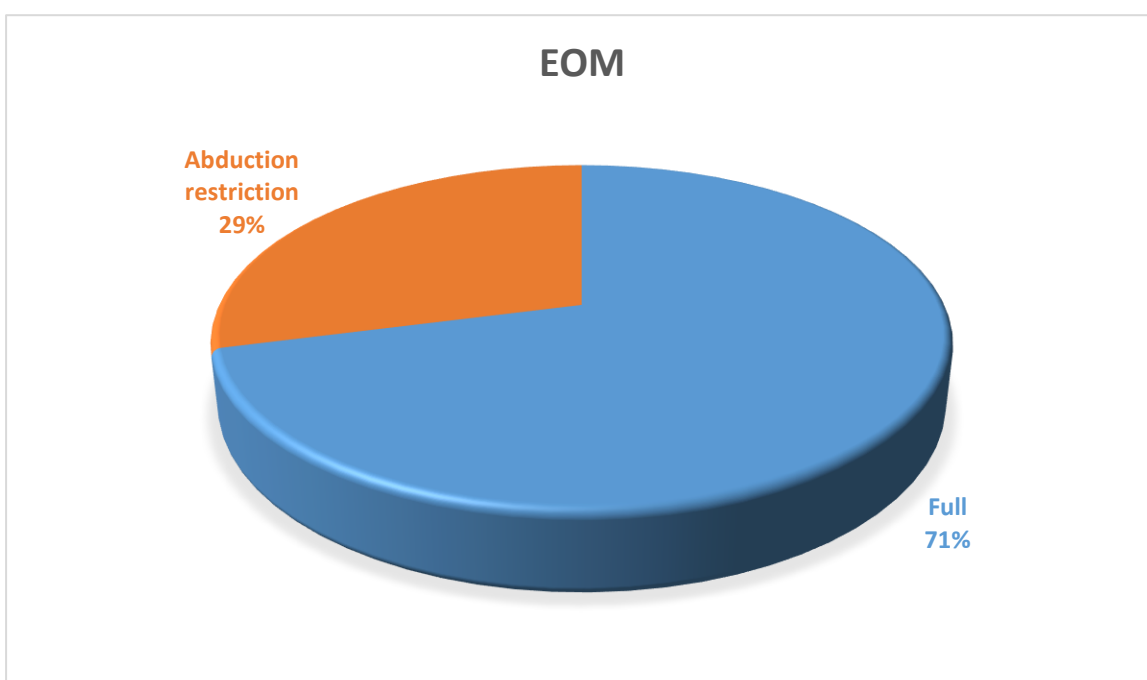


CHART 6 SHOWING EOM IN IIIH PATIENTS

INTERPRETATION

29% of patients presented with complaints of restriction of abduction in either eye.

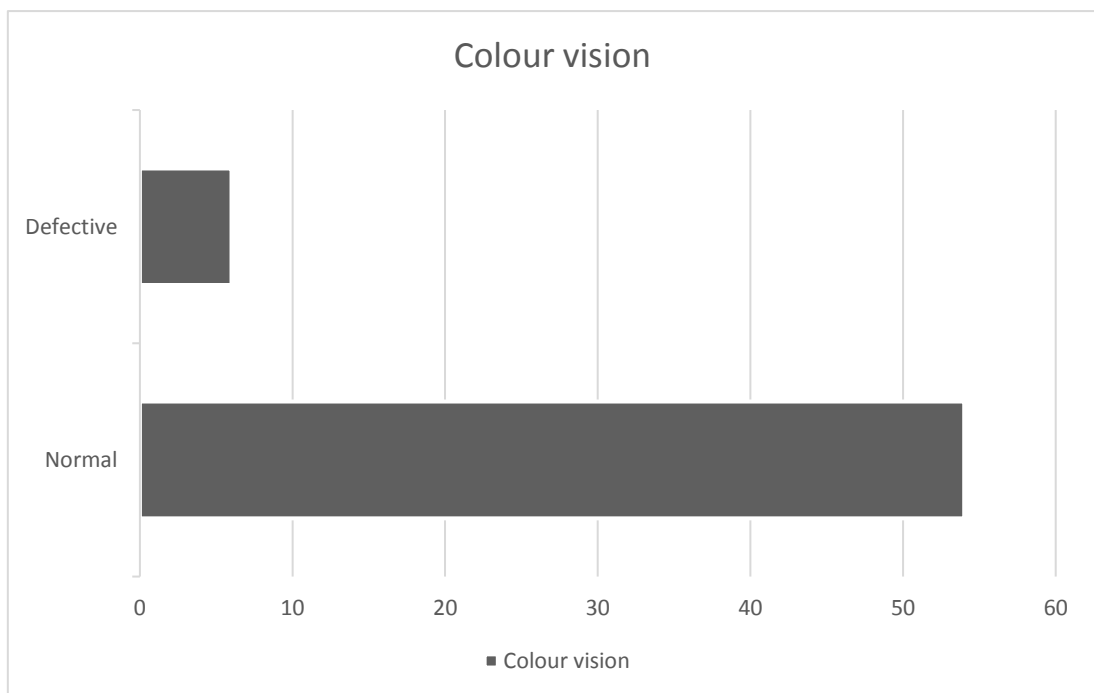
Some of these affected individuals presented with diplopia as the presenting complaint to the hospital.

9. COLOUR VISION

TABLE 10 SHOWING COLOUR VISION IN IHH PATIENTS

COLOUR VISION	FREQUENCY	PERCENTAGE
Normal	53	88%
Defective	7	12%

CHART 7 SHOWING COLOUR VISION IN IHH PATIENTS



INTERPRETATION

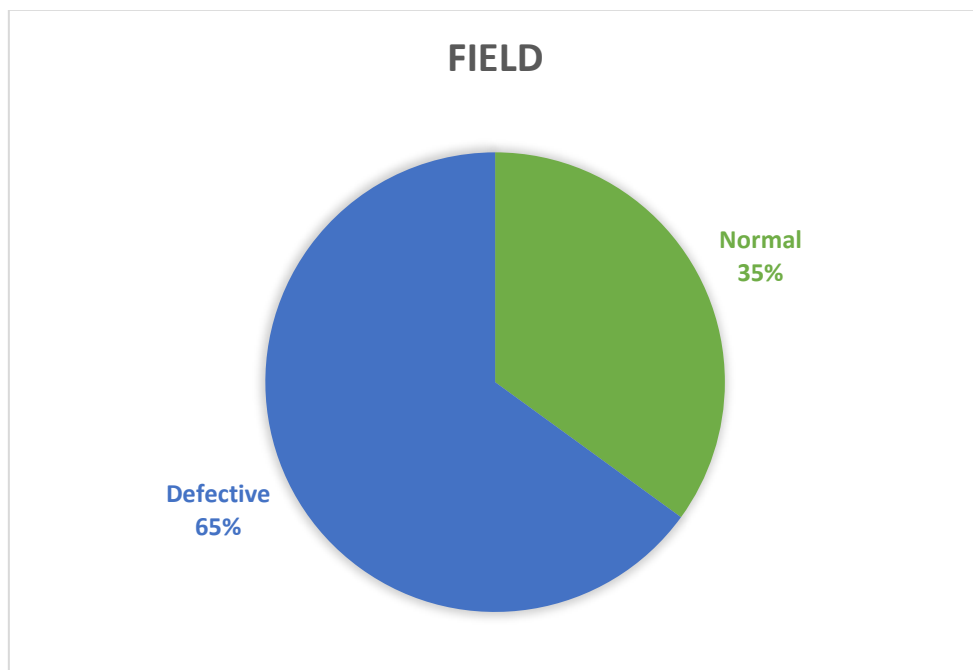
Colour vision was affected in 12% of the eyes tested.

10. FIELDS

TABLE 11 SHOWING VISUAL FIELDS IN IIH PATIENTS

FIELDS	FREQUENCY	PERCENTAGE
Normal	21	35%
Defective	39	65%

CHART 8 SHOWING VISUAL FIELDS IN IIH PATIENTS



INTERPRETATION

Visual field was defective in 65% of the eyes tested with automated perimetry.

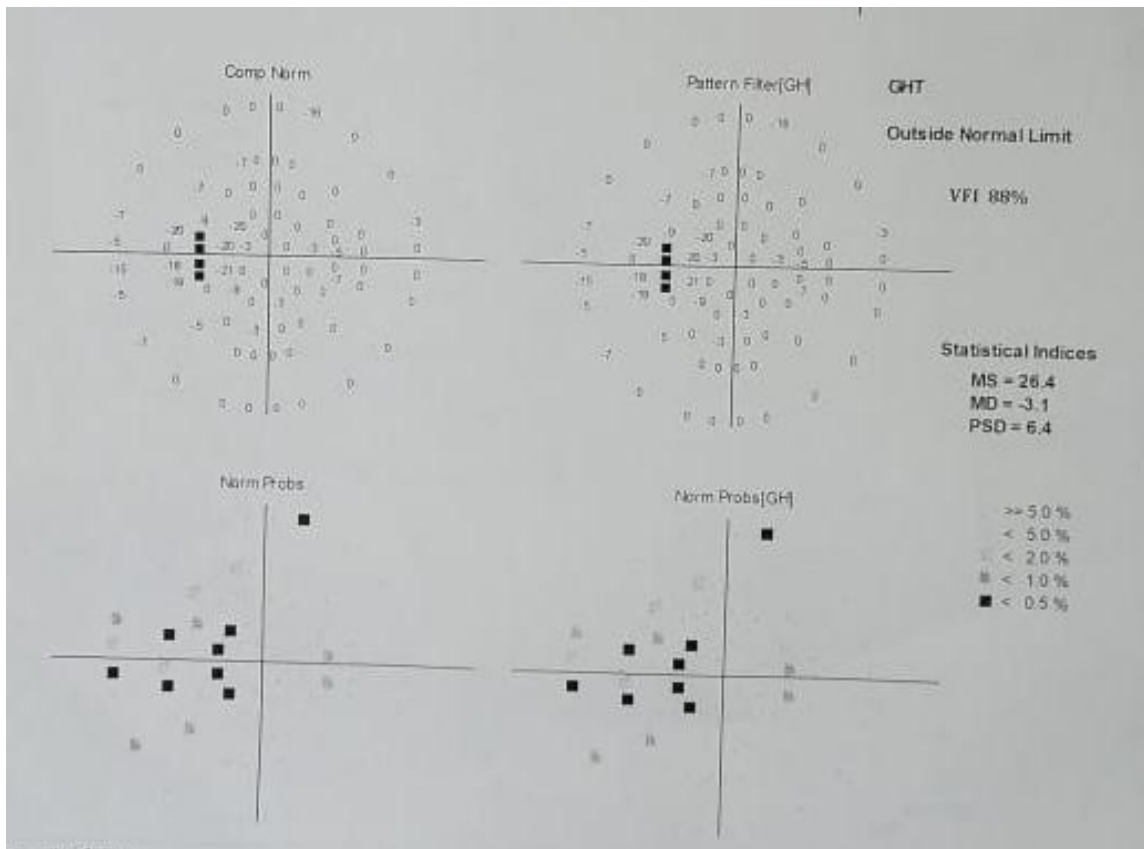


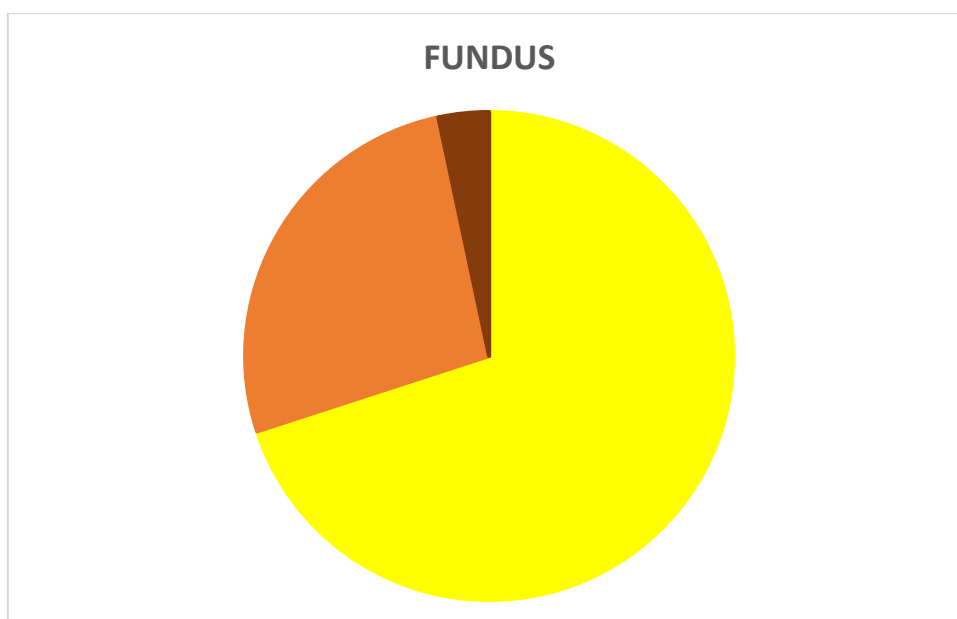
Figure 23: BLIND SPOT ENLARGEMENT

11. FUNDUS

TABLE 12 SHOWING FUNDUS FINDINGS IN 1ST VISIT

FINDINGS	FREQUENCY	PERCENTAGE
Early papilledema	42	70%
Established papilledema	16	27%
Chronic papilledema	2	3%

CHART 9 SHOWING FUNDUS FINDINGS IN 1ST VISIT



INTERPRETATION

Majority of patients presented with early papilledema [70%] followed by established papilledema [27%]



Figure 24: EARLY PAPILLEDEMA



Figure 25: ESTABLISHED PAPILLEDEMA

TABLE 13 SHOWING FUNDUS FINDINGS IN 2ND VISIT

FINDINGS at Visit 2	FREQUENCY	PERCENTAGE
Early papilledema	2	4%
Established papilledema	14	26%
Chronic papilledema	1	2%
Resolving papilledema	29	54%
Normal	8	15%

CHART 10 SHOWING FUNDUS FINDINGS IN 2ND VISIT

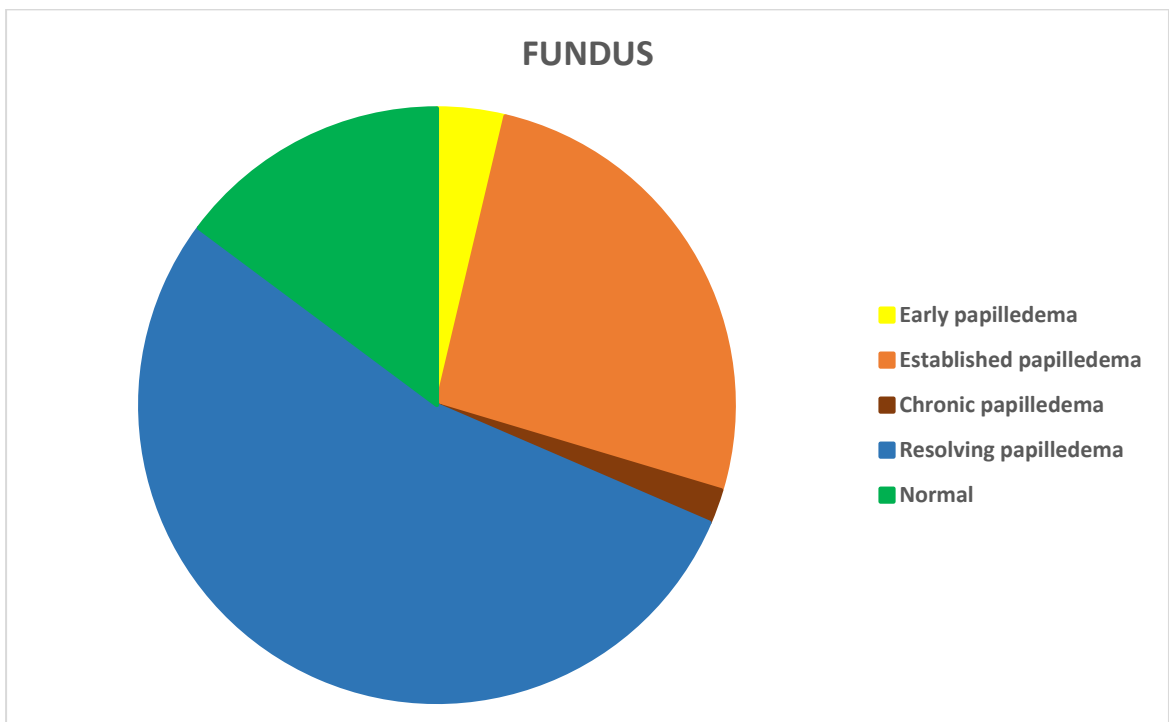


TABLE 14 SHOWING FUNDUS FINDINGS IN 3RD VISIT

FINDINGS at Visit 3	FREQUENCY	PERCENTAGE
Early papilledema	0	0
Established papilledema	2	4%
Chronic papilledema	1	2%
Resolving papilledema	19	42%
Normal	24	51%

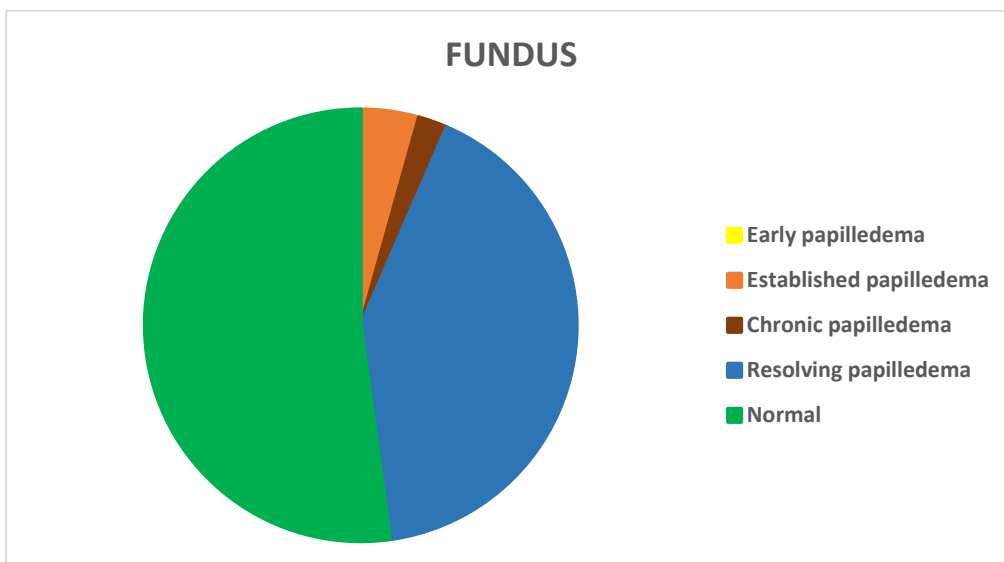


CHART 11 SHOWING FUNDUS FINDINGS IN 3RD VISIT

INTERPRETATION

During subsequent visits, the percentage of eyes with resolving papilledema increases showing the response to treatment.

At the final visit, nearly half the patients had normal fundus appearance with majority of the remaining eyes had resolving stage of papilledema.

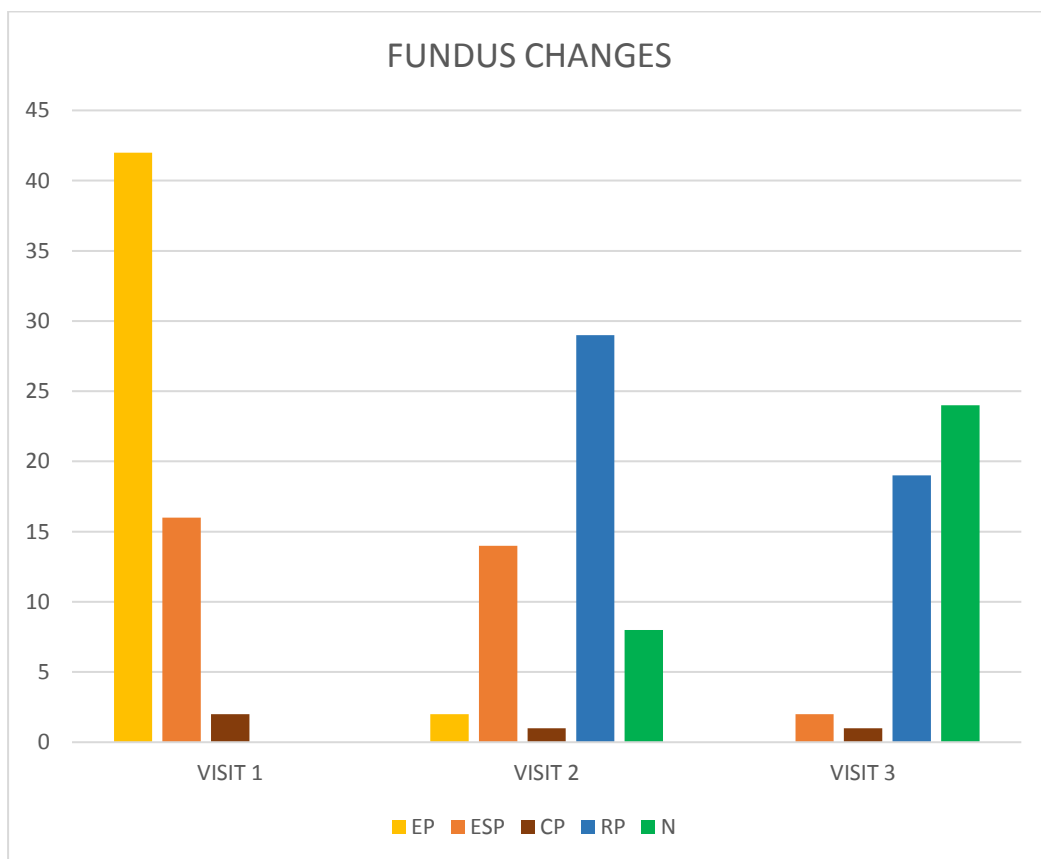


CHART 12 SHOWING FUNDUS FINDINGS IN ALL VISITS

Paired Samples Statistics

	Mean	N	Std. Deviation	Std. Error Mean
Pair 1 PRE.FUNDUS	.5435	46	.09105	.01342
POST.FUNDUS	1.0217	46	1.06602	.15718

Paired Samples Correlations

	N	Correlation	Sig.
Pair 1 PRE.FUNDUS & POST.FUNDUS	46	.077	.061

12. NEUROIMAGING

TABLE 15 SHOWING NEUROIMAGING FINDINGS IN IIH

FINDINGS	FREQUENCY	PERCENTAGE
Normal	6	20%
Partial empty sella	15	50%
Perioptic space widening	16	53%
Flattening of posterior globe	5	17%
Transverse / sigmoid sinus stenosis	8	27%
Tortuosity of optic nerve	4	13%

INTERPRETATION

Ultrasonography, CT, MRI with MRA and MRV were taken for the patients.

Half of the patients had findings of empty sella and perioptic space widening in the imaging results.

Isolated or combined Sigmoid and Transverse sinus stenosis was the next most common finding.

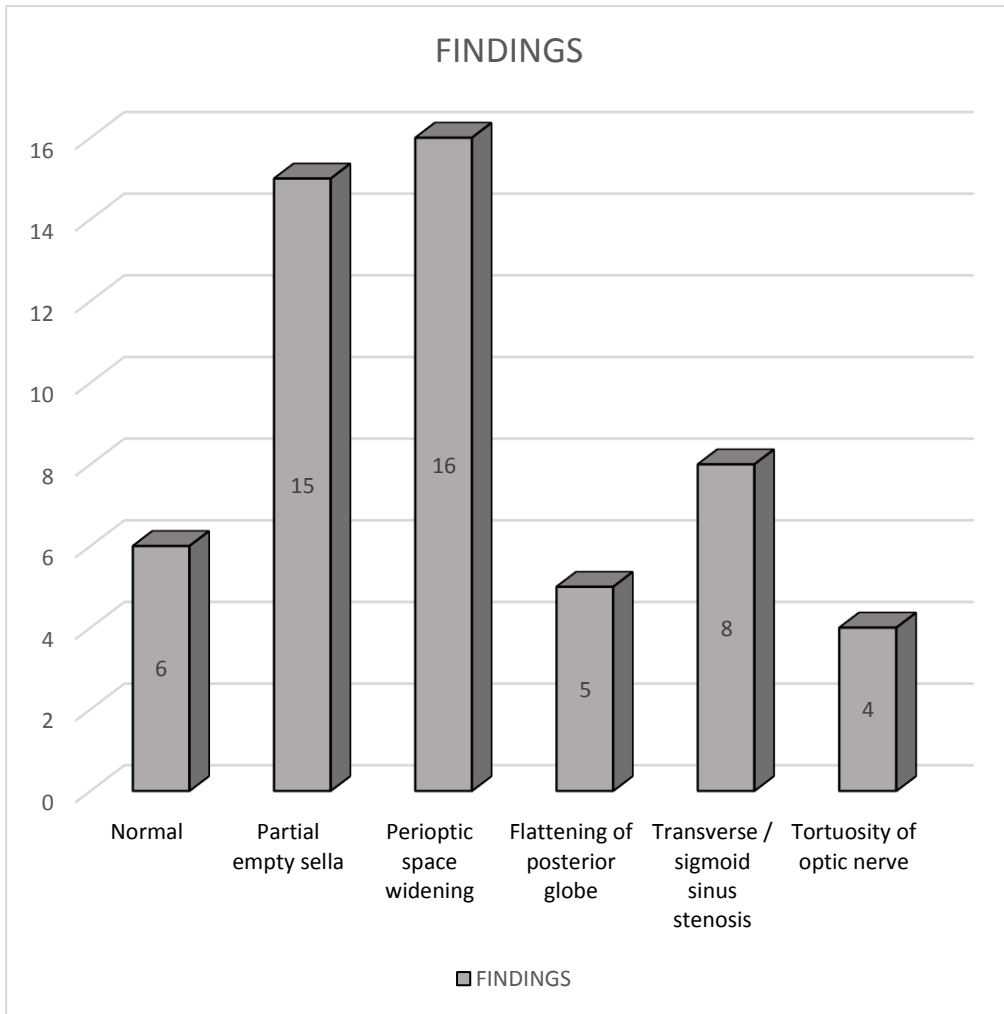


CHART 13 SHOWING NEUROIMAGING FINDINGS IN IIIH

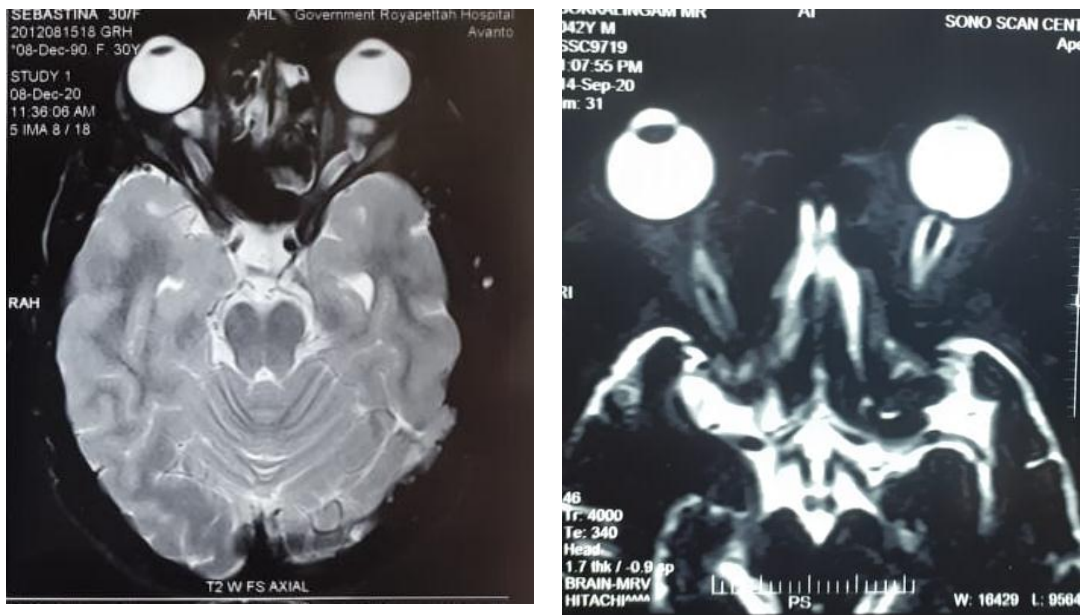


Figure 26: A] TORTUOSITY OF OPTIC NERVE, B] THICKENING OF OPTIC NERVE SHEATH

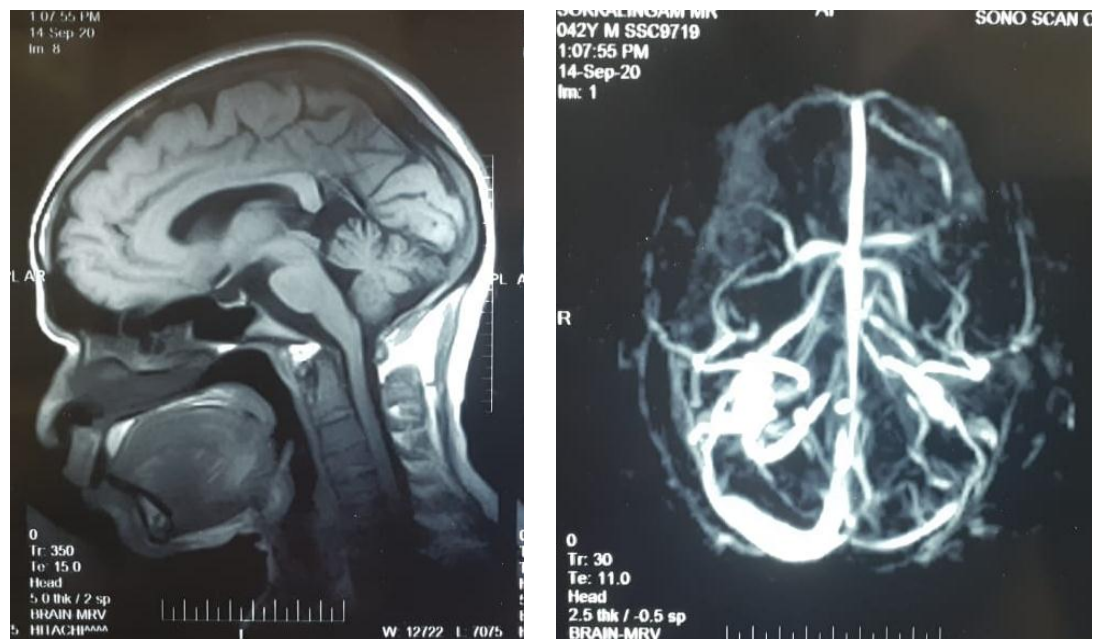


Figure 27: A] EMPTY SELLA, B] TRANSVERSE SINUS THROMBOSIS

13. TREATMENT

TABLE 16 SHOWING TREATMENT IN IIIH

TREATMENT	FREQUENCY	PERCENTAGE
WEIGHT REDUCTION & DIAMOX	17	57%
DIAMOX ALONE	13	43%

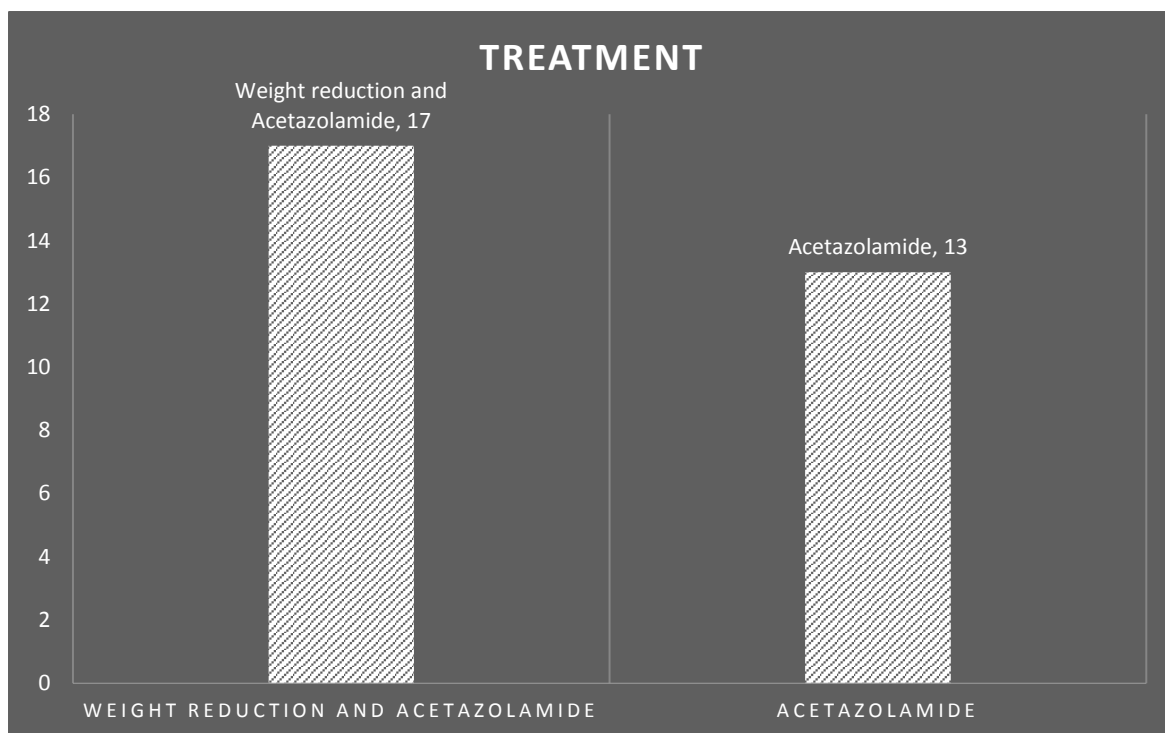


CHART 14 SHOWING TREATMENT IN IIIH

INTERPRETATION

Weight reduction was advised in 57% of the patients who were obese and overweight. Tablet Acetazolamide was started for all patients and advised to follow up at 1 week, 1 month and 6 months of treatment.

DISCUSSION

In our study, 30 patients of Idiopathic intracranial hypertension presented to Regional Institute of Ophthalmology are analyzed.

In our study, majority of the patients were between the age group of 31 to 40 years. The mean age is 34.2 years. The mean age was 32.89 years³ in a study conducted among 50 patients by **Ambika S et al.**

Among the patients 26 were female, comprising 83% of total sample. **Michael Wall et al** found that 92% of the patients were females⁴. A study by **Craig et al** (2001) showed a female to male ratio²³ of 5.7:1 which was comparable to ratio of 6.5:1

Symptoms like head ache, nausea, vomiting, diplopia, tinnitus were analyzed among 30 patients. Head ache was present in 93% of patients, being the presenting symptoms in majority of them. Transient visual obscurations was the second common symptom presenting in 63% of the patients.

A study by **Binder et al**²⁴ has supported this study with more than 90% of the patients having headache as presenting symptom. In study of IHH patients by **Michael Wall et al**, 72% of patients complained of visual obscurations.

Hypertension and polycystic ovarian disease with intake of oral contraceptive pills intake together account for 20% of the cases. **KP Kilgore et al** studied the association between oral hormonal contraceptive use²⁵ and idiopathic intracranial hypertension in 53 patients.

One patient presented with a history of fever and intake of Tab Doxycycline for the same prior to the onset of IIH. **Friedman et al** studied the occurrence of IIH²⁷ among 7 patients taking doxycycline for acne and febrile illness.

Majority of the patients presented were obese [56%] and overweight. A matched case control study by **Daniels AB et Al** found that higher BMIs²⁷ and higher weight in the recent past were associated with greater risk of IIH.

Vision was grossly unaffected in most of the patients. Only 22% of patients had vision less than 6/18.

29% of patients presented with complaints of restriction of abduction in either eye. Some of these affected individuals presented with diplopia as the presenting complaint to the hospital. **Michael Wall** has reported horizontal diplopia in 1/3rd of IIH patients and abducens nerve paresis were found in 10 to 20% of cases.

Visual field was defective in 65% of the eyes tested with automated perimetry. **FJ Rowe et al** assessed the visual function in cases of IIH²⁸ and found that 75% of the patients and 63% of the eyes had field defects on testing with automated perimetry. Commonest finding in perimetry is enlargement of blind spot.

Majority of patients presented with early papilledema [53%] followed by established papilledema [40%]. During subsequent visits, the percentage of eyes with resolving papilledema increases showing the response to treatment.

On imaging, half of the patients had findings of empty sella and perioptic space widening. Isolated or combined Sigmoid and Transverse sinus stenosis was the next most common finding. In a retrospective case series by **Brodsky MC** and **Vaphiades M**²⁹, the MRI showed flattening of posterior sclera in 80%, findings of empty sella in 70%, distension of the perioptic space in 45%, enhancement of optic nerve in 50%, intraocular protrusion of the optic nerve in 30% and tortuosity of the orbital optic nerve in 40% of the patients.

Weight reduction with Tablet Acetazolamide was advised in 57% of the patients who were obese and overweight. Tablet Acetazolamide 250mg BD was started for all other patients and reviewed periodically for assessment of visual function and fundus examination. Majority of the patients improved with medical management and weight reduction.

CONCLUSION

Idiopathic intracranial hypertension is characterized by elevated CSF pressure without an identifiable cause. Obese women of child-bearing age are more commonly affected than males. Most common presenting complaint was headache.

Commonly associated risk factors are obesity, hypertension, thyroid dysfunction, and drug intake like tetracyclines, oral contraceptives and thyroid replacement therapy. High body mass index of more than 30 is associated with IIH.

Visual acuity was normal in most eyes. Sixth nerve paresis occur as a false localizing sign of IIH. Most of the patients in our study presented with early papilledema. Most common visual field defect is an enlarged blind spot detected by Automated perimetry.

In MRI imaging, empty sella and perioptic space widening were the most common findings in our patients. In MRV study, one fourth of patients had transverse sinus stenosis and sigmoid sinus stenosis.

Patients who had high BMI were treated with Tablet Acetazolamide 250mg BD along with Weight reduction. Follow up at one month and at six months showed resolving papilledema in most cases.

Visual acuity showed better improvement post treatment with variables showing good significance.

Any patients presenting with headache should be evaluated for ocular causes of headache like refractive error, if found to be normal, they should be evaluated for Idiopathic intracranial hypertension.

Early detection with initiation of appropriate treatment prevent visual loss. Since most of the patients in our study are detected with early papilledema, we conclude that earlier the treatment either surgical or medical, the better the visual prognosis.