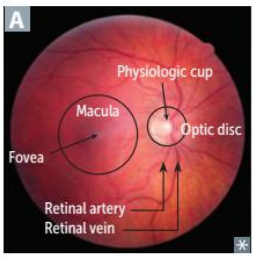
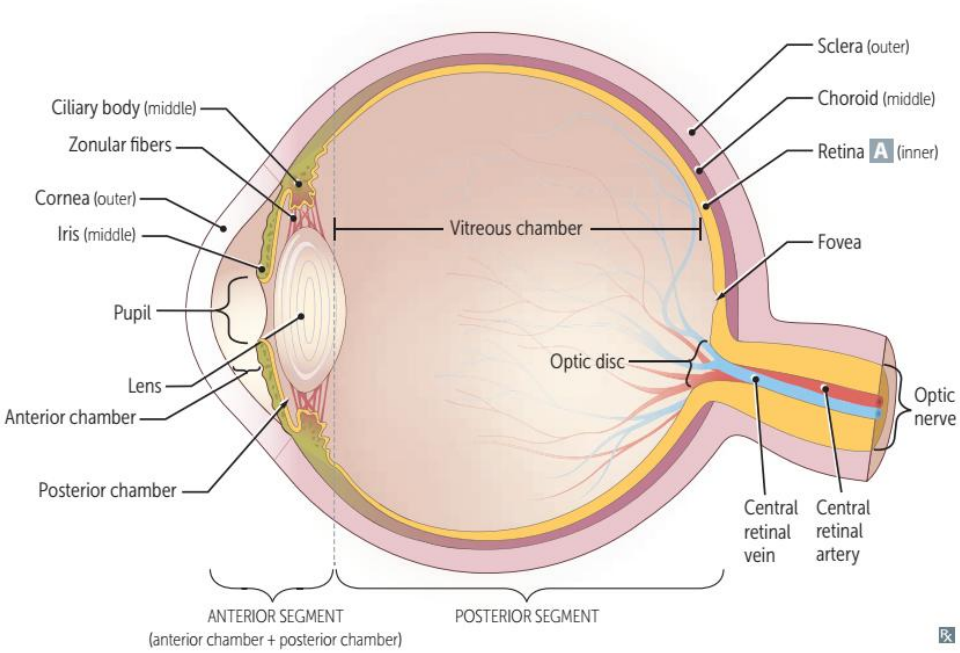
Chapter 12: Special senses

EYE & VISION

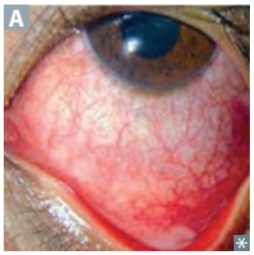
Basic eye anatomy and normal fundus





Diseases of the conjunctiva

Conjunctivitis

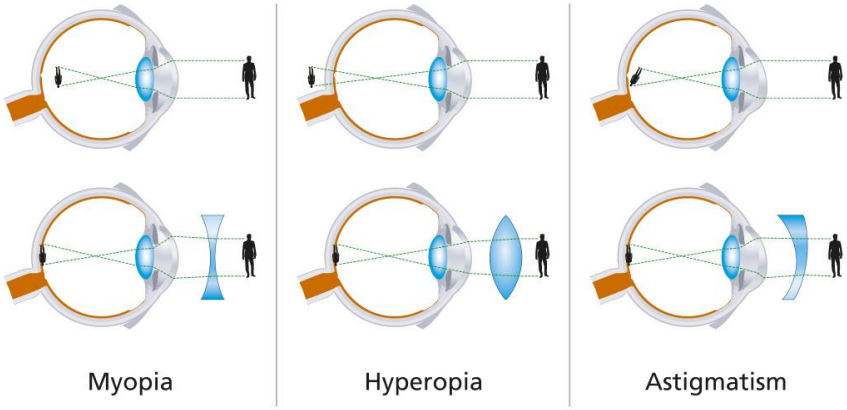


Inflammation of the conjunctiva → red eye A.
Allergic—itchy eyes, bilateral.
Bacterial—pus; treat with antibiotics.
Viral—most common, often adenovirus; sparse mucous discharge, swollen preauricular node, ↑ lacrimation; self-resolving.

*** Treatment:**
- Viral = symptomatic (decongestants)
- Bacterial = Antibiotics + decongestants
- Allergic = Antihistaminics

Diseases of the eye lens

Refractive errors	Common cause of impaired vision, correctable with glasses.
Hyperopia	Also called "farsightedness." Eye too short for refractive power of cornea and lens → light focused behind retina. Correct with convex (converging) lenses.
Myopia	Also called "nearsightedness." Eye too long for refractive power of cornea and lens → light focused in front of retina. Correct with concave (diverging) lens.
Astigmatism	Abnormal curvature of cornea → different refractive power at different axes. Correct with cylindrical lens.

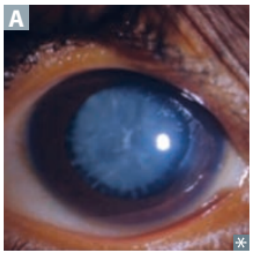


Presbyopia

Aging-related impaired accommodation (focusing on near objects), primarily due to ↓ lens elasticity, changes in lens curvature, ↓ strength of the ciliary muscle. Patients often need "reading glasses" (magnifiers).

* **Accommodation** is increased lens curvature due to contraction of ciliary muscle on bringing an object near to the eye

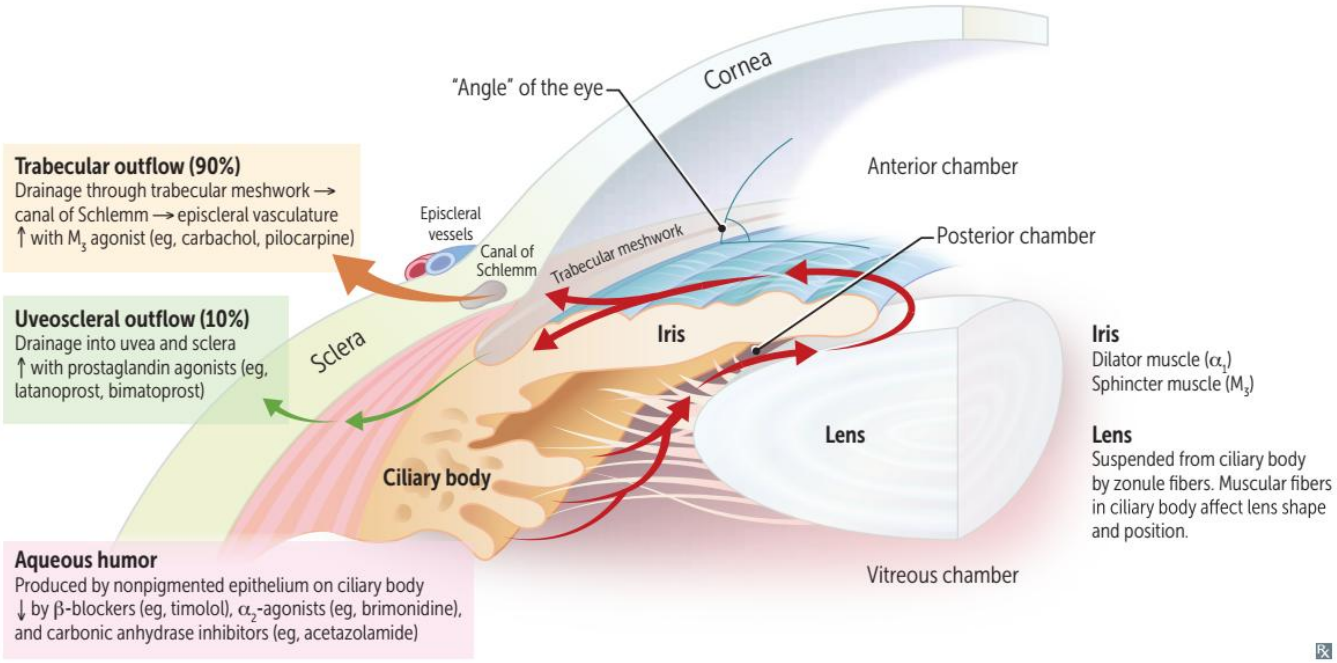
Cataract



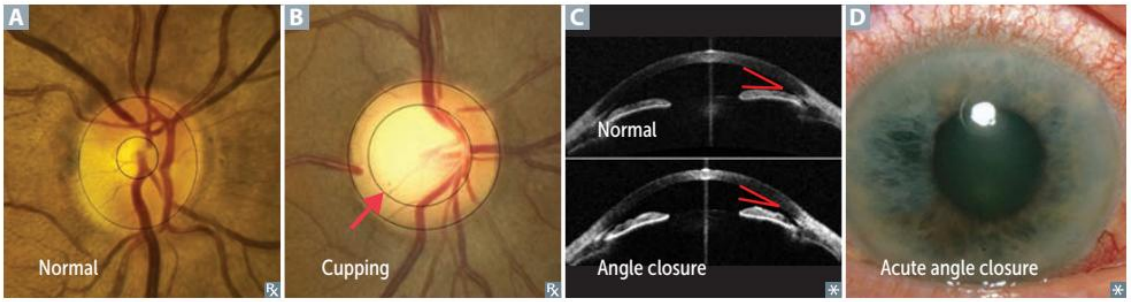
Painless, often bilateral, opacification of lens A, often resulting in glare and ↓ vision, especially at night. Acquired risk factors: ↑ age, smoking, excessive alcohol use, excessive sunlight, prolonged corticosteroid use, diabetes mellitus, trauma, infection. Congenital risk factors: classic galactosemia, galactokinase deficiency, trisomies (13, 18, 21), TORCH infections (eg, rubella), Marfan syndrome, Alport syndrome, myotonic dystrophy, neurofibromatosis 2.

* **Types of cataract =** Senile, Juvenile (congenital), Metabolic (ex. diabetic), Traumatic
* **Treatment =** Surgical replacement of opacified lens with artificial lens

Aqueous humor physiology and pathology



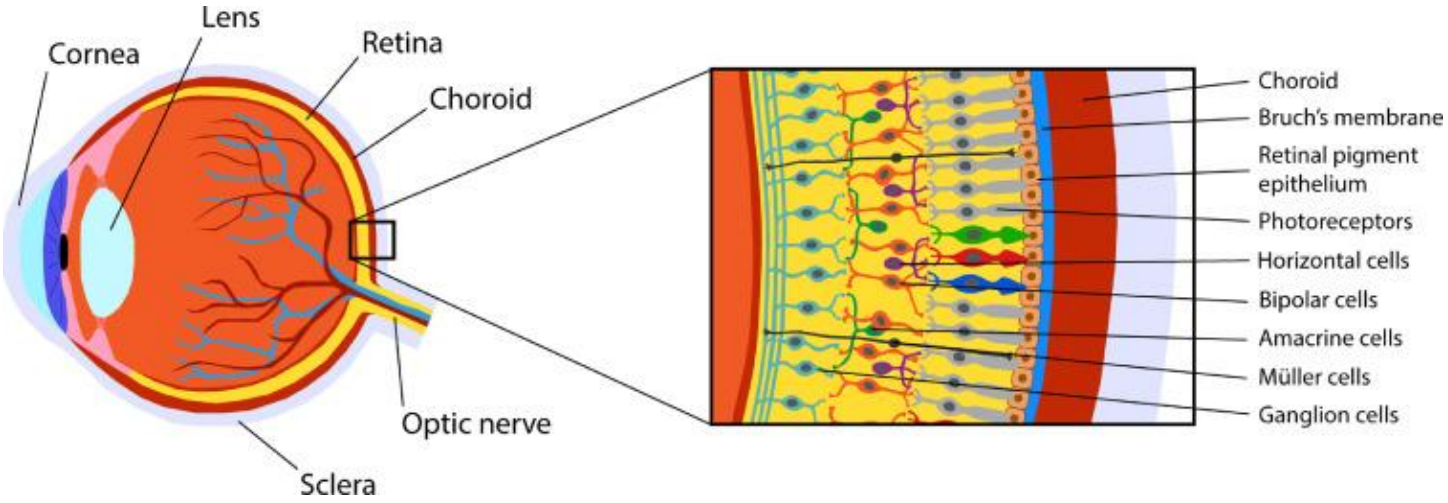
Glaucoma	Optic disc atrophy with characteristic cupping (normal A versus thinning of outer rim of optic nerve head B), usually with elevated intraocular pressure (IOP) and progressive peripheral visual field loss if untreated. Treatment is through pharmacologic or surgical lowering of IOP.
Open-angle glaucoma * The irido-corneal angle is free	Associated with ↑ age, African-American race, family history. Painless, more common in US. Primary—cause unclear. Secondary—blocked trabecular meshwork from WBCs (eg, uveitis), RBCs (eg, vitreous hemorrhage), retinal elements (eg, retinal detachment).
Closed- or narrow-angle glaucoma * The primary usually leads to acute glaucoma while the secondary leads to chronic	Primary—enlargement or anterior movement of lens against central iris (pupil margin) → obstruction of normal aqueous flow through pupil → fluid builds up behind iris, pushing peripheral iris against cornea C and impeding flow through trabecular meshwork. Secondary—hypoxia from retinal disease (eg, diabetes mellitus, vein occlusion) induces vasoproliferation in iris that contracts angle. Chronic closure—often asymptomatic with damage to optic nerve and peripheral vision. Acute closure—true ophthalmic emergency. ↑ IOP pushes iris forward → angle closes abruptly. Very painful, red eye D, sudden vision loss, halos around lights, frontal headache, fixed and mid-dilated pupil, nausea and vomiting. Mydriatic agents contraindicated.



- **Treatment:**

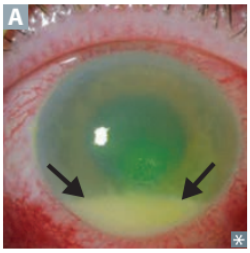
DRUG CLASS	EXAMPLES	MECHANISM	ADVERSE EFFECTS
β-blockers	Timolol, betaxolol, carteolol	↓ aqueous humor synthesis	No pupillary or vision changes
α-agonists	Epinephrine (α_1), apraclonidine, brimonidine (α_2)	↓ aqueous humor synthesis via vasoconstriction (epinephrine) ↓ aqueous humor synthesis (apraclonidine, brimonidine)	Mydriasis (α_1); do not use in closed-angle glaucoma Blurry vision, ocular hyperemia, foreign body sensation, ocular allergic reactions, ocular pruritus
Diuretics	Acetazolamide	↓ aqueous humor synthesis via inhibition of carbonic anhydrase	No pupillary or vision changes
Prostaglandins	Bimatoprost, latanoprost ($PGF_{2\alpha}$)	↑ outflow of aqueous humor via ↓ resistance of flow through uveoscleral pathway	Darkens color of iris (browning), eyelash growth
Cholinomimetics (M_3)	Direct: pilocarpine, carbachol Indirect: physostigmine, echothiophate	↑ outflow of aqueous humor via contraction of ciliary muscle and opening of trabecular meshwork Use pilocarpine in acute angle closure glaucoma—very effective at opening meshwork into canal of Schlemm	Miosis (contraction of pupillary sphincter muscles) and cyclospasm (contraction of ciliary muscle)

Anatomy of uveal tract and retinal layers



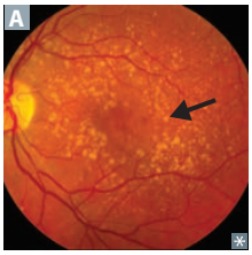
Diseases of the uveal tract and retina

Uveitis



Inflammation of uvea; specific name based on location within affected eye. Anterior uveitis: iritis; posterior uveitis: choroiditis and/or retinitis. May have hypopyon (accumulation of pus in anterior chamber A) or conjunctival redness. Associated with systemic inflammatory disorders (eg, sarcoidosis, rheumatoid arthritis, juvenile idiopathic arthritis, HLA-B27–associated conditions).

Age-related macular degeneration



Degeneration of macula (central area of retina). Causes distortion (metamorphopsia) and eventual loss of central vision (scotomas).

- Dry (nonexudative, > 80%)—Deposition of yellowish extracellular material ("Drusen") in between Bruch membrane and retinal pigment epithelium A with gradual ↓ in vision. Prevent progression with multivitamin and antioxidant supplements.
- Wet (exudative, 10–15%)—rapid loss of vision due to bleeding 2° to choroidal neovascularization. Treat with anti-VEGF (vascular endothelial growth factor) injections (eg, bevacizumab, ranibizumab).

* Wet type can happen on top of proliferative diabetic retinopathy

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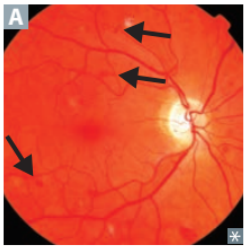
Diabetic retinopathy

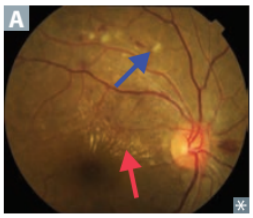


Retinal damage due to chronic hyperglycemia. Two types:

- Nonproliferative—damaged capillaries leak blood → lipids and fluid seep into retina → hemorrhages (arrows in A) and macular edema. Treatment: blood sugar control.
- Proliferative—chronic hypoxia results in new blood vessel formation with resultant traction on retina → retinal detachment. Treatment: anti-VEGF injections, peripheral retinal photocoagulation, surgery.

*** Discussed in details in endocrine pathology (Diabetic complications)**

Hypertensive retinopathy

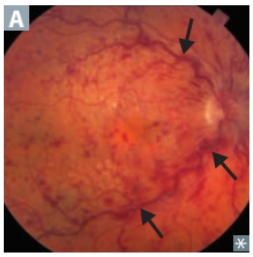


Retinal damage due to chronic uncontrolled HTN.

Flame-shaped retinal hemorrhages, arteriovenous nicking, microaneurysms, macular star (exudate, red arrow in A), cotton-wool spots (blue arrow in A). Presence of papilledema requires immediate lowering of BP.

Associated with ↑ risk of stroke, CAD, kidney disease.

Retinal vein occlusion



Blockage of central or branch retinal vein due to compression from nearby arterial atherosclerosis. Retinal hemorrhage and venous engorgement ("blood and thunder appearance"; arrows in A), edema in affected area.

* **Manifest as** subacute, gradual and progressive vision loss

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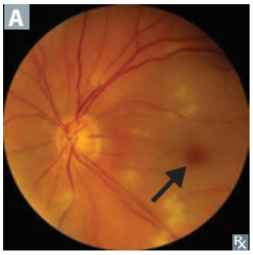
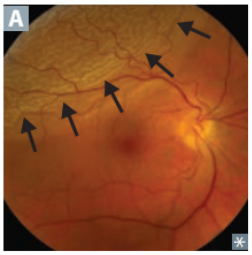
Central retinal artery occlusion



Acute, painless monocular vision loss. Retina cloudy with attenuated vessels and "cherry-red" spot at fovea (center of macula) A. Evaluate for embolic source (eg, carotid artery atherosclerosis, cardiac vegetations, patent foramen ovale).

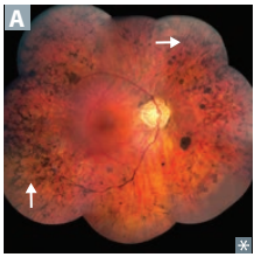
Retinal detachment



Separation of neurosensory layer of retina (photoreceptor layer with rods and cones) from outermost pigmented epithelium (normally shields excess light, supports retina) → degeneration of photoreceptors → vision loss. May be 2° to retinal breaks, diabetic traction, inflammatory effusions. Visualized on fundoscopy as crinkling of retinal tissue A and changes in vessel direction.

Breaks more common in patients with high myopia and/or history of head trauma. Often preceded by posterior vitreous detachment ("flashes" and "floaters") and eventual monocular loss of vision like a "curtain drawn down." Surgical emergency.

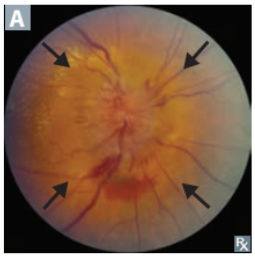
Retinitis pigmentosa



Inherited progressive retinal degeneration. Nyctalopia (night blindness) → peripheral vision loss. Bone spicule-shaped deposits A.

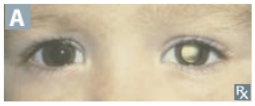
* It starts with **night blindness then, progressive vision loss** occur

Papilledema



Optic disc swelling (usually bilateral) due to ↑ ICP (eg, 2° to mass effect). Enlarged blind spot and elevated optic disc with blurred margins A.

Leukocoria



Loss (whitening) of the red reflex. Important causes in children include retinoblastoma A, congenital cataract, toxocariasis.

Pupil: Physiology and pathology

Pupillary control

Miosis

Constriction, parasympathetic:

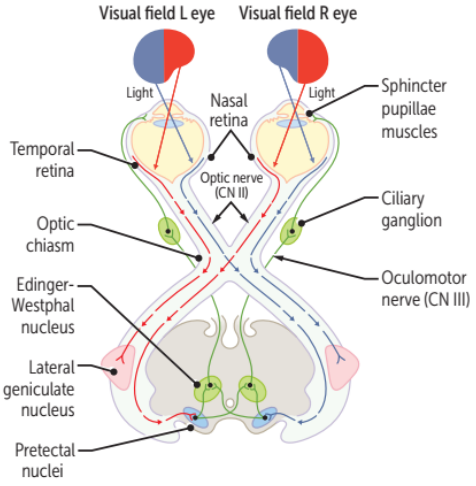
- 1st neuron: Edinger-Westphal nucleus to ciliary ganglion via CN III
- 2nd neuron: short ciliary nerves to sphincter pupillae muscles

Short ciliary nerves shorten the pupil diameter.

Pupillary light reflex

Light in either retina sends a signal via CN II to pretectal nuclei (dashed lines in image) in midbrain that activates bilateral Edinger-Westphal nuclei; pupils constrict bilaterally (direct and consensual reflex).

Result: illumination of 1 eye results in bilateral pupillary constriction.

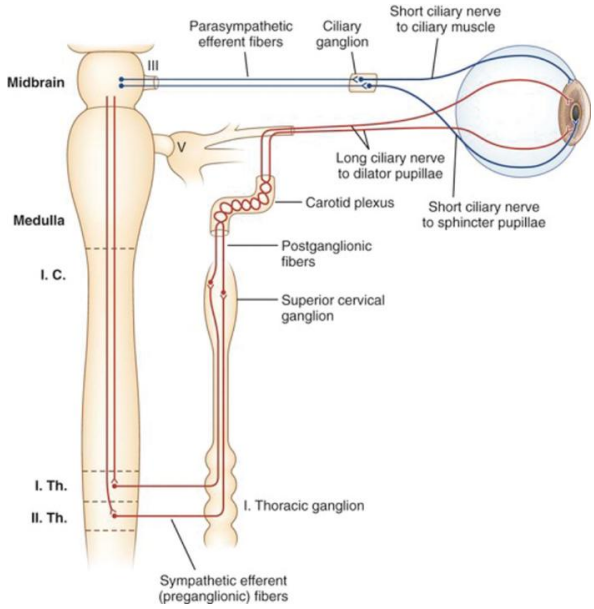


Mydriasis

Dilation, sympathetic:

- 1st neuron: hypothalamus to ciliospinal center of Budge (C8–T2)
- 2nd neuron: exit at T1 to superior cervical ganglion (travels along cervical sympathetic chain near lung apex, subclavian vessels)
- 3rd neuron: plexus along internal carotid, through cavernous sinus; enters orbit as long ciliary nerve to pupillary dilator muscles. Sympathetic fibers also innervate smooth muscle of eyelids (minor retractors) and sweat glands of forehead and face.

Long ciliary nerves make the pupil diameter longer.



Sympathetic supply of the pupil

Marcus Gunn pupil

*** Tested by swinging flash light test**

Also called relative afferent pupillary defect (RAPD). When the light shines into a normal eye, constriction of the ipsilateral (direct reflex) and contralateral eye (consensual reflex) is observed. When the light is then swung to the affected eye, both pupils dilate instead of constrict due to impaired conduction of light signal along the injured optic nerve. Associated with optic neuritis, early multiple sclerosis.

Horner syndrome

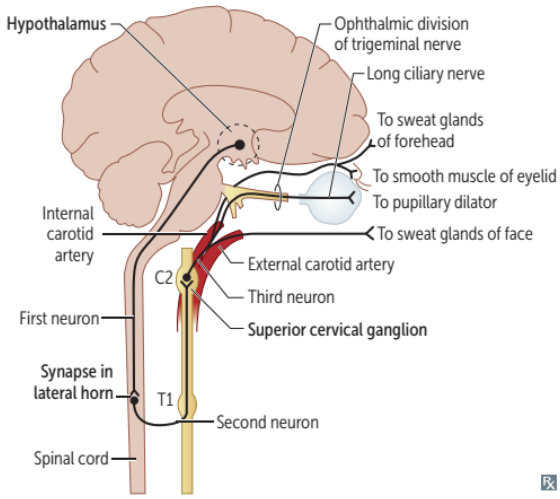
Sympathetic denervation of face →:

- Ptosis (slight drooping of eyelid: superior tarsal muscle)
- Anhidrosis (absence of sweating) and flushing of affected side of face
- Miosis (pupil constriction)

Associated with lesions along the sympathetic chain:

- 1st neuron: pontine hemorrhage, lateral medullary syndrome, spinal cord lesion above T1 (eg, Brown-Séquard syndrome, late-stage syringomyelia)
- 2nd neuron: stellate ganglion compression by Pancoast tumor
- 3rd neuron: carotid dissection (painful)

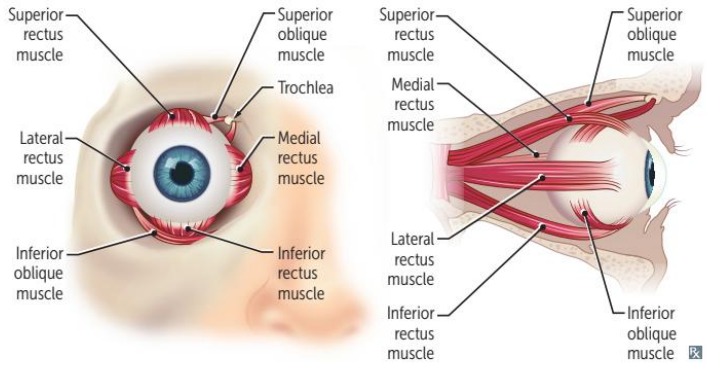
PAM is horny (Horner).



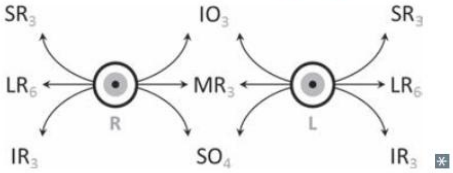
- **Argyl Robertson pupil**
  - Cause: Tertiary neuro-syphilis
  - Pathology: Destruction of pretectal nucleus of the midbrain
  - Result: Pupil constrict in response to accommodation but not to light
  - Accommodate but not react 😊
- **Addie pupil** is the same as Argyl Robertson but, unilateral due to lesion in the ciliary ganglion

Movement of the eye

Ocular motility



CN VI innervates the Lateral Rectus.
CN IV innervates the Superior Oblique.
CN III innervates the Rest.
The "chemical formula" $LR_6SO_4R_3$.



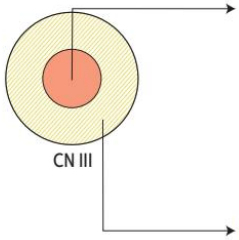
Obliques go Opposite (left SO and IO tested with patient looking right).
IOU: IO tested looking Up.

CN III, IV, VI palsies

CN III damage

CN III has both motor (central) and parasympathetic (peripheral) components.
Common causes include:

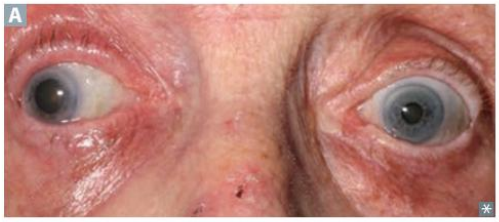
- Ischemia → pupil sparing (motor fibers affected more than parasympathetic fibers)
- Uncal herniation → coma
- PCom aneurysm → sudden-onset headache
- Cavernous sinus thrombosis → proptosis, involvement of CNs IV, V_1/V_2, VI
- Midbrain stroke → contralateral hemiplegia



Motor output to extraocular muscles—affected primarily by vascular disease (eg, diabetes mellitus: glucose → sorbitol) due to ↓ diffusion of oxygen and nutrients to the interior fibers from compromised vasculature that resides on outside of nerve. Signs: ptosis, "down-and-out" gaze.

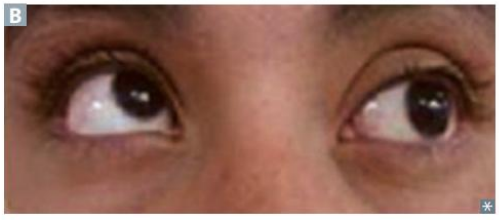
Parasympathetic output—fibers on the periphery are first affected by compression (eg, PCom aneurysm, uncal herniation). Signs: diminished or absent pupillary light reflex, "blown pupil" often with "down-and-out" gaze A.

Motor = Middle (central)
Parasympathetic = Peripheral



CN IV damage

Pupil is higher in the affected eye B.
Characteristic head tilt to contralateral/unaffected side to compensate for lack of intorsion in affected eye.
Can't see the floor with CN IV damage (eg, difficulty going down stairs, reading).



CN VI damage

Affected eye unable to abduct and is displaced medially in primary position of gaze C.

Horizontal Conjugate gaze

- **Def.:** The coordinate horizontal movement of both eyes so that the image falls on two corresponding spots on the retinas of both eyes for proper vision
- **Control: Gaze centers**
  1- Frontal eye field area = contralateral conjugate gaze
  2- Paramedian pontine reticular formation in the pons = ipsilateral conjugate gaze
- **Pathway: Let's say you want to look to the right**
  1- Left frontal eye field send an impulse to the right PPRF
  2- Right PPRF send an impulse to the right abducent nerve nucleus which stimulate the left oculomotor nerve nucleus through the MLF
  3- Right eye abducts and left eye adducts
- **MLF:**
  # Highly myelinated bundle connecting abducent nerve nucleus to oculomotor nerve nucleus
  # Affected by MS causing nystagmus on horizontal gaze in the abducting eye (internuclear ophthalmoplegia)
  # Although the adducting eye fail to move with conjugate gaze, there is intact convergence with accommodation due to intact oculomotor nerve
  # Since oculomotor nerve is intact no ptosis or miosis occur
- **Lesions:**
  1- Frontal eye field ➔ see before (cerebral cortex section) but please note that corticobulbar and corticospinal tract fibers lie in close proximity to the frontal eye field area so, usually they are severed with the frontal eye field lesion
  2- Abducent nerve nucleus ➔ ipsilateral complete horizontal conjugate gaze loss (both eyes can not look towards the affected side) + ipsilateral facial paralysis (Do you remember why ? 😊)
  3- MLF ➔ internuclear ophthalmoplegia (see below)

Vertical Conjugate gaze

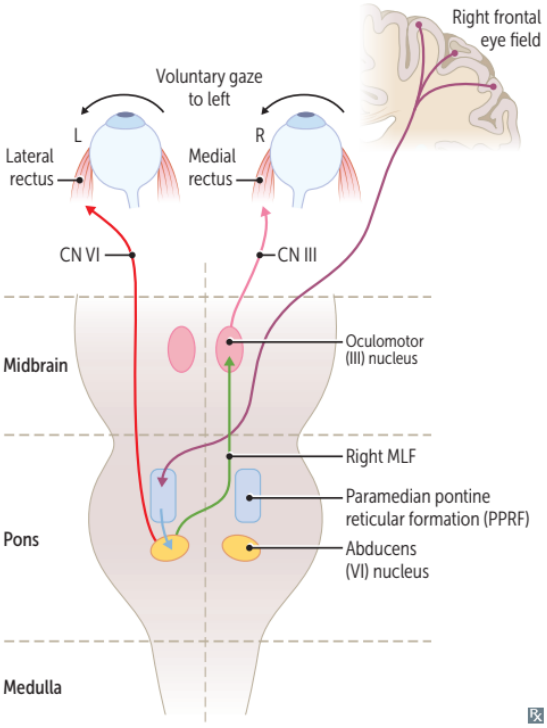
- **Def.:** The coordinate vertical movement of both eyes so that the image falls on two corresponding spots on the retinas of both eyes for proper vision
- **Control:** Frontal eye field, Superior colliculi and midbrain vertical gaze centers
- **Pathway:**
  # Frontal eye field ➔ Superior colliculi ➔ vertical gaze centers ➔ vertical gaze
  # Right eye is controlled by the right frontal eye field and left colliculus
- **Lesion:** The affected eye will not move vertically, and the unaffected eye will have a vertical nystagmus

- **PLEASE NOTE THAT VESTIBULAR TYPES OF NYSTAGMUS E.G., CENTRAL CEREBELLAR NYSTAGMUS ARE DISCUSSED LATER IN THE VESTIBULAR SYSTEM**

Internuclear ophthalmoplegia

Medial longitudinal fasciculus (MLF): pair of tracts that allows for crosstalk between CN VI and CN III nuclei. Coordinates both eyes to move in same horizontal direction. Highly myelinated (must communicate quickly so eyes move at same time). Lesions may be unilateral or bilateral (latter classically seen in multiple sclerosis, stroke).

Lesion in MLF = internuclear ophthalmoplegia (INO), a conjugate horizontal gaze palsy. Lack of communication such that when CN VI nucleus activates ipsilateral lateral rectus, contralateral CN III nucleus does not stimulate medial rectus to contract. Abducting eye displays nystagmus (CN VI overfires to stimulate CN III). Convergence normal.

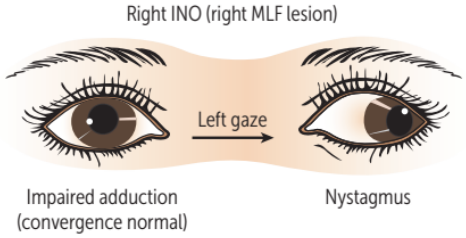


MLF in MS.

When looking left, the left nucleus of CN VI fires, which contracts the left lateral rectus and stimulates the contralateral (right) nucleus of CN III via the right MLF to contract the right medial rectus.

Directional term (eg, right INO, left INO) refers to the eye that is unable to adduct.

INO = Ipsilateral adduction failure, Nystagmus Opposite.



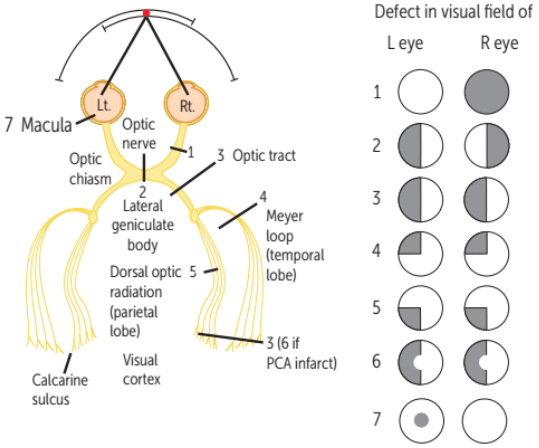
Visual pathway and field defects

Visual field defects

1. Right anopia (monocular vision loss)
2. Bitemporal hemianopia (pituitary lesion, chiasm)
3. Left homonymous hemianopia
4. Left upper quadrantanopia (right temporal lesion, MCA)
5. Left lower quadrantanopia (right parietal lesion, MCA)
6. Left hemianopia with macular sparing (right occipital lesion, PCA)
7. Central scotoma (eg, macular degeneration)

Meyer **L**oop—**L**ower retina; **L**oops around inferior horn of **L**ateral ventricle.

Dorsal optic radiation—superior retina; takes shortest path via internal capsule.



Note: When an image hits 1° visual cortex, it is upside down and left-right reversed.