



Neuroophthalmology

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Content

- **Visual pathway affection**
 - Diseases and affections of optic nerve
 - Optic chiasm pathology
 - Pathology of retrochiasmic part
- **Eye movement disorders**
 - Binocular diplopia
- **Pupillary reaction abnormalities**
 - Anisocoria
- **Combined disorders**

Examination - part I

- Medical history
 - subjective (visual loss, diplopia)
 - When it started/ how long lasts it?
 - Does it change in time/ during the day?
 - Any progression?
 - What about the fellow eye?
 - Other signs?
 - Personal medical history?
 - Pharmacological history?
 - objective (pupillary dysfunction, eye movement disorders, ptosis of upper eyelid, red eye)

Examination - part II

- **Visual acuity**
 - Without and with correction
 - Monocular vision / binocular vision
- **Basic ophthalmological examination**
 - Anterior segment (by slit lamp)
 - Posterior segment - arteficial mydriasis is essential (indirect ophthalmoscopy)
- **Visual field examination (static / kinetic perimetry)**

Examination - part III

- **Basic examination (GP)**
- **Neurological examination**
 - Intracranial conditions (including MRI)
 - neurological signs
- **Endocrinology**
 - Thyroid associated orbitopathy / ophthalmopathy
 - Pituitary dysfunction

Examination - part IV

- **Imaging techniques**

- Ultrasonography (eye bulb, orbit)
- X-ray of skull (orbit, paranasal cavities)
- Computerised Tomography of head (brain, skull bones, orbital bones)
- MRI of head (brain, orbital structures)

Optic nerve disorders

Clinical signs

- Visual acuity decrease
- Visual field defect

Division:

- **Congenital**
 - developmental anomalies
- **Acquired**
 - inflammation – optic neuritis
 - Non-inflammation – ischemic neuropathy (anterior, posterior)
- **Genetic relatives**
 - LHON (Leber hereditary optic atrophy)

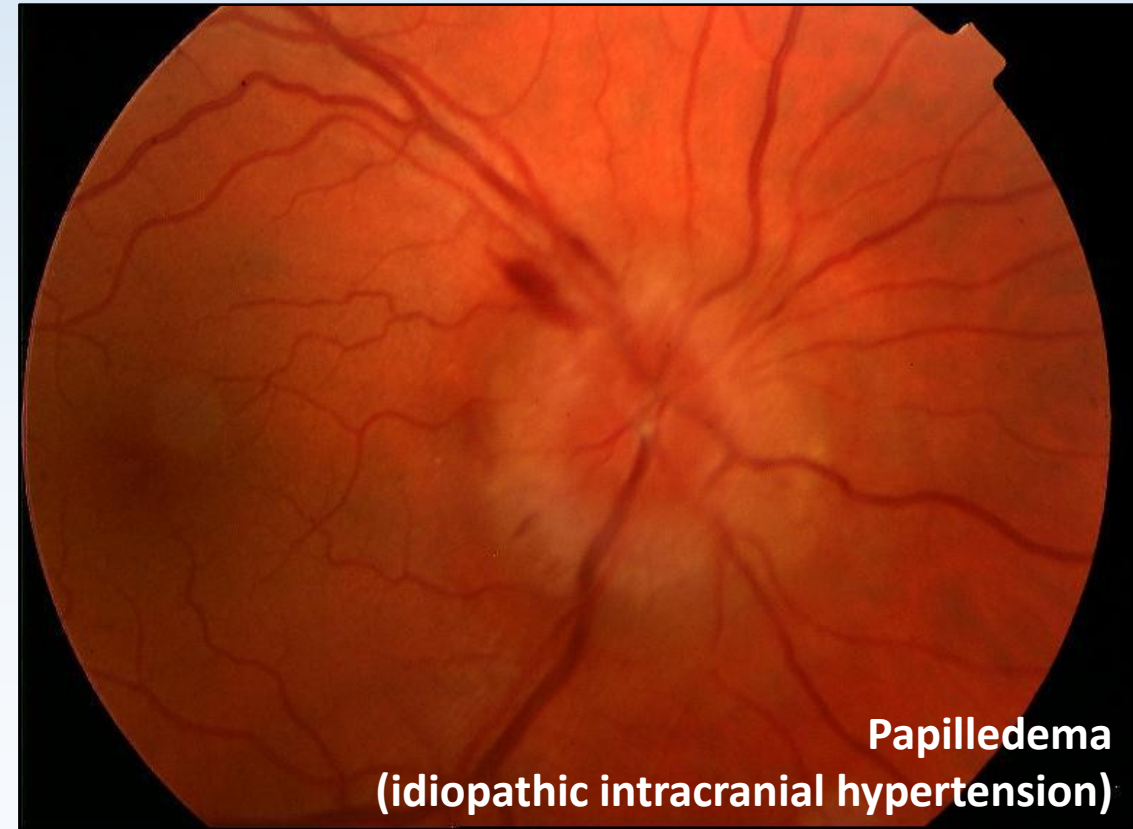
Optic disc oedema = papilledema in general

- **Causes**

- *Elevated intracranial pressure* – tumor, idiopathic intracranial pressure, hydrocephalus
- *Optic nerve affection* – optic neuritis, neuropathy

- **Clinical picture**

- elevation of optic disc, diminished margins
- loss of physiological excavation
- swelling of peripapillary retinal nerve fiber layer
- optic disc hyperemia
- dilatation and tortuosity of vessels
- Haemorrhages, cotton wool spots



Papilledema

- cause – elevated intracranial pressure
- 75% of cases – intracranial tumor!!!
- bilateral condition, often asymmetric
- faster onset in young people
- mostly without subjective signs
- sometimes blurred vision
- enlargement of blind spot



Optic neuritis

Clinical picture

- unilateral condition
- fast onset (hours)
- loss of visual acuity
- *retrobulbar pain* – pathognomical sign
- color vision defects
- visual field defects

Causes

- demyelination – most common (multiple sclerosis)
- infection / parainfection
- paraneoplastic

Types

- intraocular
- retrobulbar – most common

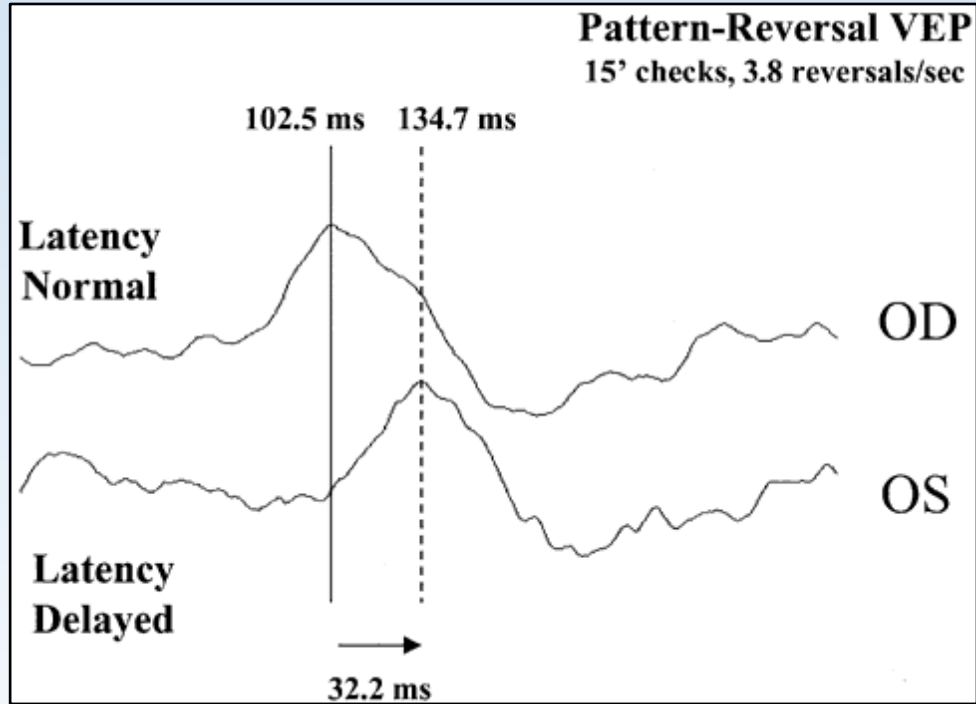
Prognosis

- usually good – regression after intravenous corticoids

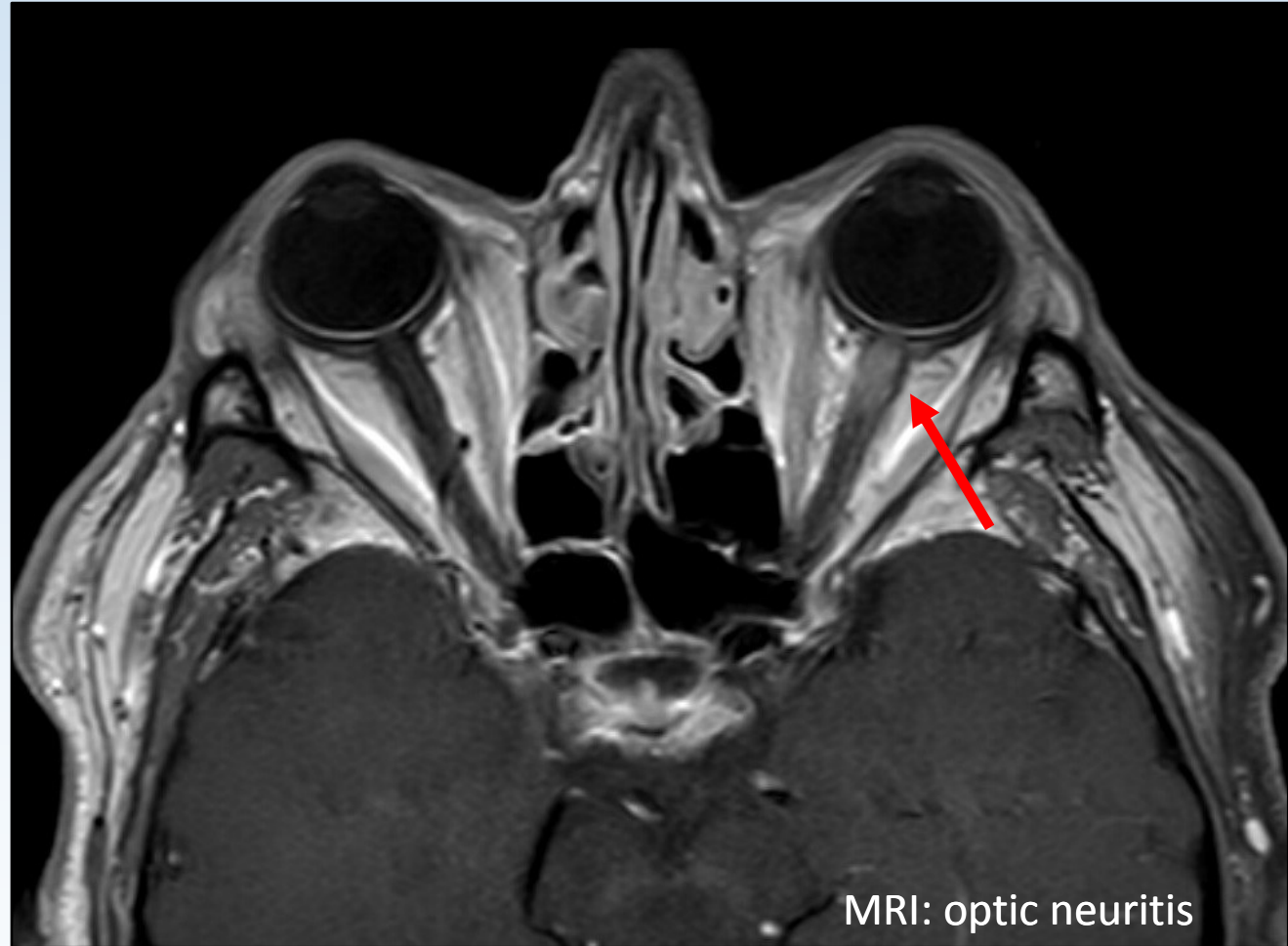
Epidemiology

- 20-40 years of age
- female/male: 2-3/1
- Strong association with MS
 - 20% of cases – first sign of MS
 - 50% patients with MS – manifestation of ON during the disease

Optic neuritis diagnosis



VEP: delayed latency P100



Anterior ischemic optic neuropathy

- most common optic nerve affection in advanced age
- unilateral condition

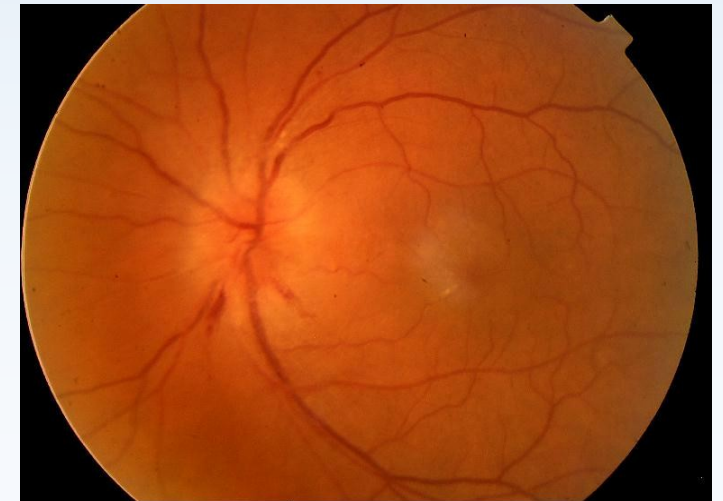
Cause – affection of short ciliar arteries

Epidemiology

- 50 years of age and more

Clinical picture

- loss of visual acuity - fast onset, painless (light perception to almost normal values)
- monocular visual field defect – altitudinal scotoma
- unilateral ischemic optic disc oedema



Anterior ischemic optic neuropathy

Arteritic form (10 – 15 % of all cases) – less common, more serious

- Risk factors: association with systemic vasculitis (giant-cell arteritis = Horton disease)
- Clinical picture: loss on weight, headache, jaw claudication, tenderness and sensitivity on the scalp)
- very high sedimentation rate - over 100 per hour, temporal artery biopsy
- **High risk of affection of fellow eye** (days, weeks) – **immediate therapy!!!**
- *Therapy*: high dosage of intravenous corticoids

Nonarteritic form (85 – 90 % of all cases)

- Risk factors: hypertension, diabetes, dyslipidemia, smoking, obesity
- *Therapy*: N/A, compensation of all systemic diseases



Optic nerve atrophy

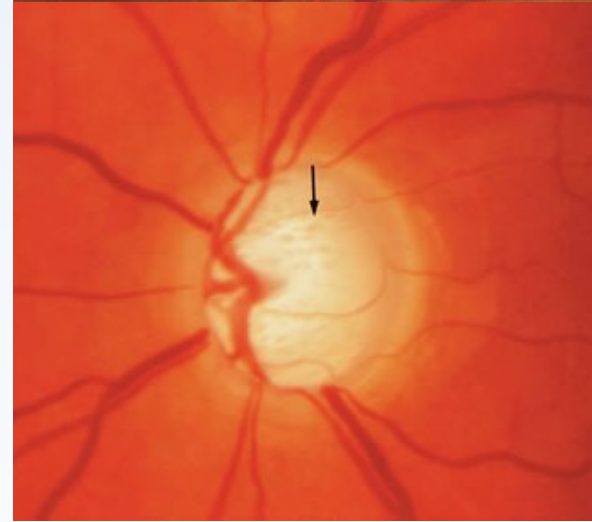
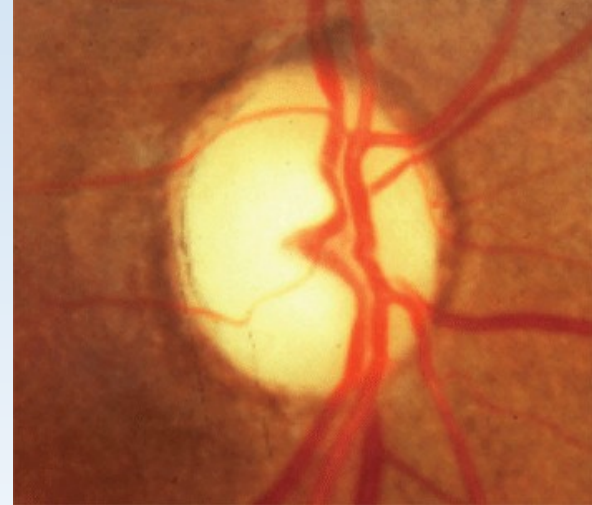
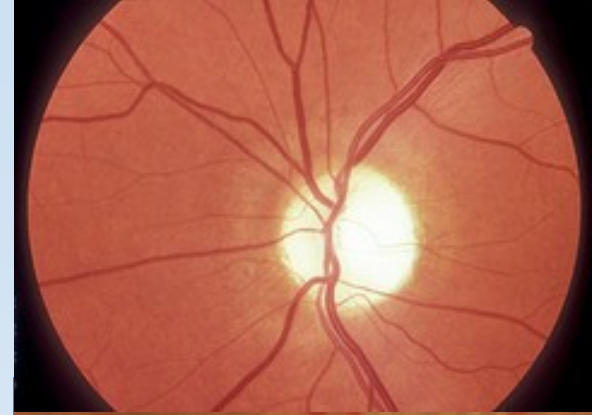
- Irreversible loss of axons
- After various optic nerve affections

Etiology

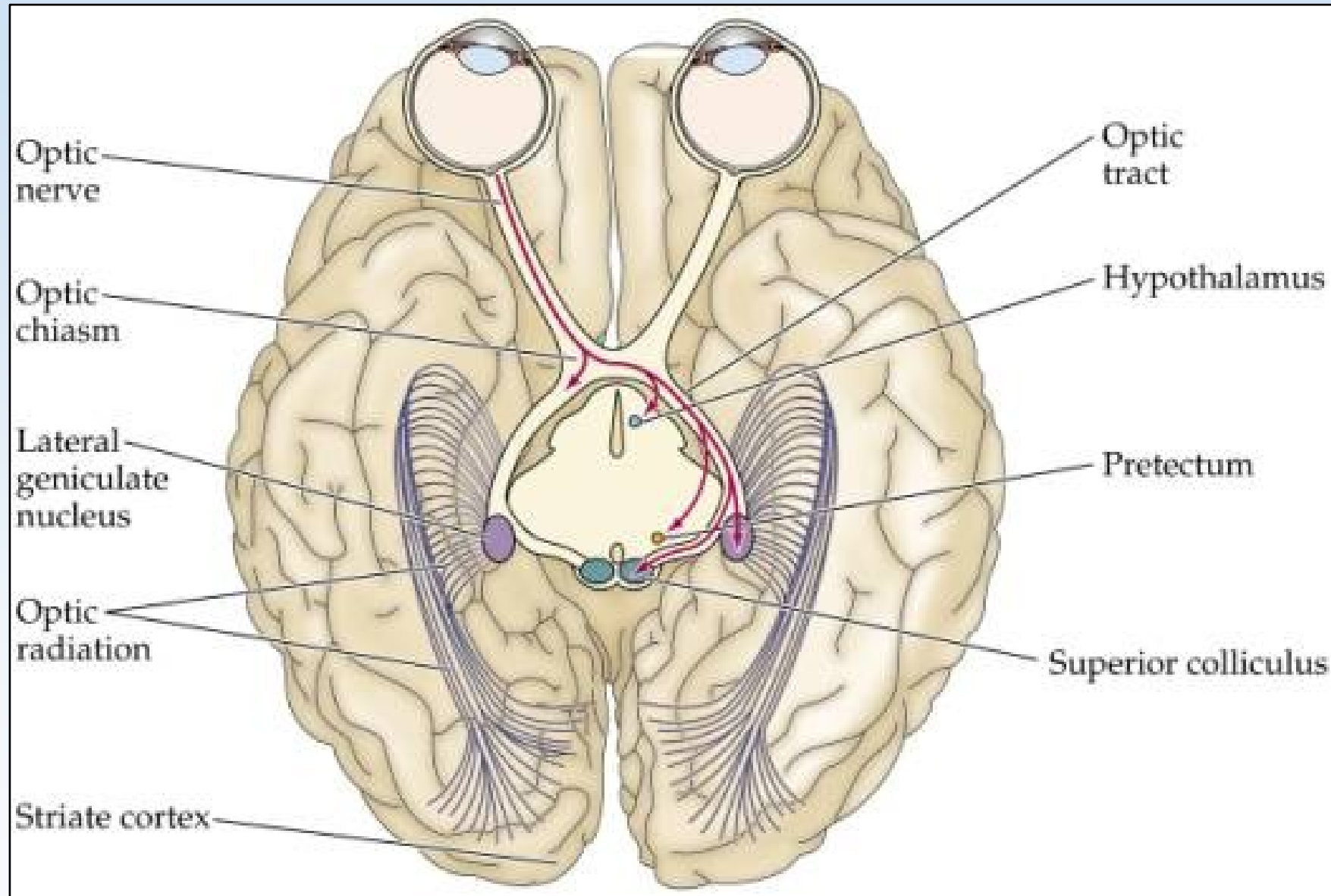
- *Primary* – posttraumatic, by direct pressure of tumor
- *Secondary* – affection of optic nerve (ischemia, inflammation)
- *Glaucomatous* – elevates intraocular pressure

Clinical picture

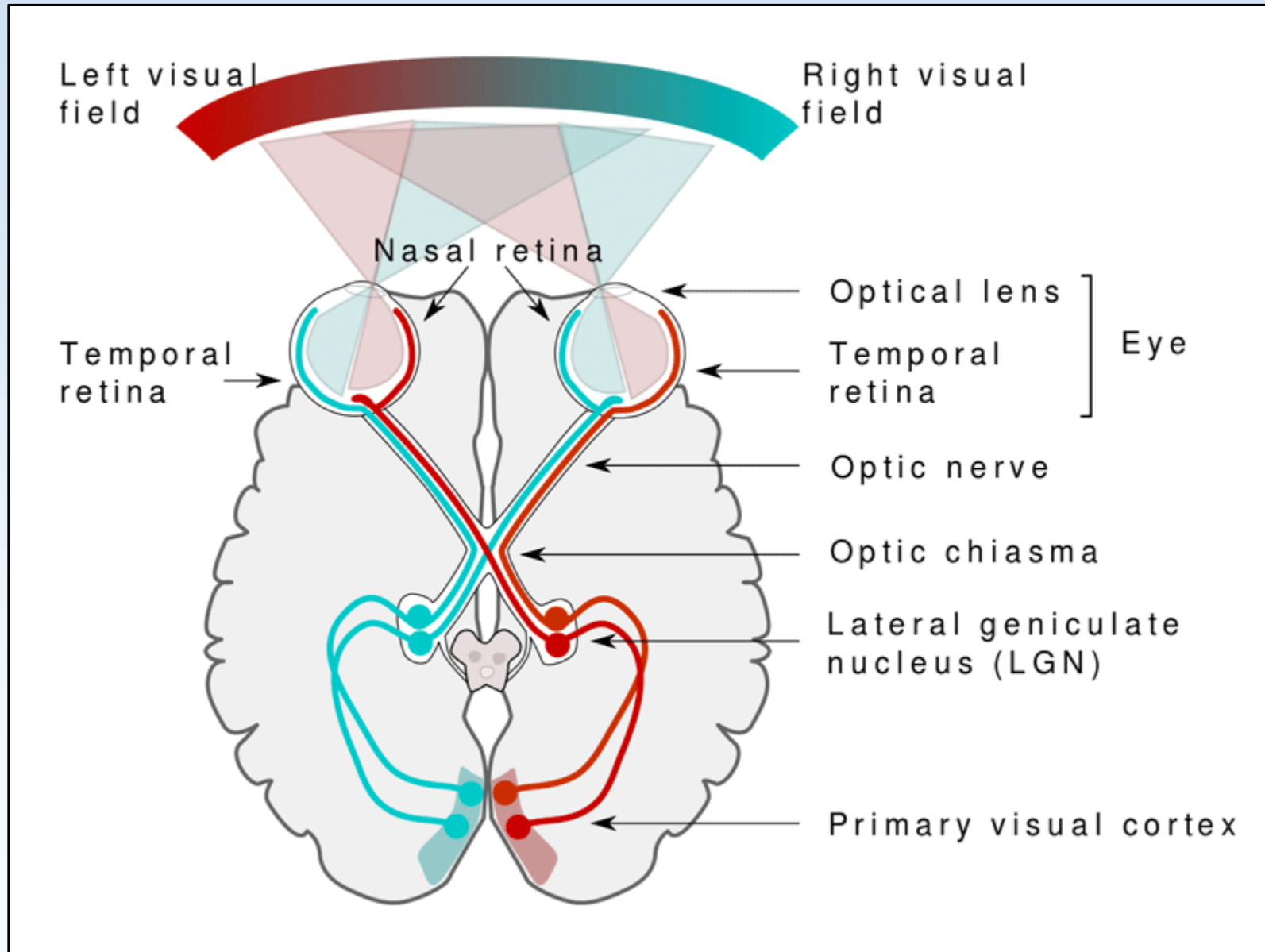
- Pale optic disc
- Reduction of smaller vessels



Anatomy of visual pathway



Anatomy of visual pathway



Visual field defects

Optic nerve

- monocular visual field defects

Optic chiasm

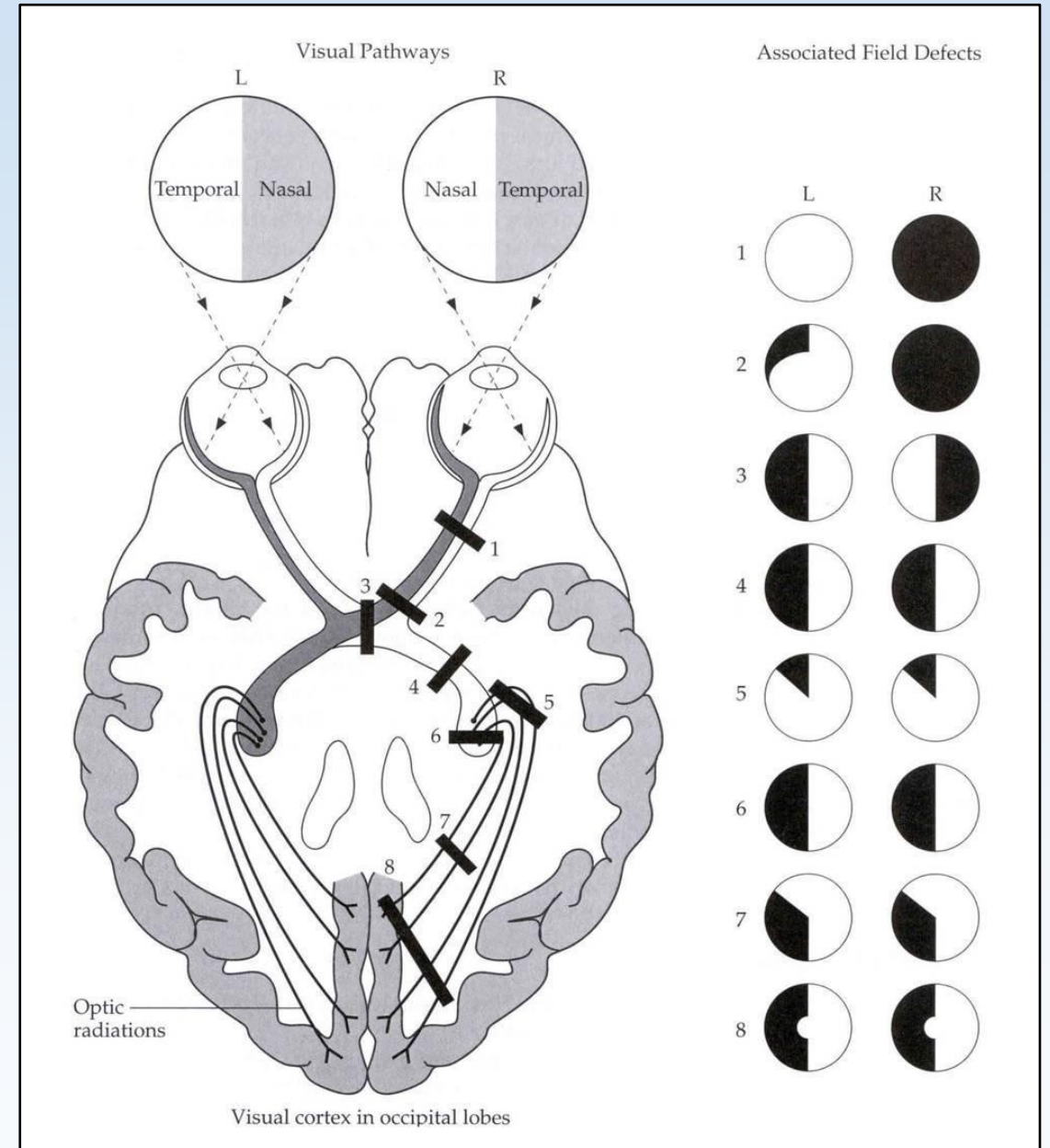
- bilateral heteronym visual field defects

Optic tract

- bilateral homonym visual field defects

Geniculocalcarine tract

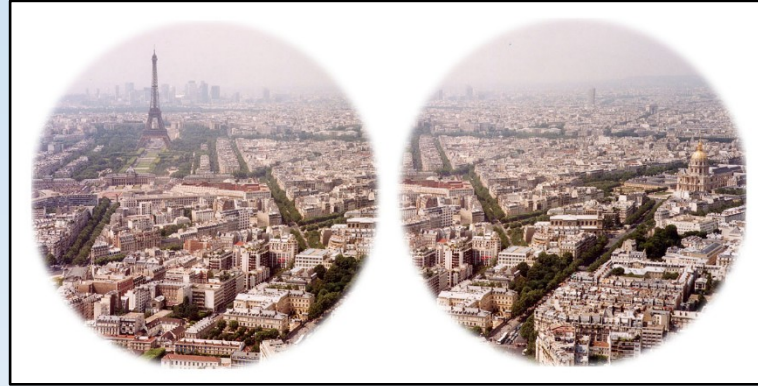
- bilateral visual field defects often with central sparing



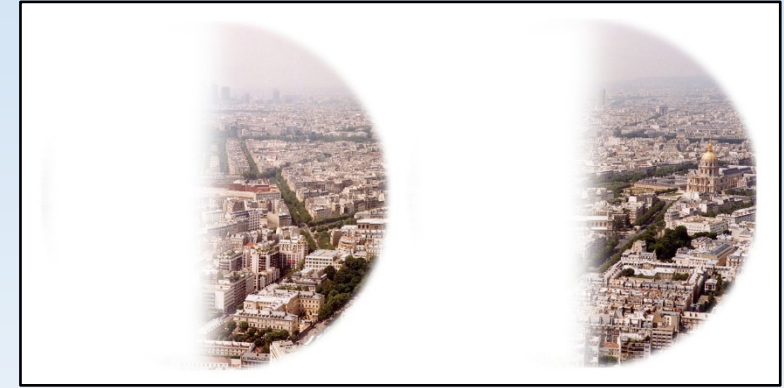
Specific visual field defects with correlation to visual pathways



Bitemporal hemianopsia



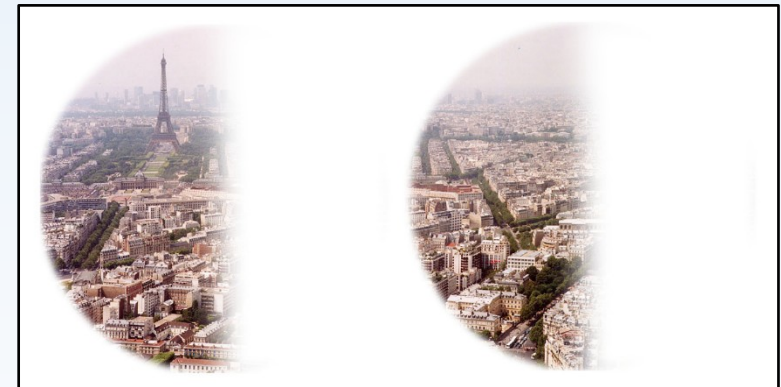
Physiological visual field



Left sided homonymous hemianopsia



Binasal hemianopsia



Right sided homonymous hemianopsia

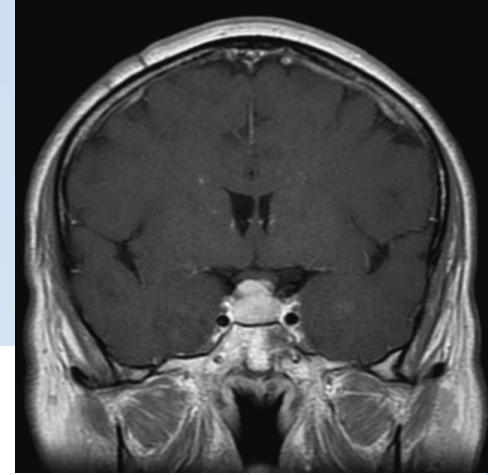
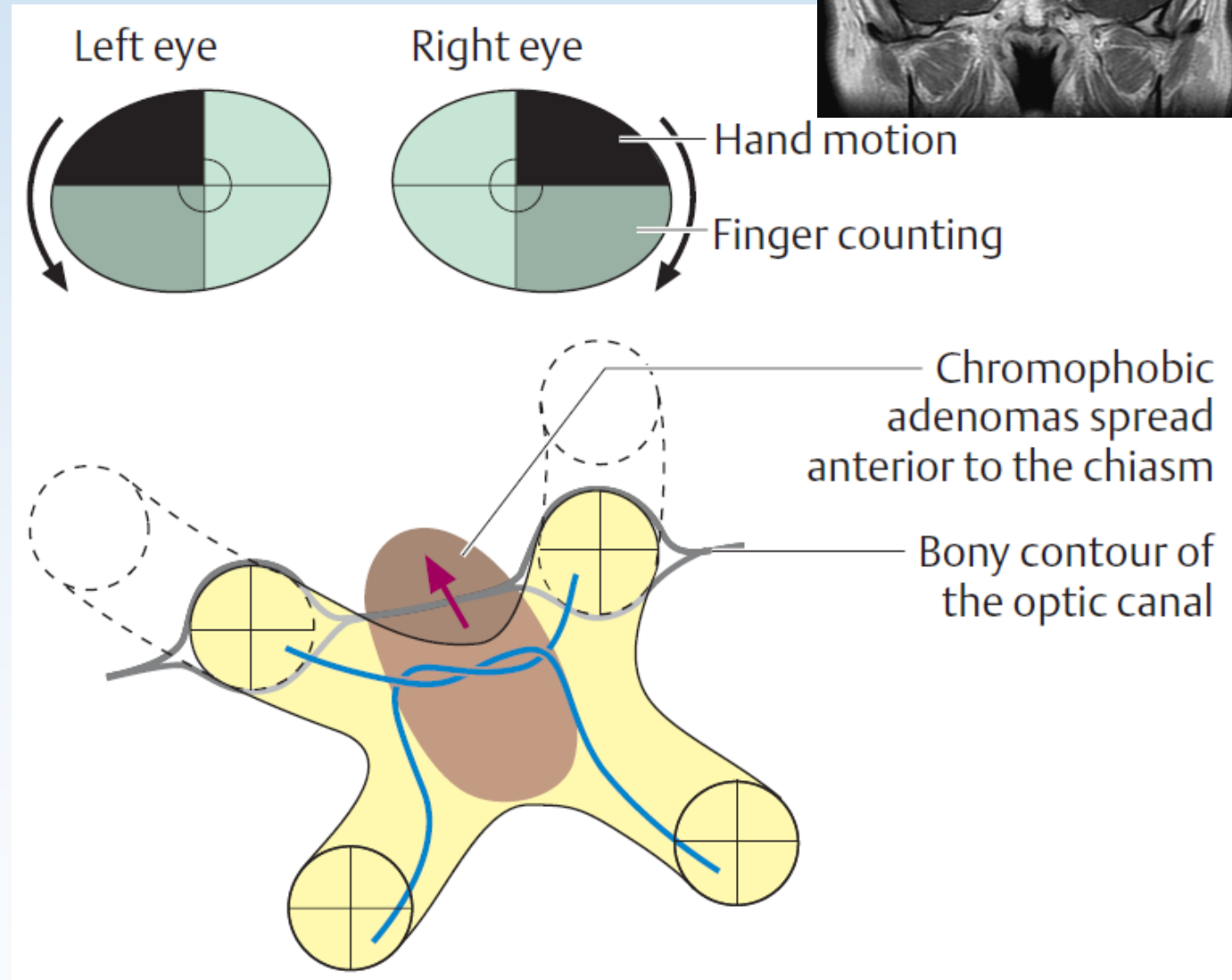
Chiasmal syndrome

- lesions in chiasmal area
- typically compressive, expansive condition
- typical visual field defects – use in diagnosis

- causes:
 - Pituitary adenomas
 - Craniopharyngioma
 - Meningioma
 - Aneurysm

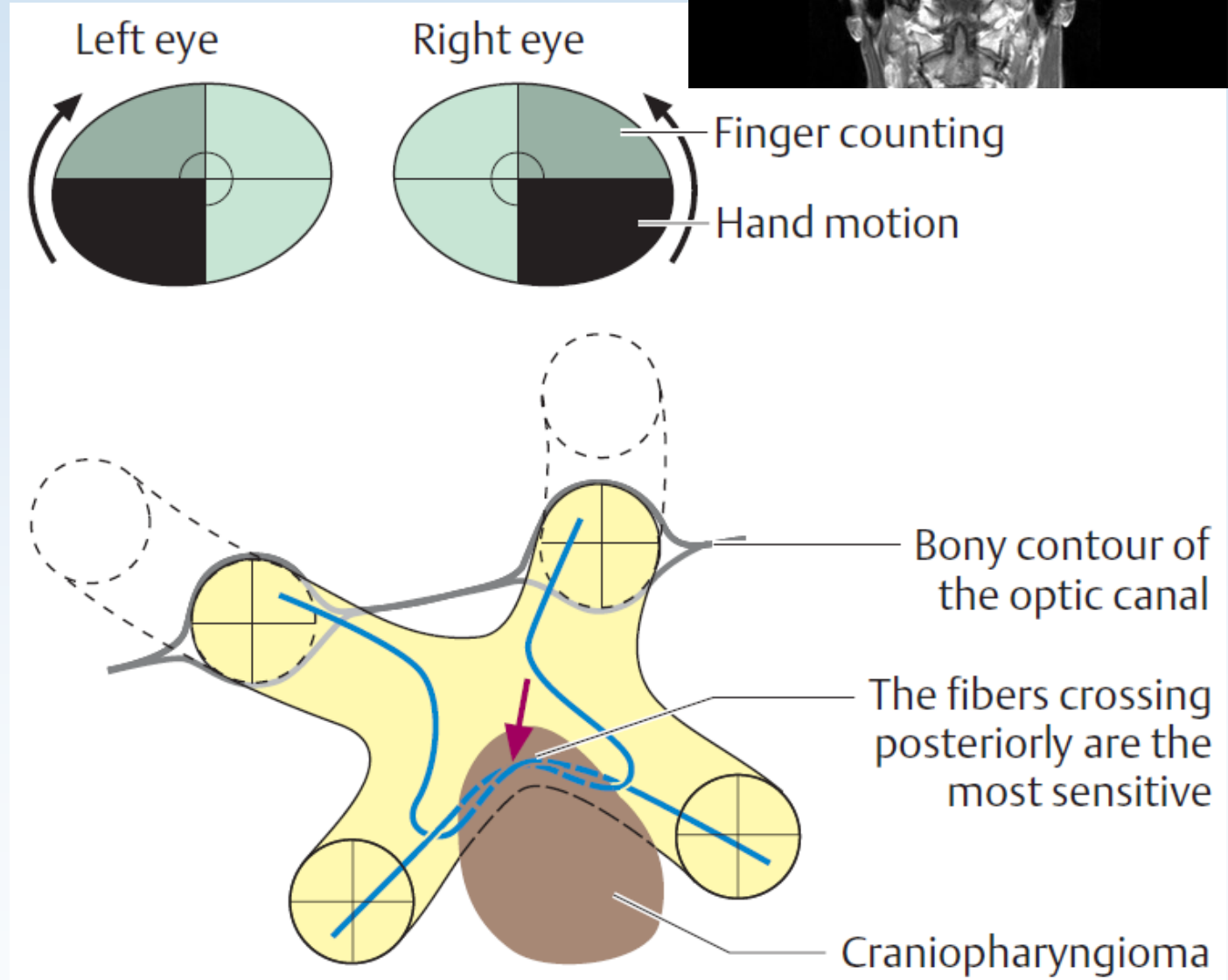
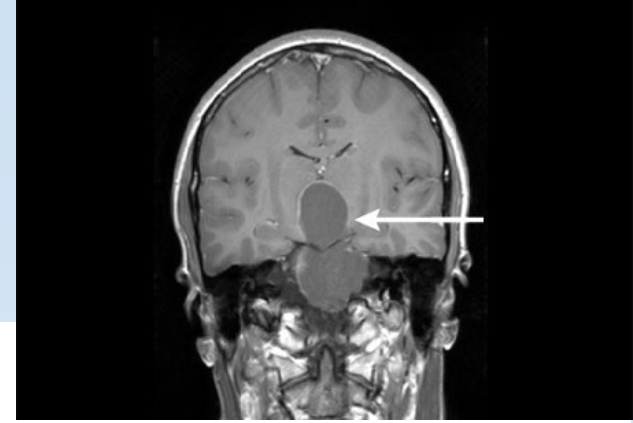
Pituitary adenoma

- benign tumor of pituitary gland
- classification
 - by size – microadenoma (up to 10mm), macroadenoma (more than 10mm)
 - biological activity – benign adenoma, invasive adenoma, adenocarcinoma
- possibility of metabolic activity (e.g. prolactinoma)
- compression and lesion of optic chiasm by tumor growth – **bitemporal hemianopsia** – starting as upper kvadrantanopsia
- therapy
 - *conservative* – hormone inhibition (Cabergolin, Octreotid)
 - *surgical* - resection (endonasal, transsphenoidal adenectomy)



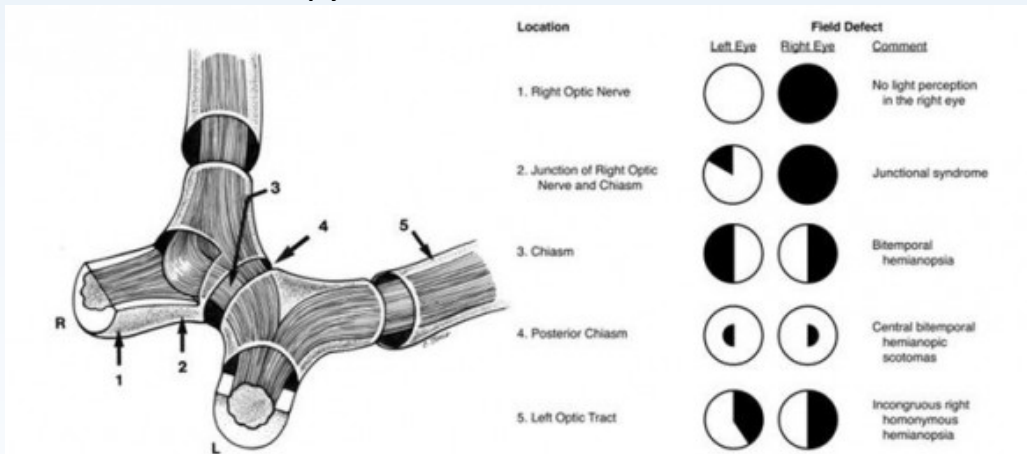
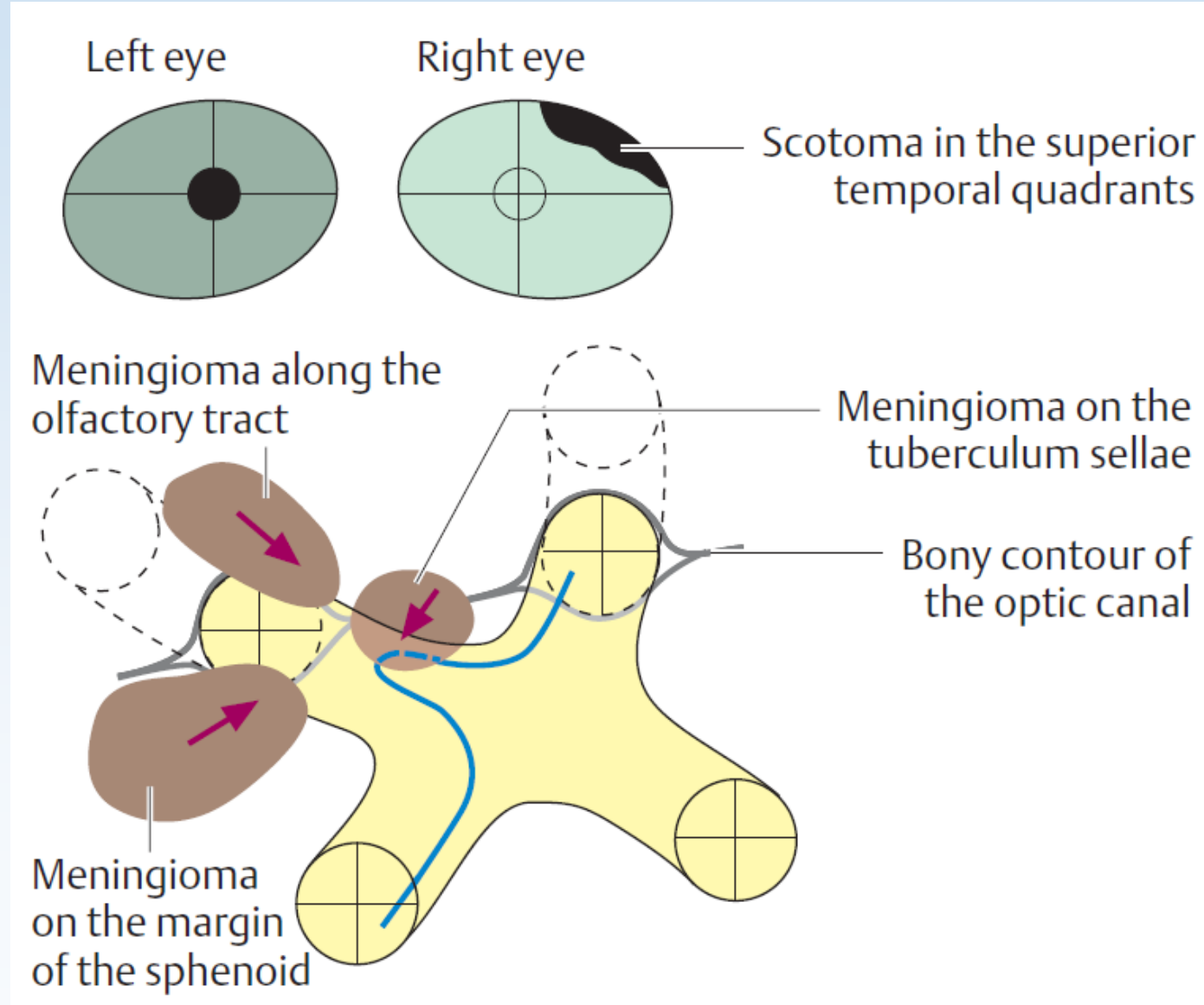
Craniopharyngioma

- benign rare type of tumor from pituitary gland embryonal tissue
- pressure on nearby tissue, typical visual field defects – **bitemporal hemianopsia** – first starting as lower quadrantanopsia
- therapy
 - *surgical* - transsphenoidal adenectomy)
 - *radiotherapy*



Meningioma

- slow growing tumor from meninges
- tumor growth, pressure on nearby tissue, typical visual field defects depending on location
- therapy
 - *surgical*
 - *radiotherapy*



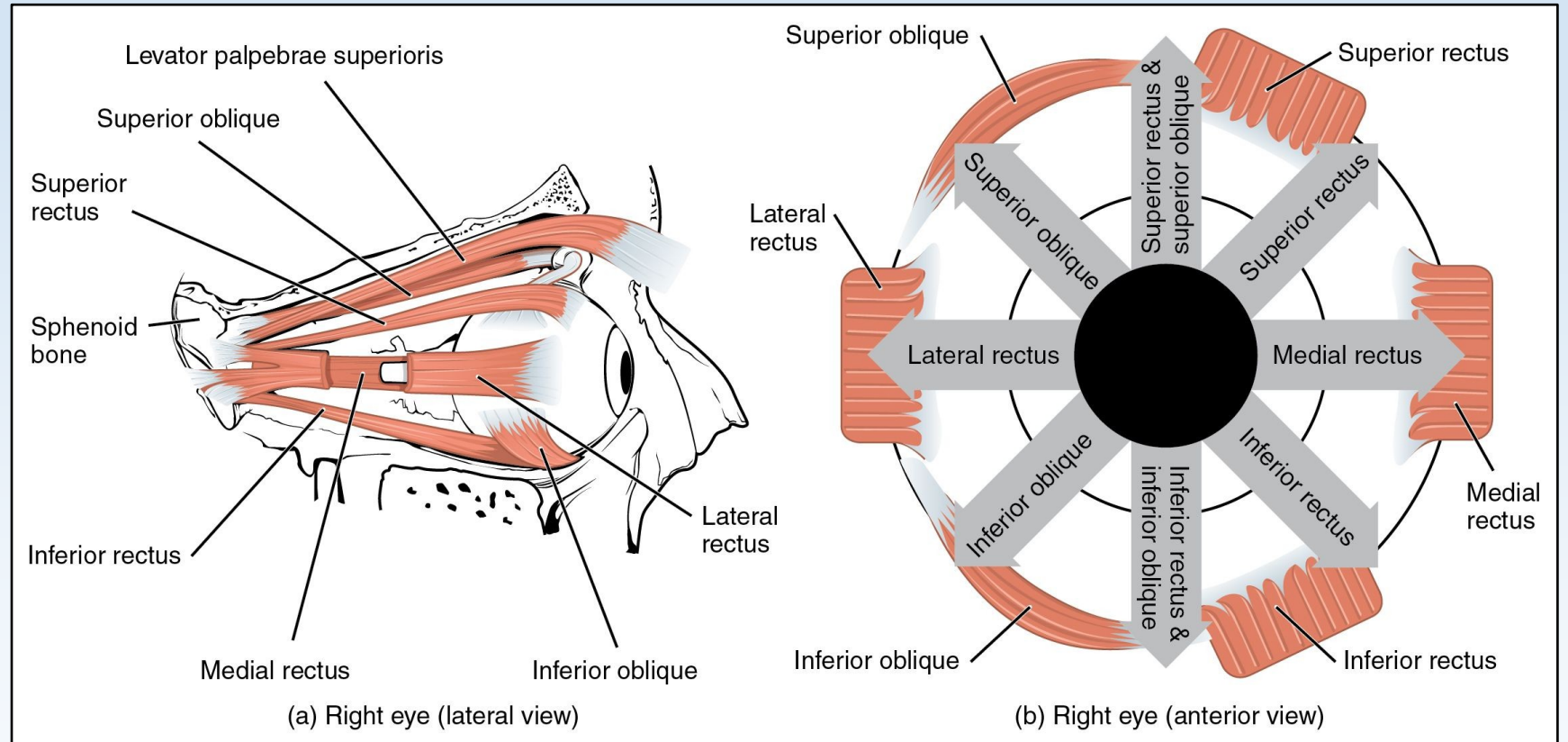
Anatomy of eye movement system

- **4 recti muscles:**

- medial rectus m.
- lateral rectus m.
- inferior rectus m.
- superior rectus m.

- **2 oblique muscles:**

- superior oblique m.
- inferior oblique m.



Eye movement disorders

- **Isolated palsies**

- oculomotor nerve palsy
- trochlear nerve palsy
- abducent nerve palsy

- **Ophthalmoplegia**

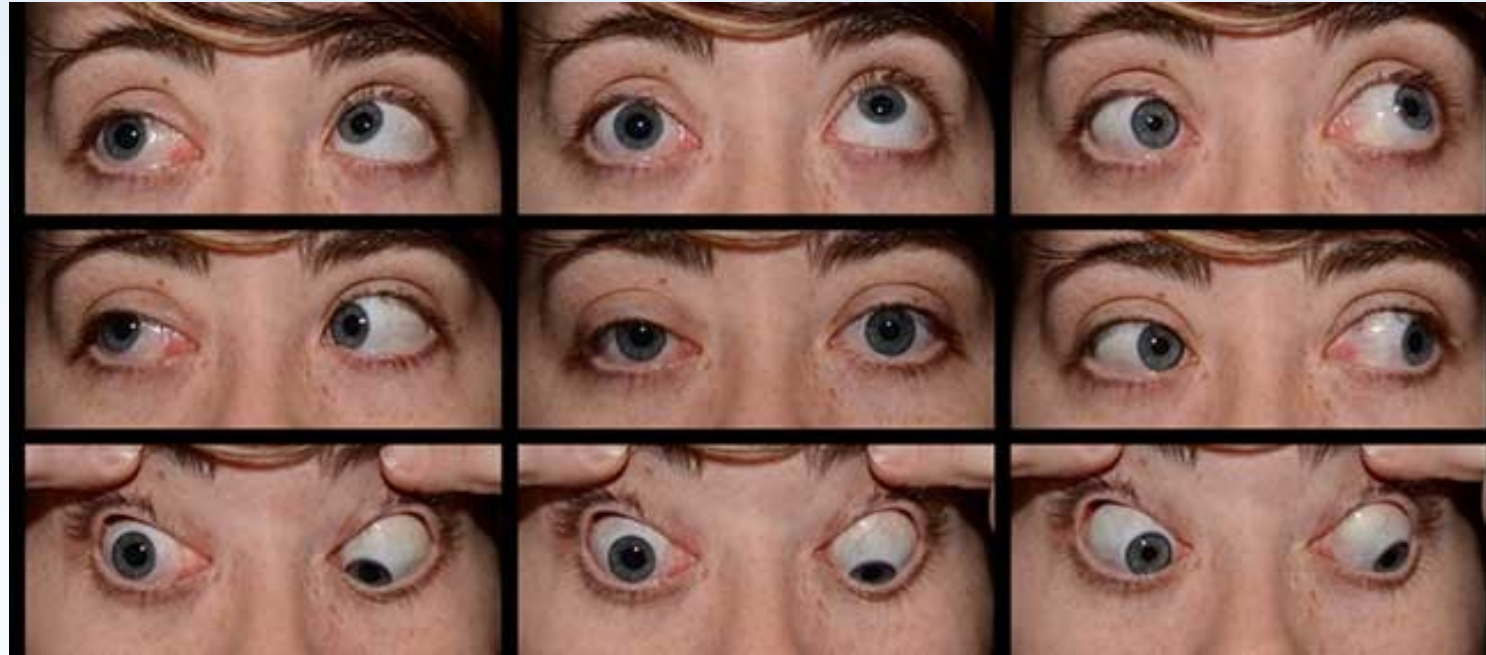
- combined disorders (affection of 2 or 3 nerves)
 - cavernous sinus syndrome
 - orbital apex syndrome
 - carotido-cavernous fistula

Isolated palsies

- **Oculomotor nerve palsy**– *aneurysm* (most common), less common tumor, trauma, ischemia
- **Trochlear nerve palsy** – most common *trauma* (fall on head), less common ischemia of brainstem, tumor, half of cases idiopathic
- **Abducent nerve palsy** – mostly trauma, ischemia (diabetes), intracranial hypertension (sometimes first manifestation), less common tumor or idiopathic

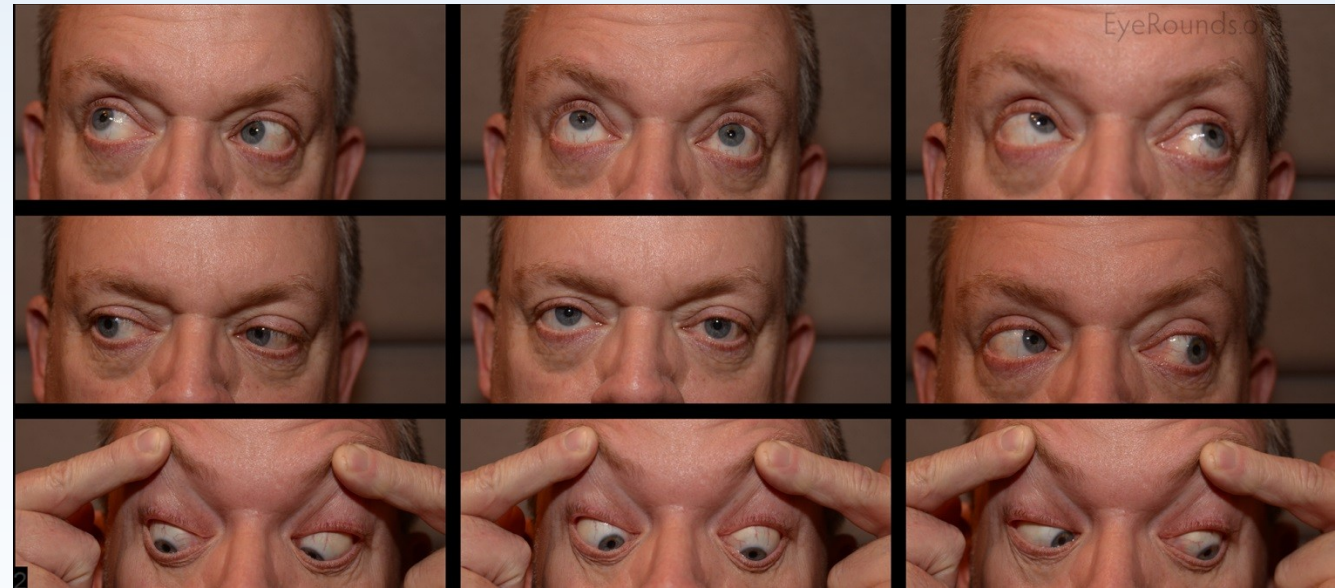
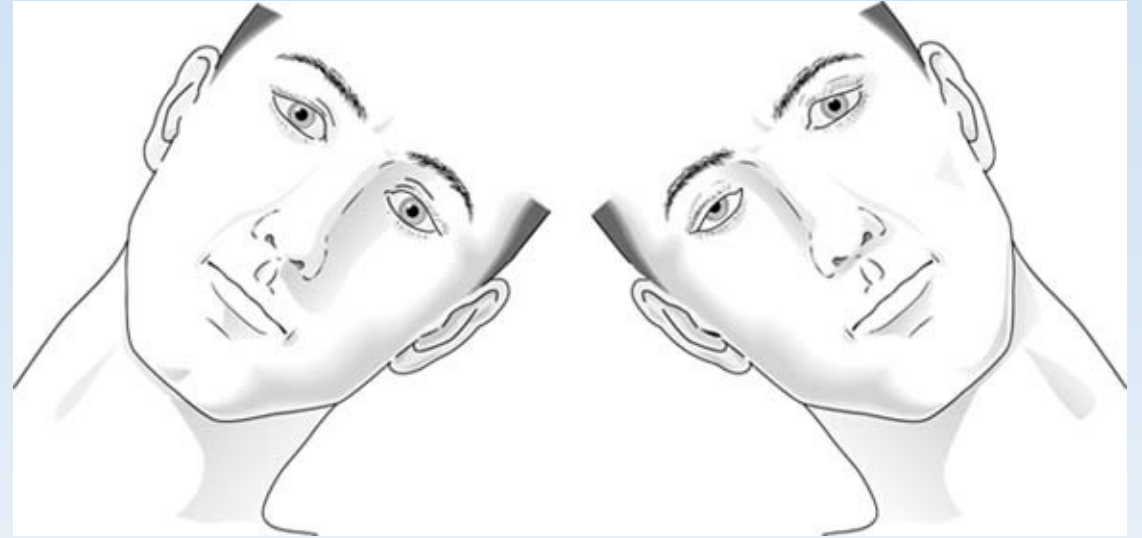
Oculomotor nerve palsy

- **Upper eyelid ptosis**
- **Abduction** of affected eye
- **Eye movement disorder** – multiple sizes of gaze (nasal, up, down)
- **Diplopia** (mixed – horizontal and vertical)
- **Anisocoria** (mydriasis on affected size)



Trochlear nerve palsy

- **Diplopia** – vertical, major manifestation in downgaze, bigger turning back, walking downstairs)
- **Compensation head posture** (Torticollis) – chin turning down, head posture at non affected size
- **Eye movement disorder** – affected eye upgaze, not necessary visible!



Abducent nerve palsy

- **Diplopia** – typically horizontal, major manifestation in gaze to affected side
- **Compensation head posture** – head turned on affected side
- **Eye movement disorder** – adduction of affected eye o straight position, abduction insufficiency on affected side



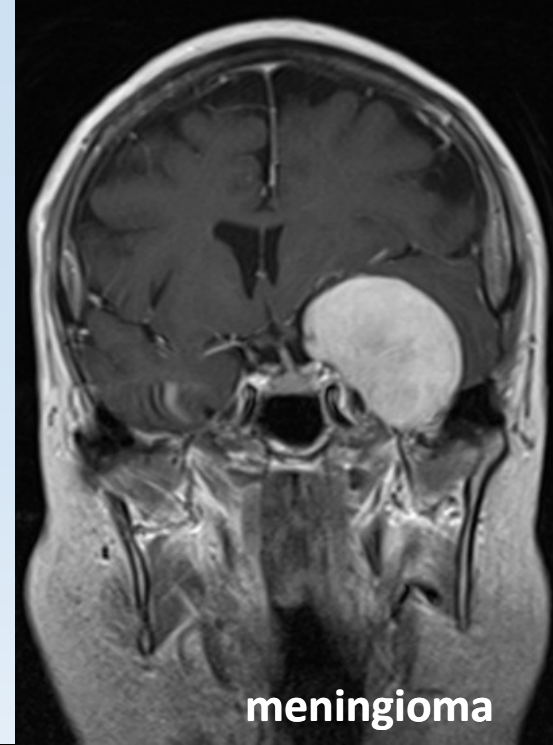
Cavernous sinus syndrome

- **Etiology**

- Expansive / infiltrative condition in cavernous sinus (thrombosis, tumor, metastasis, aneurysm)
- Affection of oculomotor, trochlear, abducent, trigeminal nerve (1. or 2. branch)

- **Clinical picture**

- Upper eyelid ptosis
- Ophthalmoplegia (incomplete / complete)
- Diplopia
- Plegic pupil / mydriasis
- Exophthalmus
- pain / hypesthesia



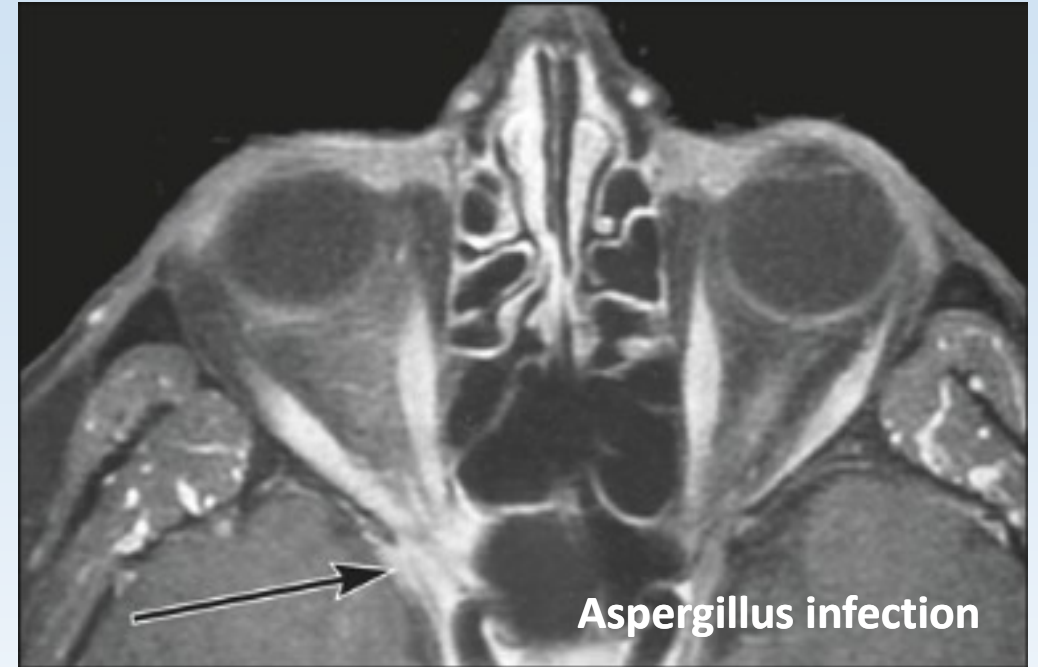
Orbital apex syndrome

- **Etiology**

- expansive / infiltrative condition in orbital apex (tumor, metastasis, orbitocellulitis, infection)
- Affect oculomotor, trochlear, abducent, trigeminal nerve and also optic nerve

- **Clinical picture**

- Upper eyelid ptosis
- Ophthalmoplegia (incomplete / complete)
- Visual acuity decrease – diplopia (indirectly to level of visual acuity)
- Exophthalmus
- Hypoesthesia (inervation area of 1. and 2. branch of trigeminal nerve)



Carotid-cavernous fistula

• Etiology

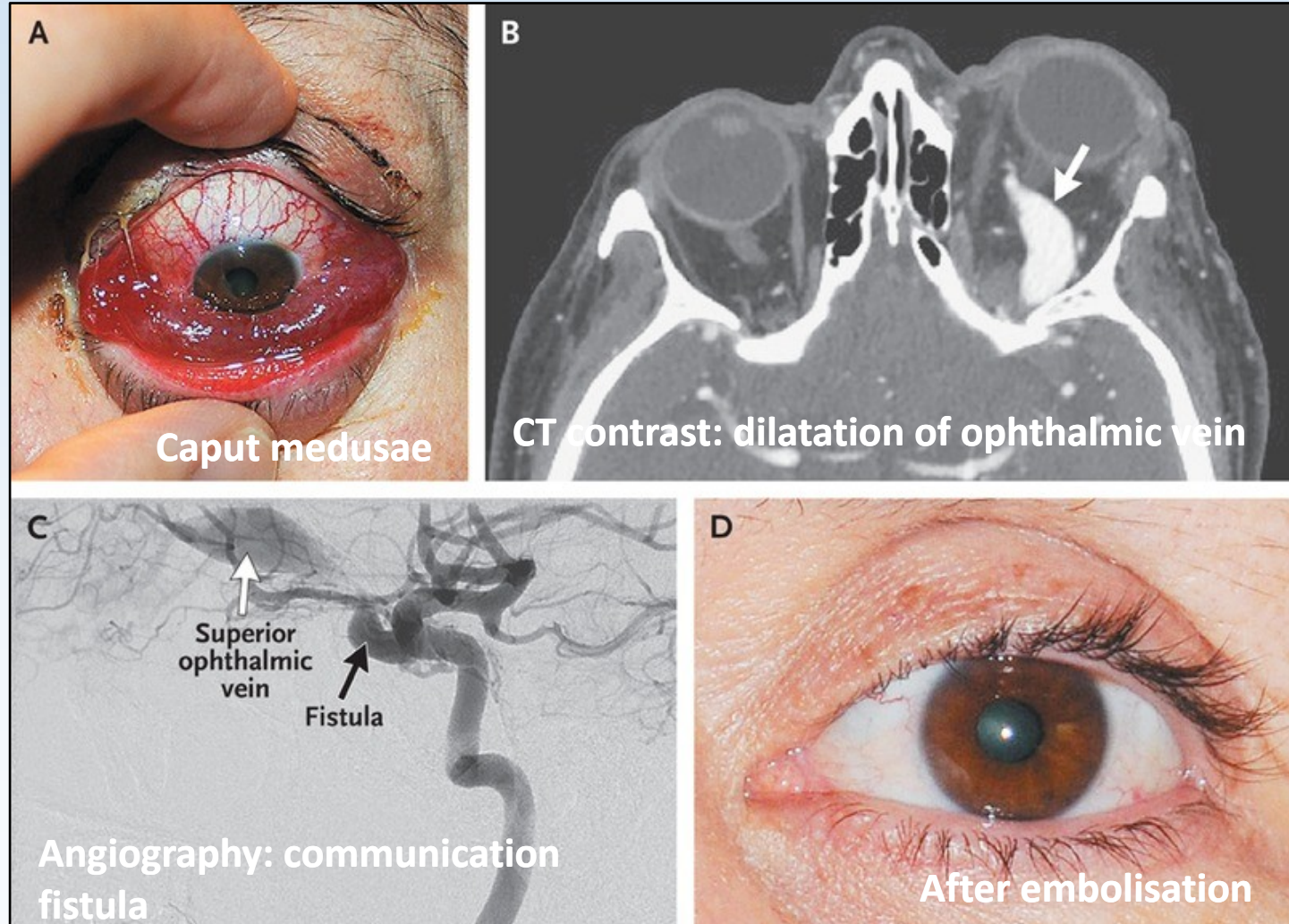
- abnormal communication between arterial and venous system within the cavernous sinus
- *direct* – post traumatic; rupture of ICA wall
- *indirect* – rupture of smaller vessels

• Clinical picture

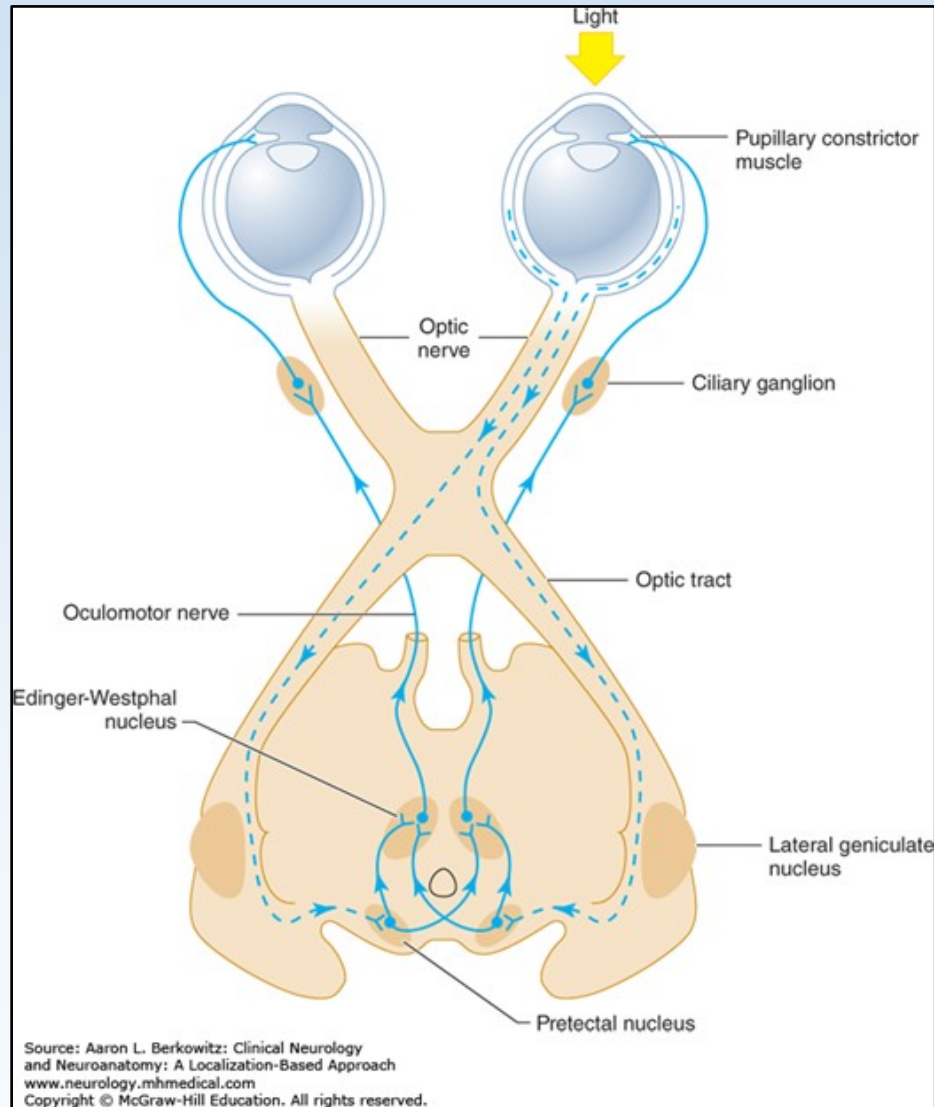
- **Caput medusae**, bruit, feeling of pulsation, eye movement disorder, diplopia, exophthalmus, elevated intraocular pressure, decreased visual acuity

• Therapy

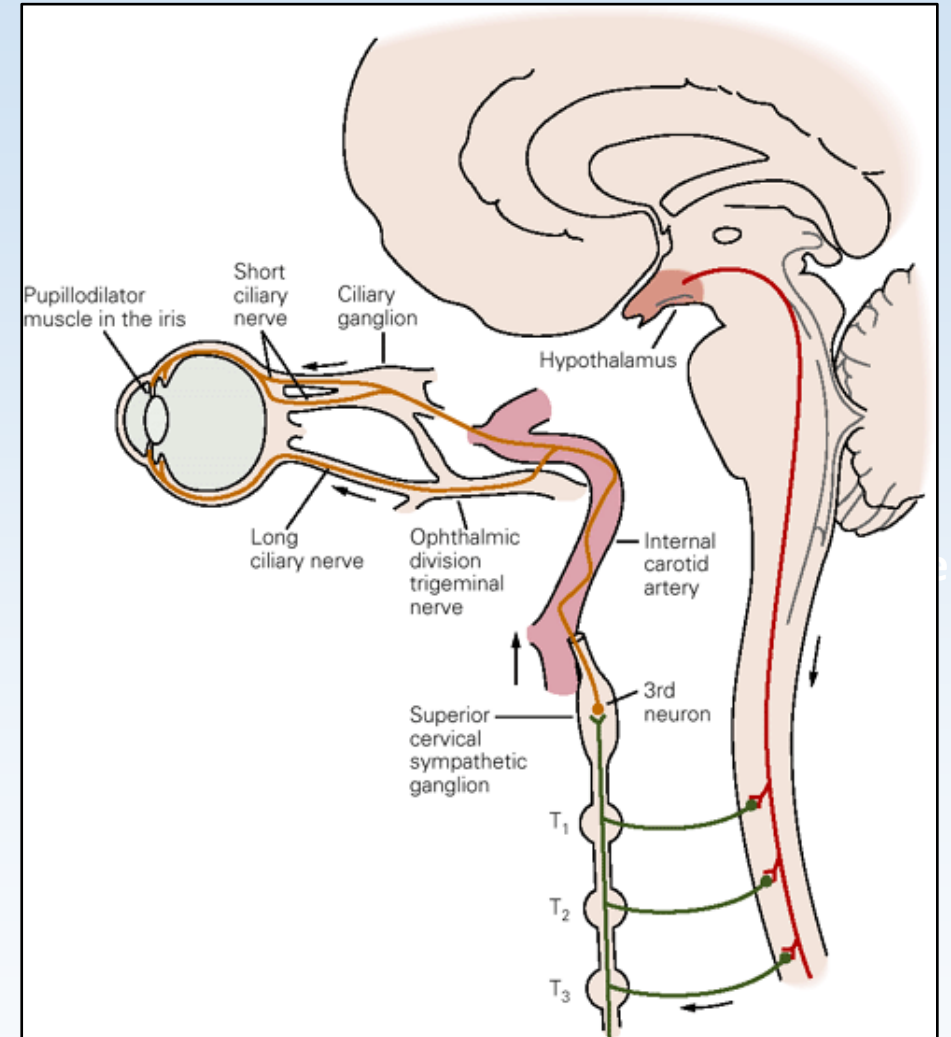
- endovascular – transarterial or transvenous (stent, coil, balloons)



Pupilmotoric pathway



Parasympathetic pathway



Sympathetic pathway

Pupillary reactions

- **Basic reactions**

- **Mydriasis** – sympathetic innervation (m. dilatator pupillae)
- **Miosis** – parasympathetic innervation (m. sphincter pupillae)

- **Diagnostic tests**

- **Direct and undirect shine** (action-reaction) – physiological miosis of both pupils
- **Near response test** – physiological miosis associated with accommodation
- **Pharmacological testing**

Pupillary reactions

- **Typical pupil appearance**

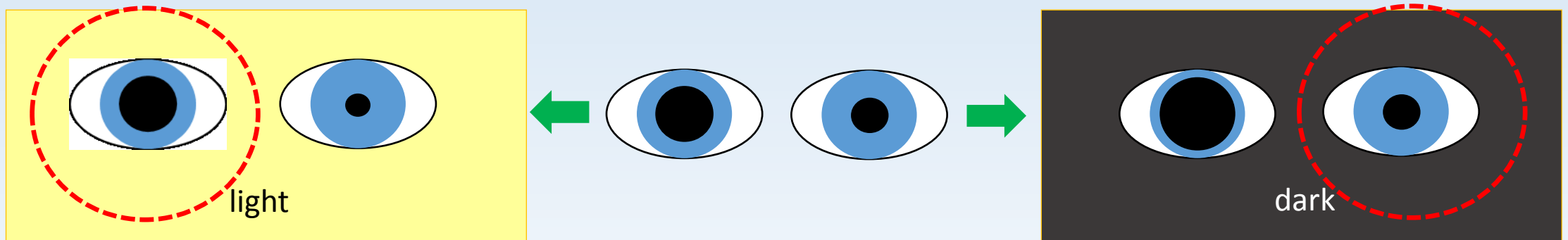
- **isocoria** (physiological anisocoria up to 1mm – same size in various illuminance)
- **consensual reaction** of both pupils
- **size** – diameter 3mm, dependence on autonomic nervous system

- **Atypical appearance or pupilar reaction**

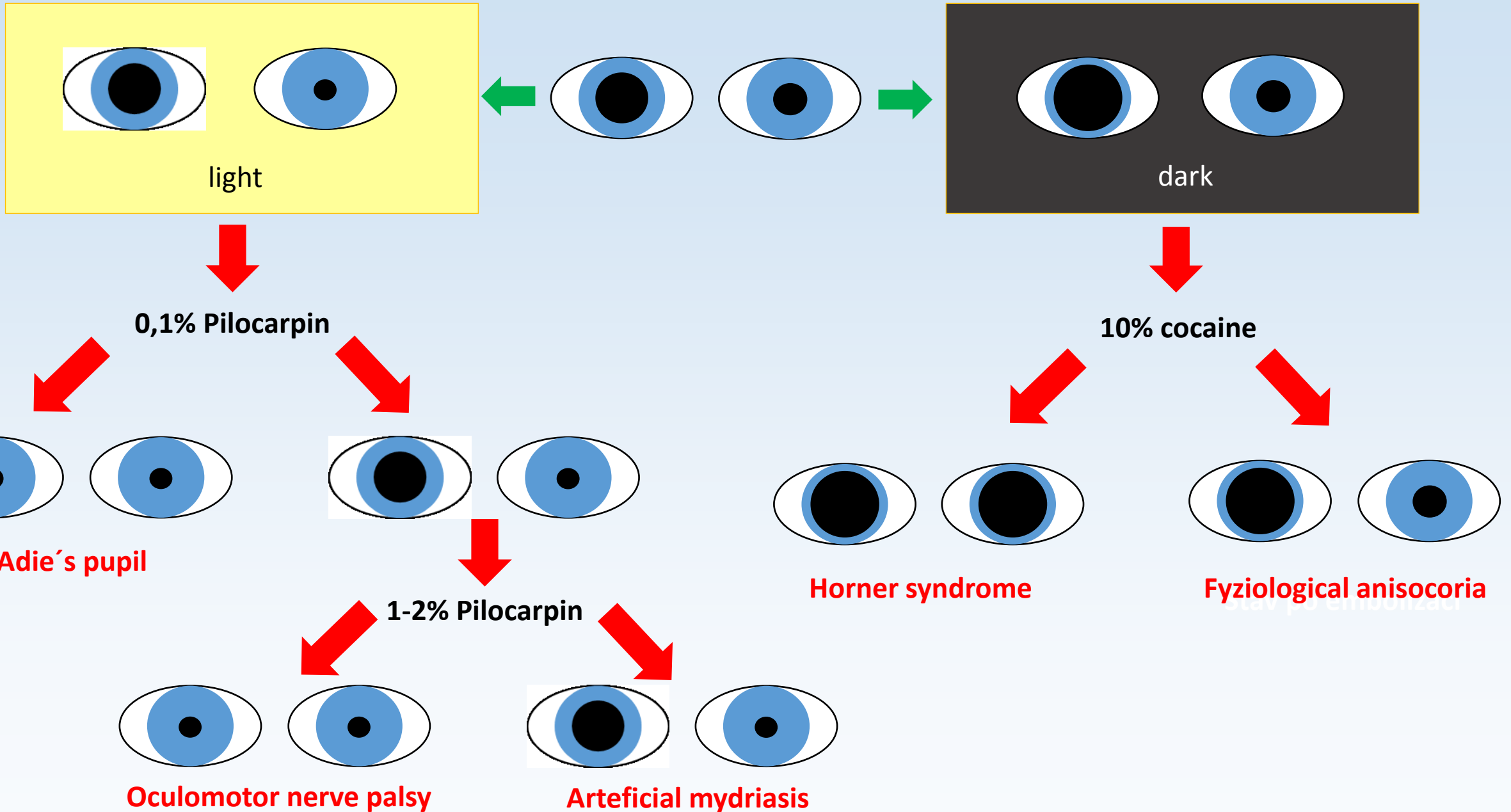
- *Pupilotonia (Adie's pupil)* – dilated pupil not responding to light, worm-like movement within the pupil, accommodation disorder, diminished patellar reflex and Achilles tendon reflex
- *Argyll-Robertson pupil* – narrow pupil with no reaction to light, preserved accommodation (syphilis, neuropathy, diabetes)
- *Anisocoria* – unequal size of pupils (more than 1mm), often random finding, may be physiological (up to 1mm)

Anisocoria – diagnostic scheme

Which pupil is pathological?



Anisocoria – diagnostic scheme



Horner syndrome

Signs

- Miosis (no mydriasis in dark)
- Ptosis of upper eyelid
- Pseudoenophthalmus
- Anhidrosis (diminished sweating of half part of face)
- Heterochromia (congenital form only)

Etiology

- Trauma, dissection of internal carotid artery, ischemia of brain stem, multiple sclerosis, intracranial tumor, syringomyelia, Pancoast tumor (lung apex tumor), goiter, thyroid carcinoma... **but mostly idiopathic!**



Thanks for your attention!

