PAPILLEDEMA

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Papilledema –
A Medical Emergency

LACK OF TIMELY / PROPER DIAGNOSIS AND WORK-UP OF PAPILLEDEMA IS A REASON FOR LAWSUITS AGAINST OPTOMETRISTS

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Symptoms of Increased Intra-Cranial Pressure

- Headache
- Nausea
- Vomiting
- Diplopia (Abduction deficit – CN VI)
- Pulsatile tinnitus
- Transient Visual Obscurations (TVOs)
  - Last few seconds (uni or bi-lateral)
  - Transient optic nerve ischemia

Features of Papilledema

Bilateral/Asymmetric (anatomic difference in lamina)

RARELY Unilateral

ICP (Intracranial pressure) greater than 200 - 250 mm H2O (20 – 25 cm H20)

Spontaneous Venous Pulsation

- Best viewed with a direct ophthalmoscope
- Presence of SVP means ICP normal (at that moment - can fluctuate)
- 10-20 % of normal patients may not have SVP
- **SVP will NOT be present in papilledema**

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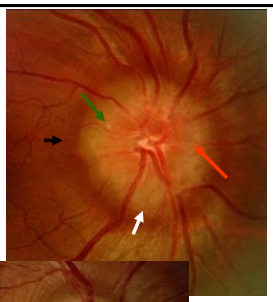
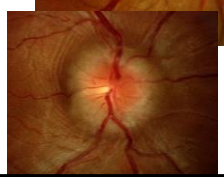
Features of edema

- Axoplasmic stasis in pre-laminar optic nerve
- Obscuration of retinal vessels coursing over the disc margin
- Paton's lines temporally
- Extruded axoplasm (in chronic papilledema)
- Can have hemorrhages





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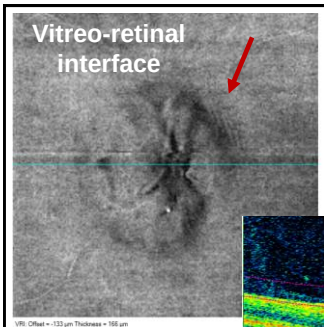
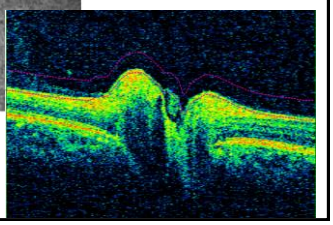


En face OCT can help with detection of subtle Paton lines



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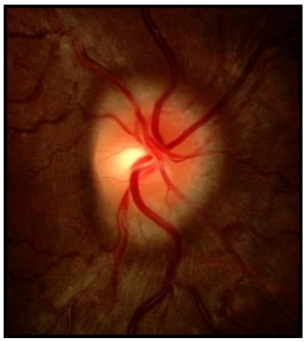
Pattern of Edema

- Corresponds with NFL thickness
- Superior, Inferior > Nasal > Temp
- Superior and Inferior NFL swell first
- Last to swell is Temporal NFL



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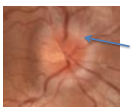
Modified Frisén Papilledema Scale

Grade 0

Grade 1

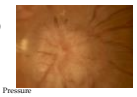
Grade 2

Grade 3



Grade 4

Grade 5



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The Modified Frisén Scale
Diagnosis and Grading of Papilledema in Patients with Raised Intracranial Pressure Using OCT Compared to Clinical Expert Assessment using a Clinical Staging Scale
Scott CJ, Kardon RH, Lee AG, Frisén L, Wall M. Arch Ophth (in press).

CAUSES OF PAPILLEDEMA

- Brain Tumor or Spinal Cord Tumor
- Venous Sinus Thrombosis
- Arteriovenous Malformation / Dural A-V Fistula
- Subdural or Subarachnoid Hemorrhage
- Meningitis
- Other Infectious / Inflammatory Etiology
- Idiopathic Intracranial Hypertension (aka Pseudotumor Cerebri) - *dx of exclusion*

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Papilledema Work-Up

- Neuro-Imaging
  - **MRI of brain with contrast and MRV**
  - **(less often MRA)**
- Lumbar Puncture (***only after imaging***)
  - Opening pressure
  - Analysis of CSF (r/o infection, meningitis, etc)

Normal values (CSF):
CSF opening pressure: 50–180 mmH$_2$O
Glucose: 40–65 mg/dL
Protein (total): 15–45 mg/dL
Lactate dehydrogenase: 1/10 of serum level.
Lactate: less than 35 mg/dL
Leukocytes (WBC): 0–5/μL (adults / children); up to 30/μL (newborns)
Gram stain: negative.
Culture: sterile.
Specific gravity: 1.006–1.009.
Syphilis serology: negative.
Gross appearance: Normal CSF is clear and colorless.
Differential: 60–70% lymphocytes; up to 30% monocytes and macrophages; other cells 2% or less.

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Why is Papilledema Considered a Medical Emergency?

1

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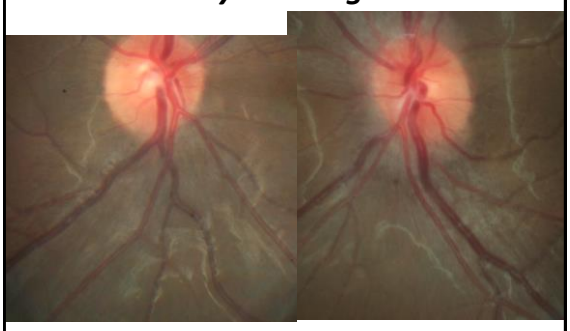
18 year-old girl



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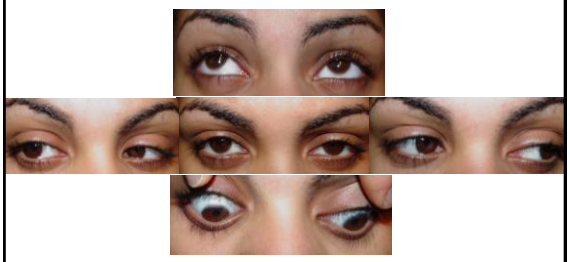
18 year-old girl



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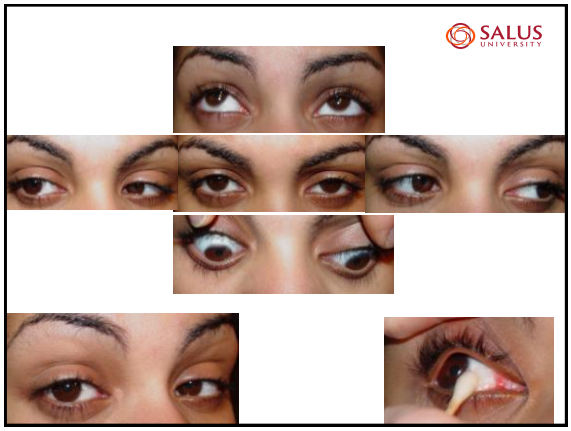

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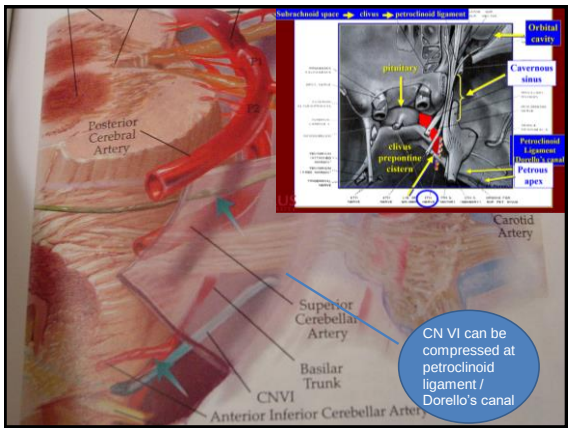

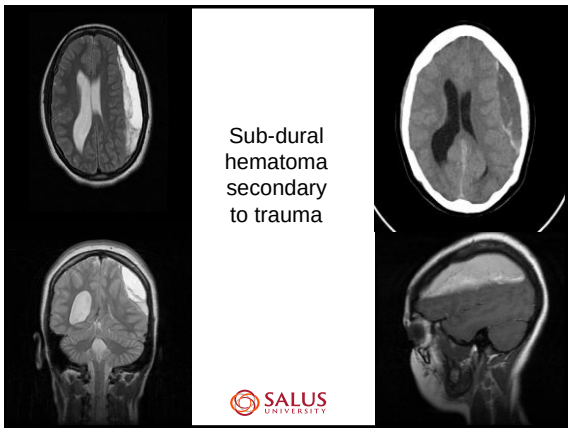

Sub-dural hematoma secondary to trauma

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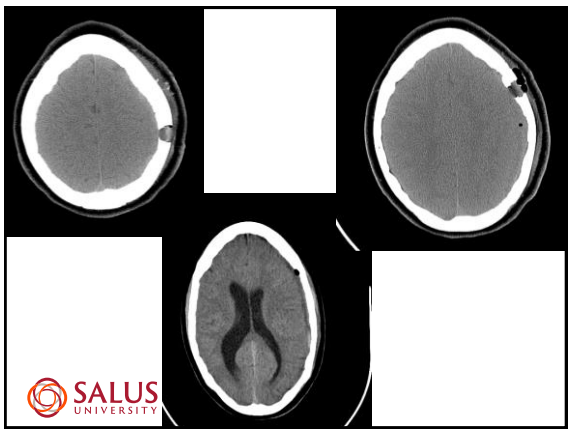

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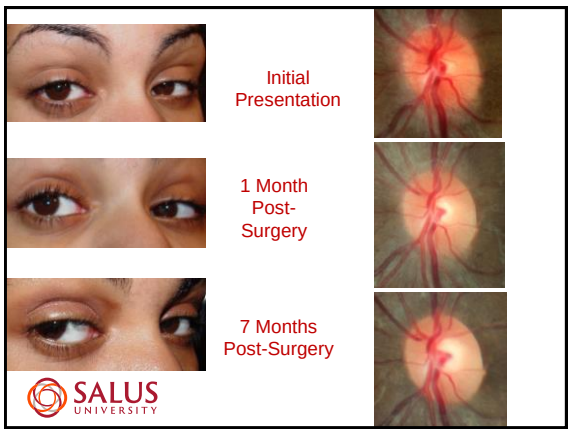

Initial Presentation

1 Month Post-Surgery

7 Months Post-Surgery

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Why is Papilledema Considered a Medical Emergency?
2

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63 year old mechanic

- c/o fatigue since hitting hear on car
- worst HA of life x 3 days
- episode of nausea / vomiting
- Pulsatile tinnitus
- All symptoms now improving
- Sys Hx: DM, HTN
- BCVA: OD 20/30 OS 20/30
- Color: 14/14 OD, 14/14 OS
- PERRL (-) RAPD

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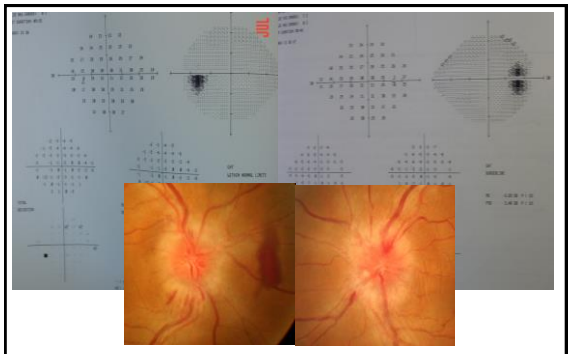

WHAT IS THE CONCERN?

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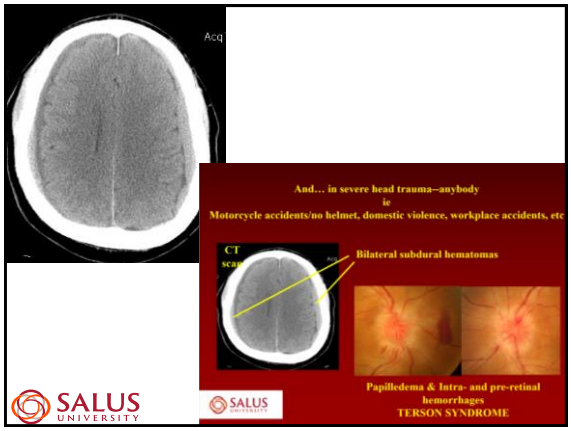
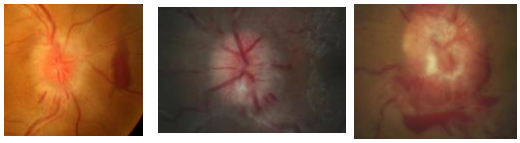


Hemorrhages In Papilledema

- Severe Papilledema (From any etiology, even IIH)
- Acute Increase in Intracranial Pressure
- Trauma
- Terson Syndrome
  - (Intracranial Hemorrhage + Intraocular Hemorrhage)



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Why is Papilledema Considered a Medical Emergency? 3

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20 year-old girl

- Visited an ER 5 x in last 2 months
- Was given separate medications for headache, vomiting, and neck pain/stiffness over last 2 months
- Now she feels that her vision is getting blurry, and she is seeing double

Remember to ask about neck stiffness. It is an important symptom of meningitis.

Meningitis can be:
Bacterial
Viral
Fungal
Parasitic
Carcinomatous
Other aseptic forms

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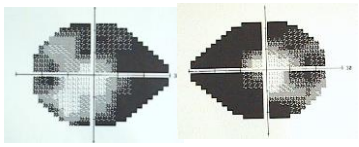
Examination Results

- VA: OD 20/25 OS 20/20
- Color (Ishihara): OD 9/14 OS 12/14
- Pupils: PERRL (+ 0.3-0.6 log) APD OD
- CF: nasal defects OU
- Normal anterior segment exam
- IOP: OD 18 mm Hg OS 18 mm Hg
- BP: 126/90



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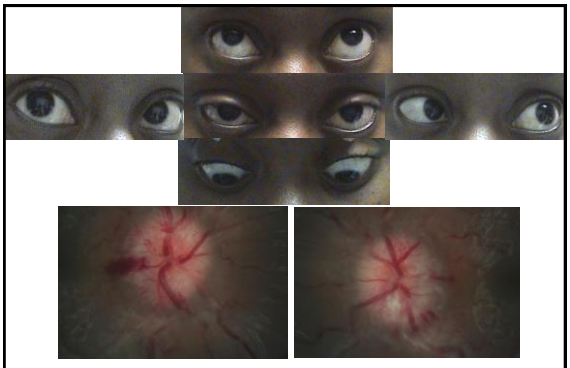

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Dx: Meningitis

Why is Papilledema Considered a Medical Emergency? 4

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31 year-old woman

- 5'2", 270lb
- routine eye exam
- blurry VA x 2 mos (attributed to old CLs)
- denies any eye / head pain, diplopia
- denies nausea, vomiting, pulsatile tinnitus, fever
- Sys hx: asthma, depression, sinusitis
- Meds: Effexor, Advair, recently d/c birth control med due to weight-gain

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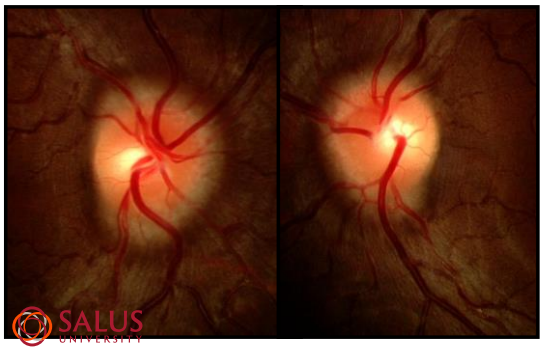
(-) SVP OU



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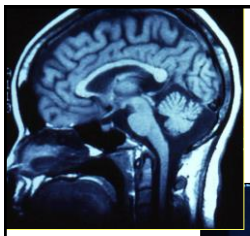
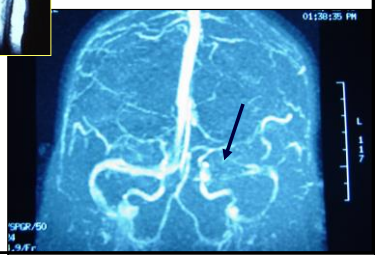


Venous Sinus Thrombosis



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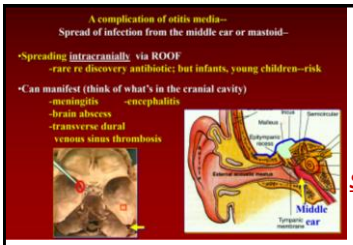

LATERAL SINUS THROMBOSIS

SEPTIC THROMBOSIS (infectious)

- MAY HAVE ASSOCIATED RETRO-AURICULAR EDEMA (GREISINGER'S SIGN)
- AUSCULTATE OVER MASTOID BONES

- CHRONIC OTITIS MEDIA
- SCALP INFECTIONS
- PHARYNGEAL ABSCESS
- INFECTION INVOLVING SINUSES, MASTOIDS, LEPTOMENINGES
- MALIGNANCIES

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LATERAL SINUS THROMBOSIS

ASEPTIC THROMBOSIS (non-infectious)

- ACUTE DEHYDRATION
- CYANOTIC CONGENITAL HEART DISEASE
- HYPERCOAGULABLE STATES & COAGULOPATHIES
- MALNUTRITION
- PREGNANCY / ORAL CONTRACEPTIVES
- TRAUMA

Factors that increase risk of clotting

- **Factors that increase clot risk**
  - Birth control
  - Tobacco use
  - Age >35
  - Hx of Migraine
  - Hypercoagulable states

Increased clot risk

WE NEED TO EDUCATE OUR PATIENTS ABOUT THIS RISK!

IMPORTANCE OF DFE IN WOMEN ON BIRTH CONTROL, REGARDLESS OF SYMPTOMS

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Causes of Papilledema

- Intracranial mass or hemorrhage (mass effect) ✓
- Venous Sinus Thrombosis (VST) ✓
- Meningitis ✓
- Dural AVM or AVF
- Spinal cord tumors

- Pseudotumor cerebri (PTC) or Idiopathic Intracranial Hypertension (IIH) – ***dx of exclusion (Dx when all testing is negative!)***

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PAPILLEDEMA IN MEN?

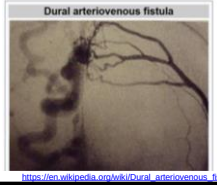
- If MRI normal
- Before considering PTC or IIH, get MRA
- DURAL ARTERIOVENOUS FISTULAS NEED TO BE RULED OUT

If your patient is a man, and you are thinking IIH, think again!!

IIH is RARE in MEN

LOOK FOR AN ALTERNATE CAUSE OF PAPILLEDEMA

GET CTA OR MRA TO LOOK FOR DURAL A-V FISTULA



https://en.wikipedia.org/wiki/Dural_arteriovenous_fistula

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What if the work-up is negative?

Idiopathic Intracranial Hypertension
or
Pseudotumor Cerebri

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Modified Dandy's Diagnostic Criteria (applies for adults)

***ALL* of these criteria must be met in order to diagnose IIH**

- Patient Must Be Awake & Alert
- Signs & Symptoms Of Increased Intracranial Pressure
- No Neurologic Signs Except CN VI Paresis
- Normal Neuro-Imaging (MRI, MRV)
  - *MUST be done PRIOR to lumbar puncture*
- CSF Opening Pressure > 200mm to 250 mm H20 & Normal Composition of CSF

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What if the work-up is negative?

Idiopathic Intracranial Hypertension
or
Pseudotumor Cerebri
1

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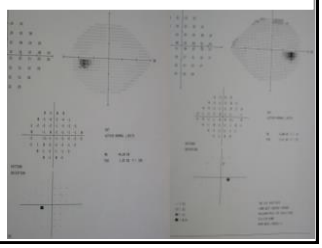
50 year-old woman

- headaches, transient visual obscurations
- VA OD 20/20 OS 20/20
- PERRLA (-) APD
- color: 14/14 OD, 14/14 OS
- CF: full OU



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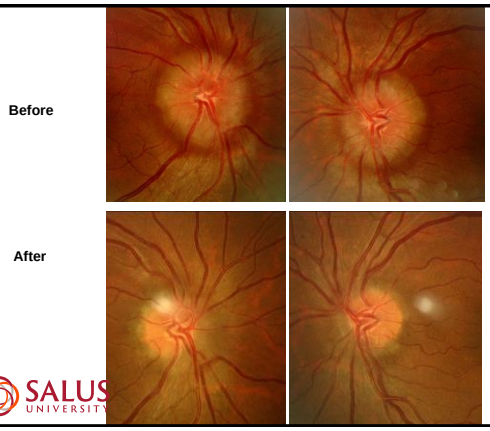
Work-up

- MRI performed – normal
- MRV performed – normal
- LP: 320 mm H2O opening pressure
- with normal contents

- Treatment:
  - Diamox
  - Weight loss (successfully!)

IIH TREATMENT

- WEIGHT LOSS (at least 6-10% of body weight)
  - Diet / healthy eating habits, exercise
- CARBONIC ANHYDRASE INHIBITORS
  - acetazolamide (Diamox); usual starting dose of 1000 mg
  - reduces CSF by 50% but may be unsustained!
  - Contraindicated in renal disease



IF AFTER LP…

- Patient complains of worsening headache
- Headache is worse when upright

- Why is this? What does this mean?

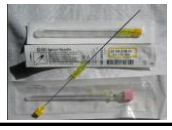
Lumbar Puncture

- May be difficult to get a successful traditional (in fetal position) LP due to patients weight/size
  - May need to be done under fluoroscopy
    - Lying on stomach
- Results of OP can vary depending on position of patient

BENEFITS OF DOING IMAGE GUIDED (FLUOROSCOPIC) LP IN OBESE PATIENTS

- Successful 1st tap
- Avoid multiple taps
- Less chance of complications, such as CSF leak and spinal headache

LP UNDER FLUOROSCOPY



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POST LP INSTRUCTIONS

- **For the first 24 hours after the test:**
- Remain flat on your back as much as possible.
- Drink at least 2 1/2 quarts of fluid.
- Avoid heavy exercise or heavy lifting for 24 hours after the procedure.
- Notify your doctor if you have any persistent drainage from the puncture site, or headache.

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SPINAL HEADACHE

- The spinal headache often is described as "a headache like no other."
- Spinal headaches are much more severe when the person is in an upright position.
- They improve when the person lies down.

- We need to recognize the symptoms
- Need to refer the patient back to the physician who did the LP
- May be in need of a **blood patch**

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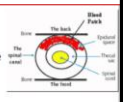
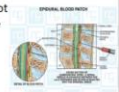
BLOOD PATCH

- An epidural **blood patch** is a surgical procedure that uses autologous **blood** in order to close one or many holes in the dura mater of the spinal cord, usually as a result of a previous lumbar puncture.
- Can be used to relieve post dural puncture headaches caused by lumbar puncture.

BLOOD PATCH

- An epidural needle is inserted into the epidural space at the site of the cerebrospinal fluid leak and blood is injected.
- The clotting factors of the blood close the hole in the dura.
- As such, the autologous blood does not "repair" the leak, but rather treats the patient's symptomology.





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What if the work-up is negative?

Idiopathic Intracranial Hypertension
or
Pseudotumor Cerebri
2

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36 YEAR OLD WOMAN

- Headaches and episodes of transient vision loss
- Ultimately went to ER and was found to have disc edema
- Weight was 293 lbs
- Had MRI and MRV, which were unremarkable
- Had LP, which found an OP of >30cmH2O
- She was prescribed Diamox
  - She felt sick when taking Diamox and discontinued it

- Systemic Health
  - Hypertension and Sleep Apnea
- Medications
  - Depo Provera
- Family History
  - Mother had IIH, and subsequently lost a lot of weight

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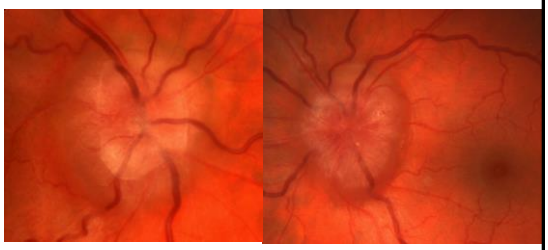
Initial Presentation



VA: 20/20 OD and OS
0.6 log RAPD OD

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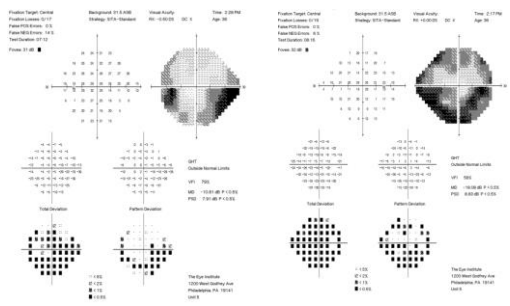
Initial Presentation



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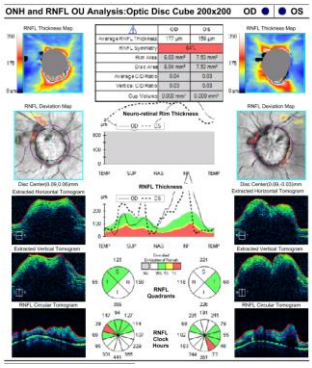
Initial Presentation



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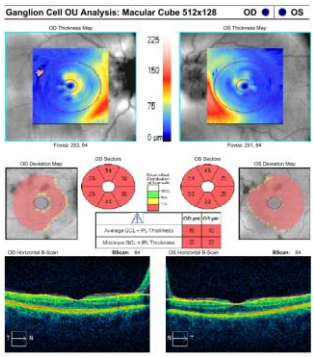

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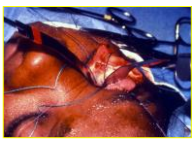
- Educated on need for weight loss
- Asked patient to try Diamox again
- Scheduled patient with neurosurgery for consideration of VP shunt

- Since her headache was improved, and she was mainly complaining of vision, she first had an Optic Nerve Sheath Fenestration (ONSF) OD



- Due to persistent symptoms, including transient visual obscurations, she then had a VP shunt on right side

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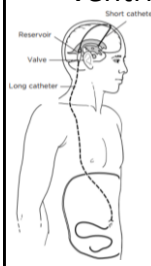
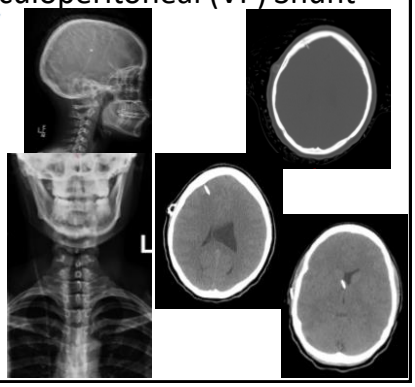
Ventriculoperitoneal (VP) Shunt





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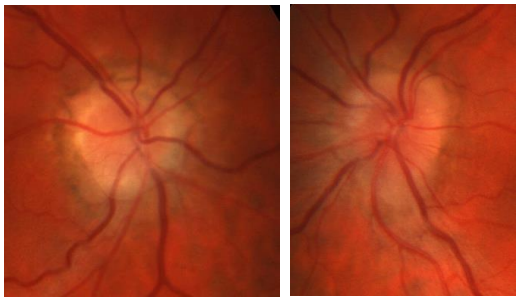
s/p ONSF and 1 wk s/p first VP shunt



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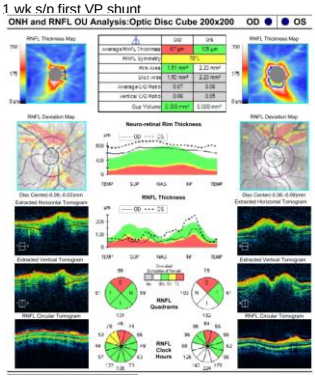
s/p ONSF and 1 wk s/p first VP shunt



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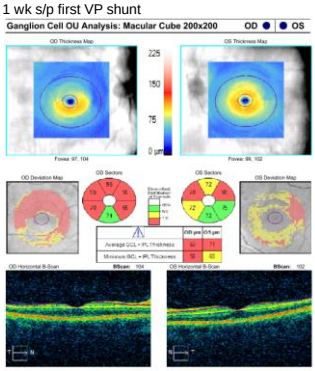
s/p ONSF and 1 wk s/p first VP shunt



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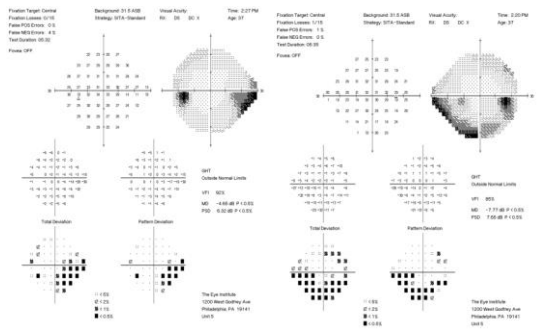
s/p ONSF and 1 wk s/p first VP shunt



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- She developed an infection in the shunt, and it needed to be removed.
- Once the infection was treated, she then underwent a second LP shunt placement on the left side

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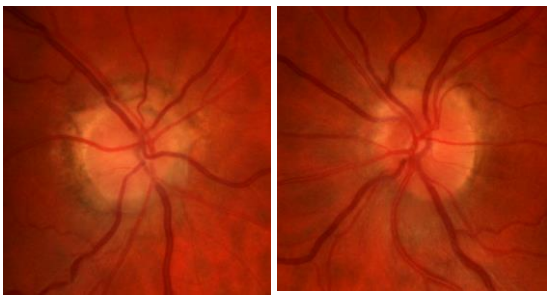
s/p ONSF and 1 wk s/p Second VP shunt



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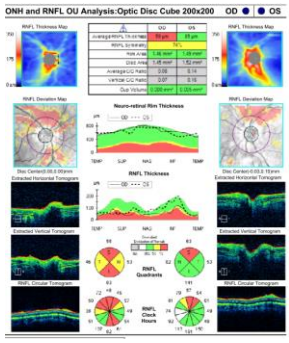
s/p ONSF and 1 wk s/p Second VP shunt



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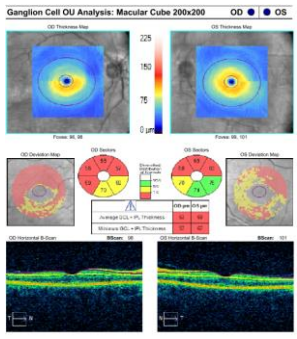
s/p ONSF and 1 wk s/p Second VP shunt



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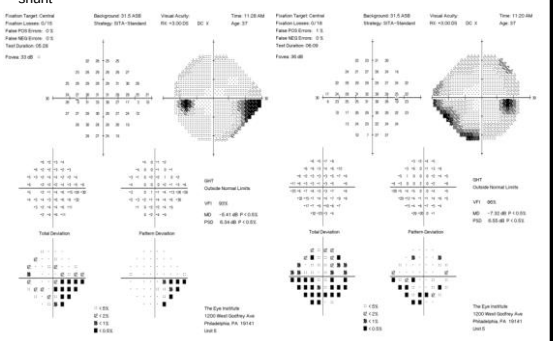
s/p ONSF and 1 wk s/p Second VP shunt



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What if the woman is NOT significantly overweight?

Can it still be IIH?

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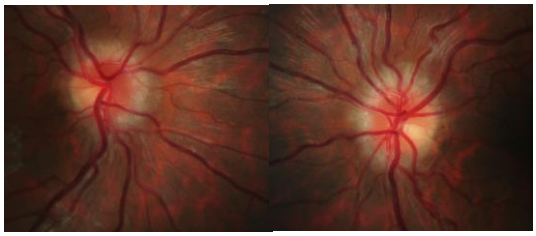
SLENDER 25 YO WOMAN



MRI and MRV were normal!

LP showed normal CSF analysis and an elevated opening pressure…

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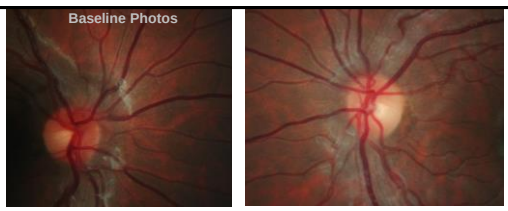

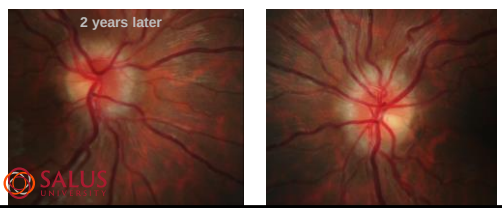

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Should we be getting baseline ONH photos /OCTs on all new patients?

Then we could more easily assess for interval change

Can't rely on chart documentation to assess for change

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74

All the previous cases were adults…

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IIH / PTC is possible in children, BOTH male and female

But, children can also have pathology as the causative etiology of papilledema

PAPILLEDEMA IS A MEDICAL EMERGENCY AT ANY AGE!!!!

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Why is Papilledema Considered a Medical Emergency in Children?
1

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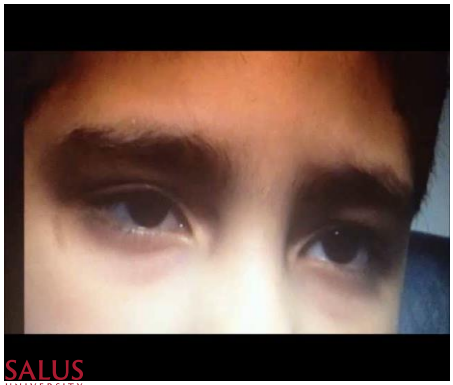

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GAZE EVOKED NYSTAGMUS:

	PHYSIOLOGIC	PATHOLOGIC
•SYMPTOMS:	NO	YES
•PRIMARY:	NO	YES
•SYMMETRY:	YES	NO ←
•30/45 DEG:	NO	YES
•REBOUND:	NO	YES

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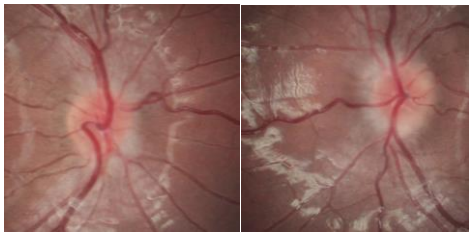
10 year-old boy with nystagmus



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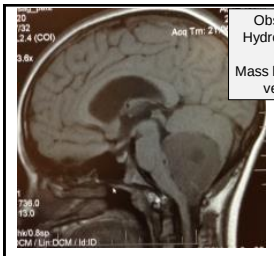
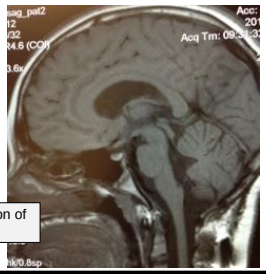


Obstructive Hydrocephalus

Mass blocking 4th ventricle



s/p resection of mass

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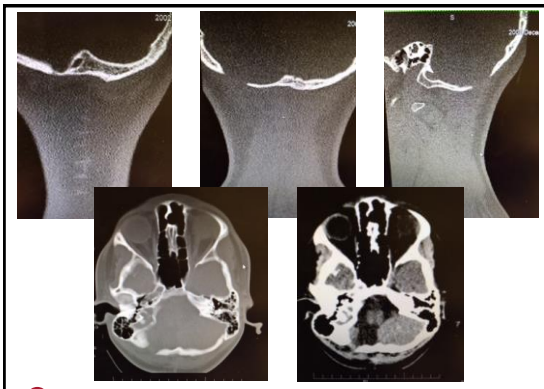

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Why is Papilledema Considered a Medical Emergency in Children? 2

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5 year-old girl

- Had routine eye exam 3 months ago, but was unable to stay for dilation.
- She is just here for the DFE
  - She has been getting occasional frontal headaches
  - They have not affected her daily activities
  - Already mentioned it to her pediatrician
  - Normally complains of headaches when at school (1x/month)
  - Only complained of headache once on a weekend, but she was in the mall and hungry

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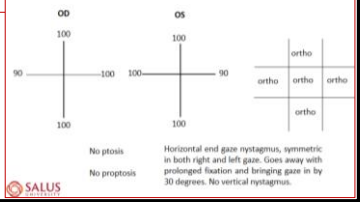
Afferent Exam Findings

- VA: OD 20/20 -
  OS 20/20-
- Pupils isocoric, (-)RAPD
- CF: full to finger count and red targets OU
- Color: OD 13/14 OS 13/14
- Unable to do HVF

Efferent Exam Findings



No ptosis

No proptosis

Horizontal end gaze nystagmus, symmetric in both right and left gaze. Goes away with prolonged fixation and bringing gaze in by 30 degrees. No vertical nystagmus.

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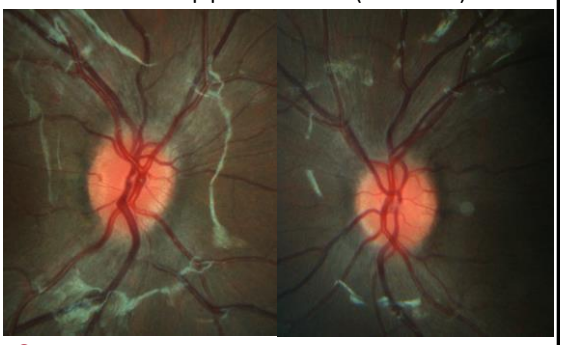
ONH Appearance (no SVP)



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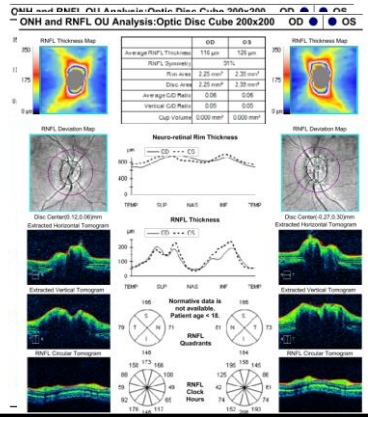
ONH OCT

There are no age-matched norms for children.



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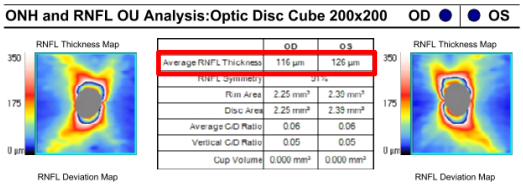

Average RNFL Thickness
OD 116um, OS 126um

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Yanni et al Am J Ophthalmol. 2013 February ; 155(2): 354–360

- 83 healthy North American **children aged 5-15**
  - attempt to determine normative data using the Spectralis SDOCT.
- Taking the 5th to 95th percentile data from these children as representing normal values
  - **mean peripapillary RNFL thickness 107.6um.**
- this is significantly higher than normative data for adults
- This difference is explained by the fact that even healthy individuals experience RNFL thinning with age.

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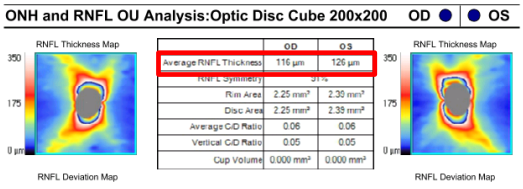

Average RNFL Thickness
OD 116um, OS 126um

Normal mean peripapillary RNFL thickness 107.6um

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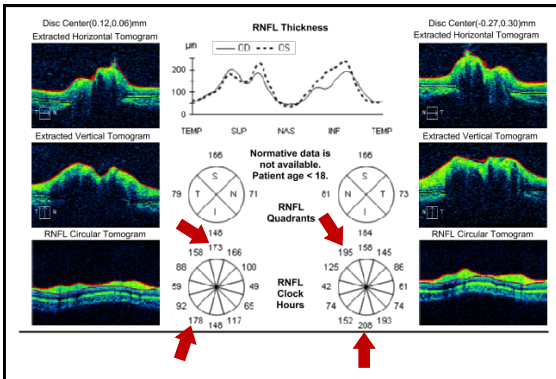

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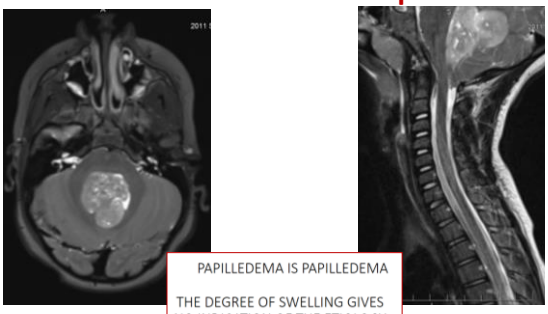
Inferior & superior nerve fibers swell first

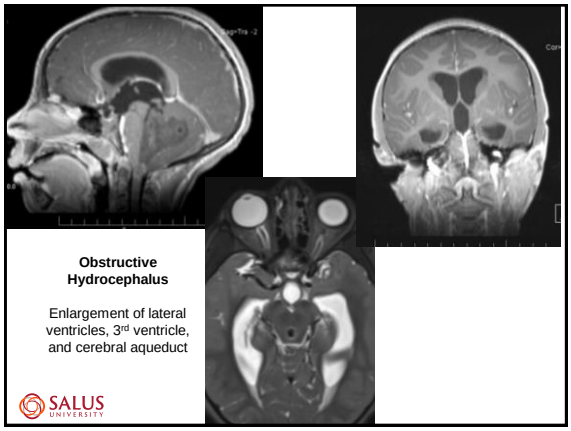
Medulloblastoma with Drop Mets



PAPILLEDEMA IS PAPILLEDEMA

THE DEGREE OF SWELLING GIVES NO INDICATION OF THE ETIOLOGY

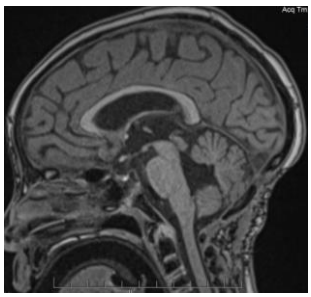
SO, EVEN MILD PAPILLEDEMA IS A MEDICAL EMERGENCY



Obstructive Hydrocephalus

Enlargement of lateral ventricles, 3rd ventricle, and cerebral aqueduct

5 year old: One year post-surgery, radiation and chemo



Why is Papilledema Considered a Medical Emergency in Children?

3

6 Year Old Boy

CHIEF COMPLAINT

History of headaches and lethargy

Vision seems to be fine

PAST OCULAR HISTORY

His last eye exam was 1 year ago, and was unremarkable

BCVA: 20/20 OD, 20/20 OS

Color Vision: (Ishihara) 14/14 plates OD & 14/14 OS

Pupils isocoric, (-) RAPD

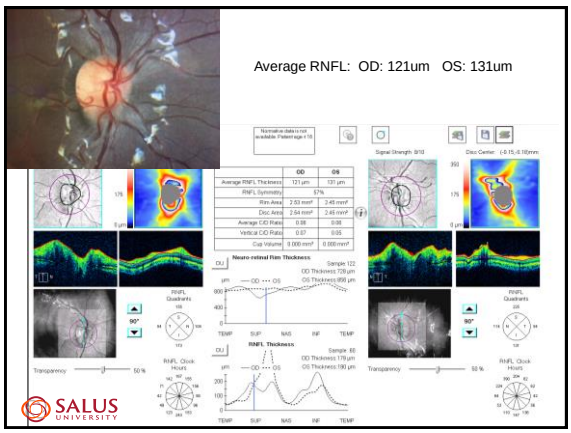
CF: OD full to finger count, no red desaturation

OS full to finger count, no red desaturation

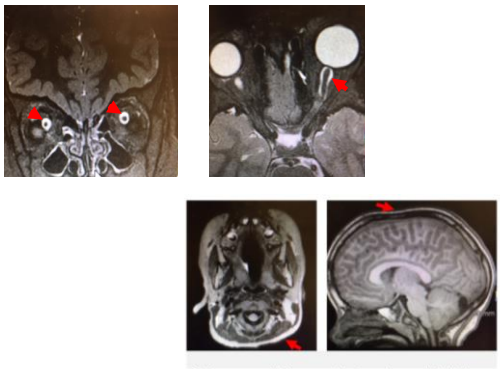
No ptosis or proptosis

No ductional limitations, no abduction deficit

Cover testing: comitant small angle exophoria



Average RNFL: OD: 121um OS: 131um



Red arrows point to areas showing abnormal skull shape due to early closure of multiple cranial sutures.

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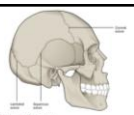
Craniosynostosis



- Early closure of a skull suture.
- May occur with just one, or multiple, sutures.
- Early closure results in lack of ability of brain and skull expansion during infancy and childhood. Head or facial deformity is often seen, but not always present.
- Premature suture closure can result in increased intracranial pressure, requiring surgical skull vault expansion.

Craniosynostosis

- Should be considered in cases of papilledema in children
  - Craniosynostosis has been documented as the cause of papilledema in 18.4% of children aged 3-13.
- Traditional neuro-imaging may not detect craniosynostosis.
  - Head CT with 3-D reconstruction is the preferred imaging method to make the diagnosis.

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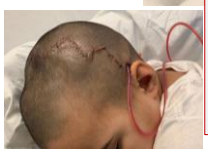
Craniosynostosis

Craniosynostosis is often diagnosed and treated in infancy.
However, delayed presentations are possible, and can manifest as:

- debilitating headaches
- developmental delays
- head shape abnormalities
- Chiari malformation
- papilledema



TREATMENT: skull vault expansion surgery

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What if the work-up is negative?

Idiopathic Intracranial Hypertension or Pseudotumor Cerebri in Children

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PEDIATRIC PSEUDOTUMOR CEREBRI EPIDEMIOLOGY

- = GENDER RATIO
- OBESITY NOT AS GREAT A FACTOR
- INSTEAD OF HA
  - IRRITABLE, APATHETIC, SOMNOLENT
- HIGHER PREVELANCE OF CNVI
- CNIII, CNIV, LMN VII, SKEW
- DANDY'S DOES NOT APPLY

PEDIATRIC PSEUDOTUMOR CEREBRI: RELATIONSHIP OF AGE AND OBESITY
(Balcer et al 1998 NANOS)

	3-11 yrs	12-14 yrs	15-17 yrs
OBESE	3 (33%)	8 (89%)	6 (86%)
NON-OBESE	6 (67%)	1 (11%)	1 (14%)
Total	9	9	7

Regardless of Patient Age

IIH /PTC IS ALWAYS A DIAGNOSIS OF EXCLUSION!

PAPILLEDEMA IS ALWAYS A MEDICAL EMERGENCY!

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THANK YOU.
ANY QUESTIONS?
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