PSEUDOTUMOR CEREBRI

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WCUCOM

MECHANISM

- Technically Unknown
- Thought to be changes in cerebral venous outflow
- Obesity causing increased intrabdominal pressure
- Sleep Apnea
  - Hypercarbia increasing ICP via Vasodilation

EPIDEMIOLOGY

- Incidence 1-2 per 100,000
- Females>Male
- Obese>health BMI

RISK FACTORS

- Growth hormone therapy
- Tetracyclines
- Hypervitaminosis A
- Anemia
- Sleep Apnea
- Autoimmune (SLE, Addison's)
- PCOS
- Coagulation disorder

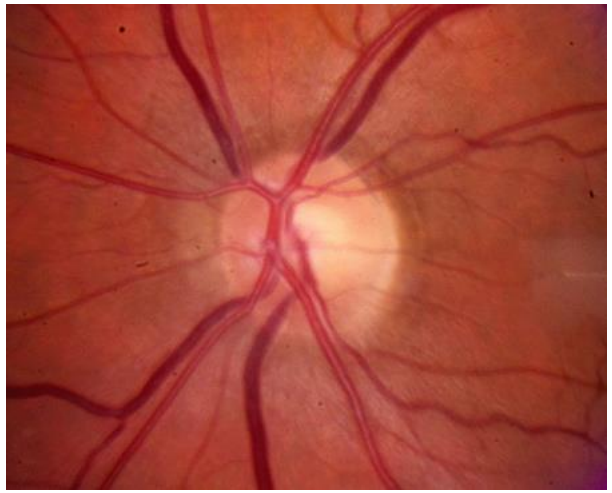
CLINICAL FEATURES

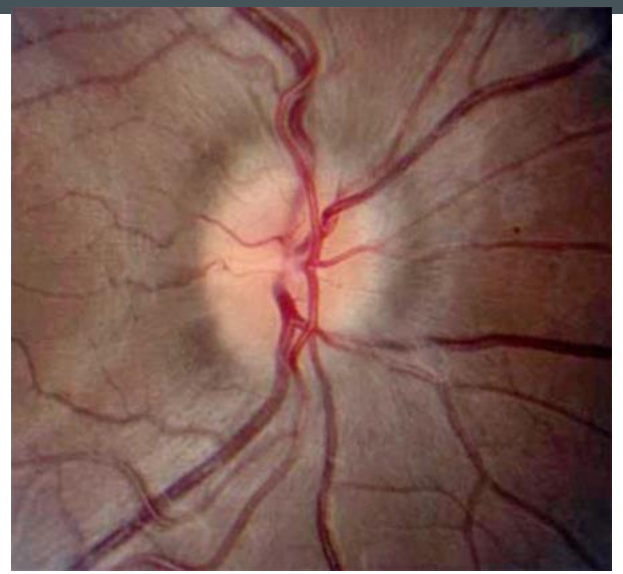
- Pulsatile tinnitus
- Headache (throbbing/pulsatile, but severe)
- Diplopia
- Sustained Vision loss
- Retrobulbar Pain (pain with eye movement)
- Photopsia
- Backpain
- Neck Pain

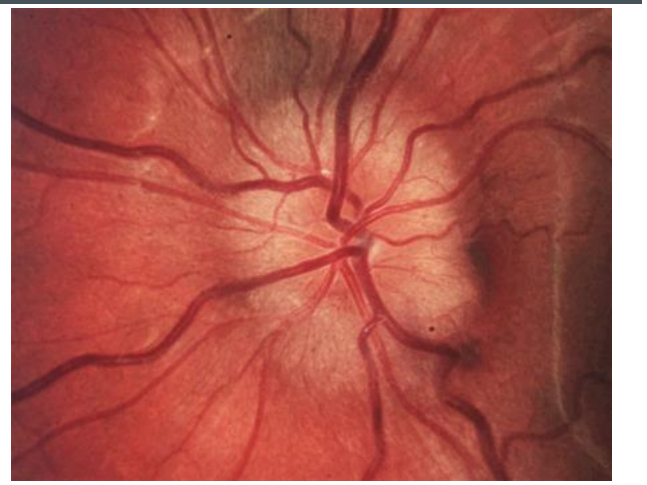
PHYSICAL EXAM

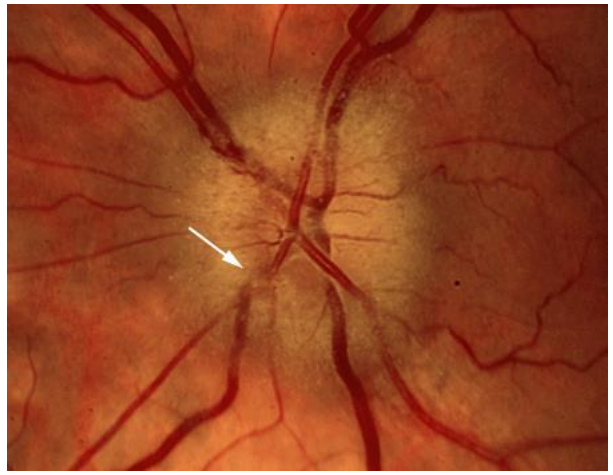
- Papilledema
- Visual field loss
- CN VI palsy

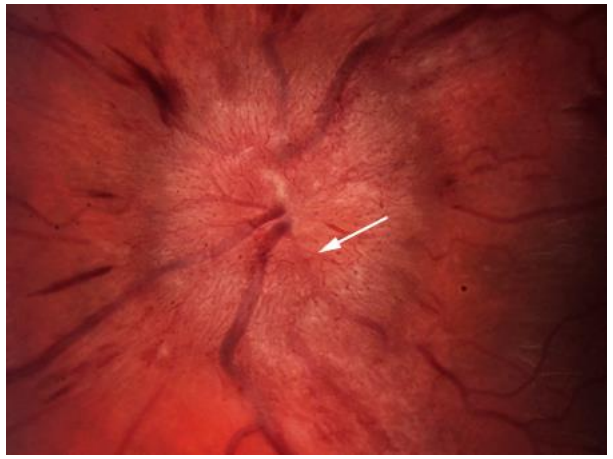
PHOTOS OF PAPILLEDEMA (**FRISÉN GRADING SYSTEM**)

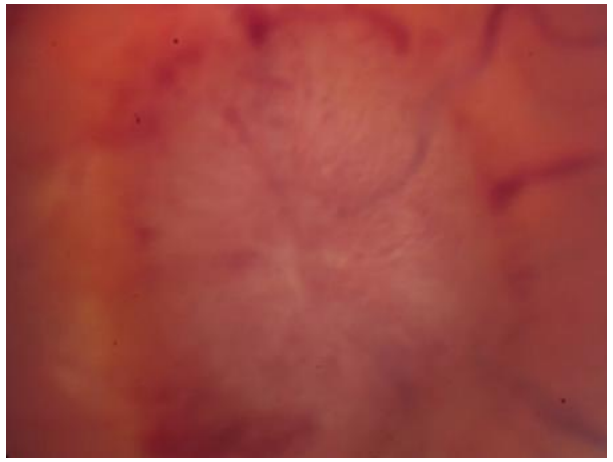




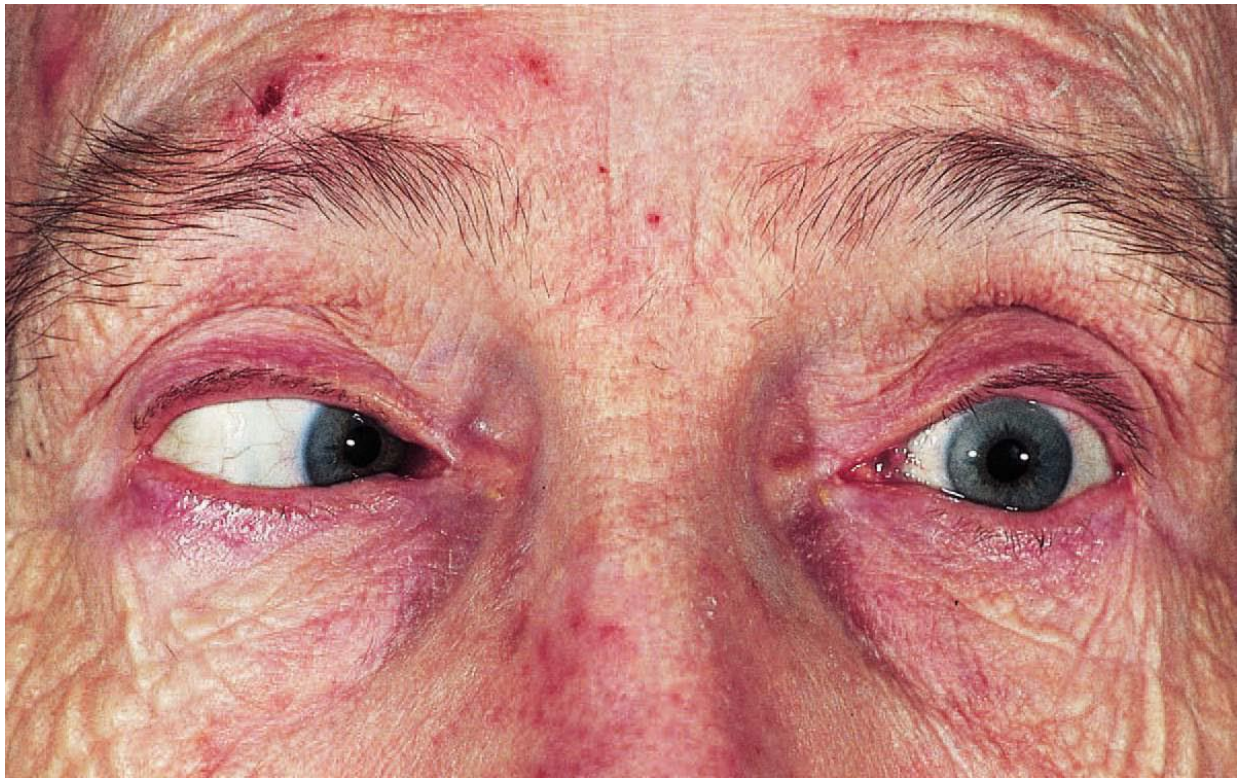








CN VI PALSY PHOTO

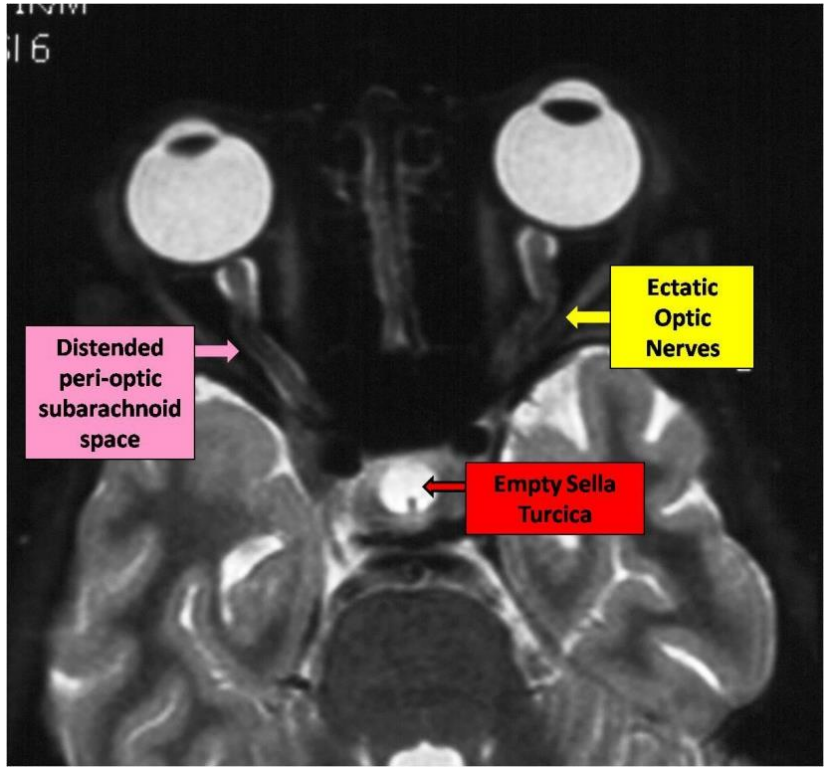


EVALUATION

- Urgent Imaging (MRI)
- Ophthalmic evaluation (Orbital US, Fluorescein angiography, Visual field testing)
- Lumbar puncture
- BP check

WHAT WE SEE ON MRI

- Distension/enhancement of peri-optic subarachnoid space
- Empty Stella
- Intraocular protrusion of pro-laminar optic nerve
- Tortuosity of the orbital optic nerve
- Saccular dilatation of the optic nerve sheath



DIFFERENTIAL

- Mass/Abscess
- Venous outflow obstruction
- Obstructive hydrocephalus
- Decreased CSF absorption (scarring from meningitis/granulations in arachnoids space)
- Increased CSF production due to Choroid Plexus Tumor

DIAGNOSTIC CRITERIA

Diagnostic criteria

- Papilledema must be present
- Normal neurological examination except for cranial nerve abnormalities
- Neuroimaging: normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
- Normal CSF composition
- Elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF if the child is not sedated and not obese]) in a properly performed lumbar puncture
- If papilledema is not present, then there must either be a CN6 palsy (unilateral or bilateral) or ≥ 3 neuroimaging criteria satisfied:
- Empty sella
- Flattening of the posterior aspect of the globe
- Distention of the peri0optic subarachnoid space with or without a tortuous optic nerve
- Transverse venous sinus stenosis

TREATMENT

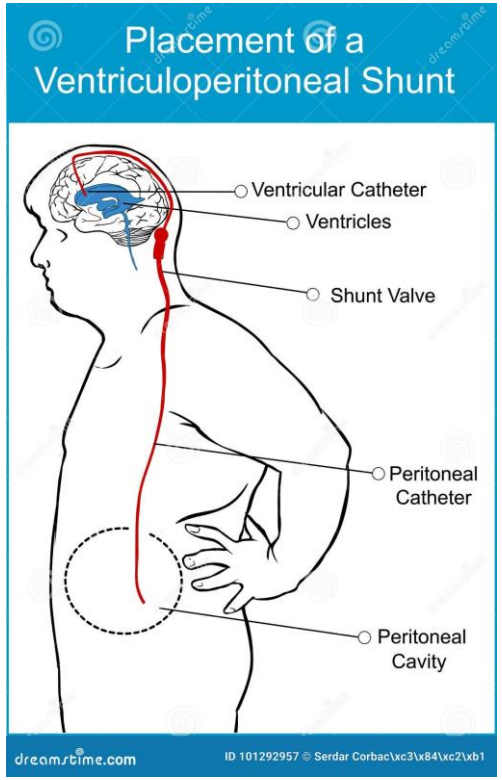
Mild

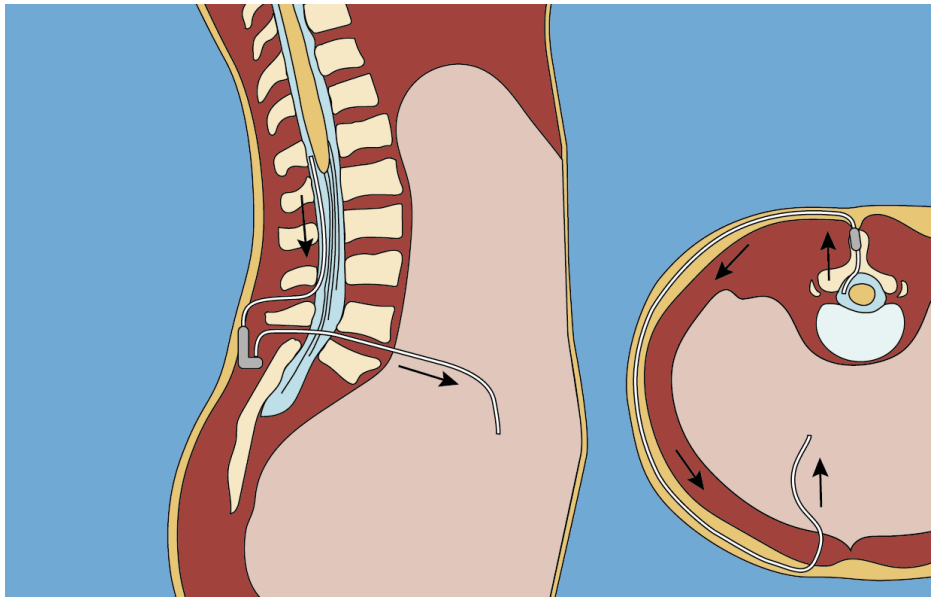
- Acetazolamide 500mg BID
  - Can increase to 2-4g/Day
  - Kids 100mg/kg or 2g/day
- Weight lose + low Na diet
  - 6% lose recommended
- Check for Sleep apnea
- Furosemide
  - Adults 20-40mg/day
  - Kids 1-2mg/kg

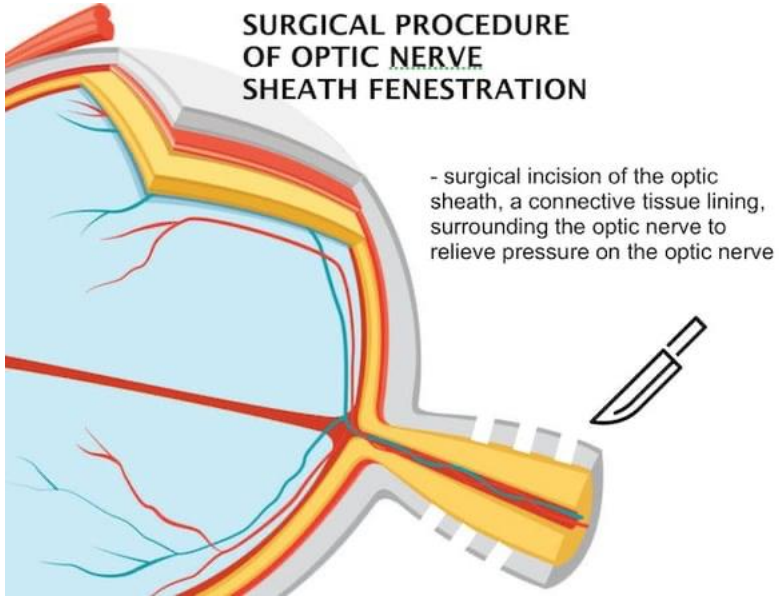
Urgent

- Acetazolamide titrated to 4g/day rapidly
- Glucocorticoid therapy
- Serial lumbar Punctures
- Neurosurgical eval
  - Ventricular peritoneal shunt
  - Lumbar peritoneal shunt
  - Optic nerve sheet fenestration

SURGERY PICTURES







FOLLOW UP

- Frisen Grade 2 Papilledema: 2-6 months
- Frisen Grade 1 Papilledema: yearly

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