


Idiopathic Intracranial Hypertension: Updates on Imaging Findings, Pathophysiology and Treatment

Charif Sidani. MD



Learning objectives

- Classic imaging findings IIH
- Pathophysiology  Imaging
- Venous Stenting
- IIH vs sCSF leak “spectrum”
- IIH vs Spaceflight-Associated Neuro-Ocular Syndrome (SANS)

Intracranial Hypertension

American Family Physician:

- Normal opening pressure ranges:
 - 10 to 100 mm H₂O in young children
 - 60 to 200 mm H₂O > 8 years of age
 - up to 250 mm H₂O in obese patients

Intracranial Hypertension

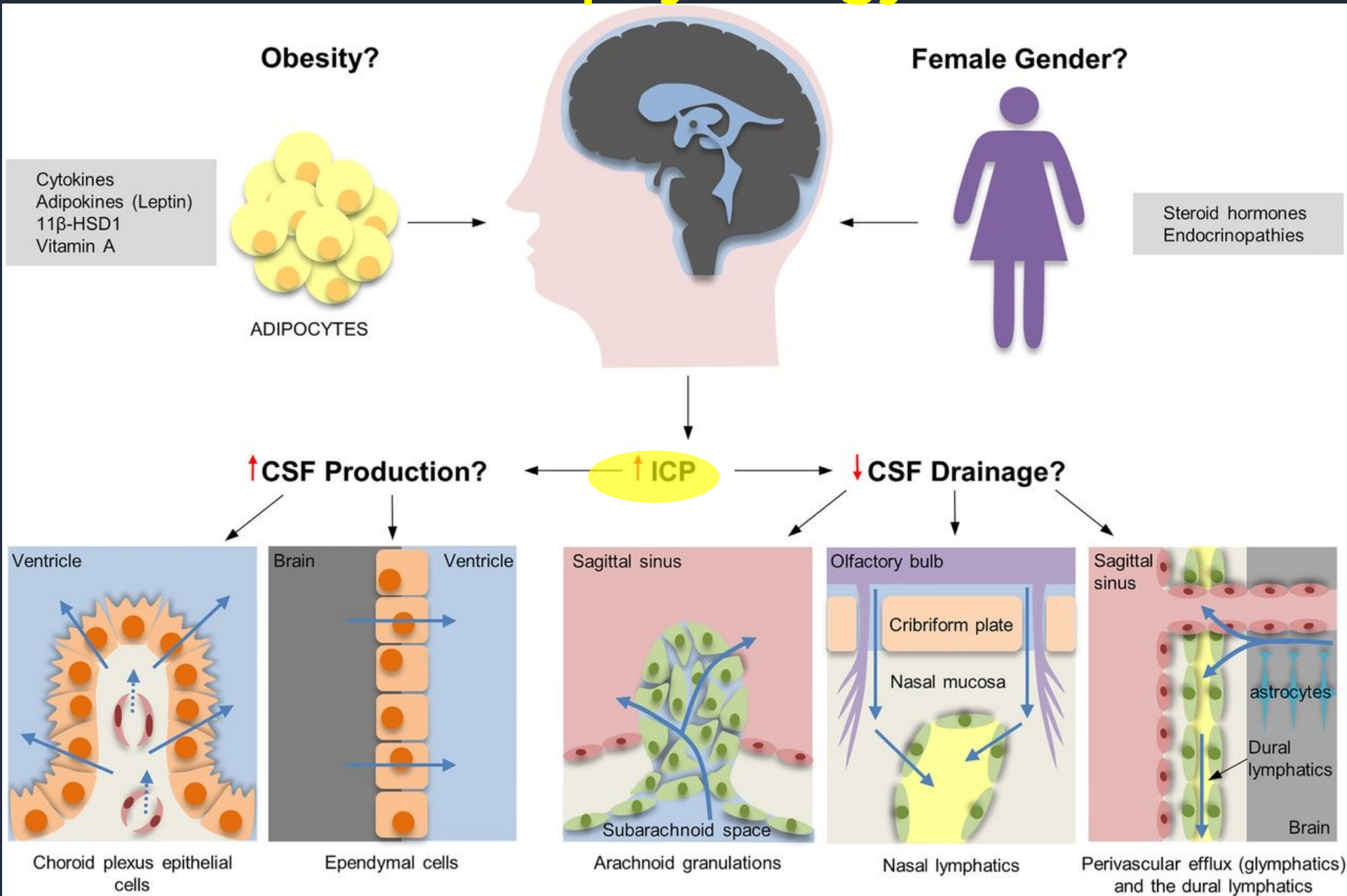
- Secondary
 - Mass
 - DVST
 - Edema
 - Infection
 - Hydrocephalus....
- Idiopathic IIH

Pseudotumor Cerebri. → IIH Diagnosis

Table 2 Criteria for diagnosing idiopathic intracranial hypertension

1. If symptoms present, they may only reflect those of generalized intracranial hypertension or papilledema.
2. If signs present, they may only reflect those of generalized intracranial hypertension or papilledema.
3. Documented elevated intracranial pressure measured in the lateral decubitus position.
4. Normal CSF composition.
5. No evidence of hydrocephalus, mass, structural, or vascular lesion on MRI or contrast-enhanced CT for typical patients, and MRI and MR venography for all others.
6. No other cause of intracranial hypertension identified.

Pathophysiology



↑CP activity1
Vit A (Aquaporin 1
rec
Mineralocorticoid
rec

Mollan et al J
Neurol Neurosurg
Psychiatry 2015

Role of Obesity

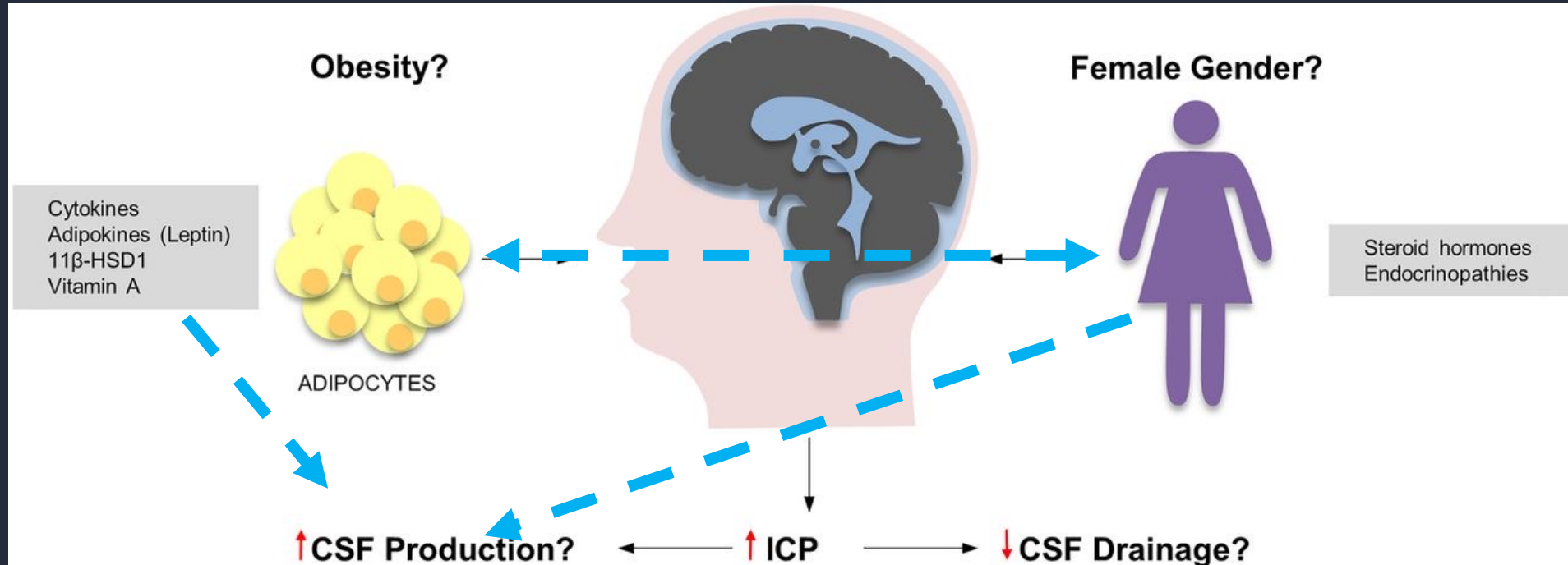
- Consistent risk factor for development of IIH
- Correlation: BMI & risk of IIH
- Association:
 - ↑ weight & disease recurrence
 - induce weight loss → improvements in ICP, papilledema, VF, headache

Daniels AB. Am J Ophthalmol 2006

Ko MW *et al.* . Weight gain and recurrence in Idiopathic Intracranial Hypertension: a case-control study. Neurology 2011

3Sinclair AJ, *et al.*. BMJ 2010

Obesity: Cause Vs Effect ?



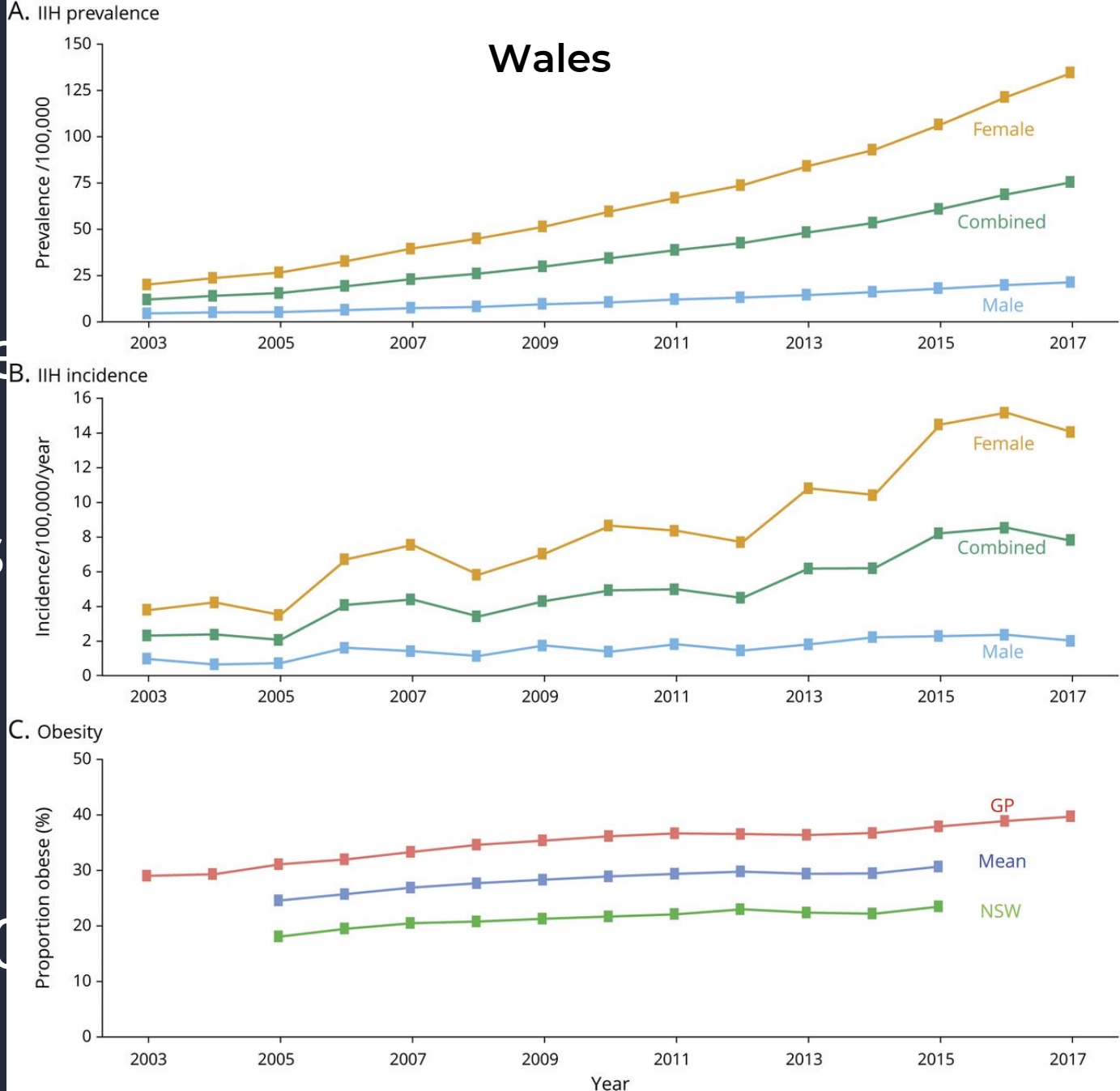
- \uparrow MC (mineralocorticoid) \rightarrow \uparrow CP \rightarrow \uparrow CSF production*
 - Obesity
 - Hyperaldosteronism
 - Hypercortisolism
 - Hypervitaminosis A
 -

? No Hydrocephalus

*Salpietro V et al Endocrinol Lett 2012

*Andrews et al Jormone Research in Pediatrics 2014

- Incidence
- Statistics
- 2X
- 3X
- In 210



~ Obesity

➔ Record

IIH: Socioeconomic Burden, USA

- 320% ↑ in new CSF shunt procedures 1998-2002
- Total economic costs ~ \$444 million/ year
- 57% of patients report significant lost earnings
- 31% changing occupation
- ↑ depression & ↓ levels of quality of life

1. Curry et al. Neurosurgery 2005

2. Friesner D et al. Obes Rev 2011

3. Kleinschmidt, J et al. Idiopathic intracranial hypertension: Relationship to depression, anxiety, and quality of life. Neurology 2000

IIH: Socioeconomic Burden, UK

- Jan 2002 – Dec 2016
- IIH incidence is rising (>>100%) highest in areas of social deprivation and mirroring obesity trends.
- Admission rates rose by 442%

IIH In MEN

- IIH typically occurs in young, obese women
- Rare in men
- 721 patients
 - 66 men (9%)
 - 655 women (91%).

Idiopathic intracranial hypertension in men

[B.B. Bruce, MD](#), [S. Kedar, MD](#), [G.P. Van Stavern, MD](#), [D. Monaghan, BS](#), [M.D. Acierno, MD](#), [R.A. Braswell, MD](#), [P. Preechawat, MD](#), [J.J. Corbett, MD](#), [N.J. Newman, MD](#), and [Y. Bioussé, MD](#)

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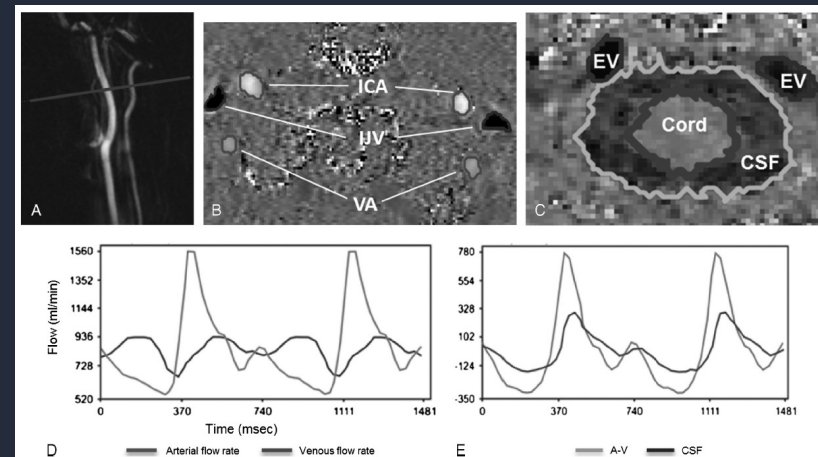
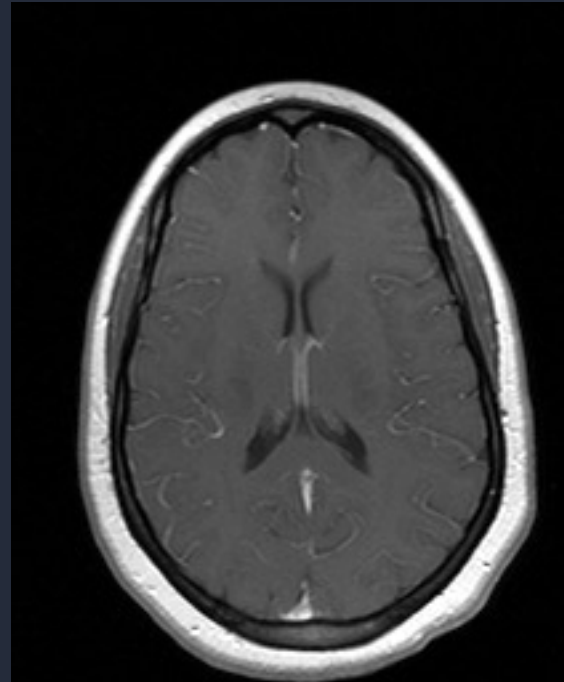
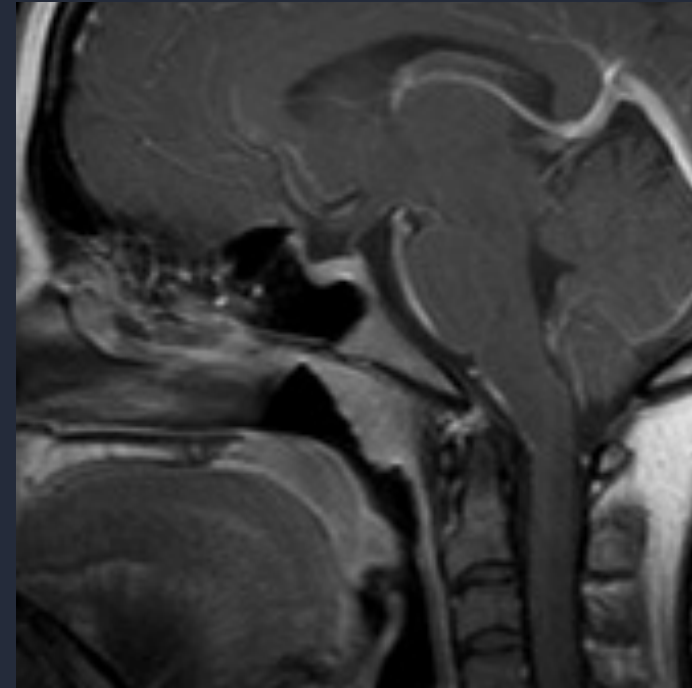
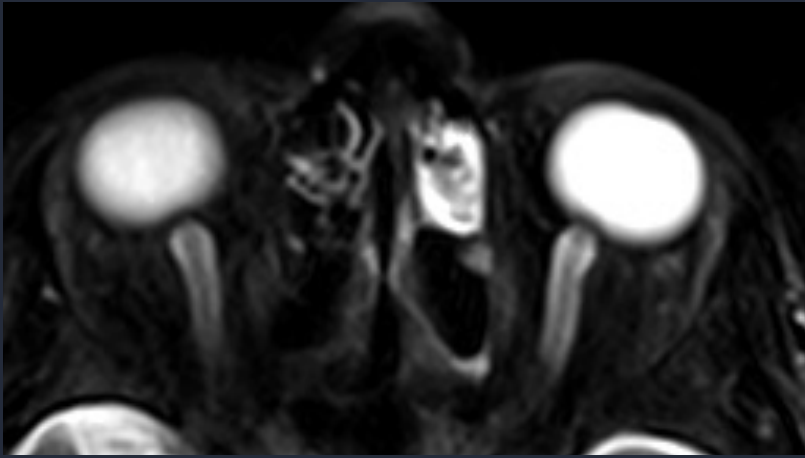
IIH. MEN VS WOMEN

- Men were more likely to have **sleep apnea**
- Were **older** (37 vs 28 years) $p = 0.02$
- As their 1ST symptom of IIH, men were less likely to report headache but more likely to report **visual disturbances**
- Men continued to have less headache
- Visual acuity and visual fields at presentation and last follow-up were significantly worse among men.

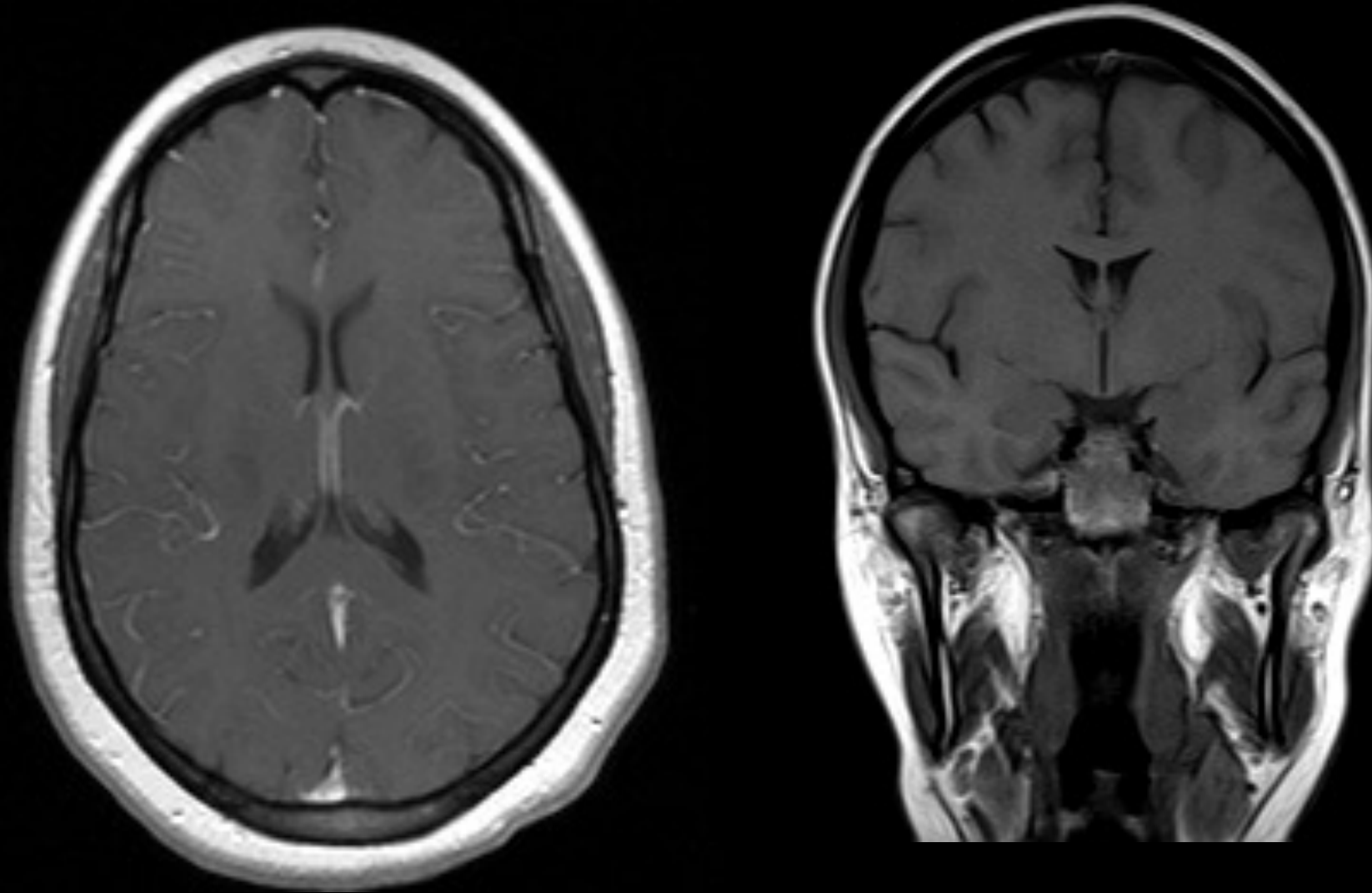
IIH: Treatment

- Acetazolamide: ↓ CFS production
- Weight Loss
- ON Fenestration
- CSF Shunting : ↓ CSF volume
- TS stenting: ↓ Venous pressure

IIH: Radiology

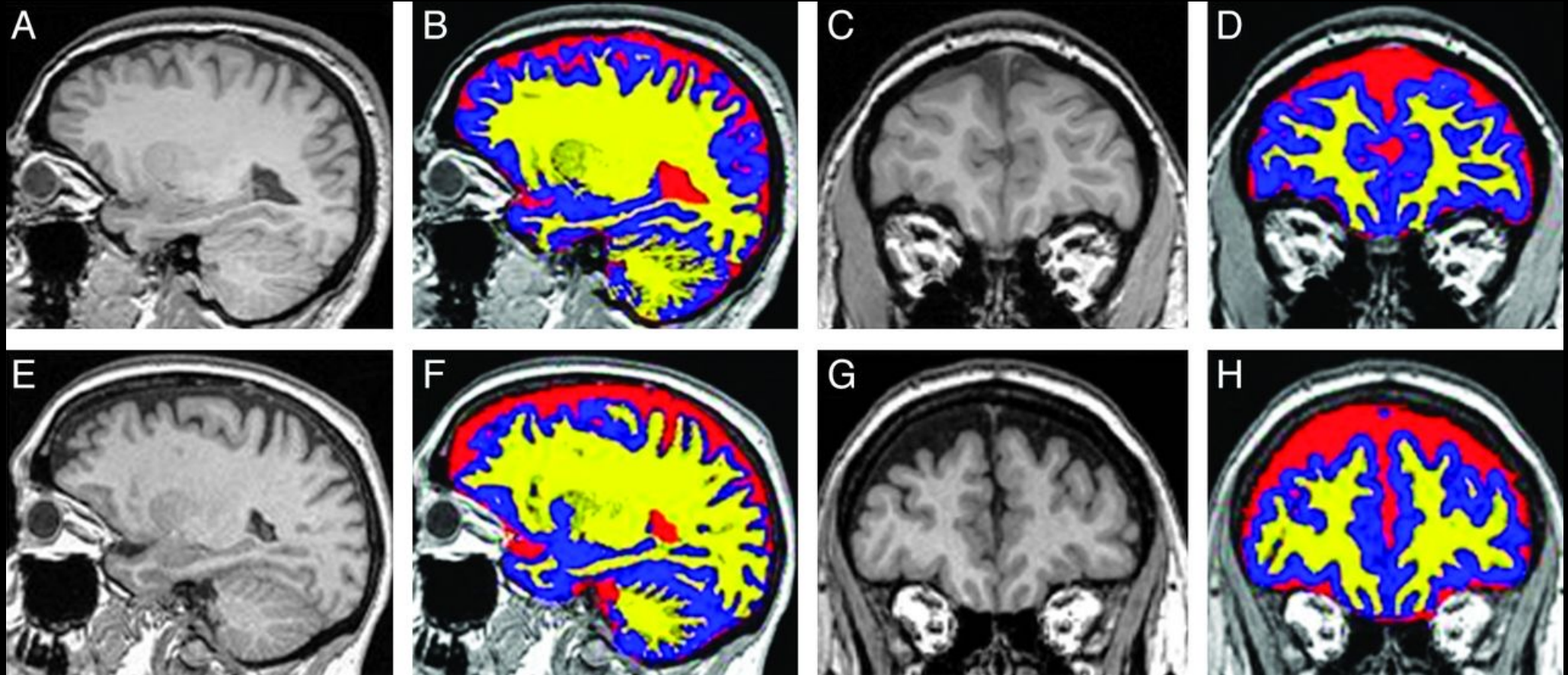


Ventricles & SAS



?? ↑ CSF production / ↓ CSF absorption ??

Ventricles & SAS



Segmentation:

Parenchyma

CSF

-GM

-ICCSF

-WM

-IVCSF

Ventricles & SAS

Table 1: Mean and SD values of cranial volumetric measures

	ICCSF (mL)	VCSF (mL)	EVCSF (mL)
Control (<i>n</i> = 11)		19 ± 6	
IIH (<i>n</i> = 11)		19 ± 7	
<i>P</i> value		0.950	

Nor ICCSF (%)	Nor VCSF (%)	Nor EVCSF (%)
18 ± 2	1.4 ± 0.5	16 ± 1
21 ± 3	1.3 ± 0.4	20 ± 2
0.0014	0.511	0.0007

CONCLUSIONS: Increased IC CSF volume that accumulates in the EV SAS provides direct evidence for impaired CSF homeostasis in obesity-associated IIH

Ventricles & SAS

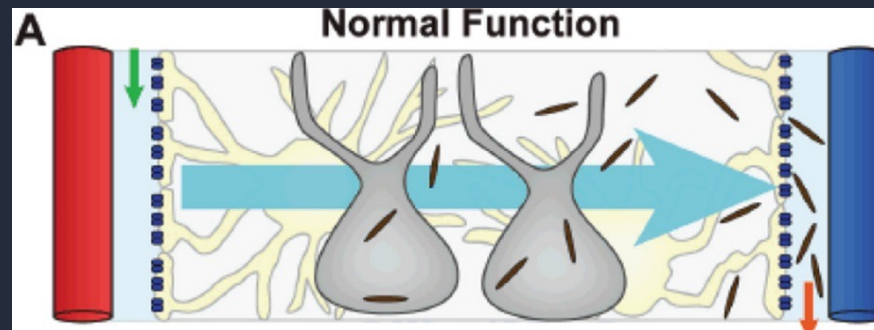
Table 1: Mean and SD values of cranial volumetric measurements

	ICCSF (mL)	VCSF (mL)	EVCSF (mL)	GM (mL)	WM (mL)	ICV (mL)
Control (<i>n</i> = 11)	238 ± 25	19 ± 6	220 ± 24	557 ± 31	537 ± 47	1332 ± 81
IIH (<i>n</i> = 11)	309 ± 56	19 ± 7	290 ± 52	602 ± 57	528 ± 35	1438 ± 124
<i>P</i> value	0.002	0.950	0.0011	0.038	0.616	0.029

Note:—EVCSF indicates extraventricular CSF volume; ICCSF, intracranial CSF volume; Nor, normalized; VCSF, ventricular

CONCLUSIONS: Increased GM Volume ~ Interstitial fluid
 -Likely secondary to increased resistance to cerebral venous drainage

Glymphatic ?



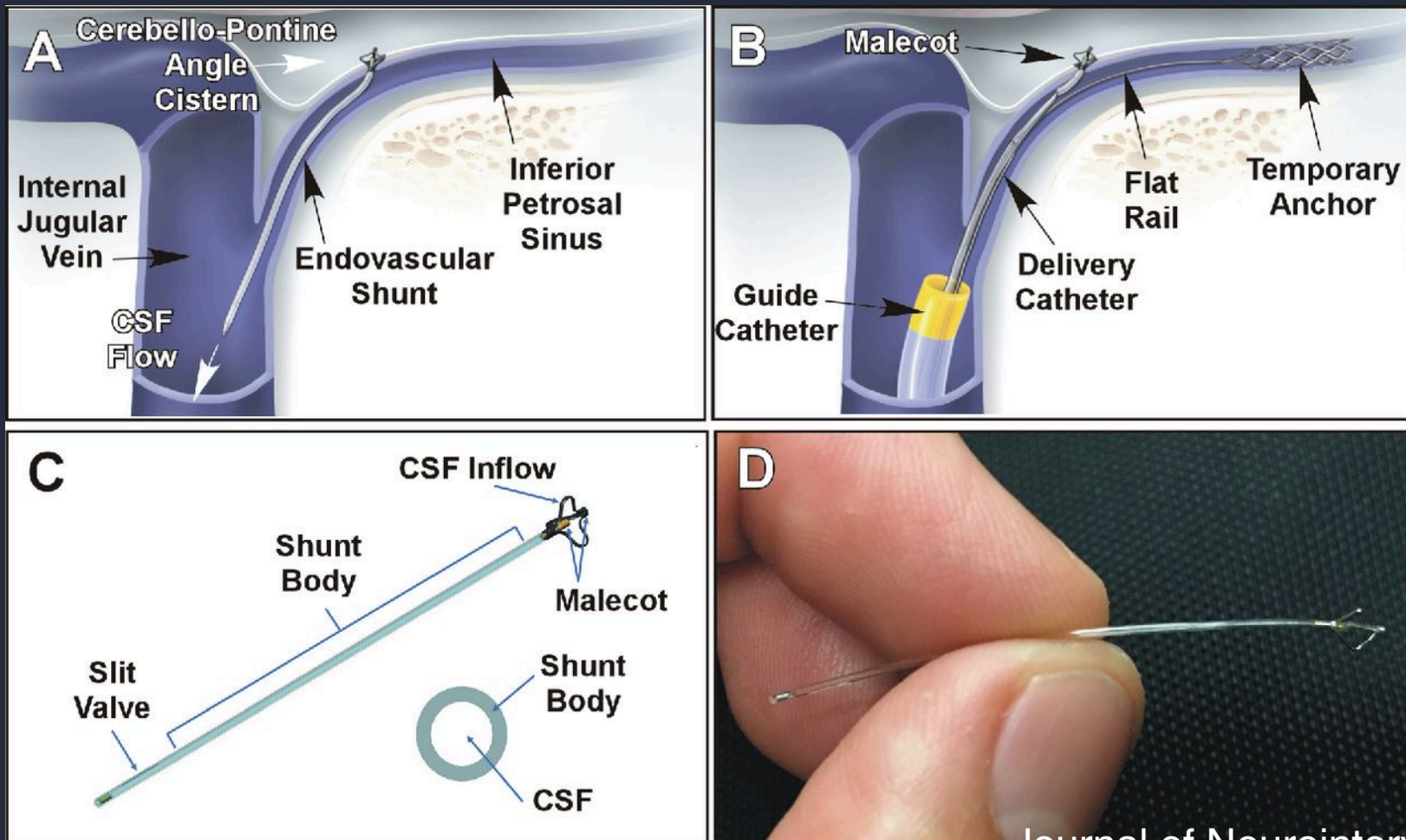
IIH: Treatment

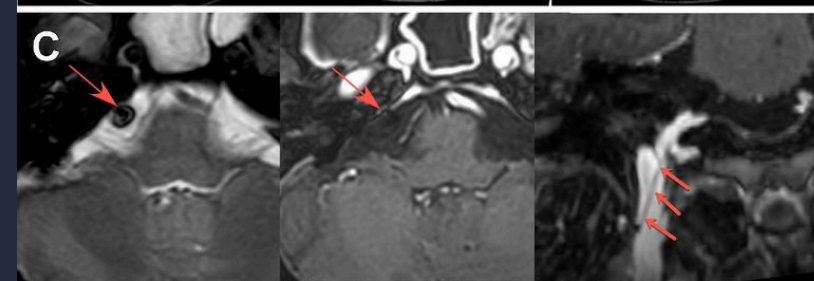
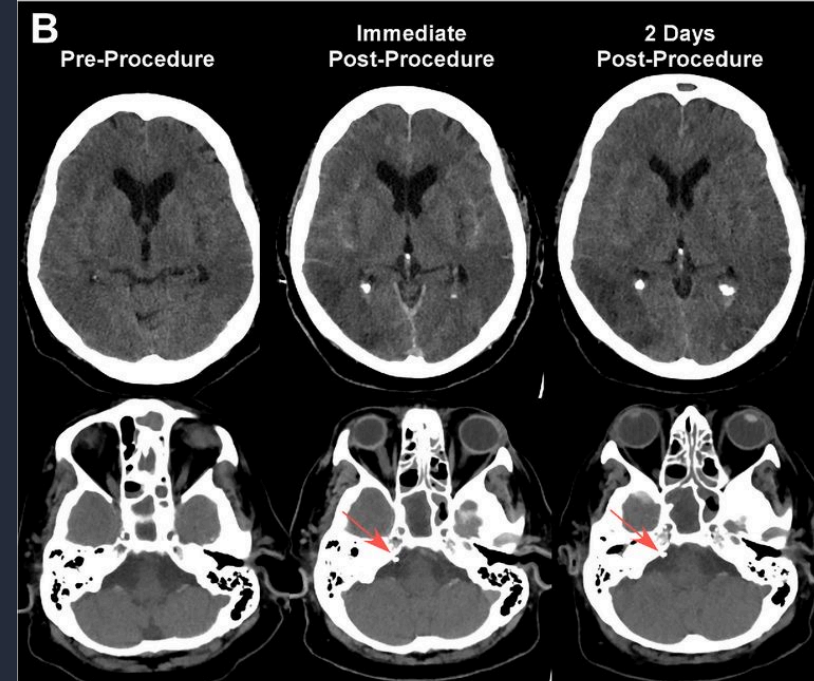
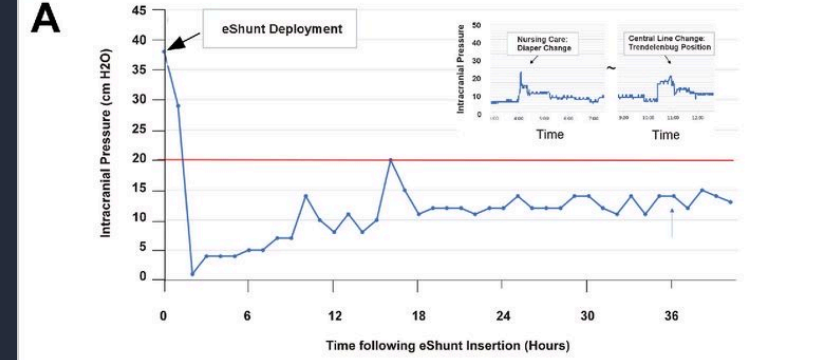
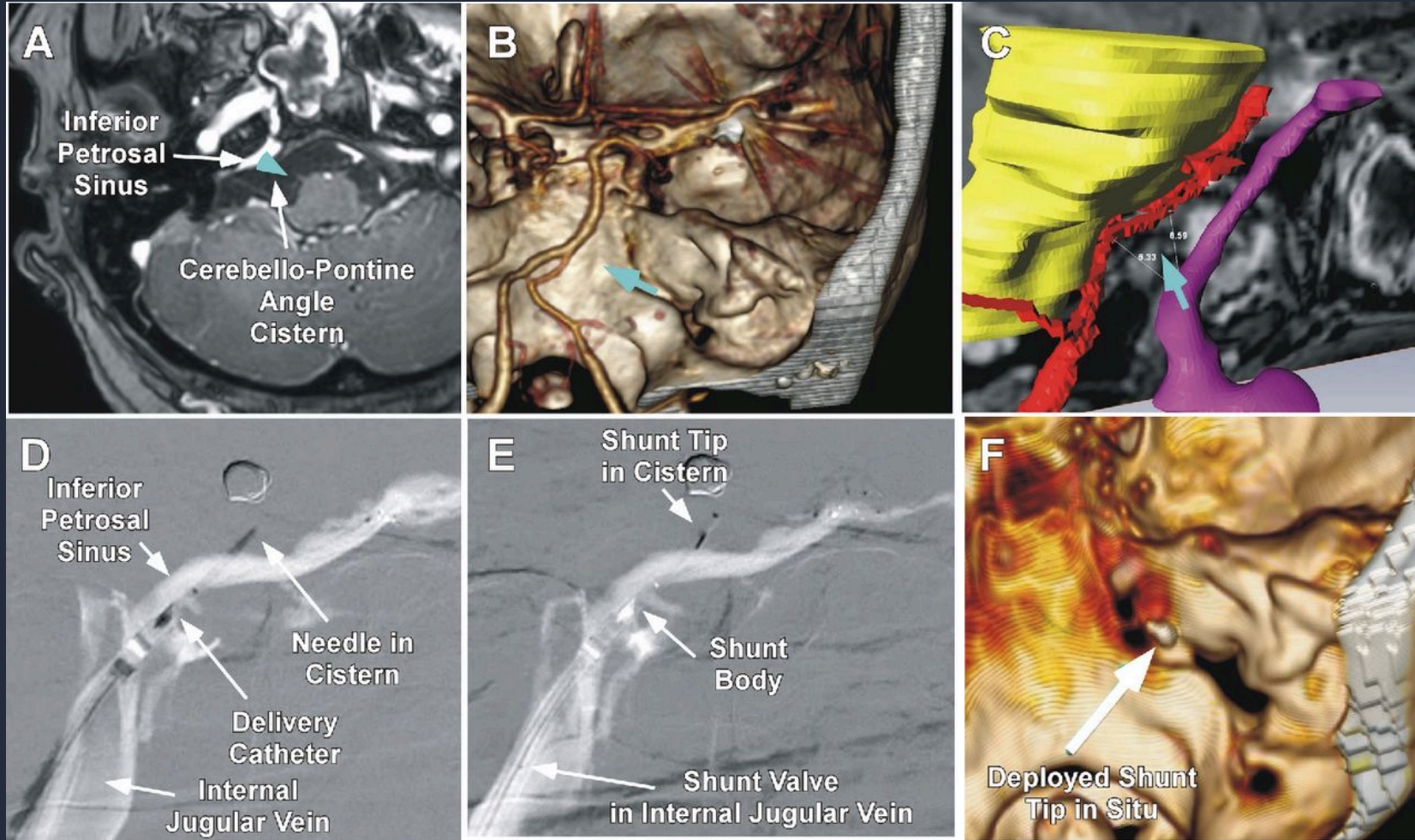
- Acetazolamide: ↓ CSF production
- Weight Loss
- ON Fenestration
- CSF Shunting : ↓ CSF volume
- TS stenting: ↓ Venous pressure

First-in-human endovascular treatment of hydrocephalus with a miniature biomimetic transdural shunt

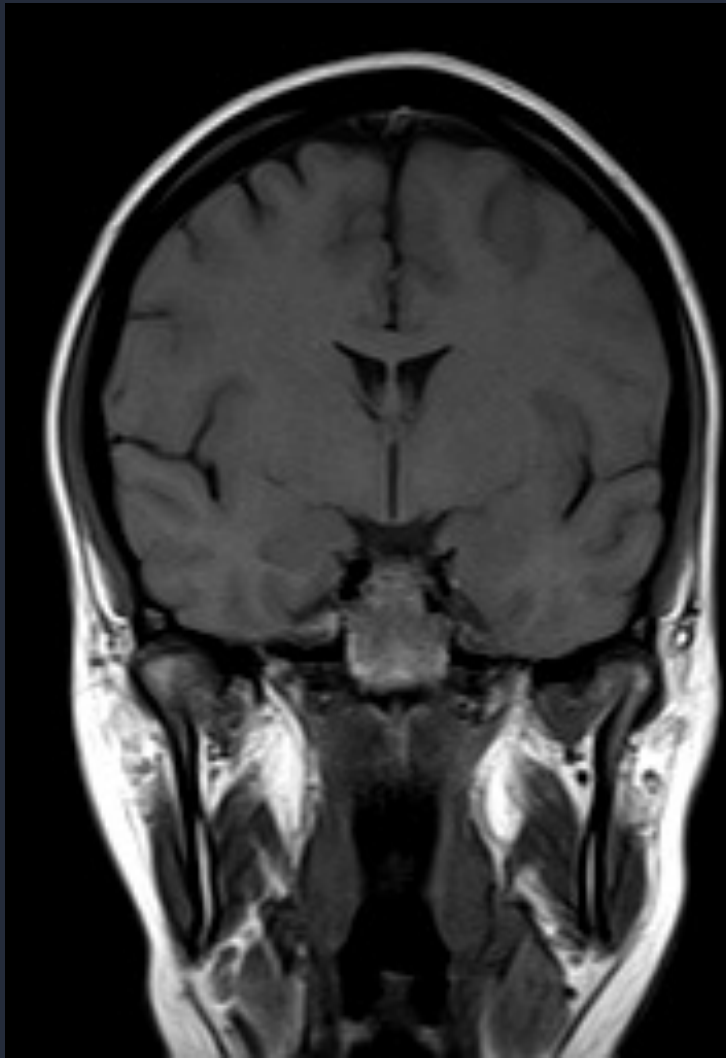
Pedro Lylyk¹, Ivan Lylyk², Carlos Bleise¹, Esteban Scrivano¹, Pedro Nicolas Lylyk¹, Brandon Beneduce³, Carl B Heilman⁴, Adel M Malek⁴

Correspondence to Dr Adel M Malek, Neurosurgery, Tufts Medical Center, Boston, MA 02111, USA;



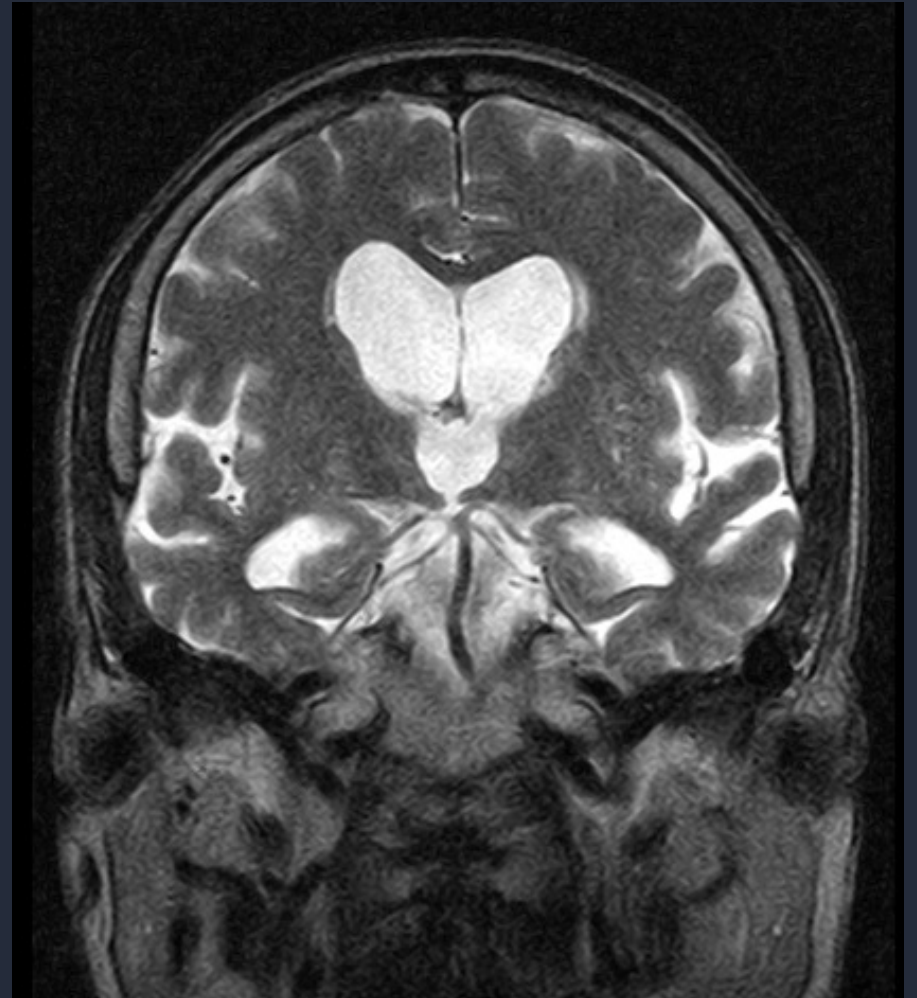


ICP and VENTRICULAR SIZE



normal to small vent
Inc ICP

VS



Large Vent
Normal ICP

Optic Nerve Sheath Complex

Findings seen in IIH, SIH and in Astronauts:

1. Tortuosity/Enlargement ONS 45%
2. Flattening of the posterior sclera 80%
3. Inward protrusion of the ON head

2 & 3 have Sens & Spec ranging between 80-100% *

*Agid et al . Neuroradiology 2006
Rohr et al. AJNR 2011

SPACEFLIGHT-ASSOCIATED NEURO-OCULAR SYNDROME (SANS)

The image is a screenshot of a NASA website article. At the top, the NASA logo is on the left, and navigation links for Topics, Missions, Galleries, NASA TV, Follow NASA, Downloads, About, and NASA Audiences are in the center. A search bar is on the right. Below the navigation is a 'Latest' sidebar with three article teasers: 'NASA Invites Media to Briefing on Research Launching to Space Station in 11 hours', 'Experiments Riding 24th SpaceX Cargo Mission to Space Station Study Bioprinting, Crystallization, Laundry a day ago', and 'Space Station Science Highlights: Week of November 29, 2021 5 days ago'. The main article is titled 'International Space Station Research Keeps an Eye on Vision Changes in Space' and is dated 'Aug 3, 2020'. It features a large photo of Commander Anton Shkaplerov using a microscope. Social media icons for Facebook, Twitter, LinkedIn, Pinterest, and a plus sign are at the bottom right.

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NASA Invites Media to Briefing on Research Launching to Space Station *in 11 hours*

Experiments Riding 24th SpaceX Cargo Mission to Space Station Study Bioprinting, Crystallization, Laundry *a day ago*

Space Station Science Highlights: Week of November 29, 2021 *5 days ago*

Commander Anton Shkaplerov works on a space physics experiment *7 days ago*

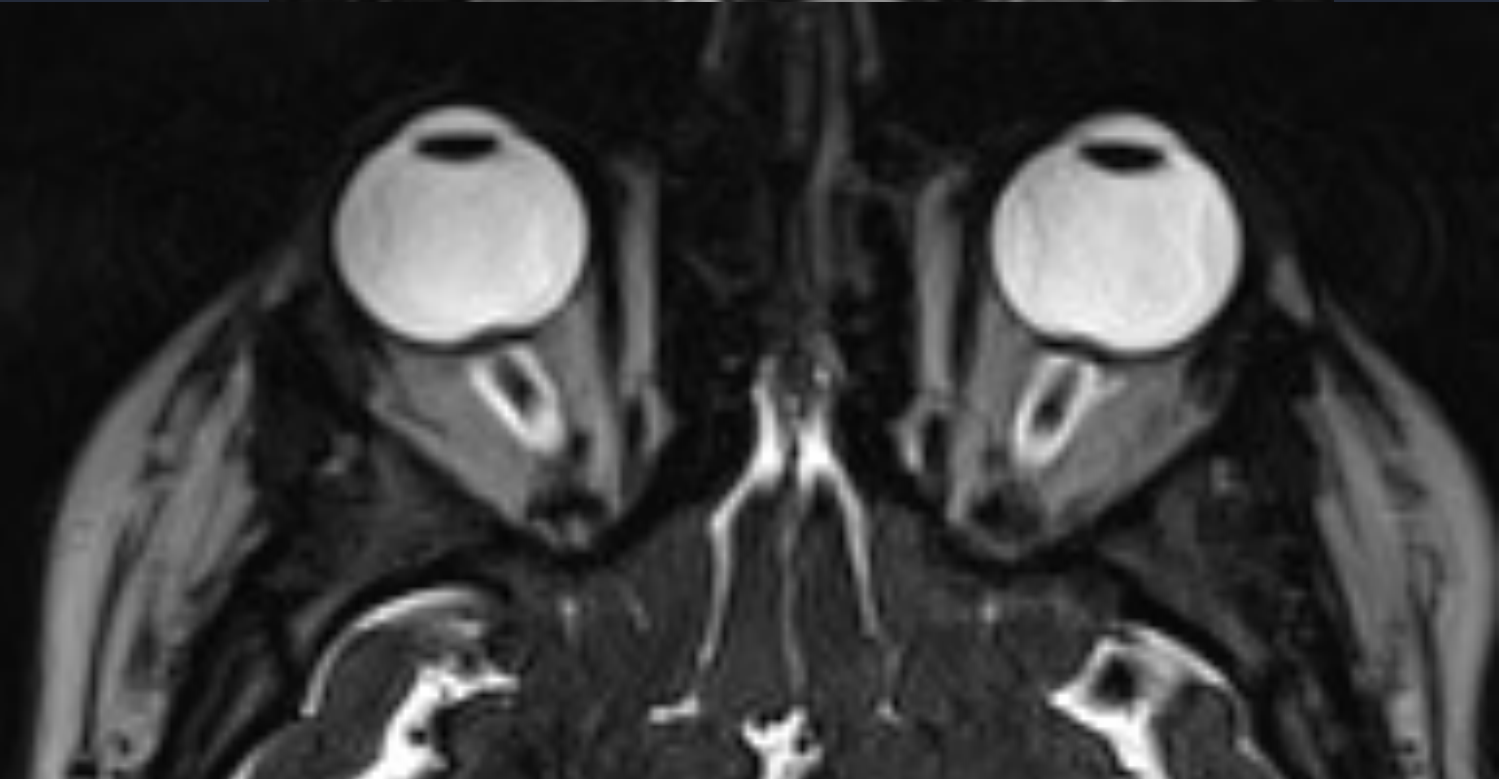
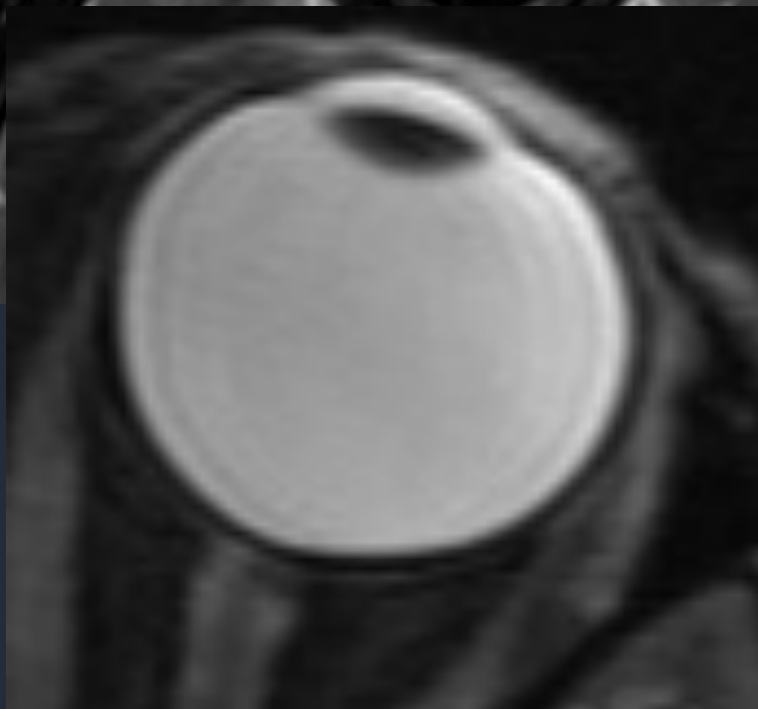
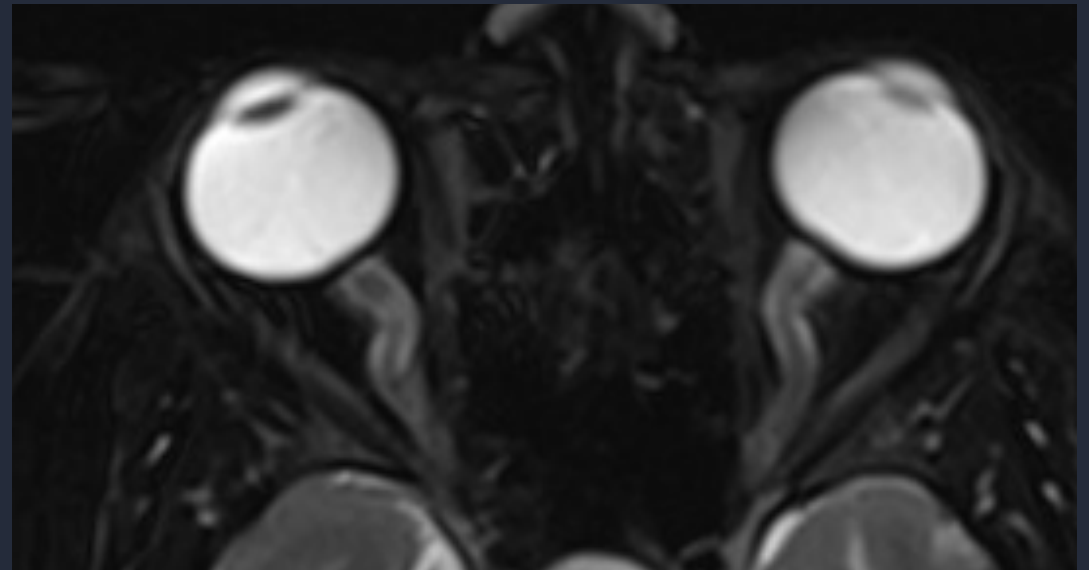
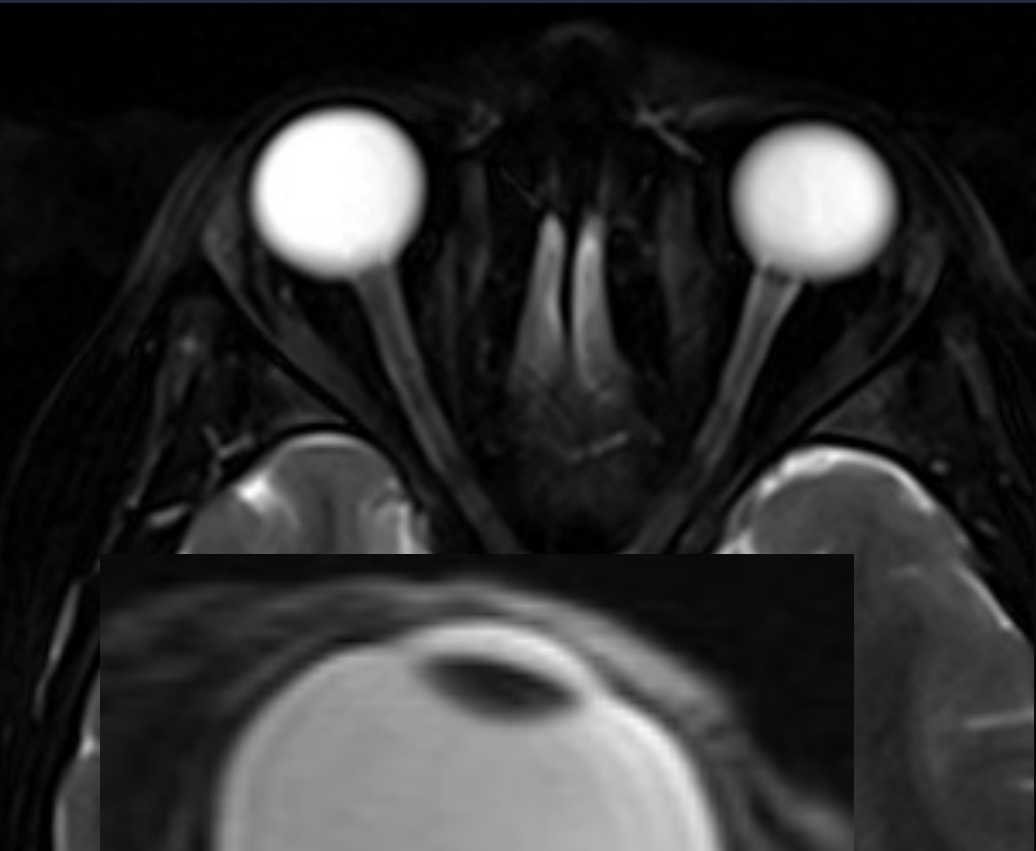
Space Station Research

Aug 3, 2020

International Space Station Research Keeps an Eye on Vision Changes in Space

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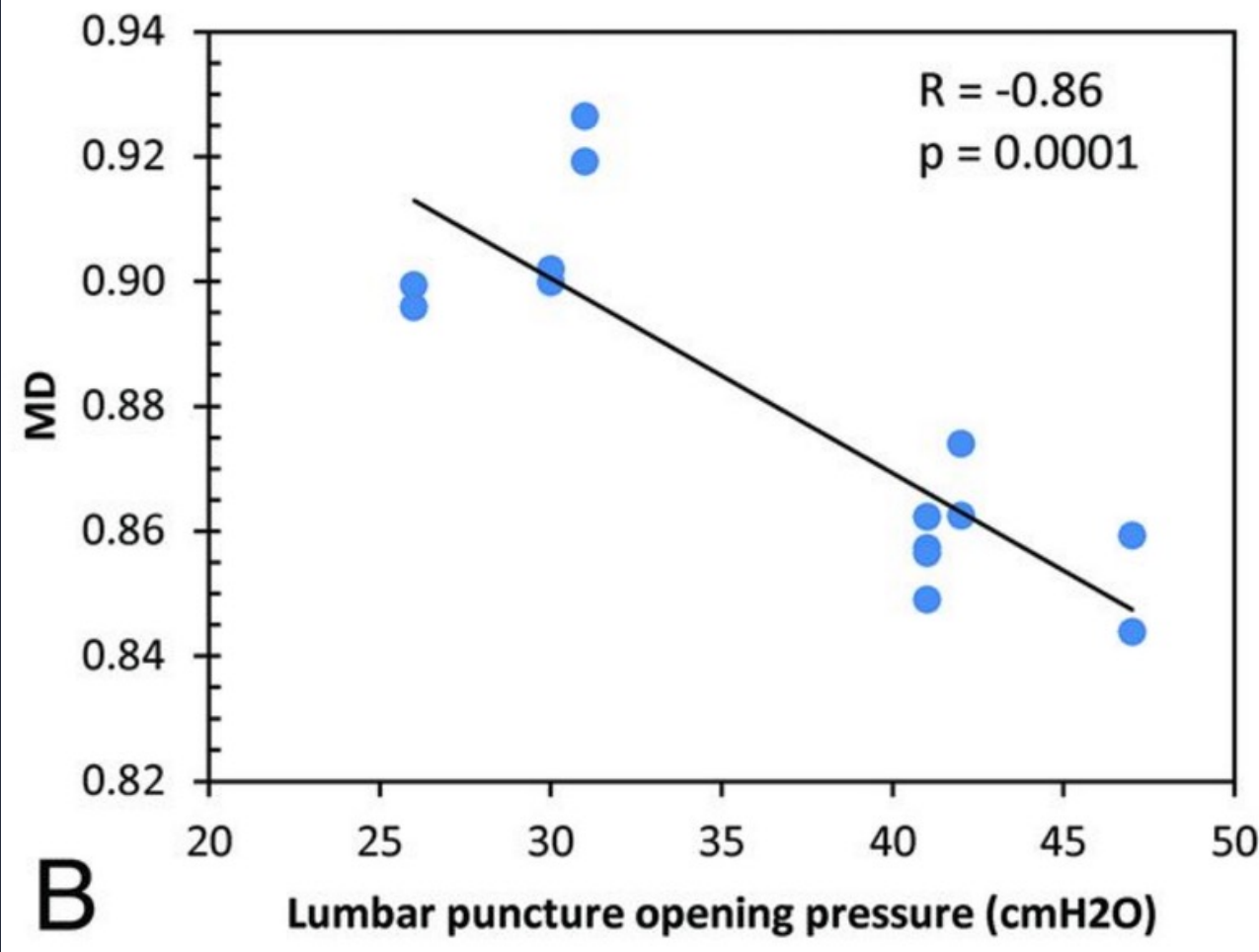
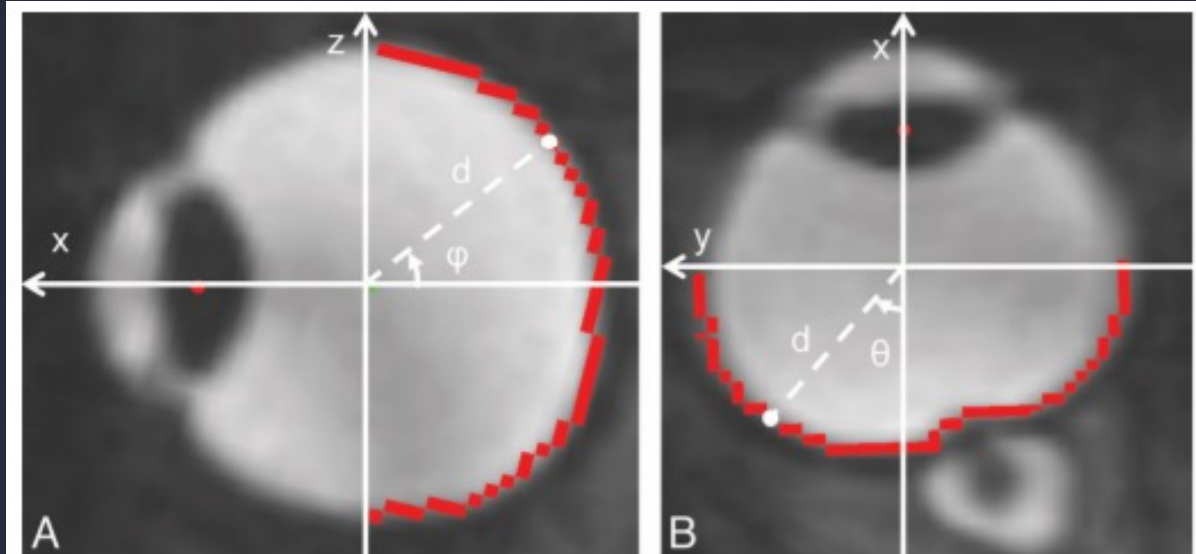
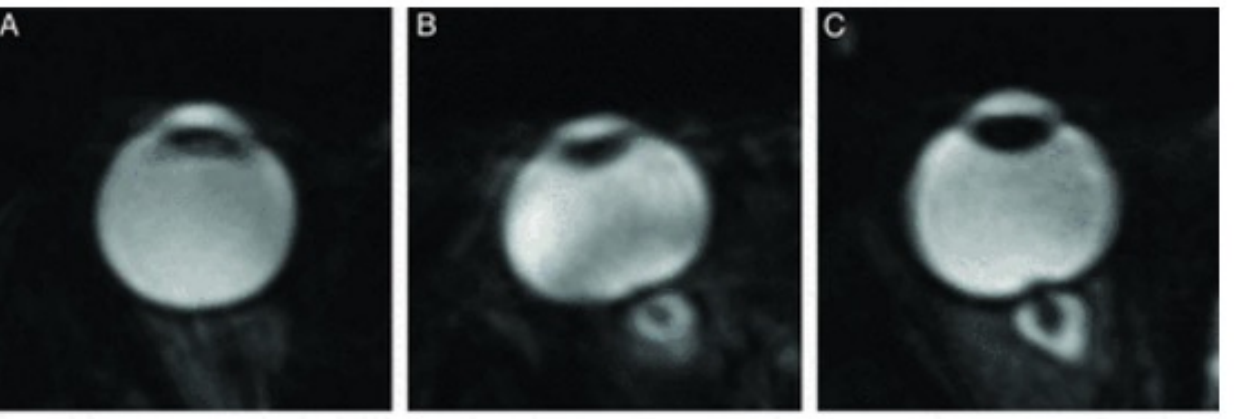
Normal ONS



Automated Quantitation of the Posterior Scleral Flattening and Optic Nerve Protrusion by MRI in Idiopathic Intracranial Hypertension

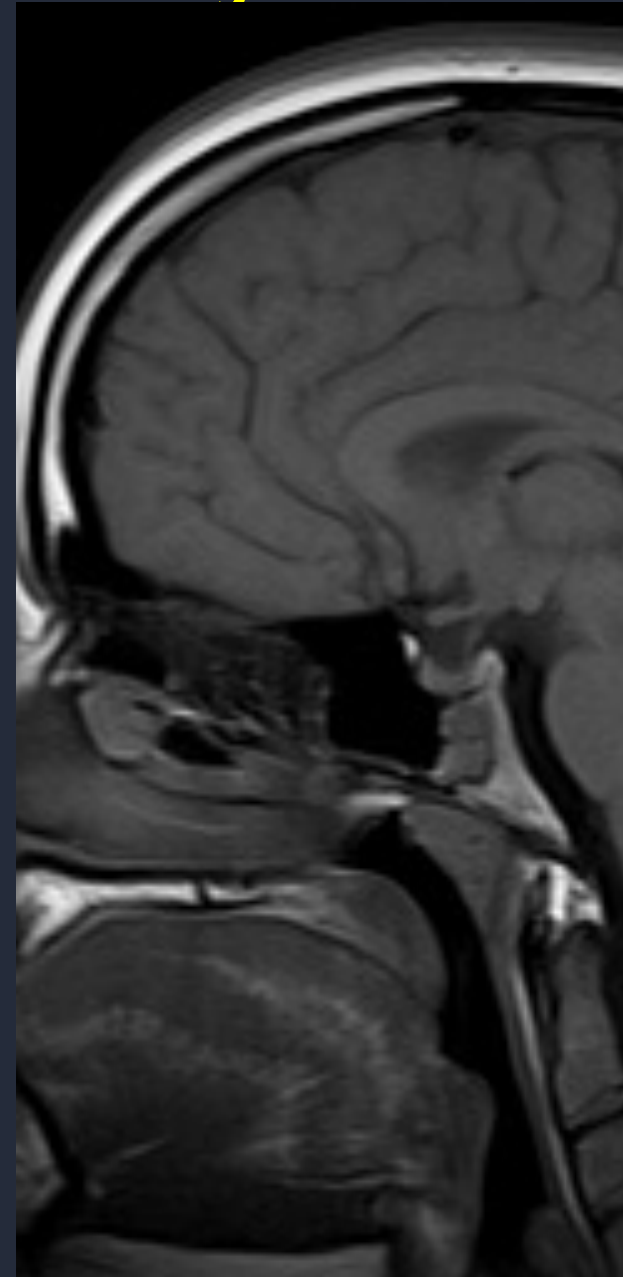
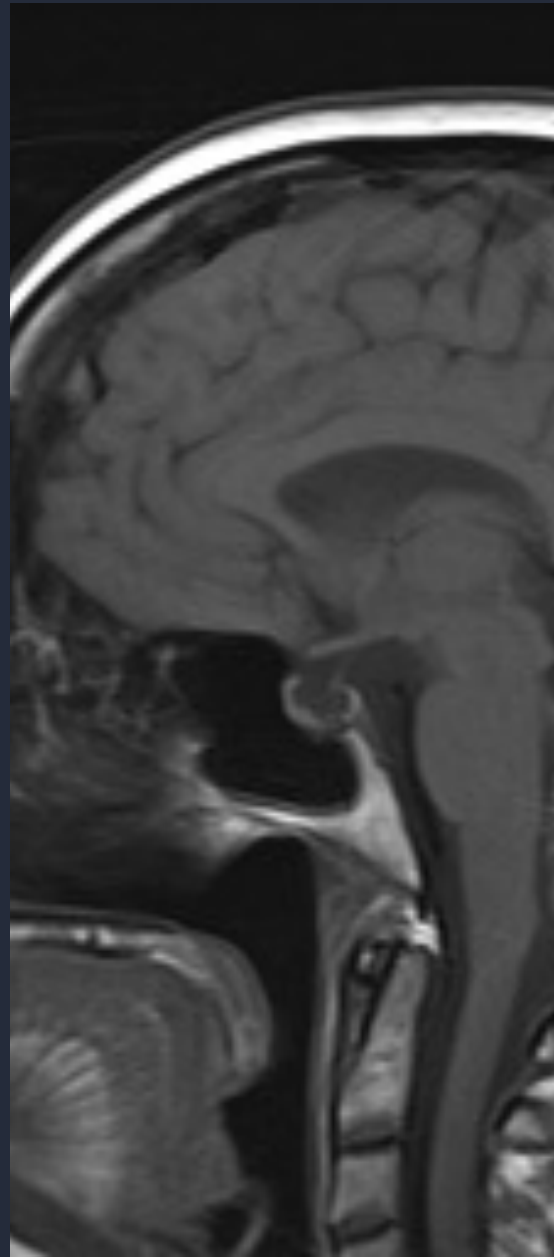
AJNR 2013

N. Alperin, A.M. Bagci, B.L. Lam, and E. Sklar

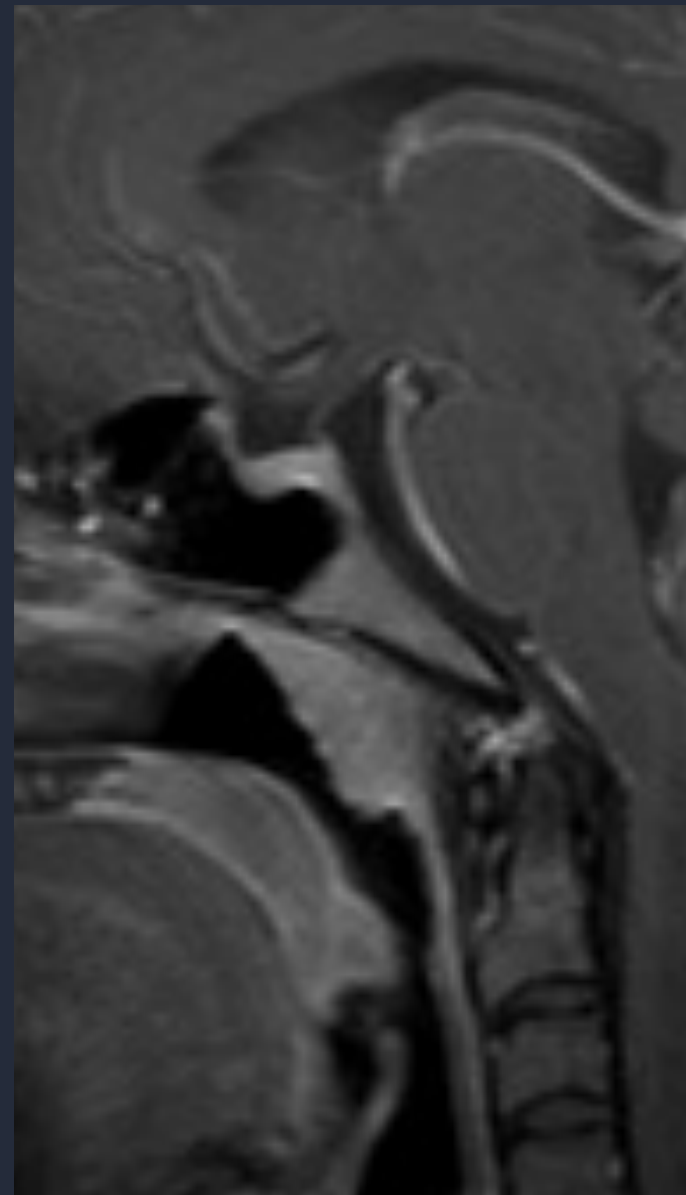
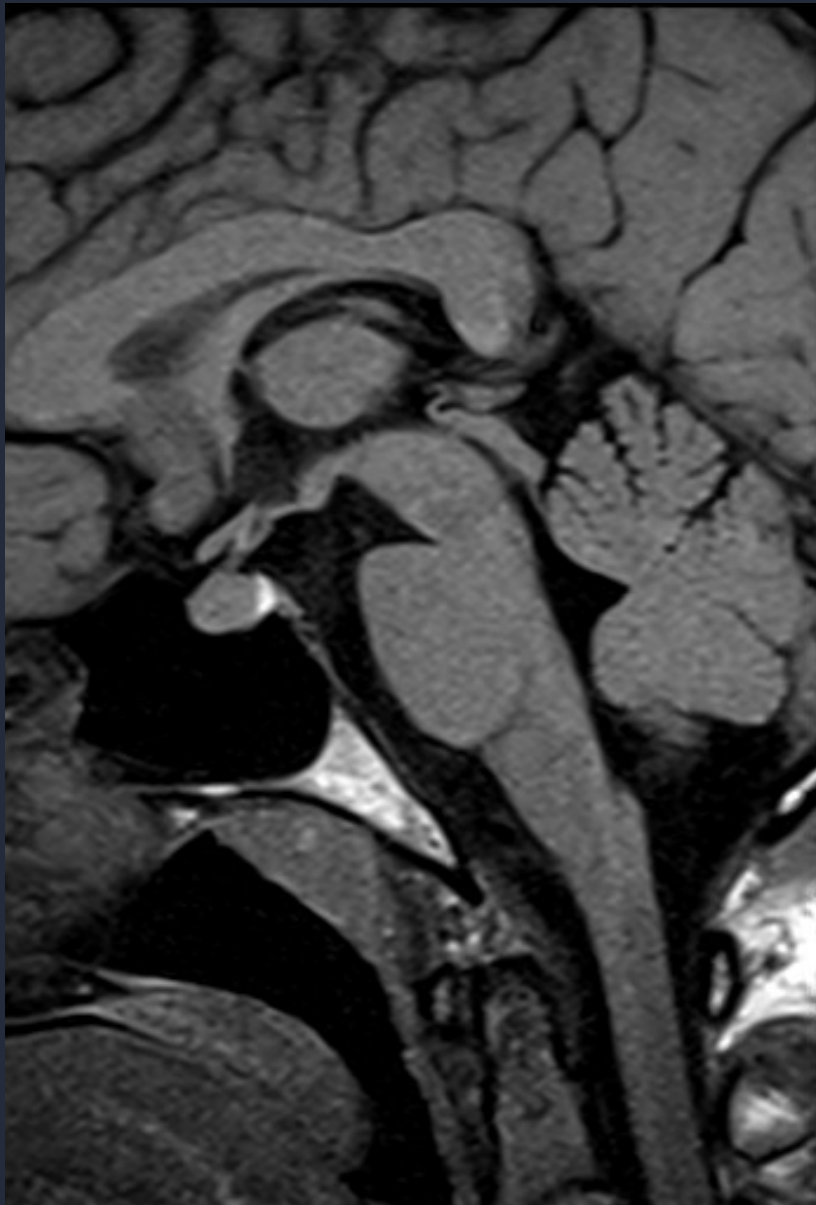


MD: depicting the combined deformation due to the flattening and the nerve protrusion

Sella & Pituitary



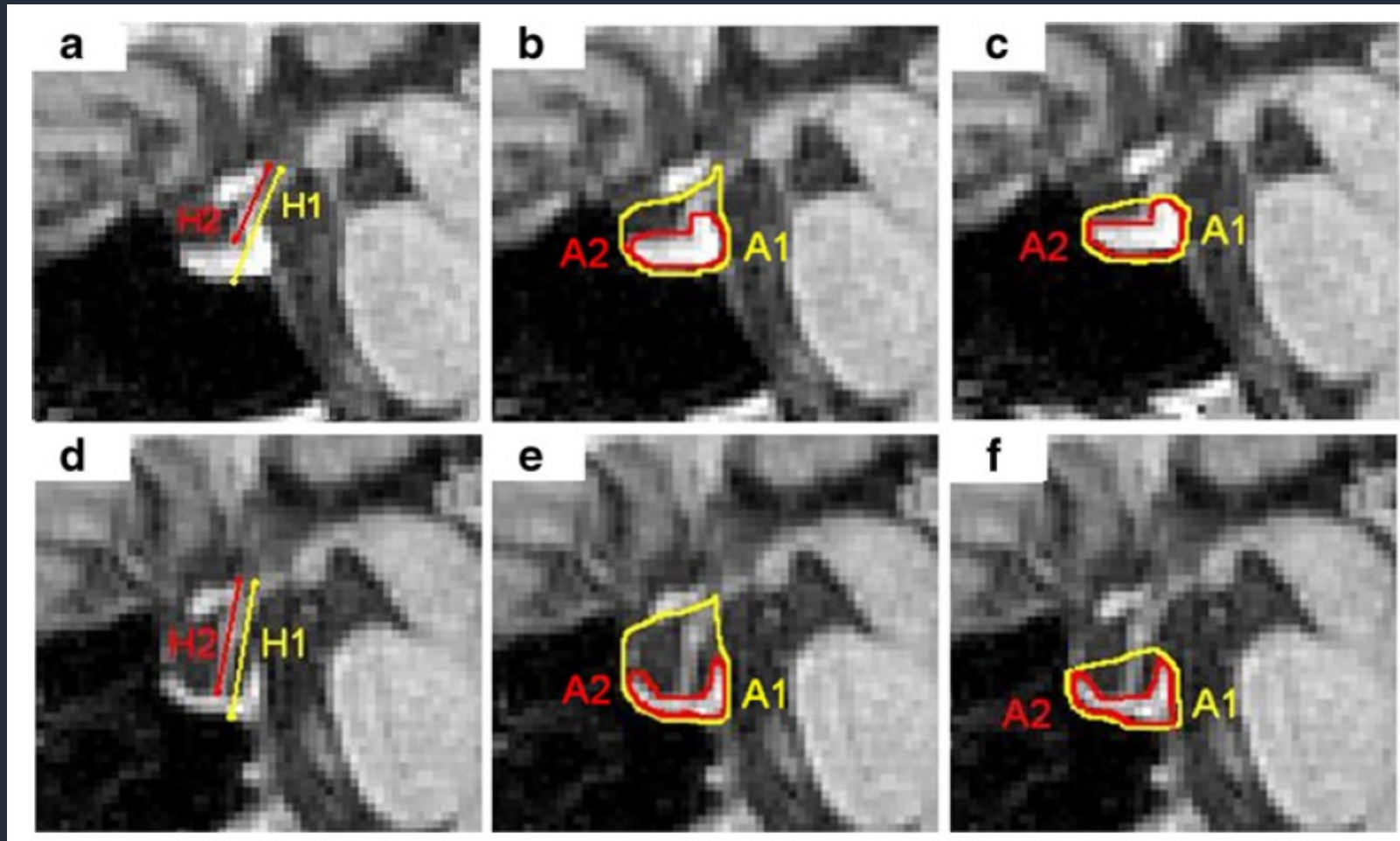
Sella & Pituitary



Magnetic resonance imaging finding of empty sella in obesity related idiopathic intracranial hypertension is associated with enlarged sella turcica

15 May 2013

Sudarshan Ranganathan • Sang H. Lee • Adam Checkver • Evelyn Sklar • Byron L. Lam Gary H. Danton • Noam Alperin

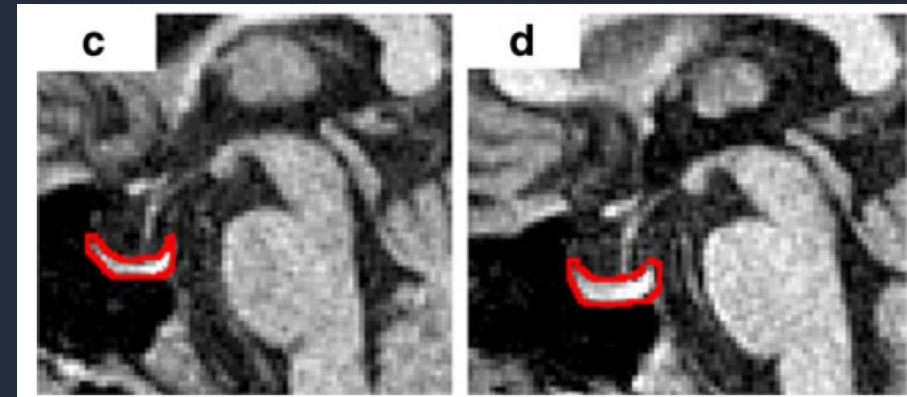
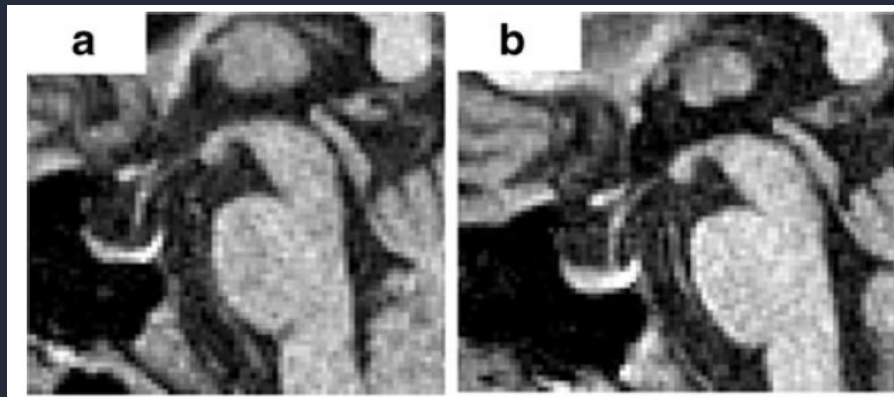


Sella and Pituitary

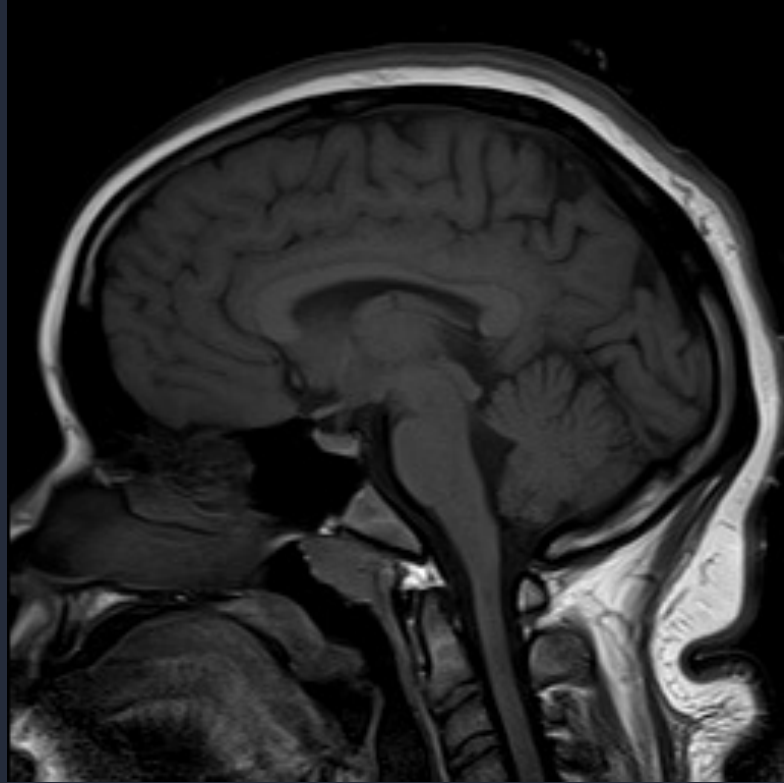
Measurement	Controls (<i>n</i> =11) (mean±SD)	IIH patients (<i>n</i> =10) (mean±SD)	<i>p</i> value
H1 (mm)	14.83±2.44	16.27±2.69	0.173
H2 (mm)	9.94±1.82	12.46±2.11	0.009 *
H1-H2 (mm)	4.89±1.36	3.81±1.30	0.067
(H1-H2)/H1	0.33±0.06	0.23±0.06	0.004 *
M1_A1 (mm ²)	124.15±24.63	200.39±24.18	<0.0001 *
M2_A1 (mm ²)	71.97±22.25	116.09±25.01	0.001 *
A2 (mm ²)	57.20±14.45	62.02±16.74	0.557
A2/M1_A1	0.47±0.11	0.31 ±0.07	0.003 *
A2/M2_A1	0.82±0.15	0.54±0.11	0.0004 *

Sella and Pituitary: Post treatment

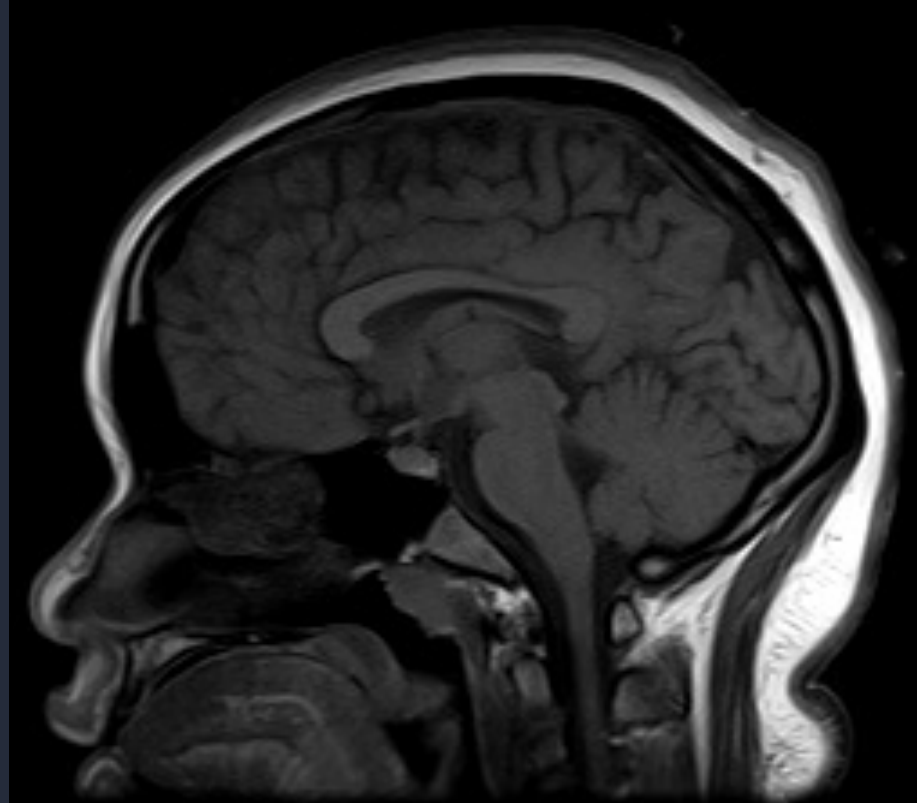
IIH patient (n=8)	Sella area (A1) (mm ²)		Gland area (A2) (mm ²)		Normalized gland area (A2/A1)	
	Pre	Post-1 week	Pre	Post-1 week	Pre	Post-1 week
P2	212.0	218.0	56.3	56.0	0.27	0.26
P3	198.0	206.0	52.8	68.5	0.27	0.33
P4	216.0	221.0	48.5	69.0	0.22	0.31
P6	183.0	175.0	49.3	53.0	0.27	0.30
P7	182.0	185.0	64.3	65.0	0.35	0.35
P9	224.0	227.0	58.0	65.0	0.26	0.29
P11	216.9	219.1	82.8	85.7	0.38	0.39
P12	152.0	146.0	41.8	54.5	0.27	0.37
Mean	197.9	199.6	56.7	64.6	0.29	0.33
SD	24.4	28.4	12.6	10.6	0.05	0.05
<i>p</i> value	0.547		0.016*		0.031*	



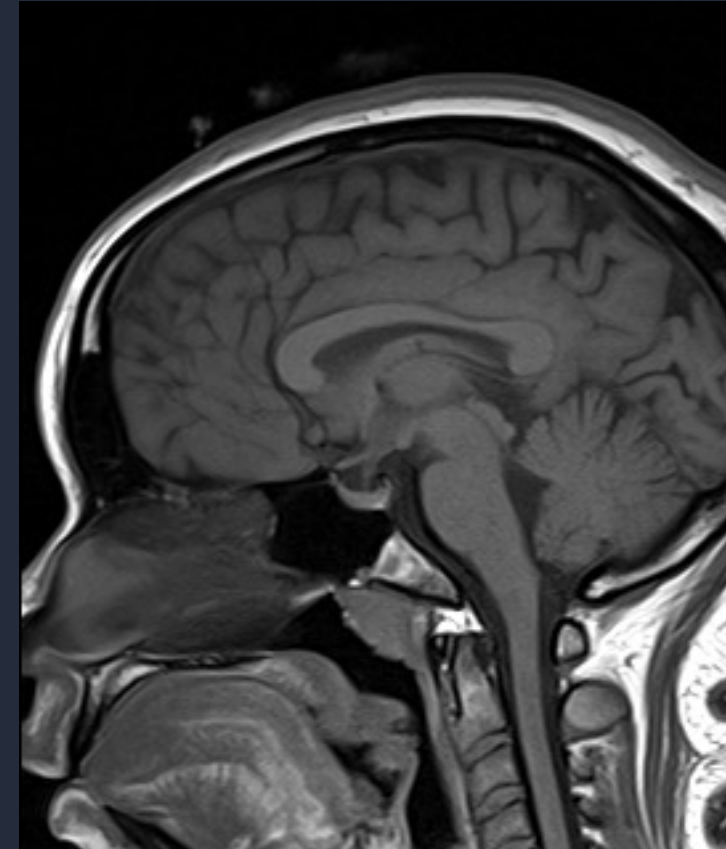
13yo F with decreased vision
ICP 45



Presentation



D2 after large volume
tap



2 years after LP shunt

Venous Sinuses & Venous Drainage

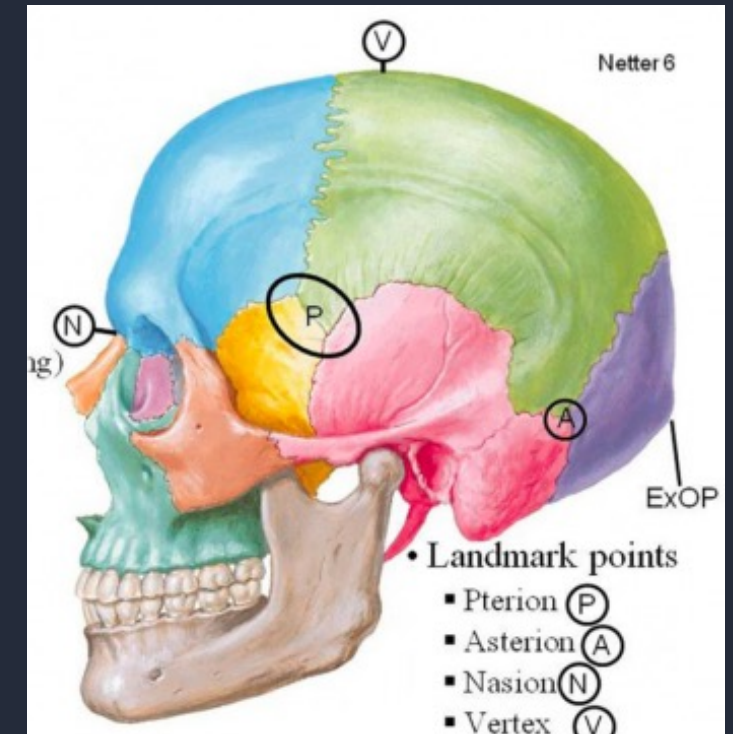


- Why does it happen: Cause &/or Effect?
- Why at this location?
- Correlation with severity?
- Correlation with outcome?
- Stenting?

STENOSIS LOCATION

1. Why is stenosis in TS at a fixed anatomic point?

- Distal TS is collapsible. Autopsy
- Acts as a “dynamic resistor” (Starling resistors)
- Location of TSS is consistent and is above the Asterion



TSS: Cause Vs Effect

- Manometry*:
 - Elevated Venous pressure in SSS
 - Significant pressure gradient across the “stenosis”

Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis

B Riggeal et al Neurology 2013

- Evaluate the effect of TSS on course of IIH. 51 patients

Results

- TSS ~ 100% & almost always bilateral
- Always same location

TSS and Disease course/Outcome

- No correlation between % TSS and
 - Opening Pressure
 - Patient w highest ICP (60 cm):lowest % stenosis ~ 19%
 - Clinical course and outcome
 - VF
 - Clinical Course
 - Having unilateral TSS did not correlate with the clinical course
 - Incidentally: Patients with highest stenosis → good outcome & normal VFs

→ Clinical features, not the degree of TSS, should be used to determine management in IIH

TSS: Cause Vs Effect

Transverse Sinus Stenting for Idiopathic Intracranial Hypertension: A Review of 52 Patients and of Model Predictions

AJNR
2011

INTERVENTIONAL

Transverse Sinus Stenting for Idiopathic Intracranial Hypertension: A Review of 52 Patients and of Model Predictions

AJNR
2014



G. Michael Halmagyi

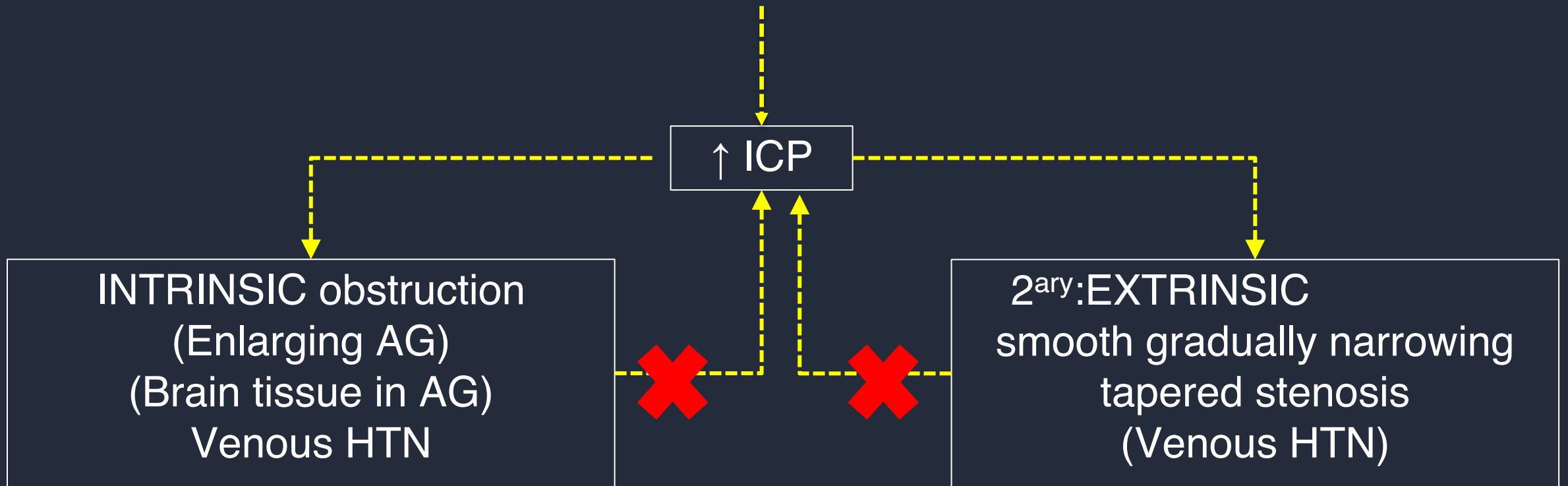


Geoffrey D. Parker

Department of Neurology,
Royal Prince Alfred Hospital,
Missenden Rd, Camperdown,
Sydney, NSW, Australia

TSS: Cause Vs Effect

????



“It became clear that when one is faced with a positive feedback loop or a “vicious cycle,” the loop just needs to be broken and the answer to the question of causation becomes irrelevant—rather like the question of which came first: the chicken or the egg”.

Table 1: Clinical parameters in patients with IIH before and after transverse sinus stent placement

Clinical Parameter	Before Stent			After Stent		
	Mild	Moderate	Severe	Mild	Moderate	Severe
Papilledema	11 ^a	27	7	0 ^b	0	0
Visual acuity loss	4	4	5	3	1	0
Visual field loss	19 ^c	6	5	5	2	0
Headache	6	22	15	0	8 ^d	0
Transient visual obscurations	3	10	6	0	0	0
Pulsatile tinnitus	6	9	2	0	0	0
Diplopia	3	2	1	0	0	0

Table 4: Venous pressures in 40 patients with IIH with papilledema requiring only 1 stent placement procedure versus 6 (all with papilledema at first stent) requiring a second stent placement procedure^a

	Superior Sagittal Sinus Pre-stent (mm Hg)	Superior Sagittal Sinus Post-stent (mm Hg)	Gradient Pre-stent (mm Hg)	Gradient Post-stent (mm Hg)
Single stent (<i>n</i> = 40, all with papilledema)	31 (15–94) (<i>n</i> = 40)	17 (11–33) (<i>n</i> = 39)	19 (6–36) (<i>n</i> = 40)	0.4 (0–5) (<i>n</i> = 40)
Repeat stents, on 1st stent (<i>n</i> = 6, all with papilledema prior to first stent)	43 (18–56) (<i>n</i> = 6)	18 (11–32) (<i>n</i> = 5)	29 (15–41) (<i>n</i> = 6)	2.8 (0–14) (<i>n</i> = 6)

^a Ranges indicated in parentheses.



Pascal Jabbour MD FAANS, FACS, FAHA • 2nd

The Angela and Richard T. Clark Distinguished Professor of ...

2mo • 🌐

+ Follow ...

For the non believers :

Progressive blindness with headaches

1- Venous manometry 98 with a gradient of 70!

2- Severe T-S stenosis stented with Zilver 9x40 with post plasty

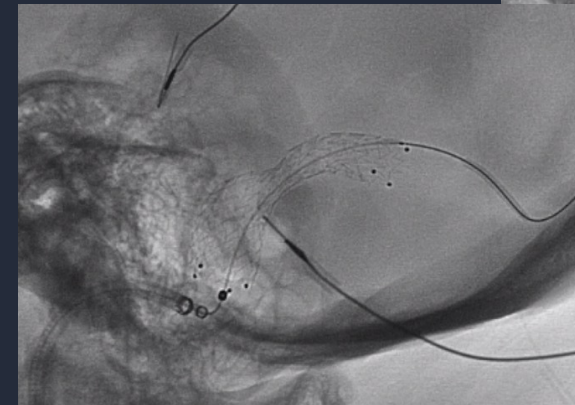
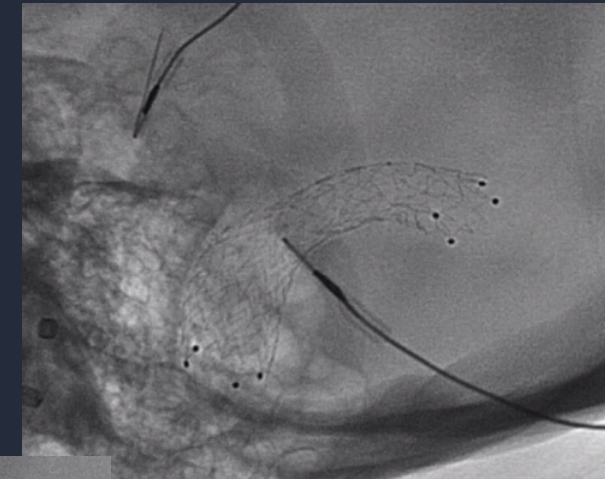
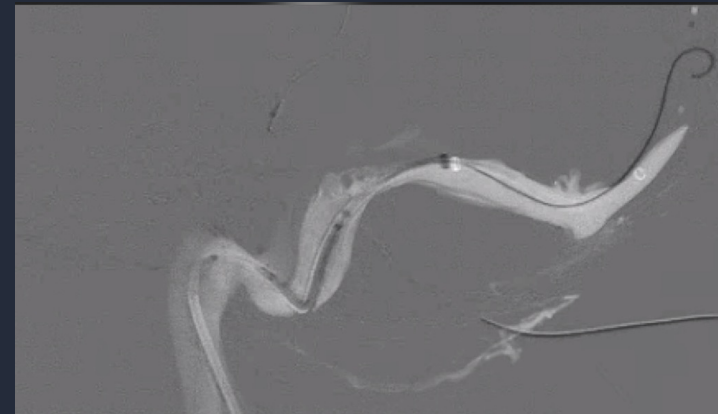
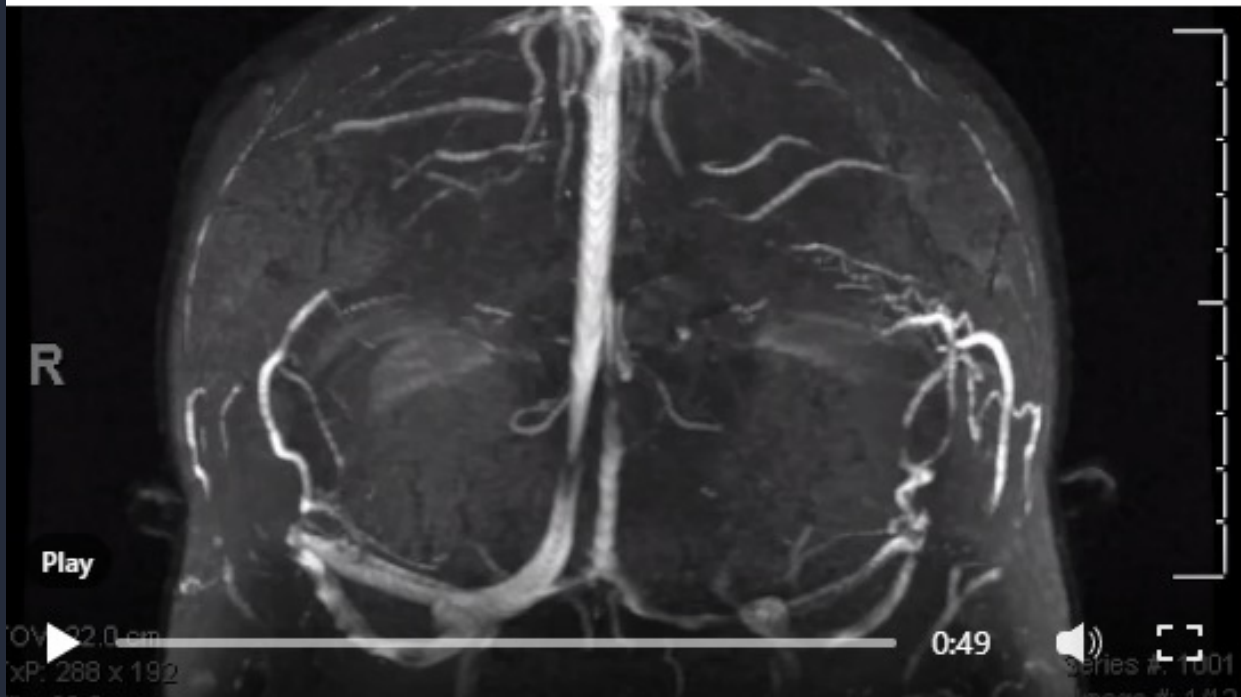
3- Pressure down to 18 !

4- significant vision improvement in 24 hours

#IIH #SinusStenting

Jefferson Neurosurgery JEFFERSON HOSPITAL FOR NEUROSCIENCE Thomas

Jefferson University Hospitals




March 2023



Medical Imaging—Original Article

Intracranial venous stenting for idiopathic intracranial hypertension

Frederick Ong , Timothy Phillips, Gregory Selkirk, William McAuliffe

First published: 16 January 2023 | <https://doi.org/10.1111/1754-9485.13505>

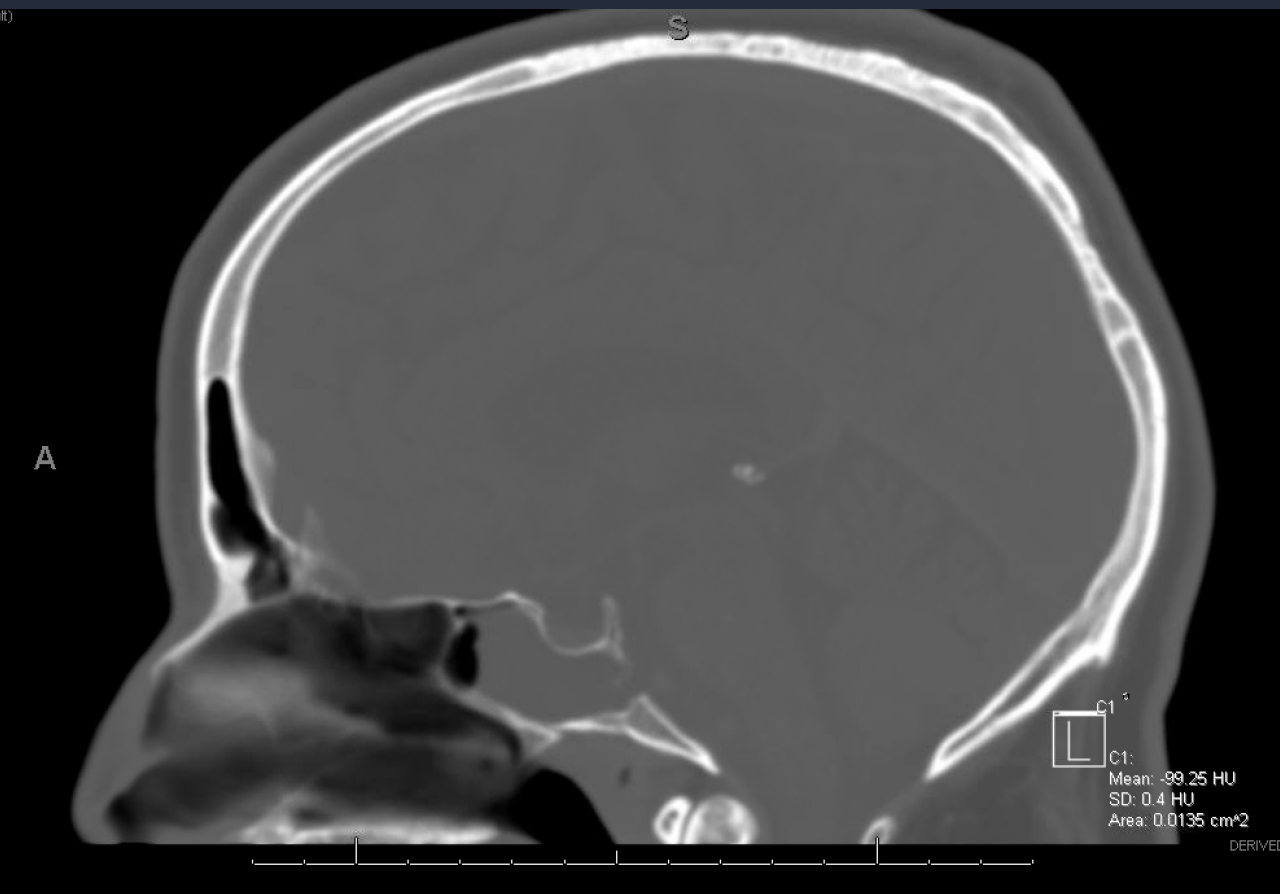
Results

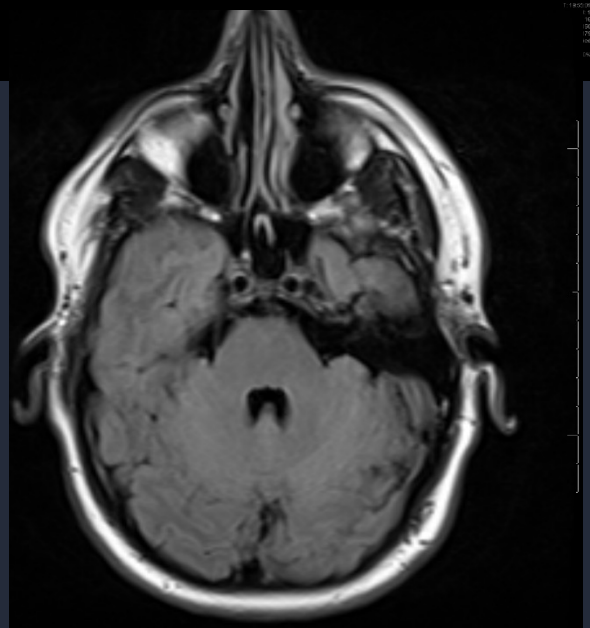
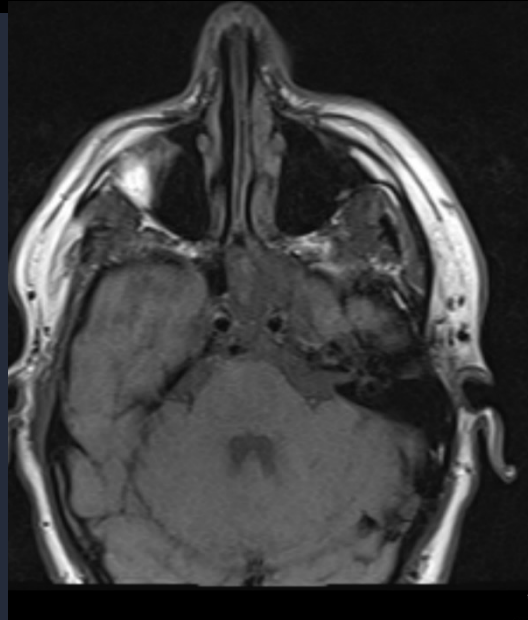
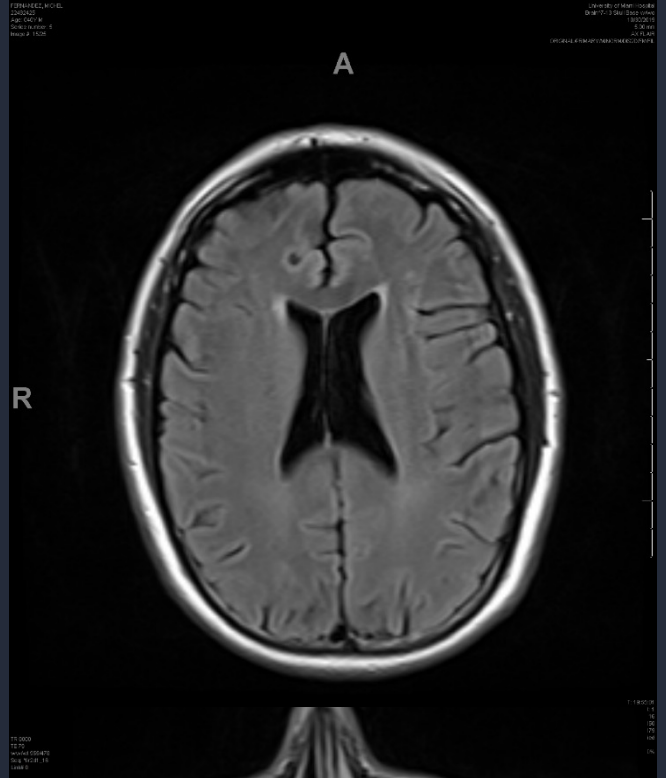
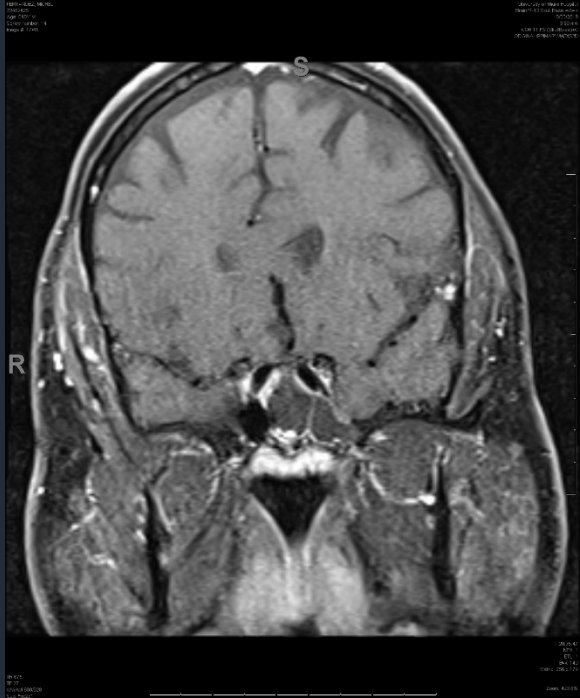
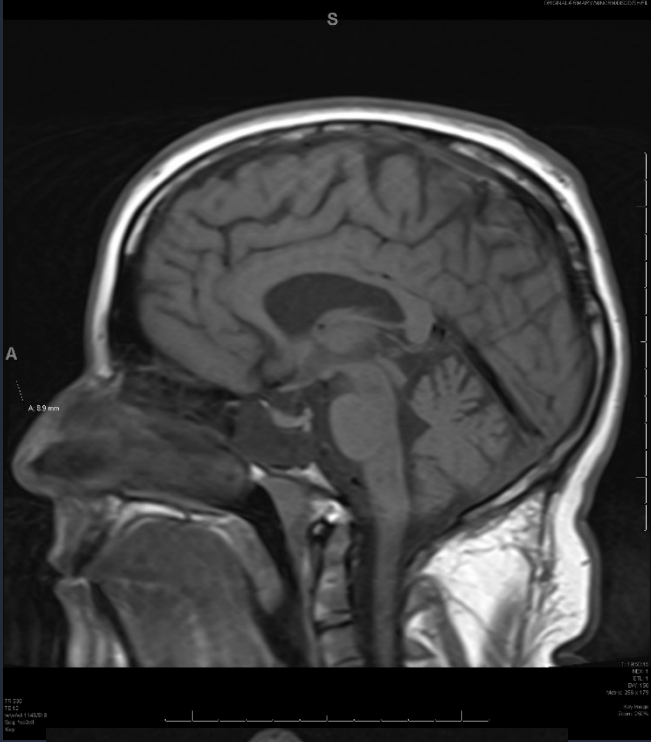
89.2% patients were able to cease Acetazolomide post stenting. 78.3% patients reported resolution of headaches. 84.3% patients demonstrated resolution of their papilloedema. 91.6% patients demonstrated improvement of their visual acuity. Compared with a recent meta-analysis by Satti and Chaudry in 2015, our results demonstrated a higher technical success rate and lower complication rates.

**Is There a Different Imaging/Clinical
Presentation for IIH ?**

Case presentation

- 40 y.o Male
- 4 weeks prior to presentation: rapid onset of severe headaches when traveling followed by constant dripping of clear fluid from the left nostril.
- BMI 32, questionable weight gain over the past year
- PMH none
- SH none
- Meds none





T1 W

T2 W

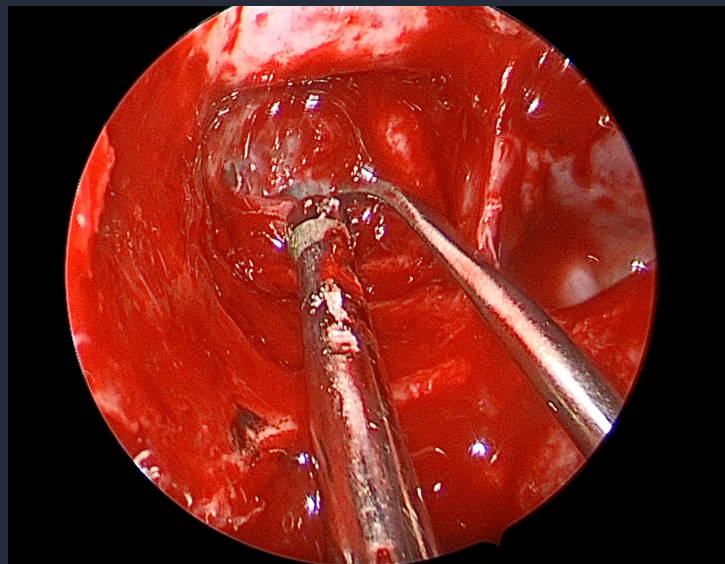
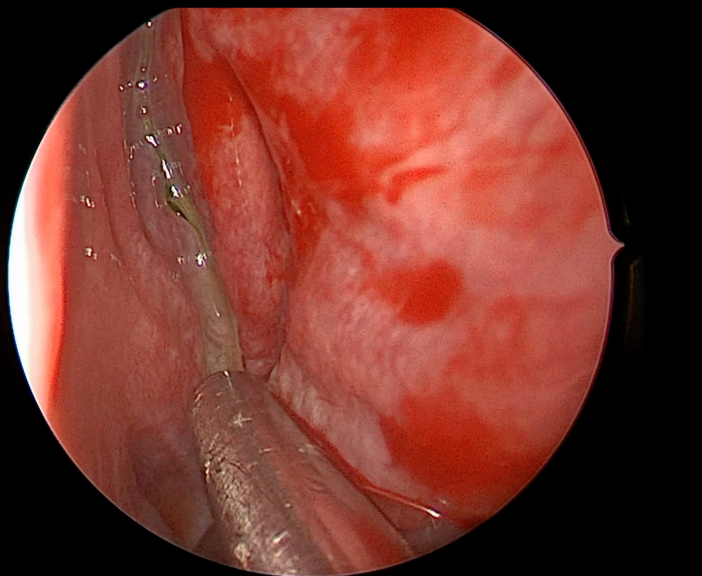
FLAIR

Case

- Ophthalmology: No Papilledema, No VF defect
- 2 team surgery for resection of meningocele and repair of CSF leak with Lumbar drain placement and intrathecal fluorescein injection
- Lumbar drain placed preoperatively, normal opening pressure, Fluorescein injected intrathecally.

Case presentation

- Reconstruction done using multilayer technique (Allograft + left sided nasal septal flap)
- Lumbar drain kept open at 5 ml an hour for 4 days.
- LD clamped on POD 4, patients started leaking on POD 6.



**C/O Zoukaa Sargi, MD MPH
NASBS 2020**

Case presentation

- Taken back to the OR one week later for revision repair:
- Abdominal fat graft and nasal septal flap
- Repair challenged intraoperatively
- Lumbar Drain kept managed similarly
- with every clamp attempt, the patient starts leaking again. ICP measures start going up.
- Starts developing intermittent leaking despite the drain in place
- → VP shunt

sCSF leak Vs IIH

- S-CSF leak is a form of IIH, occurring in a subset of patients ¹⁻³
- Evidence: S-CSF leak patients post surgical repair of skull base defect develops elevated ICP and signs and symptoms of IIH⁴

1. Goddard JC, et al. Otol Neurotol 2010
2. Lloyd KM et al. Radiology 2008
3. Silver RI et al. Am J Rhinol. 2007
4. Mokri B et al. Mayo Clinic Proc. 2002

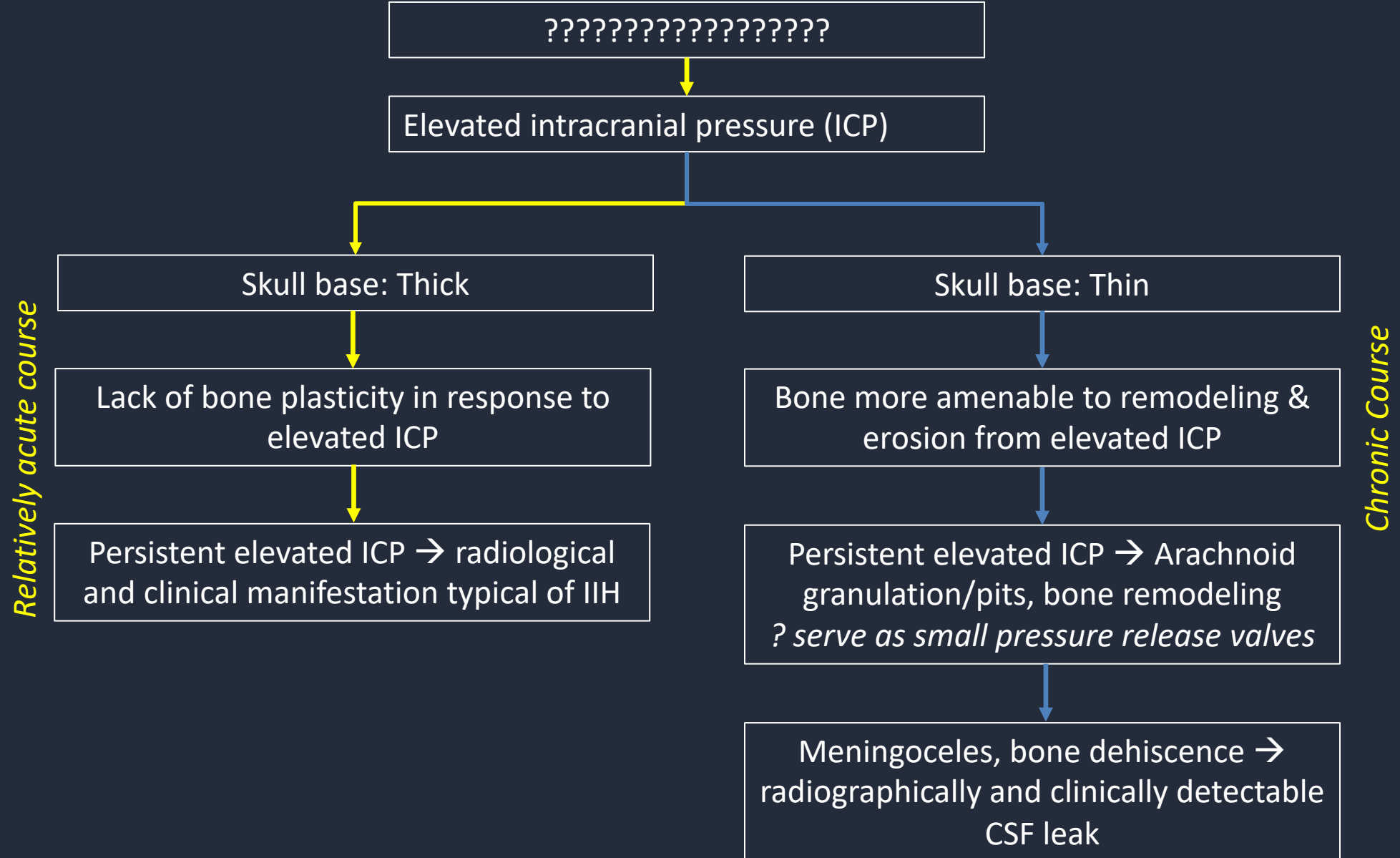
Skull Base Thickness: A Proposed Factor Linking the Pathogenesis of Spontaneous CSF Leak & IIH*

Two Different Diseases

Vs

One Disease with Two Manifestations?

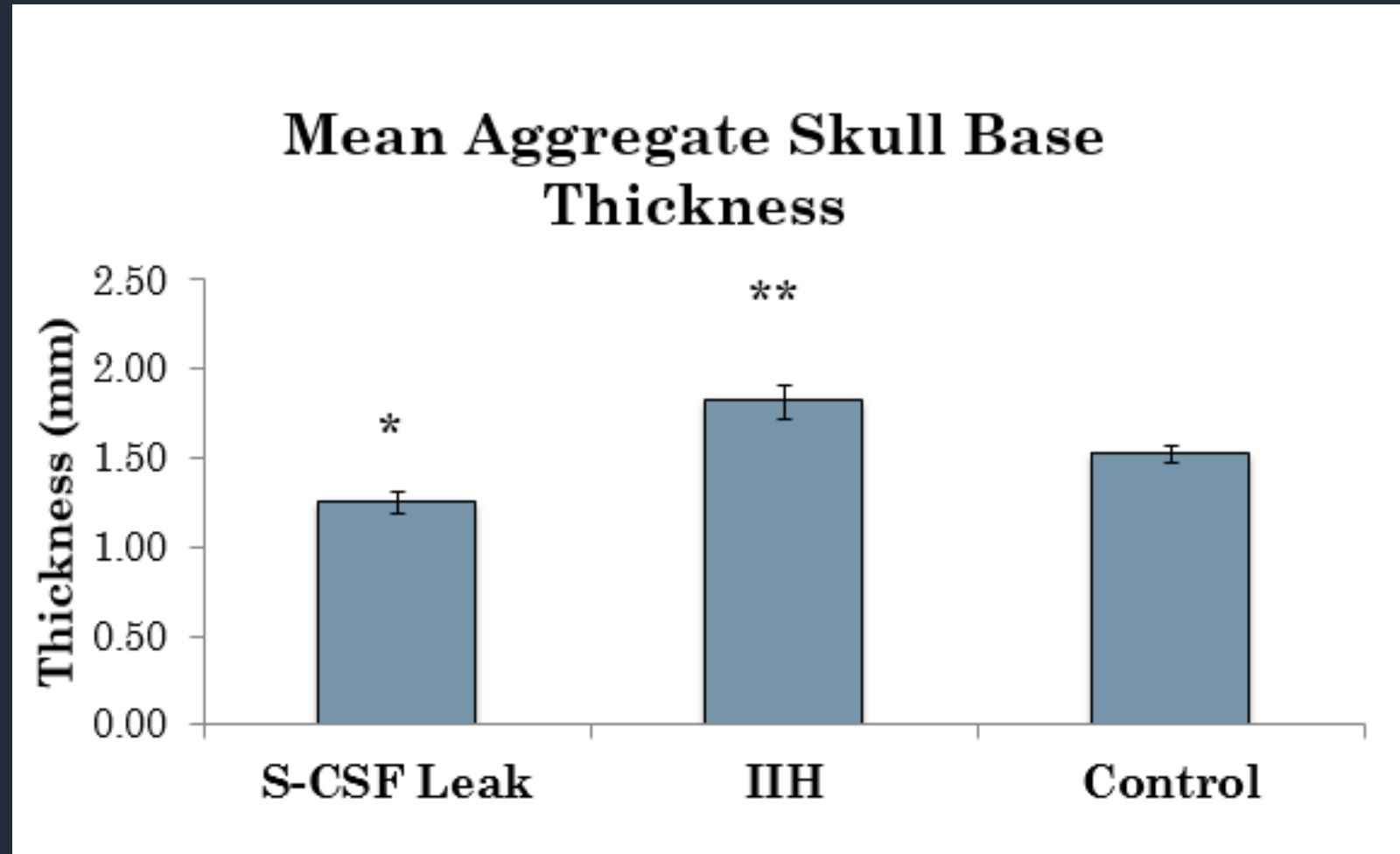
One Disease: Two Manifestations



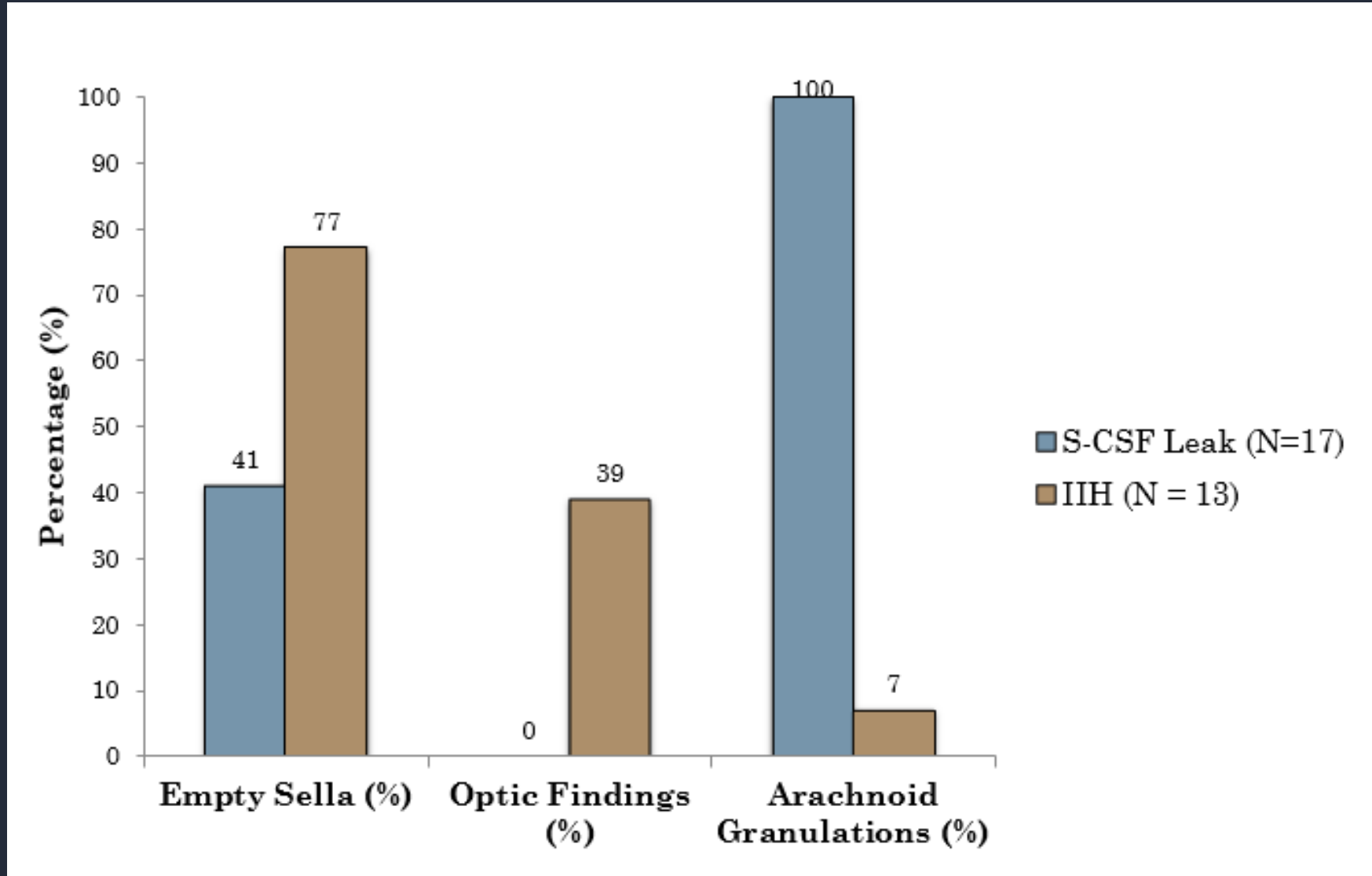
Statistically Significant Results

Statistically Significant Results


- IIH: younger age
- Female
Predominance in
Both groups
- Av BMI > 30 in both
groups



Statistically Significant Results



Intervention for Elevated Intracranial Pressure Improves Success Rate After Repair of Spontaneous Cerebrospinal Fluid Leaks

William Teachey, BS; Jessica Grayson, MD; Do-Yeon Cho, MD; Kristen O. Riley, MD;
Bradford A. Woodworth, MD, FACS  Laryngoscope . 2017

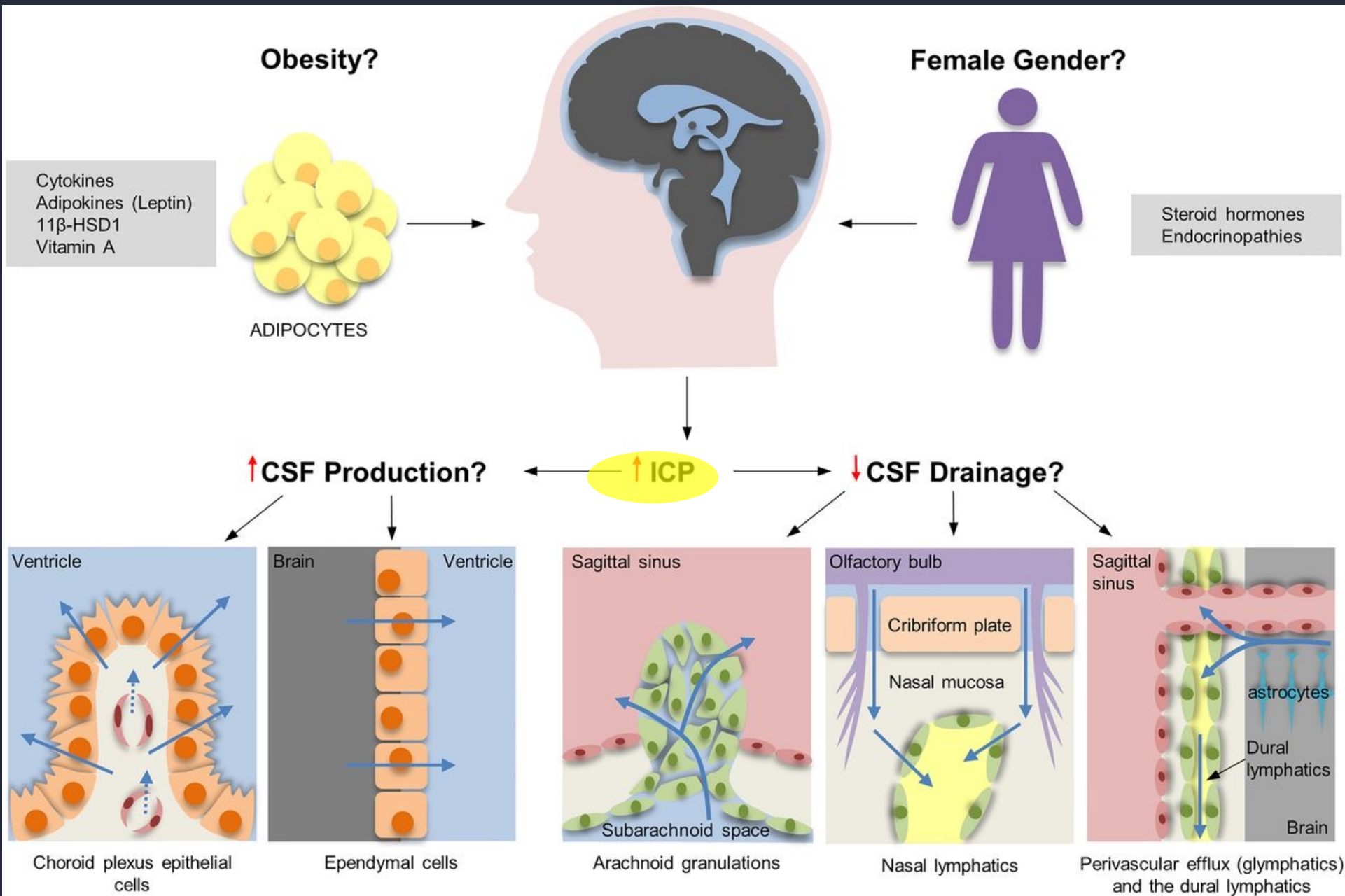
56 articles, 679 patients

TABLE III.
Comparison of ICP Intervention.

Patient Cohort	No. Treated	No. of Failures	Success Rate	Mean Follow-up, mo
ICP intervention	348	25	92.82%	24.1 ± 2.9
No ICP intervention	331	60	81.87%	26.9 ± 3.7

ICP = intracranial pressure.

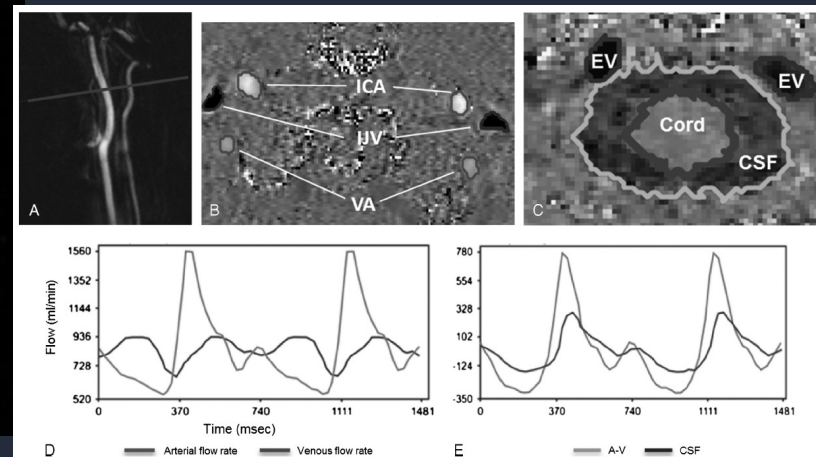
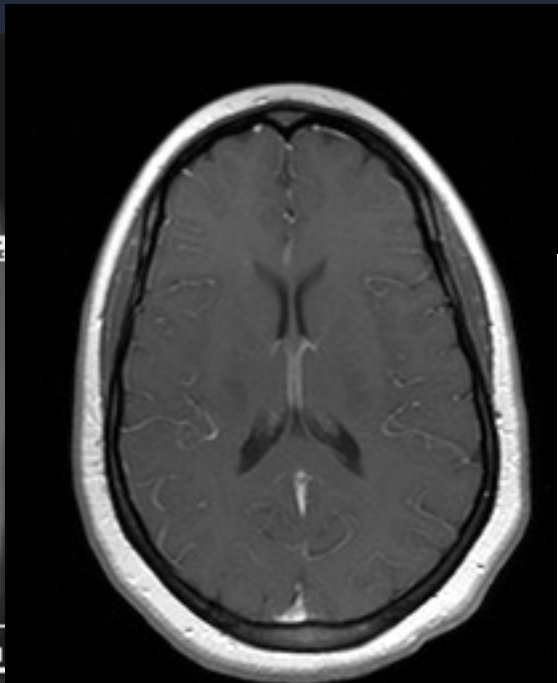
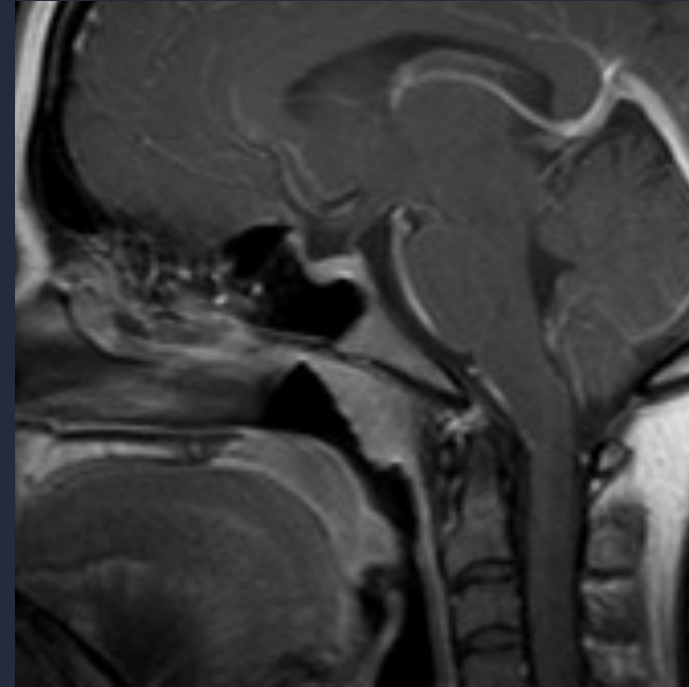
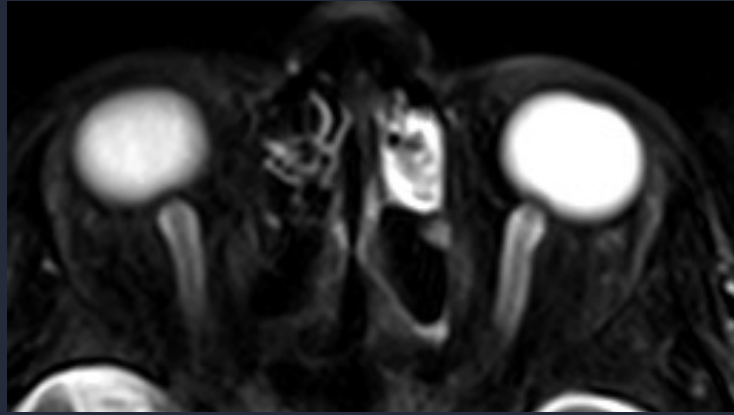
Take home points



↑CP activity1
Vit A (Aquaporin 1
rec
Mineralocorticoid
rec

Mollan et al J
Neurol Neurosurg
Psychiatry 2015

Take Home Points



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