

# IDIOPATHIC INTRACRANIAL HYPERTENSION

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# IDIOPATHIC INTRACRANIAL HYPERTENSION

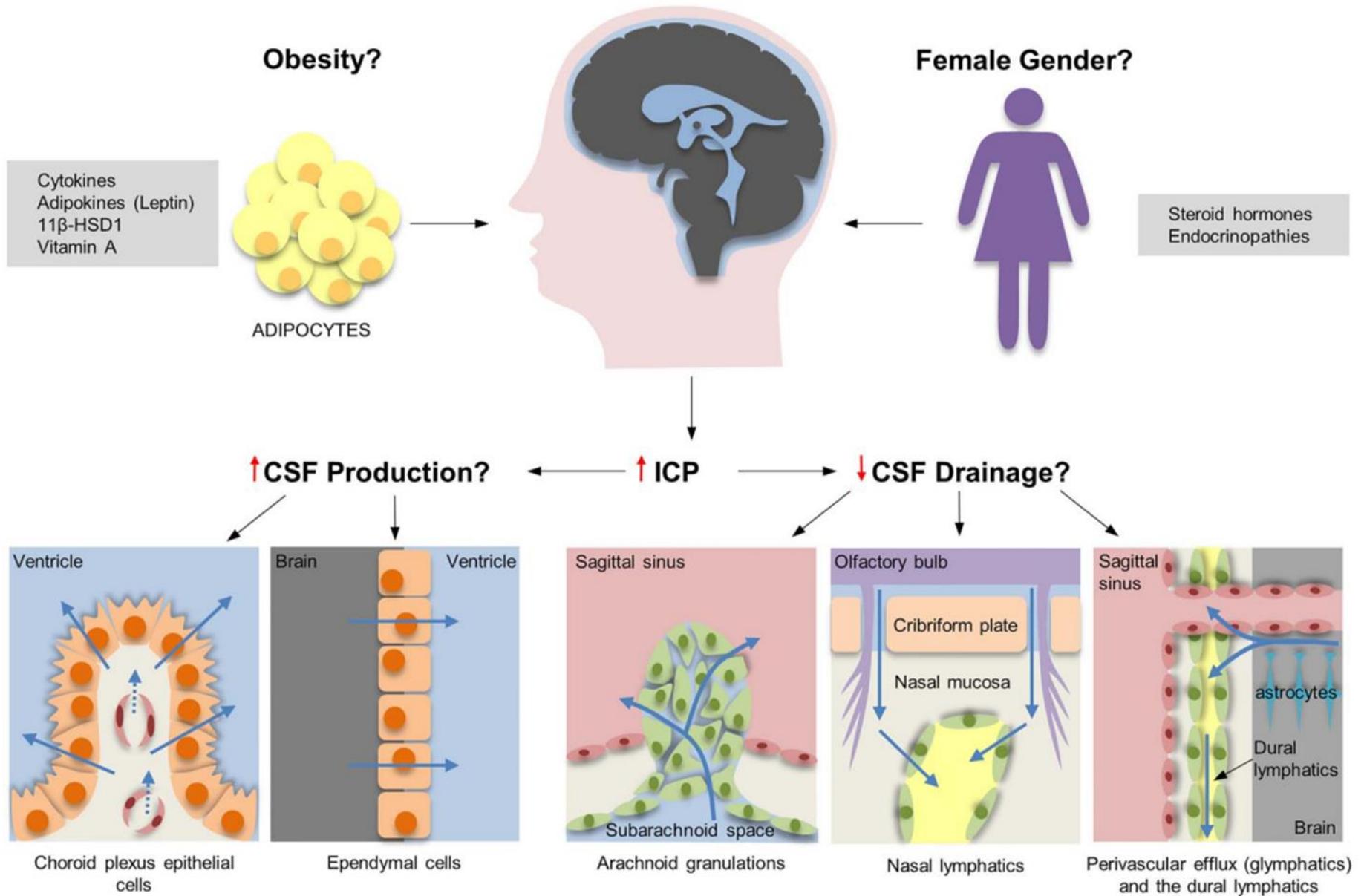
**Idiopathic intracranial hypertension (IIH)** is a syndrome characterized by elevated intracranial pressure that most commonly affects obese women during their reproductive years.

## Epidemiology

1 and 3/100 000/year in the general population. When stratified for reproductive age, female gender and weight, the incidence rises by 12–28/ 100 000/year.

## Pathophysiology (underlying pathogenesis is uncertain)

- 1) Due to increased CSF production (increased activity of choroid plexus)
- 2) Reduced CSF drainage: Increased CSF outflow resistance due to decreased absorption via the arachnoid granulation.



## Clinical Features

The symptoms of IIH patients are

headache (94%)

transient visual obscurations (68%)

pulse synchronous tinnitus (58%)

photopsia (54%)

retrobulbar pain (44%)

Diplopia(38%)

visual loss (30%)

**Headache** :- severe daily pulsatile headaches may awaken the patient and usually last hours. The headache is often reported as the worst head pain associated with nausea.

**Visual obscurations** :- transient blurred vision that usually last less than 30 seconds, followed by visual recovery to baseline.  
monocular or binocular presentation.  
Transient ischemia of the optic nerve head due to increased tissue pressure.

**Diplopia** :- Abducent nerve palsy

**Pulsatile intracranial noises** or **pulse synchronous tinnitus** The sound is often unilateral.  
Jugular compression or head turning ipsilateral to the sound abolishes it.  
Transmission of intensified vascular pulsations, increased turbulence through smooth walled venous stenoses related to transverse sinus collapse from high CSF pressure

## Signs:-

### **Papilledema**

Optic disc edema due to increased intracranial pressure is the **cardinal sign** of IIH.

**Horizontal diplopia** due to sixth nerve palsies (esotropia)

### **Sensory visual function**

Visual acuity usually remains normal in patients with papilledema, when the condition is long-standing and severe or if there is a serous retinal detachment and optic disc edema extends to the macula.

# Evaluation

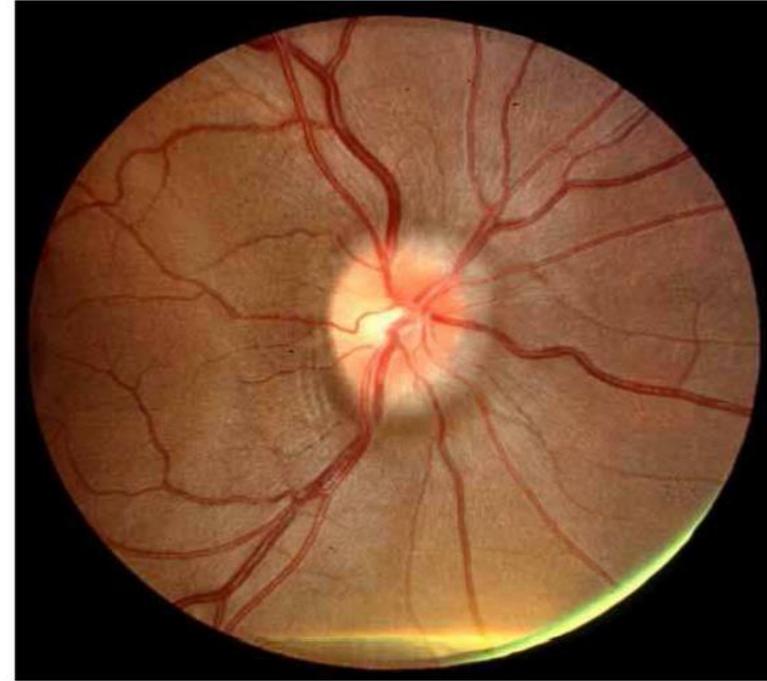


# OPHTHALMOSCOPY:- FRISEN GRADES

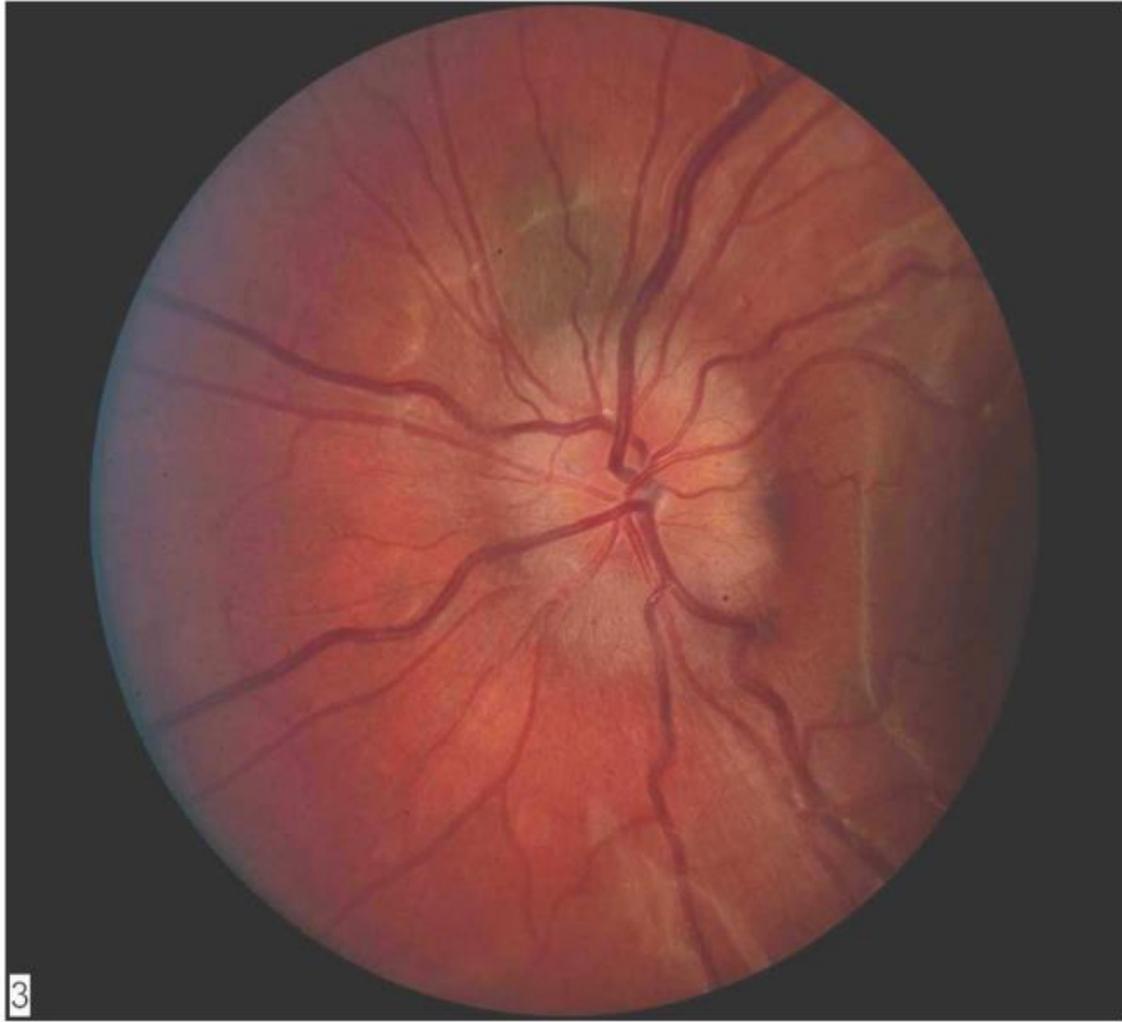
## Modified Frisén Papilledema Scale



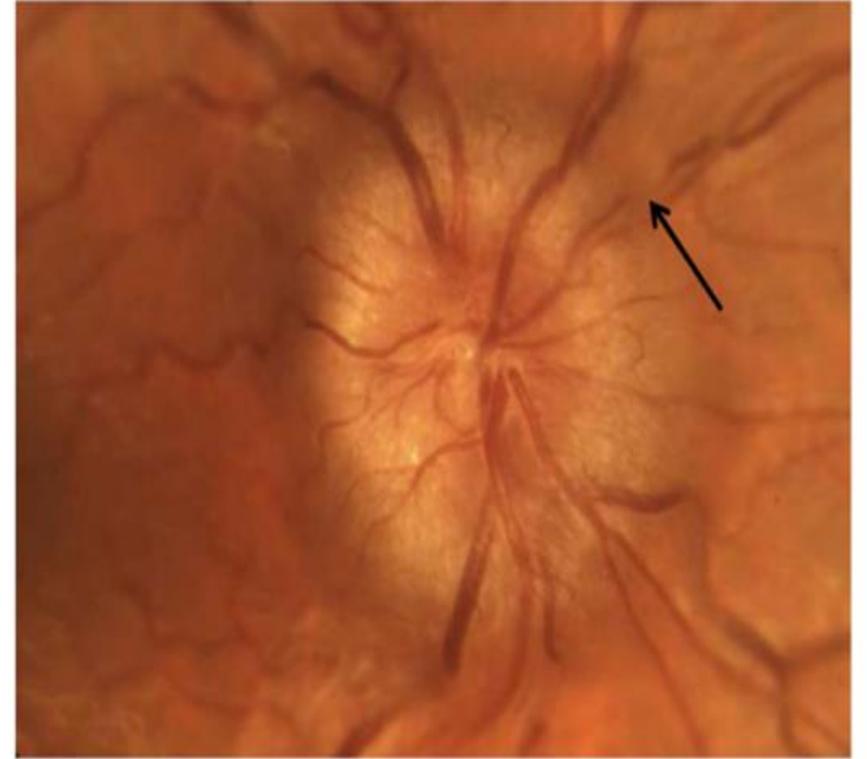
Grade 0: No disc edema



**Figure 3.** characteristic "C-shaped halo" with a temporal gap surrounding the disc of early of (Frisén grade 1) papilledema.



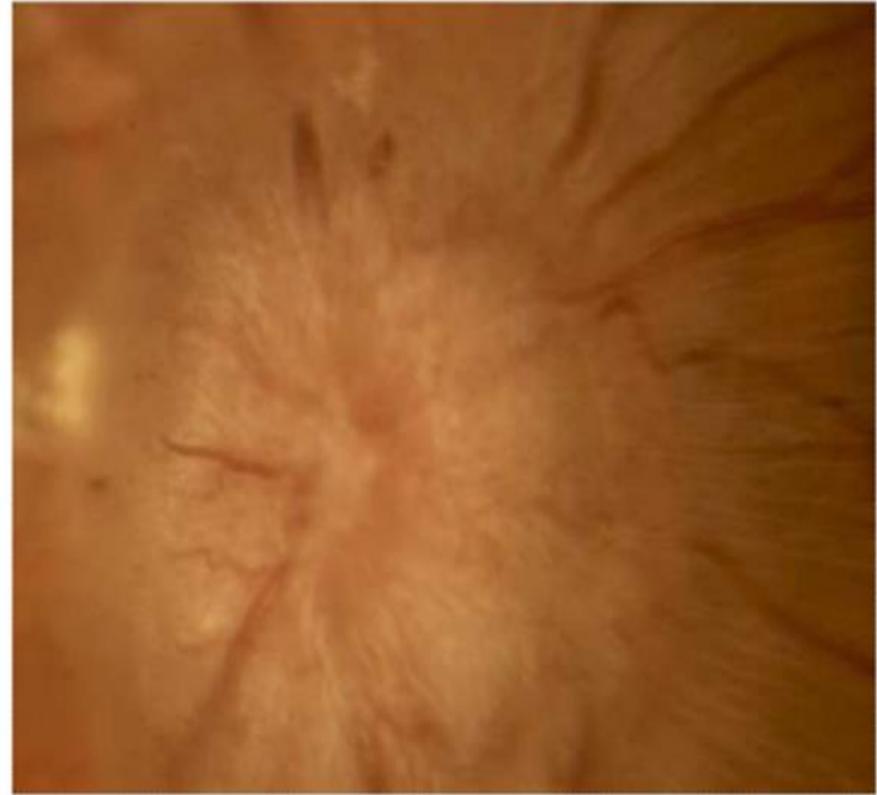
**Figure 4.**  
With grade II papilledema the halo becomes circumferential.



**Figure 5.**  
Grade III papilledema is characterized by Loss of major vessels as they leave the disc (arrow).



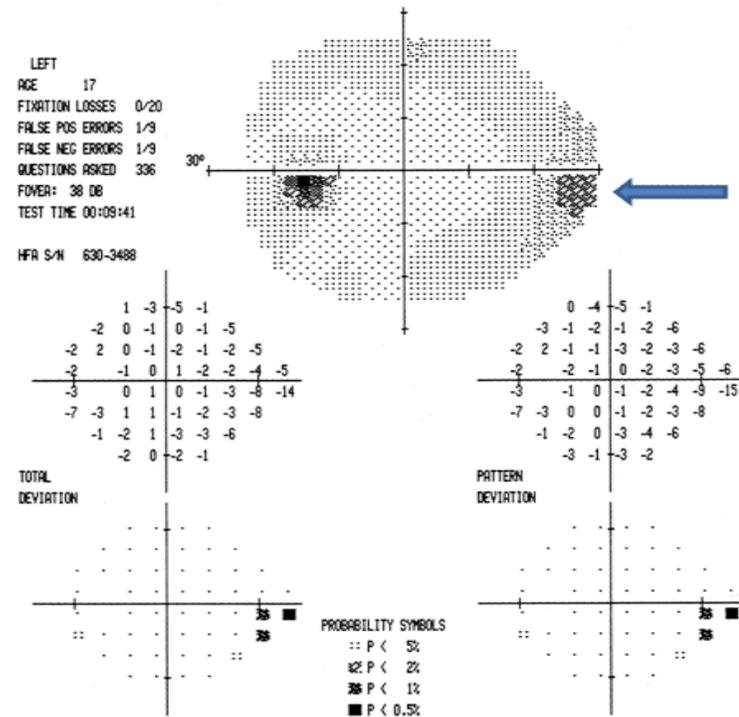
**Figure 6.**  
Grade 4 papilledema is characterized by Loss of major vessels on the disc.



**Figure 7.**  
Grade V has the criteria of Grade IV plus partial or total obscuration of all vessels on the disc.

# Perimetry

Enlargement of the physiologic blind spot  
Loss of inferonasal portions of the visual field



**Figure 8.**  
A typical inferonasal step defect (arrow) of early optic disc edema in IIIH.

## Grades of visual field progression

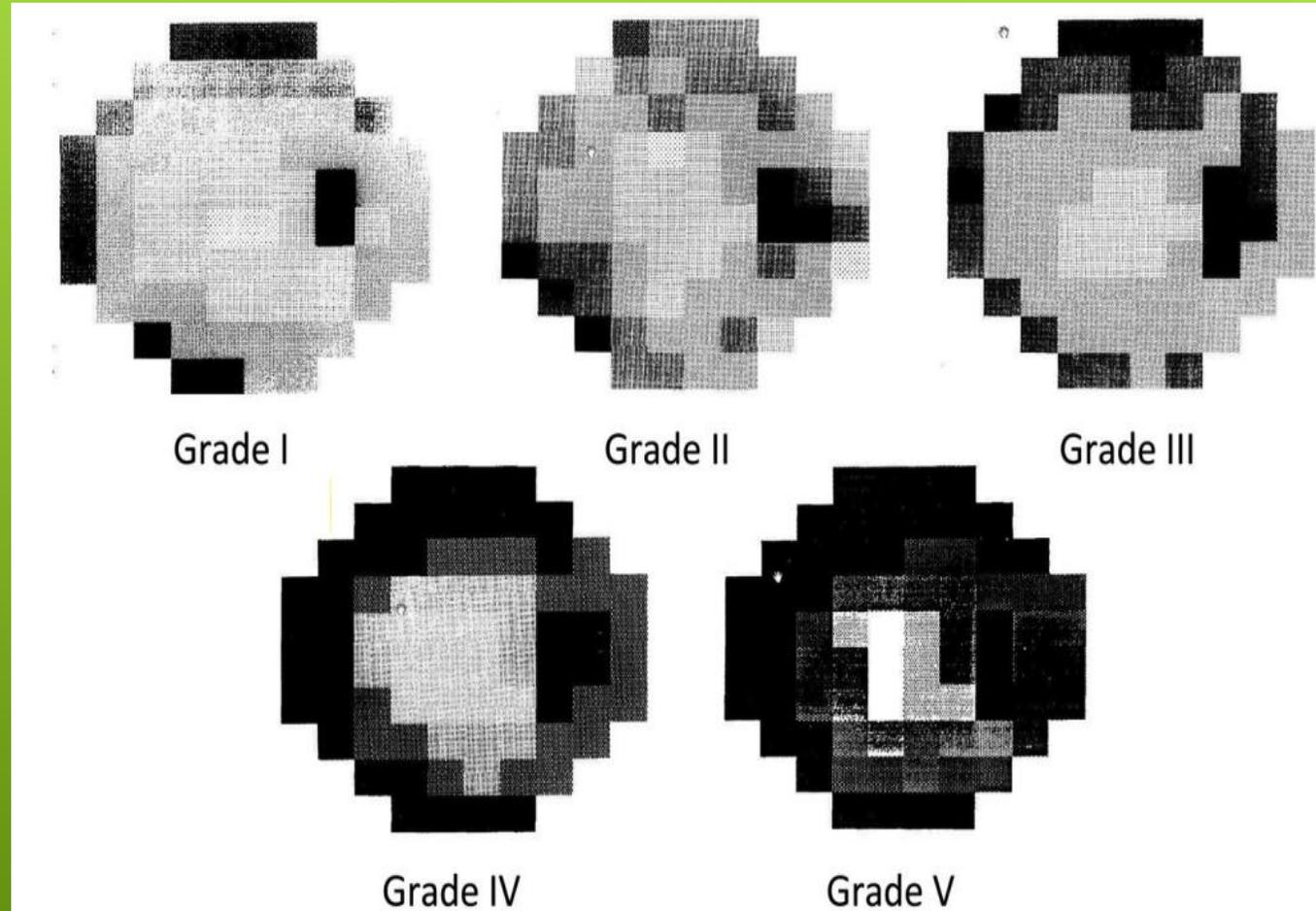
Grade 1:- Enlarged blind spot

Grade 2:- Inferior nasal step

Grade 3:- Arcuate scotomas

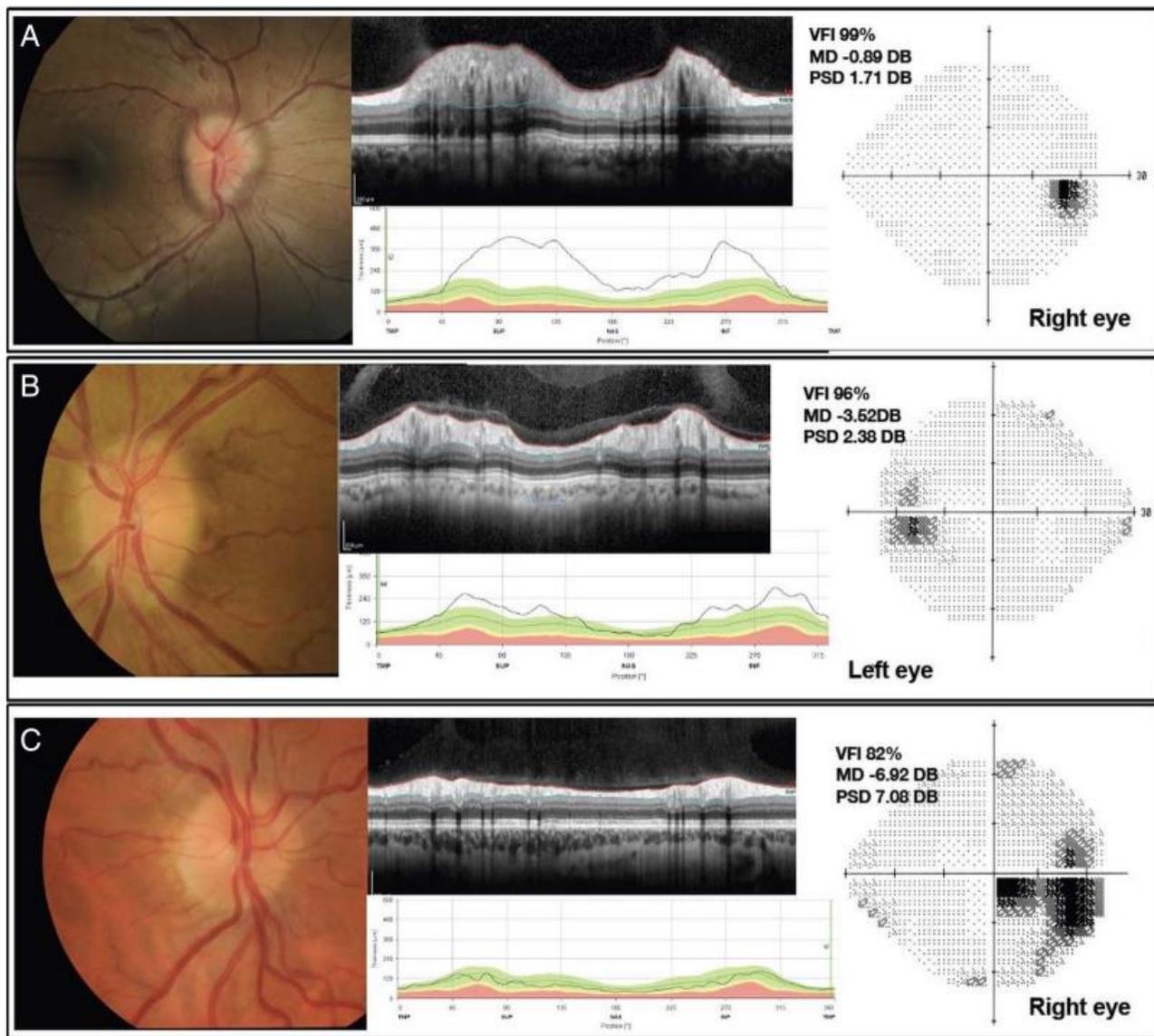
Grade 4:- Constriction

Grade 5:- General depression

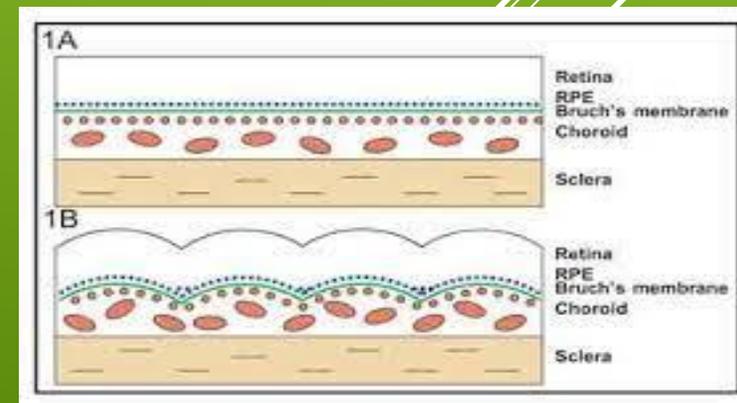


**Figure 9.**

Grades of visual loss in IIH found by grading the visual field examinations and then averaging the values from within each grade.



The fine black line depicting the patient's data, and the block colors (green, yellow, red) showing the normative data. **Patient A** with Frisen grade 3, there is obscuration of more than one major vessel leaving the disc, there is a circumferential halo and elevation of all borders. Paton's lines (curvilinear chorioretinal folds adjacent to the temporal (left hand side) of the optic disc margin) are evident. **Patient B**, with IIH with Frisen grade 2. Circumferential halo and there is no major vessel obscured. **Patient C**, a treated patient with IIH who has resolved papilledema (Frisen grade 0). There is **reduction** in the RNFL thickness which is due to resolution of swelling, but also axonal loss. There is a centrocecal visual field defect. Optical chromatography scan can identify the subtle changes in the retina i.e. frisen grade 1 and grade 2



## OPTICAL COHERENCE TOMOGRAPHY



**Figure 11.**

Gross pathologic specimen of optic nerve (central core), optic nerve sheath and arachnoid trabeculations in between (from Sergott et. al.<sup>73</sup>) Note the well-developed series of arachnoid trabeculations and the fully unfolded optic nerve sheaths.

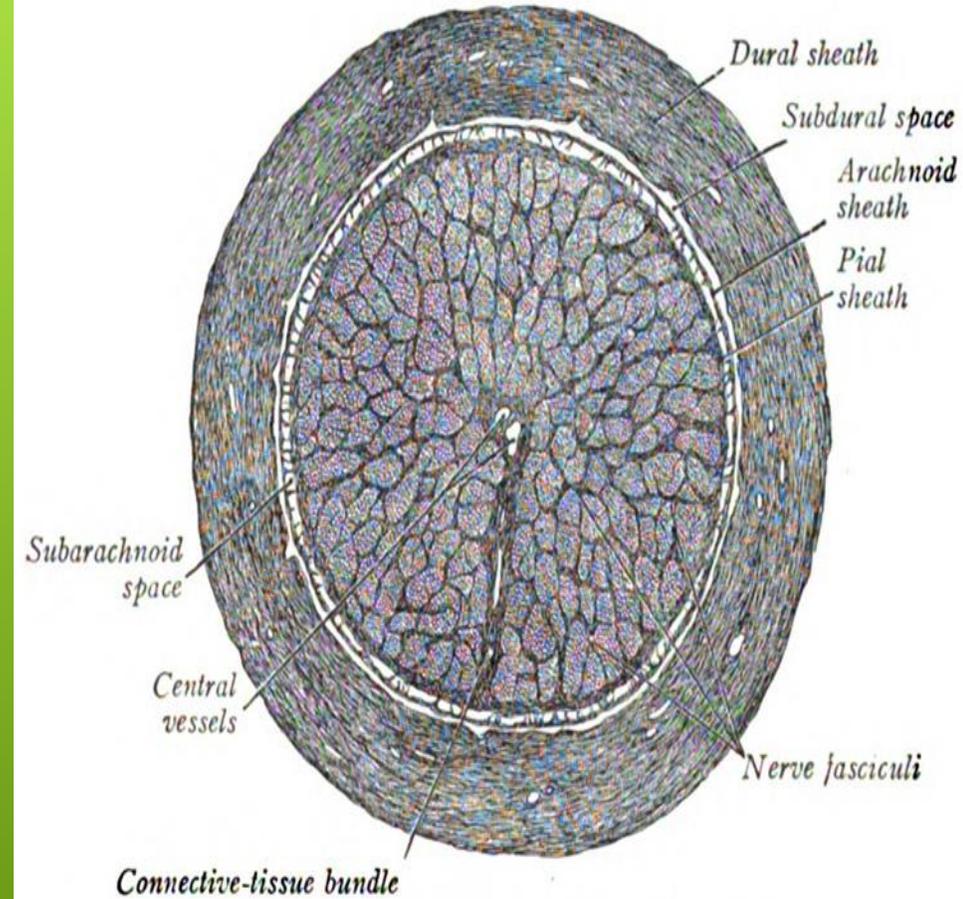


FIG. 745.—A cross-section of the optic nerve.

## MRI signs of increased ICP

Flattening of posterior sclera

Distention of peri optic subarachnoid space

Enhancement of prelaminar optic nerve



Vertical tortuosity of the orbital optic nerve (40%)

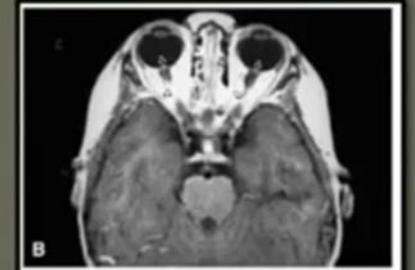


Empty sella (70%)

## MRI signs of increased ICP



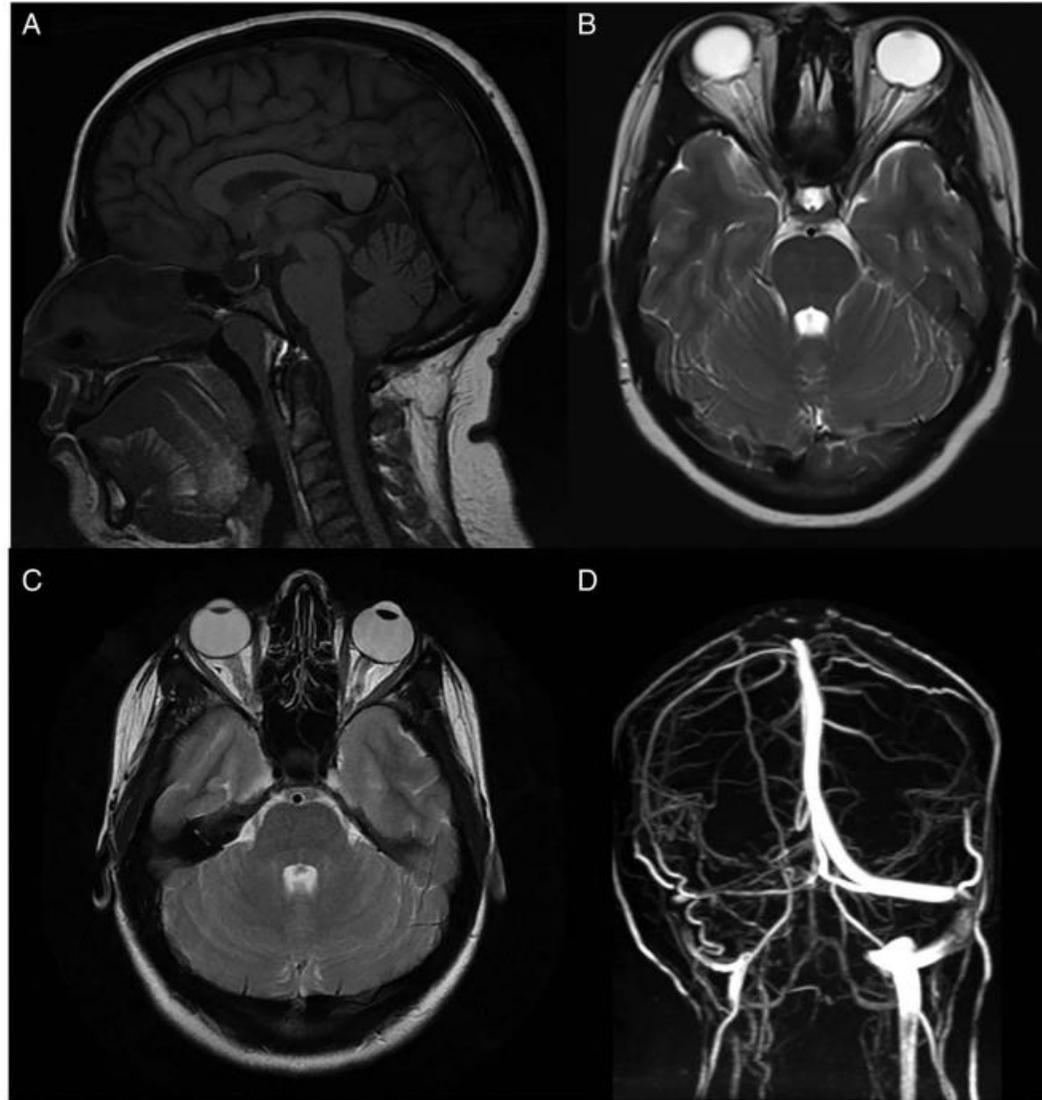
Flattening of posterior sclera (80%)



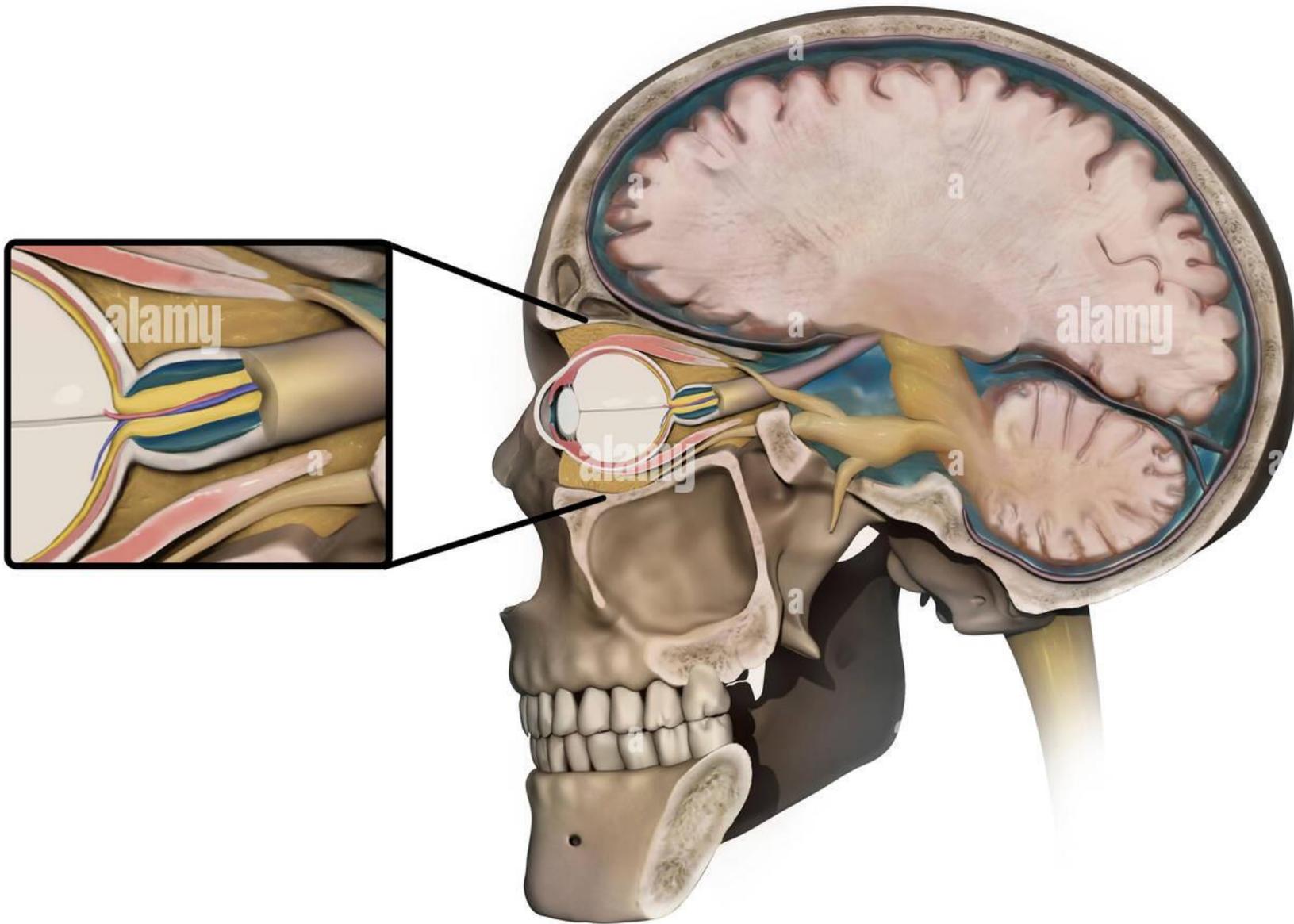
Enhancement of prelaminar optic nerve (45%)



Distention of perioptic subarachnoid space (50%)



**Figure 2** (A) MRI T1-weighted sagittal imaging demonstrating an empty sella (the pituitary gland has been flattened against the wall of the sella). (B) MRI T2-weighted axial image demonstrating flattening of the posterior globes at the insertion of the optic nerves, protrusion of the optic nerve head into the vitreous and increased fluid in the optic nerve sheath complex bilaterally. (C) MRI T2-weighted axial image demonstrating tortuosity (kinking) of the intraorbital optic nerve on the left with fluid in the associated optic nerve sheath complex. (D) MR venography (posterior view) demonstrating a longitudinal extensive left transverse sinus stenosis (extraluminal appearance).



**Table 2** Conditions that may cause intracranial hypertension

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**Pharmacological agents**

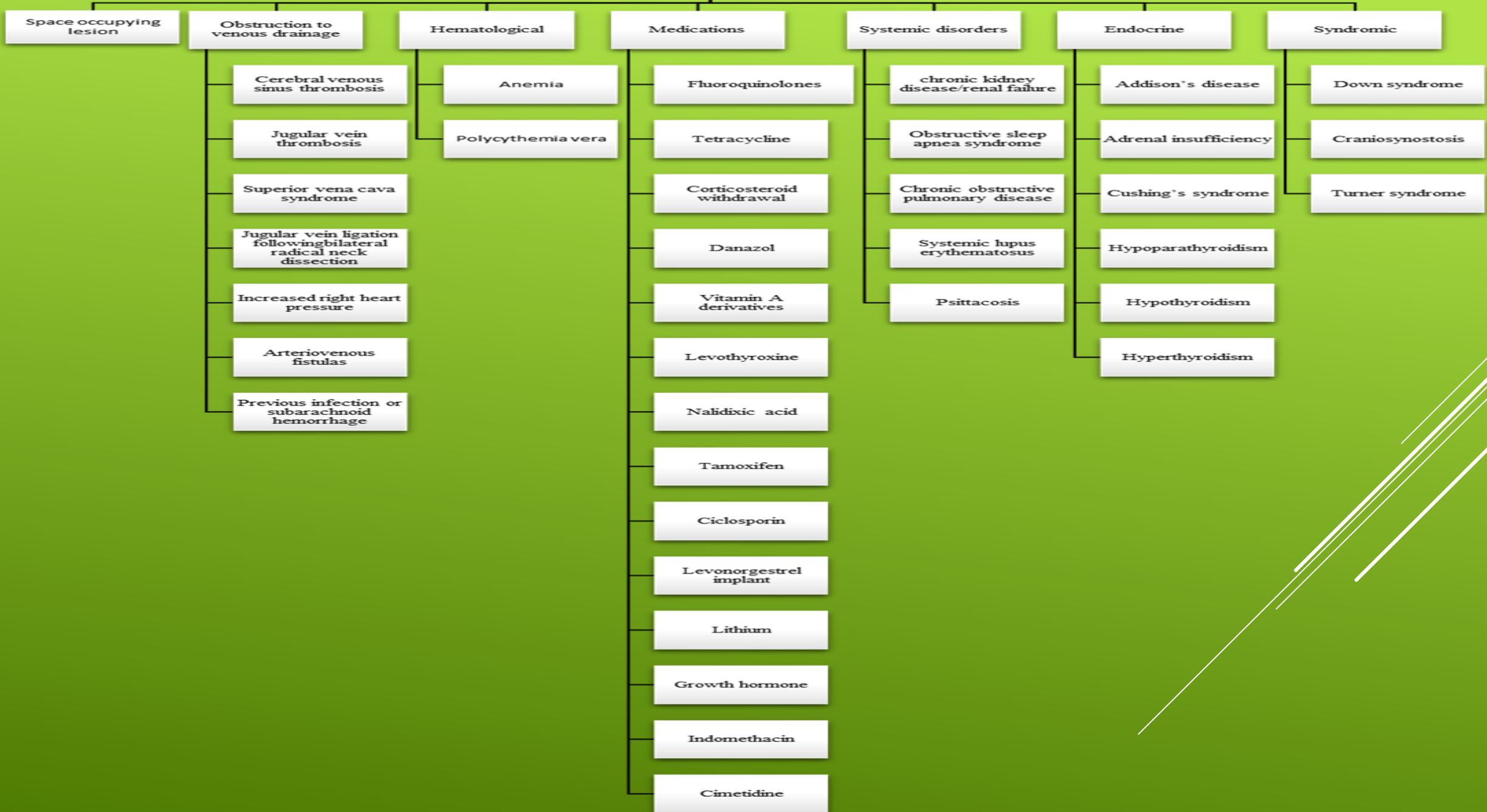
Antibiotics: tetracycline and derivatives, vitamin A derivatives: isotretinoin, all-trans-retinoic acid (for acute promyelocytic leukaemia)  
Hormonal agents: corticosteroid withdrawal, growth hormone, thyroxine replacement in children  
Other agents: lithium, nalidixic acid, rofecoxib, cimetidine

**Systemic conditions**

Haematological: anaemia  
Respiratory: obstructive sleep apnoea  
Renal: renal failure  
Endocrine: obesity, weight gain, polycystic ovarian syndrome, Cushing's disease, Addison's disease, hypoparathyroidism  
Genetic: turner syndrome  
Autoimmune: systemic lupus erythematosus  
Nutritional: hypervitaminosis A  
Venous: cerebral venous sinus thrombosis, superior vena cava obstruction, increased right-sided heart pressure

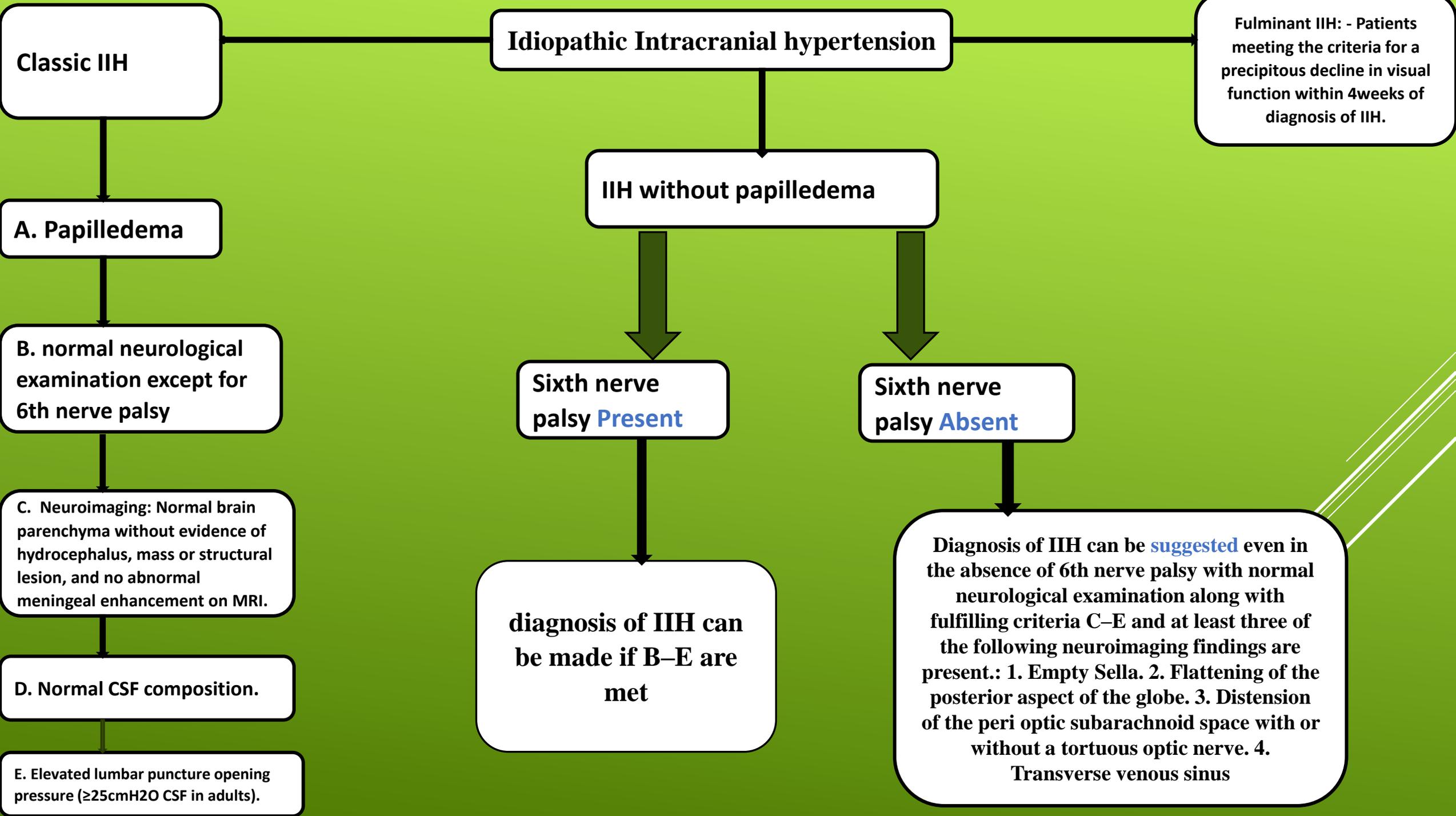
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Secondary aetiology of ICH



# MODIFIED DANDY CRITERIA FOR IIH

- ❖ Signs and Symptoms only reflect those of generalized high icp or papilledema (headache, visual disturbances, papilledema, 6<sup>th</sup> nerve palsy)
- ❖ Documented elevated ICP in lateral decubitus CSF pressure >25cmH<sub>2</sub>o
- ❖ Normal csf composition (r/o infection, inflammatory, neoplastic)
- ❖ No evidence of hydrocephalus, mass, structural or vascular lesion on MRI/MRV or contrast enhanced CT for typical patient and atypical patients
- ❖ No other causes of increased ICP identified



**Table 1** Diagnostic criteria for idiopathic intracranial hypertension (IIH) adapted from Friedman *et al*<sup>1</sup>

Diagnosis of IIH	Diagnosis of IIH without papilloedema
<p>Diagnosis of IIH is definite if the patient fulfils A–E</p> <p>A. Papilloedema.</p> <p>B. Normal neurological examination except for sixth cranial nerve abnormalities.</p> <p>C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass or structural lesion, and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used.</p> <p>D. Normal CSF composition.</p> <p>E. Elevated lumbar puncture opening pressure (<math>\geq 250</math> mm CSF in adults) in a properly performed lumbar puncture.</p>	<p>In the absence of papilloedema, a diagnosis of IIH can be made if B–E are satisfied, and in addition the patient has unilateral or bilateral abducens nerve palsy.</p> <p>In the absence of papilloedema or sixth nerve palsy, a diagnosis of IIH can be suggested but not made if B–E are satisfied, and in addition at least 3 of the following are present on neuroimaging:</p> <ol style="list-style-type: none"><li>1. Empty sella.</li><li>2. Flattening of the posterior aspect of the globe.</li><li>3. Distension of the perioptic subarachnoid space with or without a tortuous optic nerve.</li><li>4. Transverse venous sinus stenosis.</li></ol> <p>(See <a href="#">figure 2</a>: MRI findings in IIH)</p>

The diagnosis of IIH is considered probable if A–D are met, but the cerebrospinal fluid pressure is below 250 mm.

## Papilloedema Identified

Record:

1. Visual acuity
2. Formal Visual fields
3. Dilated fundoscopy\*

NB Regular assessment of vision if affected

*\*Confirm papilloedema with a experienced clinician if any doubt or fundoscopy*

Check blood pressure and exclude malignant hypertension [ref]

**Brain Imaging within 24 hours (CT/MRI) AND venography**

No lesions identified.

**Lumbar puncture**

Opening pressure >25cm CSF; normal contents.

**Exclude secondary causes (table 2)**

**Idiopathic Intracranial Hypertension**

Is the vision at imminent RISK?

**Fulminant IIH**

Is this a woman, of reproductive age, with a BMI >30kg/m<sup>2</sup>?

**Typical IIH**

Is this person not female, not of reproductive years and with a BMI <30kg/m<sup>2</sup>?

**Atypical IIH**

### ❖ Medical management

Weight loss

Low sodium diet

Low calorie diet

Bariatric surgery

Acetazolamide 2-4g/day

Topiramate- weight loss, headache.

Furosemide, methazolamide if weight loss difficult or rebound edema on steroid withdrawal

**Malignant ITH:-** rapid vision loss, rapid onset, treat with short course of steroids and surgery

Serial LP - Short lasting. **Complication** are post LP headache. Recommended in pregnant woman who cannot take meds

### ❖ Surgical management indication

Treatment Failure, not tolerant or noncompliant to medications, headache unresponsive to treatment, worsening visual acuity/visual field, severe vision loss on presentation and severe Headache not responding to meds- **lumbo peritoneal shunt or ventriculoperitoneal shunt**. **Complication** are shunt failures requiring shunt revision and infection.

**Optic nerve sheath decompression** reserved for pts who have vision loss as their primary complaint complication—diplopia, tonic pupil, vision loss.

**Transverse venous stenosis:** Transverse sinus stenting.

# Idiopathic Intracranial Hypertension

Weight management advice for all with a BMI  $\geq 30\text{kg/m}^2$  \*(Question 2)

No immediate threat to vision

Consider medical therapy with acetazolamide (Question 8,9)

Headache assessment:

1. Evaluate the headache phenotype (Question 11,12)
2. Eliminate medication overuse elements (Question 14)
3. Treat (Question 13)

Ophthalmology assessments (Question 23, Table 5)

If significant deterioration of visual function, consider diagnostic lumbar puncture

Vision threatened (Fulminant IIH)

Temporising lumbar drain if surgery planned >24 hours

CSF diversion (Question 4)

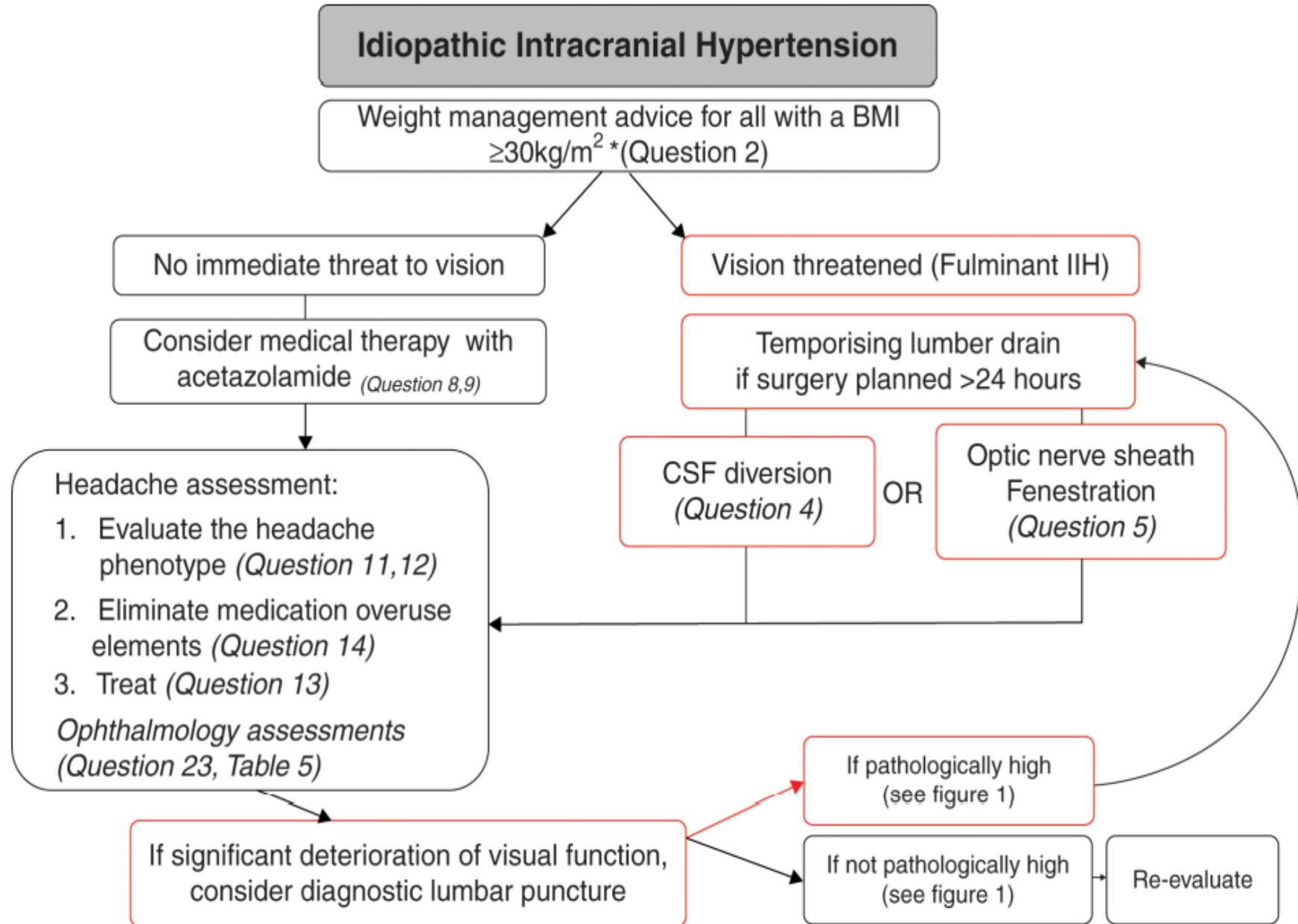
OR

Optic nerve sheath Fenestration (Question 5)

If pathologically high (see figure 1)

If not pathologically high (see figure 1)

Re-evaluate



## Prognosis-

- ❖ Self limited.
- ❖ 26% develop  $<20/200$  in worse eye and irreversible changes such as disc pallor, vascular sheathing and optic atrophy.
- ❖ Risk factors for poor prognosis:- systemic HTN, Age, not following up with an ophthalmologist.

## Acetazolamide adverse events

Renal stones, tramaminitis, pancreatitis, diverticulitis, allergic reaction, hypokalemia, paresthesia, dysgeusia, fatigue, N, V, diarrhea, tinnitus, avoided during the first 20 weeks of pregnancy.

## Topiramate side effects

Cognitive slowing, memory impairment, paresthesia, renal stones.

## **References:**

- 1) Mollan SP, Ali F, Hassan-Smith G, Botfield H, Friedman DI, Sinclair AJ. Evolving evidence in adult idiopathic intracranial hypertension: pathophysiology and management. *J Neurol Neurosurg Psychiatry*. 2016 Sep;87(9):982-92. doi: 10.1136/jnnp-2015-311302. Epub 2016 Feb 17. PMID: 26888960; PMCID: PMC5013119.
- 2) Wall M. Idiopathic intracranial hypertension. *Neurol Clin*. 2010 Aug;28(3):593-617. doi: 10.1016/j.ncl.2010.03.003. PMID: 20637991; PMCID: PMC2908600.
- 3) Mollan SP, Davies B, Silver NC, Shaw S, Mallucci CL, Wakerley BR, Krishnan A, Chavda SV, Ramalingam S, Edwards J, Hemmings K, Williamson M, Burdon MA, Hassan-Smith G, Digre K, Liu GT, Jensen RH, Sinclair AJ. Idiopathic intracranial hypertension: consensus guidelines on management. *J Neurol Neurosurg Psychiatry*. 2018 Oct;89(10):1088-1100. doi: 10.1136/jnnp-2017-317440. Epub 2018 Jun 14. PMID: 29903905; PMCID: PMC6166610.

**Thank You**

