

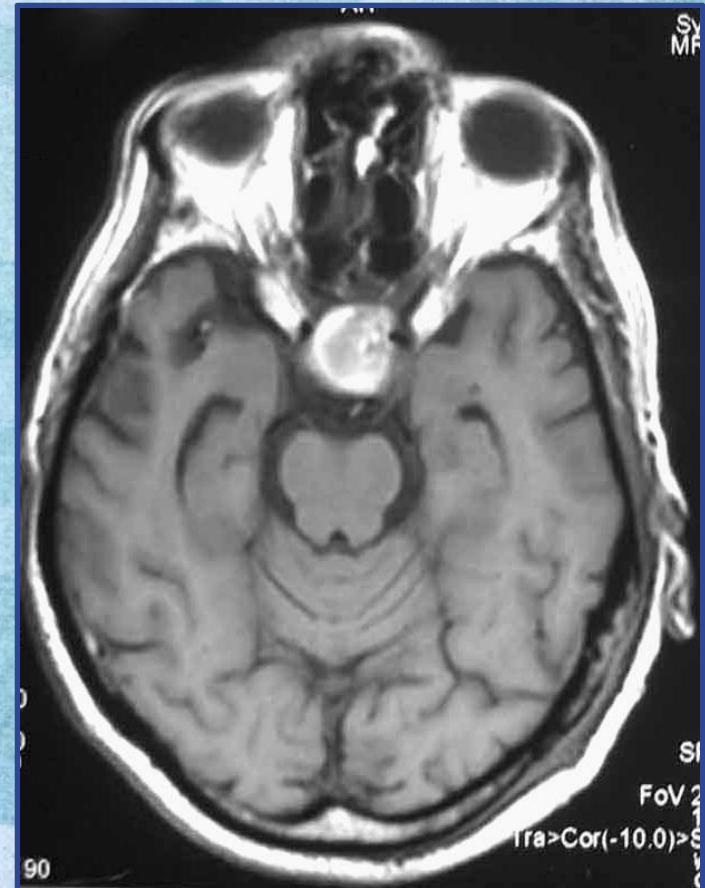
Everything You Always Wanted to Know About **PITUITARY ADENOMA**

But Were Afraid to Ask!

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Chiasmal Syndrome

- Online notes
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- Disclosures
 - None



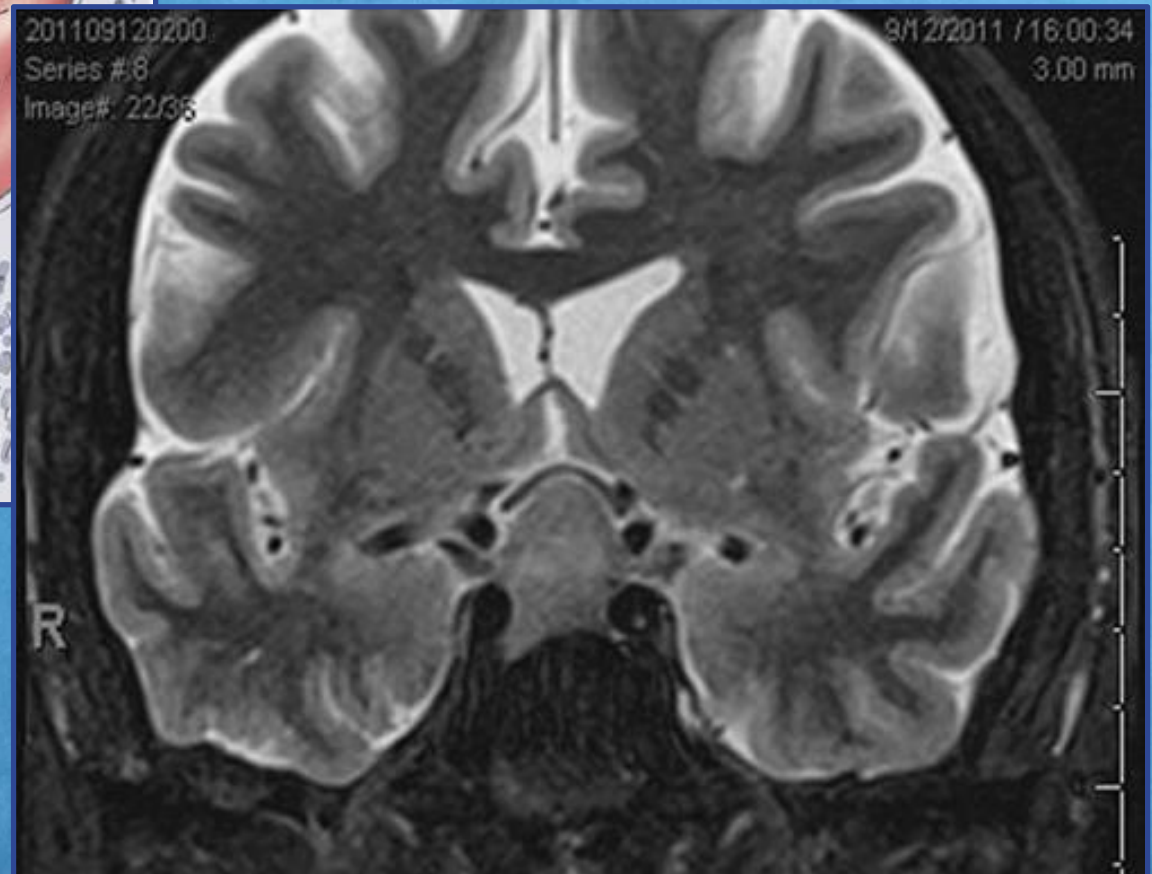
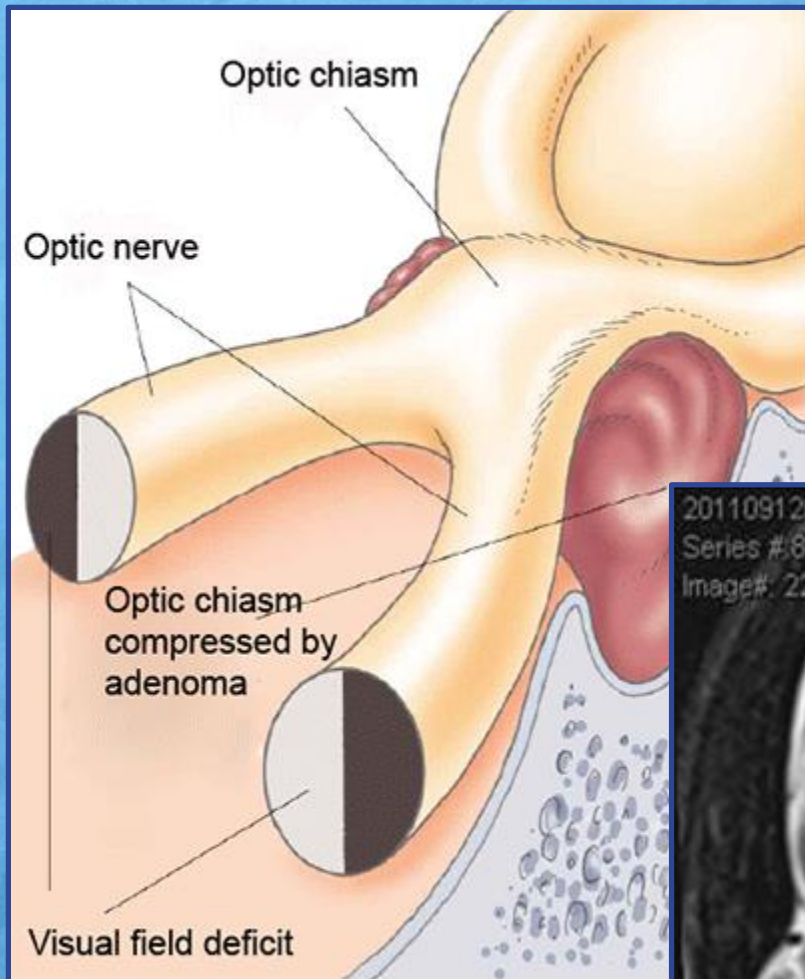
Chiasmal Syndrome

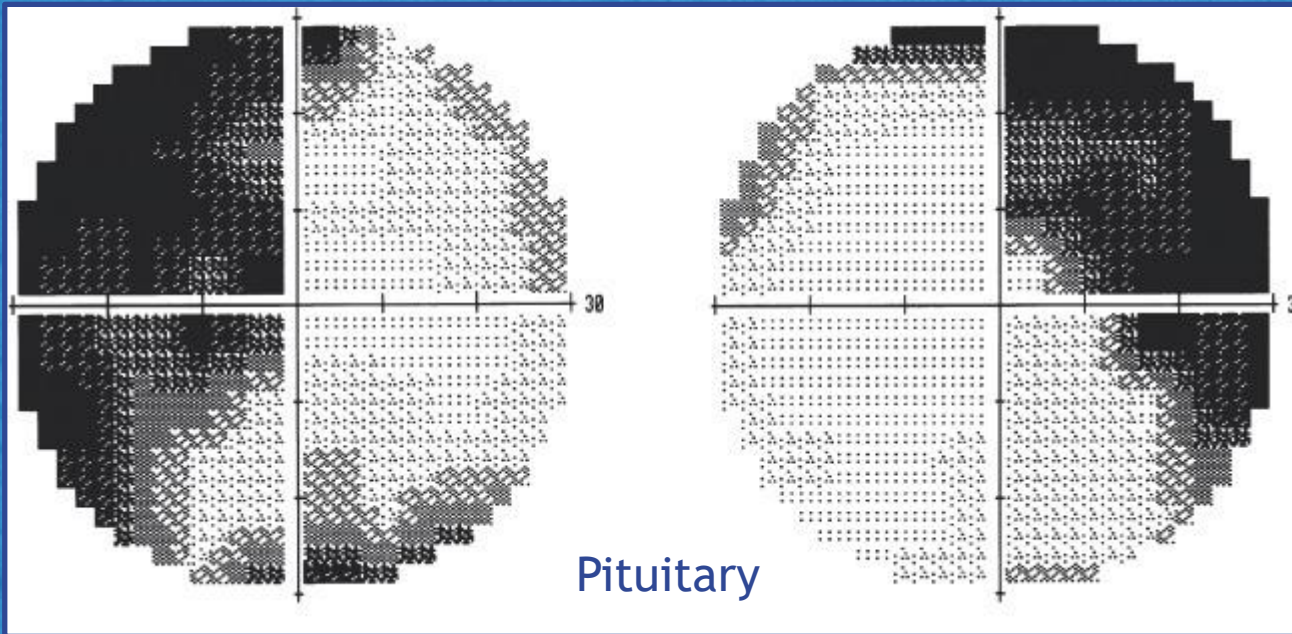
- Chiasmal syndrome is the constellation of signs and symptoms associated with lesions of the optic chiasm
 - Pituitary adenoma is the most common cause
- **25% of all brain tumors occur in this region**
 - 50% are pituitary adenomas
 - *Visual disturbance is common*
- Patients with chiasmal lesions may present c/o headache and/or visual disturbances

Chiasmal Syndrome

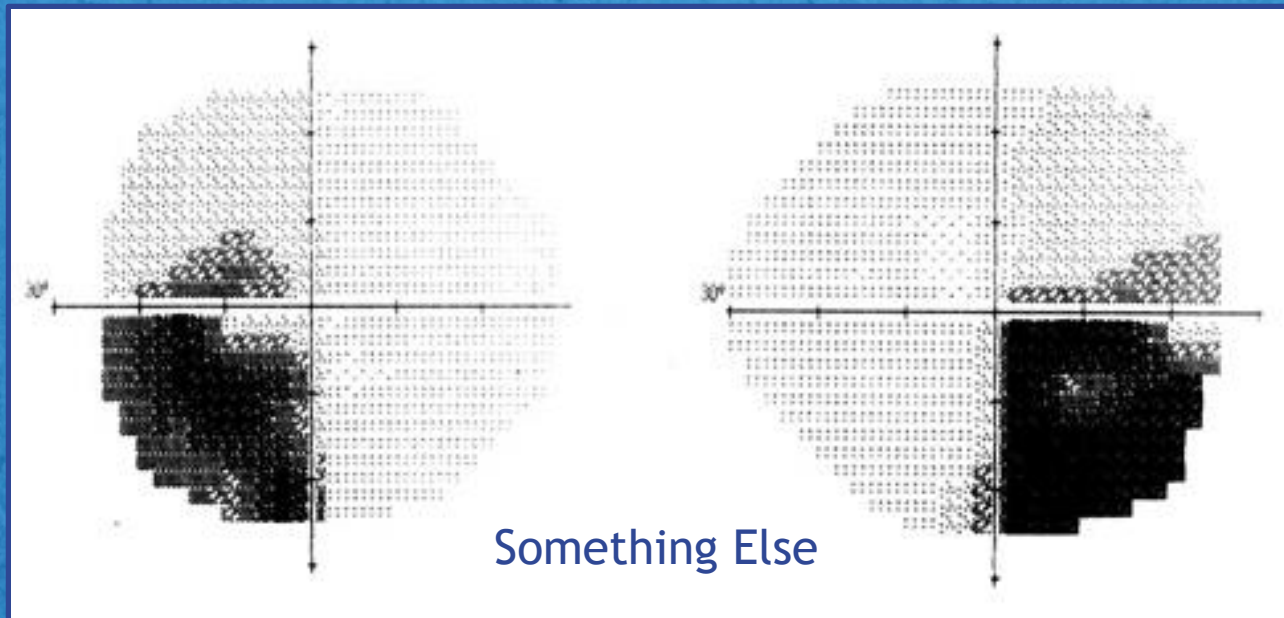
- Causes of chiasmal syndrome include tumor, inflammation, and ischemia
- Findings suggestive of an etiology other than pituitary adenoma:
 - **Visual sx**s (blur or difficulties with side vision)
 - Younger age
 - Unilateral optic disk pallor
 - RAPD
 - A complete hemianopic VF defect
 - **VF defect greater inferiorly than superiorly**

Pituitary adenoma compresses the optic chiasm from below





Incomplete
bitemporal
hemianopic defect
greater above
than below - highly
suggestive of
pituitary adenoma



Incomplete
bitemporal
hemianopia
greater below
than above - highly
suggestive of
something other
than pituitary
adenoma

Chiasmal Syndrome

- Anatomy Review
- All About Pituitary Adenomas
- Clinical Features of Chiasmal Syndrome
- Clinical Pearls
 - Red Flag Warning Signs
 - Chiasmal Work-up

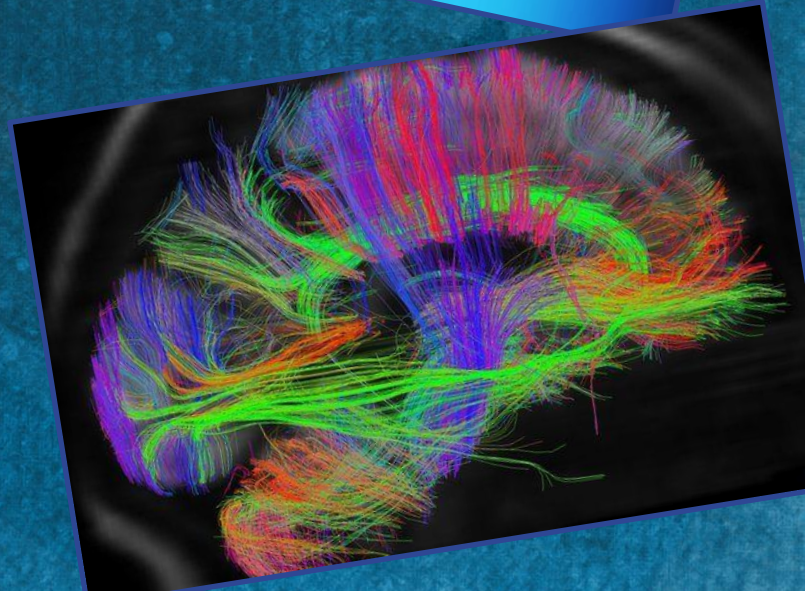
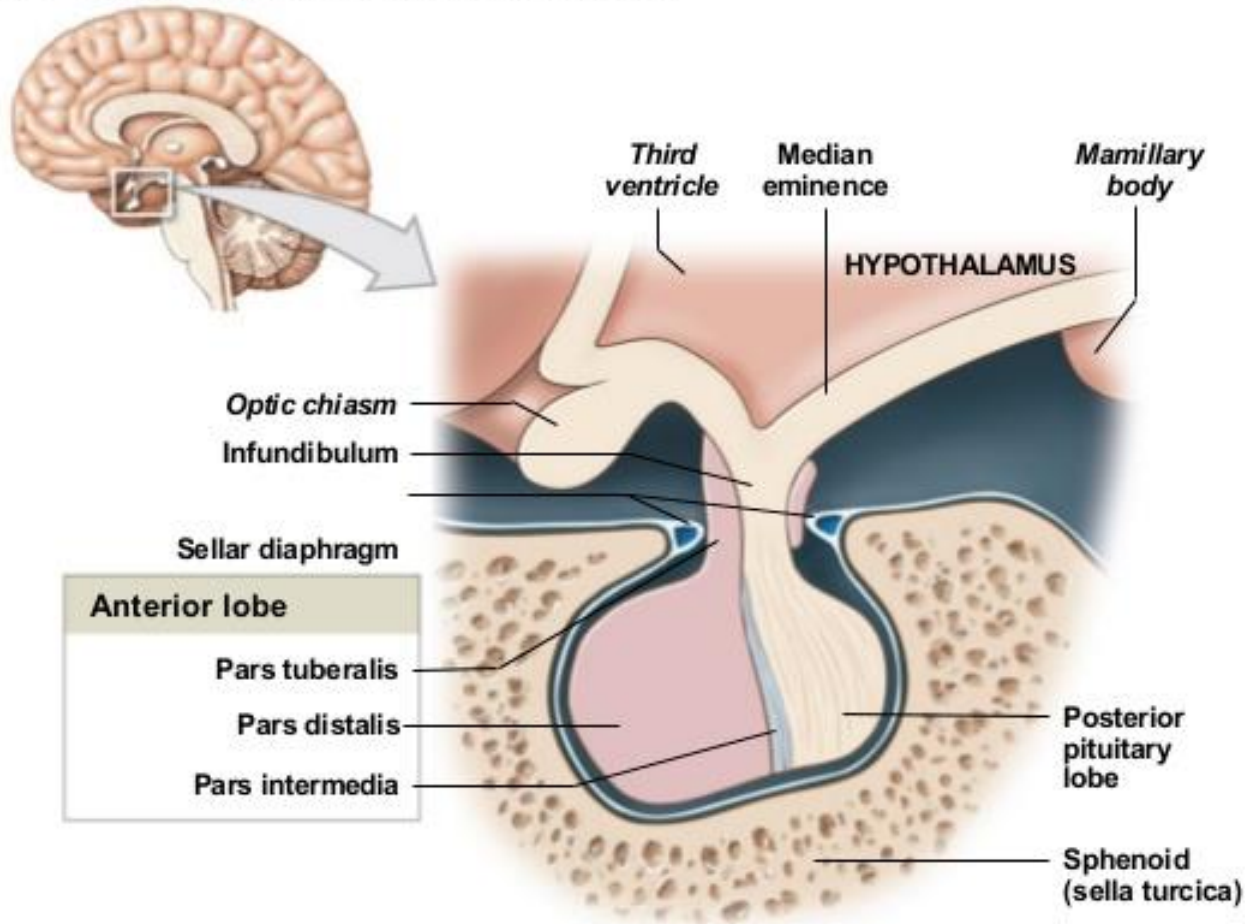
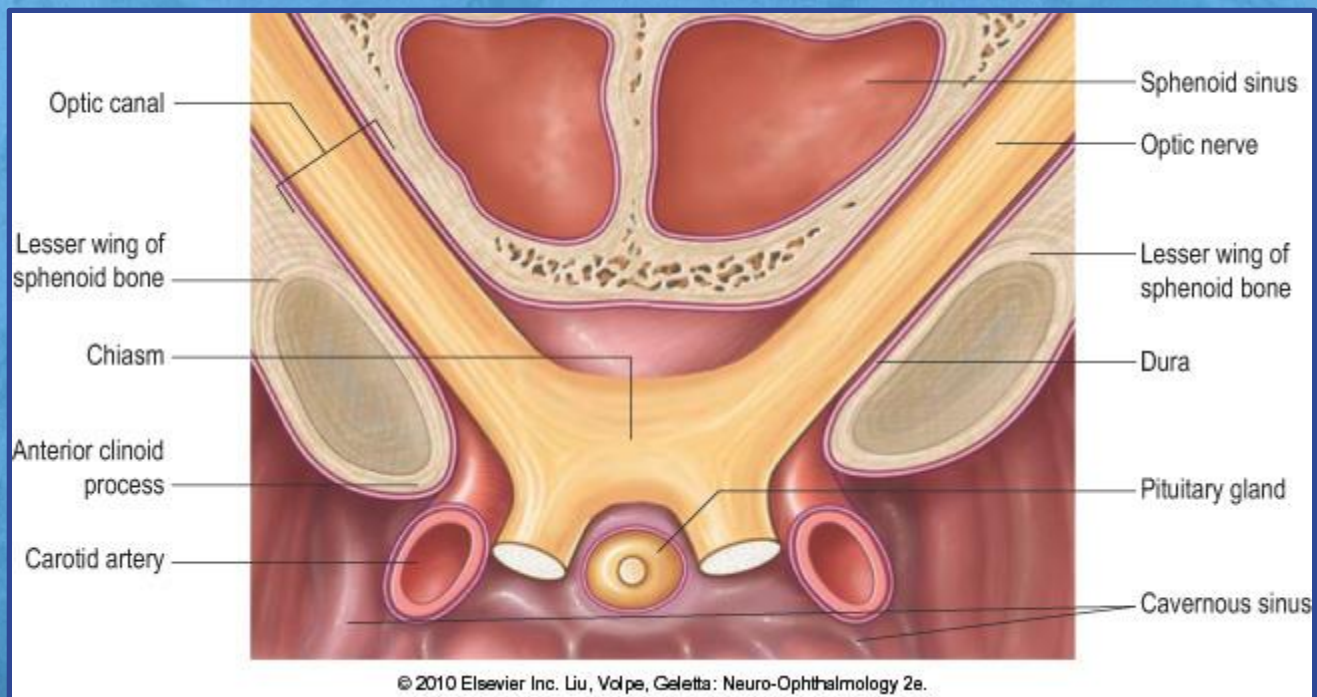
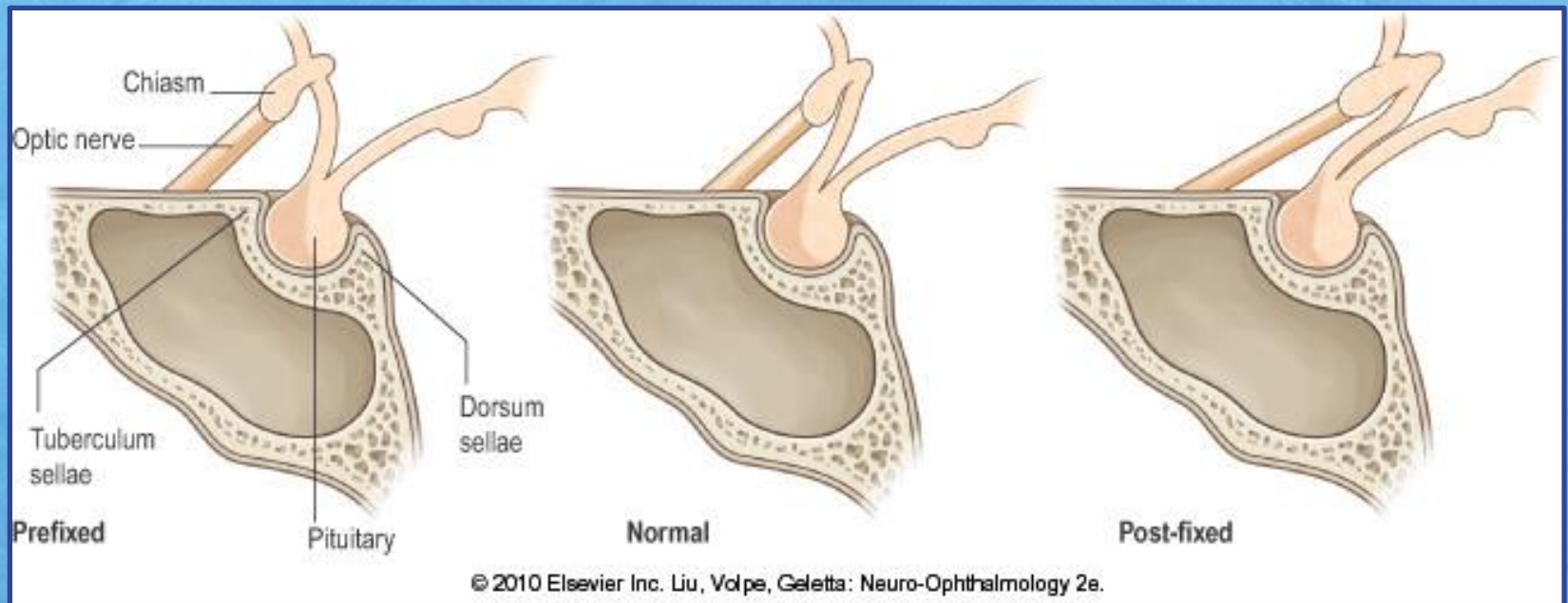
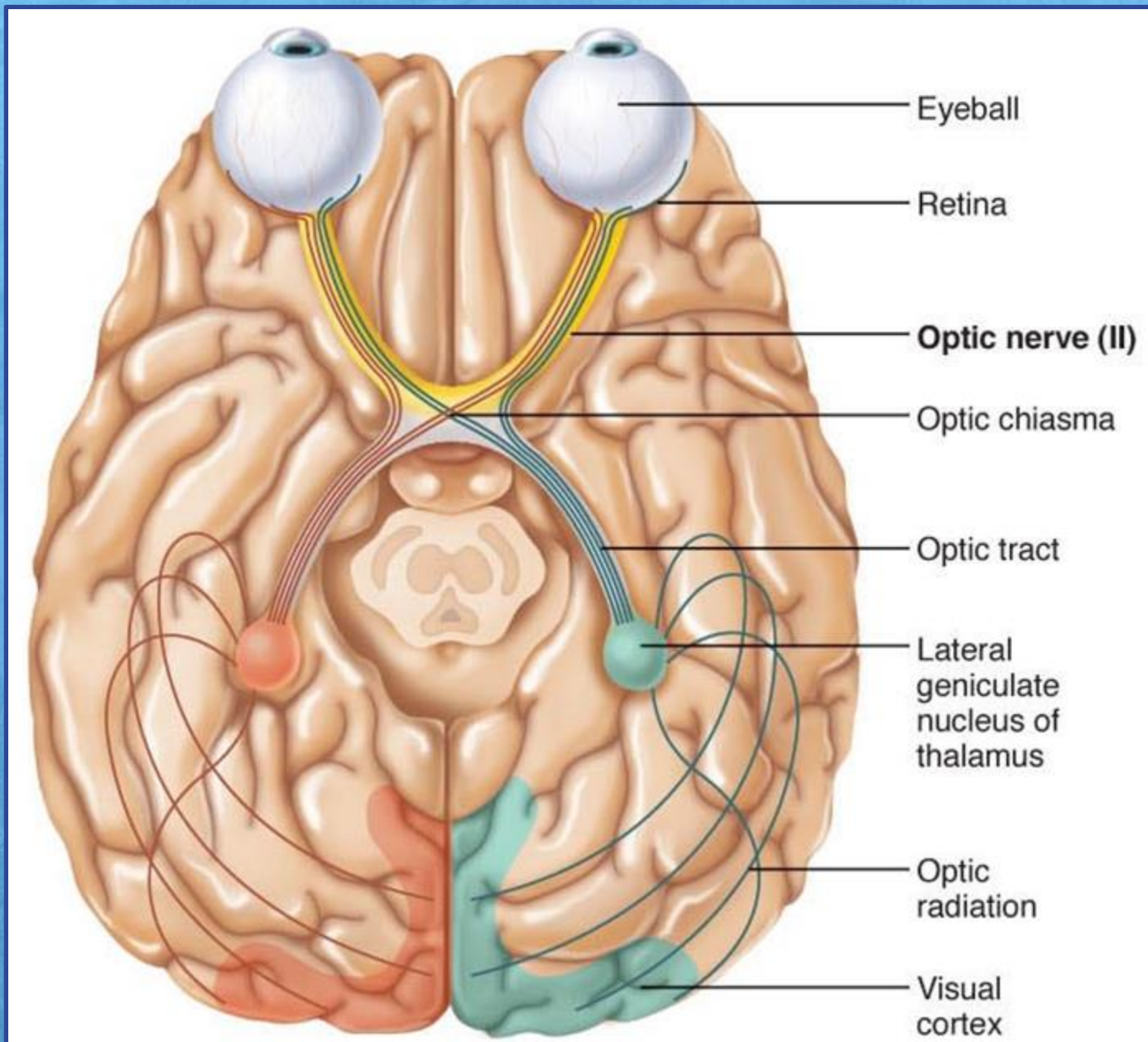


Figure 18-6a The Anatomy and Orientation of the Pituitary Gland

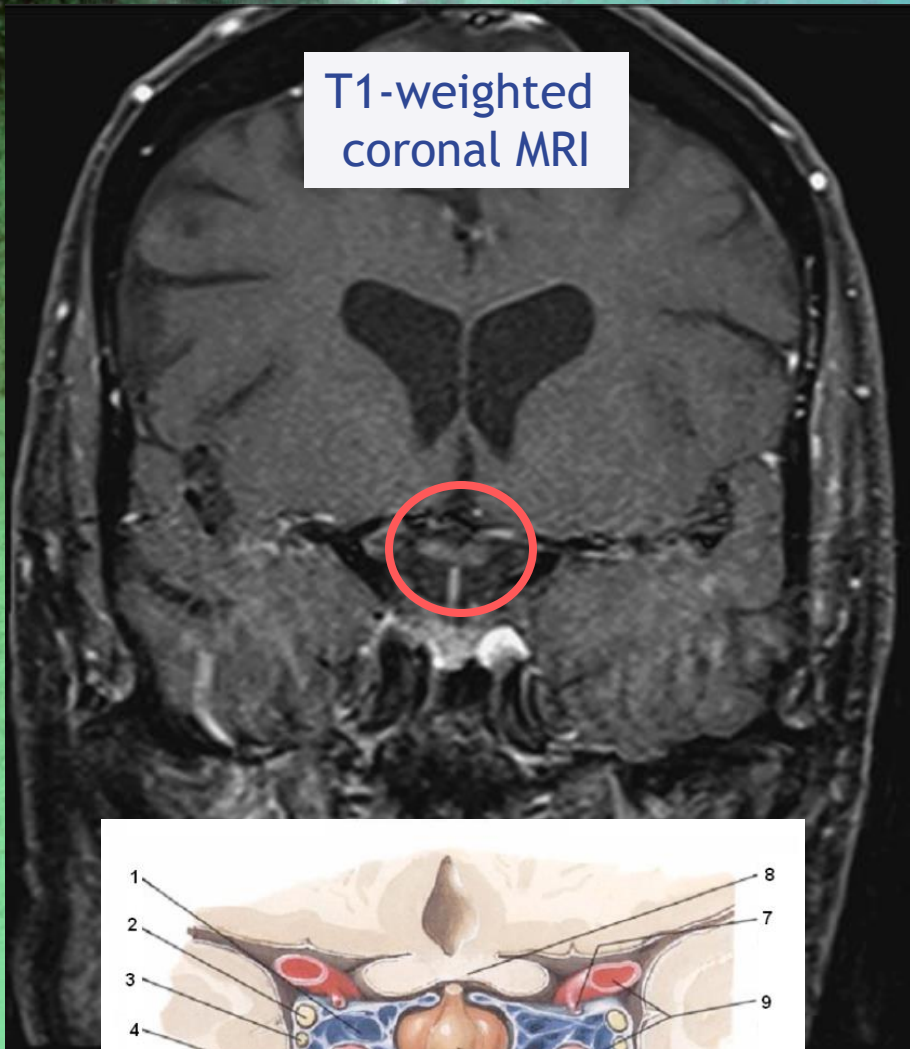


The pituitary gland is located 10mm immediately below the optic chiasm

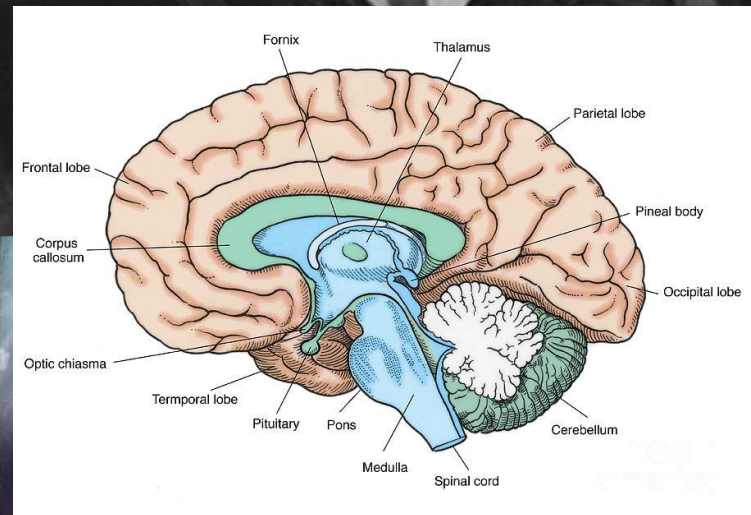
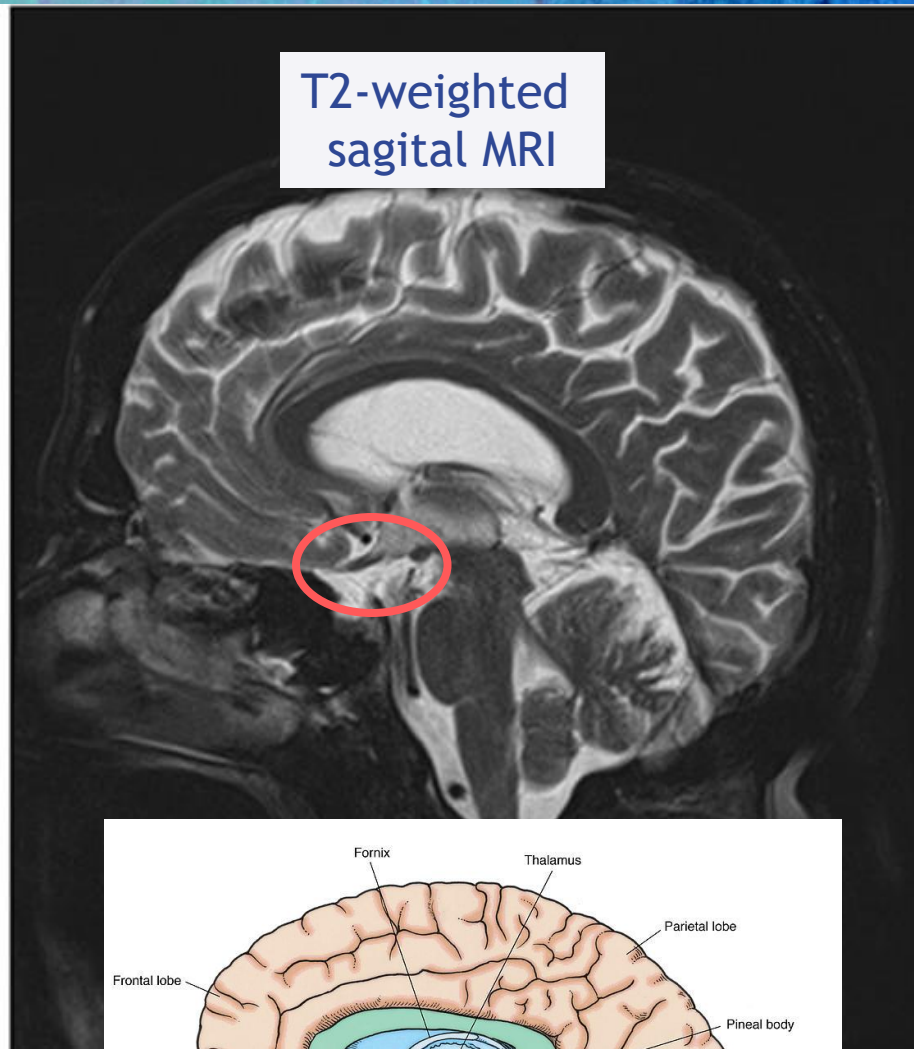




T1-weighted
coronal MRI



T2-weighted
sagittal MRI



Pituitary Adenoma

- **Benign slow-growing tumor**
- Epidemiology
 - 10-25% of all brain tumors
 - Incidence highest 30-45yo age group
 - No racial or sex difference
- Classification
 - **Hormone producing (75%) or non-functioning**
 - Most common (25%) produce prolactin
 - Signs & symptoms determined by hormone secreted, if any

Pituitary Adenoma

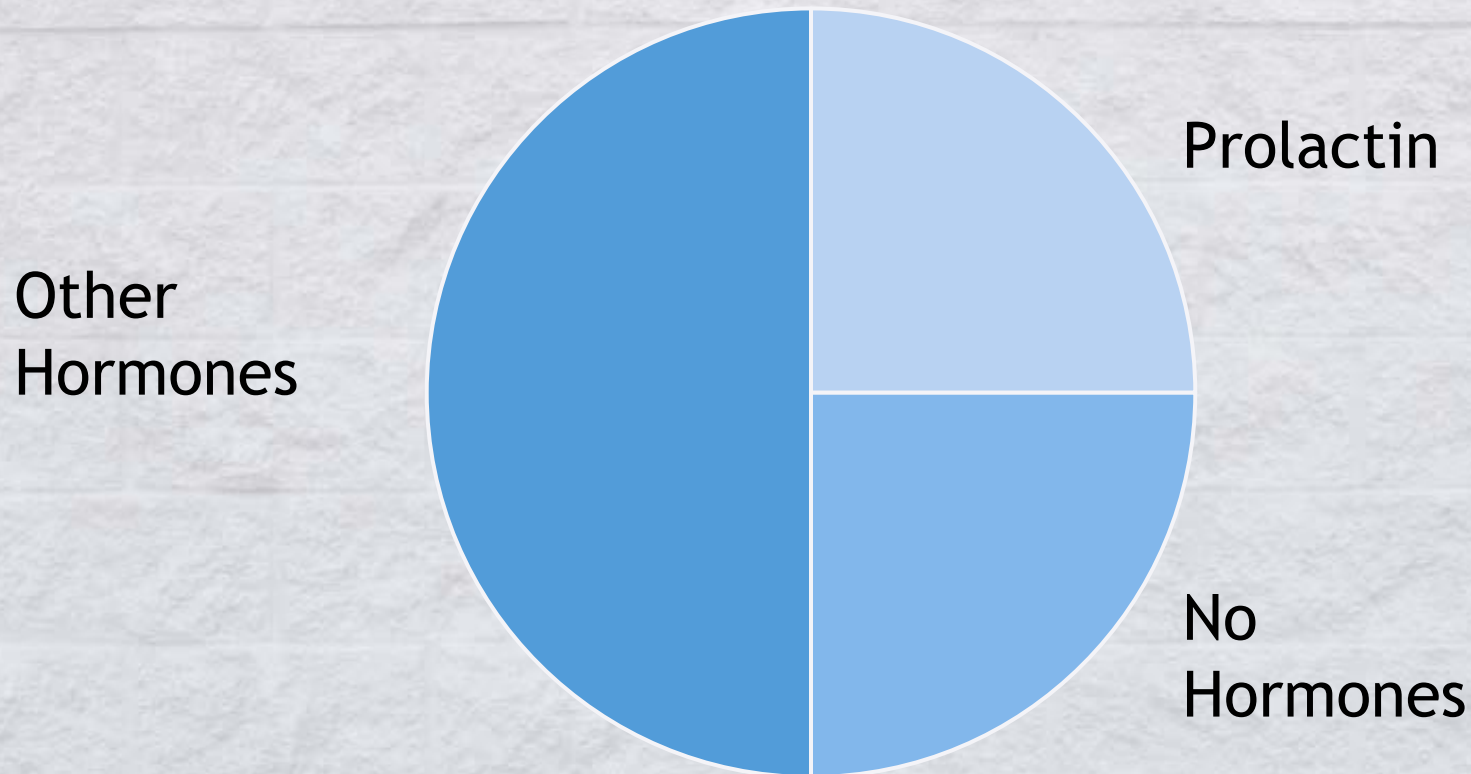
- Nonfunctioning adenomas
 - 25% of cases
 - Most common cause of chiasmal syndrome
 - Only non-specific manifestations, such as **headache**, prior to onset of vision loss
 - May lead to hypopituitarism by compression of adjacent normal gland
 - Findings include diabetes insipidus, fatigue, weight loss, hypothyroidism, sexual dysfunction

Pituitary Adenoma

- Prolactin-secreting adenomas
 - 25% of cases, “prolactinoma”
 - Women: Galactorrhea-amenorrhea syndrome
 - Men: Impotence, loss of libido, infertility
 - Women typically seek care earlier and hence the tumor is discovered while still small
 - Men often wait longer, resulting in **more severe clinical manifestations, including vision loss**

Pituitary Adenoma

Hormone Production

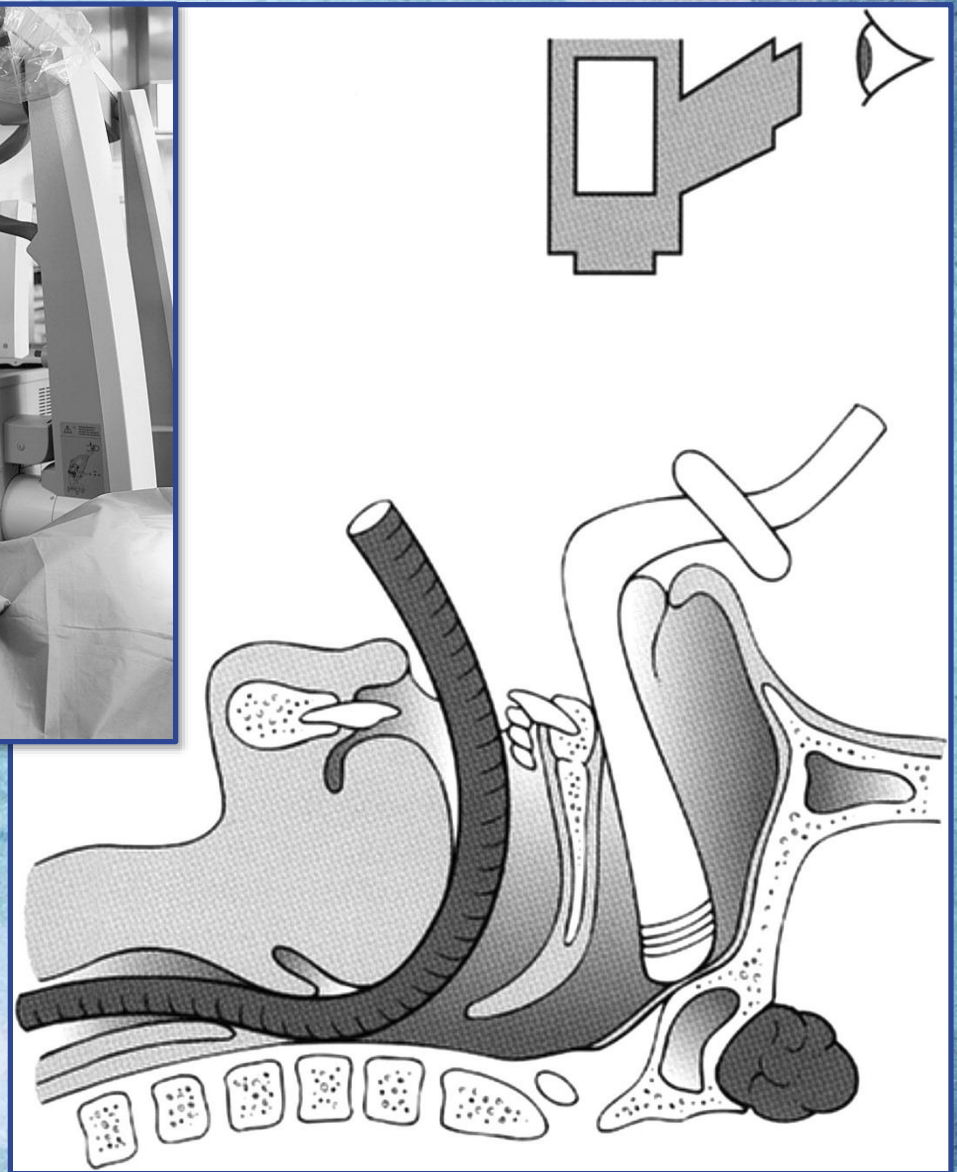


Pituitary Adenoma

- Pituitary Apoplexy
 - The most serious, potentially life-threatening complication of pituitary adenoma
 - Acute **ischemic or hemorrhagic infarction** of the adenoma
 - Rare, with estimated incidence of 1.6% of pituitary adenomas
 - Abrupt onset of symptoms and signs including headache, nausea and vomiting, visual disturbances, oculomotor paresis, confusion and/or coma.

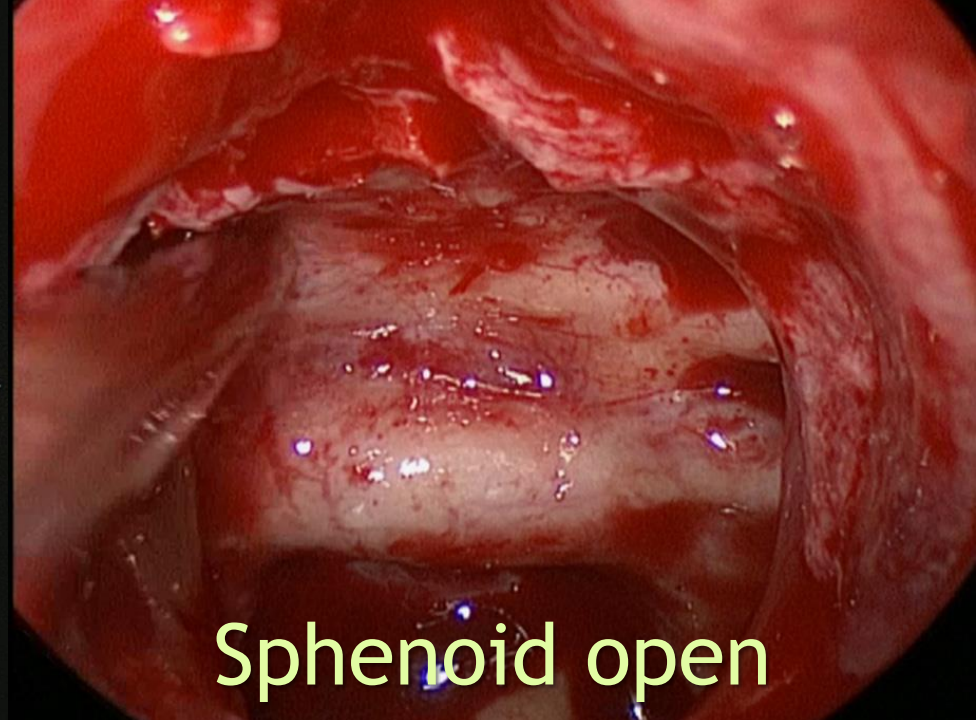
Pituitary Adenoma

- Treatment
 - Medical
 - Treatment-of-choice for smaller hormone-secreting tumors
 - Surgery
 - Treatment for **larger non-secreting tumors** and smaller tumors resistant to medical therapy
 - Endonasal transsphenoidal endoscopic approach used in >90% of cases





Nasal cavity



Sphenoid open



Sella open



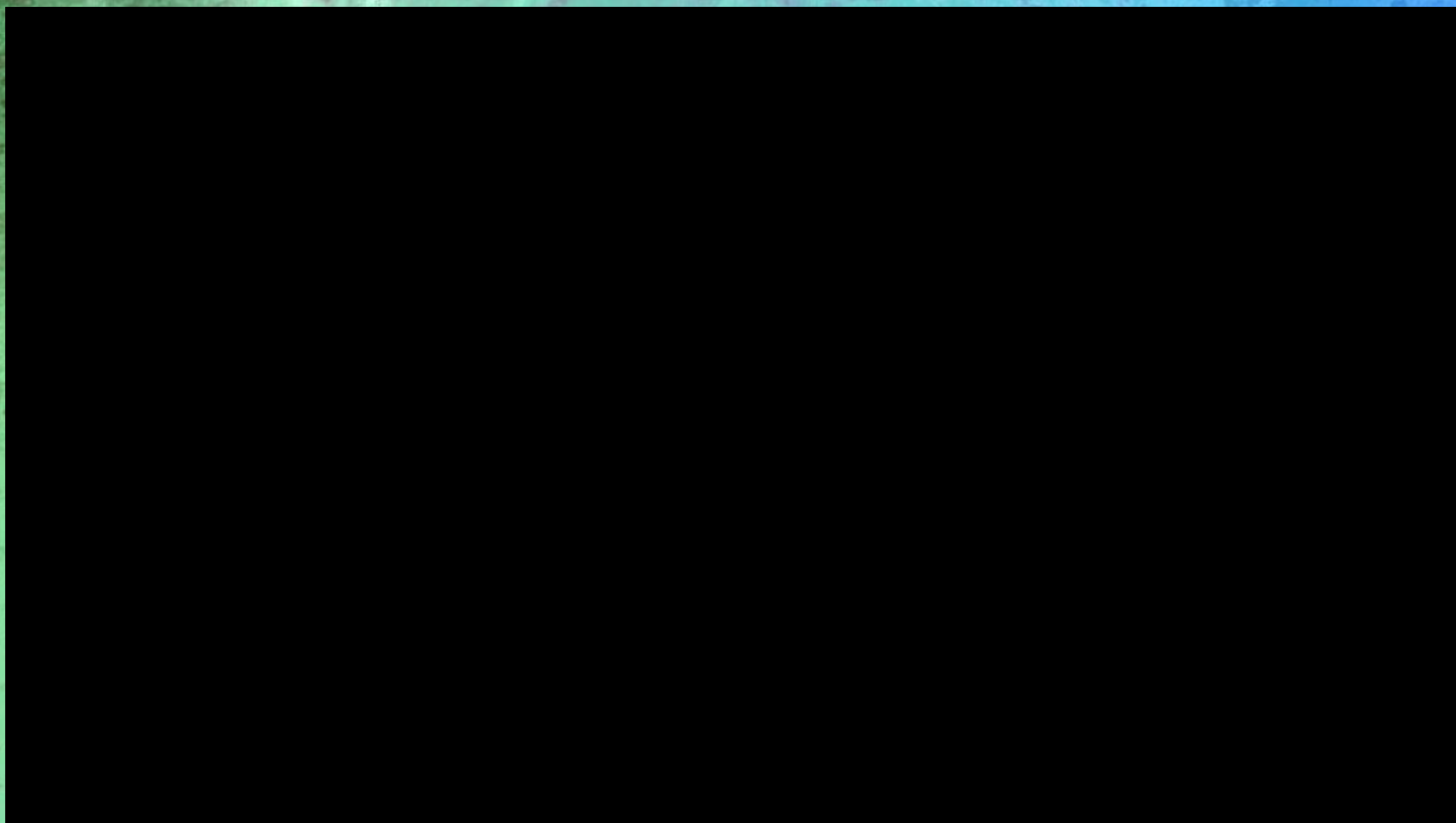
Adenoma

UPMC

Endoscopic Endonasal Approach (EEA)

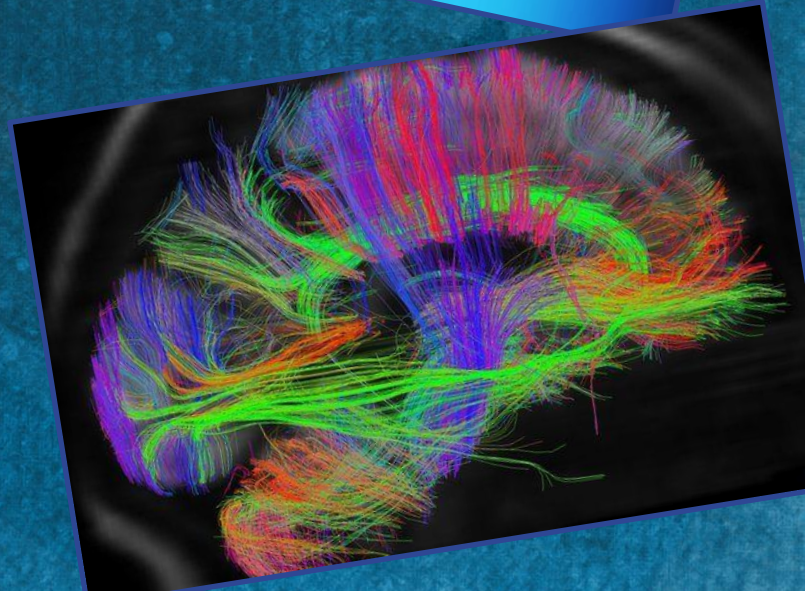
*A Pioneering Surgical Approach for
Skull Base Tumors and Lesions*

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Chiasmal Syndrome

- Anatomy Review
- All About Pituitary Adenomas
- Clinical Features of Chiasmal Syndrome
- Clinical Pearls
 - Red Flag Warning Signs
 - Chiasmal Work-up



Chiasmal Syndrome

SYMPTOMS

- Headache
- Visual loss
- Diplopia
- Loss of depth perception
- Endocrine dysfunction

SIGNS

- Visual field defects
- Optic disc pallor and cupping
- OCT abnormalities
- Oculomotor pareses
- Nystagmus
- Cerebrospinal fluid rhinorrhea

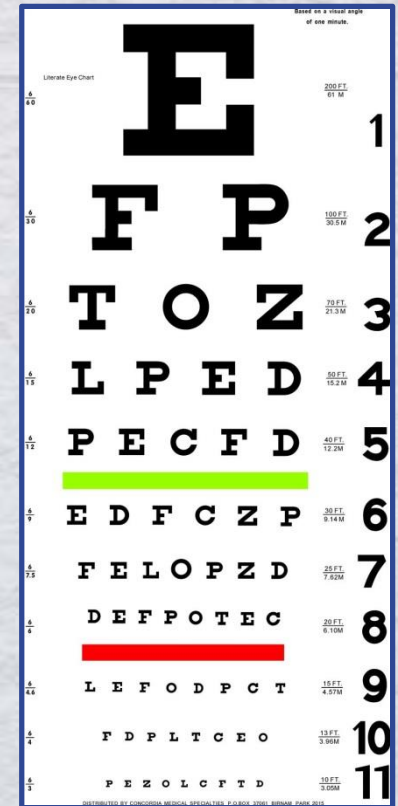
Headache

- 50%-70% of patients with pituitary adenoma
- **Often the presenting symptom**
- May be mild or severe
- HA severity not related to tumor size
 - May be related to hormonal imbalance caused by tumor



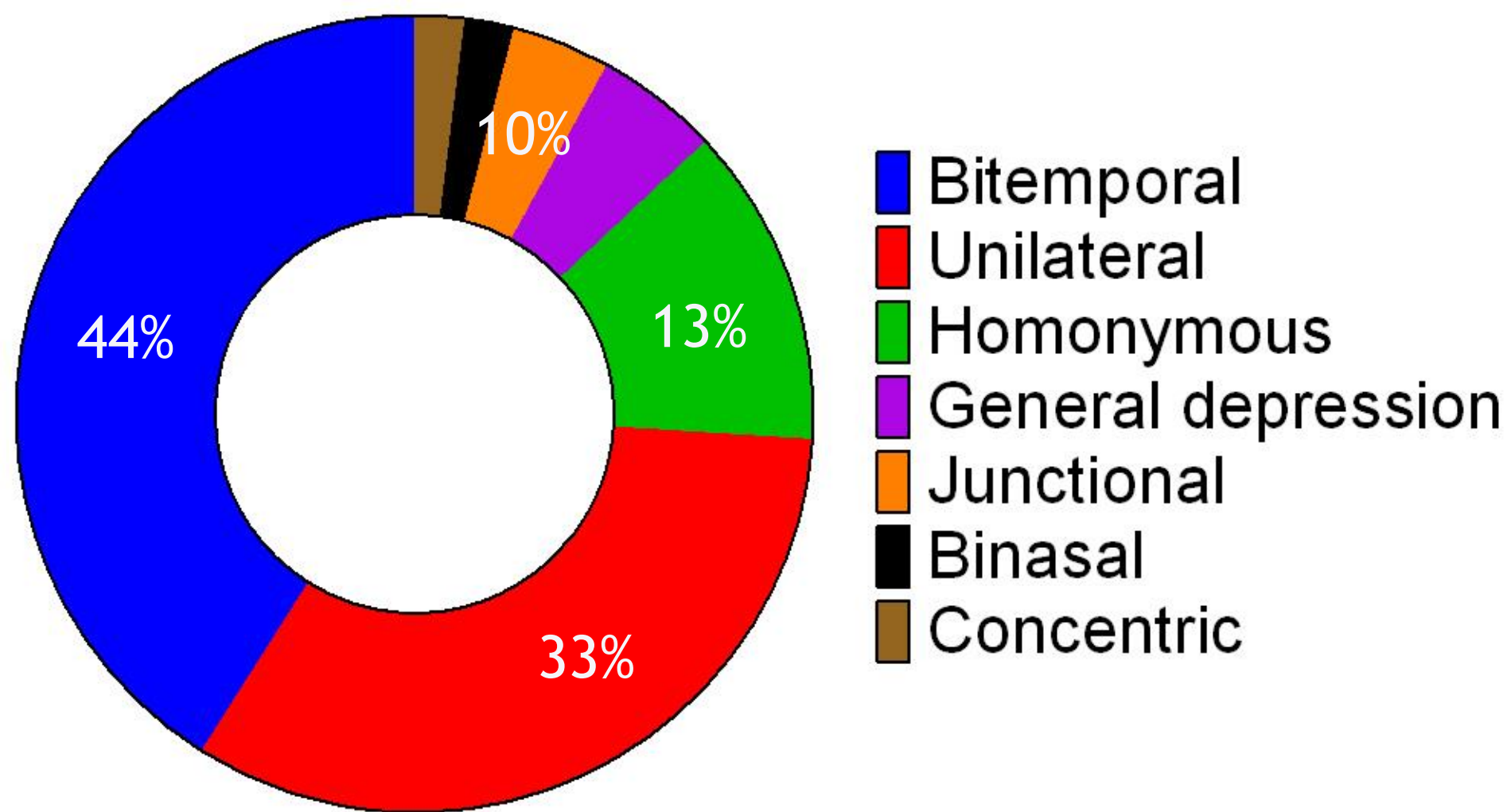
Visual Loss

- **VA is typically normal** in patients with chiasmal lesions
- Depression of central acuity is rare with bitemporal VF defects
- Anterior chiasmal lesions (“junctional scotoma”) are the exception
- Apoplexy is associated with acute vision loss

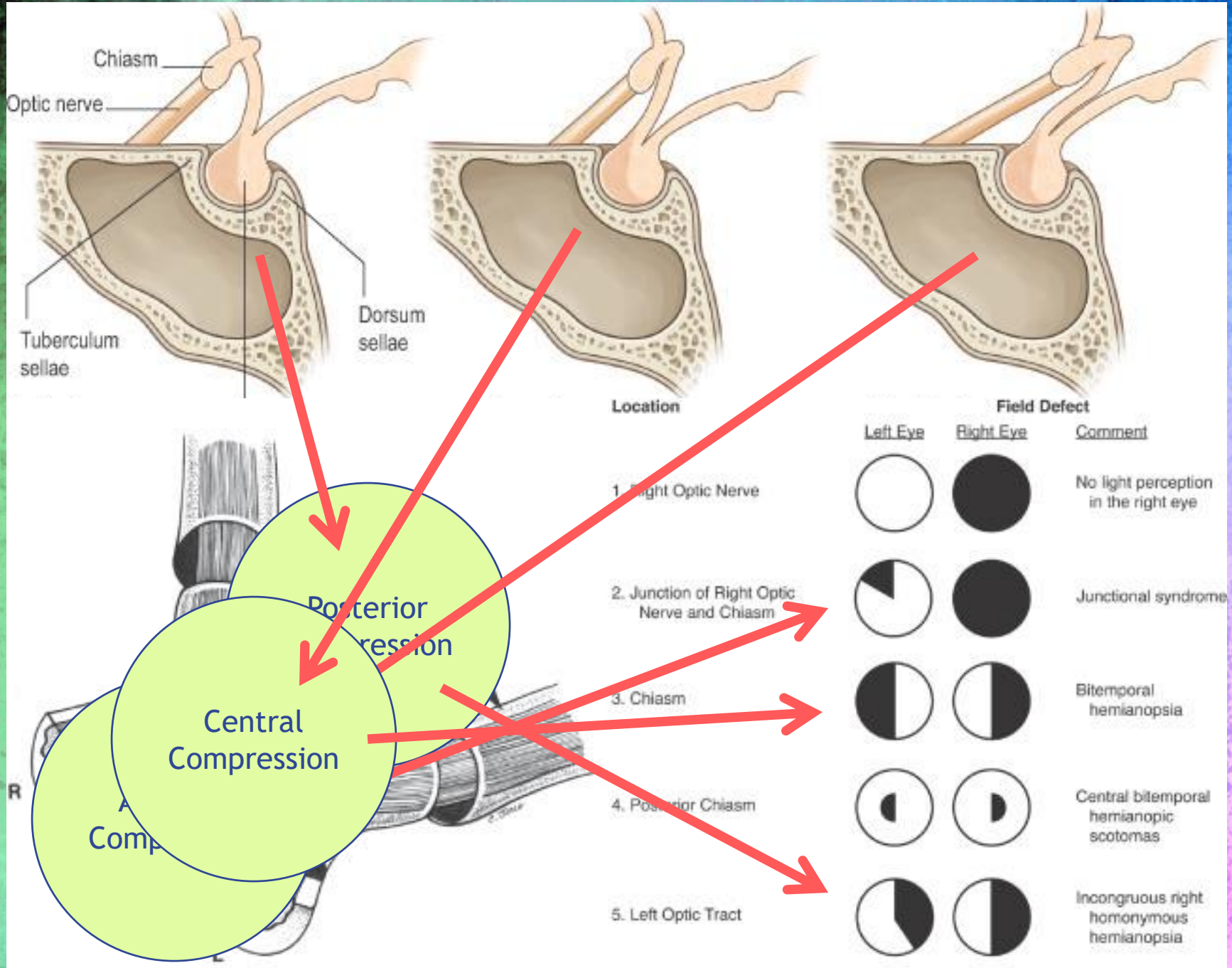


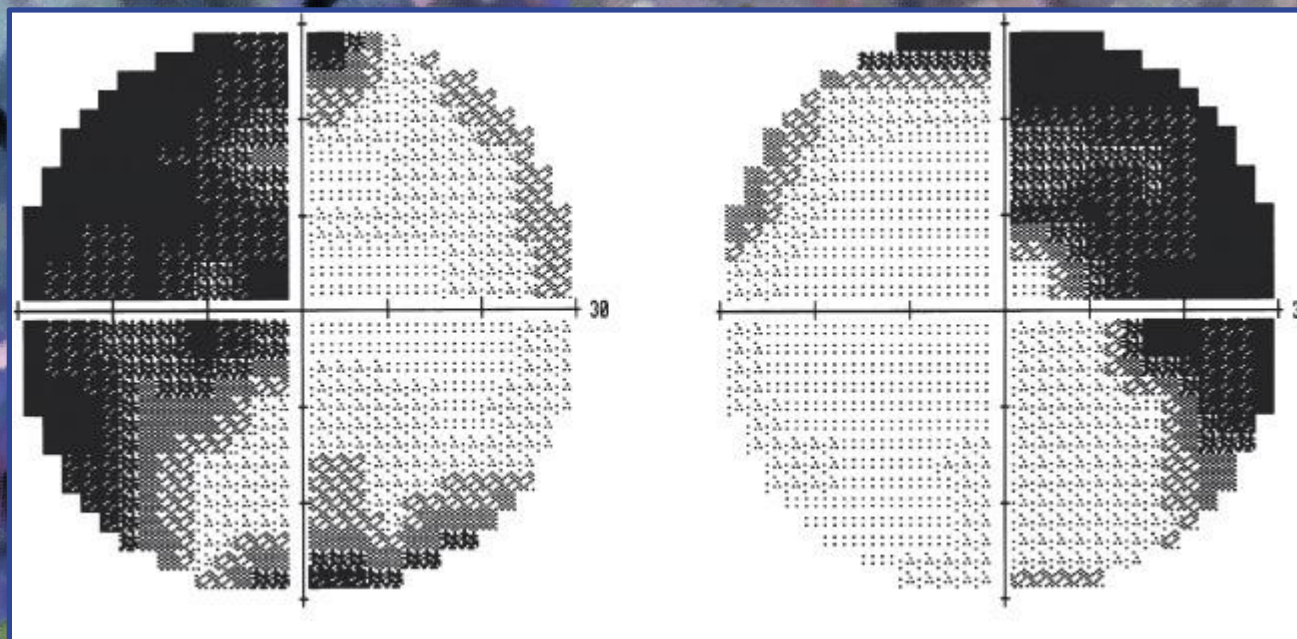
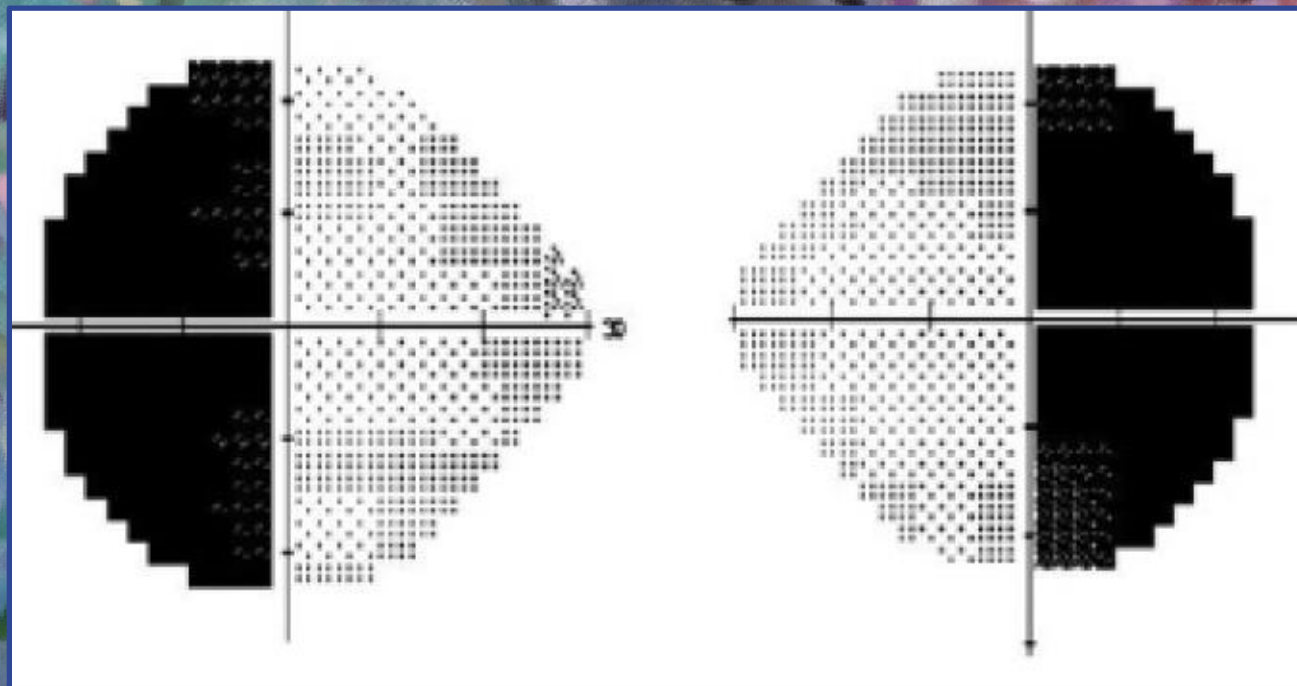
Visual Field Defects

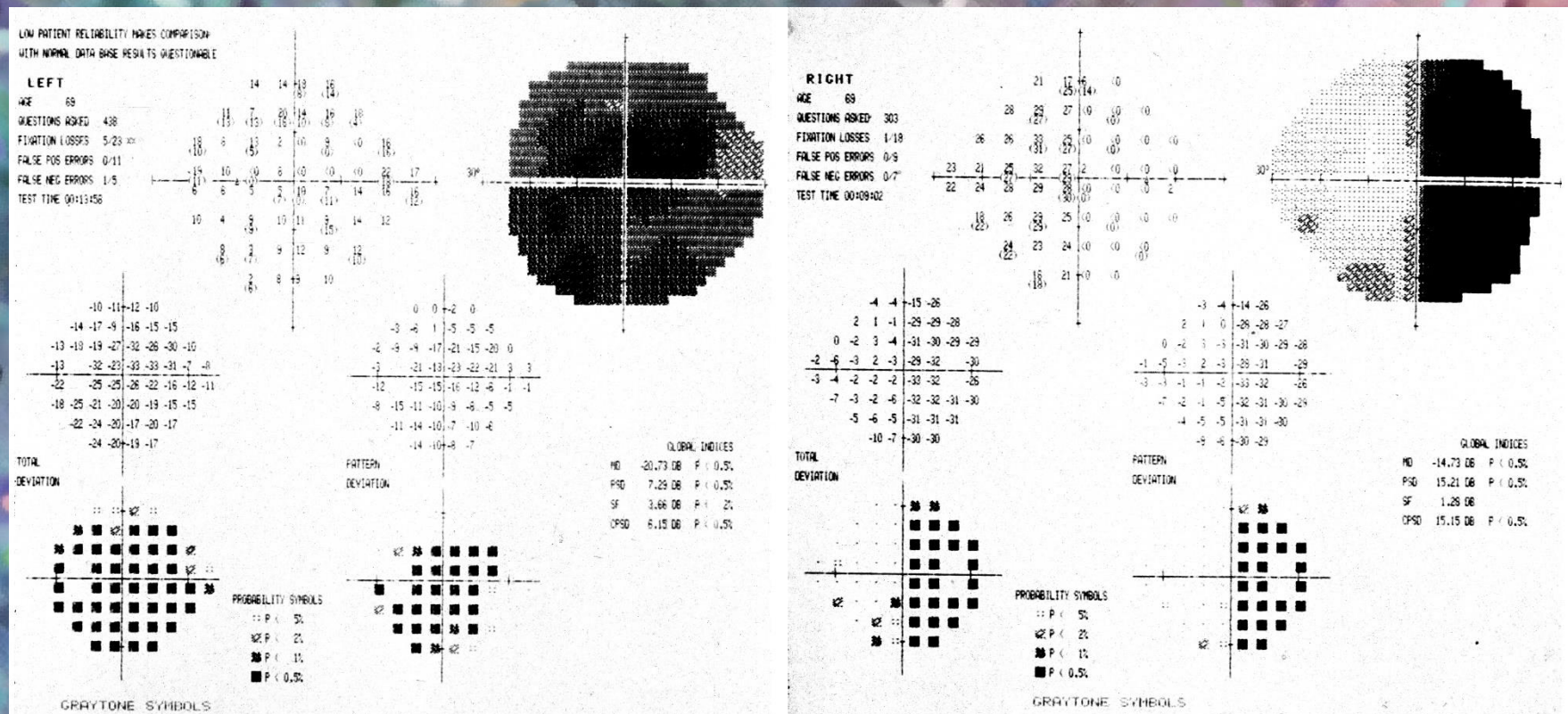
- Perimetry is a key test for detecting chiasmal lesions
- Prior to acuity loss, VF defects may be the *only* clinical sign of a chiasmal lesion
- VF defects occur most often in patients with non-functioning tumors
- Age at presentation is 10yrs older than patients with functioning tumors
 - Mean age at presentation: 54yo (non-function)



Visual field defects in 103 consecutive patients presenting to neurosurgery with pituitary adenoma

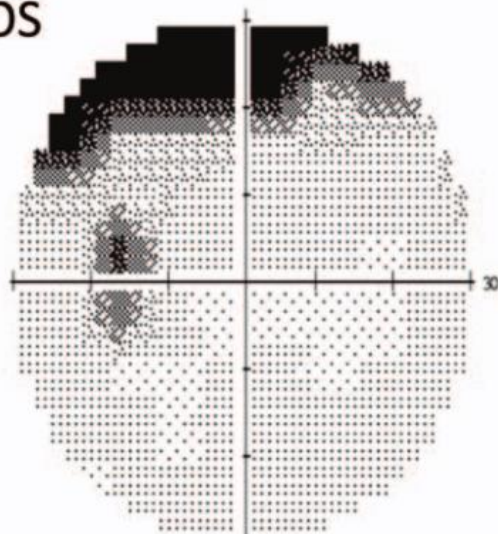




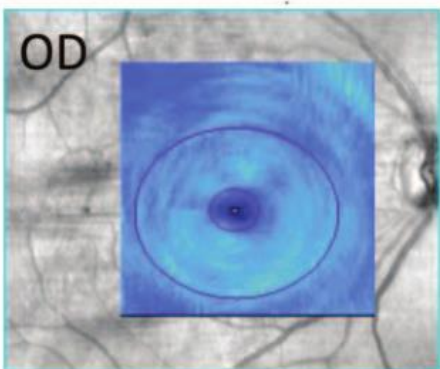
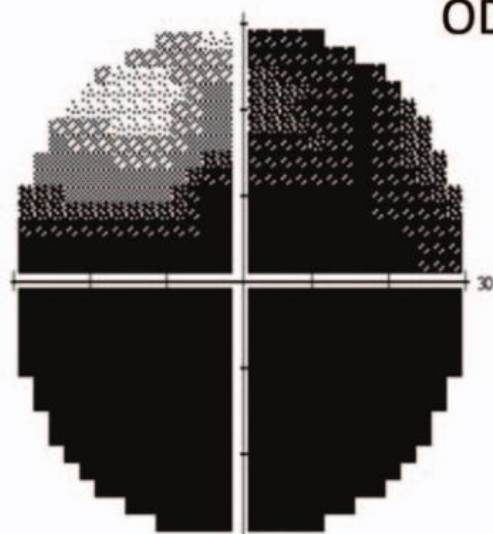


Junctional scotoma of the left eye. This 69yo man presented with c/o vision loss OS x 4 weeks. BVA was 20/25 OD and FC OS. +APD OS. CT scan revealed a pituitary adenoma.

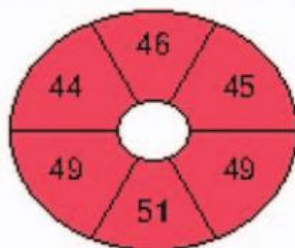
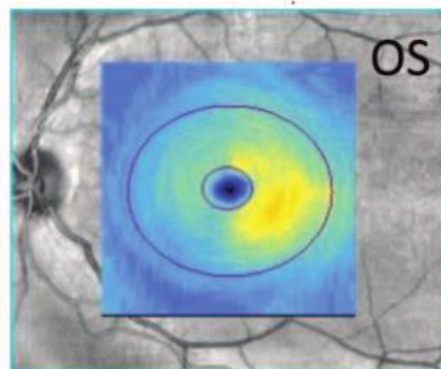
OS



OD

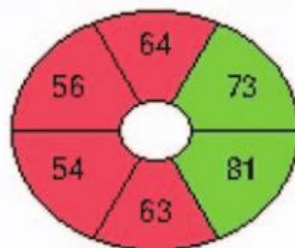


225
150
75
0 μm

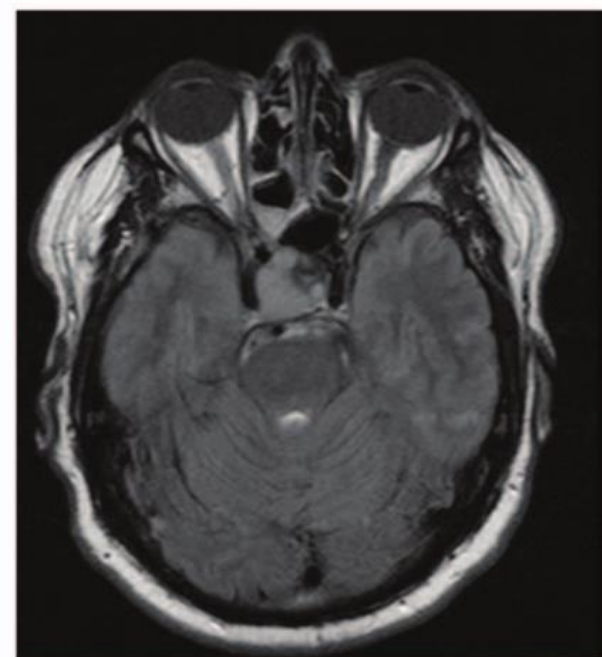


Diversified:
Distribution
of Normals

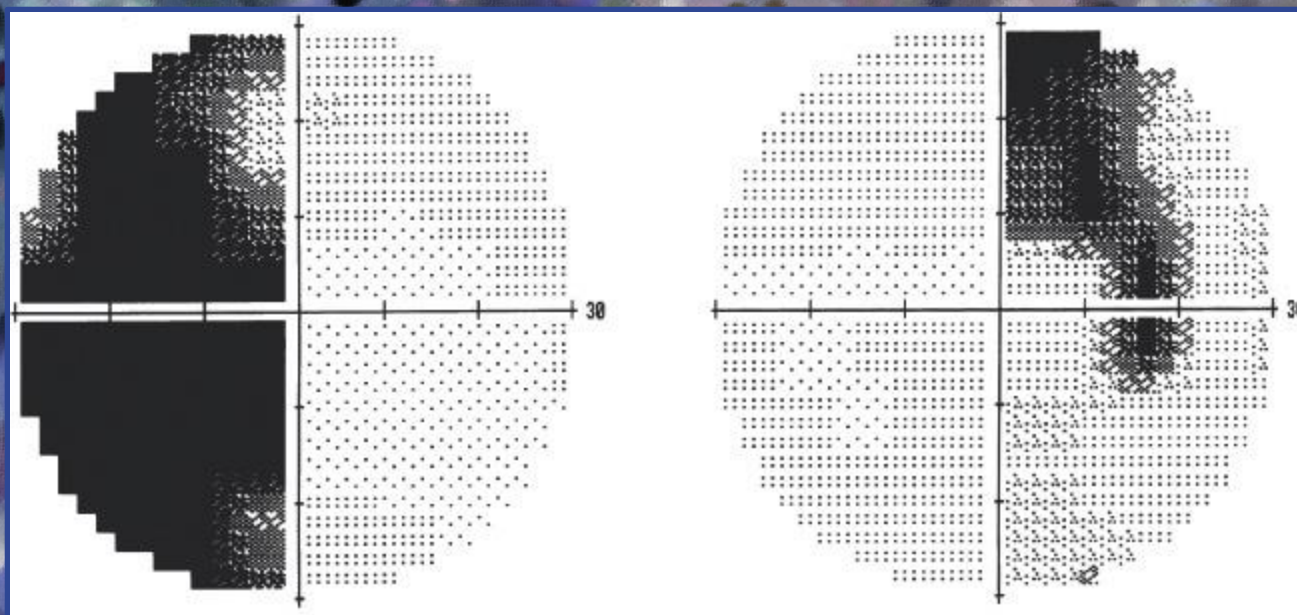
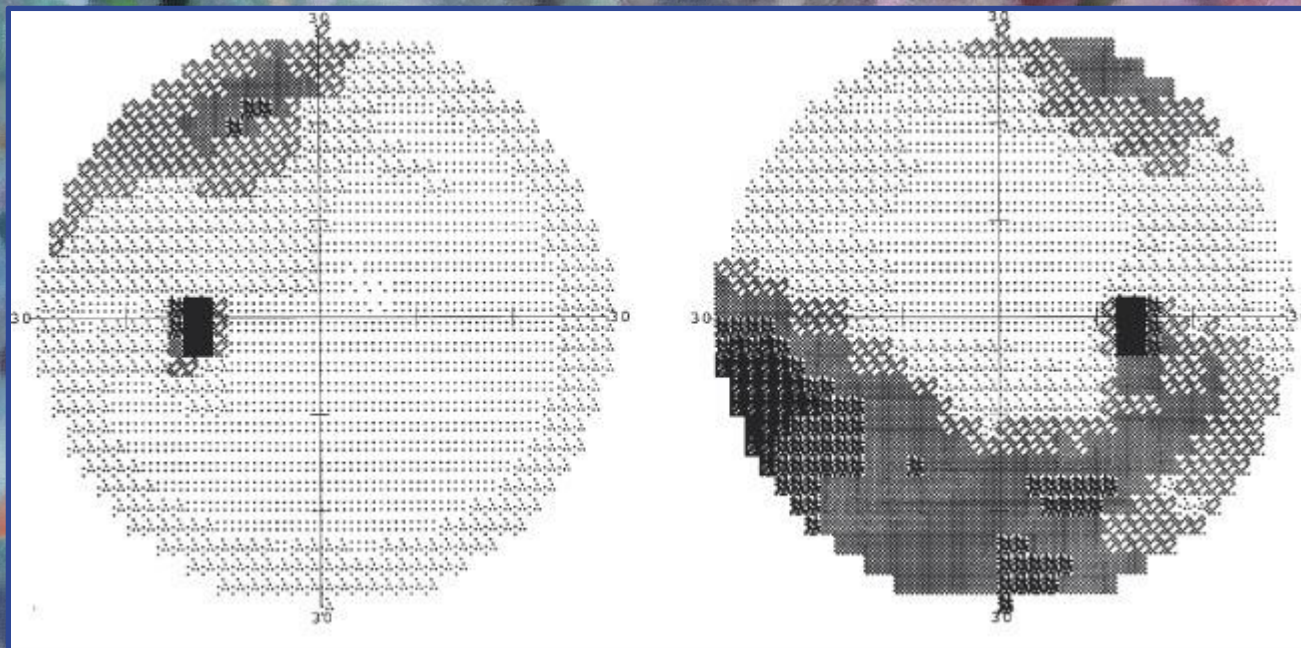
95%
5%
1%



	OD μm	OS μm
Average GCL + IPL Thickness	47	65
Minimum GCL + IPL Thickness	42	53

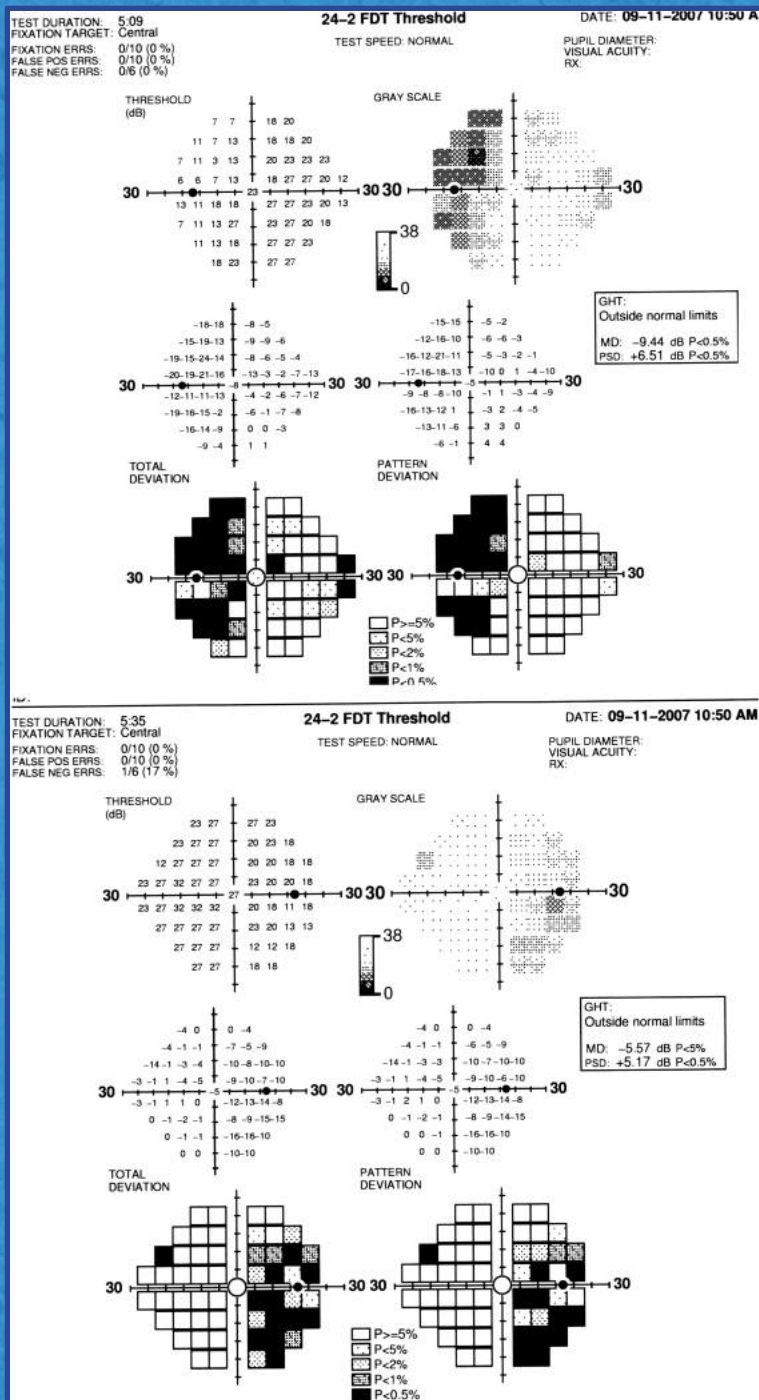


Vuong LN, Hedges TR. Curr Opin Ophthalmol 2017;28:573-8.



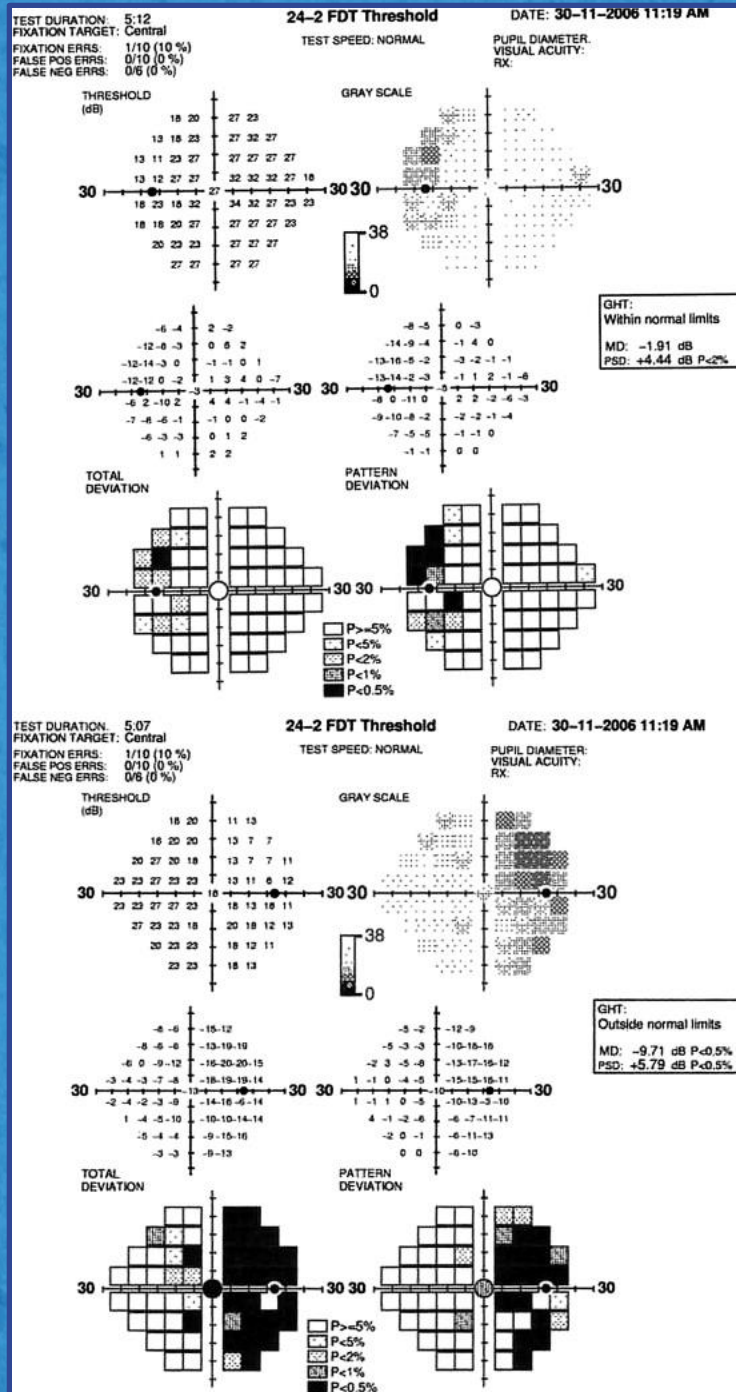
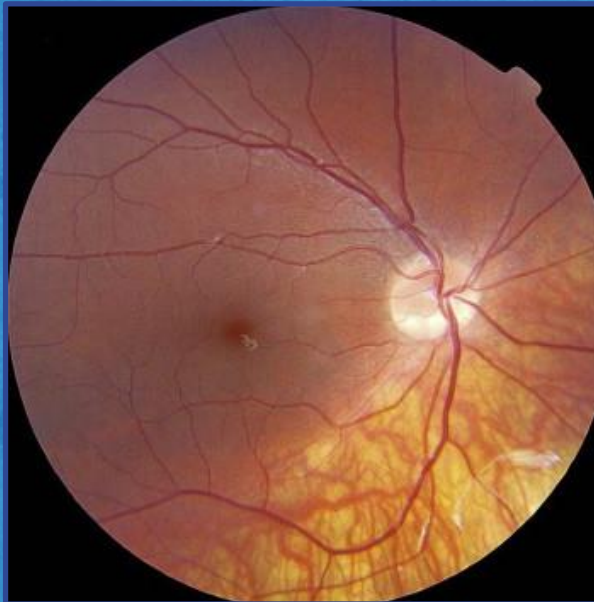
Visual Field Defects

- Bitemporal hemianopia is NOT pathognomonic for chiasmal syndrome
- Other conditions that can give rise to bitemporal vision loss
 - Tilted disc syndrome
 - Overhanging redundant upper lid tissue
 - Enlarged blind spots
 - Bilateral medullation of nasal nerve fibers



Tilted disc
 syndrome
 simulating
 bitemporal
 hemianopia

Sowka JW, Luong V
 V. Optometry
 2009;80:232-42.

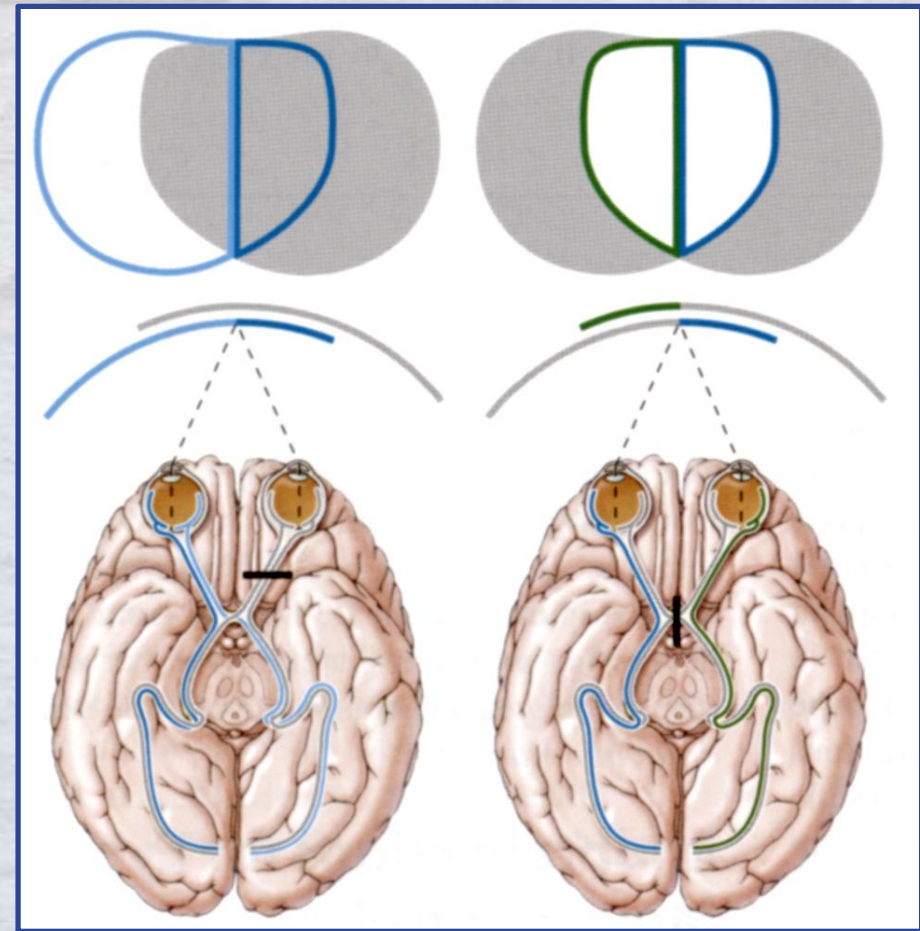


Tilted disc
syndrome
simulating
bitemporal
hemianopia

Sowka JW, Luong V
V. Optometry
2009;80:232-42.

Bitemporal Hemianopia

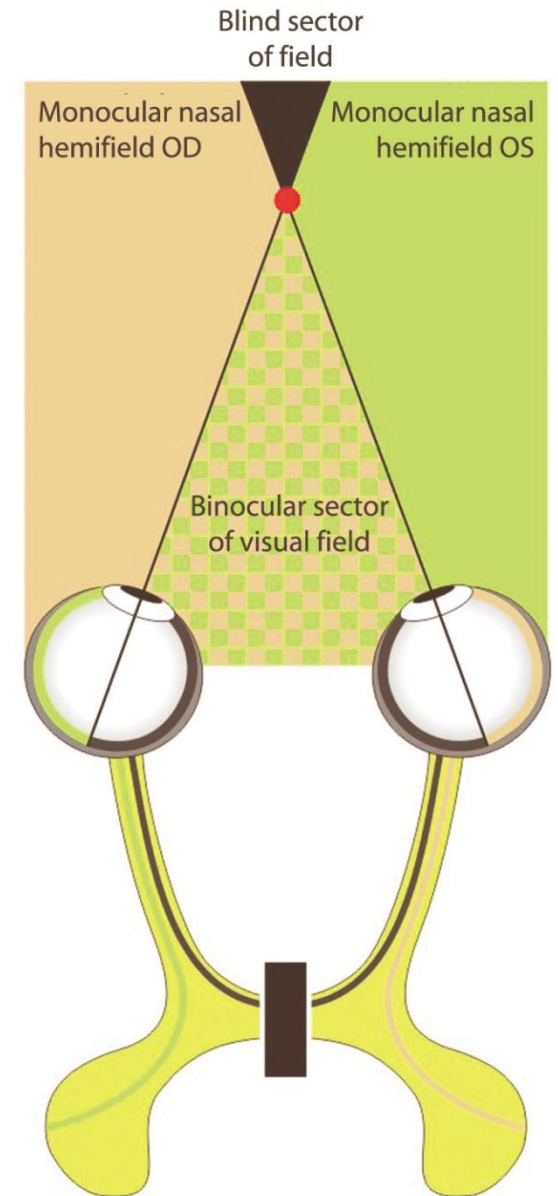
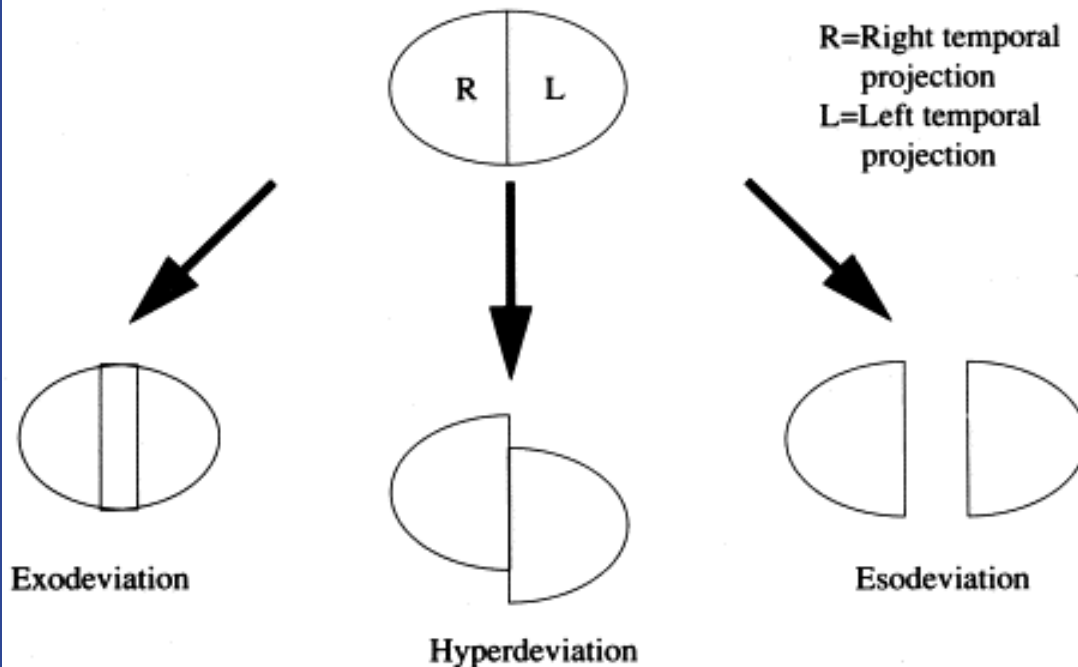
- The temporal crescents are the only part of the binocular VF that is lost
- A central 110-120° remains but there are **no overlapping VF elements**
- Lack of fusion lock decompensates any pre-existing phoria into a tropia



Absence of fusion lock allows hemifields to slide.

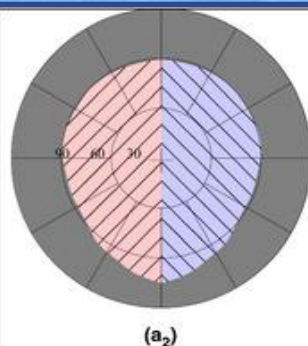
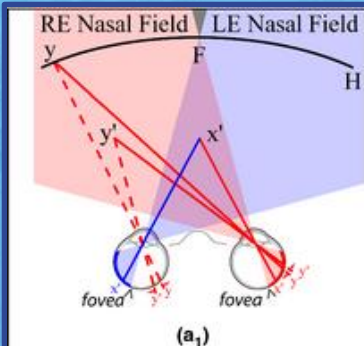
When converged at near, there is overlap before the target and blindness behind it.

KIRKHAM'S "HEMIFIELDS SLIDE PHENOMENON"



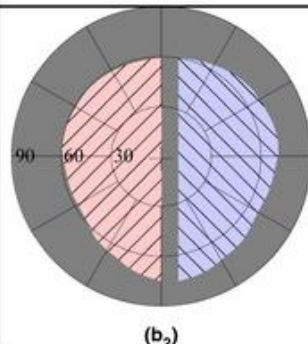
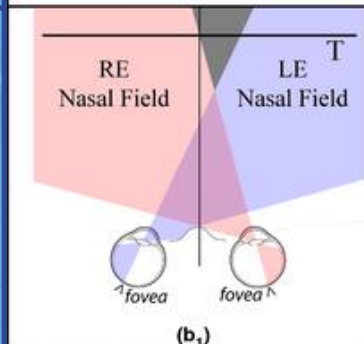
Orthophoria.

Orthotropic fixation results in mild loss of peripheral vision (temporal crescents) and a normal percept.



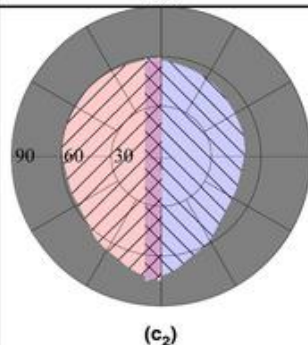
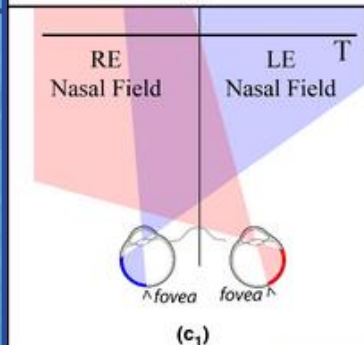
Left esotropia.

Left esotropia shifts the left nasal field to the right, leaving a vertical strip of central scotoma between the two nasal hemi-fields



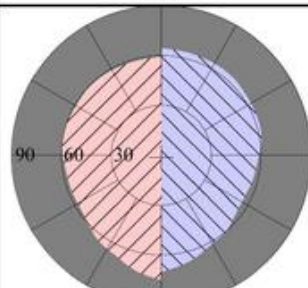
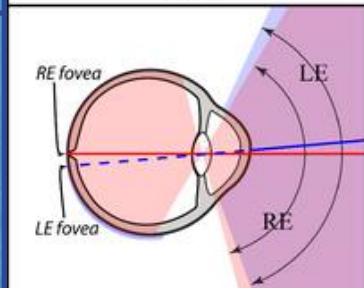
Left exotropia.

Left exotropia shifts the left nasal field to the left, overlapping the right nasal field (crosshatched area), resulting in diplopia.



Left hypertropia.

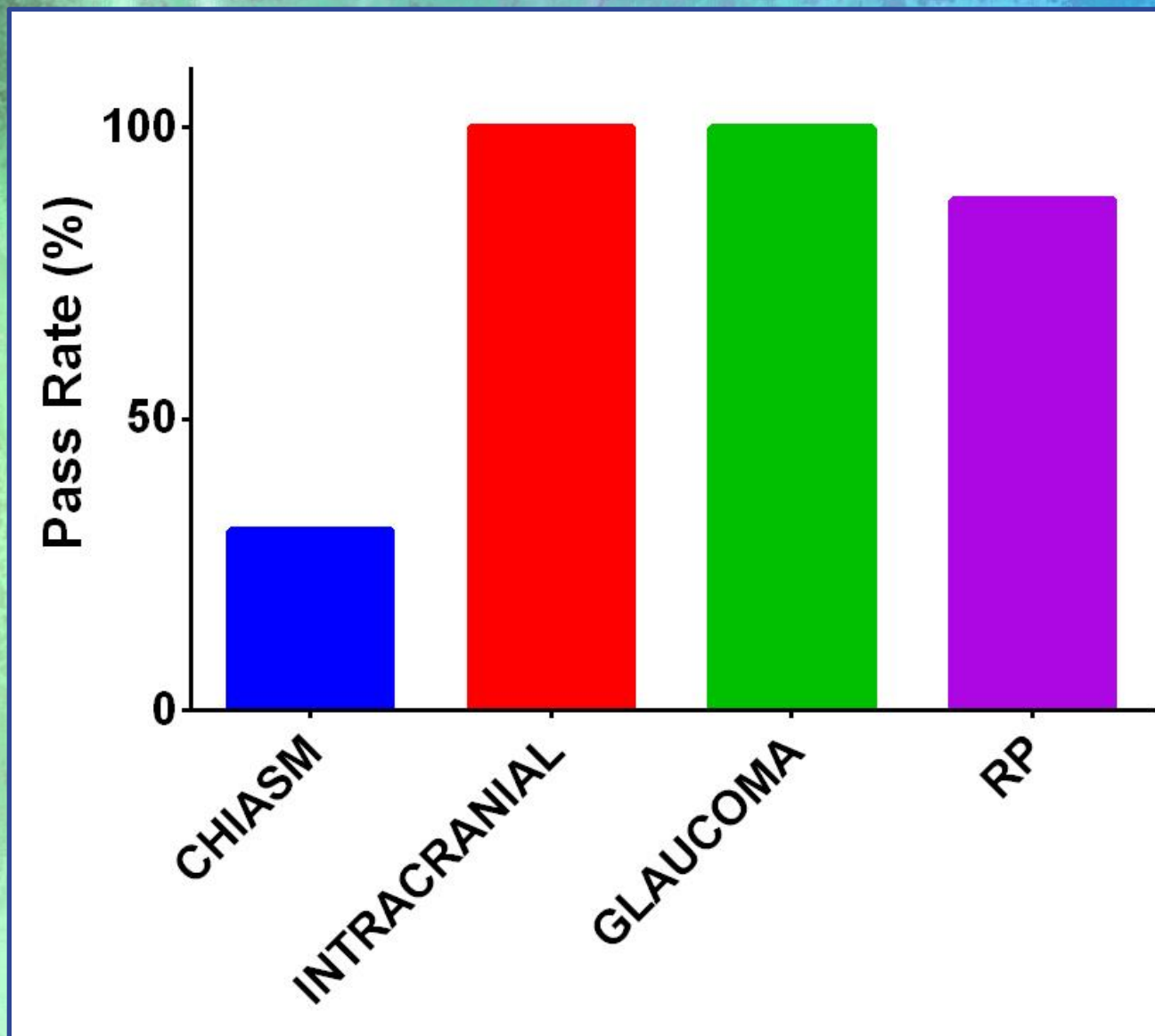
Left hypertropia slides the left nasal hemifield upward causing the right image to be perceived as lower. This may be reported as double vision (split diplopia)



Diplopia & Stereopsis

- Intermittent diplopia occurs due to decompensating exophoria and vertical imbalance
- *Poor depth perception is an important symptom of chiasmal syndrome*
- Loss of overlapping VF at fixation results in severe loss of stereopsis, even when VF loss is minimal and VA is preserved.
- **Stereo tests are a simple, easy, and quick screening test for chiasmal disease**

Effect of various disease conditions on Titmus Stereo Test



Chiasmal Syndrome

SYMPTOMS

- Headache
- Visual loss
- Diplopia
- Loss of depth perception
- Endocrine dysfunction

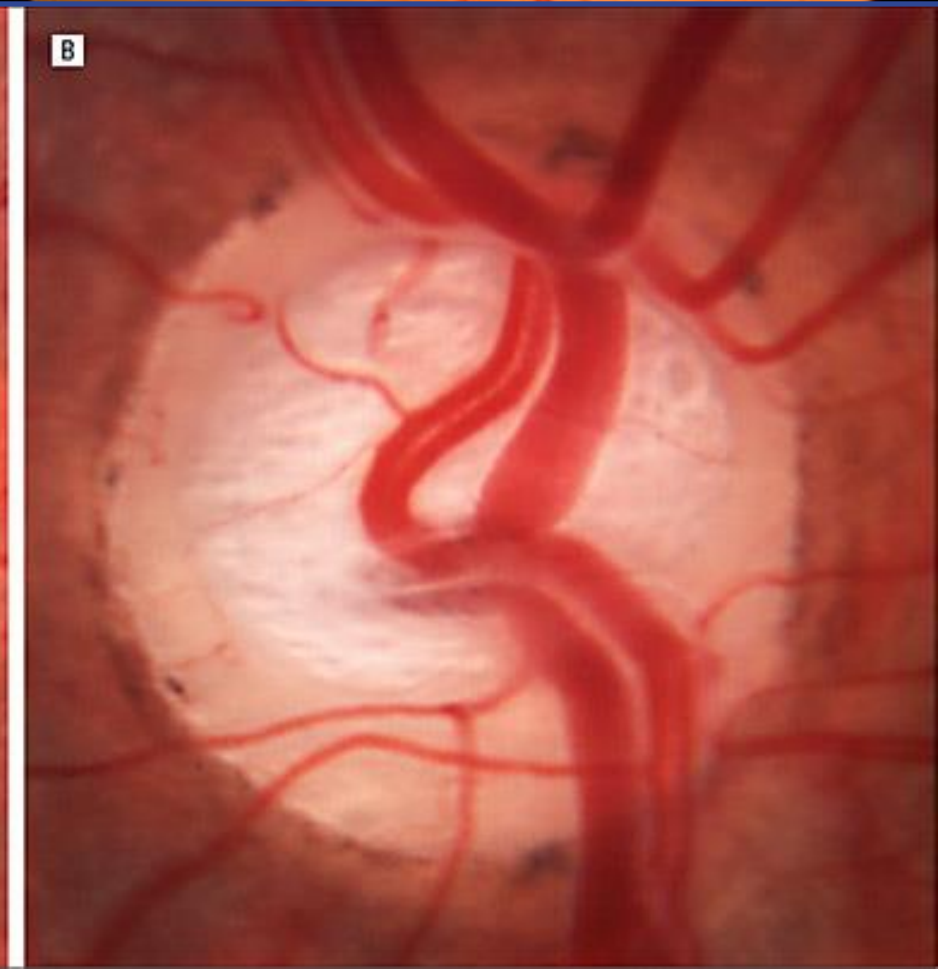
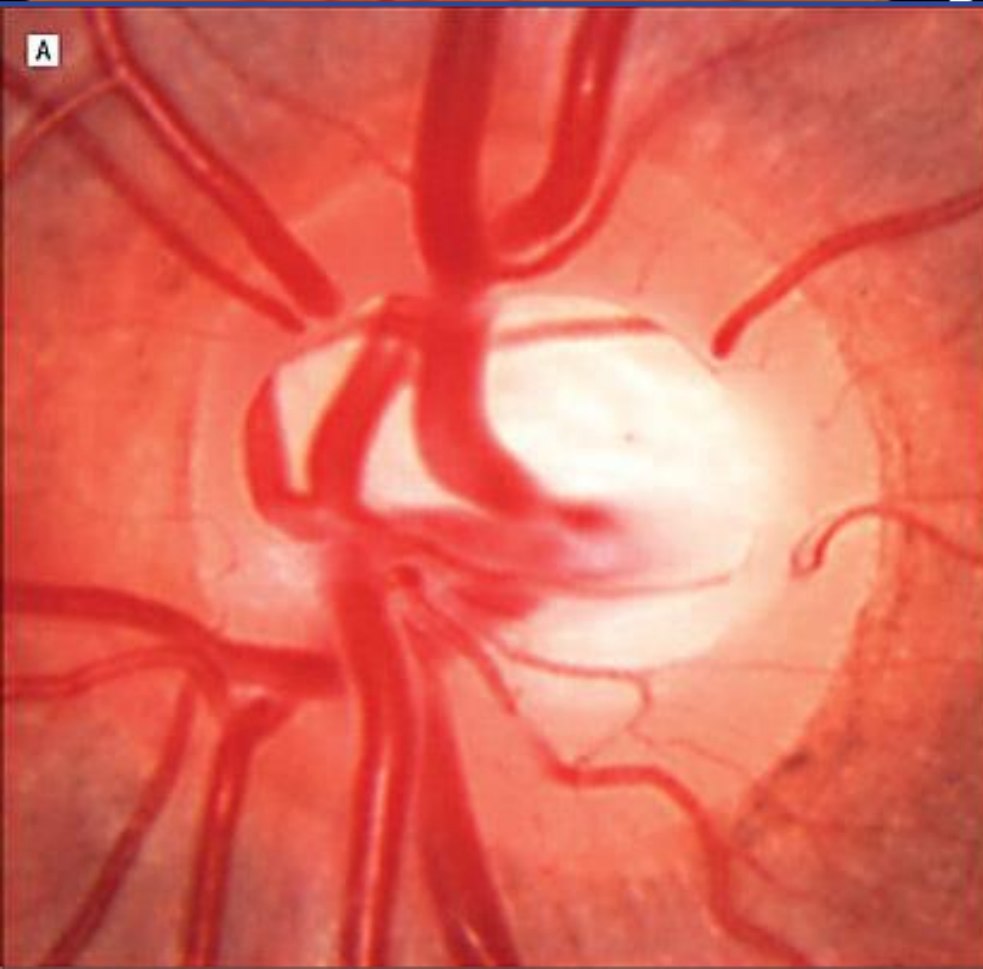
SIGNS

- Visual field defects
- Optic disc pallor and cupping
- OCT abnormalities
- Oculomotor pareses
- Nystagmus
- Cerebrospinal fluid rhinorrhea

Optic Disc

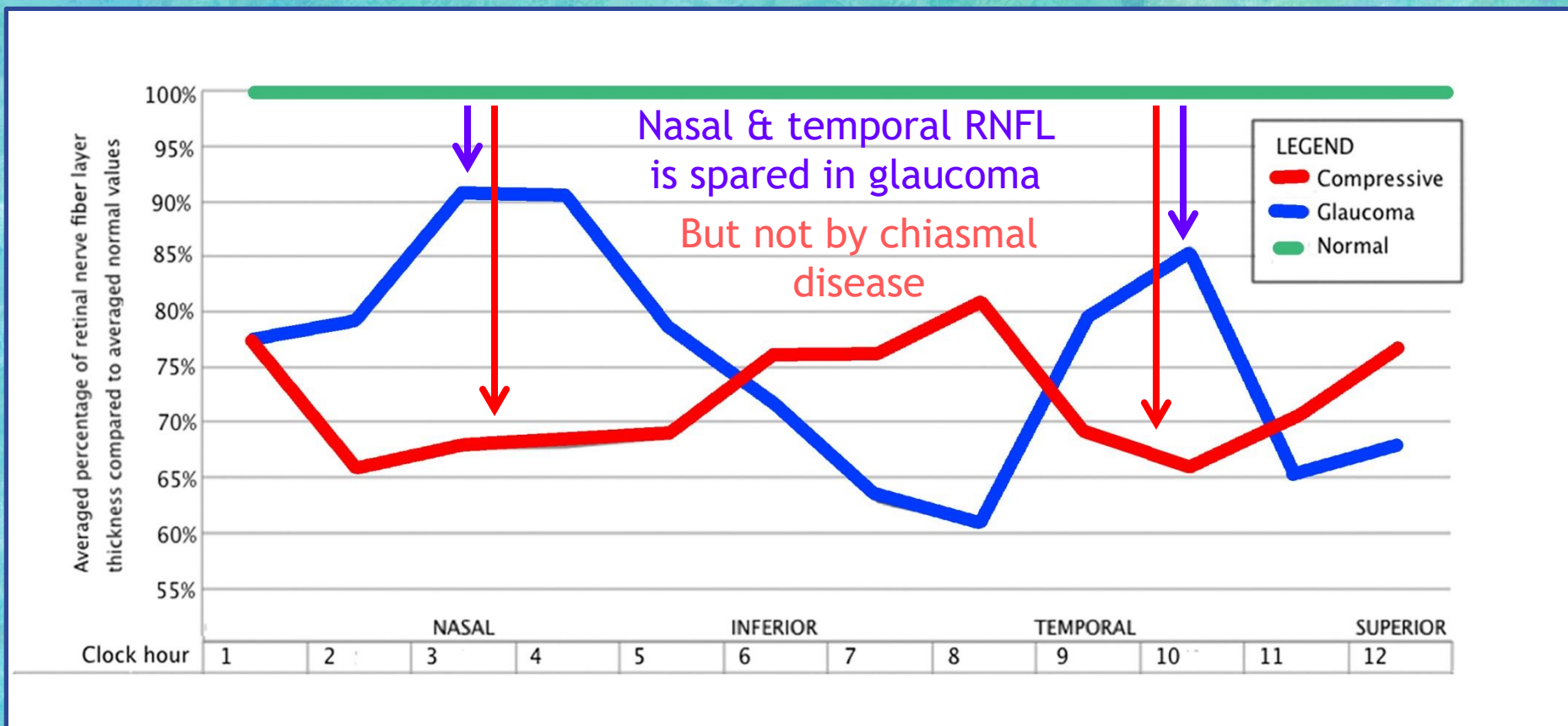
- Pituitary adenoma is an important cause of non-glaucomatous optic disc cupping
 - Compression of the optic nerve and chiasm can produce enlargement of the cup
- Chiasmal compression preferentially affects the nasal and temporal rim, resulting in a horizontal **band of pallor** (“bow-tie”)
- Chiasmal lesions do not cause papilledema



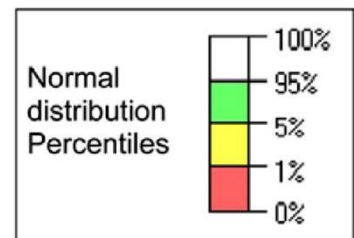
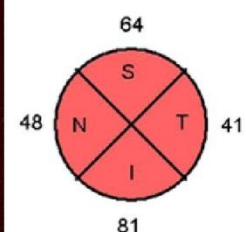
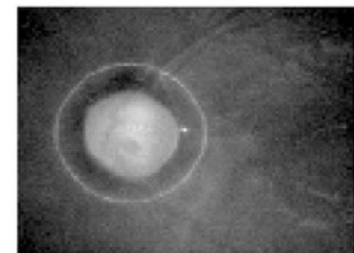
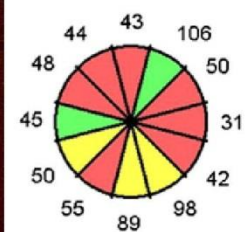
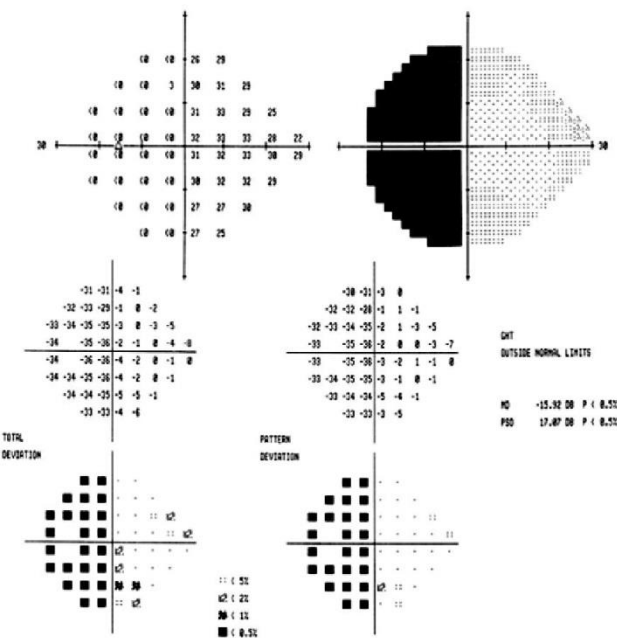
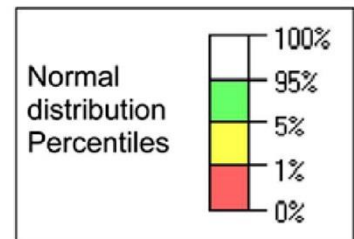
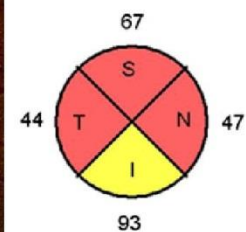
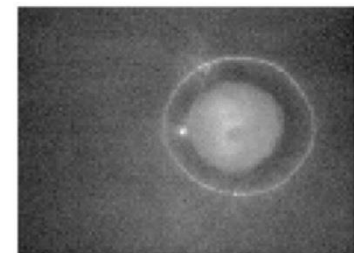
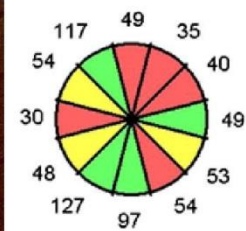
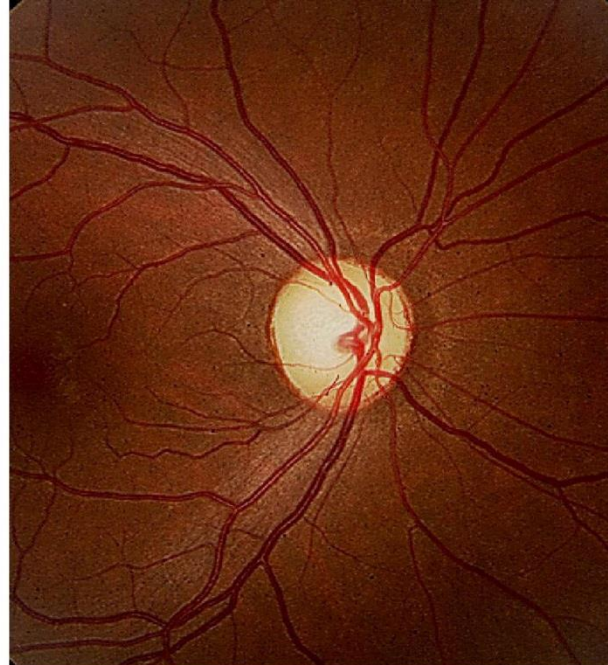
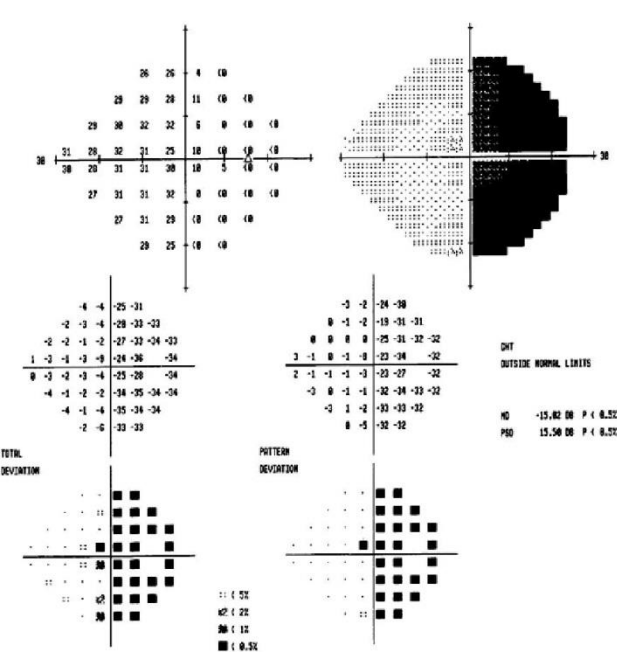


OCT Findings

- Chiasmal compression results in RNFL and GCC thinning on OCT
 - Unlike glaucoma, RNFL thinning is fairly uniform in all meridians
 - Thinning of the GCC (often in a binasal pattern) may be detected before RNFL loss
 - *More severe RNFL/GCC loss is associated with less VF recovery following tumor excision*

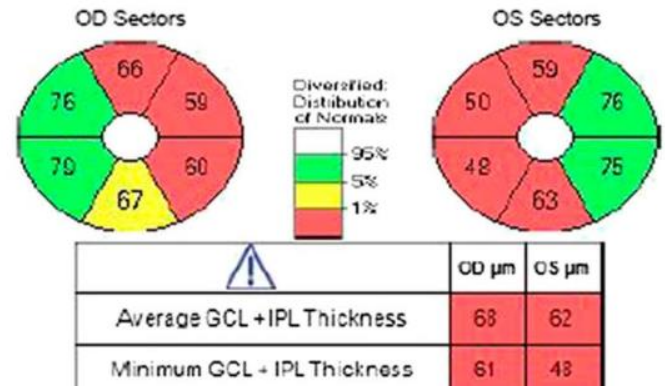
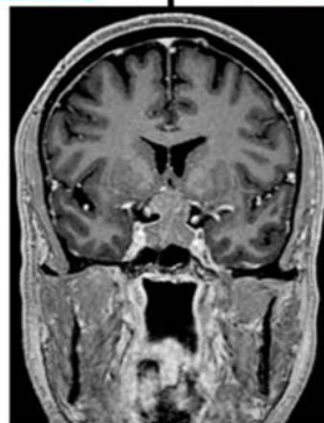
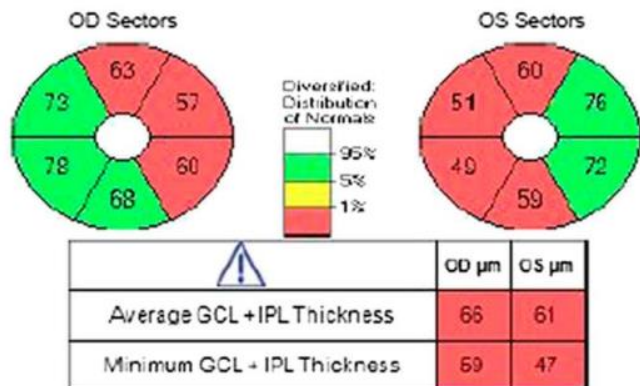
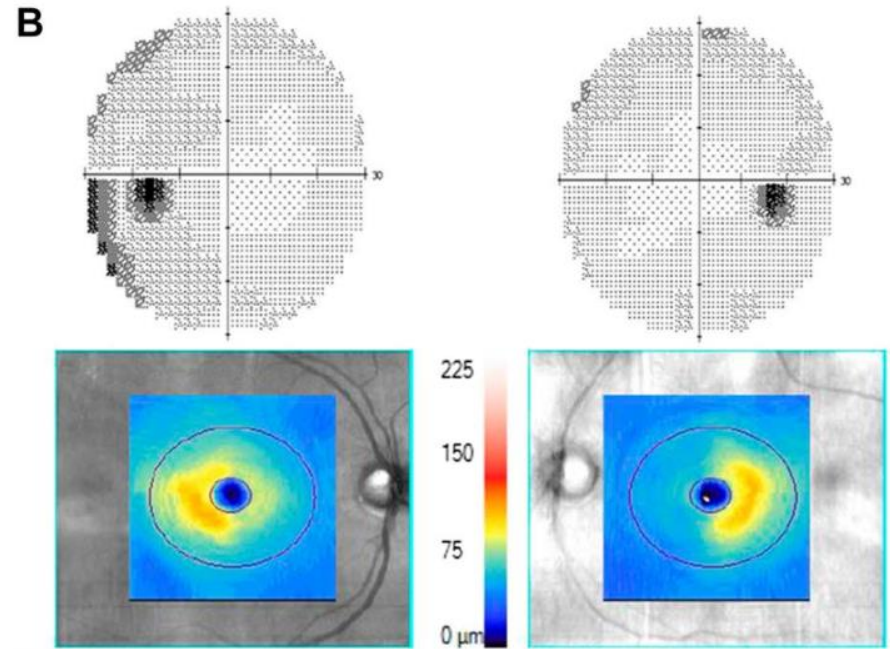
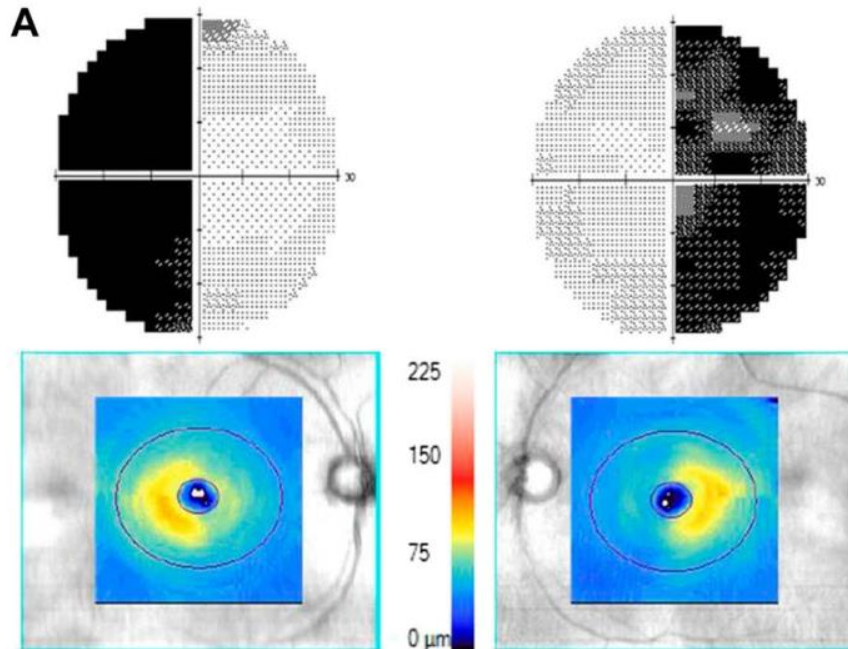


There is relative sparing of the nasal and temporal RNFL with glaucomatous optic neuropathy but not with chiasmal compression.



Pre-Op

Post-Op



OCT Findings

- RNFL analysis is less sensitive than perimetry in detecting chiasmal compression
- GCC thinning is more sensitive than perimetry in detecting chiasmal compression
- **OCT can detect macular ganglion cell complex (GCC) thinning before visual field loss occurs**

Optical coherence tomography retinal ganglion cell complex analysis for the detection of early chiasmal compression

Richard J. Blanch^{1,2,3} · Jonathan A. Micieli¹ · Nelson M. Oyesiku⁴ · Nancy J. Newman^{1,4,5} · Valérie Biousse^{1,5}

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Abstract

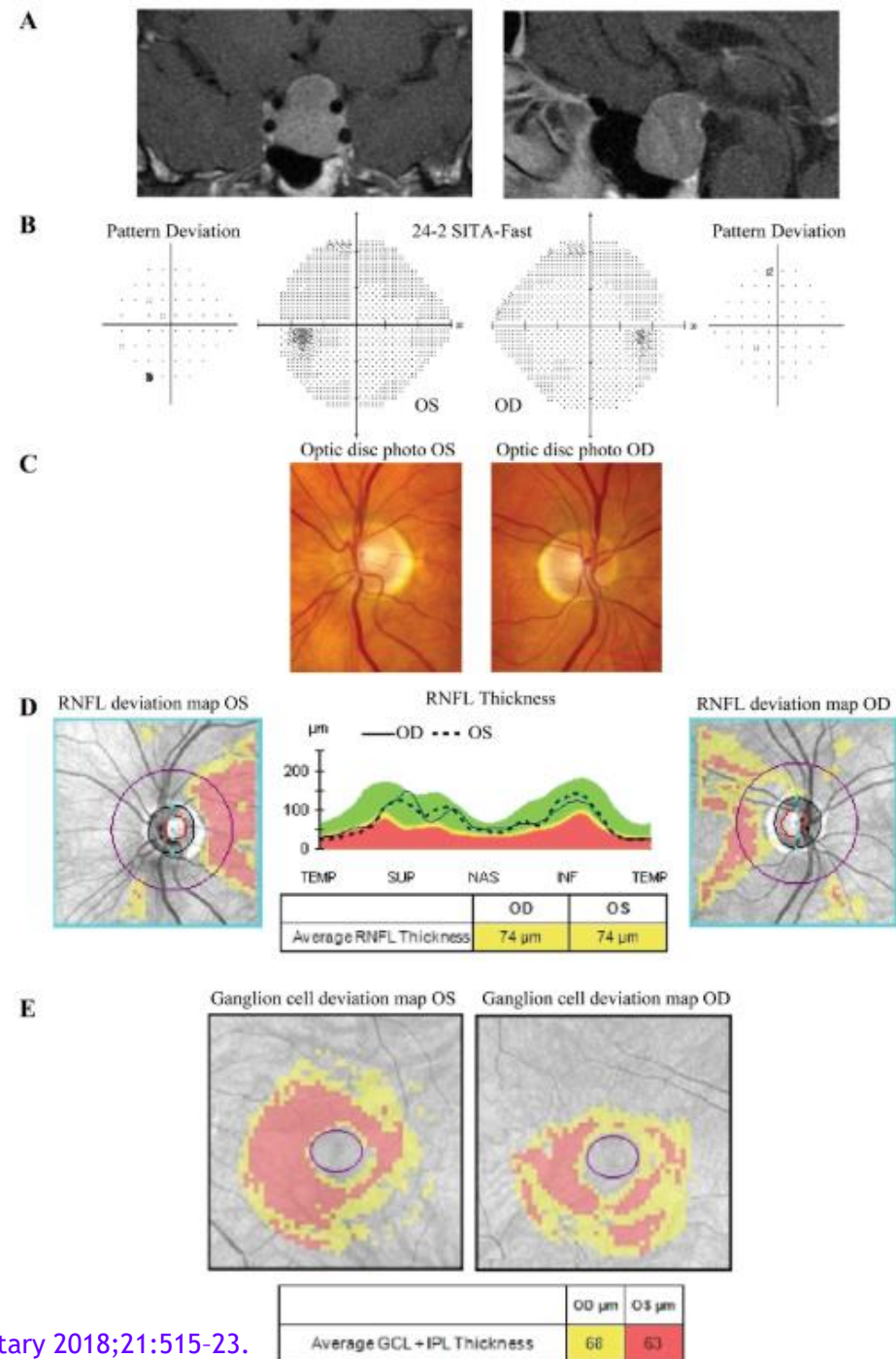
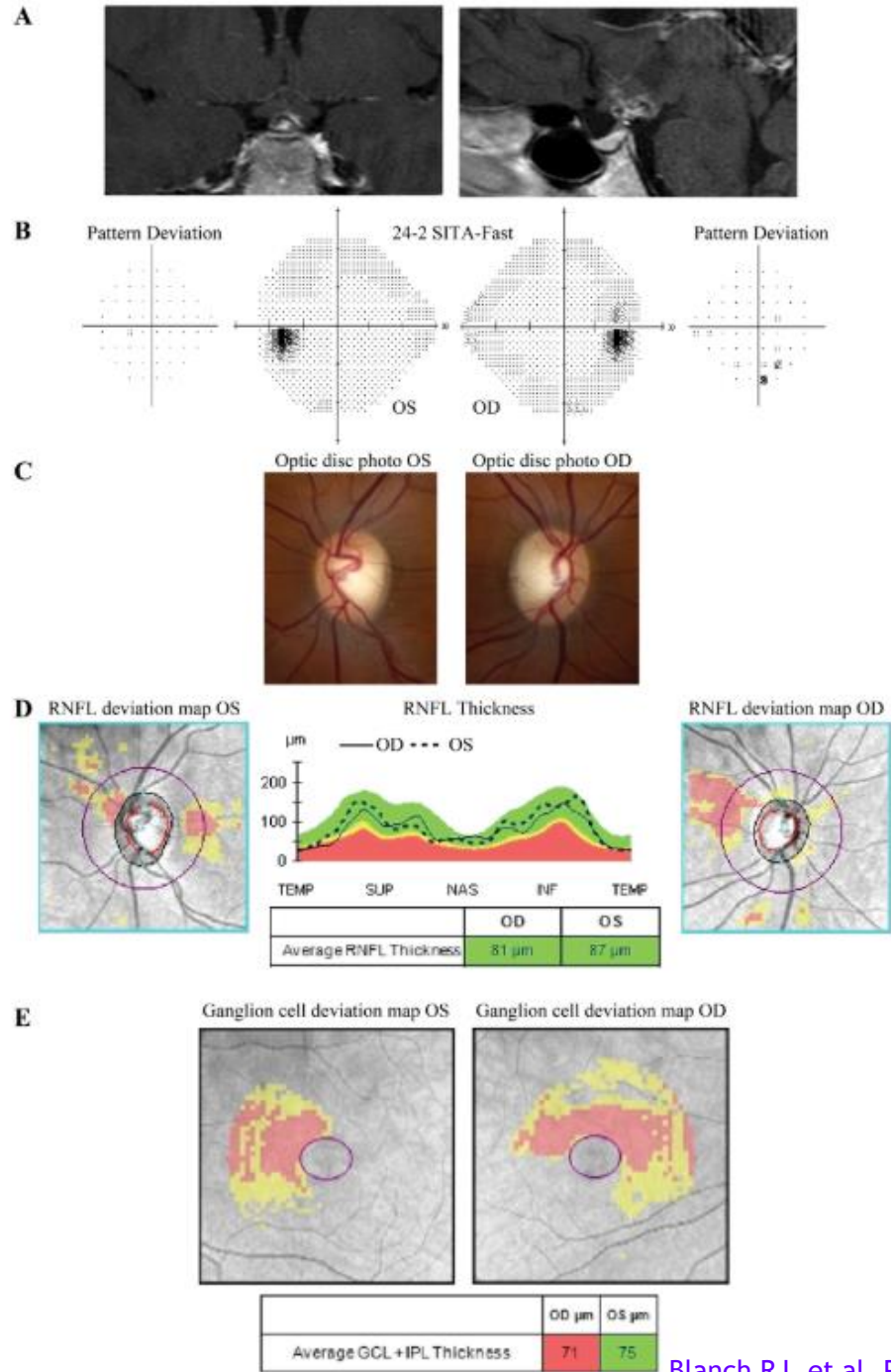
Purpose To report patients with sellar tumors and chiasmal compression with normal visual fields, who demonstrate damage to the retinal nerve fiber layer (RNFL) and ganglion cell complex (GCC) on optical coherence tomography (OCT).

Methods Seven patients with sellar tumors causing mass effect on the optic chiasm without definite visual field defect, but abnormal GCC are described. GCC/RNFL analyses using Cirrus-OCT were classified into centiles based on the manufacturer's reference range.

Results In seven patients with radiologic compression of the chiasm by a sellar tumor, OCT-GCC thickness detected compressive chiasmopathy before visual defects became apparent on standard automated visual field testing. Without OCT, our patients would have been labelled as having normal visual function and no evidence of compressive chiasmopathy. With only OCT-RNFL analysis, 3/7 patients would still have been labelled as having no compression of the anterior visual pathways.

Conclusions These patients show that OCT-GCC analysis is more sensitive than visual field testing with standard automated perimetry in the detection of compressive chiasmopathy or optic neuropathy. These cases and previous studies suggest that OCT-GCC analysis may be used in addition to visual field testing to evaluate patients with lesions compressing the chiasm.

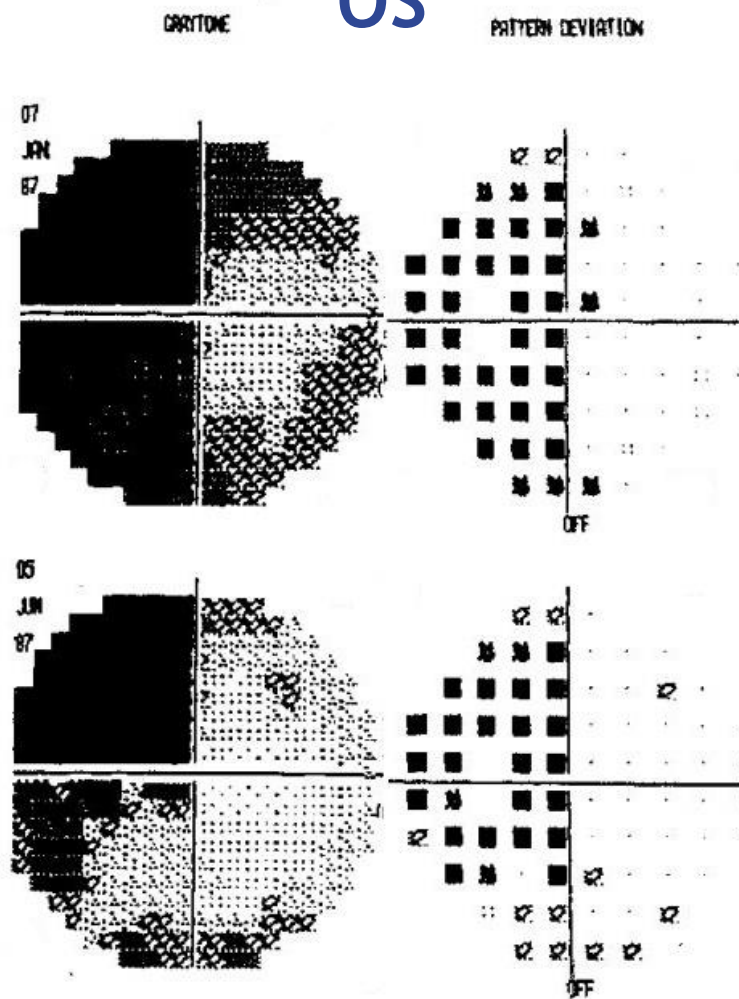
OCT can detect chiasmal compression
before VF loss occurs



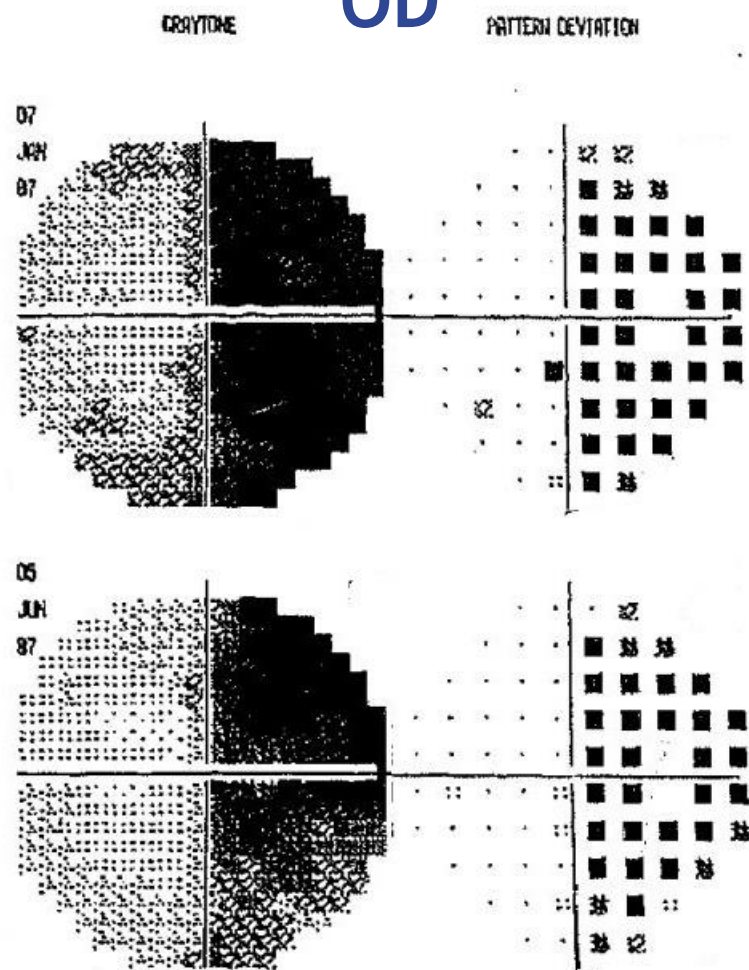
Prognosis

- Visual outcome following pituitary adenoma surgery is highly variable
 - RNFL/GCC thickness, duration of symptoms, disc pallor, and age influence recovery
 - **Most patients will experience some recovery**, and many will experience complete resolution of VF defects
 - Most of the recovery occurs within the first 3 months following surgery

OS



OD



Chiasmal Syndrome

SYMPTOMS

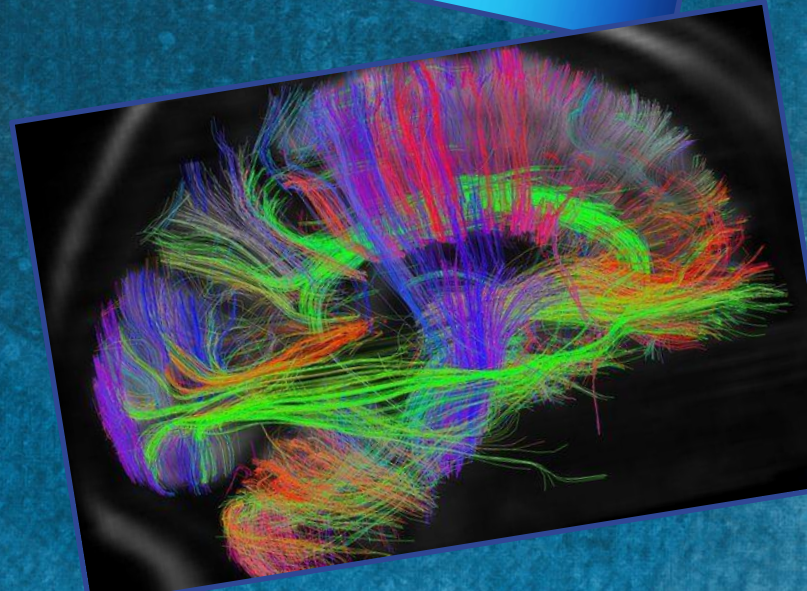
- Headache
- Visual loss
- Diplopia
- Loss of depth perception
- Endocrine dysfunction

SIGNS

- Visual field defects
- Optic disc pallor and cupping
- OCT abnormalities
- Oculomotor pareses
- Nystagmus
- Cerebrospinal fluid rhinorrhea

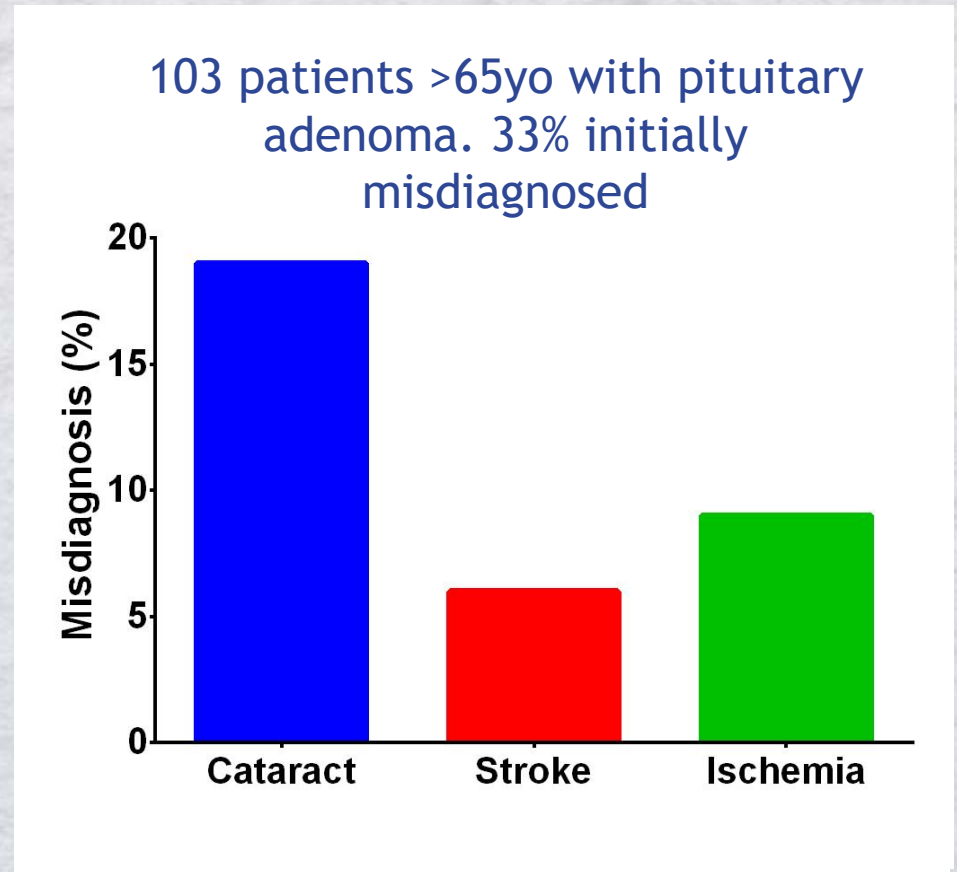
Chiasmal Syndrome

- Anatomy Review
- All About Pituitary Adenomas
- Clinical Features of Chiasmal Syndrome
- Clinical Pearls
 - Red Flag Warning Signs
 - Chiasmal Work-up



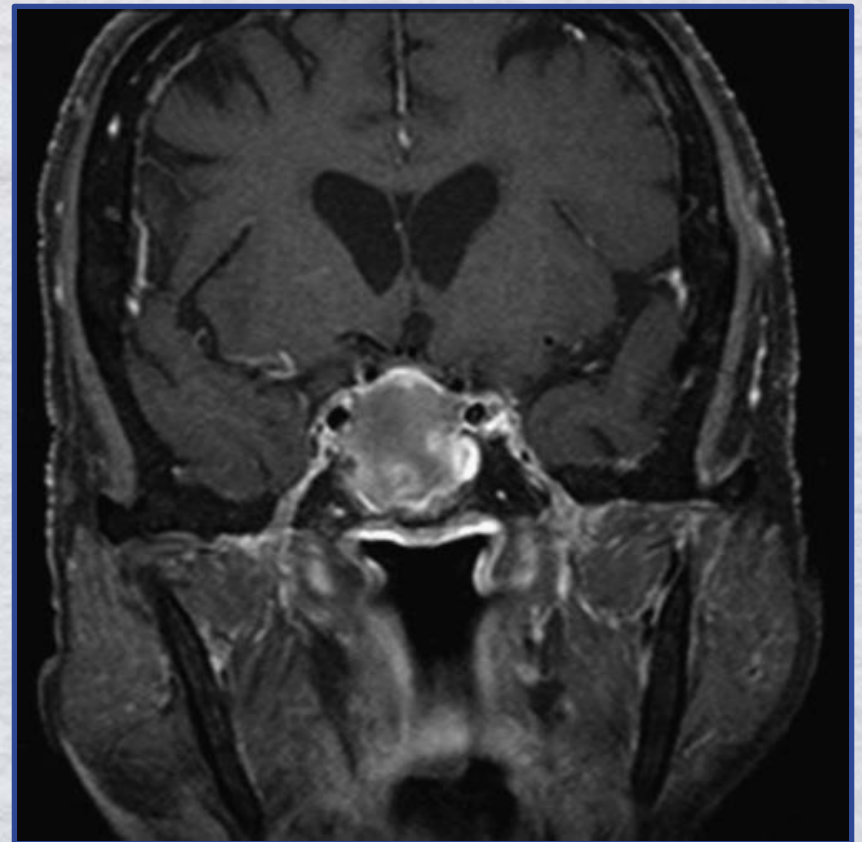
Red Flags

- Headaches
- Normal tension glaucoma
- Unexplained poor visual acuity
- Poor depth perception/stereo
- Intermittent diplopia



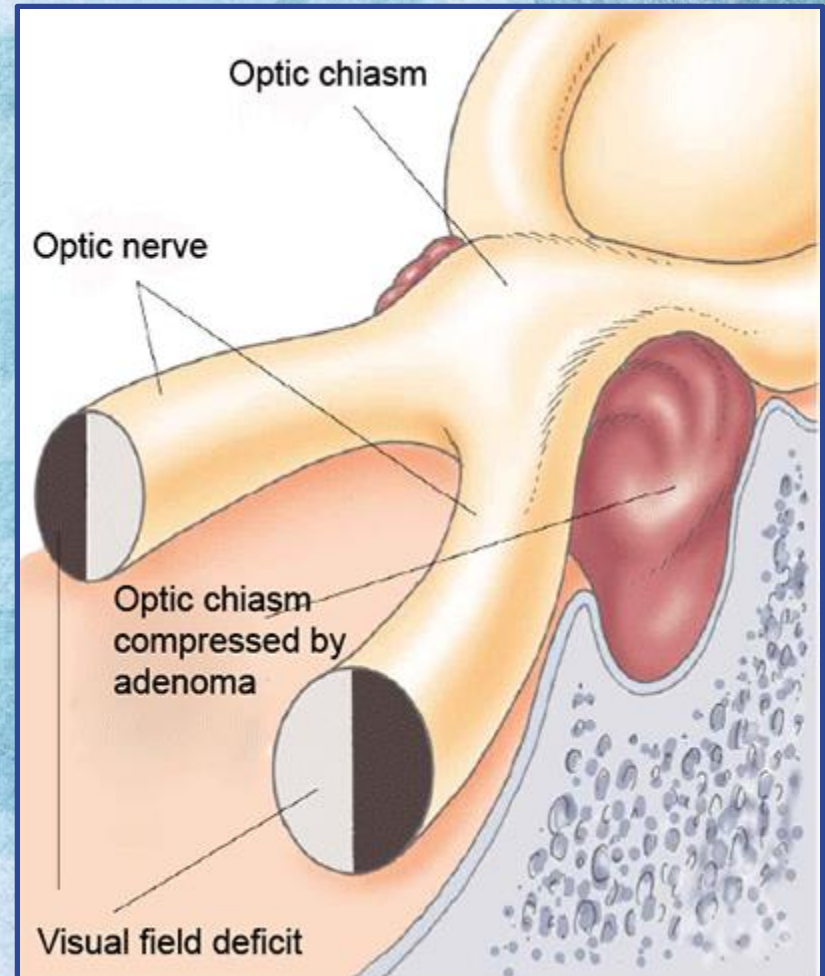
Red Flags

- When should I order an MRI on my NTG suspects?
 - Age younger than 50 years
 - VA less than 20/40
 - Optic nerve pallor
 - **Vertically aligned visual field defects**
 - Focal neurologic signs (eg. **headache**)



Chiasmal Work-up

- Headache history
- Confrontation VF
- Stereopsis
- Pupils & color vision
- Ophthalmoscopy & **OCT**
- Perimetry
- MRI



Headache

- Specifically inquire about any new or unusual headaches
- **Positive headache history increases risk** of pituitary tumor
 - Regardless of severity or nature of headache
- Absence of headache does not rule out tumor



Confrontation VF

FCCF testing of patient with left hemianopia



Neuro-Ophthalmology Diagnosis and Management. 2nd Ed. 2010

Stereopsis

- Stereo tests are a simple, easy, and quick screening test for chiasmal disease
- Mild, incomplete bitemporal hemianopia can result in loss of stereopsis that is detectable with routine stereo tests
- **Fail: <6/9**

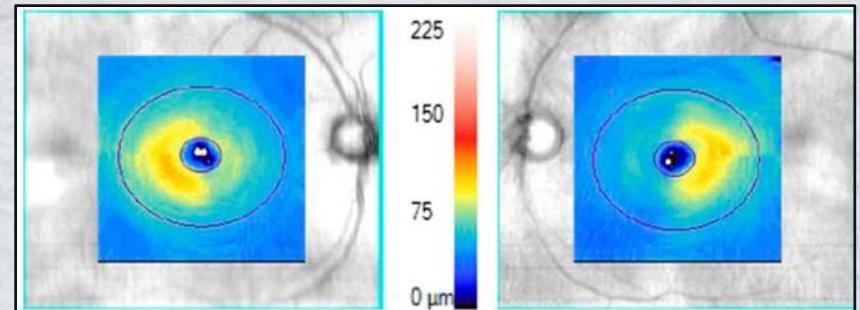


Pupils & Color Vision

- An APD suggests a lesion other than pituitary adenoma
- Color vision is a more sensitive indicator of afferent system damage than VA
 - Mild-moderate VA loss with significant color vision loss = optic neuropathy is likely
 - Color vision preserved and VA is poor = optic neuropathy is unlikely

Ophthalmoscopy & OCT

- Early
 - GCC abnormality (nasal loss > temporal loss) despite little or no VF loss
 - Compression impairs axon function prior to ganglion cell death (VF recovery possible)
- Late
 - Bow-tie pallor (nasal & temporal quadrants)
 - Disc cupping
 - Diffuse RNFL thinning w/wo band atrophy

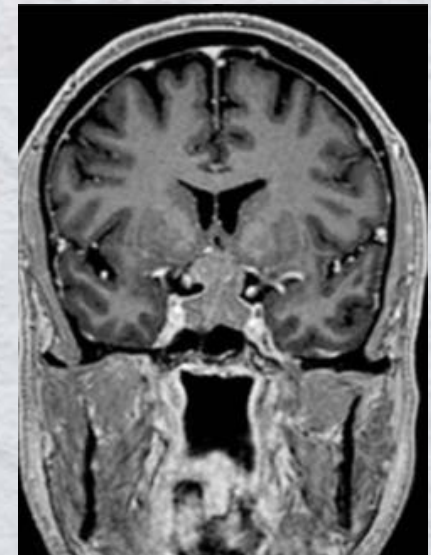


Perimetry

- Perimetry is a key test for detecting chiasmal lesions
- Inadequate VF assessment is the chief cause of misdiagnosis of chiasmal lesions
- Standard automated perimetry is the “gold standard”
 - SAP can find defects missed with FCCF

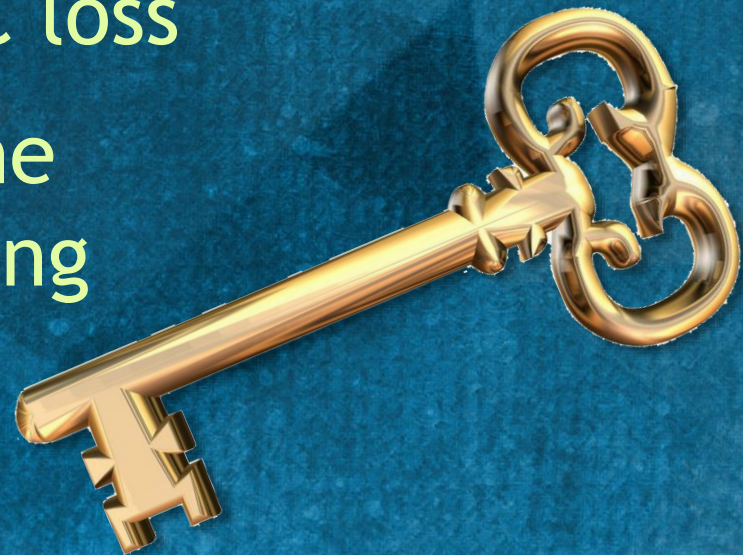
MRI

- **Required to confirm diagnosis** and plan treatment
- Order MRI of optic chiasm with and without contrast
- The exploration protocol is with T1-weighted sagittal sections, then T1- and T2-weighted coronal sections with and without contrast



Key Points

- Chiasmal syndrome is a subtle, easily missed condition
- Headache and BV complaints are common
- Be suspicious of all NTG suspects
- Look for binasal OCT GCC loss
- Threshold perimetry is the test of choice for detecting chiasmal syndrome



Thank you!