

Pseudotumor Cerebri

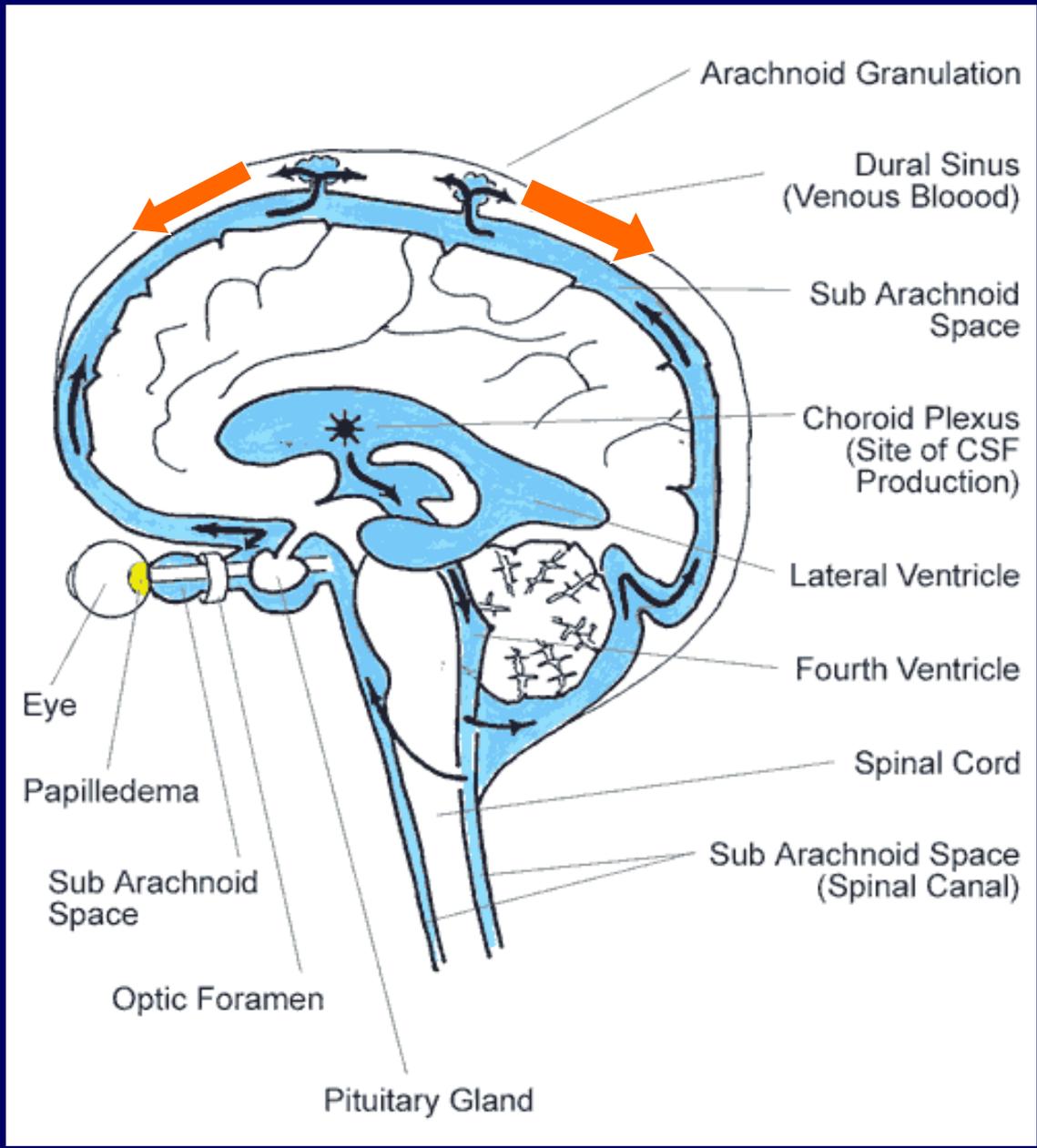
Hydrocephalus Association 12th National Conference
Bethesda MD
June 27-July 1 2012.

Michael Altman MD

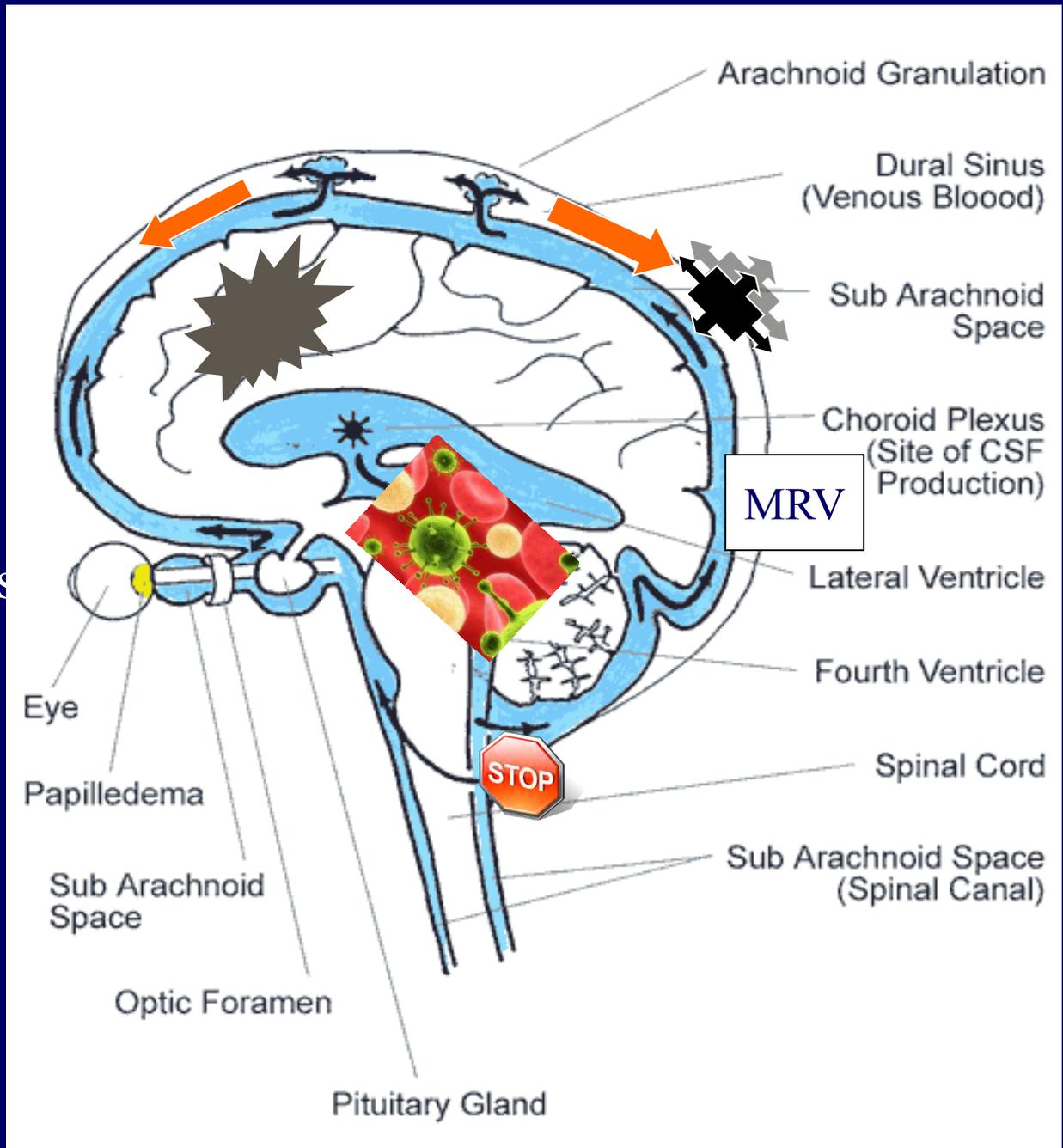
Director Pediatric Ophthalmology
Director Neuroophthalmology
Krieger Eye Institute
Sinai Hospital of Baltimore

Cerebrospinal Fluid (CSF)

- The ventricles produce special fluid which bathes and cushions the brain and spinal cord within their bony confines.
- Protects the brain , provides buoyancy
- **Flows freely** from the ventricles into the subarachnoid space
- Then into the large veins
- Veins finally drain blood and CSF out of the brain



- Space occupying
- Obstruction CSF
- Obstruction Vein
- Comm. Hydrocephalus
- None of the above ?
- Pseudotumor Cerebri



Have Symptoms of High Pressure ?

- Everyone should be evaluated promptly for the cause
- MRI
- MRV
- Spinal Tap
- Eye Exam
- All causes of high pressure can lead to visual loss

High Intracranial Pressure

Symptoms

- Nausea, Vomiting , Lethargy, Neck / Back pain
- Pulsatile Tinnitus (ringing in the ears)
- Change in personality, memory, cognition
- Headache
- Double Vision
- Visual loss
 - Transient
 - Permanent
 - Slowly
 - Quickly

Pseudotumor Cerebri

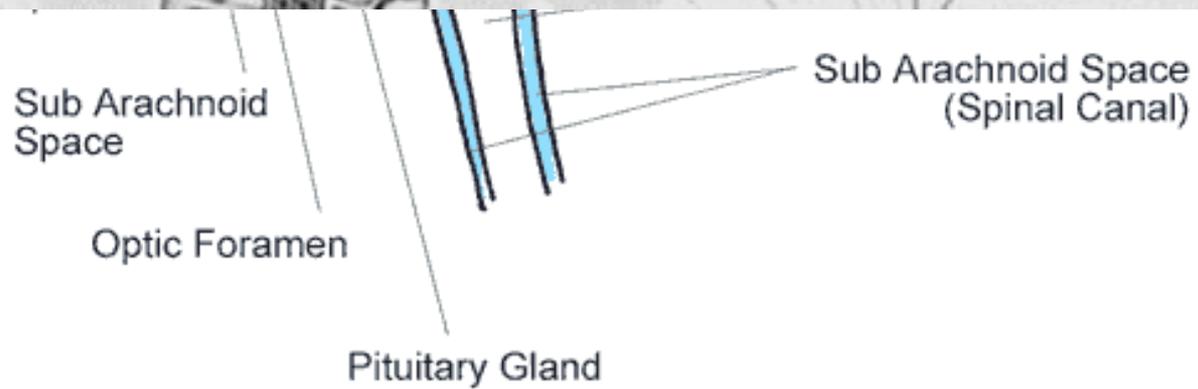
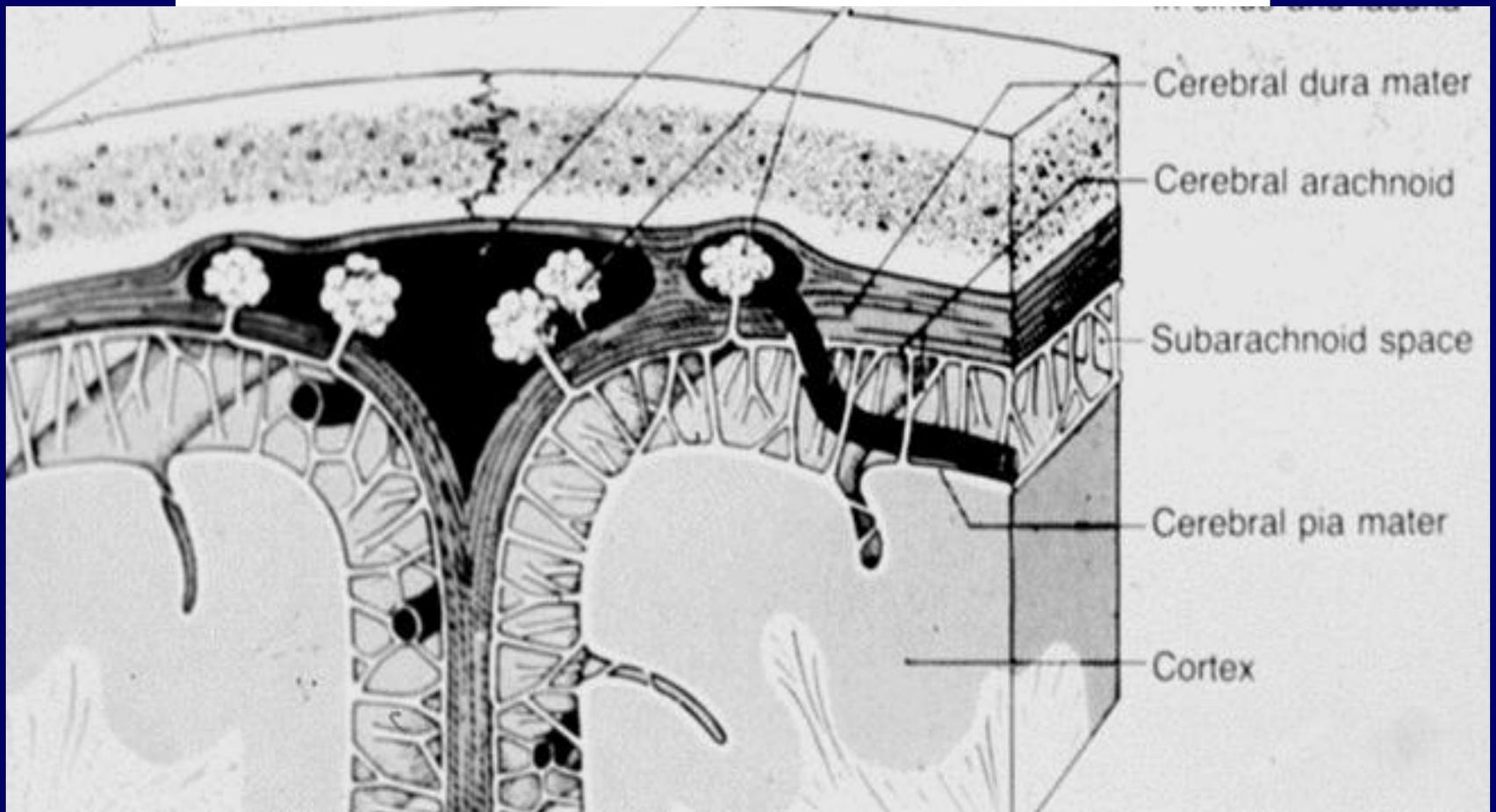
- Symptoms/Signs of increased pressure
- No Mass/Tumor (MRI is normal!)
- Ventricles of normal size (MRI is still Normal !)
- CSF healthy composition (Spinal fluid is normal)
- Yet – *CSF pressure is high*

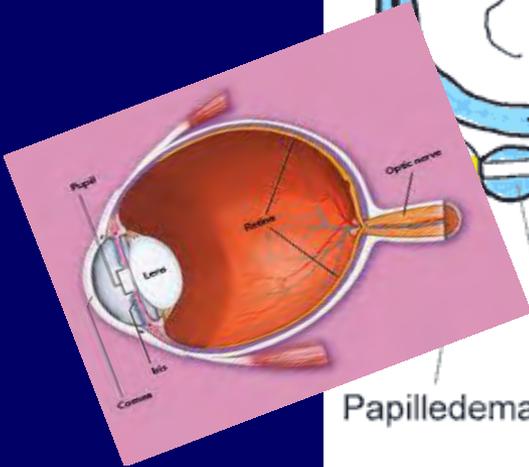
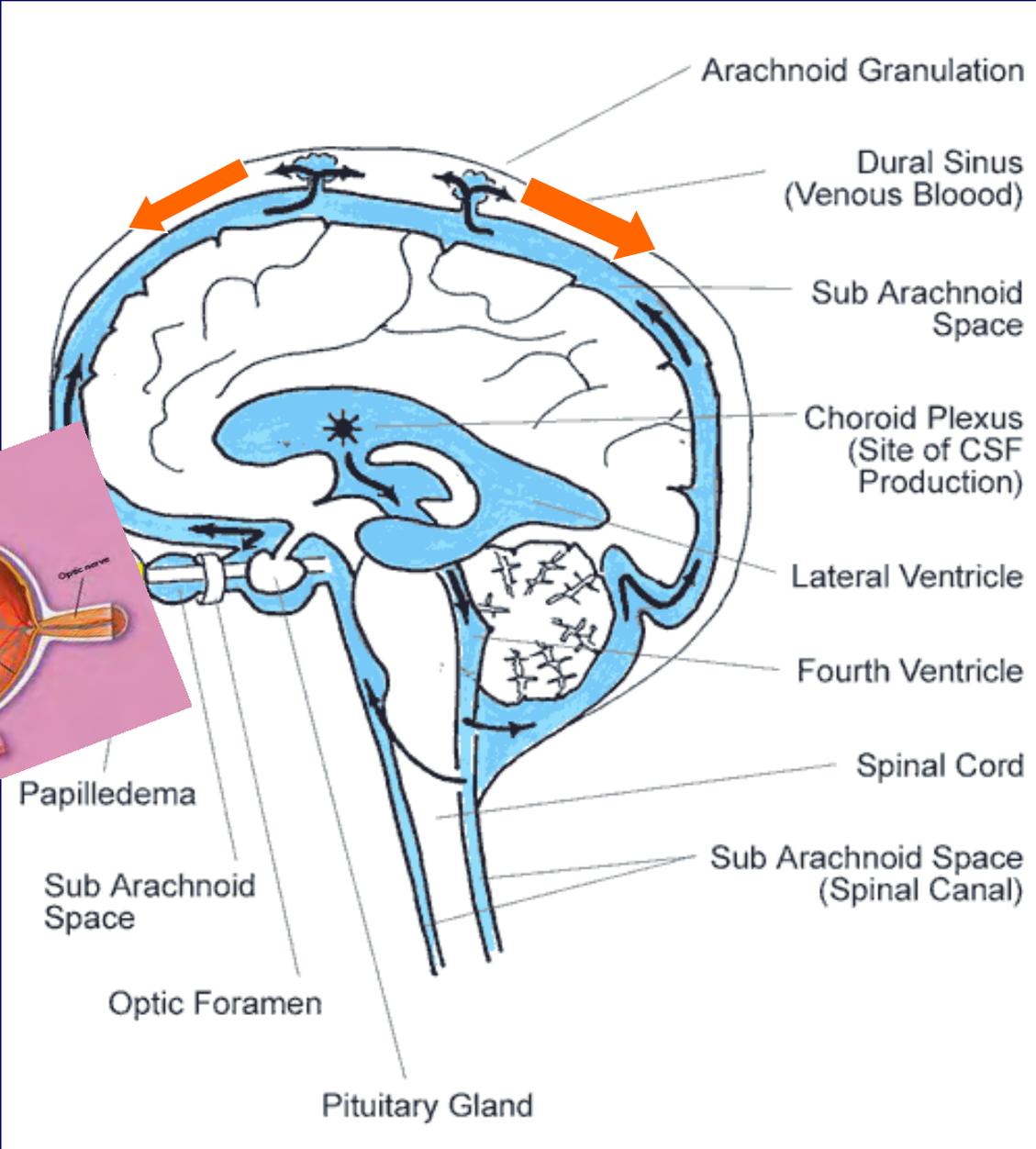


"It's Lyme disease again."

Etiology ?





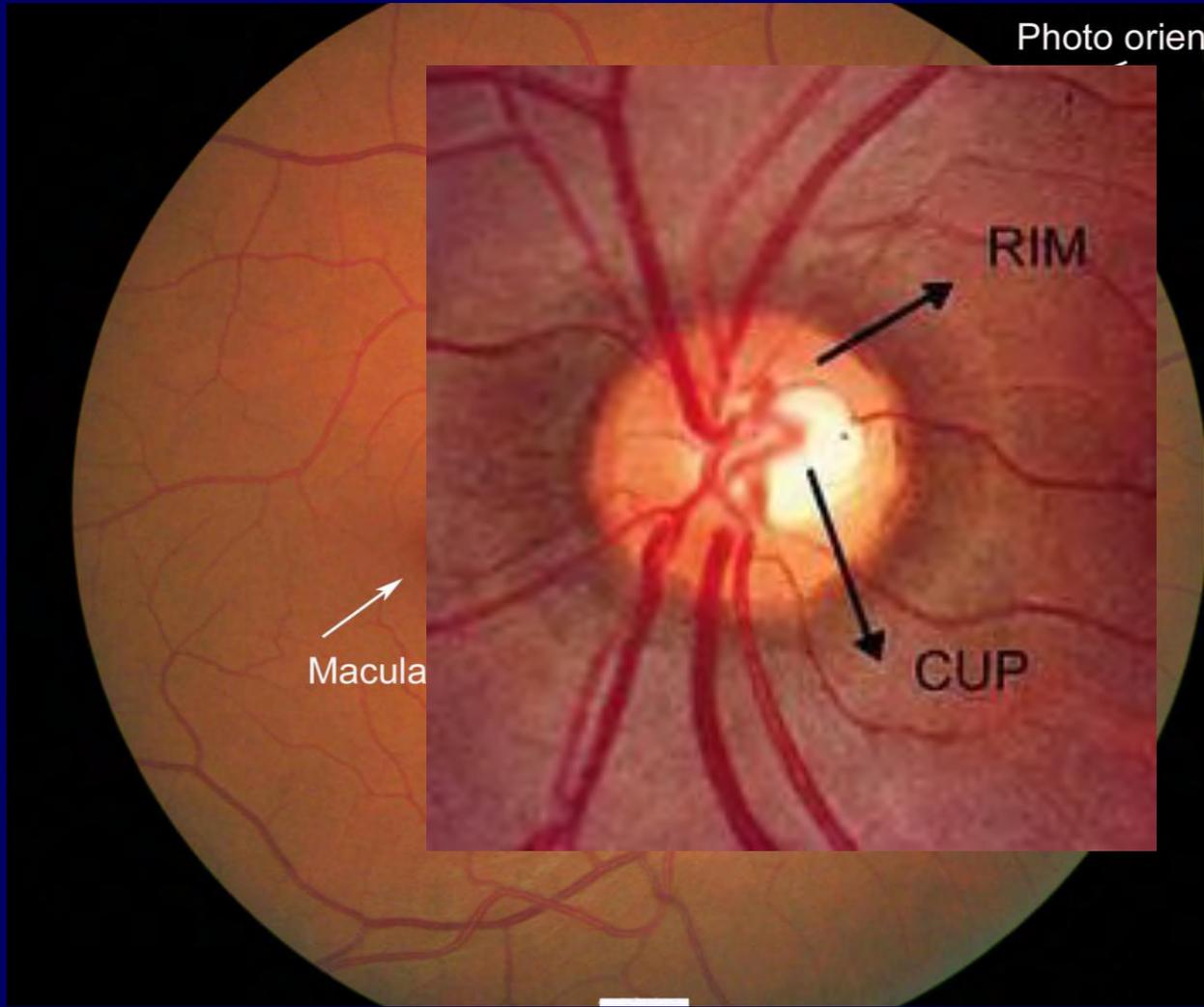


High Intracranial Pressure Papilledema

- Definition :
- Optic Nerve swelling due to elevated intracranial pressure



Photo orient



Macula

RIM

CUP





Photo orien



For Example

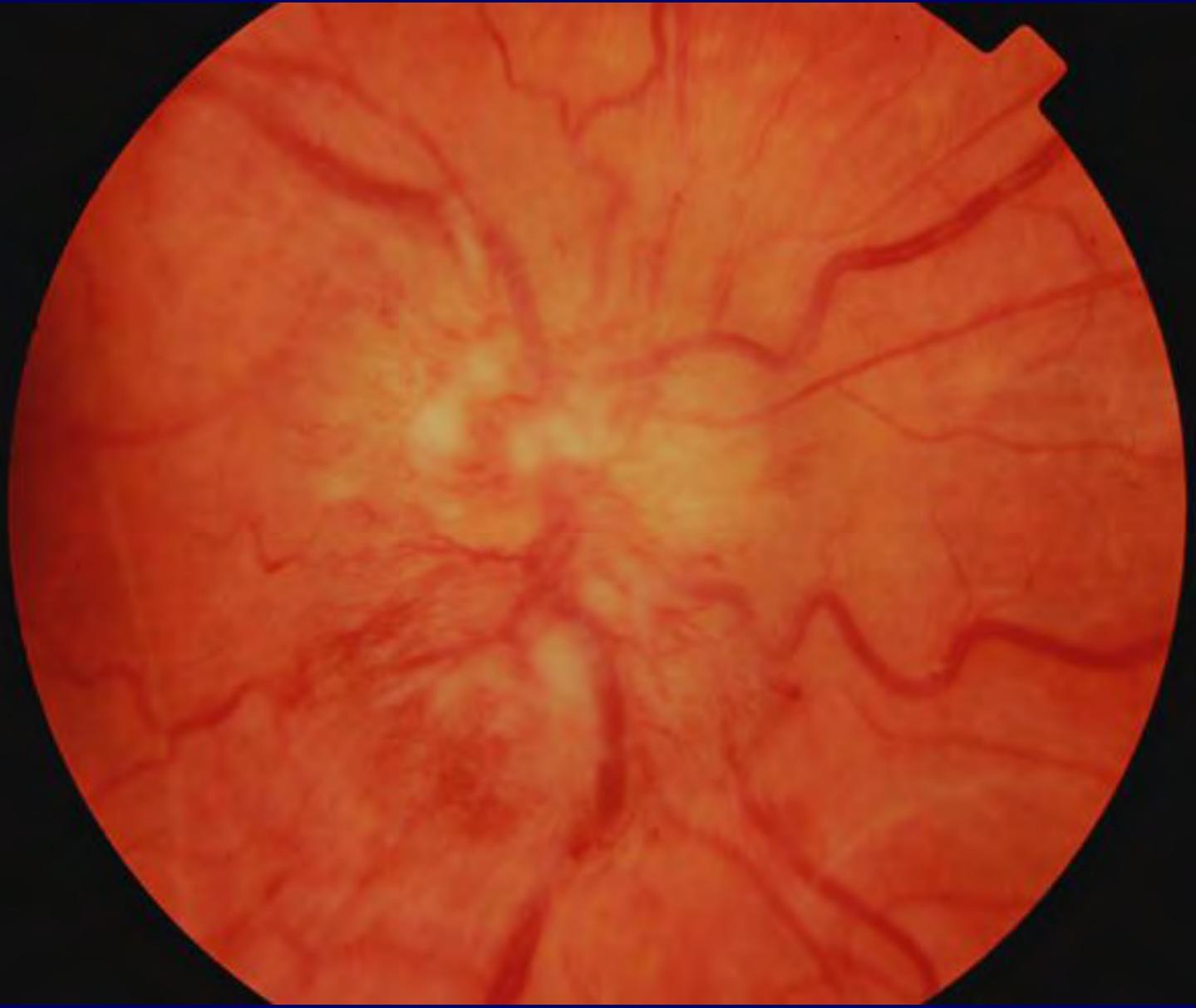


- Healthy 12 yo with HA for one month
- Minocycline has cleared her acne
- Began to lose vision the past week
- “Everything is getting dim”
- Nausea

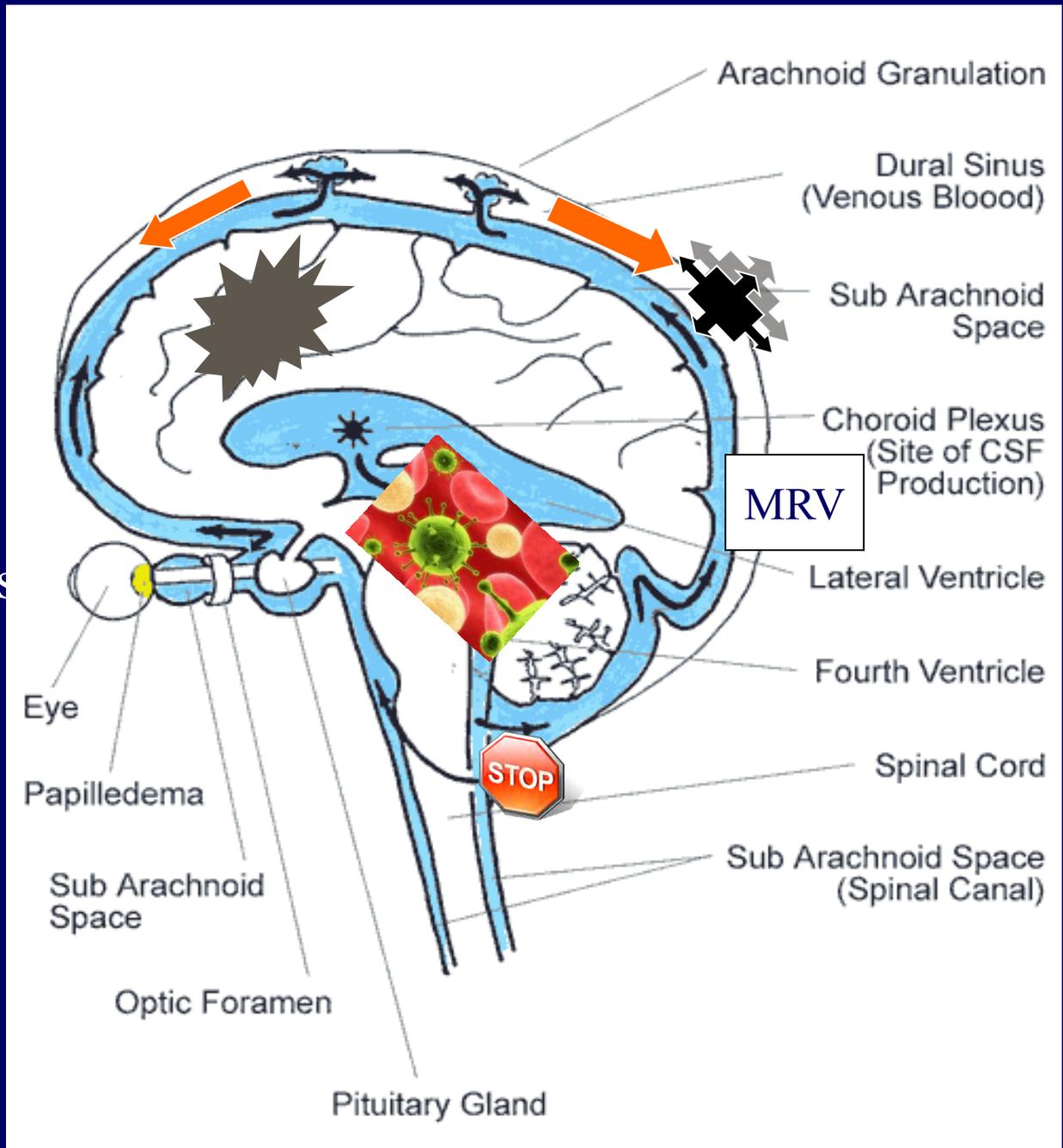
“...There is a Stomach Virus Going Around...”

- Seen in the ER with severe headache
- 20/200 OD and 20/80 OS
- Double Vision
- Irritable
- Feeling sick 4 weeks





- Space occupying
- Obstruction CSF
- Obstruction Vein
- Comm. Hydrocephalus
- None of the above ?
- Pseudotumor Cerebri



Doing A Bit Better



- MRI/MRV normal
- Spinal Tap
- Opening pressure off the manometer
- Happy go lucky child again!
- HA all but gone
- 20/50 OD and 20/25 OS

Pseudotumor Cerebri

- Symptoms/Signs of increased pressure
- No Mass/Tumor (MRI is normal!)
- Ventricles of normal size (MRI is still Normal !)
- CSF healthy composition (Spinal fluid is normal)
- Yet – *CSF pressure is high*

Pseudotumor Cerebri

Epidemiology

- 0.9 per 100,000
- 1.6 per 100,000 women
- 3.3 per 100,000 women age 15-44
- 7.9 per 100,000 obese women age 15-44
- F:M = 8:1
- Pre-puberty M=F

Mayo clinic 1976-1990

Pseudotumor Cerebri



- HA
- N/V
- Pulsatile Ringing in the ears
- Neck / Back pain
- Double vision
- Transient visual darkening
- Permanent visual loss

Pseudotumor Cerebri



- Tetracyclines
- Vitamin A
- Growth Hormone
- Cyclosporine
- Craniosynostosis
- Venous Sinus Thrombosis
- Arnold Chiari malformation
- Steroid use / withdrawal

Pseudotumor Cerebri

Treatment

- Weight reduction
- Treat Sleep Apnea if present
- Diamox
- Topamax
- Discontinue 'causative' medication
- *Optic nerve sheath decompression*
- LP / VP shunt

Surgical Treatment Pseudotumor Cerebri

- HA > Vision
 - CSF shunting procedure (LP/VP)

Advantages:

Decompress entire system
Relieves Headache well
Improve all symptoms

Problems:

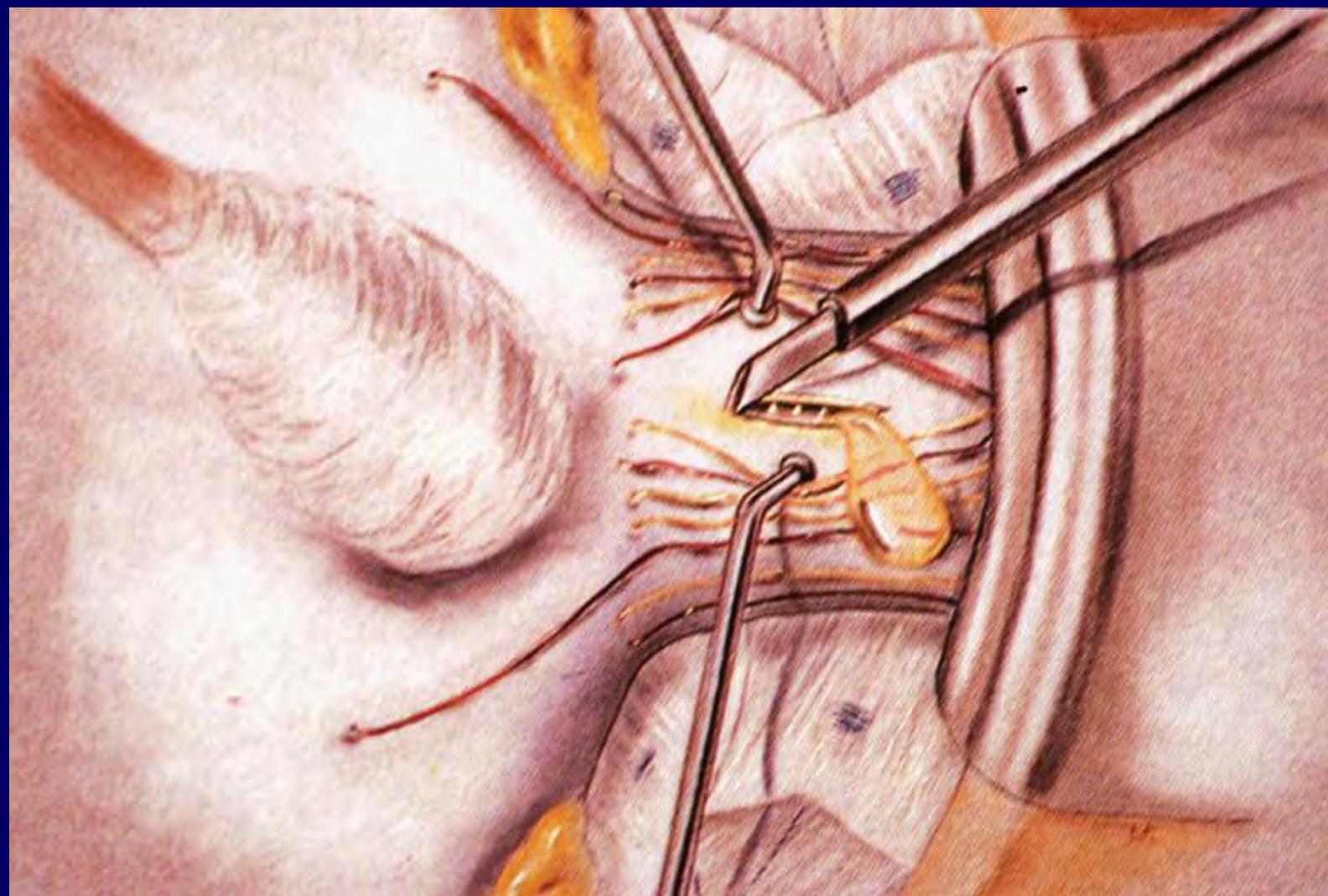
-Shunt dysfunction
-Over drainage
-Shunt revisions
-Abdominal pain

■ ? Stenting of Venous Sinuses

Pseudotumor Cerebri

Optic Nerve Decompression

- Visual loss > HA
- Powerful procedure for visual loss
- Relieves HA about 50-60%
- Can achieve long lasting results



Multiple Personality



- Overnight, Symptoms return
- Total of 3 LP's thru the night
- VP shunt in the morning
- Vision stabilizes at 20/80 and 20/40
- But over the ensuing 10 days
- Losing side vision



CENTRAL 30-2 THRESHOLD TEST

FUNCTION MONITOR: GAZE-BLIND SPOT
 FUNCTION TARGET: CENTRAL
 FUNCTION LENSES: 4/12 BX
 FALSE POS ERRORS: 0 X
 FALSE NEG ERRORS: 0 X
 TEST DURATION: 04:11

STIMULUS: III - WHITE
 BACKGROUND: 31.5 DB
 STRATEGY: STD-FAST

PUPIL DIAMETER:
 VERTICAL: 6.5 MM
 HORIZONTAL: 6.5 MM

DATE: 02-01-2011
 TIME: 04:04 AM
 REG: 43

POWER: 27 DB



0	0	-5	-4					
-1	0	-1	-2	-3	-7			
2	-2	-1	-4	-2	-5	-4	-3	
1	0	0	0	-2	-2	-2	-4	-2
-2	-2	1	0	-1	-1	-1	-2	-2
-1	-2	2	1	-1	0	-2	-5	-5
-1	-2	-3	-1	0	1	-1	-2	0
-1	-3	0	-1	-4	-2	-1	1	
-3	-1	0	-1	-1	0			
2	0	0	-2					

-1	-1	-3	-4					
-2	0	-1	-2	-4	-7			
1	-2	-2	-4	-2	-5	-4	-3	
0	0	0	0	-2	-2	-2	-4	-2
-3	-2	1	0	-1	-1	-2	-2	-2
-2	-2	2	1	-1	0	-2	-5	-5
-3	-3	-1	-2	0	1	-2	-2	0
-1	-3	-1	-1	-4	-2	-1	0	
-3	-1	0	-1	-1	0			
1	0	0	-2					

MIN LOW TEST RELIABILITY: 66
 CRT
 OTHER NORMAL LIMITS

NO: -1.42 DB F / 100
 PD: 1.00 DB

TOTAL
 DEVIATION



PATTERN
 DEVIATION



○ 1.5
 ● 1.2
 ■ 1.1
 ■ 0.5

SAVED X AXIS: NO
 FROM OLD DEVIATION NO
 SLOPE C-28
 WETHERS, NO 20014
 301-548-2786/301-548-1522

CENTRAL 30-2 THRESHOLD TEST

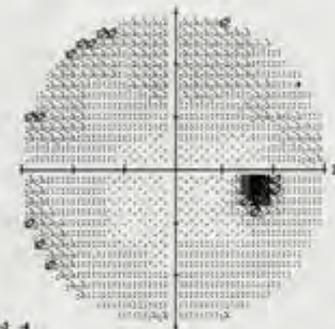
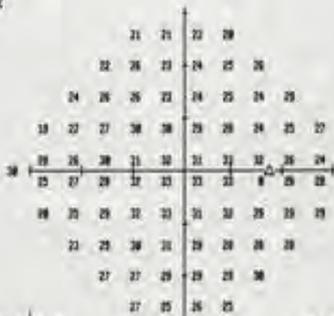
FUNCTION MONITOR: GAZE-BLIND SPOT
 FUNCTION TARGET: CENTRAL
 FUNCTION LENSES: 5/12 BX
 FALSE POS ERRORS: 13 X
 FALSE NEG ERRORS: 12 X
 TEST DURATION: 04:11

STIMULUS: III - WHITE
 BACKGROUND: 31.5 DB
 STRATEGY: STD-FAST

PUPIL DIAMETER:
 VERTICAL: 6.5 MM
 HORIZONTAL: 6.5 MM

DATE: 02-01-2011
 TIME: 04:04 AM
 REG: 43

POWER: 27 DB



-4	-5	-3	-5				
-4	-5	-6	-4	-2	-1		
-3	-4	-3	-6	-5	-6	0	
-3	-5	-2	-3	-3	-6	-5	-3
0	-3	-2	-2	-2	0	-5	-7
-2	-2	-4	-3	0	0	0	-2
-3	-3	-1	-1	-2	-1	-3	-2
-4	-2	-1	-3	-4	-3	-1	
-2	-3	-1	-1	-2	-1		
-2	-4	-4	-4				

-5	-5	-2	-4					
-4	-2	-5	-2	0				
-4	-3	-4	-7	-2	-4	-5	0	
-4	-3	-4	-2	-2	-2	-4	-4	-2
1	-4	-2	-1	-2	1	-4	-4	
-2	-3	-3	0	1	0	1	-4	-2
-7	-4	-2	0	0	-1	0	-1	-1
-4	-1	-1	0	-2	-2	-2		
-1	-2	0	-1	0				
-1	-2	-3	-4					

MIN EXCESSIVE HIGH FALSE POSITIVES: 66
 CRT
 OTHER NORMAL LIMITS

NO: -2.97 DB F / 25
 PD: 2.24 DB F / 100

TOTAL
 DEVIATION



PATTERN
 DEVIATION



○ 1.5
 ● 1.2
 ■ 1.1
 ■ 0.5

SAVED X AXIS: NO
 FROM OLD DEVIATION NO
 SLOPE C-28
 WETHERS, NO 20014
 301-548-2786/301-548-1522

CENTRAL 24 - 2 THRESHOLD TEST

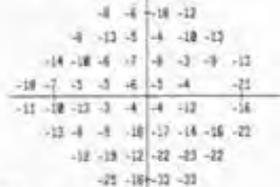
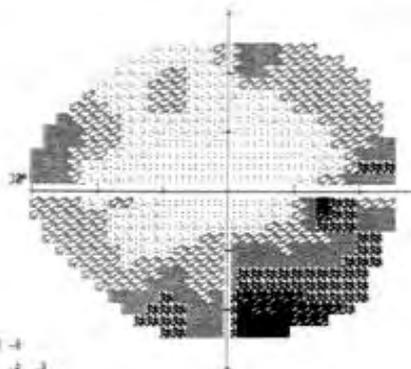
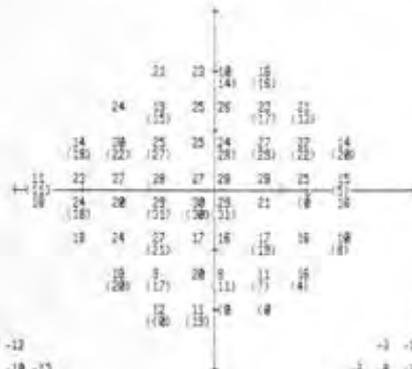
NAME

RIGHT

STIMULUS III, WHITE, SCAND 31.5 408 8.0MS SPOT CHECK SIZE III
STRATEGY FULL THRESHOLD

BIRTHDATE 04-22-68 DATE 03-28-97
FIXATION TARGET CENTRAL 03 TIME 08:27:46 AM
EX OSCL 8.00 OS DCI DEG Pupil DIAMETER 3.8 MM VA 19/20

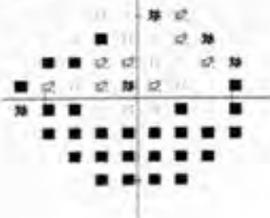
AGE 28
FIXATION LOSSES 0/25
FALSE POS ERRORS 0/13
FALSE NEG ERRORS 0/12
QUESTIONS ASKED 471
FOURAT 14 38 11
TEST TIME 13:11
HFA 0/4 048-1633



GLAUCOMA HEMIFIELD TEST (GHT)
OUTSIDE NORMAL LIMITS

TOTAL
DEVIATION

PATTERN
DEVIATION



PROBABILITY SYMBOLS

- P < .1%
- ◻ P < .2%
- ◻ P < .3%
- ◻ P < .5%

MD -11.27 DB P < 0.1%
P50 -7.72 DB P < 0.2%
SP -3.43 DB P < 2%
CPSD -3.08 DB P < 0.1%

GRAYTONE SYMBOLS

REV 8.2 X2

SYM								
ASB	16	25	36	50	70	100	160	256
DB	0	1	2	3	4	5	6	7

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SINAI HOSPITAL
2411 N. BELVEDERE AVE
BALTO, MD, 21215

HUMPHREY INSTRUMENT
A CARL ZEISS COMPANY

Handwritten signature

CENTRAL 24 - 2 THRESHOLD TEST

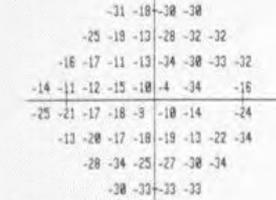
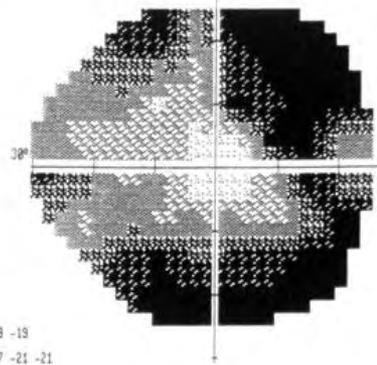
NAME

RIGHT

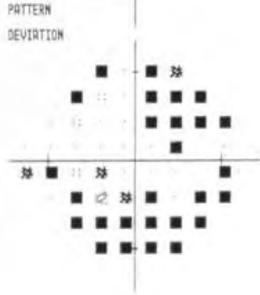
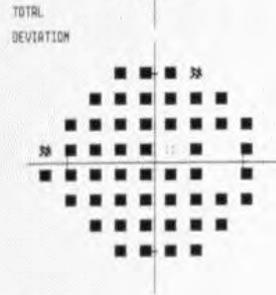
STIMULUS III, WHITE, BKGDND 31.5 RSB BLIND SPOT CHECK SIZE III
 STRATEGY FULL THRESHOLD

BIRTHDATE 04-22-68 DATE 03-19-97
 FIXATION TARGET CENTRAL ID TIME 04:16:59 PM
 RX USED + 0.00 DS OCK DEG PUPIL DIAMETER 5.8 MM VA 20/30

AGE 29
 FIXATION LOSSES 0/22
 FALSE POS ERRORS 0/13
 FALSE NEG ERRORS 2/12
 QUESTIONS ASKED 406
 TEST TIME 12:56
 HFA S/N 640-3635



GLAUCOMA HEMIFIELD TEST (GHT)
 OUTSIDE NORMAL LIMITS



PROBABILITY SYMBOLS
 □ P | 5%
 ○ P | 2%
 ⊗ P | 1%
 ■ P | 0.5%

MD -20.63 DB P | 0.5%
 PSD 9.74 DB P | 0.5%
 SF 2.20 DB P | 100%
 CPSD 9.46 DB P | 0.5%

GRAYTONE SYMBOLS REV 8.2 X2

SYM	1	2	3	4	5	6	7	8	9	10
ASB	0	2.5	8	25	79	251	794	2512	7943	?
	.1	1	3.2	10	32	100	316	1000	3162	10000
DB	41	36	31	26	21	16	11	6	1	10
	50	40	35	30	25	20	15	10	5	10

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 BALTO, MD, 21215

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CENTRAL 24 - 2 THRESHOLD TEST

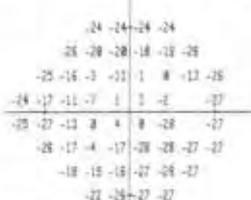
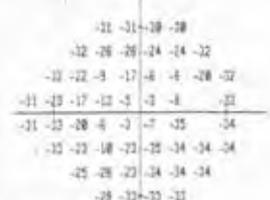
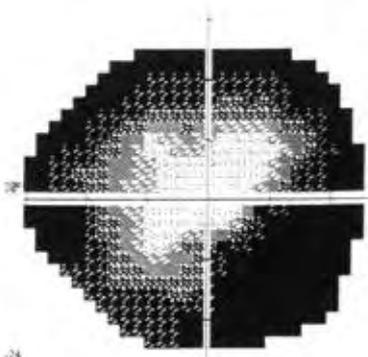
NAME

RIGHT

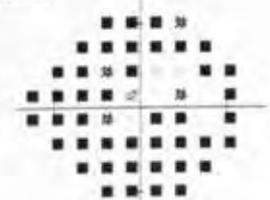
STIMULUS III, WHITE, BOXING 11.2 AND 16.2ND SPOT CHECK SIZE III
 STRATEGY FULL THRESHOLD

BIRTHDATE 04-22-68 DATE 03-21-97
 FIXATION TARGET CENTRAL 10 TIME 01:40:53 P
 EX. USE - 8.00 300 SEC PPD, 31MMETER 16

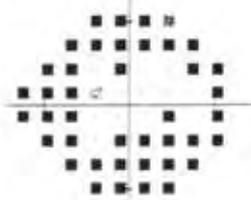
LOW PATIENT RELIABILITY



GLAUCOMA HEMIFIELD TEST (OBT)
 OUTSIDE NORMAL LIMITS



PROBABILITY SYMBOLS
 ○ = 1.00
 ◐ = 2.00
 ◑ = 3.00
 ◒ = 4.00



MD 21.38 DB P 8.00
 PD 12.22 DB P 8.00
 SF 3.88 DB P 10
 CPO 11.45 DB P 8.00

GRAYTONE SYMBOLS REV 9.2 K2

SPH										
REF	0.8	2.0	5	25	75	251	794	2512	7943	0
	1	3	10	32	100	316	1000	3162	10000	
DB	4.1	36	71	28	21	18	14	9	1	0.49
	6.9	48	105	38	28	28	18	10	5	1

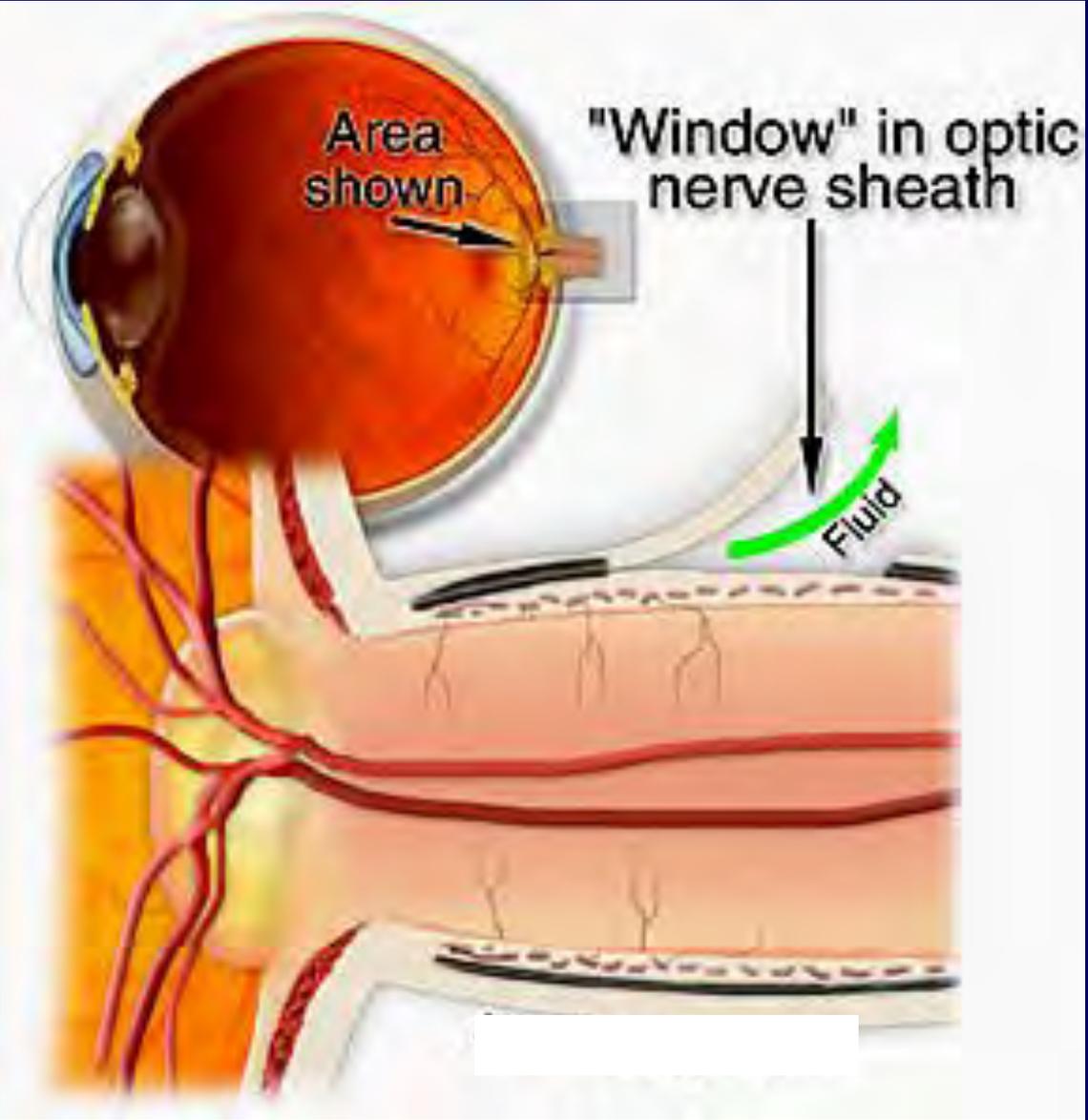
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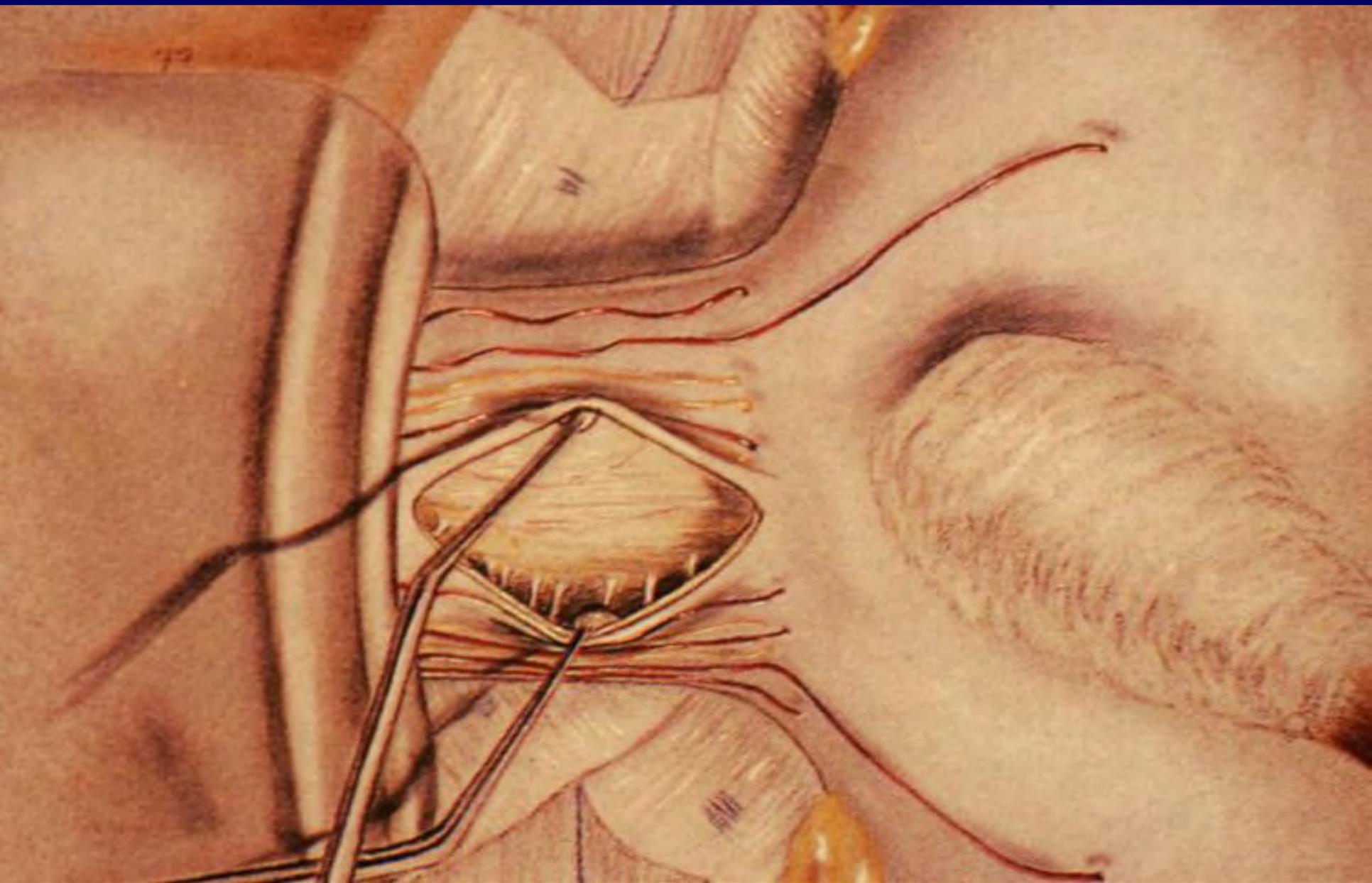
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Our Young Lady



- Optic Nerve Sheath Decompression both eyes
- So lessons learned -
 - Sometimes need a Shunt
 - Sometimes need Optic Nerve surgery
 - Rarely need both

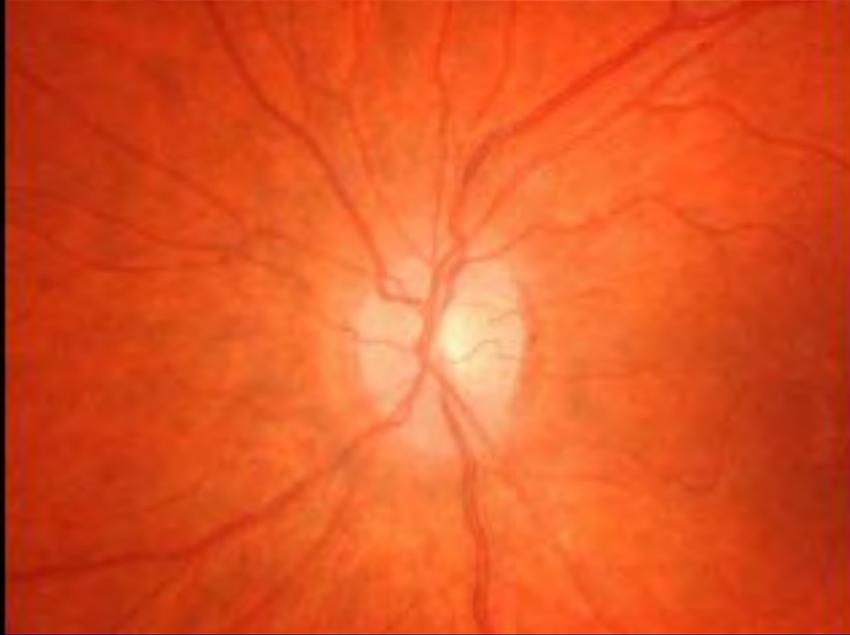
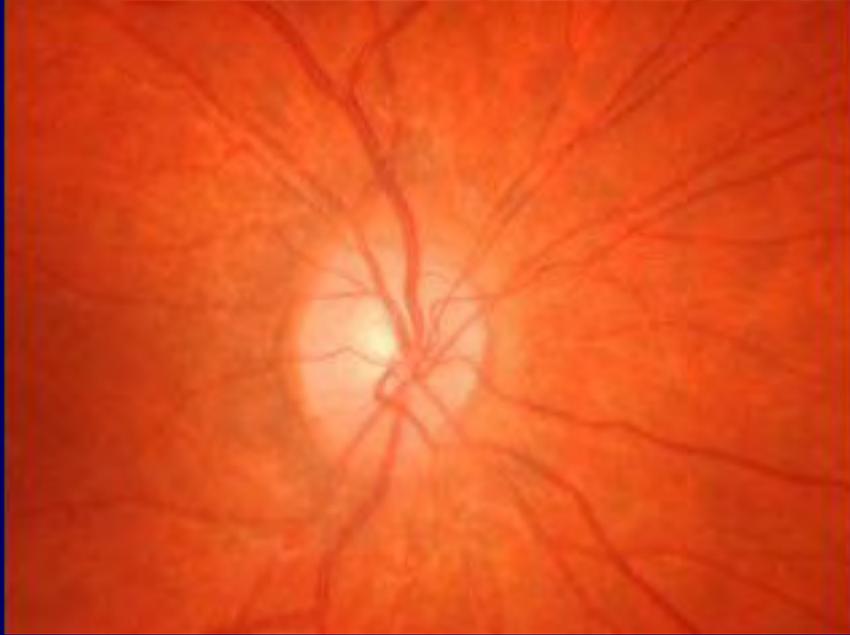




That's Our Girl



- Final result 20/30 and 20/25
- Marked resolution of Optic nerve swelling
- Visual Fields and Colors return
- HA gone



Pseudotumor Cerebri

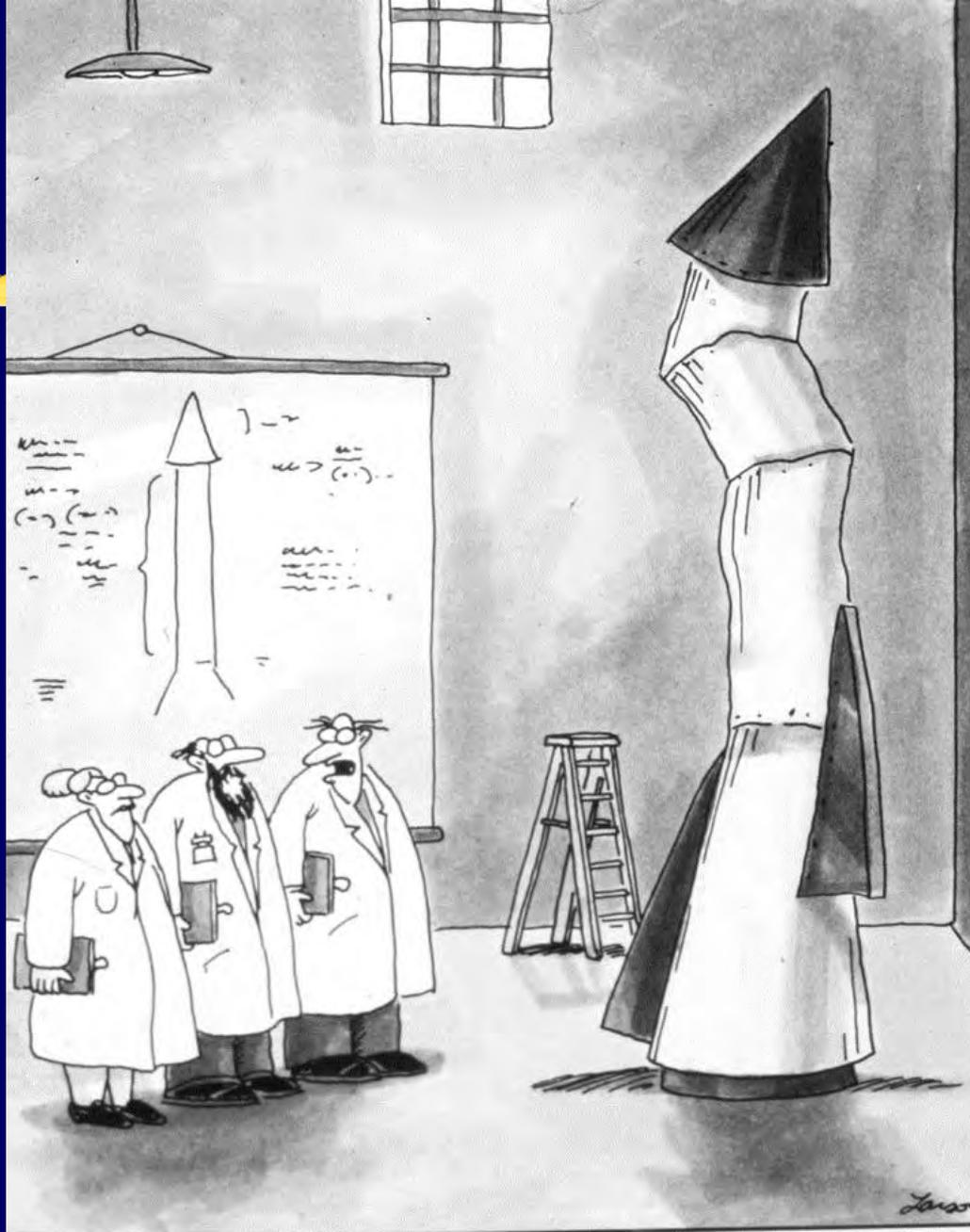


- Causes increased intracranial pressure
- All causes are ruled out except high CSF pressure
- Leads to papilledema (Optic Nerve swelling)
- Can lead to vision loss and double vision
- Optic nerve surgery is effective for visual loss
- May be required even with a shunt in place
- Often only one required; Sometimes both
- Team approach is best

Lastly

- All people with a history of :
 - Hydrocephalus
 - Shunt placement (past or present)
 - Elevated intracranial pressure – Any Cause
 - Symptoms thereof

*Must see an ophthalmologist
comfortable with these conditions
on a very regular basis*



"It's time we face reality, my friends. ...
We're not exactly rocket scientists."