University Hospitals Birmingham NHS Foundation Trust

Sandwell and West Birmingham NHS Trust

TRANSLATIONAL BRAIN SCIENCE

UNIVERSITY OF BIRMINGHAM

Idiopathic Intracranial Hypertension:

how to recognise and manage

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Headache Academy 2025

Overview

Diagnosis

Headache

Comorbidities /Differentials

Management

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Diagnosis

Case 1

- 30F presents with worsening headaches
- Worse in the morning, 8/10 and throbbing.
- Associated photophobia, phonophobia and nausea.
- Ongoing for 6 months, gradual increase in frequency – now daily
- No visual changes.
- Other symptoms: high pitched tinnitus and back pain but possibly known fibromyalgia
- BMI 35

Diagnosis based on history?

- **Migraine**
- Medication overuse headache
- Raised ICP headache
  - IIH
  - Space occupying lesion
  - CVST (less likely given duration)
- Obstructive sleep apnoea
- Hypertension related
- Spontaneous intracranial hypotension

Tips for History:

- Ask about posture, Valsalva, and movement effects
- Timings of exacerbations
- Visual disturbances, tinnitus, double vision
- Headache in female with obesity ≠ IIH

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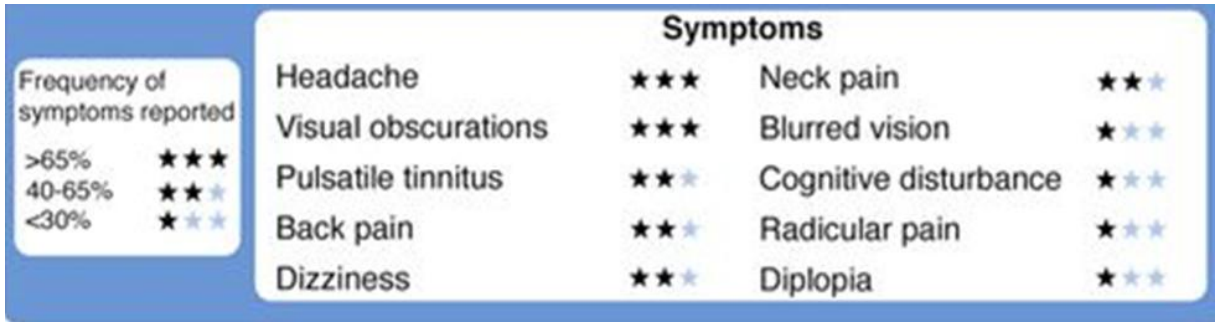
Idiopathic Intracranial Hypertension



Susan P Mollan et al. J Neurol Neurosurg Psychiatry 2018;89:1088-1100

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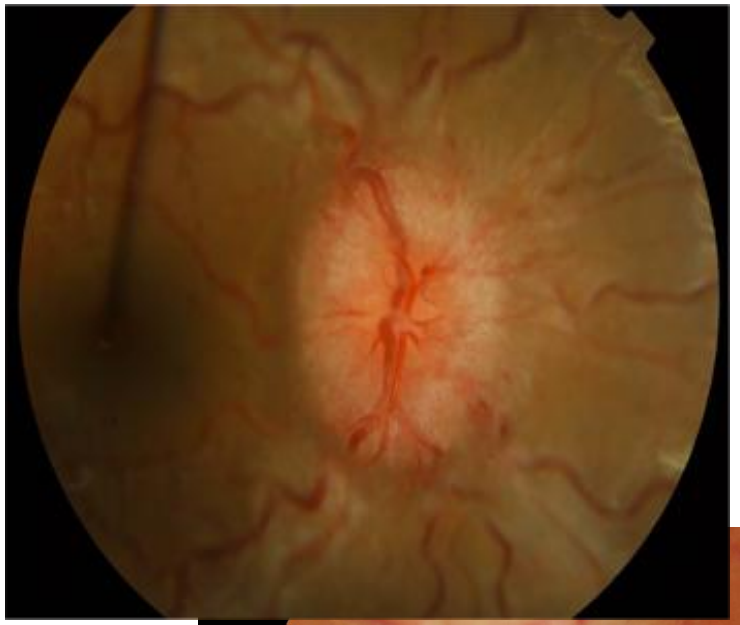
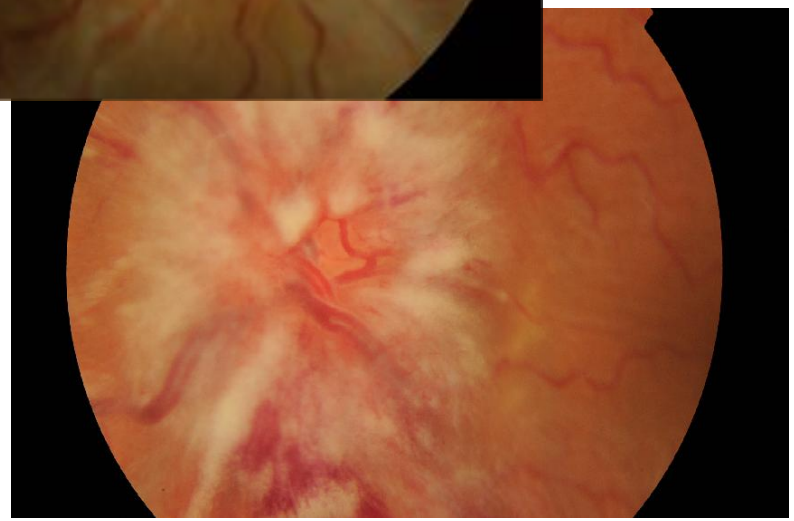
Examination

- “Normal neurological examination”
  - What was tested?
    - Cranial nerves
    - Eye exam
      - Visual acuity
      - Pupil reflexes
      - RAPD
      - Eye movements
      - Saccades and pursuits
    - **Fundoscopy**
    - Upper limb
    - Lower limb
    - Cerebellar
    - Etc.

Look at the discs





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ARCLIGHT MEDICAL

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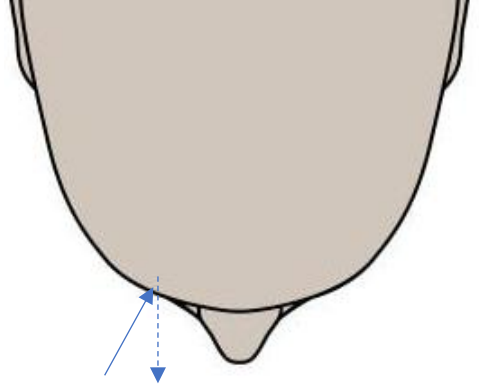
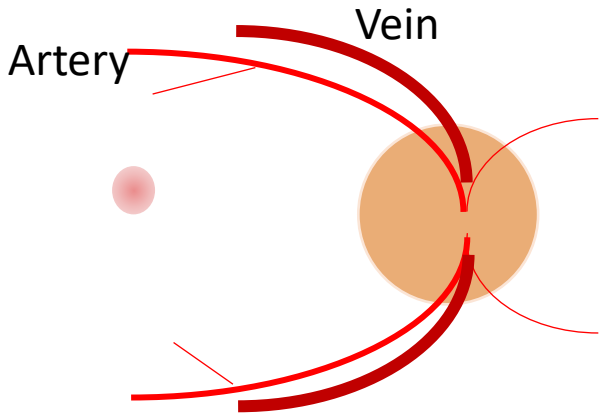
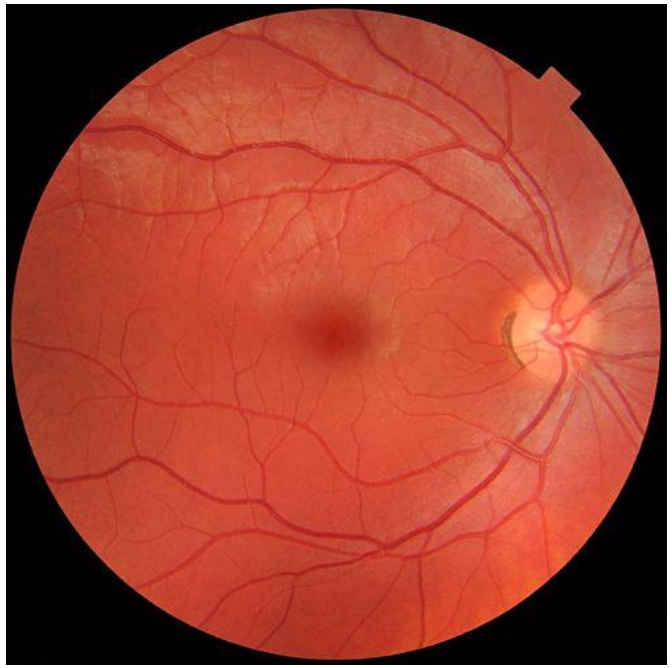
Fundoscopy Tips





- Technique
  - Start at 30 degree angle
  - Patient need to fix on a point
  - It is a big X but disc is more nasal
  - If seeing branching vessels, they are arrows in direction of nerve head
- Quantify severity
  - Normal v abnormal (practice)
  - Mild swelling v Severe /haemorrhages etc
- Clinical context



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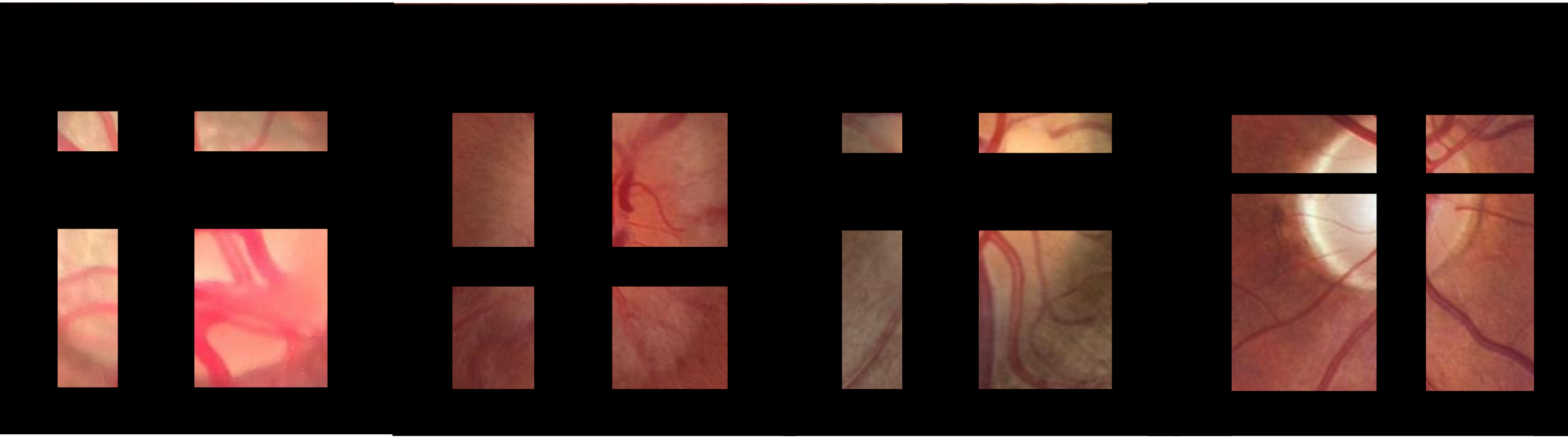
Fundoscopy – Test yourself



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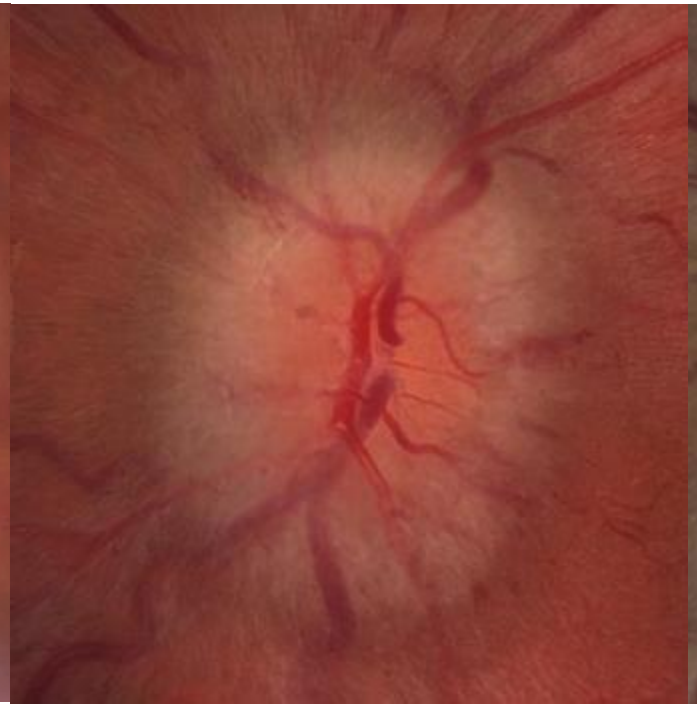
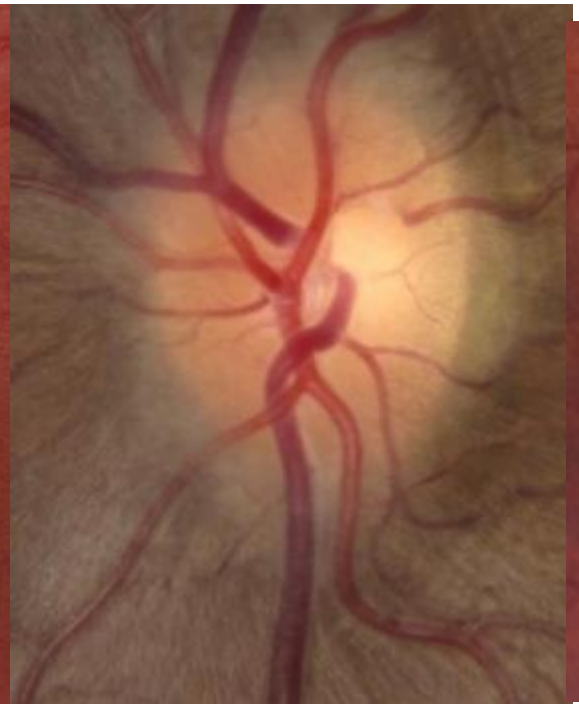
Fundoscopy – Test yourself answers

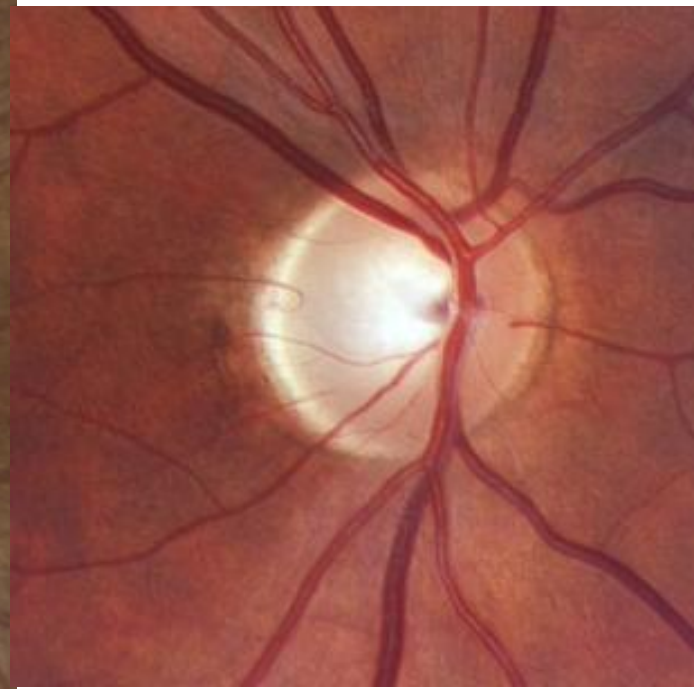
Myopic tilted disc



Papilloedema G3



Papilloedema G1



Optic atrophy

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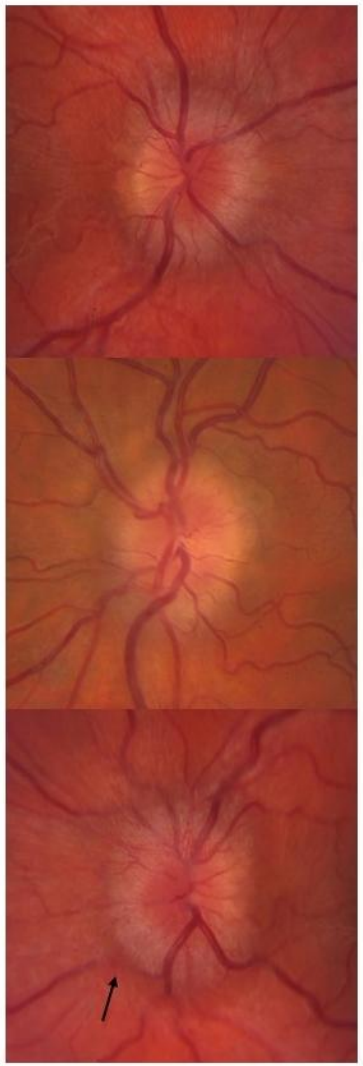
Frisen grading



Grade 1

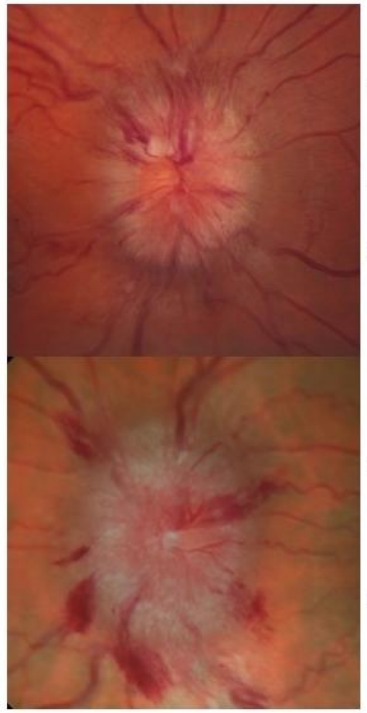
- Grayish C-shaped halo surrounding the disc*
- Sparing of the temporal disc margin
- Radial nerve fiber striation disruption

Grade 2

- Halo becomes circumferential*
- Nasal border elevation
- No major vessel obscuration

Grade 3

- Obscuration of at least one vessel *leaving the disc** (arrow)
- Elevation of all borders
- Circumferential halo

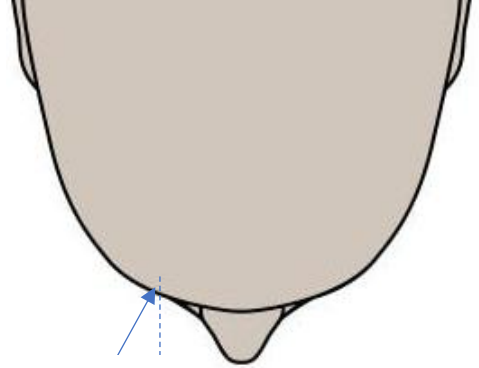


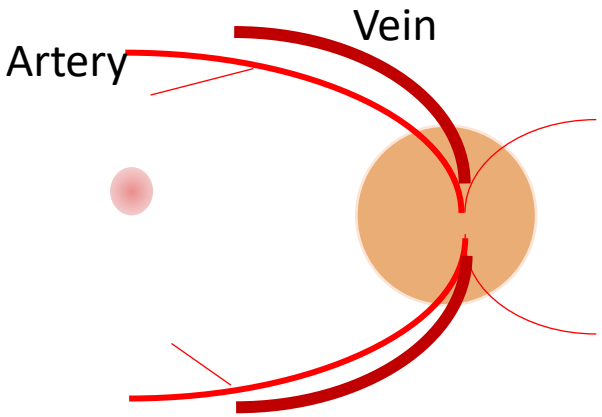
Grade 4

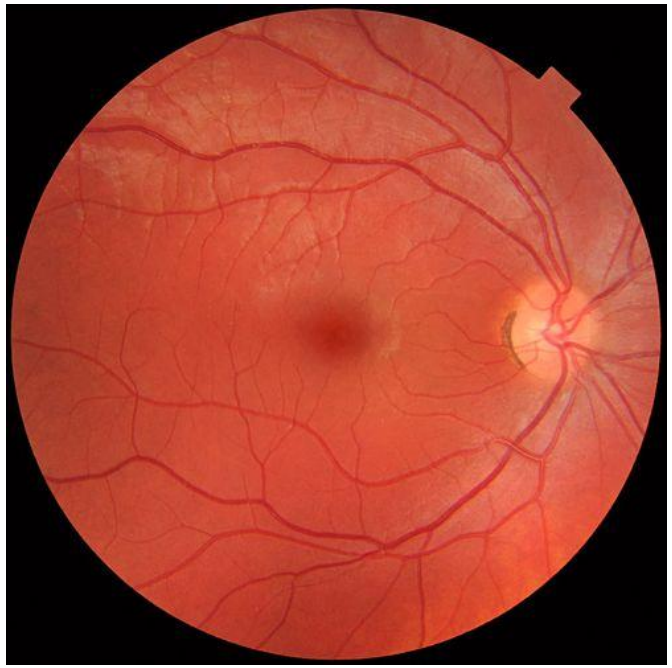
- Obscuration of a major vessel *on the disc**
- Complete elevation including the cup
- Circumferential halo

Grade 5

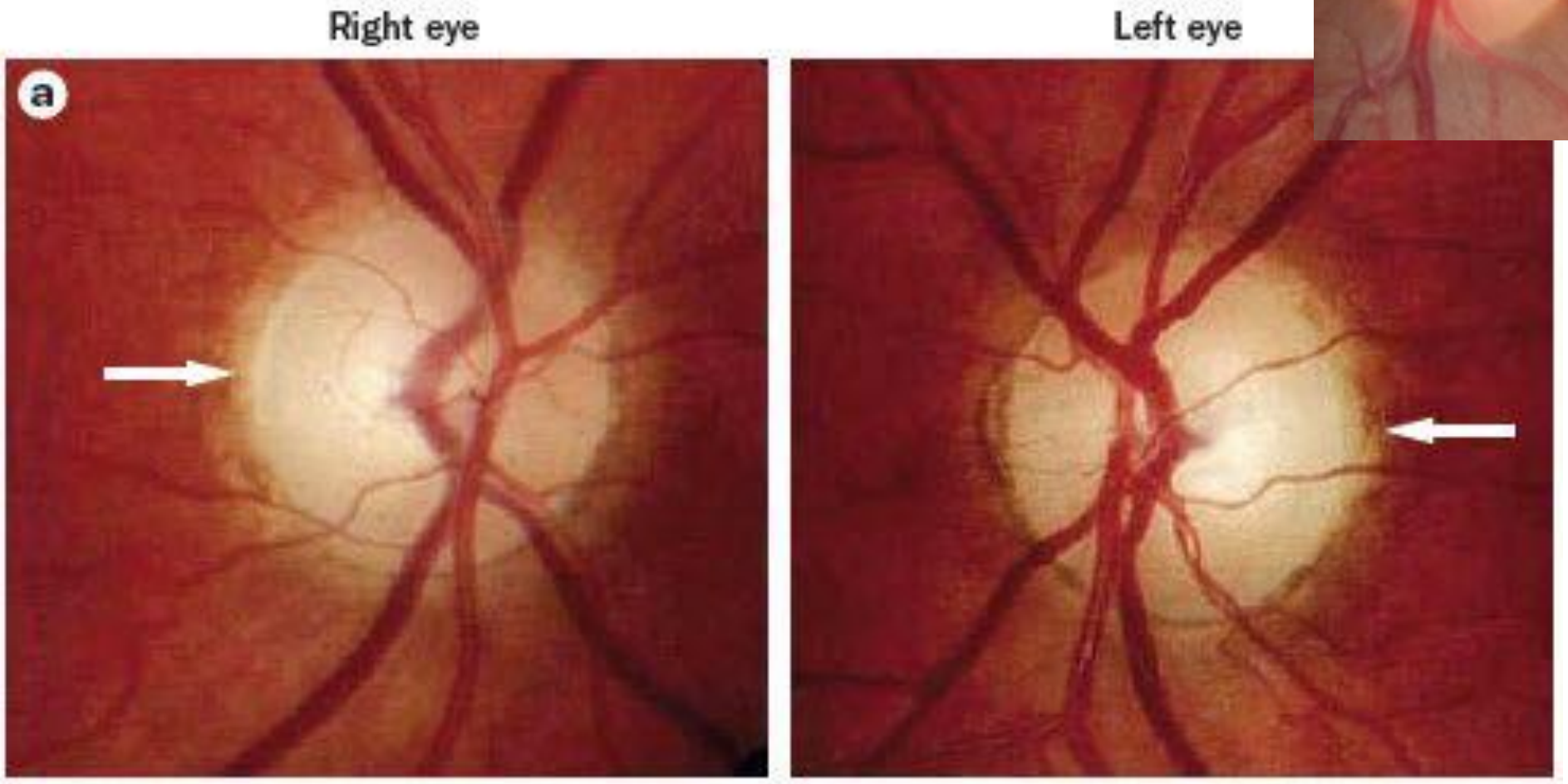
- Obscuration of all vessels on the disc *and* leaving the disc*
- All features of Grade 4



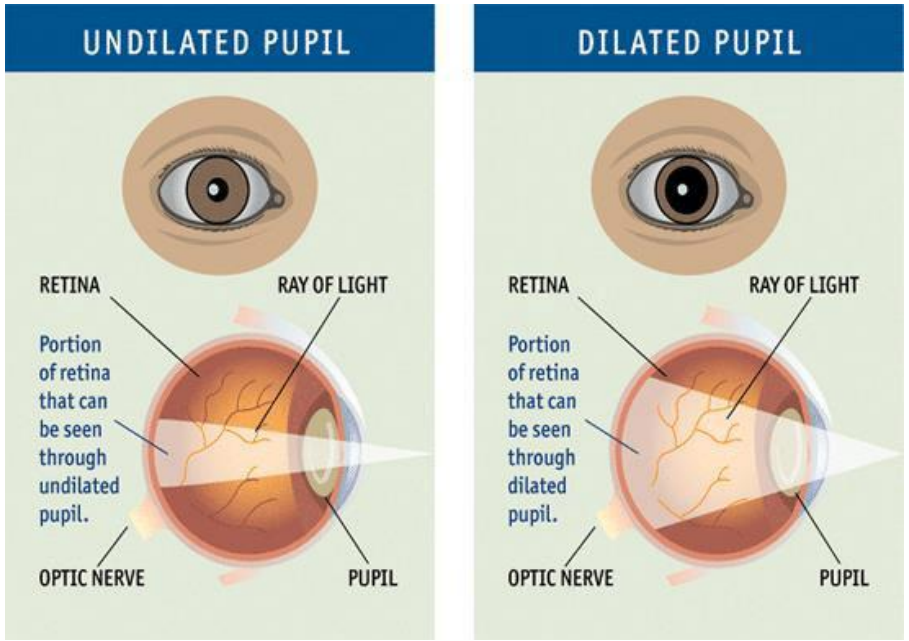




What is atrophy?



Having problems….must dilate



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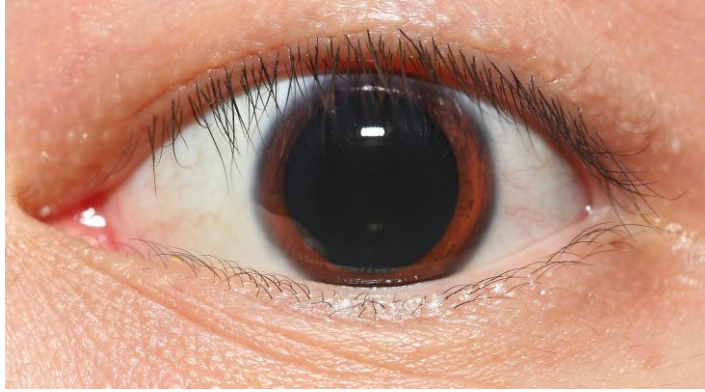
Dilation - Is it safe? YES



- Risk of acute angle-closure is very low
  - (1 to 6 per 20 000 in study/review including nearly 600 000 patients).
- Patient should be warned of the symptoms (e.g., pain, progressive blurring of vision, headache, nausea) and advised to seek immediate care should they occur.
- Maximal dilation with tropicamide 0.5% is reached after 20–30 minutes and reverses over 3-6 hours.
  - *Make sure you document in notes if used and expected reversal time*
- Sunglasses can be worn to increase comfort in bright surroundings while the pupils are dilated.
- Advised not to drive following dilation, until accommodation has returned to normal

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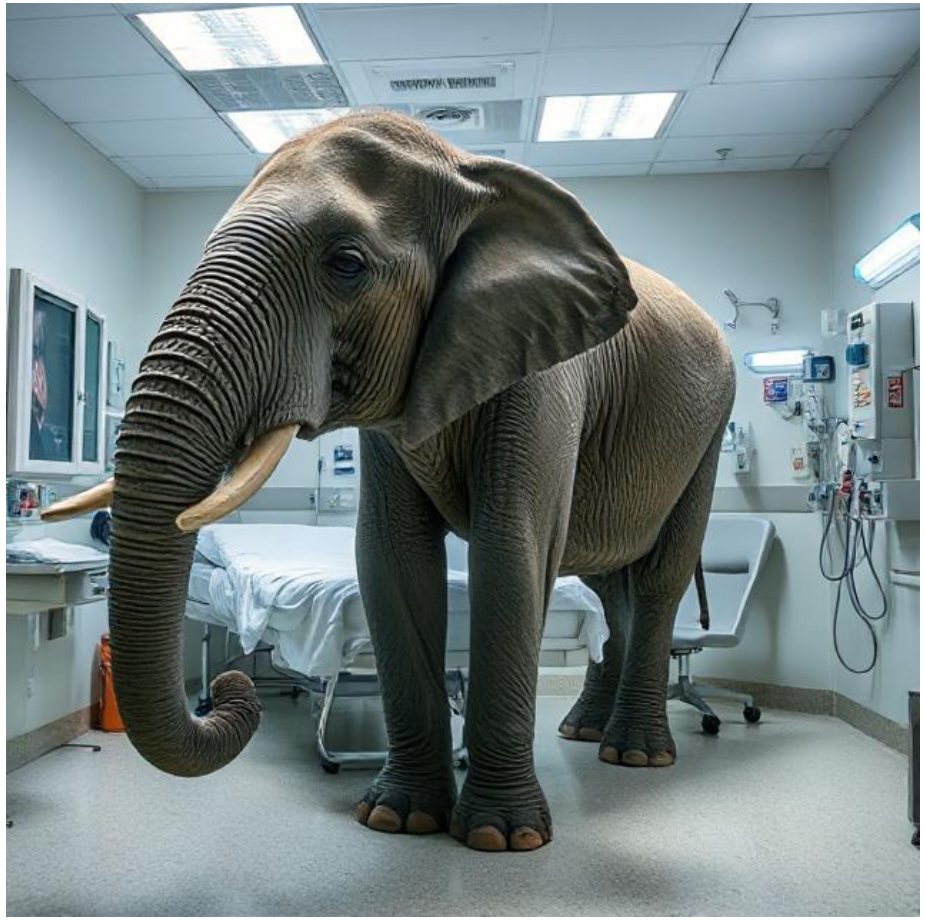
Pandit RJ, Taylor R Mydriasis and glaucoma: exploding the myth. A systematic review. Diabet Med.2000;17(10):693-699.
Liew G, Mitchell P, Wang JJ, et al. Fundoscopy: To dilate or not to dilate? BMJ 2006;332:3.

Rapid visual loss = Visual threatening Optic Nerve Swelling

- Rapid onset of symptoms
- Visual acuity loss is a LATE sign
- Visual fields is there a new impairment?
- Papilloedema severity?
  - Try to grade on fundoscopy
  - Slit lamp
  - Optical coherence tomography
- Cause?
  - Recent weight gain - Avoid obesity stigma, Medication, Anaemia etc.



IIH diagnostic criteria?

IIH diagnosis

- Papilloedema.
- Normal neurological examination (except VI nerve(s) palsy).
- Normal neuroimaging: no evidence of hydrocephalus, mass or structural lesion, and no abnormal meningeal enhancement on magnetic resonance imaging. Venous sinus thrombosis excluded.
- Normal cerebrospinal fluid composition.
- Raised lumbar puncture opening pressure (≥25 cmCSF in adults and ≥28 cmCSF in children (25 cmCSF if the child is not sedated and not obese)) in a properly performed lumbar puncture.

Susan P Mollan et al. J Neurol Neurosurg Psychiatry 2018;89:1088-1100.

Thaller et al. J Neurology. 2022

Mollan S et al. Eye 2019

Tips for LP:

- Always measure opening pressure
- Always do it in lateral position if need LP OP
- Always take a second manometer and use if needed
- Have someone on hand for positioning/additional equipment

30

25

IIH without papilloedema (IIHWOP)

IIH without Papilloedema (IIHWOP) Diagnostic criteria

Presence of criteria B-E for IIH plus:

- Unilateral or bilateral sixth cranial nerve palsy

Suggestion of possible IIHWOP if:

Presence of criteria B-E for IIH plus:

- 3 neuroimaging finding suggestive of raised intracranial pressure
  - Empty sella
  - Flattening of posterior aspect of the globe
  - Distention of the perioptic subarachnoid space ± a tortuous optic nerve
  - Transverse venous sinus stenosis

- Rare
- Over diagnosed in absence of 6th nerve palsy
- Why is this not migraine / other headache disorder?
- Subject of debate

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Imaging?

- All new presentations with papilloedema (diagnostic criteria)
  - CT head
  - CT Venogram
  - MRI head
- What is abnormal?
- Opinion: Increasingly overcalled by radiology!
  - More likely if ≥3 MRI features
  - Never diagnose on a CT head

MRI signs of raised ICP ≥3 features:

- Partially/empty sella
- Increased tortuosity of optic nerve
- Enlarged optic nerve sheath
- Flattening of posterior globe
- Venous sinus stenosis

If get an imaging diagnosis of IIH, ask

1. Does clinical history fit?
2. Is there papilloedema?
3. Is there a 6th nerve palsy?

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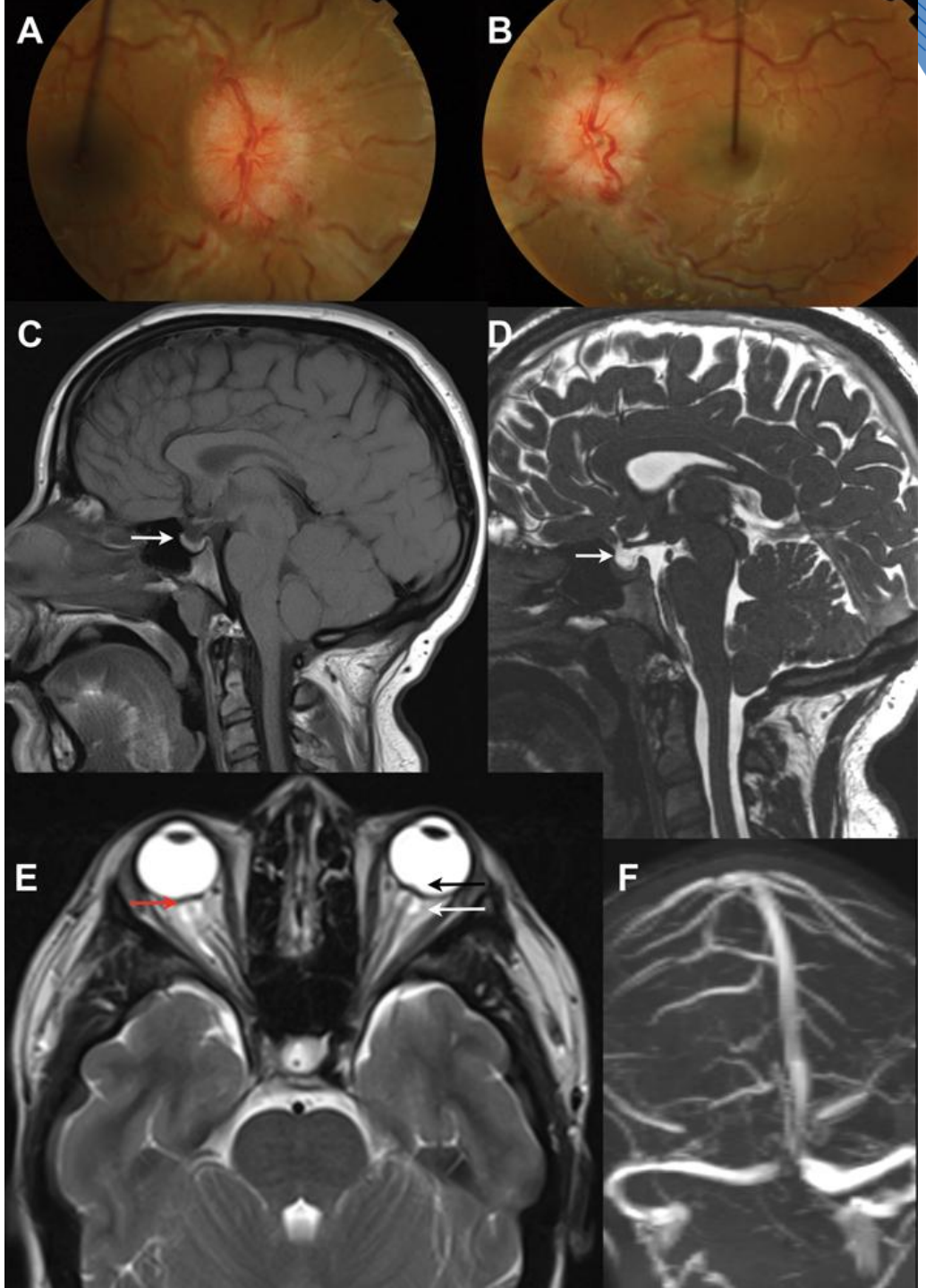
Imaging

1A and 1B show colour fundus photos of bilateral disc swelling secondary to raised intracranial pressure (papilloedema).

The neuro-imaging features are non-specific and not diagnostic of IIH:
1C and 1D show MRI T1 and T2 sagittal images showing a partially empty sella.
1E is MRI showing distension of optic nerve sheaths (white arrow), flattening of the posterior globes (red arrow) and optic nerve head protrusion (black arrow).
IF is MR venography showing no cerebral sinus thrombosis.



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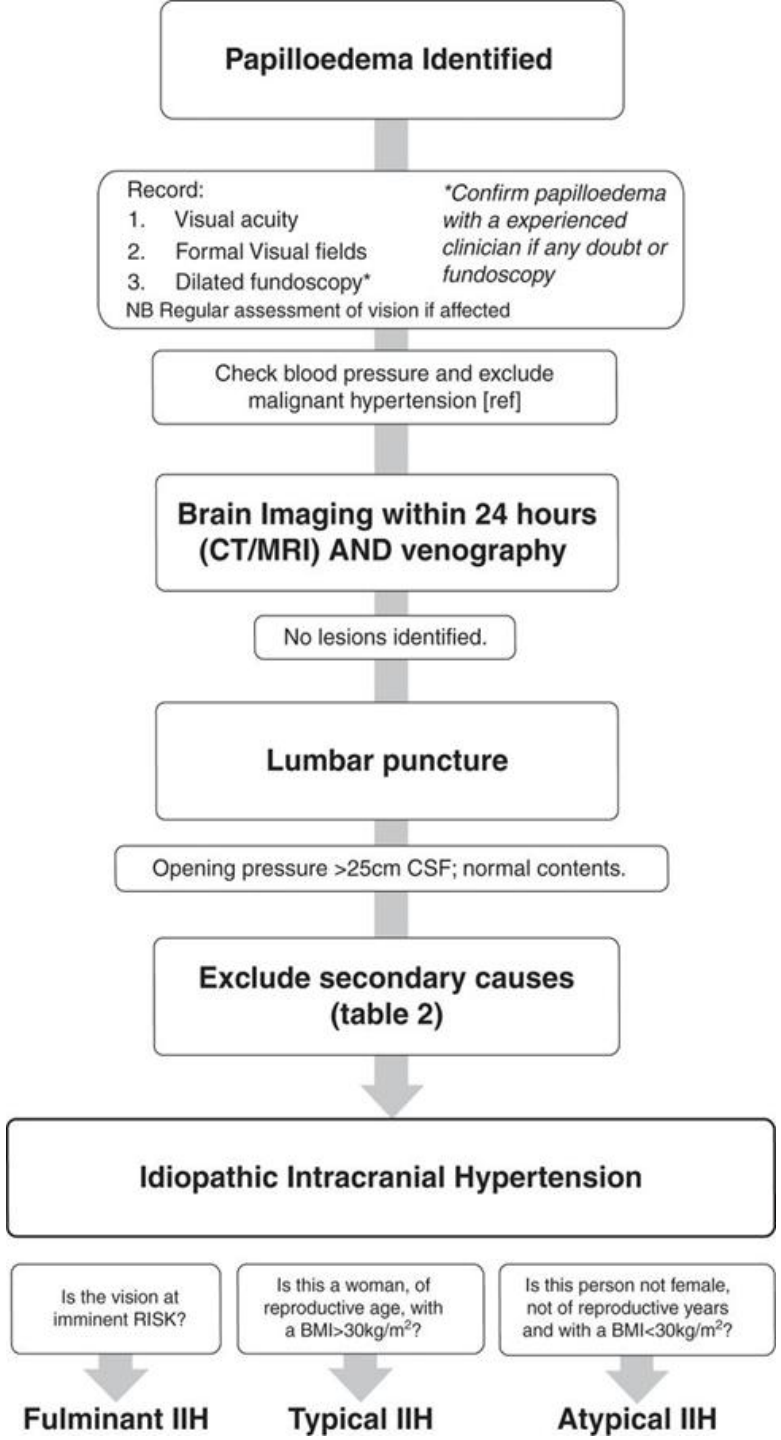
Flowchart



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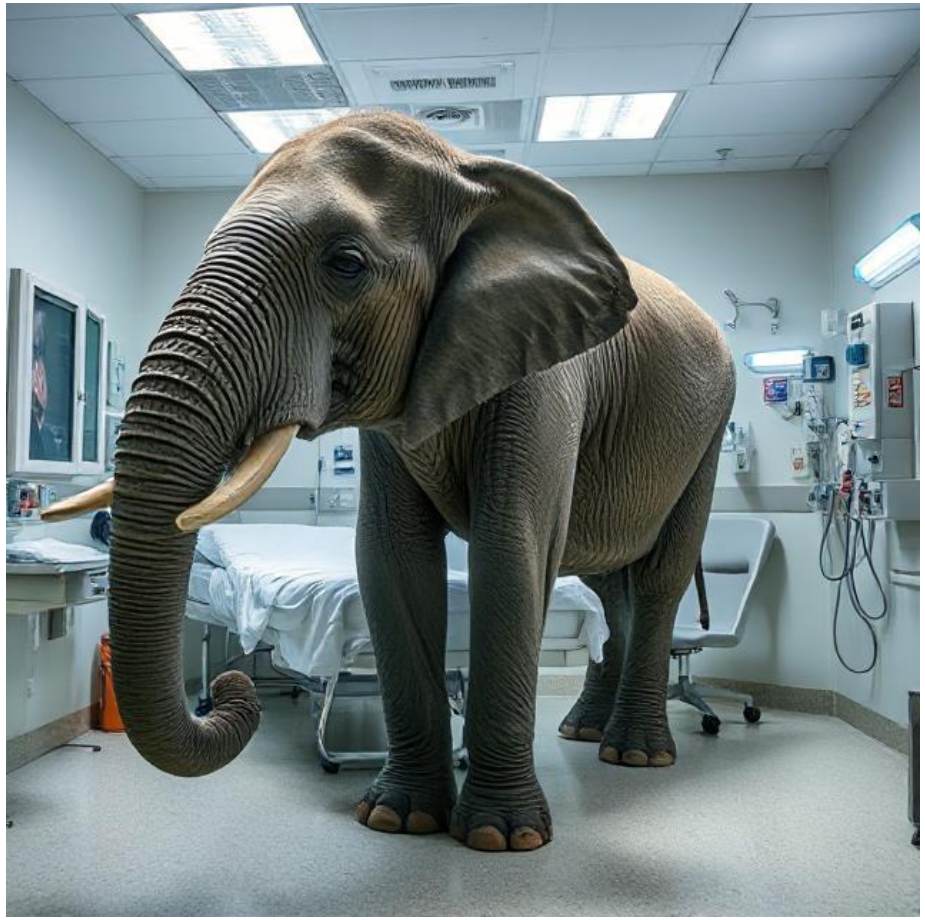
Visual threatening Optic Nerve Swelling (Papilloedema if ICP raised)

- Rapid onset of symptoms
- Visual acuity loss is a LATE sign
- New visual field impairment?
- Papilloedema severity?
  - Try to grade on fundoscopy
  - Slit lamp /Optical coherence tomography
- Cause?
  - Recent weight gain - Avoid obesity stigma, Medication, Anaemia etc.



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Headache

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What is a "typical IIH headache"

"Typical feature"

- Postural – worse on lying down
- Morning
- Valsalva triggered
- Headache improves after LP
- Chronic headache with migraine features

Differential

- Cervicogenic. Migraine.
- Medication overuse headache. Apnoea related (OSA). Cluster. Migraine
- Chiari related. Migraine. SIH. SOL. Primary cough headache
- Helpful not diagnostic, don't LP for headache, could be migraine
- Most commonly, additional IIH features?

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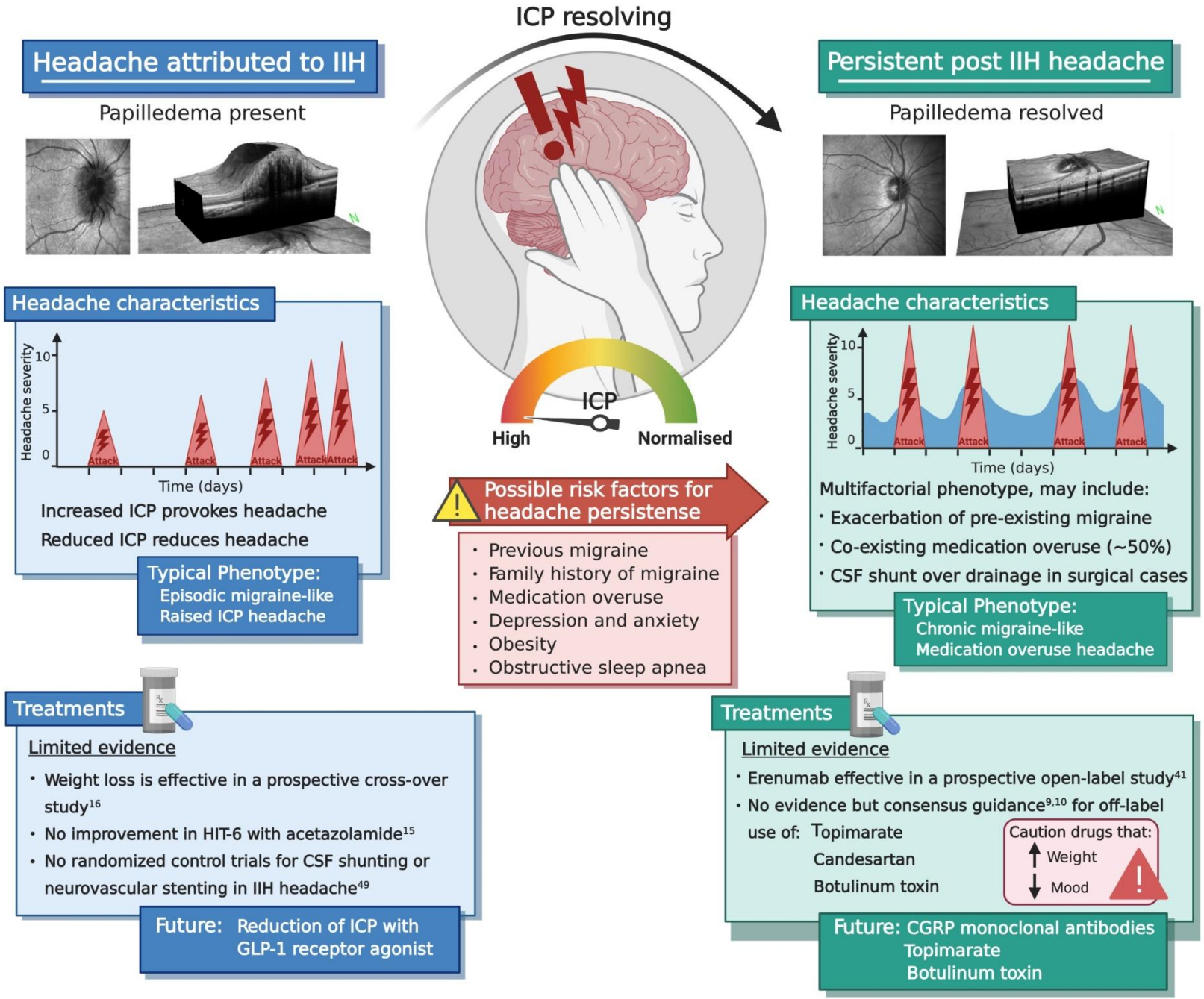
High headache burden

High opiate burden
(2-fold increase v mig, 3 v general)



Mollan et al Headache 2021

Adderley et al 2022 Neurology

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Differentials / Comorbidities

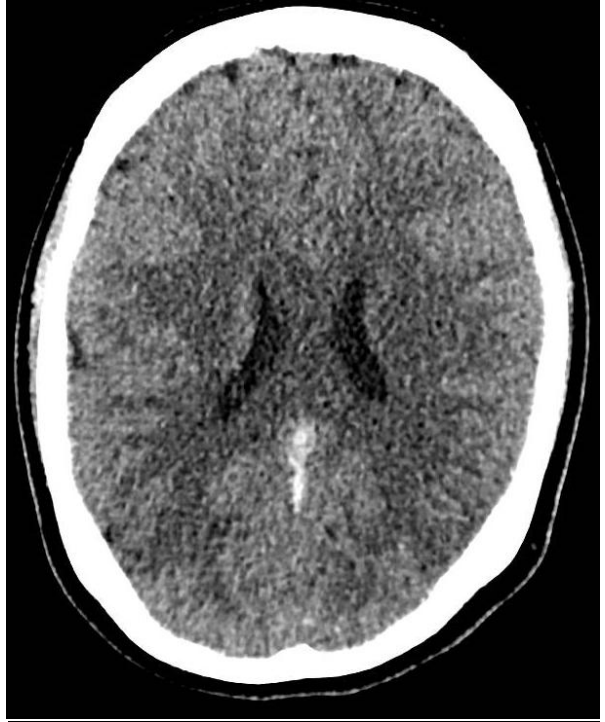
- Not all papilloedema is IIH

Case 2 (similar to case 1 with some extra history)

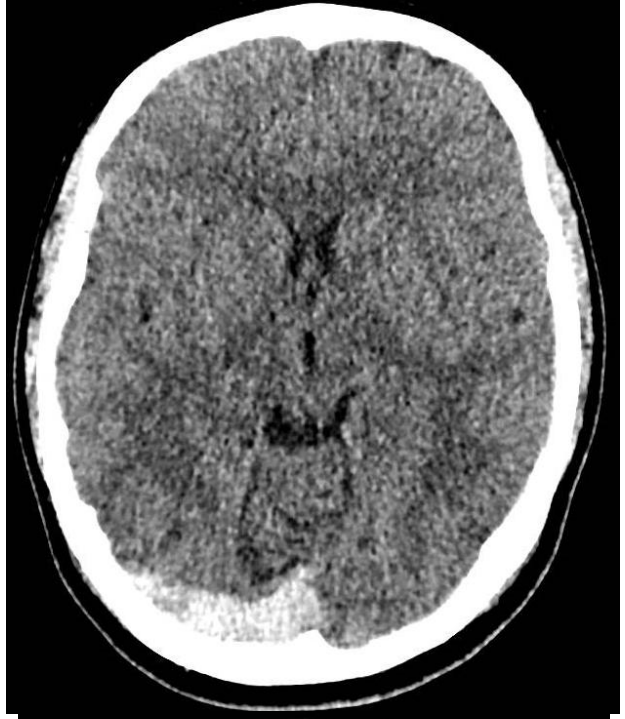
- 30F presents with worsening headaches **over last 1 week**
- Worse in the morning, 8/10 and throbbing.
- Associated photophobia, phonophobia and nausea.
- **Previous migraine. Recent flu.**
- No visual changes.
- Other symptoms: high pitched tinnitus and back pain but known fibromyalgia
- BMI 35.
- **On Combined oral contraceptive pill**
- **Fundoscopy – optic nerve swelling bilaterally Frisen 4**

Imaging

CT Head

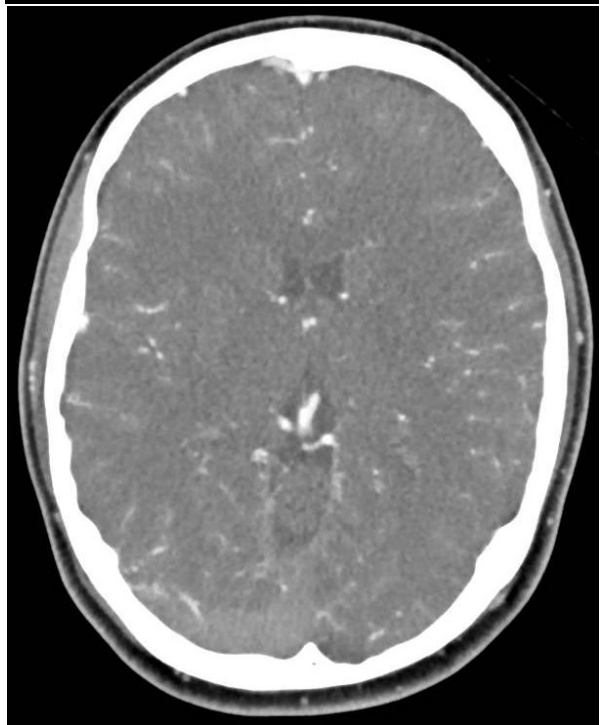


CT Head

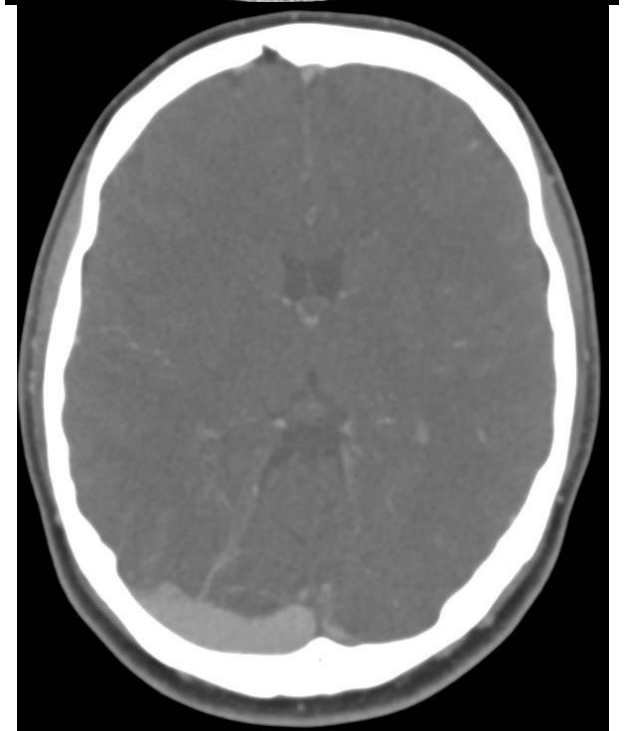


Cerebral venous sinus thrombosis
CVST

CT Venogram



CT Venogram
1m later



Management in this case

- Apixaban 5mg BD
- Acetazolamide 1g BD - Paraesthesia

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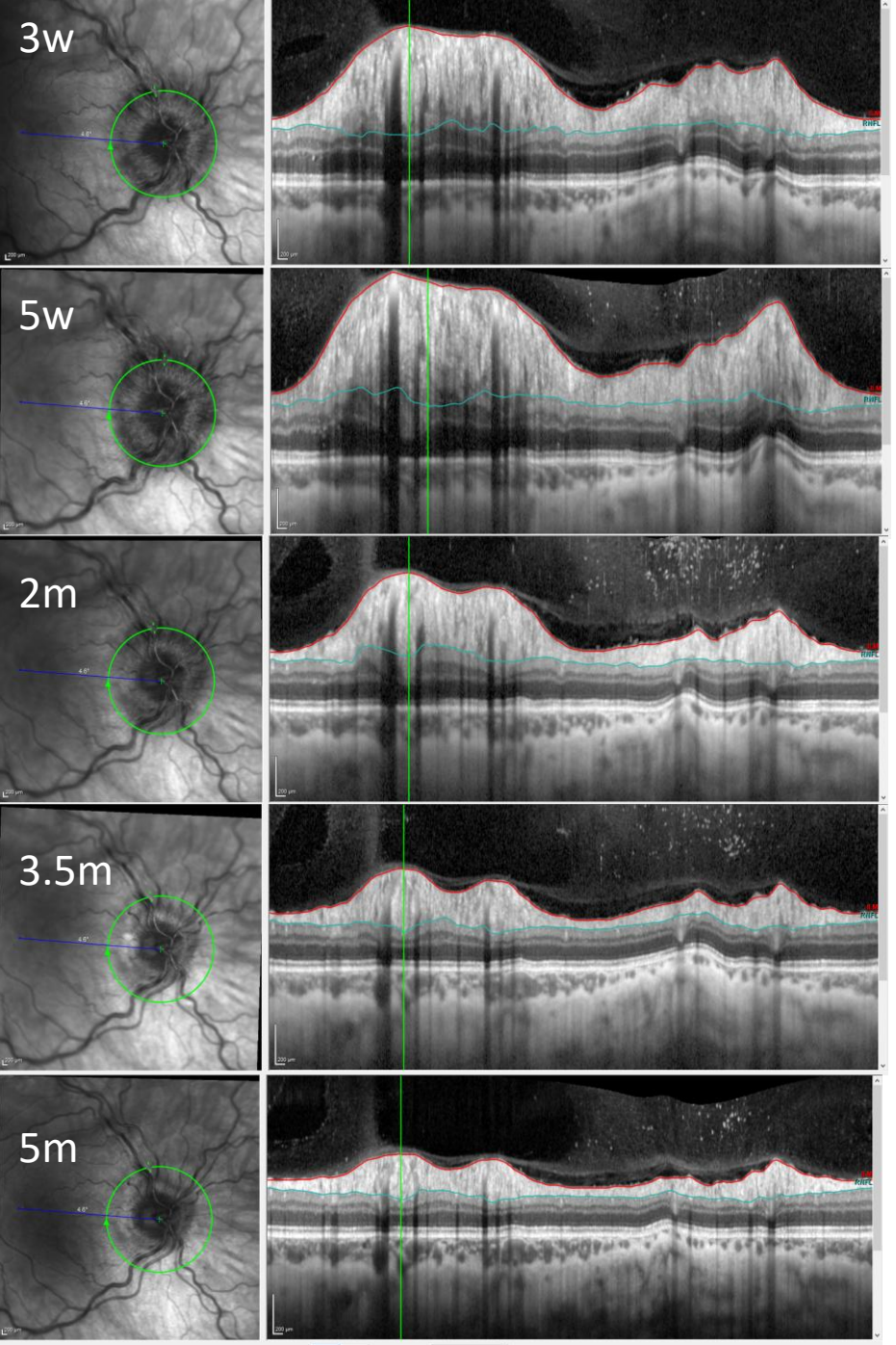
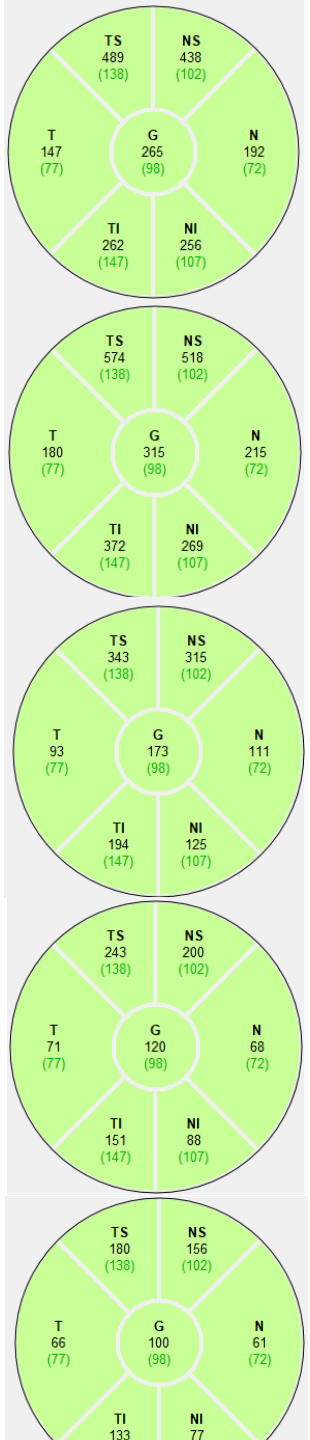
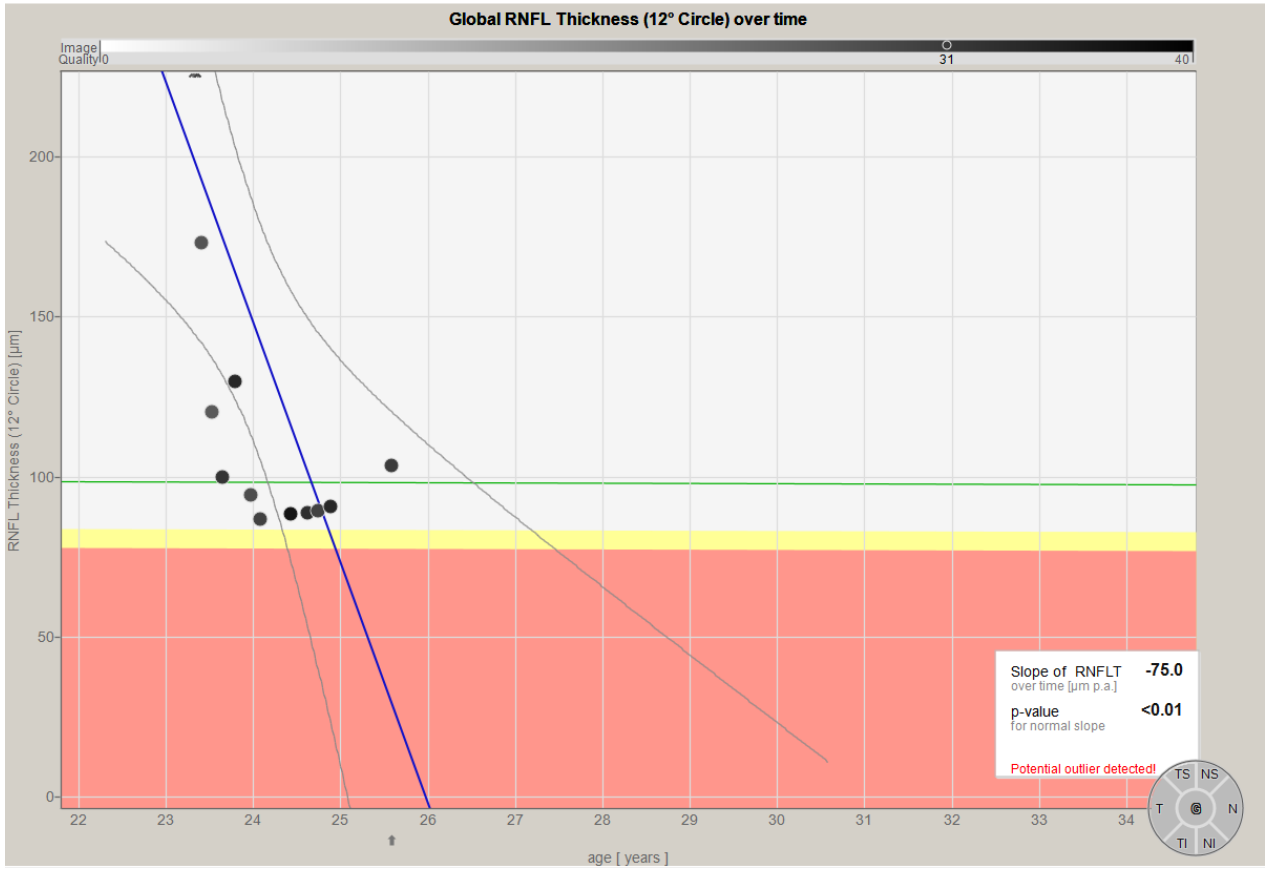
Optical Coherence Tomography (OCT)







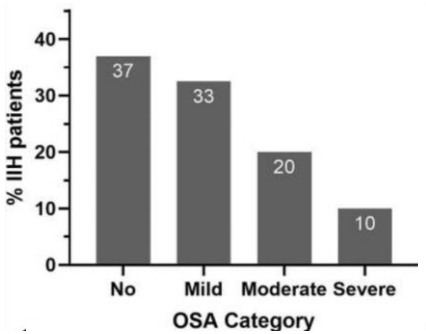
Case 3 – Do you ask about periods, sleep and meds?

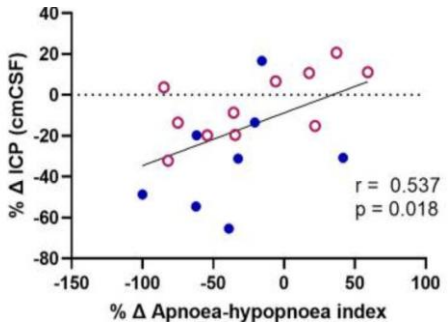
A. Irregular periods and difficulty losing weight?
   - Polycystic ovarian syndrome – 1.5-fold increased risk, 20% IIH popn
   - Management of this can benefit IIH

B. Heavy periods / endometriosis?
   - Anaemia – important to treat this but is it exacerbating predisposition?
   - Visual impairment? Rapid – i.e. fulminant - surgical intervention?

C. Snoring?
   - Obstructive sleep apnoea – STOP BANG criteria.
   - Apnoea-hypopnoea index associated with ICP
   - Management helps fatigue/headache

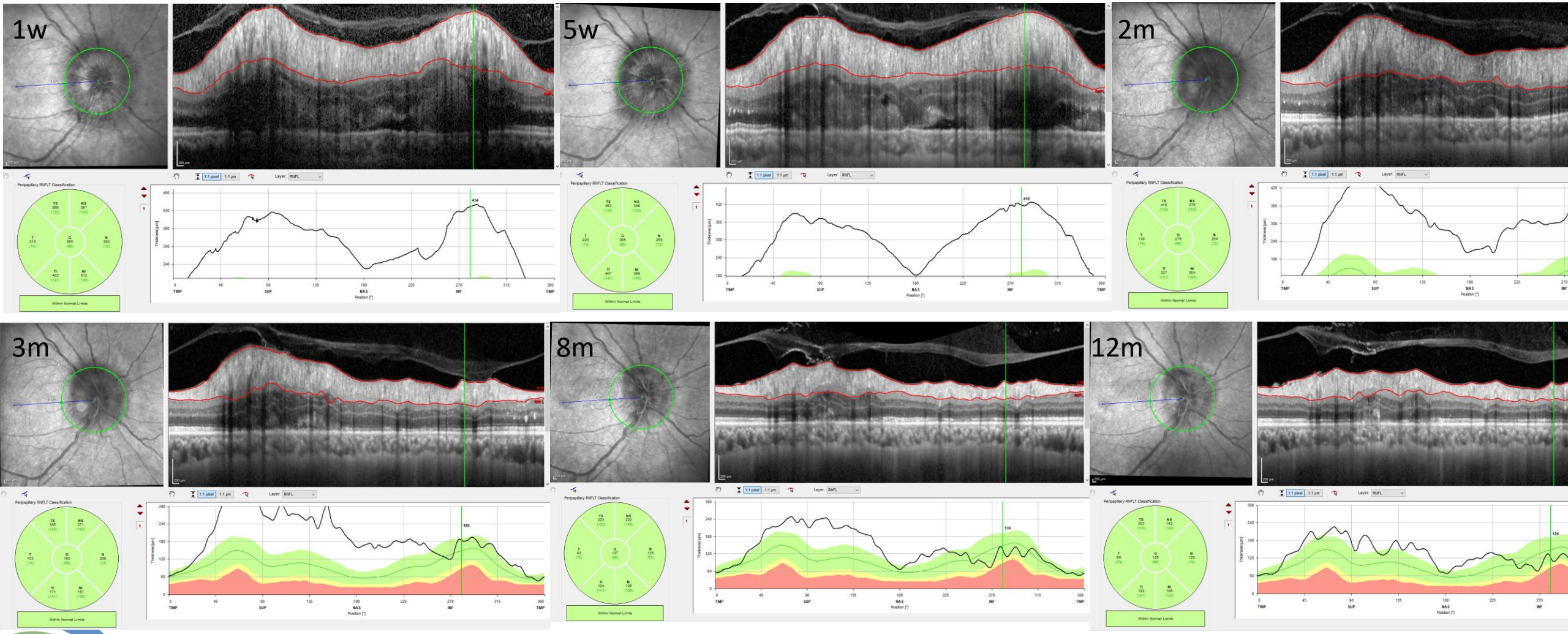
Thaller et al. 2023. Neuro-ophthalmology
Thaller et al. 2023. Eye.





Yiangou et al., J of Neurology 2021

Anaemia - OCT trend (Blood transfusions and shunt)



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Medications?

- Lots of potential correlations
- Causation however is debatable

- Impact of stopping medications
  - Don't stop OCP please – pregnancy risk
  - Interactions of medications (i.e. topiramate)

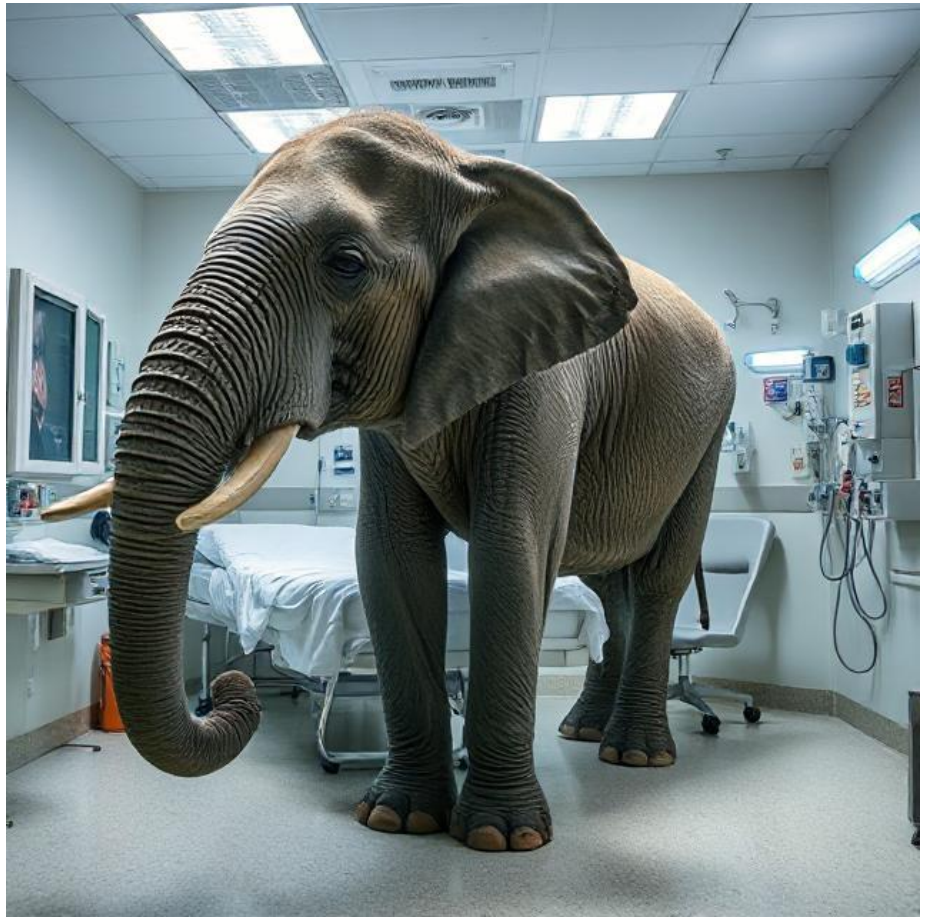
Fluoroquinolones	Systemic disorders	Chronic kidney disease/renal failure	Haematological	Anaemia
Tetracycline class antibiotics		Obstructive sleep apnoea syndrome		Polycythaemia vera
Corticosteroid withdrawal		Chronic obstructive pulmonary disease	Obstruction to venous drainage	Cerebral venous sinus thrombosis
Danazol		Systemic lupus erythematosus		Jugular vein thrombosis
Vitamin A derivatives (including isotretinoin and all-transretinoic acid)		Psittacosis		Superior vena cava syndrome
Levothyroxine	Endocrine	Addison's disease		Jugular vein ligation following bilateral radical neck dissection
Nalidixic acid		Adrenal insufficiency		Increased right heart pressure
Tamoxifen		Cushing's syndrome		Arteriovenous fistulas
Ciclosporin		Hypoparathyroidism		Previous infection or subarachnoid haemorrhage causing decreased CSF absorption
Levonorgestrel implant		Hypothyroidism		
Lithium		Hyperthyroidism		
Growth hormone	Syndromic	Down syndrome		
Indomethacin		Craniosynostosis		
		Turner syndrome		

Tips

- Optic nerve swelling ≠ Papilloedema
  - Papilloedema = optic nerve swelling due to raised ICP
- Headache in women with obesity ≠ IIH
  - Most likely migraine and manage as such
  - But do fundoscopy
- Headache with empty sella ≠ IIH
  - Most likely migraine
- Rapid visual loss with optic nerve swelling = Visual threatening IIH until proven otherwise (think about differentials)

Rapid visual loss = Visual threatening Optic Nerve Swelling

- Rapid onset of symptoms
- Visual acuity loss is a LATE sign
- New visual field impairment?
- Papilloedema severity?
  - Try to grade on fundoscopy
  - Slit lamp /Optical coherence tomography
- Cause?
  - Recent weight gain - Avoid obesity stigma, Medication, Anaemia etc



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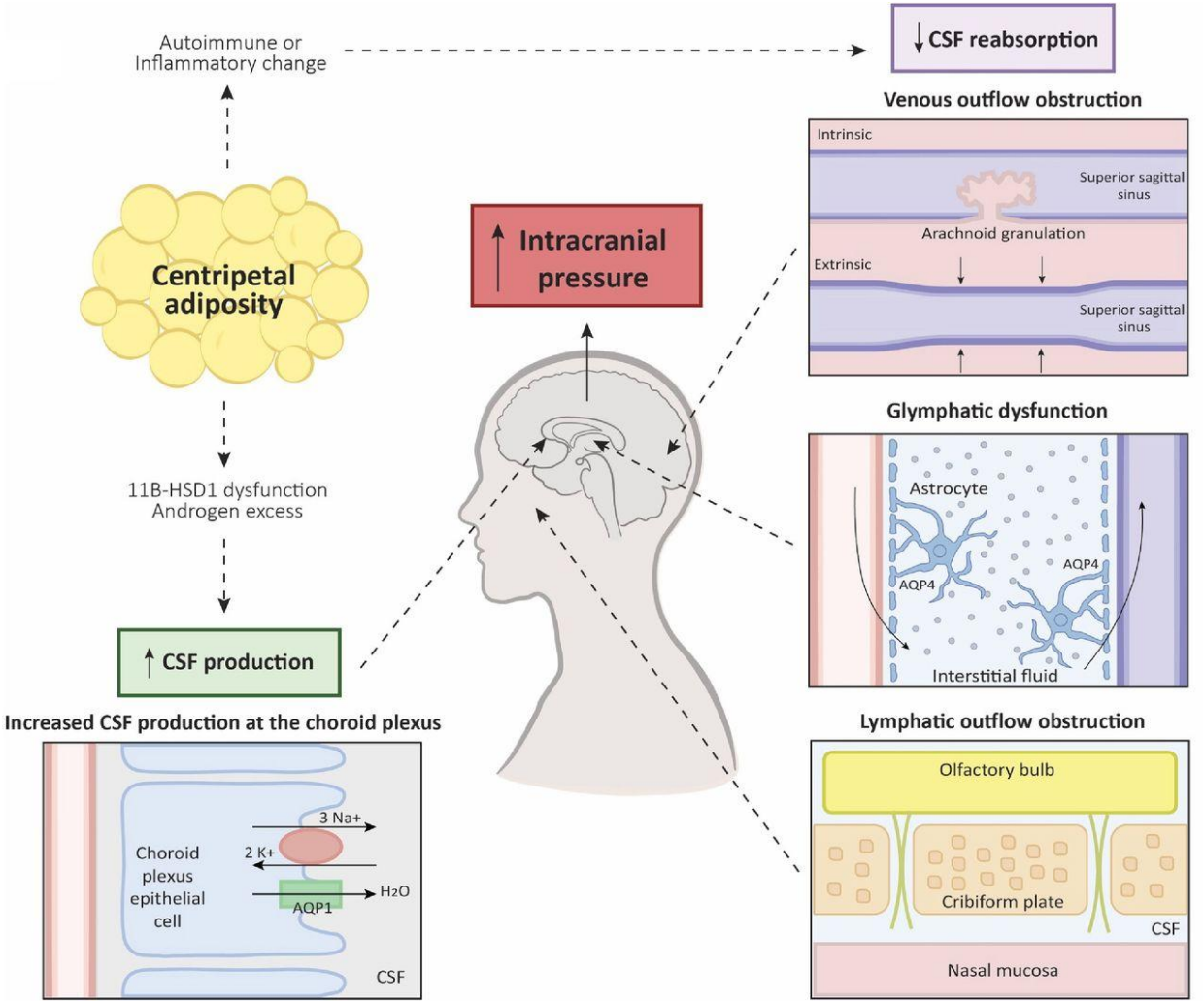
Management

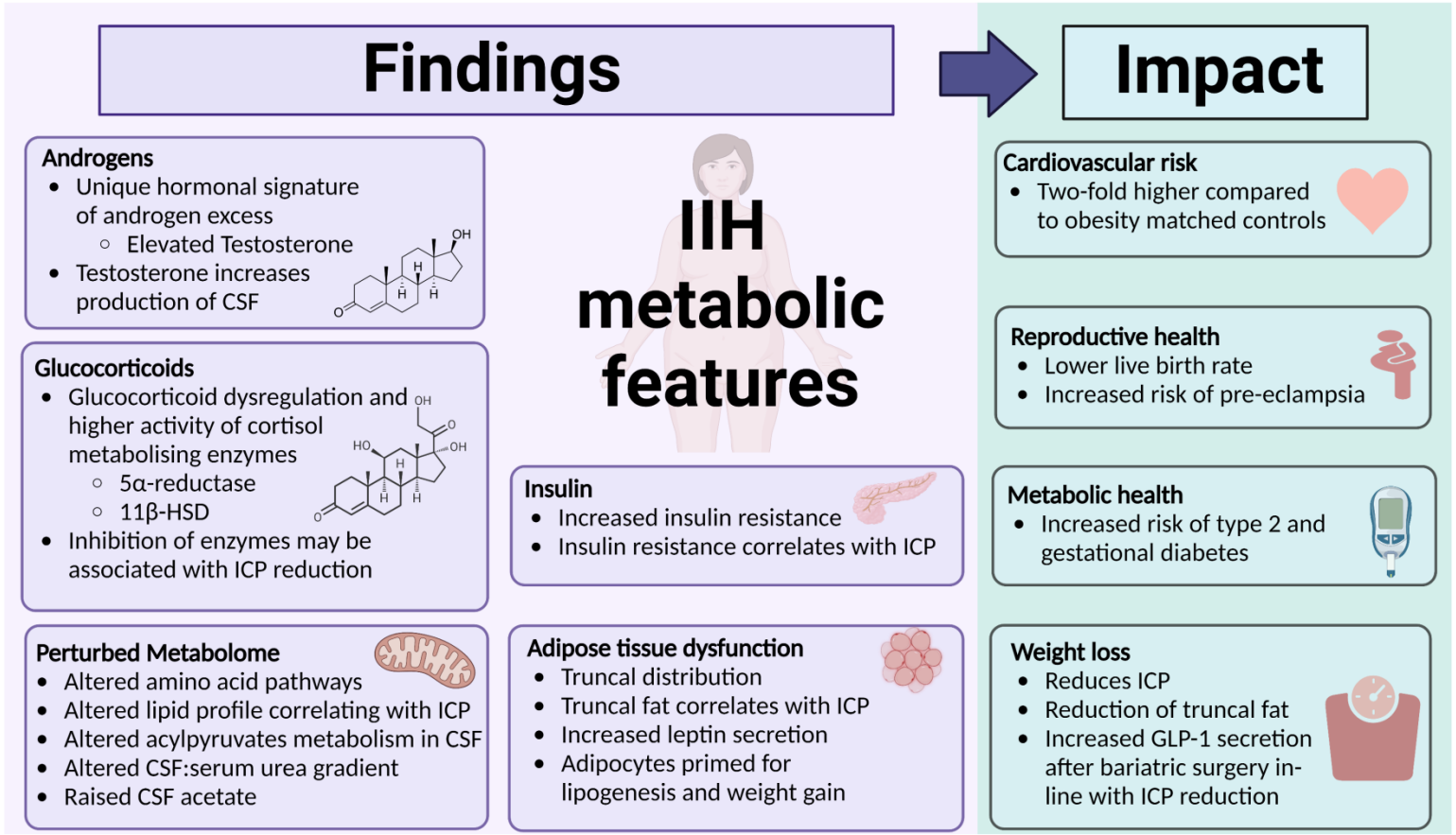
Complex mechanistic interplay



Colman et al. 2023. JNNP

Review at: Yiangou et al. Nature Reviews Neurology. 2023.

IIH metabolism summary



OSA PCOS

TRANSLATIONAL BRAIN SCIENCE

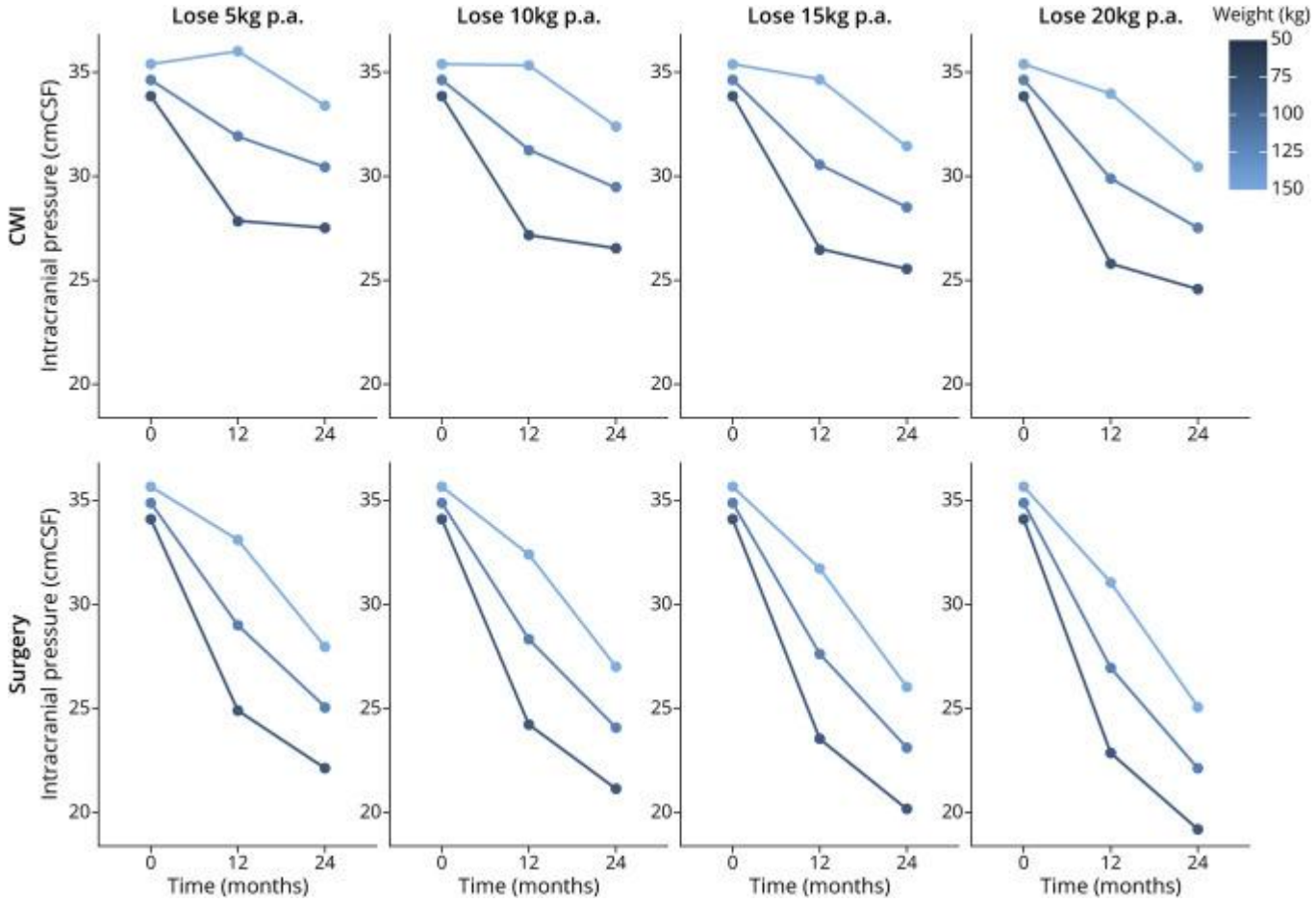
Management

1. Lifestyle/Weight management *
2. Medical
    - Acetazolamide
    - Topiramate (NB Pregnancy)
    - ?Furosemide
    - *GLP-1 Receptor agonists
    - Headache
3. Surgical
    - Ventriculoperitoneal shunt
    - Venous stenting
    - Optic Nerve Sheath Fenestration
    - *Bariatric surgery

TRANSLATIONAL BRAIN SCIENCE

Lifestyle and weight management

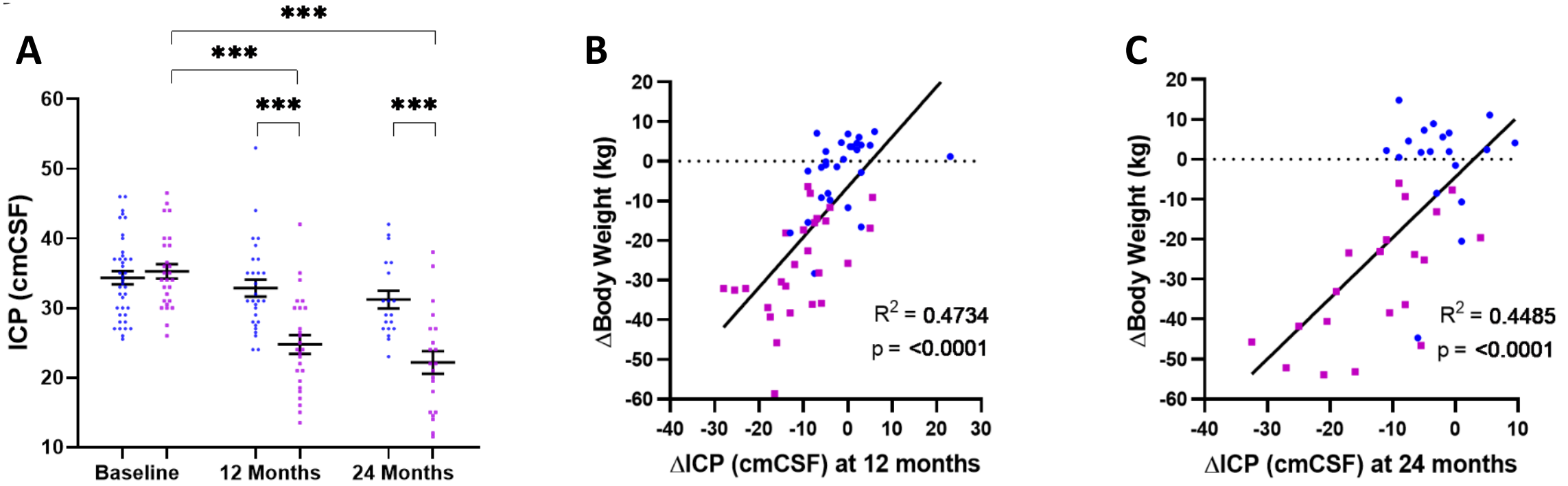
- Avoid obesity stigma
- Weight loss reduces ICP
  - Significant amounts 24% TBW
- Maintenance of loss hard



Mollan 2022. Neurology

TRANSLATIONAL BRAIN SCIENCE

Weight loss improves ICP – sig. more in Bariatric surgery



Bariatric surgery significantly reduced intracranial pressure
Weight loss correlated significantly with ICP
Often substantial weight loss is required to normalise ICP
Cost effective

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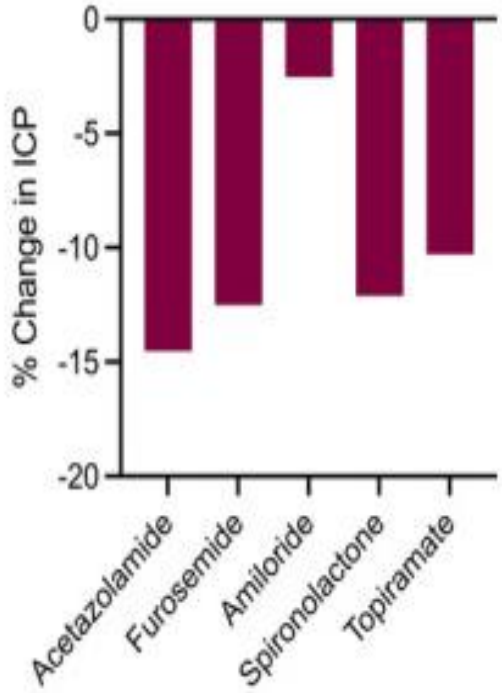
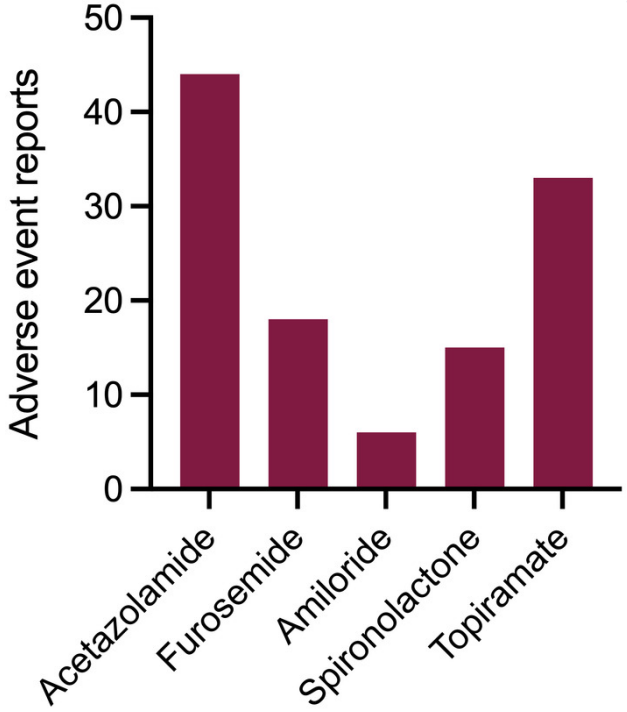
Mollan et al. JAMA Neurology 2021

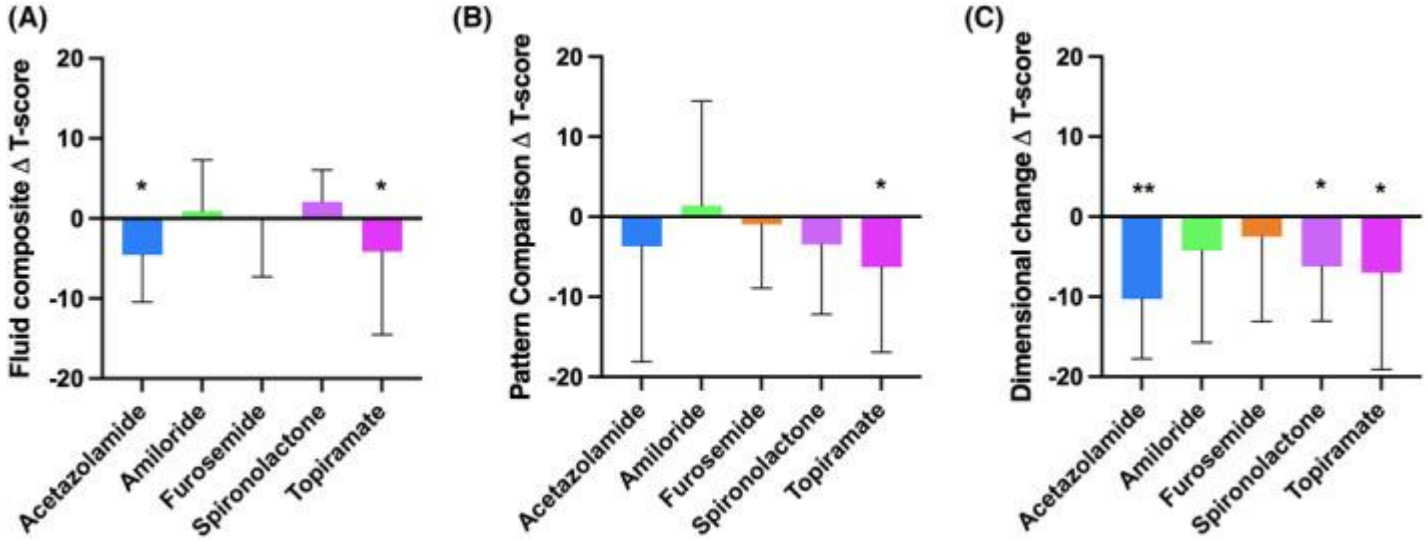
Oral Medications

- Acetazolamide
  - Target 4g/day
- Topiramate
  - Target 100mg BD
  - Contraception/PPP
- Furosemide?

- SIDE EFFECTS
  - Cognition
  - Paraesthesia
  - Weight loss







Mitchell JL. Headache. 2025

The Idiopathic Intracranial Hypertension Treatment Trial

The NORDIC Idiopathic Intracranial Hypertension Study Group Writing Committee. Effect of Acetazolamide on Visual Function in Patients With Idiopathic Intracranial Hypertension and Mild Visual Loss: The Idiopathic Intracranial Hypertension Treatment Trial. *JAMA*. 2014;311(16):1641–1651.

Objective

To determine whether acetazolamide, when added to a low-sodium weight reduction diet, is beneficial to vision in patients with IIH and mild visual loss

Design & Setting

- Randomised, double-masked, placebo-controlled study
- Multicentre: enrollment at 38 academic and private practice centres in North America
- Duration: 6 months; assessments at screening, baseline and months 1, 2, 3, 4½ and 6

317 participants assessed for eligibility

97.6% female; mean age 29; mean BMI 40

Eligible Participants

18-60 years old

Met modified Dandy criteria (1985) for diagnosis

Mild visual loss with reproducible PMD between -2 dB and -7 dB in worst affected eye

New onset and previously untreated

165 participants randomised

		Primary Outcome	Selected Secondary Outcomes			Change in Weight
		Change in PMD	Papilloedema Grade	Vision Related QoL	Headache Related QoL	
				VFQ-25	HIT-6	
Intervention n=86	Low sodium weight reduction diet + Acetazolamide 1-4g daily	**1.43** (-3.53 to -2.10)	**-1.31** (2.76 to 1.45)	**+8.33** (82.97 to 91.30)	**-9.56** (59.7 to 50.14)	**-7.5kg** (107.72 to 100.22kg)
Control n=79	Low sodium weight reduction diet + Placebo	**0.71** (-3.53 to -2.82)	**-0.61** (2.76 to 2.15)	**+1.98** (82.97 to 84.95)	**-9.11** (59.70 to 50.59)	**-3.45kg** (107.72 to 104.27kg)
		p=0.050	***p<0.001***	***p=0.003***	*p=0.77*	***p<0.001***

6 months

Limitations

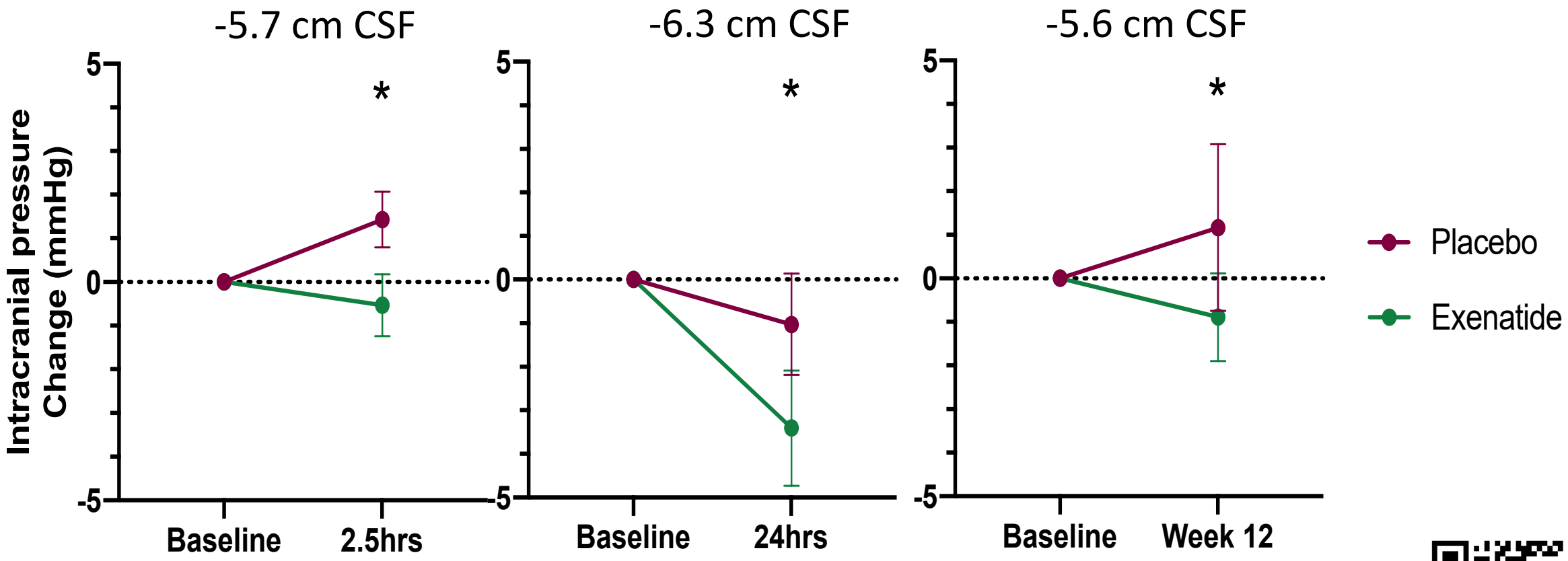
- Almost half of people screened were excluded
- Treatment effect on change in PMD is small and of uncertain clinical significance
- Greater weight loss in treatment group. However, analysis suggested effect on PMD was independent of weight loss

Importance and Practice Implications

- Supports the use of acetazolamide in conjunction with intensive lifestyle intervention in people with IIH with new onset active disease and PMD -2dB to -7dB
- Majority were able to tolerated acetazolamide at a dose above 1g per day for 6 months

Abbreviations: IIH - idiopathic intracranial hypertension; BMI - body mass index; PMD - perimetric mean deviation; QoL - Quality of Life; VFQ-25 - National Eye Institute Visual Functioning Questionnaire 25; HIT-6 - Headache Impact Test

Exenatide: GLP-1 receptor agonist

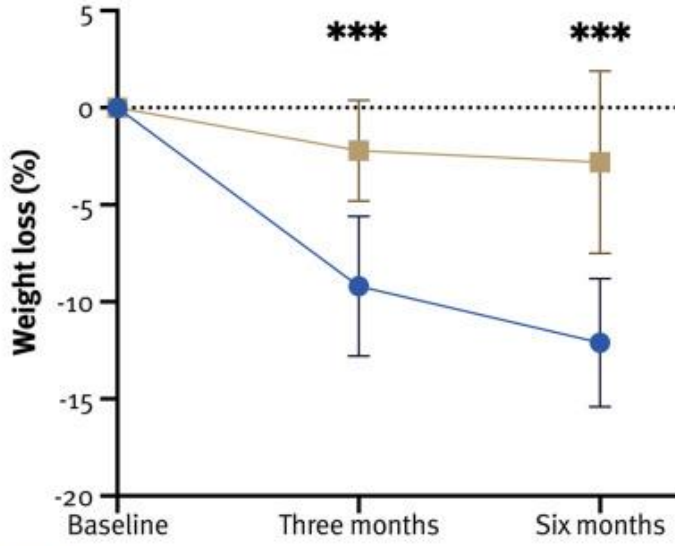


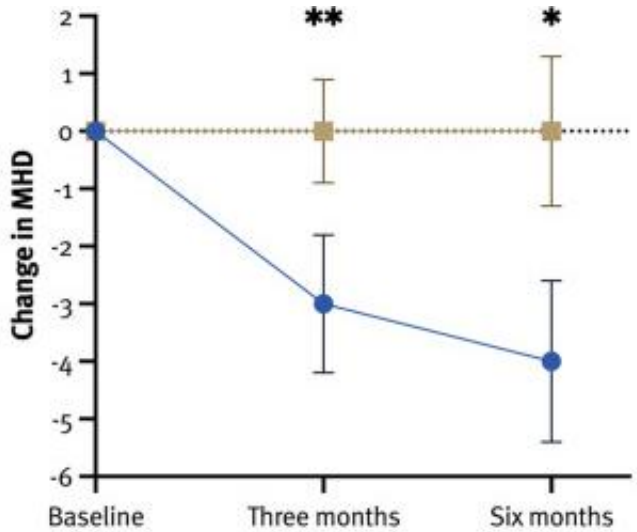
1 mmHg = 1.36 cm CSF ** p<0.1*

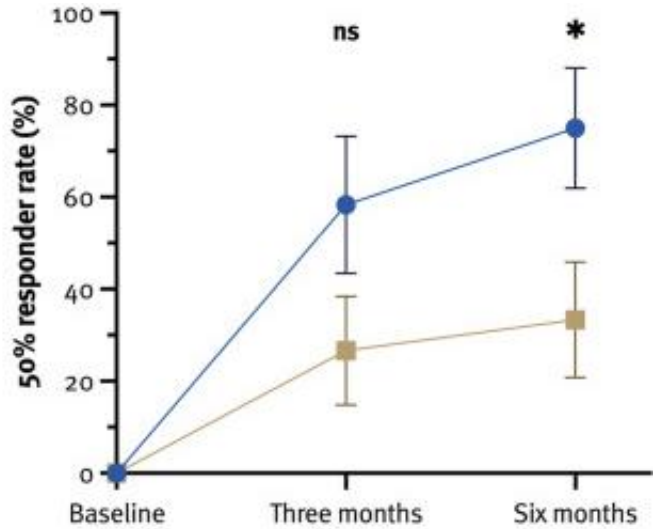
Loading: 20 microgram subcut Exenatide. Ongoing: 10 microgram BD for 12 weeks.

Mitchell et al. 2023. Brain

GLP-1 RA, weight loss and headache?



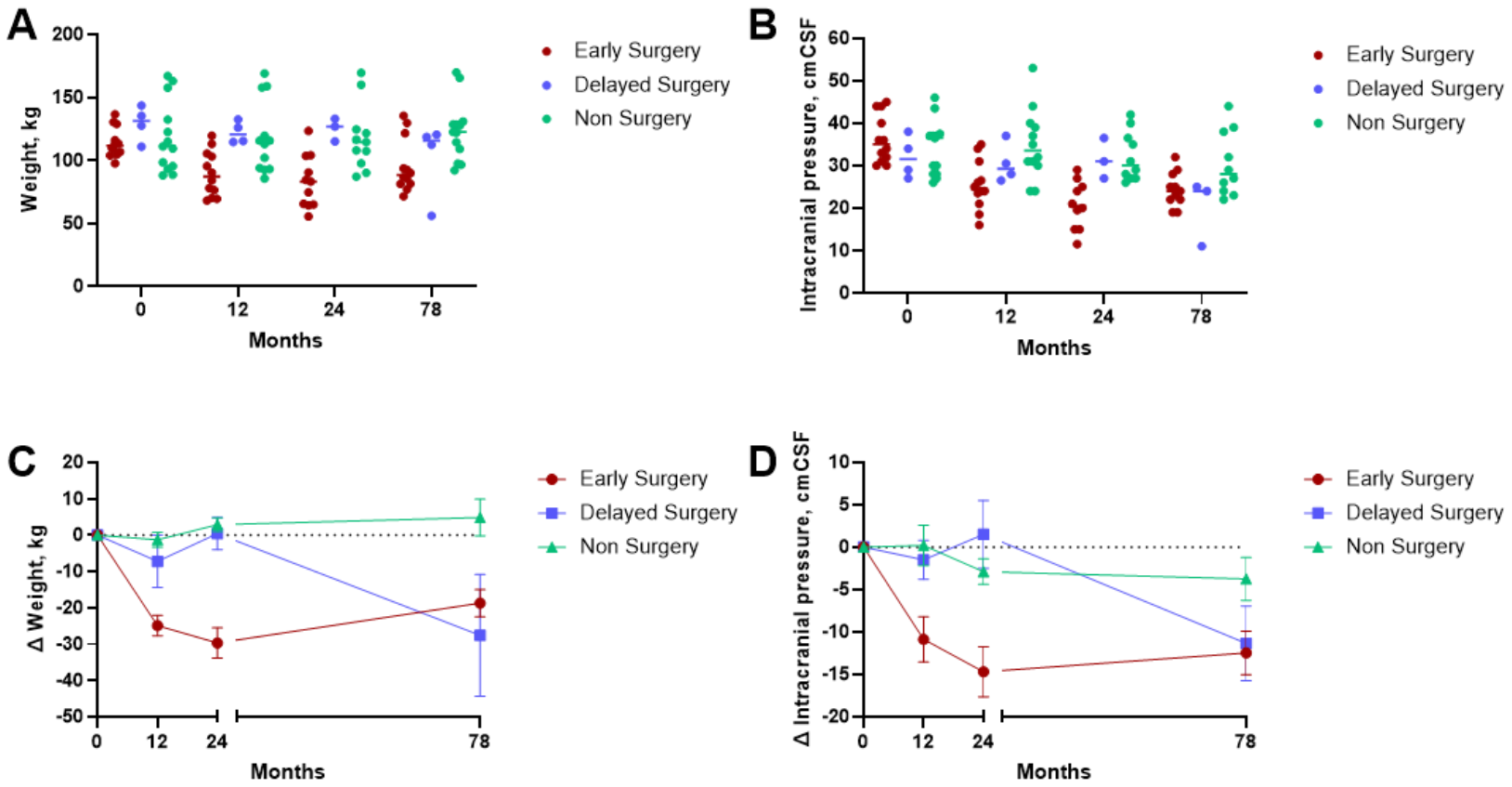




Not a specific GLP1RA and ICP outcomes not measured

Krajnc et al 2023, J Headache Pain

Bariatric surgery – very long-term outcome data shows long term significant benefits



Not yet published – for your information not for dissemination

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Headache – manage according to phenotype

- Topiramate – titrate to 50-100mg BD
  - Was very helpful when tolerated
  - CAUTION “women of childbearing potential”, need highly effective contraception
- Candesartan – titrate to 8mg BD or 16mg OD
  - Monitor BP
  - Not in pregnancy (renal)
- Propranolol - titrate to 80mg BD
  - Not in asthma
  - Possible weight gain
- Amitriptyline – not recommended due to weight gain
- Pizotifen - not recommended due to weight gain
- CGRP therapies
  - Oral antagonists – no direct studies but being increasingly used
  - Monoclonals – beneficial
- Cranial Botox is effective
- Greater occipital nerve blocks

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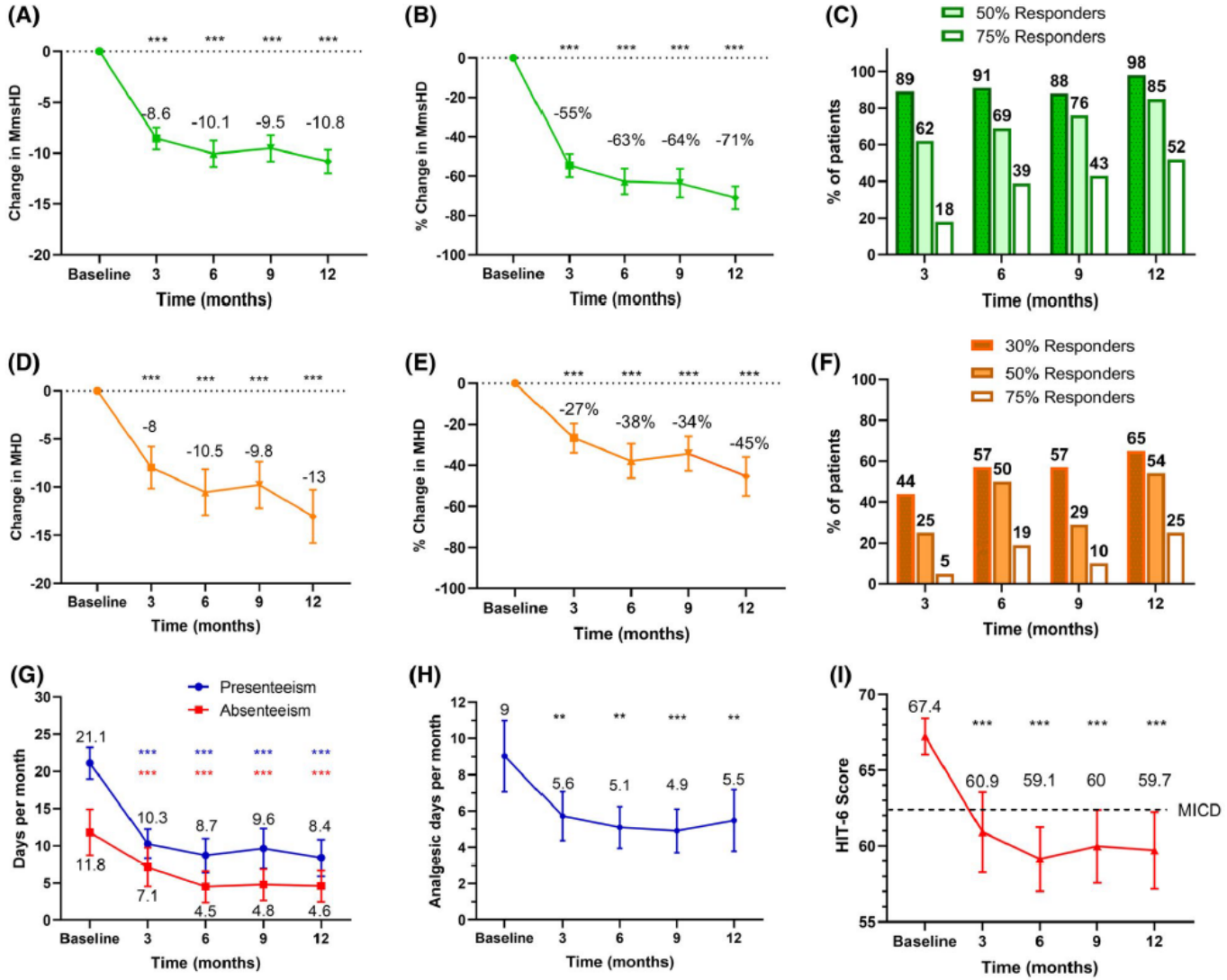
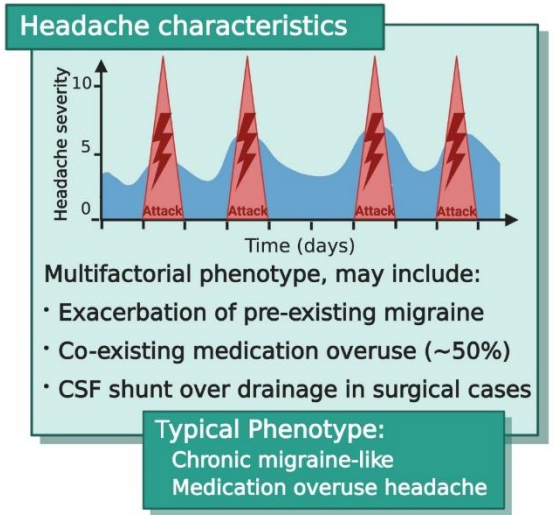
Targeting CGRP in Persistent post-IIH headache

Improvement in monthly migraine like days

Improvement in monthly headache days

Improvement in analgesic days, return to work and quality of life





CGRP monoclonal Ab, Erenumab
n=55 open label, prospective

Eligibility:
Papilloedema resolved + chronic migraine-like headache

Yiangou A et al., Headache. 2021

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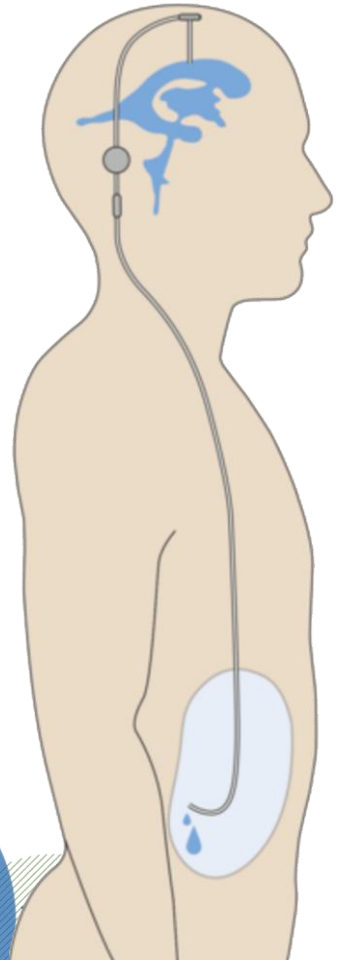
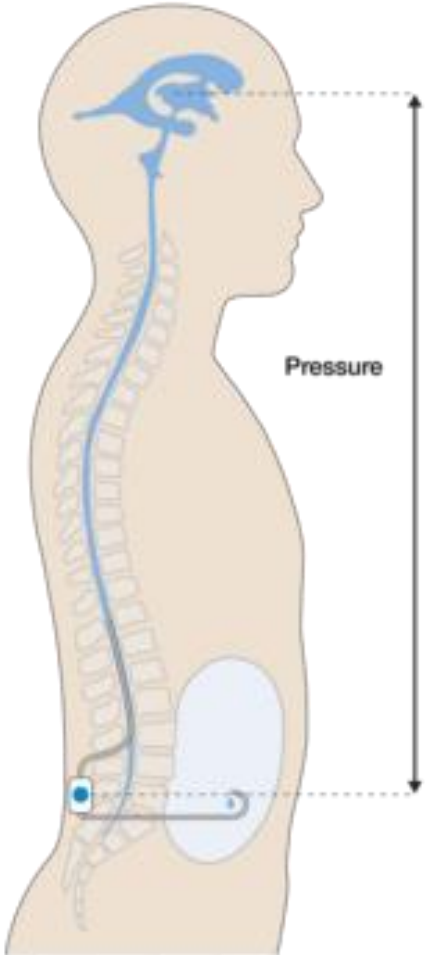
Surgical management – Sight threatening IIH

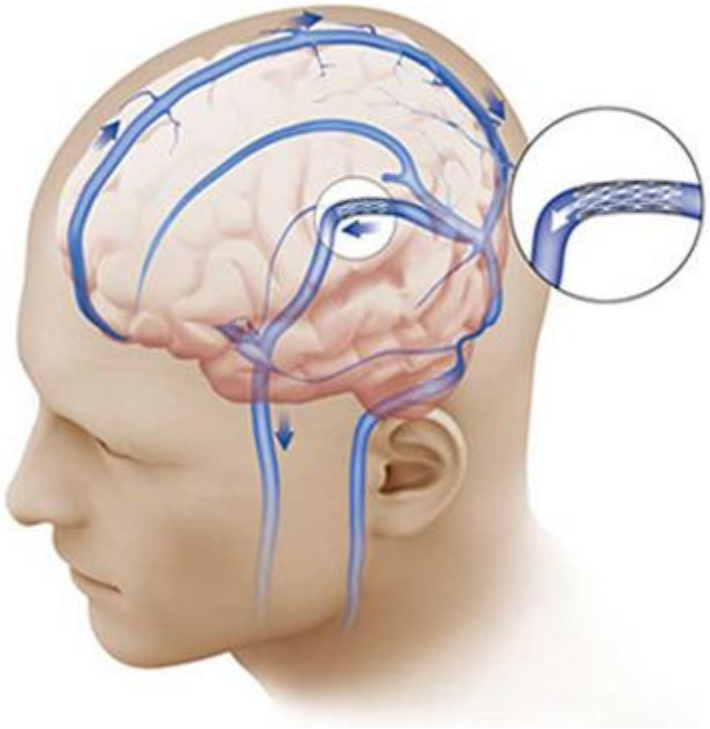
Ventriculoperitoneal VP shunt



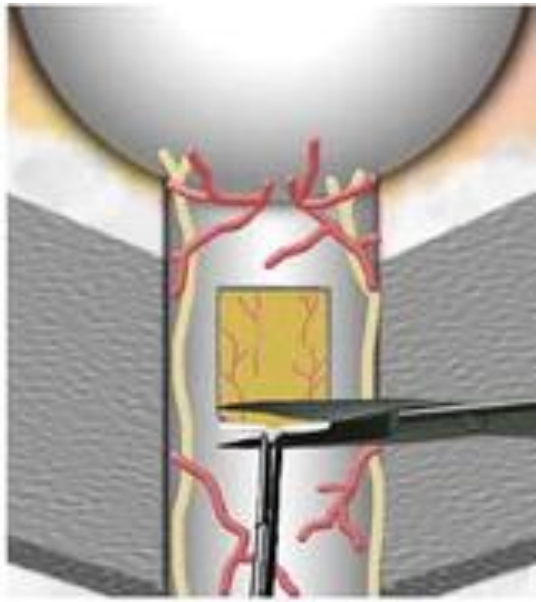
Lumboperitoneal VP shunt



Deep venous sinus stenting (transverse)

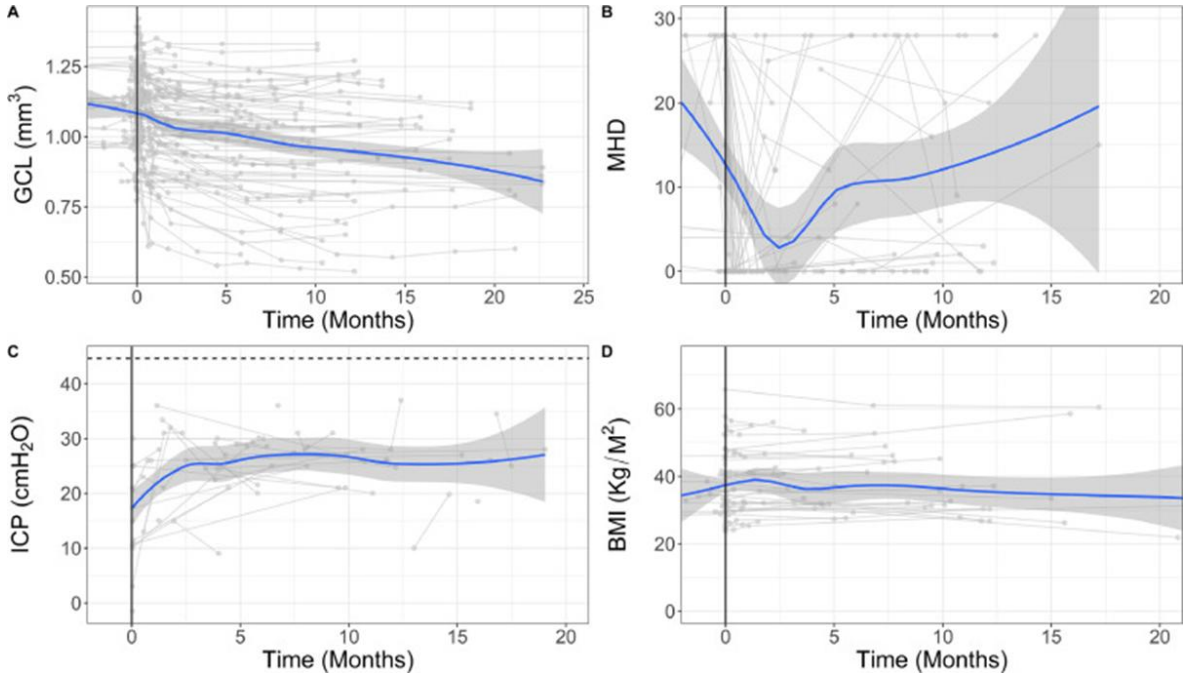


Optic nerve sheath fenestration



Which is best?

- No RCTs yet comparing Shunt to Stent
  - IIH Intervention trial currently recruiting
  - Depends on your local neuroscience centre expertise
  - Birmingham Standardized IIH Shunt Protocol – Tsermoulas 2022 – reduced revision rate
- Do not…
  - Shunt/stent for headache
  - Request serial lumbar punctures routinely
    - If needed for vision preservation -> Surgery

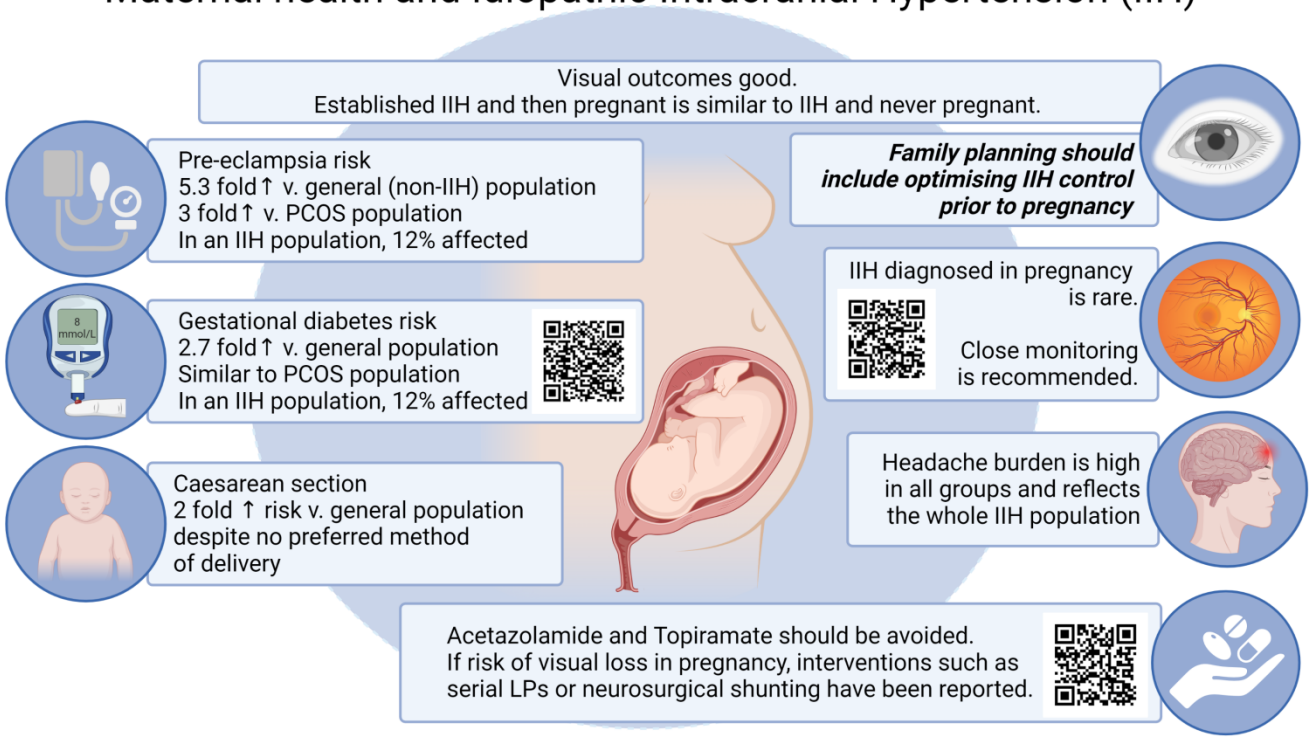


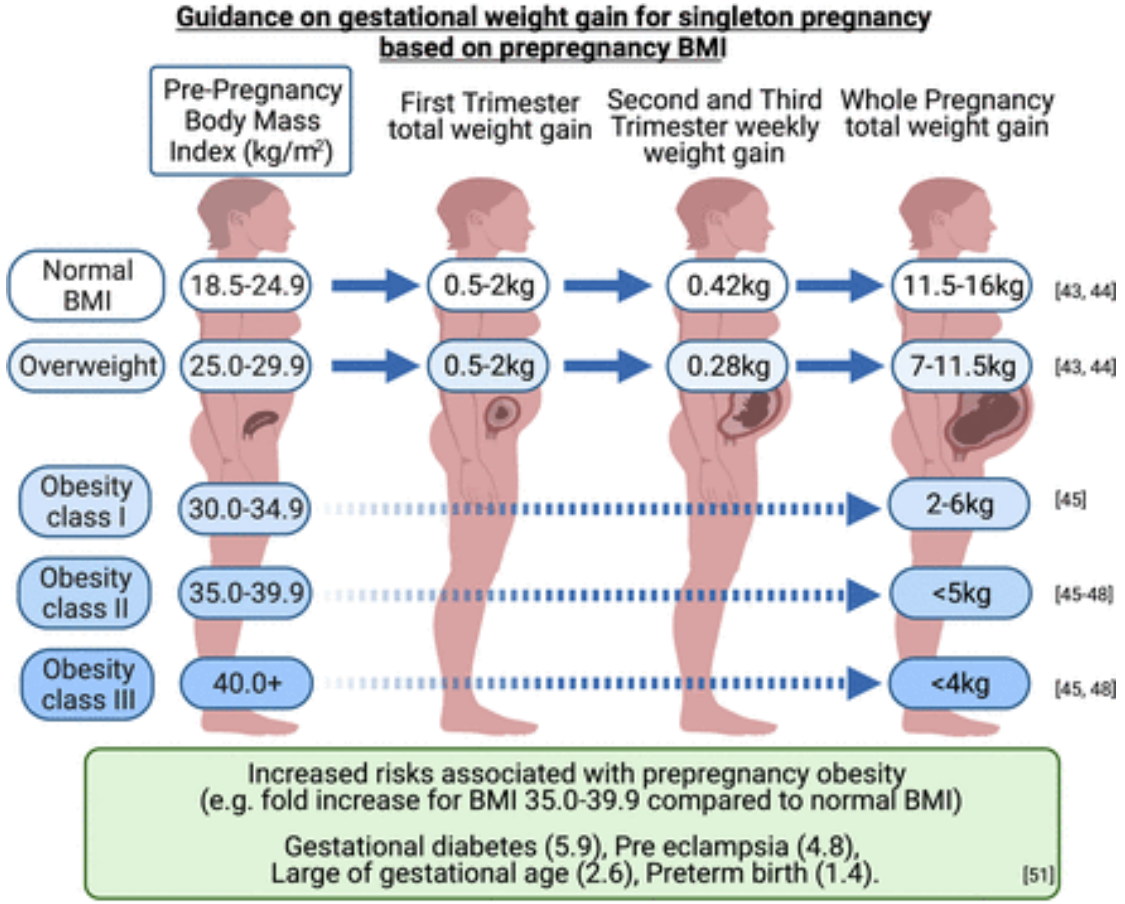
CURRENT RESEARCH

Venous stenting v Ventriculoperitoneal shunting for Sight threatening papilloedema

Pregnancy management

Maternal health and Idiopathic Intracranial Hypertension (IIH)





Active labour: Valsalva manoeuvre associated with 40 cmCSF rise in ICP but back at baseline within 16 seconds. In general, no need to avoid from ICP perspective.

Yiangou et al. 2022. Neuro-ophthalmology.

Thaller et al 2022, Practical Neurology

Idiopathic Intracranial Hypertension Conclusions

Mollan et al. Idiopathic intracranial hypertension: consensus guidelines on management. JNNP 2018.

- Need for accurate diagnosis
- Neurometabolic disorder
- Increased risk of pre-eclampsia (5x), cardiovascular risk (2x) and PCOS (1.5x)
- **Debilitating headaches** – need better options
- Visual outcomes good with appropriate escalation, monitoring and management
- Management: **Weight loss**, Acetazolamide, **GLP1RA**, anti-CGRP, Bariatric surgery, VP/DVSS/ONSF Surgery

TRANSLATIONAL BRAIN SCIENCE

Artwork by Alister McCracken IIHUK 2021 winner

Acknowledgements

Translational Brain Science team:
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Dr Hannah Lyons
Dr Yousef Hyder
Dr Matilde Sassani
Ms Jessica Hubbard

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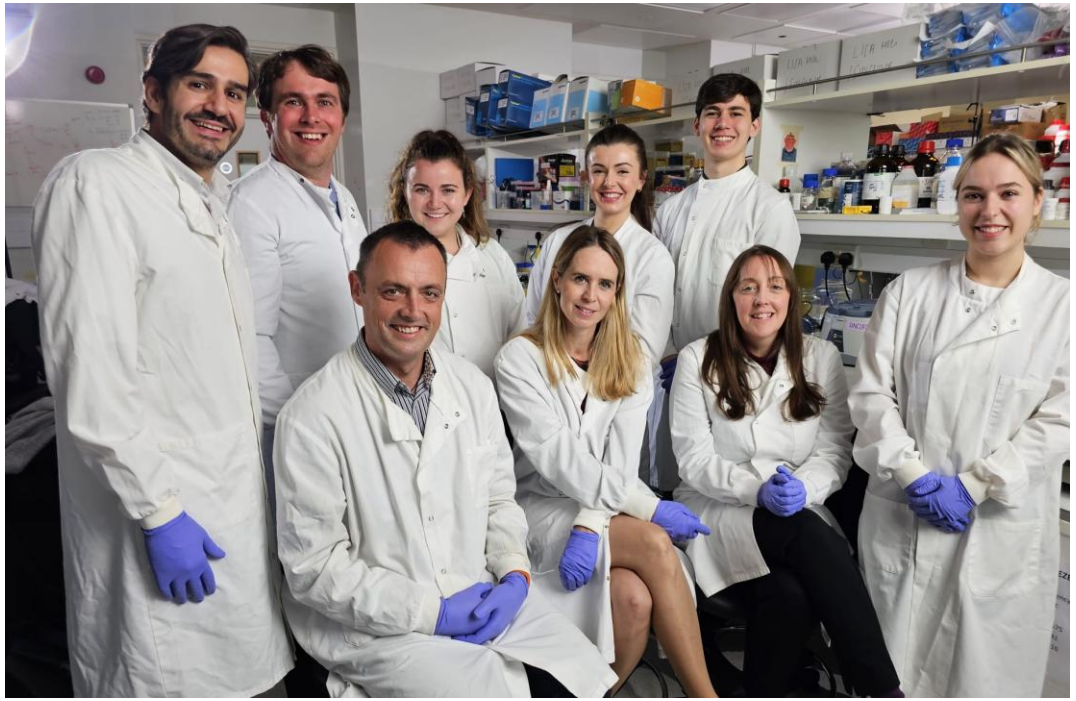
Ms Gabriele Berman
Mrs Stephanie Gregg
Mrs Sally Ware
Ms Sarah Stuart
Mrs Zee Zimdahl
Ms Claire Fisher



All researchers, staff members and patients who have contributed over the years.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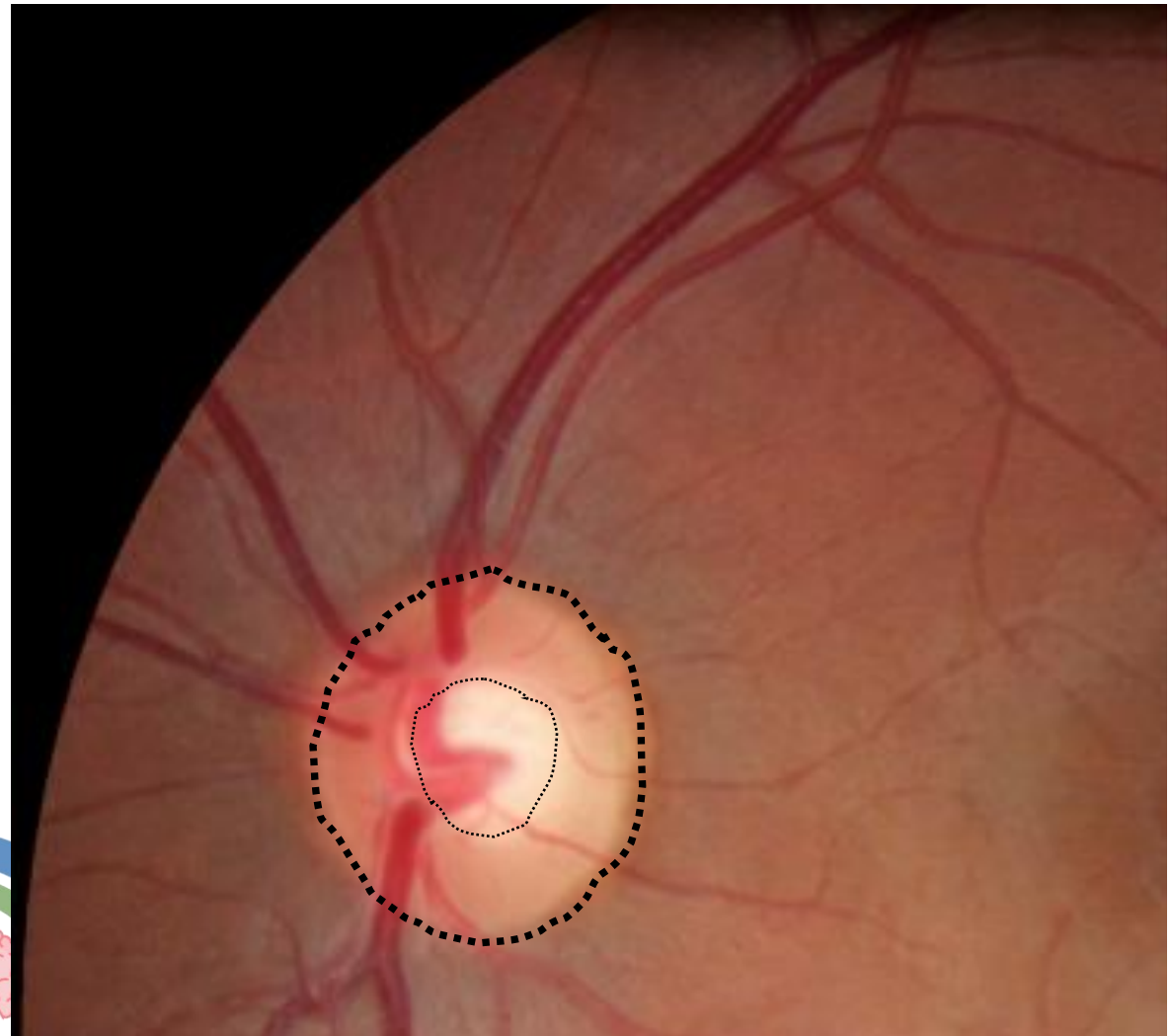
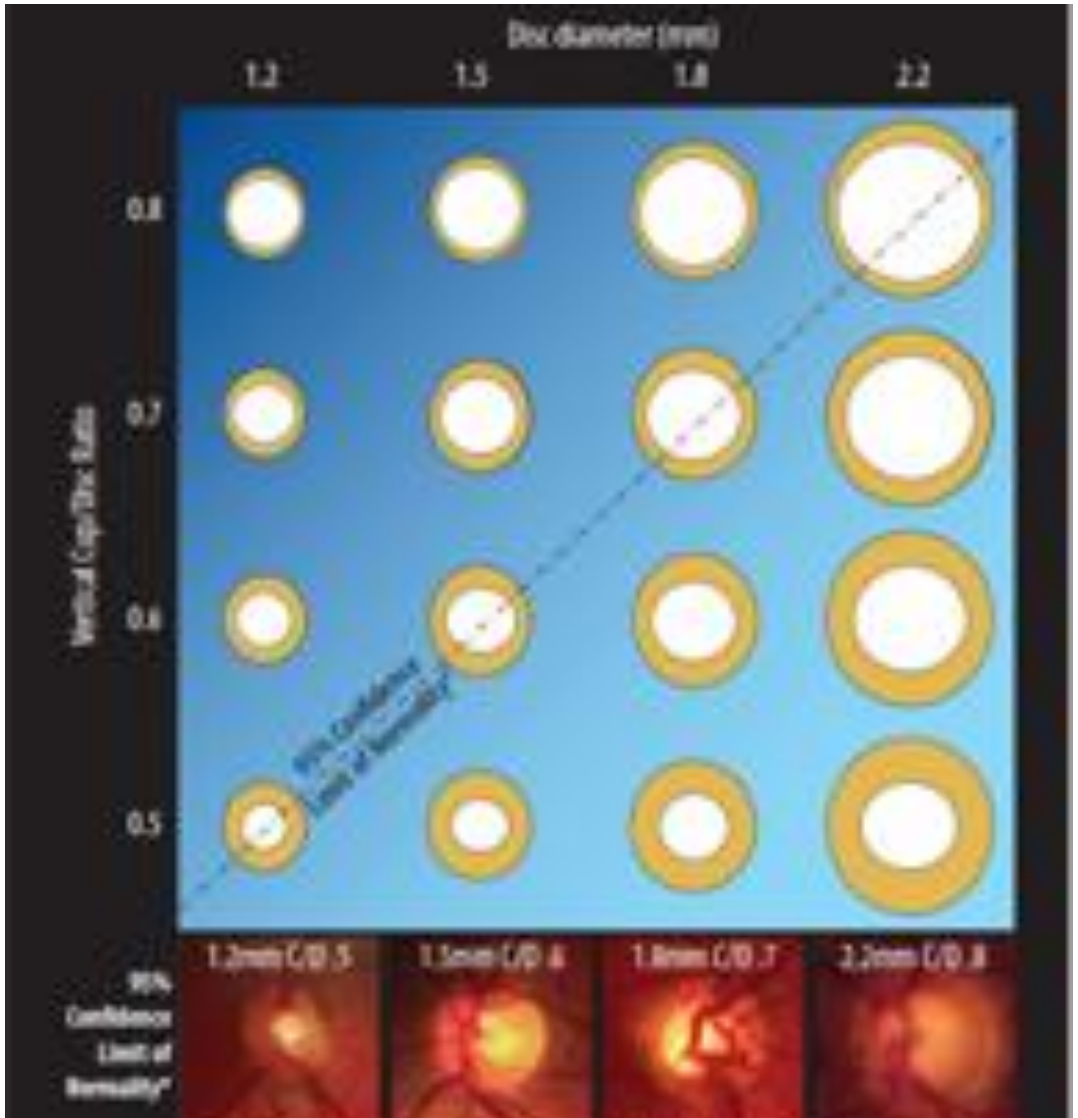
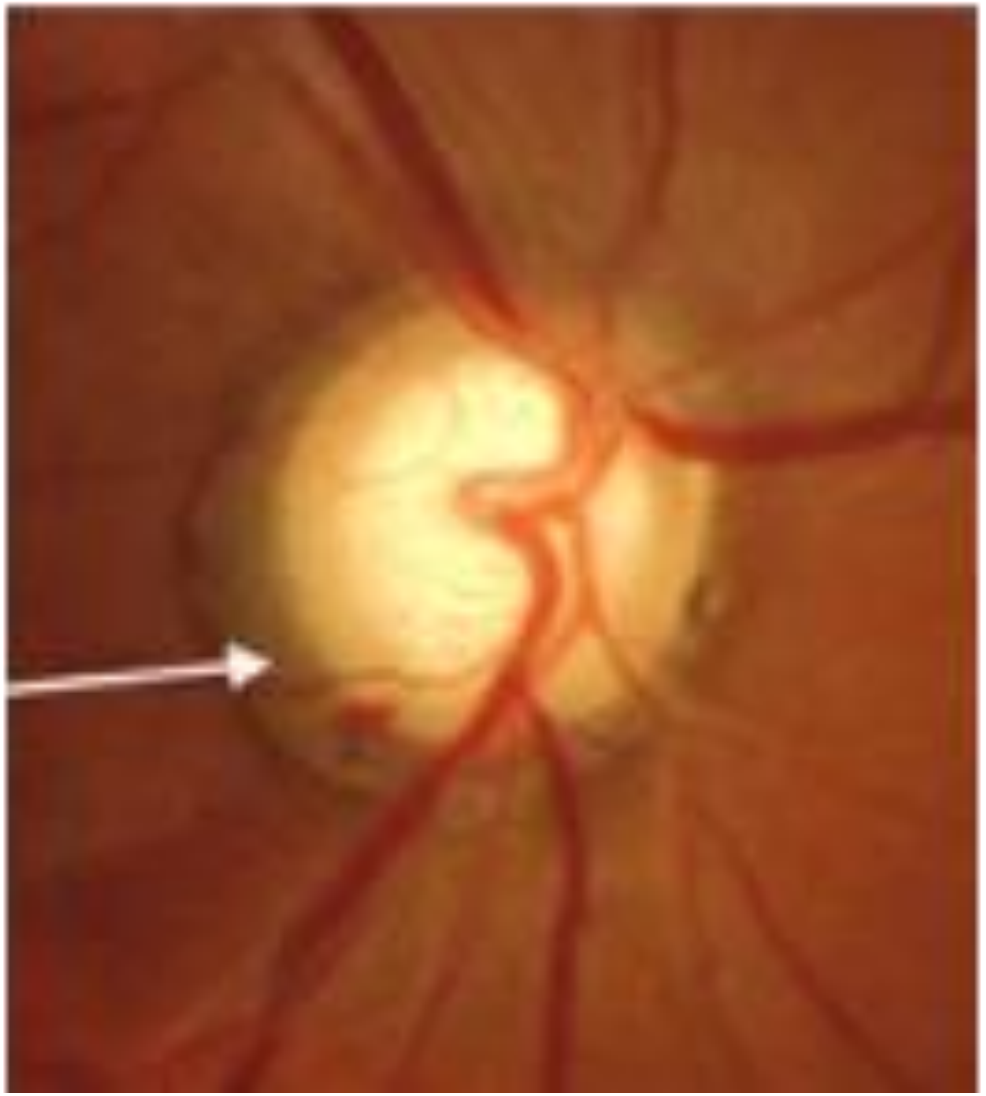
BRAIN SCIENCE

What do they mean about a cup??





What is peripapillary atrophy?



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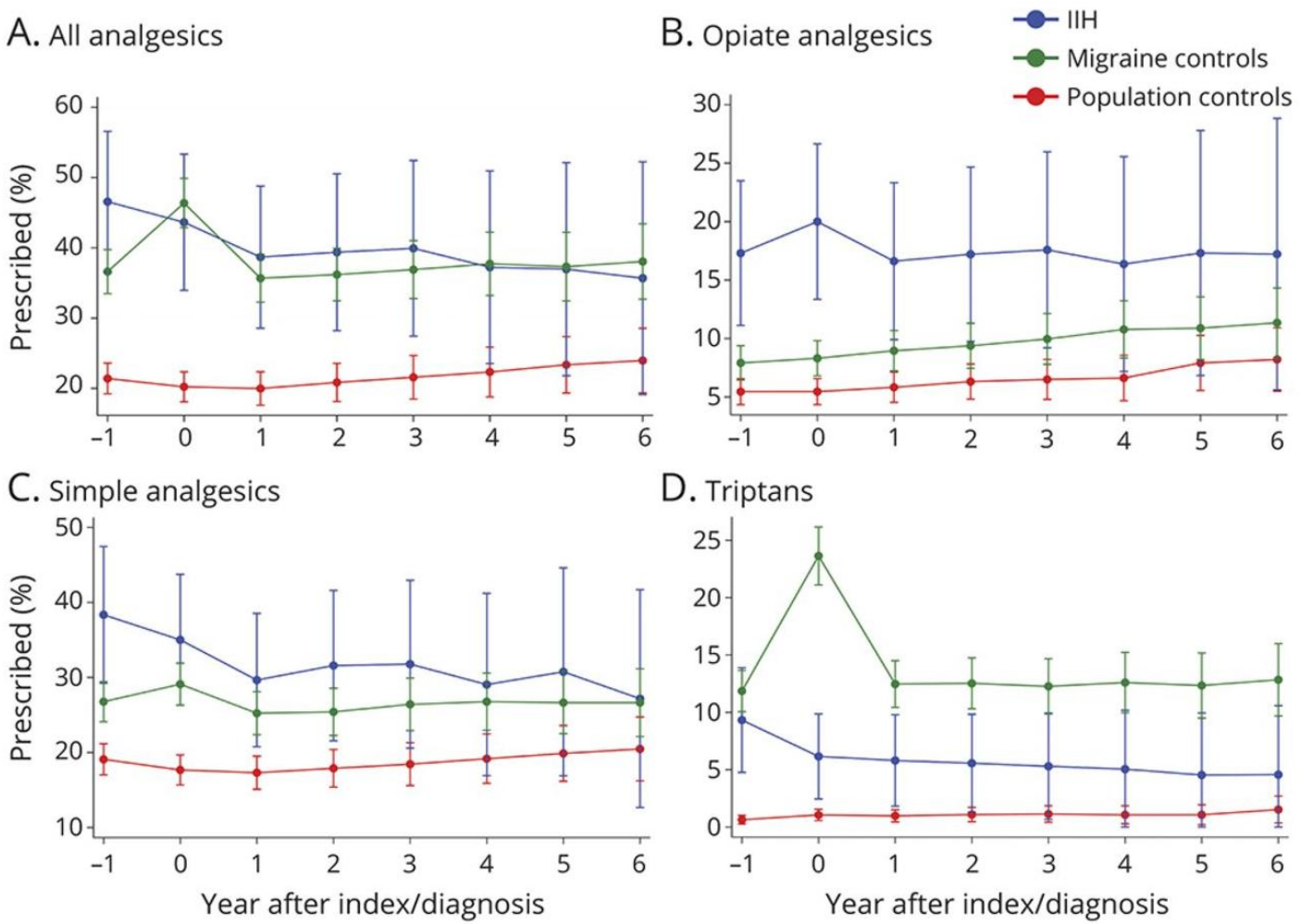
Headaches and opiates – risk of MOH



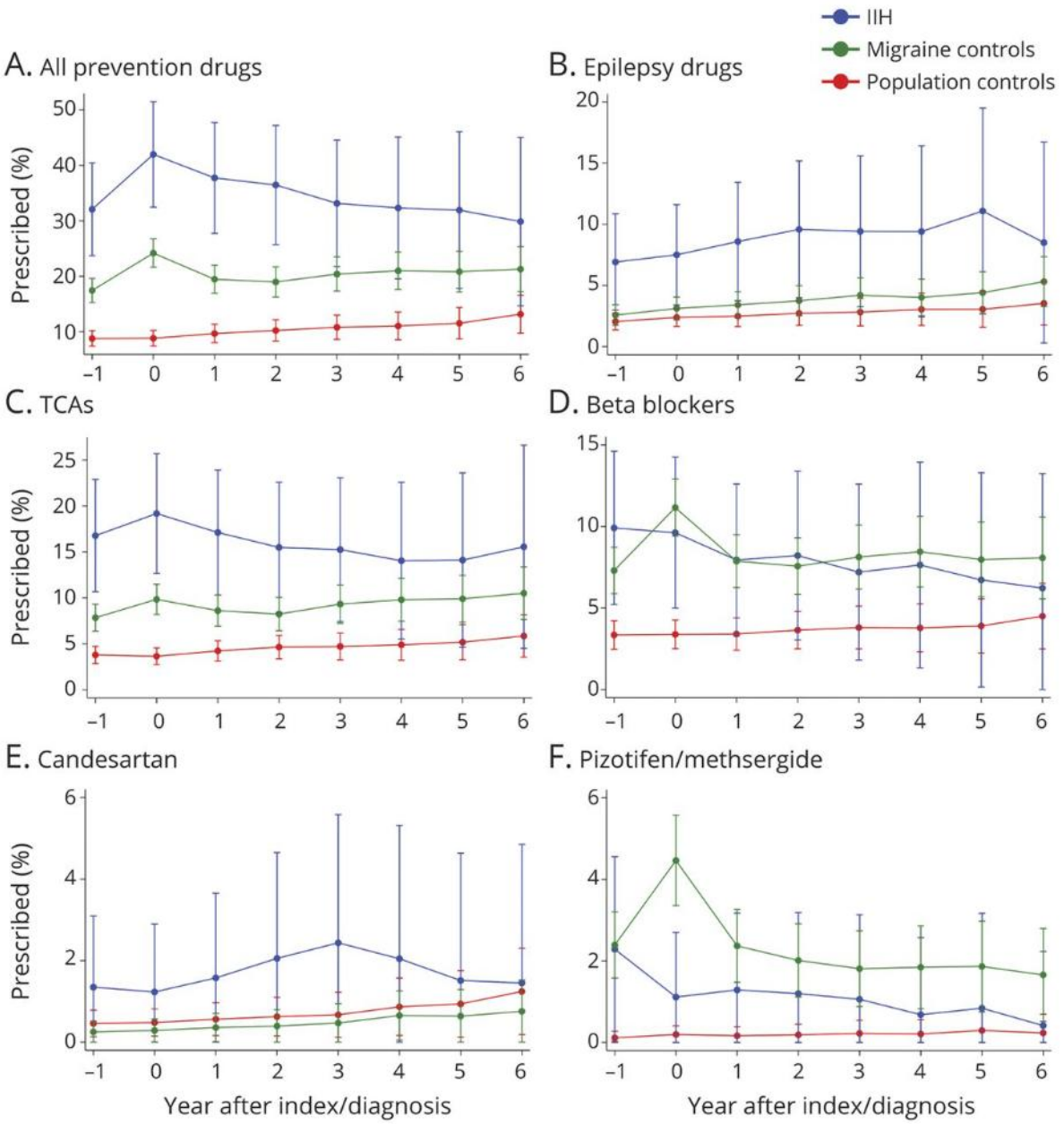
Population matched cohort study
2x ↑ in IIH versus migraine controls
3x ↑ in IIH versus population controls

Adderley et al 2022 Neurology

Headaches and preventatives

High headache burden in IIH

Preventative prescriptions
IIH > Migraine



Adderley et al 2022
Neurology

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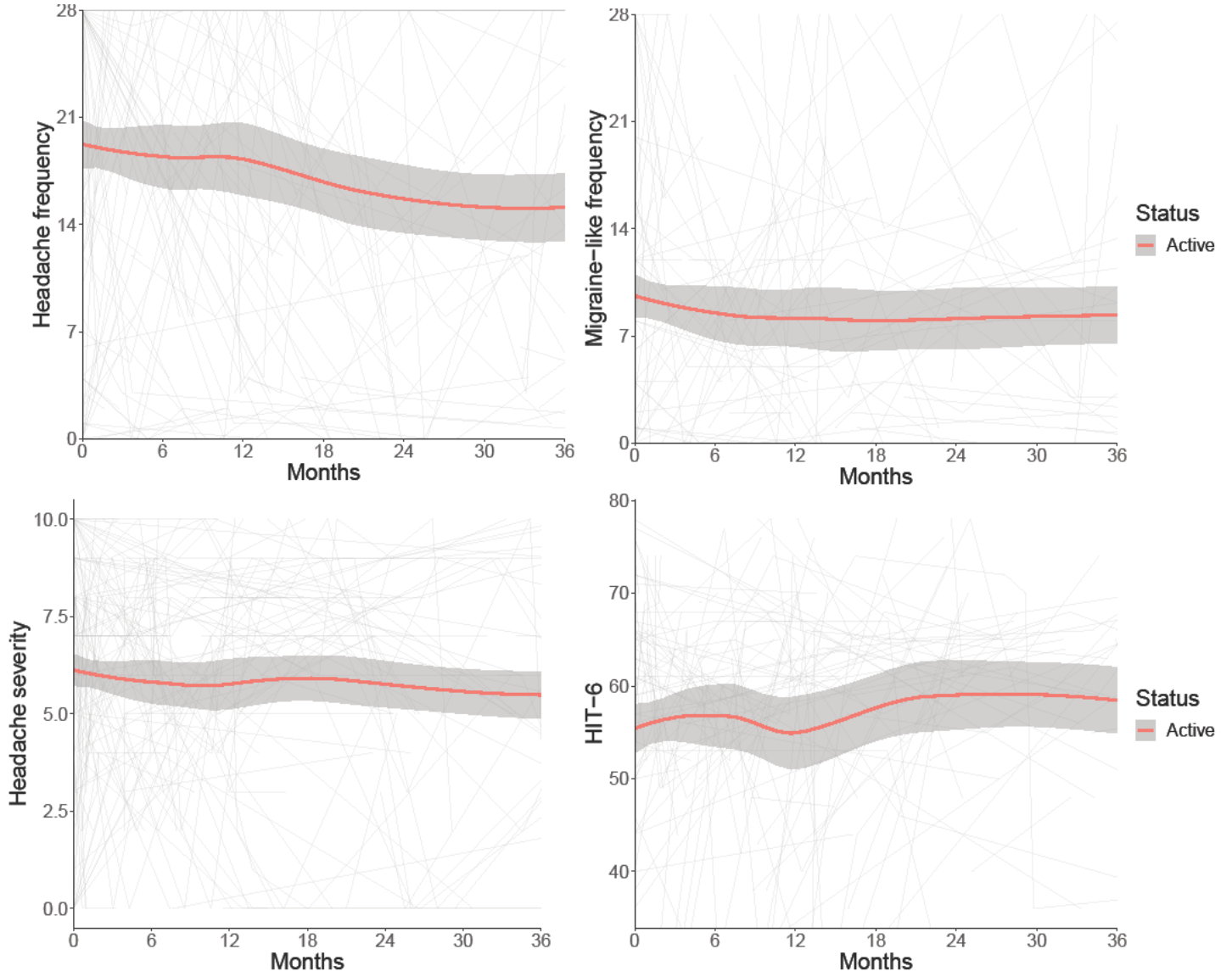
Longitudinal outcomes medically treated cohort

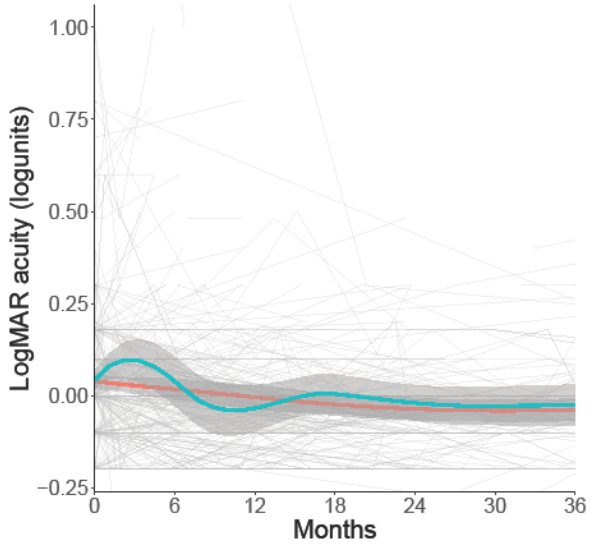
Largest IIH prospective cohort study

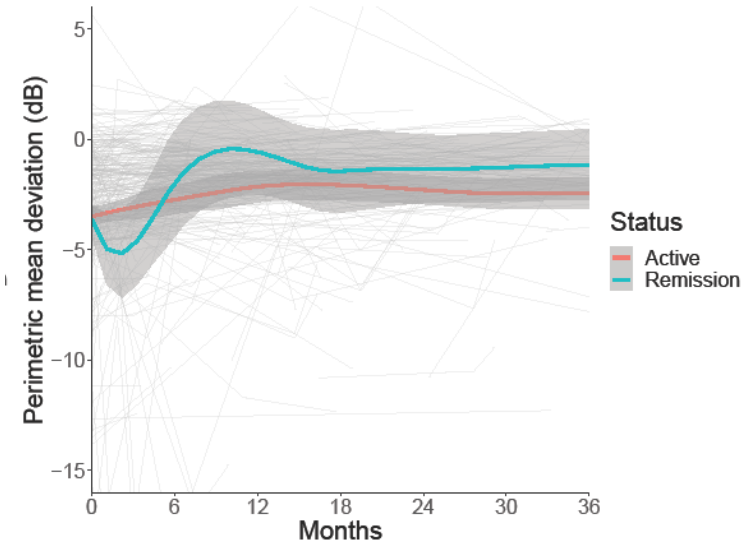
N=490

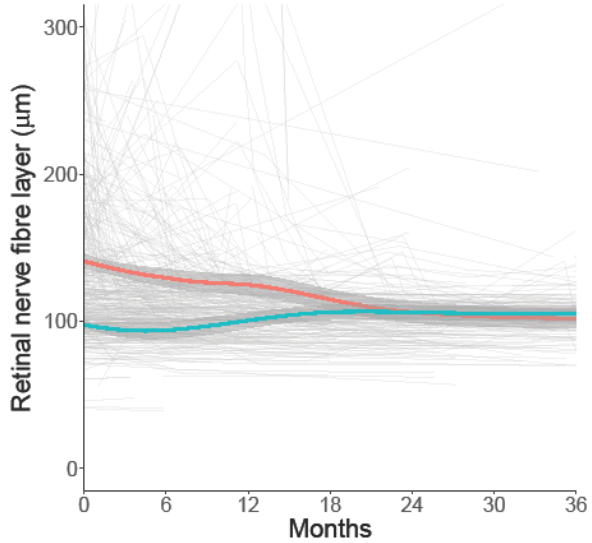


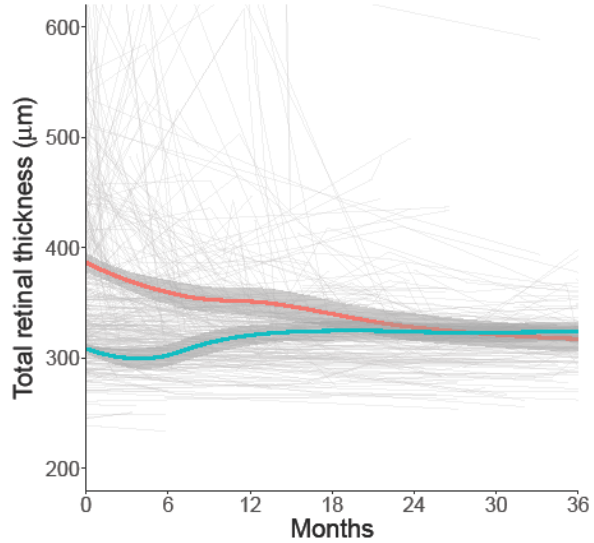
Thaller et al. J Neurology. 2022

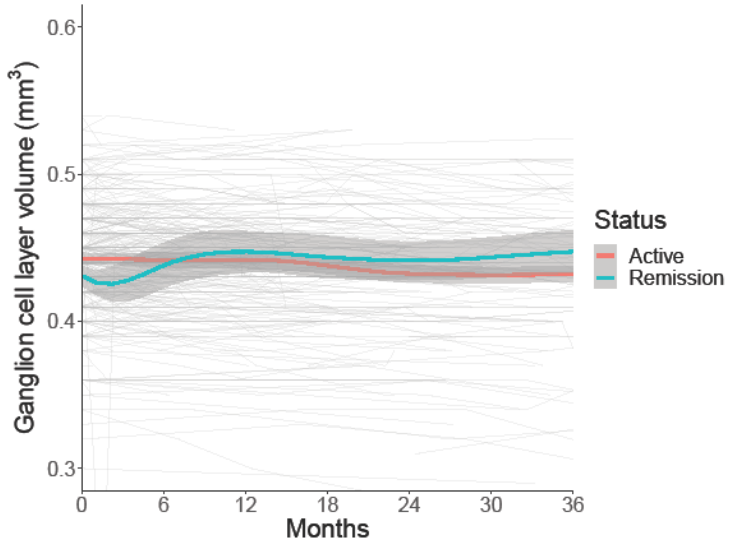
Vision - OCT better marker of disease activity











Thaller et al. J Neurology. 2022