



CHIASMAL AND RETROCHIASMAL LESIONS



Eye Learn
All about the Eye

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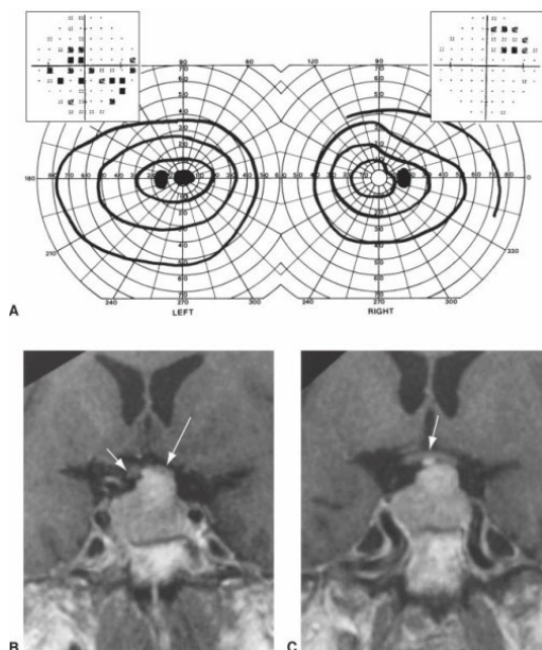
1. Causes and ocular manifestations of chiasmal lesions. 3+7 (J2018)
2. Draw a diagram of visual pathway and show visual defects in craniopharyngioma and occipital lobe lesion. J2013
3. Describe anatomy and lesions of optic tracts, chiasma and optic radiations. J2014
4. Define scotoma. How do you differentiate between positive and negative scotoma. Discuss the approach to diagnosis in a patient presenting with left hemianopia. J2012 (optic tract + radiation lesion on right)

Chiasmal Lesions

- i. With the segregation of nasal and temporal retinal fibers at the chiasm, visual field loss due to chiasmal and retrochiasmal lesions is characterized by temporal defects that align along the vertical meridian.
- ii. In chiasmal syndromes, the optic discs may appear normal initially, even with significant visual field loss.
- iii. Early on, peripapillary retinal NFL dropout and mild disc pallor develop.
- iv. With progressive damage, the optic discs show typical atrophy, often in the temporal portion of the disc.
- v. Cupping of the disc may or may not increase.
- vi. A tumor compressing the chiasm almost never produces optic disc edema.
- vii. Parasellar lesions that involve the chiasm—whether compressing or infiltrating this area—produce gradually progressive, bilateral, often asymmetric vision loss.
- viii. The peripheral (temporal) visual fields usually are involved first.
- ix. In any case of bilateral visual field loss, the clinician must carefully evaluate perimetry testing results for respect of the vertical midline.
- x. Any of the variations on bitemporal visual field loss may occur.
- xi. An affected optic nerve may produce more central loss, with impaired visual acuity, dyschromatopsia, and an RAPD on the affected side.
- xii. Markedly asymmetric visual field loss without direct optic nerve damage may also produce an RAPD.

Visual Field Loss Patterns

- Lesions that injure an optic nerve at its junction with the optic chiasm produce “junctional scotomas.”
- Diminished visual acuity and central visual field loss occur in the ipsilateral eye, and a temporal hemianopia develops in the opposite eye.



A, Visual field patterns from Goldmann perimeter testing and Humphrey 30-2 program testing (insets). Note the central scotoma in the patient's left eye along with the superotemporal depression in his right eye.

B,C, Postcontrast, T1-weighted (TR = 650 ms, TE = 14 ms) MRI scans using a slice thickness of 3 mm.

B, Coronal image of a section in front of the optic chiasm showing a tumor compressing the prechiasmatic segment of the left optic nerve (long arrow) but not the right optic nerve (short arrow).

C, Coronal image at the level of the optic chiasm showing minimal rostral displacement (arrow) but no notable direct mass effect.

- A unilateral temporal hemianopia that respects the vertical midline, with no involvement of the visual field in the opposite eye, can also indicate a chiasmal abnormality.
- Presumably, the mass compresses only the crossing nasal fibers from 1 eye.
- The most common visual field defect of chiasmal compression is a bitemporal hemianopia.
- These defects may appear relative or complete.
- They may involve the paracentral temporal field alone, and visual acuity is often unaffected until late.

Etiology of Chiasmal Disorders

Extrinsic lesions affecting the chiasm

- The most common lesions producing the chiasmal syndrome include
 - i. Pituitary adenoma
 - ii. Parasellar meningioma,
 - iii. Craniopharyngioma,
 - iv. Parasellar internal carotid artery aneurysm.
 - v. Other CNS mass lesions can produce third-ventricle dilation and secondary posterior chiasmal compression.

1. Pituitary adenomas

- ✓ Are the most common cause of chiasmal compression and may occur at any adult age; they are rare in childhood.
- ✓ Patients with non-secreting tumors typically present with vision loss, their tumors having reached a relatively large size without causing other symptoms.
- ✓ Hormonally active tumors, however, are often detected before vision loss because of systemic symptoms.
- ✓ Pituitary tumors may enlarge during pregnancy and produce chiasmal compression.
- Acute hemorrhage or infarction of the pituitary tumor, known as **pituitary apoplexy**,
 - ✓ It is a potentially life-threatening event heralded by severe headache, nausea, and altered consciousness.

- ✓ Neuro-ophthalmic findings include diplopia and loss of vision or visual field
- Sudden expansion of the tumor into the adjacent **cavernous sinuses**
 - ✓ Can cause dysfunction of CNS III, IV, V, and VI, with CN III the most commonly affected.
- **Superior extension** causes
 - ✓ Visual field loss
 - ✓ May cause central visual loss to no light perception vision.
- **Extravasation of blood into the subarachnoid space** causes numerous symptoms, including a
 - ✓ Decreased level of consciousness
 - ✓ Vasospasm with secondary stroke.
- The acute **endocrine** abnormalities may lead to numerous complications, including adrenal crisis.
- Therefore, recognition of pituitary apoplexy is crucial so treatment is initiated emergently.
- **Treatment** includes immediate administration of corticosteroids, surgical decompression of the sella, and appropriate supportive measures.
- Some authorities recommend conservative management when neuro-ophthalmic signs are absent or mild.

2. Parasellar meningiomas occur most often in middle-aged women; arise most frequently from the

- i. Tuberculum sella,
 - ii. Planum sphenoidale,
 - iii. Or anterior clinoid;
- Produce asymmetric bitemporal vision loss.
 - Parasellar meningiomas may also enlarge and produce chiasmal compression during pregnancy.

3. Craniopharyngioma

- ✓ Are common in children but may present at any age, with a second incidence peak in adulthood.
- ✓ Often arising superiorly, these tumors more frequently produce inferior bitemporal visual field loss.

4. Internal carotid artery aneurysms,

- ✓ Particularly in the supraclinoid region, may produce a markedly asymmetric chiasmal syndrome, with optic nerve compression on the side of the aneurysm.

Treatment

- Therapy of parasellar tumors is complex and depends on
 - ✓ Age of the patient;
 - ✓ The nature,
 - ✓ Location, and
 - ✓ Extent of the tumor;
 - ✓ Its hormonal activity;
 - ✓ And the severity of symptoms.
- **Observation** only, if visual fields are normal
- **Surgery**
 - ✓ Trans-frontal or trans-sphenoidal

- ✓ After surgical resection of the tumor and relief of anterior visual pathway compression, vision recovery is usually rapid (onset of improvement is within 24 hours) and may be dramatic, even with severe vision loss.
- **Medical therapy**
 - ✓ Primarily bromocriptine or cabergoline for prolactin-secreting pituitary tumors
 - ✓ Medical therapy for pituitary adenomas has a slower effect, taking days to weeks, but also produces tumor shrinkage and improved visual function in responsive cases.
- **Radiation therapy**
 - ✓ Either primary or as adjunctive therapy for incompletely resectable tumors
- **Prognosis** is poor if mean retinal nerve fiber layer thickness is less than 75 μm , as revealed on OCT scans.
- **Follow up**
 - ✓ The ophthalmologist's role in the management of parasellar tumors is crucial, in that the first sign of recurrence may be vision loss.
 - ✓ Baseline visual field and visual acuity testing should be performed 2–3 months after treatment and at intervals of 6–12 months thereafter, depending on the course.
 - ✓ Visual acuity and visual fields should be rechecked more often (immediately if necessary) if the patient notes any ongoing change.

Intrinsic lesions affecting the chiasm

- Other, infrequent causes of chiasmal disorders include intrinsic lesions of the chiasm.
 - ✓ Infectious (eg, tuberculosis, Lyme disease) and
 - ✓ Inflammatory (eg, sarcoidosis, multiple sclerosis) causes of chiasmal neuritis
 - ✓ Neoplasms can be either primary (eg, glioma) or secondary (eg, metastasis).
 - ✓ Significant closed-head trauma can injure the chiasm, resulting in a bitemporal hemianopia.
 - ✓ Chiasmal injury may result from parasellar radiation therapy as well.

Retrochiasmal Lesions

- As the fibers course in the retrochiasmal visual pathway (optic tract; lateral geniculate body; and temporal, parietal, and occipital lobe visual radiations), crossed nasal fibers from the contralateral eye and uncrossed temporal fibers from the ipsilateral eye are located together
- Retrochiasmal damage results in homonymous visual field defects that continue to respect the vertical midline.
- As fibers progress from the anterior to the posterior visual pathway, those from corresponding retinal regions of each eye tend to run closer and closer together.
- Historically, authorities have believed that anterior lesions produce dissimilar (incongruous) defects in the corresponding homonymous hemi-fields, whereas more posterior damage results in progressively more similar (congruous) defects as lesions approach the occipital lobes.
- However, this “rule” of congruity has been called into question.
- In a series of 538 patients, 59% of optic radiation lesions and 50% of optic tract lesions caused congruent homonymous hemianopia.
- Therefore, although a highly congruous homonymous hemianopia might be expected to reflect occipital disease, the possibility of a more anterior lesion should not be excluded.

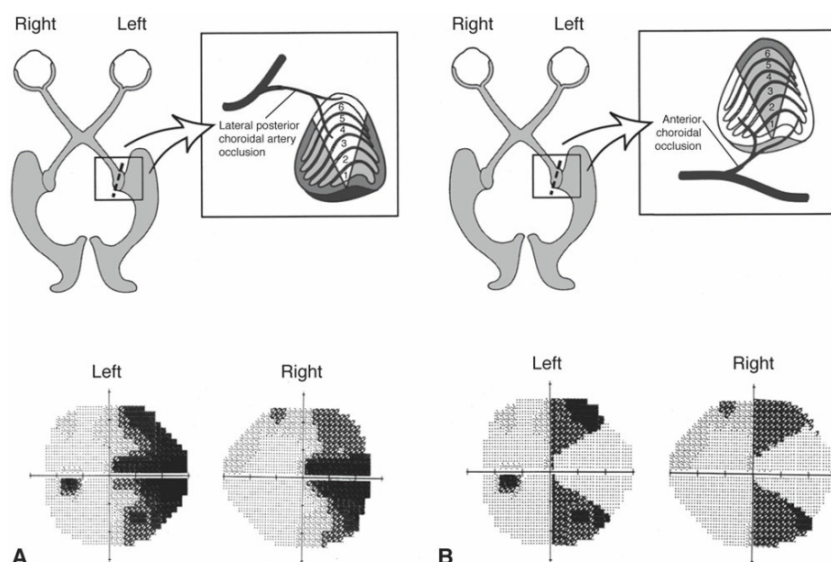
- Lesions severe enough to produce complete hemianopic defects may occur at any anteroposterior retrochiasmal location; such defects do not help localize lesions from the chiasm through the occipital cortex.
- **Stroke is the most common cause of homonymous hemianopias, followed by traumatic brain injury and tumor.**

Optic Tract

- Lesions of the optic tract produce incongruous homonymous defects in the hemi-fields contralateral to the affected optic tract
- Damage to the optic tract most commonly results from mass lesions such as
 - ✓ Aneurysms.
 - ✓ Inflammatory and demyelinating lesions occur occasionally.
 - ✓ Ischemic lesions of the tract are uncommon
 - ✓ May follow surgical disruption of the anterior choroidal artery.
- Because the fibers involved are primary neurons in the visual pathway (retinal ganglion cells), the incongruous homonymous hemianopic visual field loss is accompanied by other findings that make up the optic tract syndrome:
 - “Bow-tie” optic atrophy.
 - Because the optic tract involves crossed fibers from the contralateral eye, the corresponding atrophy of crossed retinal fibers (those nasal to the macula) involves the papillomacular fibers and the nasal radiating fibers in the contralateral eye, causing atrophy in the corresponding nasal and temporal horizontal portions of the disc (“band” or “bow-tie” atrophy)
 - Atrophy in the ipsilateral eye involves only the arcuate temporal bundles, which enter the disc at the superior and inferior poles.
 - Mild RAPD in the contralateral eye.
 - This finding stems from the presence of more crossed than uncrossed pupillary fibers in the tract, causing more pupillary fibers from the contralateral eye to be damaged by a tract lesion.

Lateral Geniculate Body

- The lateral geniculate body (LGB) is a highly organized and layered retinotopic structure; lesions in this region therefore can give highly localizing visual field defects.
 - ✓ A very congruous **horizontal sectoranopia** results from damage in the distribution of the posterolateral choroidal artery, a branch of the posterior cerebral artery.
 - ✓ Loss of the upper and lower homonymous quadrants (also called “**quadruple sectoranopia**”) with preservation of a horizontal wedge occurs with disruption of the anterior choroidal artery, a branch of the middle cerebral artery
- These visual field defects respect the vertical meridian, unlike the uncommon wedge defect observed in glaucoma.
- Very incongruous homonymous hemianopias can also occur with lesions of the LGB.
- Sectoral optic atrophy occurs with LGB lesions, and, in rare cases, bilateral LGB lesions cause blindness.



(A) a central wedge-shaped homonymous sectoranopia caused by lateral posterior choroidal artery occlusion, and

(B) a loss of the upper and lower homonymous quadrants, with preservation of the

Temporal Lobe

- Inferior visual fibers course from the LGB anteriorly in the Meyer loop of the temporal lobe (approximately 2.5 cm from the anterior tip of the temporal lobe).
- Superior fibers tend to course more directly posteriorly in the parietal lobe.
- Lesions affecting the Meyer loop thus produce superior, incongruous, homonymous defects contralateral to the lesion, which spare fixation (so called pie in the sky defects)
- Damage to the temporal lobe anterior to the Meyer loop does not cause visual field loss.
- Lesions affecting the radiations posterior to the loop produce homonymous hemianopic defects extending inferiorly.
- **Tumors** within the temporal lobe are a common cause of visual field loss
- Neurologic findings of temporal lobe lesions include seizure activity, including olfactory, and formed visual hallucinations.
- Surgical excision of seizure foci in the temporal lobes may lead to visual field defects.

Parietal Lobe

- Lesions of the parietal lobe, which usually result from **stroke**, tend to involve superior fibers first, resulting in contralateral inferior homonymous hemianopic defects.
- More extensive lesions involve the superior visual fields but remain denser inferiorly.
- Parietal lobe syndromes encompass a wide variety of other neurologic effects, including perceptual problems (agnosia) and apraxia.
- Lesions of the dominant parietal lobe cause **Gerstmann syndrome**, a combination of acalculia, agraphia, finger agnosia, and left-right confusion.
- Lesions in the non-dominant parietal lobe can produce contralateral neglect.
- Damage to pursuit pathways that converge in the posterior parietal lobes (near the visual radiations) may cause abnormalities in optokinetic nystagmus (OKN).
- The examiner elicits the impaired OKN response by moving targets toward the lesion, inducing attempts to use the damaged pursuit pathway.
- Thus, a patient with a homonymous hemianopia due to a parietal lobe lesion will have a reduced OKN response with the target moving toward the affected side, whereas a homonymous hemianopia due to a lesion of the optic tract or occipital lobe will yield an intact OKN response.

- This response was once thought to indicate a mass lesion rather than a vascular lesion but is more likely related to the extent of the lesion.
- Asymmetry in the pursuit system likely indicates involvement of area V5 or MT.
- Homonymous hemianopia should prompt OKN testing; only rarely do occipital lobe lesions cause such abnormalities.

Occipital Lobe

- As fibers approach the occipital lobes, congruity becomes more important.
- Central fibers become separate from peripheral fibers, the central ones coursing to the occipital tip and the peripheral ones to the anteromedial cortex.
- Because of the disparity in crossed versus uncrossed fibers, some of the peripheral nasal fibers leading to the anteromedial region are not matched with corresponding uncrossed fibers; they subserve a monocular “temporal crescent” of visual field in the far periphery (60°–90°).
- Finally, fibers localize within the occipital cortex superior and inferior to the calcarine fissure.
- Thus, visual field defects from occipital lobe lesions may have the following **characteristics** in the hemi-fields contralateral to the lesion:
 - i. Congruous homonymous hemianopia, possibly sparing the fixational region
 - ii. A monocular defect of the temporal crescent involving only the most anterior portion of the occipital lobe
 - iii. Homonymous lesion sparing the temporal crescent in the eye contralateral to the lesion ; Goldmann perimetry testing is required
 - iv. Homonymous hemianopia that respects both the vertical and horizontal meridians
- Most occipital lobe lesions encountered by ophthalmologists result from **stroke** and cause no other neurologic deficits.
- **A macula-sparing homonymous hemianopia** suggests a **stroke** involving the portion of the primary visual cortex supplied by the posterior cerebral artery.
- The tip of the occipital lobe receives a dual blood supply from the middle cerebral artery and the posterior cerebral artery.
- Occlusion of the posterior cerebral artery damages the primary visual cortex except for the region representing the macula at the posterior tip of the occipital lobe, which remains perfused by the middle cerebral artery.
- Systemic hypoperfusion often damages the occipital tip because the tip sits in a watershed area supplied by the endarterial branches of the posterior and middle cerebral artery systems.
- This highly vulnerable region may be the only injured area, resulting in **homonymous paracentral hemianopic scotomata**
- Such scotomata occur most commonly after surgery or other episodes involving blood loss with severe hypotension.
- **Cortical blindness** results from bilateral occipital lobe destruction.
- Normal pupillary responses and optic nerve appearance distinguish cortical blindness from total blindness caused by bilateral pre-chiasmal or chiasmal lesions.
- **Anton syndrome** (denial of blindness), though classically associated with cortical blindness, can be due to a lesion at any level of the visual system severe enough to cause blindness.
- Bilateral occipital lobe lesions occasionally permit some residual visual function.
- Disturbances of the primary visual cortex may also produce unformed visual hallucinations from tumors, migraine, or drugs.

- Formed hallucinations are usually attributed to lesions of the extrastriate cortex or temporal lobe.
- Patients with injury of the occipital cortex sometimes perceive moving targets but not static ones; this **Riddoch phenomenon** may also occur with lesions in other parts of the visual pathway.
- The Riddoch phenomenon probably reflects the fact that cells in the visual system respond better to moving stimuli than to those that are static.

Visual Rehabilitation

- Ophthalmologists are encouraged to recognize the impact of vision loss on a patient's life and advise the patient of the visual rehabilitation options available.
- Patients with homonymous hemianopia or quadrantanopia may benefit by referral to a vision rehabilitation specialist for training in visual scanning techniques, which encourage frequent exploratory saccades toward the blind hemifield.
- Orientation and mobility training may also help the patient manage daily tasks.
- Use of prisms has been suggested to compensate for visual field loss, and research is currently evaluating the effectiveness of these devices.

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