# Neuroophthalmology

## for medical students

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# Content of neuroophthalmology

- **Visual pathways affections**
  - disorders and affections of optic nerve chiasmatic lesions, postchiasmatic lesions
  - typical manifestation – decrease of visual acuity and visual field defect
- **Disorders of oculomotor balance**
  - neurogenic ofi myogenic etiology
  - typical manifestation - binocular diplopia and paralytic strabismus
- **Disorders of pupillary reactions**
  - any disorder in any part of pupilomotoric pathway
  - typical manifestation – anisocoria or abnormality in reaction of pupils

**Note: all disorders mentioned above can be present at same time in various combinations**

# Neuroophthalmological examination I.

## Medical history

- crucial – complete medical history = nearly an half of diagnostic success

- Questions about **subjective signs:**
  - Questions: when troubles started, how long last it, any change during time (intermittent / progressive), presence of troubles on fellow eye, personal history, medical history
  - Common complaints – dominant signs from the patient´s view (i.e. visual acuity decrease, diplopia)
- Searching for **objective signs:**
  - i.e. pupillary reaction disorders, oculomotor function disorders, ptosis of upper eyelid, red eye
  - Poor complaints – pacient itself does not be aware with all the signs / troubles

# Neuroophthalmological examination II.

## General ophthalmological examination

- **Visual acuity assesment** (distance) = central visual acuity (VA)
  - natural (with no correction = UCVA) and using the best correction of refractive errors (BCVA), monocular, binocular assesment
- **Basic eye examination**
  - Examination of anterior segment (slit lamp) and posterior segment (direct / indirect ophthtalmoscopy / biomicroscopy)
  - Focus to exclude possible ophthalmological causes of VA decrease (i.e. corneal scars or opacities, cataract, hemophthalmus, retinal pathologies)
- **Visual field examination (VF)** = perimetry
  - Crucial examination in neuroophthalmology
  - Important even with normal VA (*good VA does not exclude visual field defects!!!)*

# Neuroophthalmological examination III.

## Accessory (interdisciplinary) examination

- **Internal**
  - basic + specific samling
- **Neurological**
  - to exclude possible intracranial causes (intracranial expansive lesions, thrombosis, stroke)
  - to exclude neurological sign of possible neurological diseases (multiple sclerosis)
- **Endocrinnne**
  - to exclude thyroid disorders (thyroid associated orbitopathy / ophthalmopathy)
  - to exclude hormonal activity of pituitary gland (pituitary adenoma, chiasmal lesions)

# Neuroophthalmological examination IV.

## Imaging techniques

- **Ultrasonography**
  - ***Targeted tissues:*** eye bulb, soft orbital tissues (including extraocular muscles)
  - ***indications:*** examination of orbital tissues, thickness of extraocular muscles
  - ***Advantages:*** fast, widely available, cheap
  - ***Disadvantages:*** depth limitation (up to one third of orbit)
- **X-ray**
  - ***Targeted tissues:*** orbital bones, paranasal sinuses
  - ***Indications:*** bone fracture exclusions, dislocation of fragmentsexclusion of contrast orbital foreign bodies, exclusion of pathological content in paranasal cavities
  - ***Advantages:*** fast, widely available, cheap
  - ***Disadvantages:*** poor reliability of bone fractures without dislocations, poor assesment of soft orbital tissues

# Neurooftalmological examination IV.

## Imaging techniques

- **Computerized tomography (CT)**
  - ***Targeted tissues:*** brain, orbit (soft tissues, bone part)
  - ***Indication:*** exclusion of cerebral stroke, recent ischemia, intraorbital, intracranial lesion or expansions
  - ***Advantages:*** available, fast
  - ***Disadvantages:*** more expensive than X-ray, higher level of radiation
- **Magnetic resonance imaging (MRI)**
  - ***Targeted tissues:*** brain, soft orbital tissues
  - ***Indications:*** exclusion of small or intracranial / intraorbital lesions with poor contrast
  - ***Advantages:*** no possible harmfull radiation level, good imaging of soft tissues (better than CT)
  - ***Disadvantages:*** worse imaging of bony tissues, incompatibility with older metal implants or metal foreign bodies, more expensive than CT

# Optic nerve

## characteristics, physiology

### Characteristics

- 2nd cranial nerve
- Embryologically part of CNS (together with neural part of retina)
- Anterior part of visual pathway

### Physiology

- Pure senzoric nerve
- Every optic nerve = 1.2 million of nerve fibers
    - 80% of nerve fibers - **sensoric visual information**
    - 20% of nerve fibers - **pupilomotoric information** (major part of  afferent pupilomotor pathway)

# Optic nerve

## anatomy

### Composition

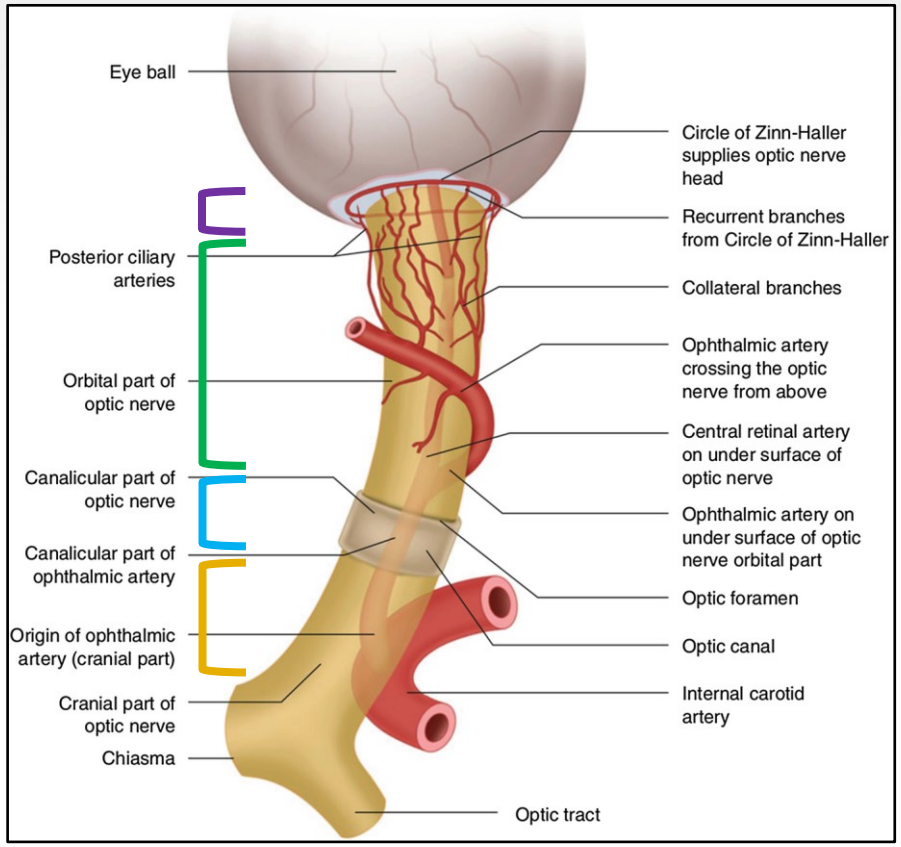
- **axons of ganglionic cells**
- neuroglia
- Optic nerve sheaths = sheats of CNS (pia mater, arachnoidea, dura mater)

### Anatomical division

- **Intraocular part**
- **Intraorbital part**
- **Intracanalicular part**
- **Intracranial part**

### Blood support

- **Posterior cilliary artery**
  - **Ophthalmic artery**
    - **Internal carotid artery**



# Optic nerve affections:

## General characteristics

**Typical clinical signs:**

- **Decrease of visual acuity** – most prominent sign, usually unilateral, represents the amount of lesion of nerve fibers predominantly from central part of retina (macula)
- **Visual field defect** – represents the amount of lesion of nerve fibers in general
- **Color vision defect** – less prominent but almost always present sign; but not specific (acquired disorder)
- **Relative afferent pupillary defect (RAPD)** – typically unilateral, present of size of affection; due to the affection of afferent part of pupillomotoric pathway

# Pathologies of optic nerve

## division

**1) congenital anomalies**

hypoplasia, coloboma, Morning glory syndrom, tilted disc, fibrae medullares, optic disc drusen

**2) inflammatory affections - neuritis**

demyelinisating, infection, paraneoplastic

**3) non-inflammatory affecions - neuropathies**

ischaemic, toxic, nutritive

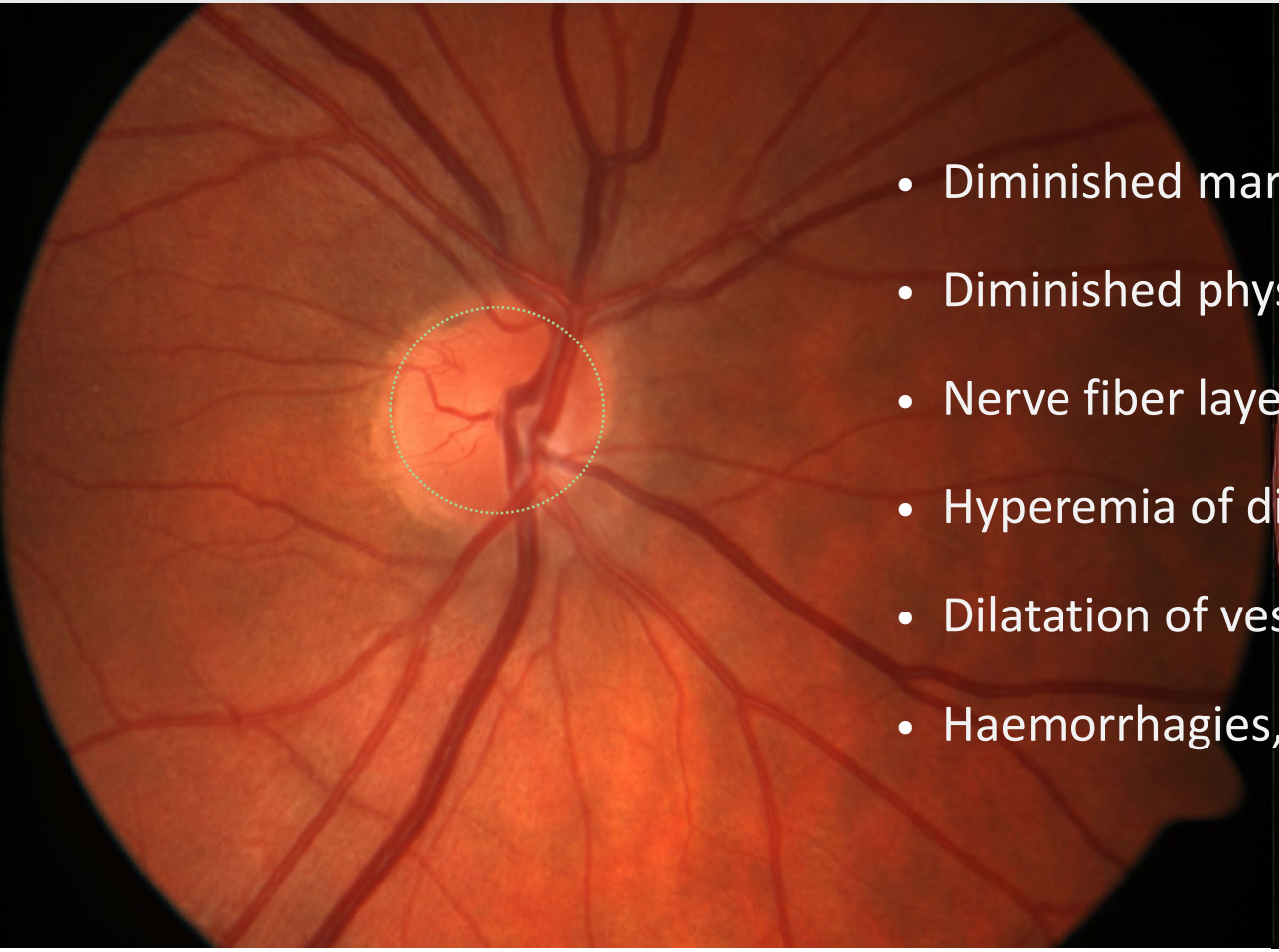
**4) Bilateral papiledema**

elevation of intracranial pressure

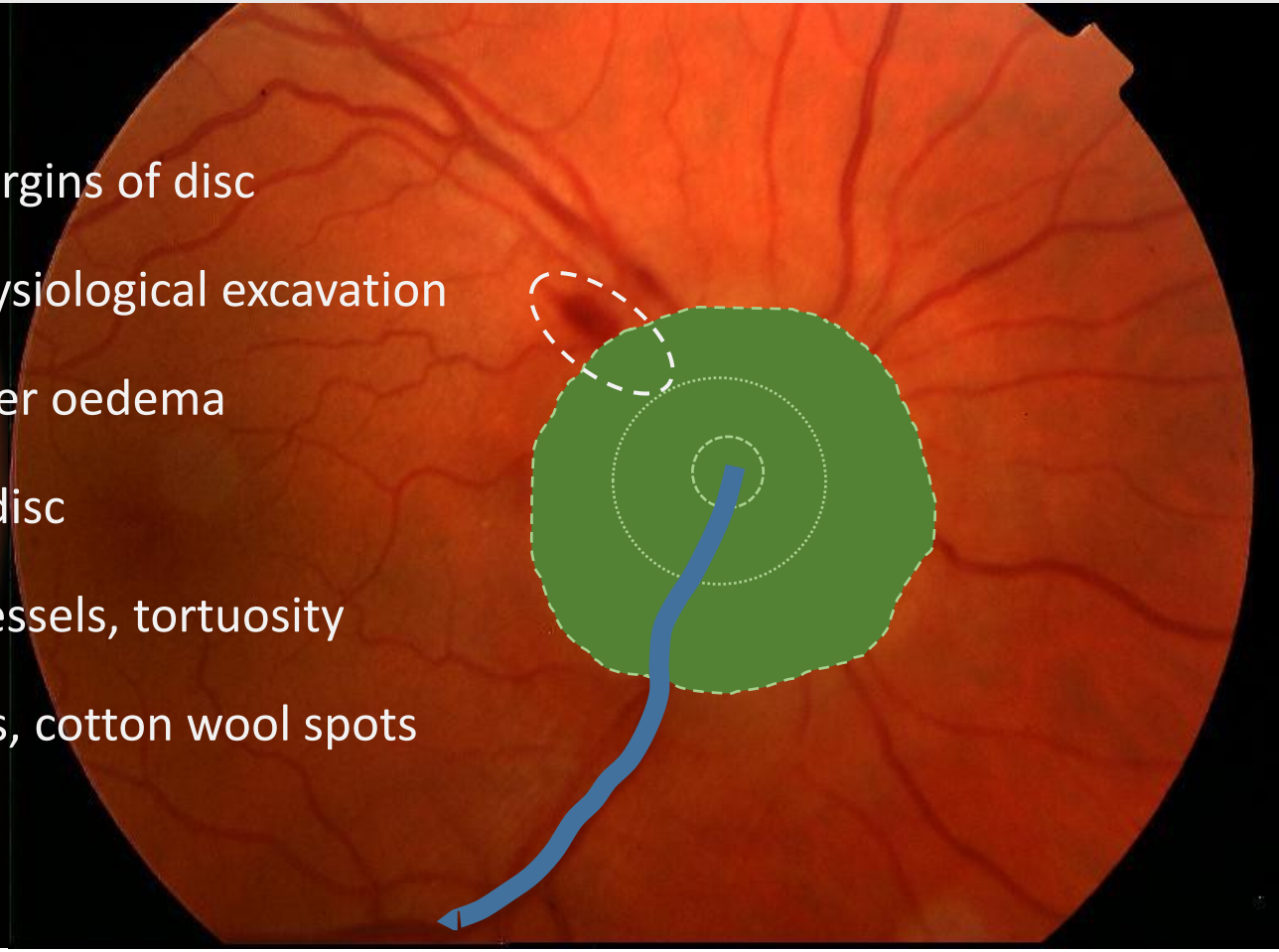
# Papiledema

## clinical picture

- Diminished margins of disc
- Diminished physiological excavation
- Nerve fiber layer oedema
- Hyperemia of disc
- Dilatation of vessels, tortuosity
- Haemorrhagies, cotton wool spots



**Physiological appearance**



**Papilloedema**

# Papiledema

## Clinical picture during time

- Dynamic state during time
- Depends of cause, duration, and therapy

**Example:** development in intracranial hypertension (young female), treatment by using acetazolamide.

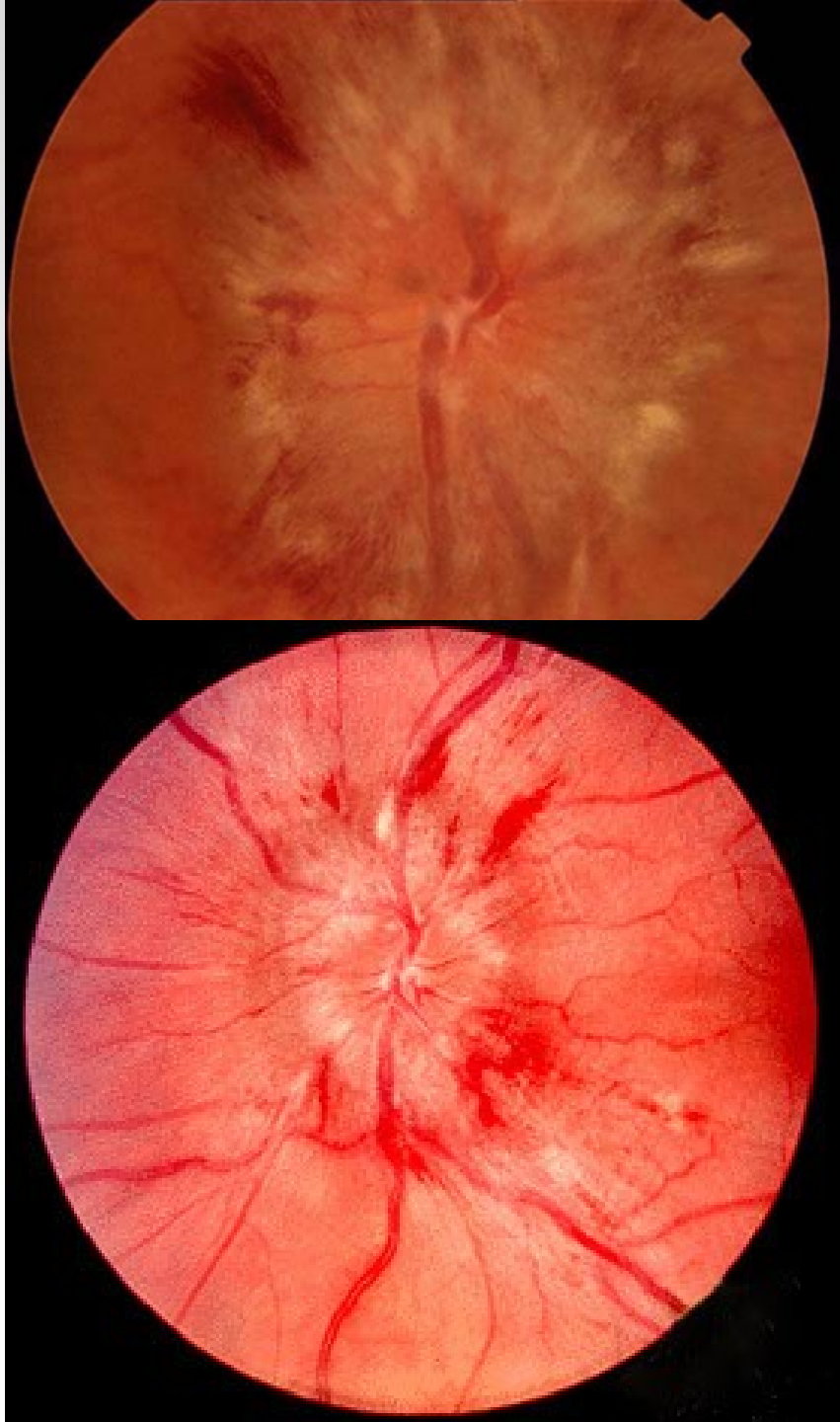
# Bilateral papiledema

## Etiology

- Increase of intracranial pressure and spreading within sheaths
  - 75% of cases – **intracranial tumor!!!** (frontal lobr or chiasmal area)
  - Rest of cases – pseudotumor, meningitis, AV malformation, thrombosis

## Clinical pisture

- **Subjective signs:**
  - Short lasting visual impairment (obnubilations)
  - Headaches (worse in horizonal position)
- **Objective signs:**
  - Bilateral oedema of optic nerve disc
  - Enlargement of blind spot – in visual field testing



# Optic neuritis

## Clinical picture

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

## Causes

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

## Types

- intraocular
- retrobulbar – most common

## Prognosis

- usually good – regression after intravenous corticoids

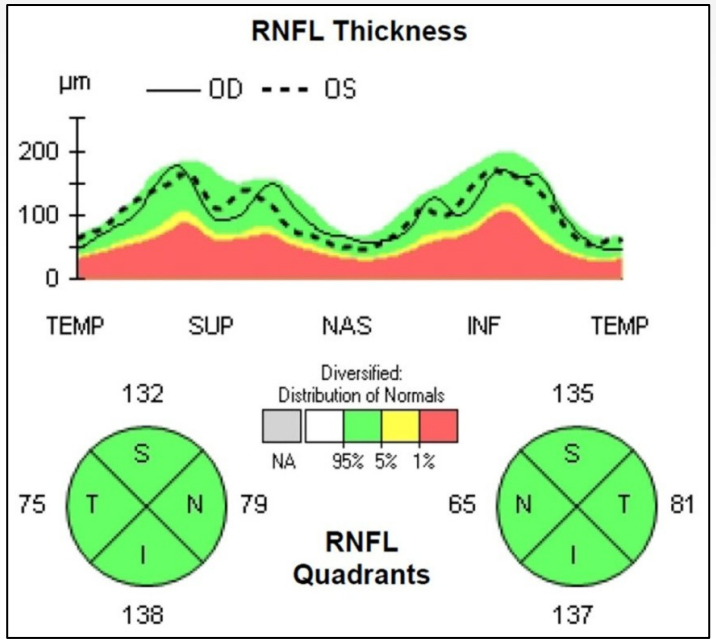
## Epidemiology

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

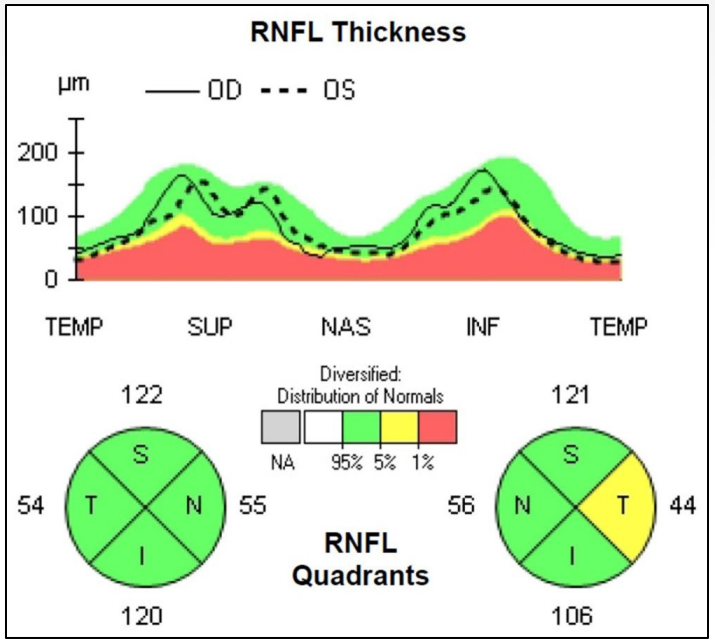
# Optic neuritis
## diagnostics

### Optical coherence tomography

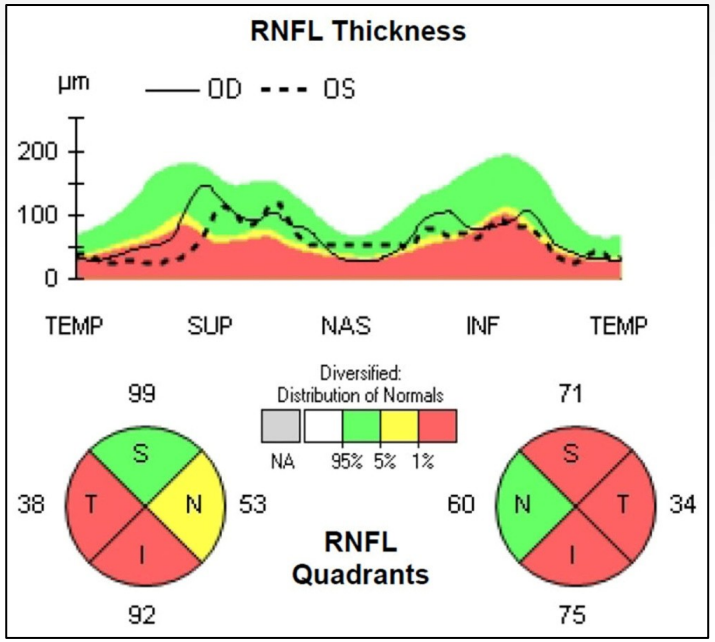
- Accessory examination of MS
- Dynamics of changes of RNFL during time
- Decrease of RNFL is **objective proof of postneuritic atrophy of optic nerve**




**Physiological findings**
Healthy pacient

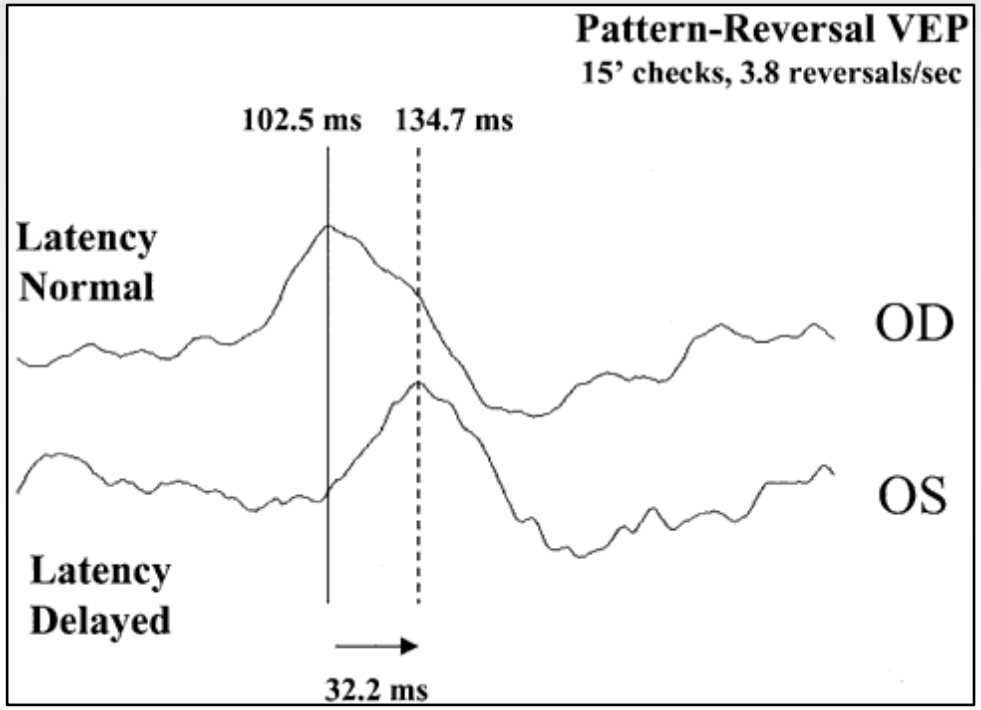



**Condition after optic neuritis**
Left eye (clinically isolated syndrome)

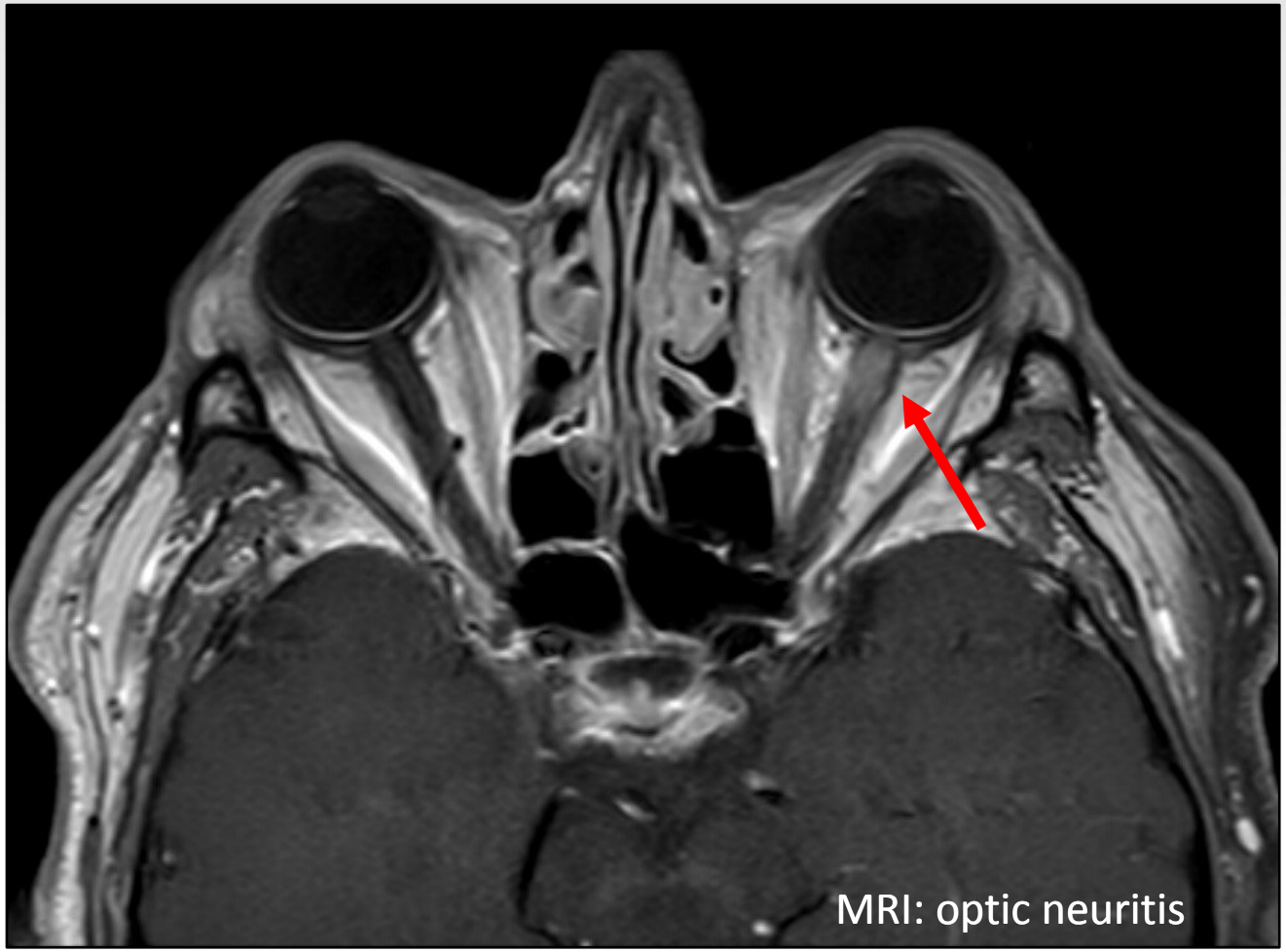



**Multiple sclerosis**
Contition after bilateral optic neuritis

# Optic neuritis
## diagnosis



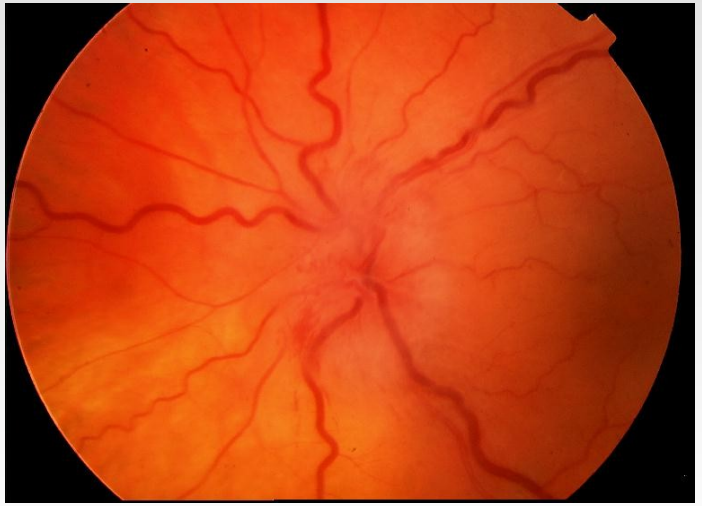

VEP: delayed latency P100



# Anterior ischemic optic neuropathy

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

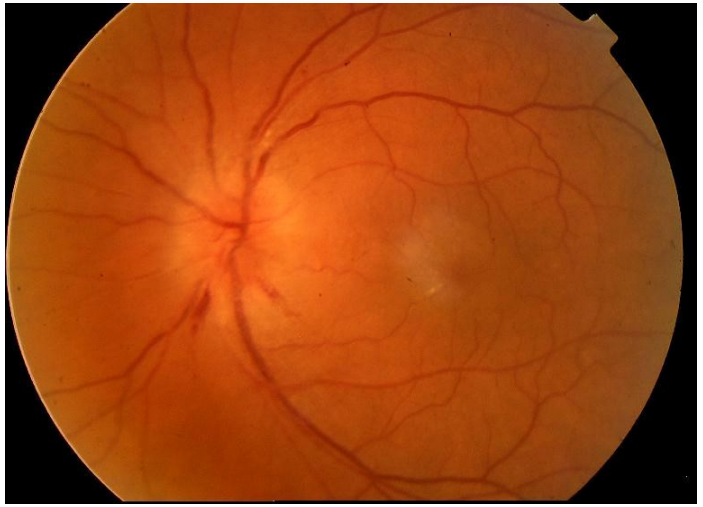
**Cause** – affection of short ciliary arteries



## Epidemiology

- 50 years of age and more

## Clinical picture

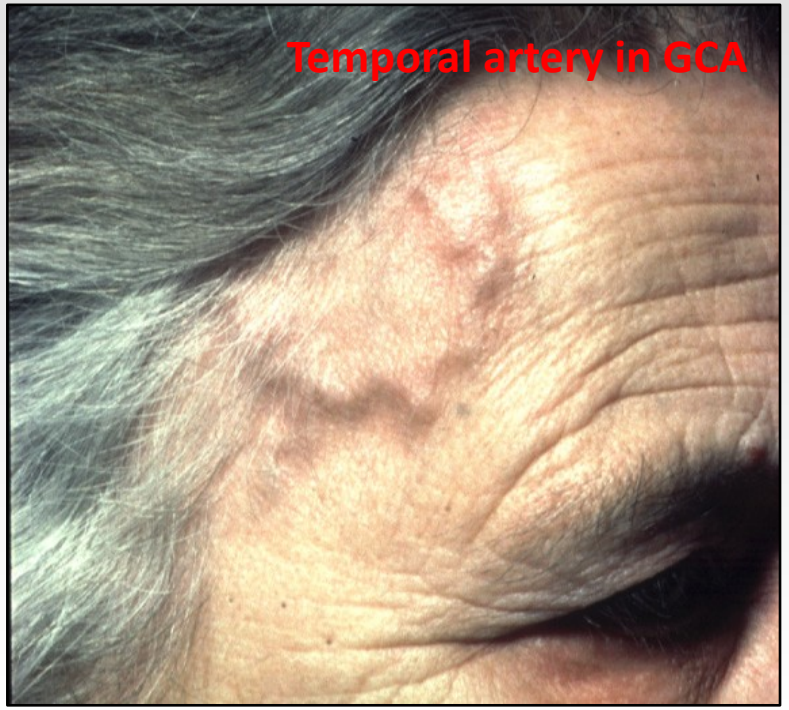


- loss of visual acuity - fast onset, painless (light perception to almost normal values)
- monocular visual field defect – altitudinal scotoma
- unilateral ischemic optic disc oedema

# 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

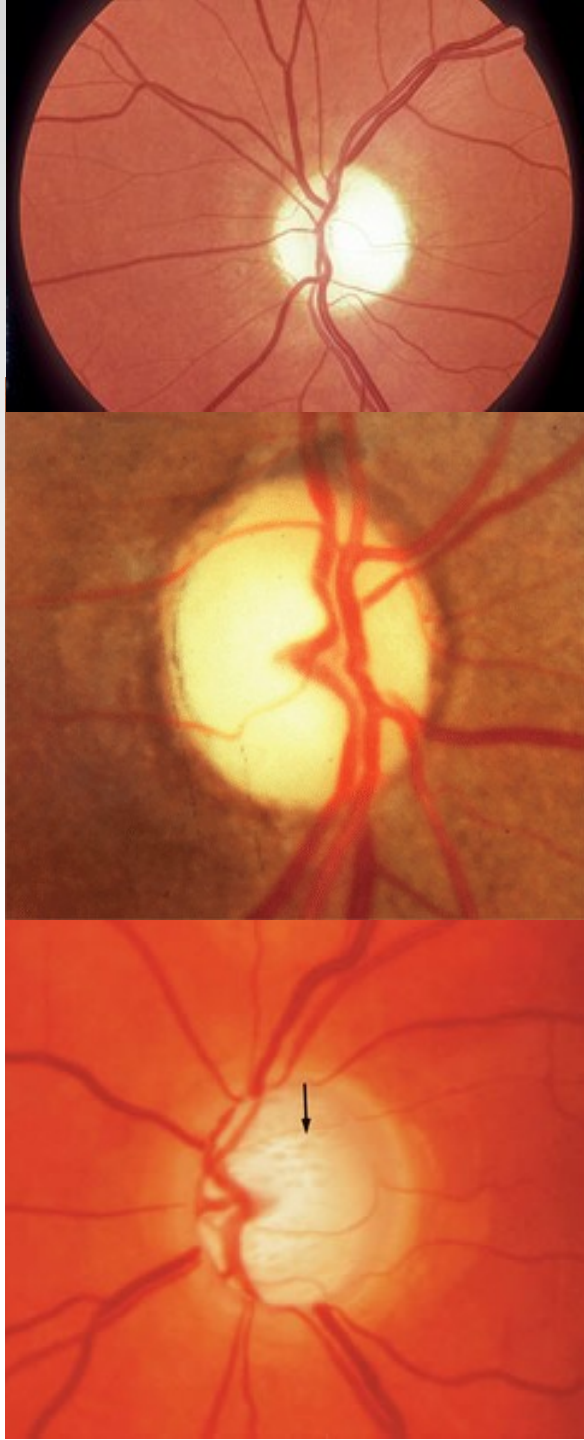
- Irreversible loss of axons
- After various optic nerve affections

**Etiology**

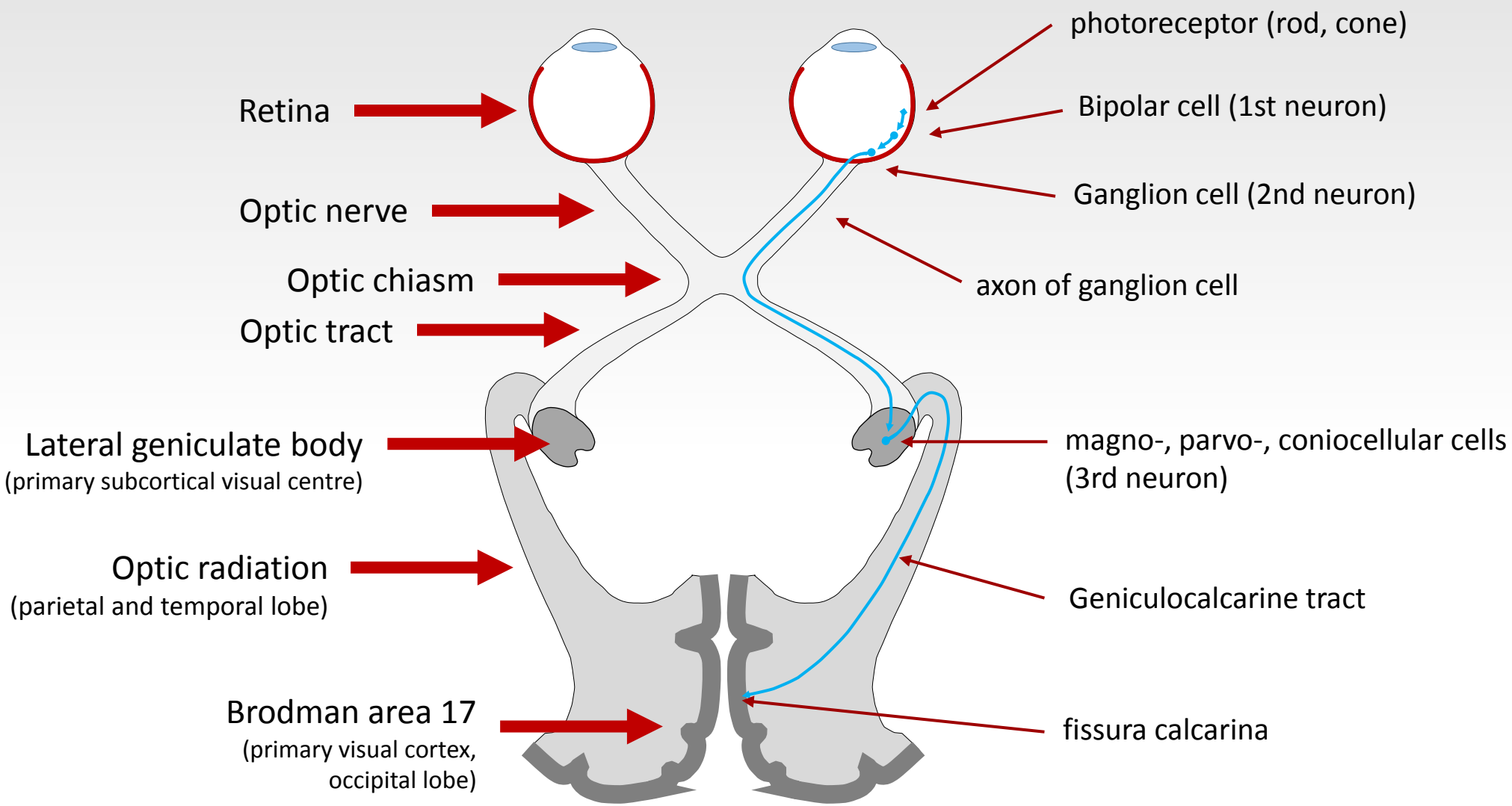
- *Primary* – posttraumatic, by direct pressure of tumor
- *Secondary* – affection of optic nerve (ischemia, inflammation)
- *Glaucomatous* – due to elevated intraocular pressure

**Clinical picture**

- Pale optic disc
- Reduction of smaller vessels



# Anatomy of visual pathway



# Anatomy of visual pathway – clinical notes

**Optical apparapart of eye switch the picture**, therefore:

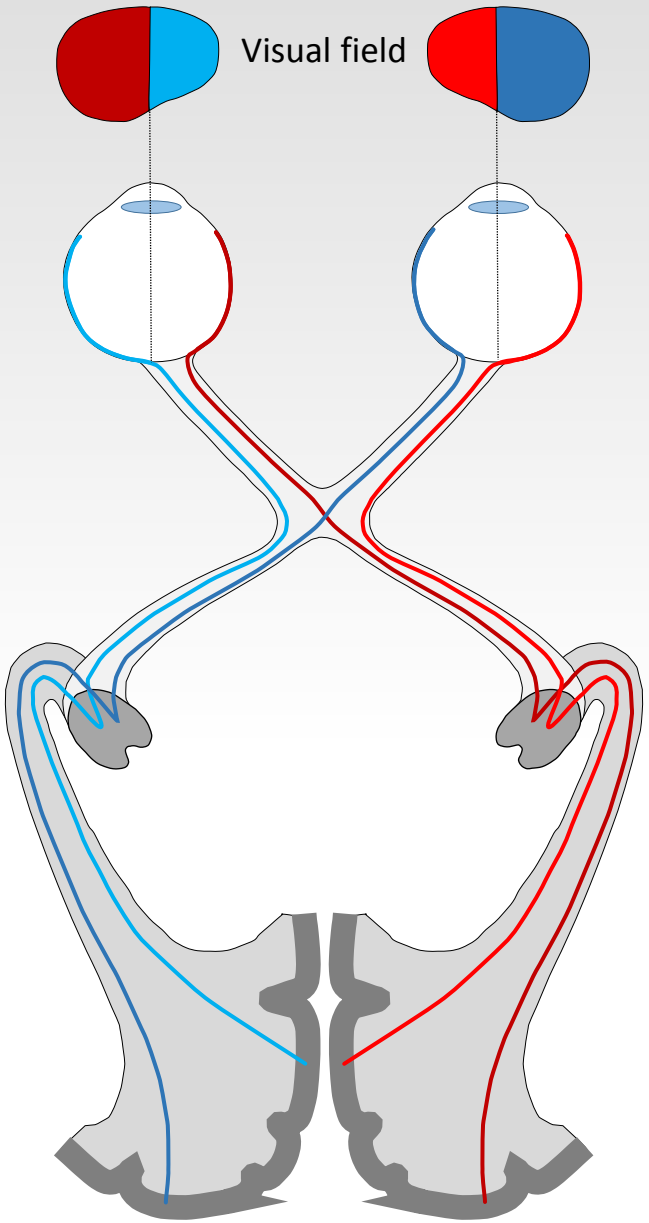
- **Temporal part of retina** process information *from* ***nasal part of VF***
- **Nasal part of retina** process information from ***temporal part of VF***

**Partial crossing** of nerve fibers in **optic chiasm**:

- **Nerve fibers originating from temporal part of retina are <u>not crossing</u>** and continue on same size of CNS
- **Nerve fibers originating from nasal part of retina are <u>crossing</u>** and continue on opposite size of CNS

**Size lateralisation** of visual information in visual cortex:

- **Right hemisphere** process information from ***left part of VF*** of both eyes
- **Left hemisphere** process information from ***right part of VF*** of both eyes



# Visual pathway lesion and visual field defects

**Optic nerve lesions**

- Monocular defects of VF

**Lesions in chiasmal area**

- Usually bilateral heteronymn defects of VF

**Lesions of optic tract**

- Bilateral (incongruent) homonymn defects of VF

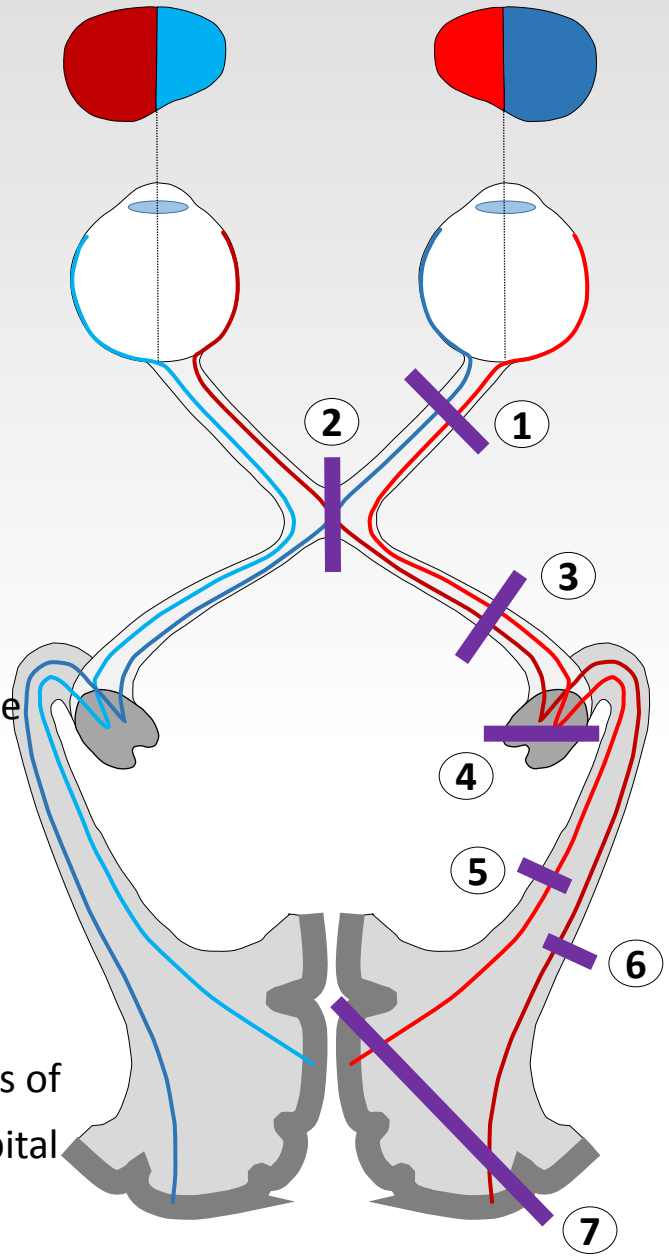
**Lesions of lateral geniculate body**

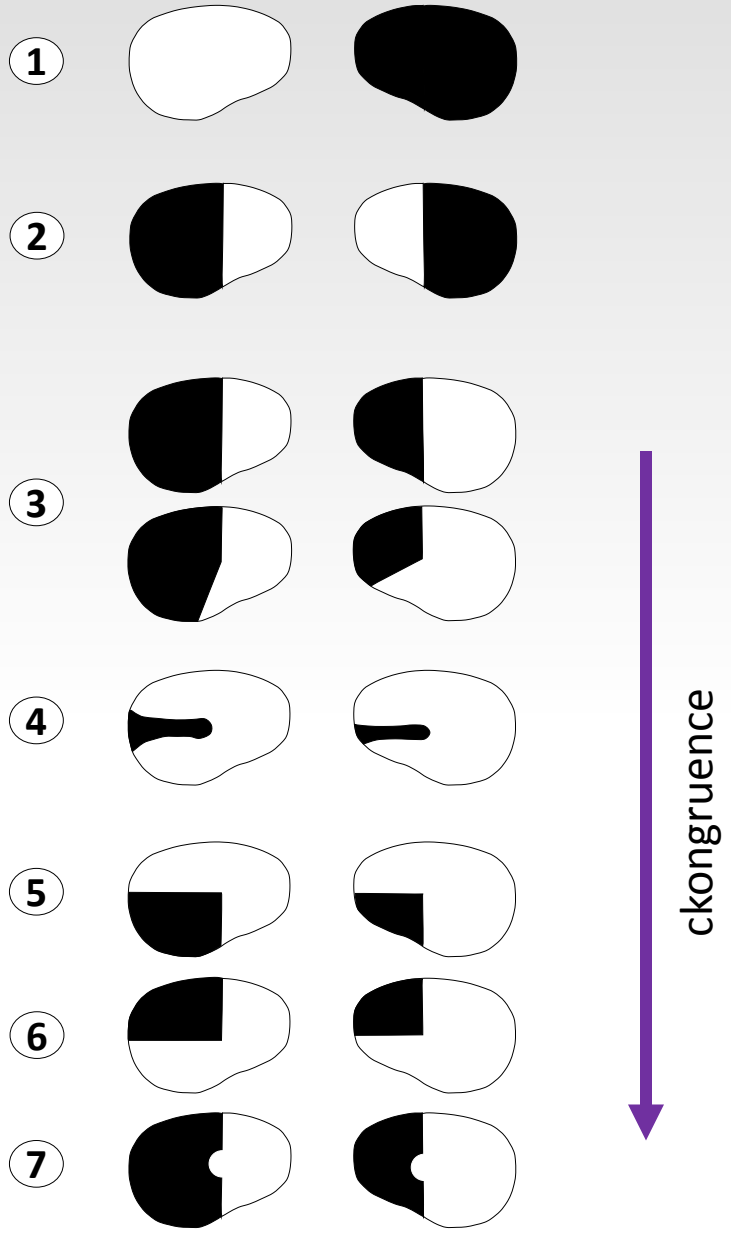
- Bilateral homonymn defects  along horizontal line

**Geniculocalcarine tract lesions**

- bilateral congruent* defects of VF

* **Pozn.:** ***congruence*** – bilateral symmetry of defects of visual field increasing with closer proximity to occipital cortex



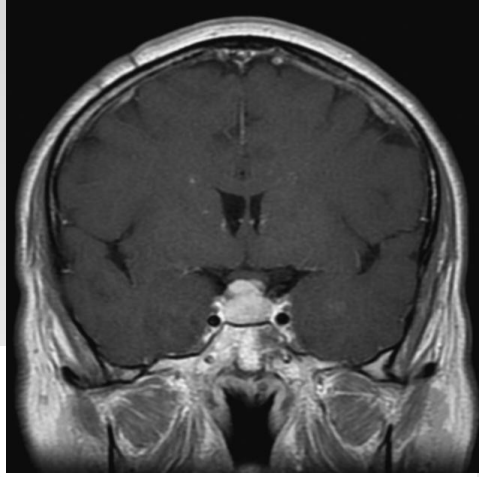



# Chiasmal syndrome

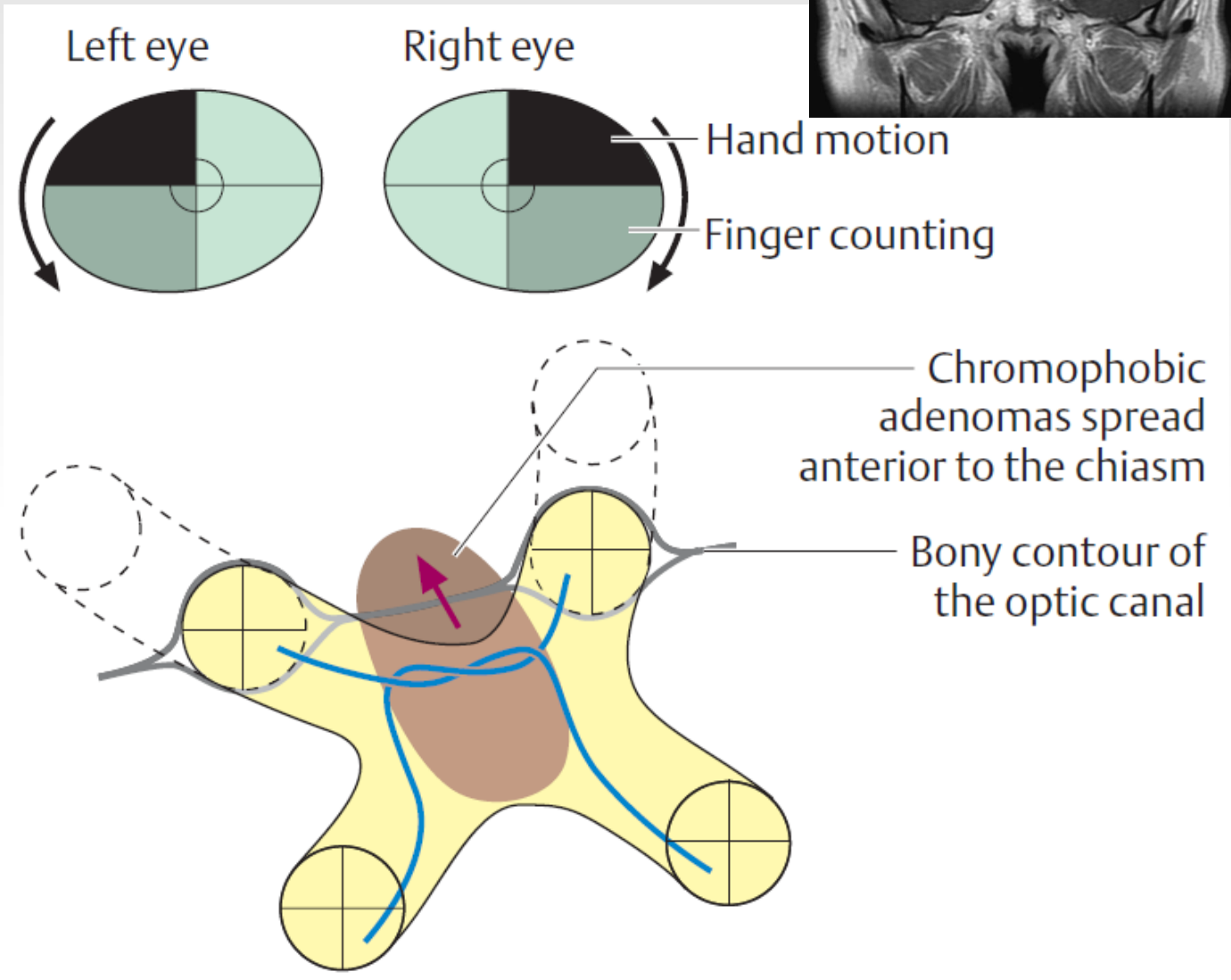
- lesions in chiasmal area
- typically compressive, expansive condition
- typical visual field defects – use in diagnosis

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

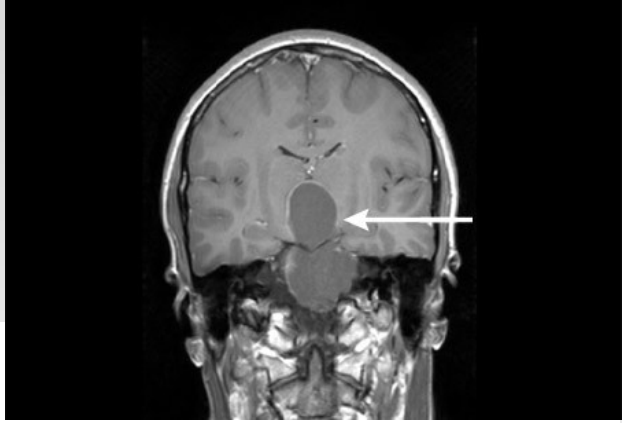
# Pituitary adenoma



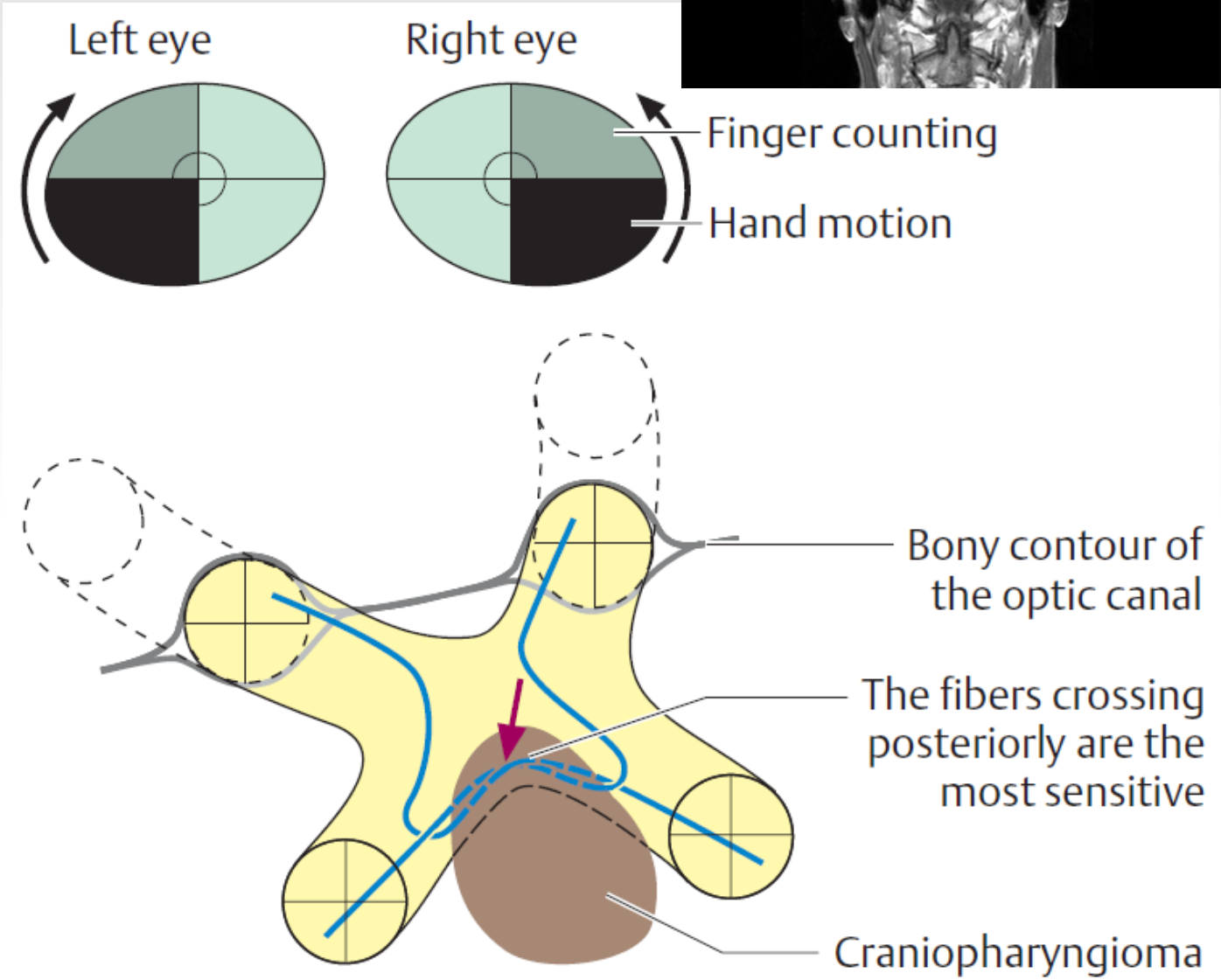
- benign tumor of pituitary gland
- classification
  - by size – microadenoma (up to 10mm), macroadenoma (more than 10mm)
  - biological activity – benign adenoma, invasive adenoma, adenocarcinoma
- possibility of metabolical activity (e.g. prolactinom) a
- compression and lesion of optic chiasm by tumor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



# Craniopharyngioma

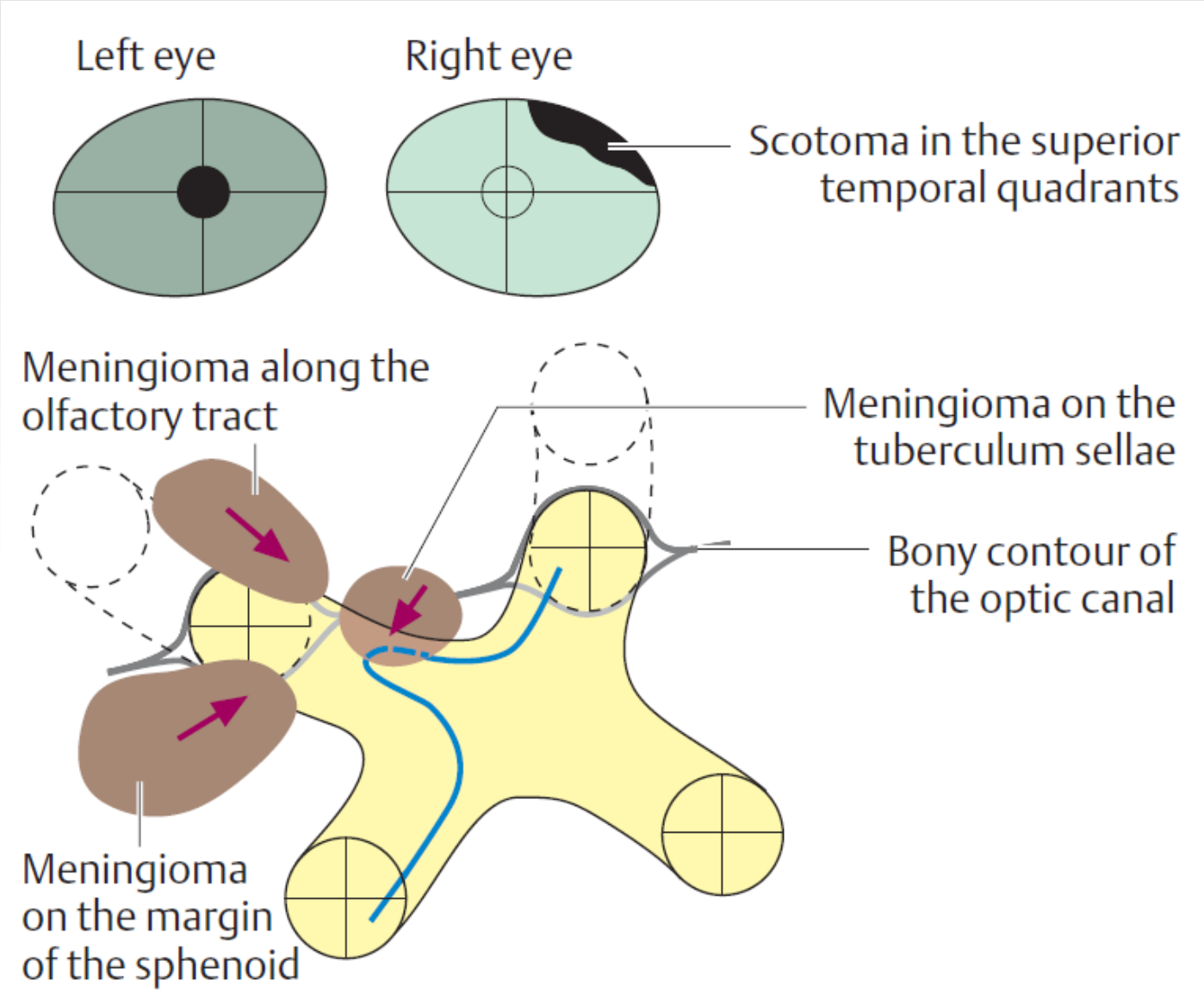


- benign rare type of tumor from pituitary gland embryonal tissue
- pressure on nearby tissue, typical visual field defects – **bitemporal hemianopsia** – first starting as lower quadrantanopsia
- therapy
  - *surgical* - transsphenoidal adenectomy)
  - *radiotherapy*



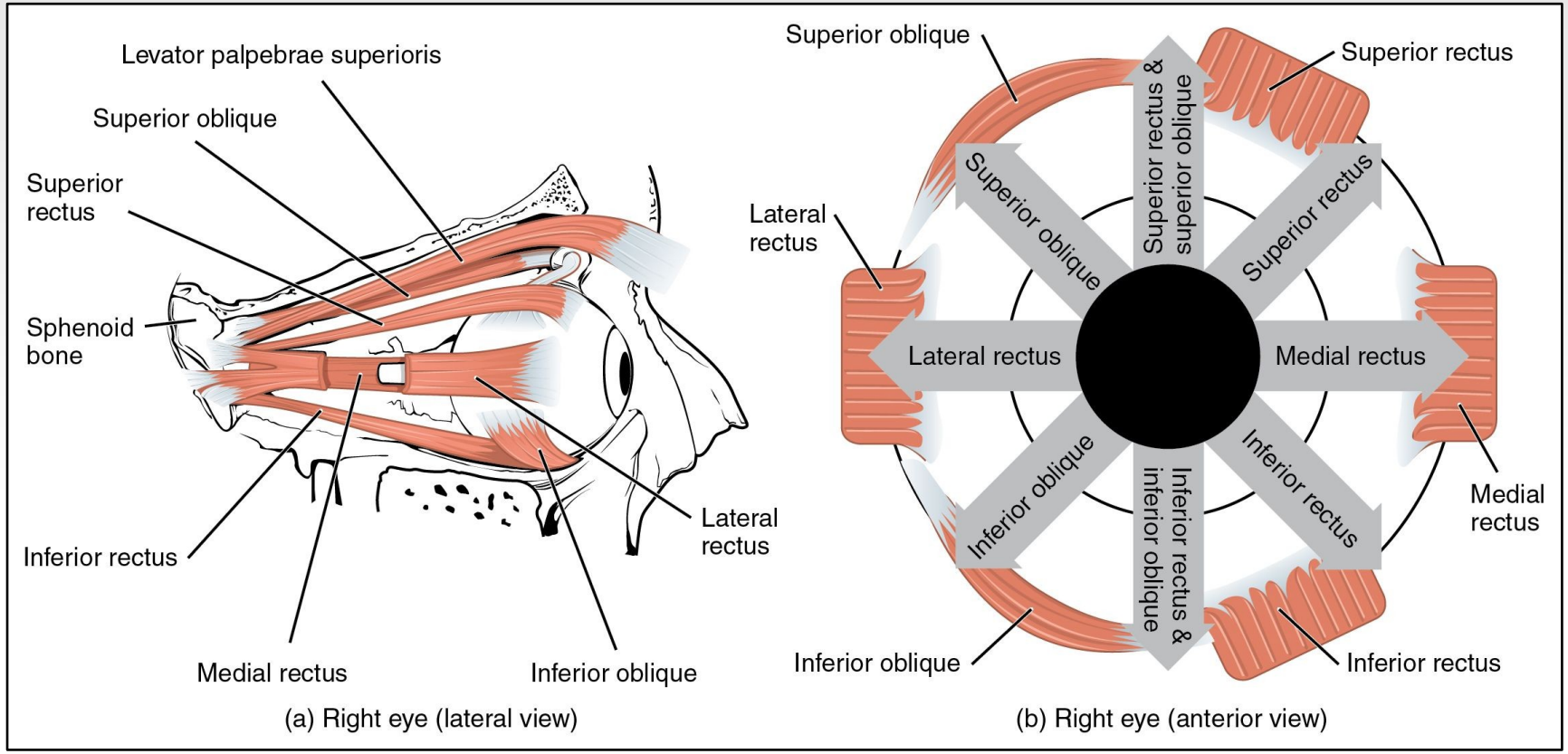
# Meningiomas

- 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

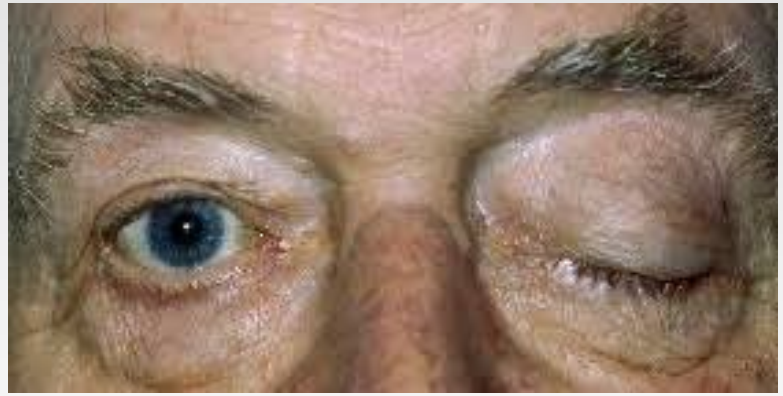
- **Isolated palsies**
  - oculomotor nerve palsy
  - trochlear nerve palsy
  - abducent nerve palsy
- **Ophthalmoplegia**
  - Combination of affection of 2 or 3 nerves
    - cavernous sinus syndrome
    - orbital apex syndrome
    - carotido-cavernous fistula

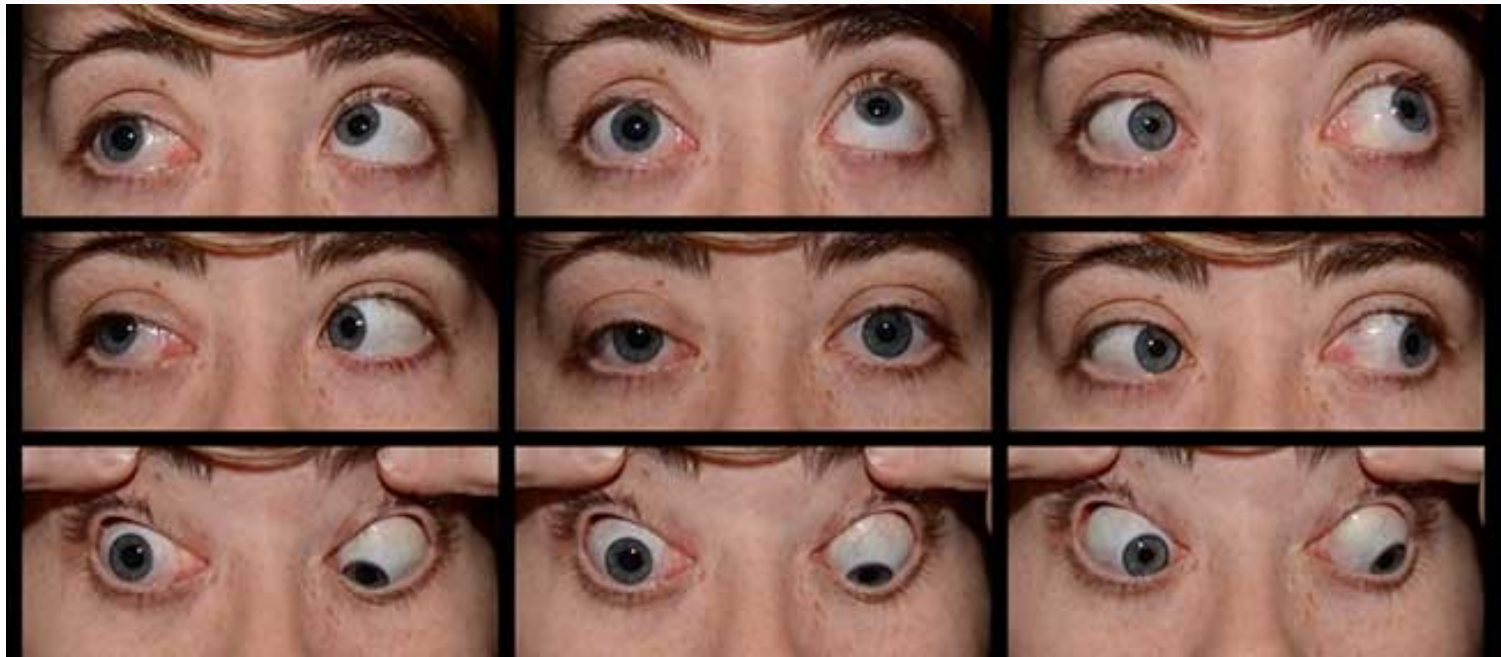
# Isolated palsies - causes

- **Oculomotor nerve palsy** – *aneurysm* (most common), less common tumor, trauma, ischemia (diabetic neuropathy)
- **Trochlear nerve palsy** – *trauma* (fall on head; most common sign), ischemia of brainstem, tumor, half of cases idiopathic
- **Abducens nerve palsy** – *trauma* (most common), ischemia (diabetic neuropathy), intracranial hypertension (sometimes first manifestation), meningitis, tumor in close proximity of brainstem, idiopathic

# Oculomotor nerve palsy

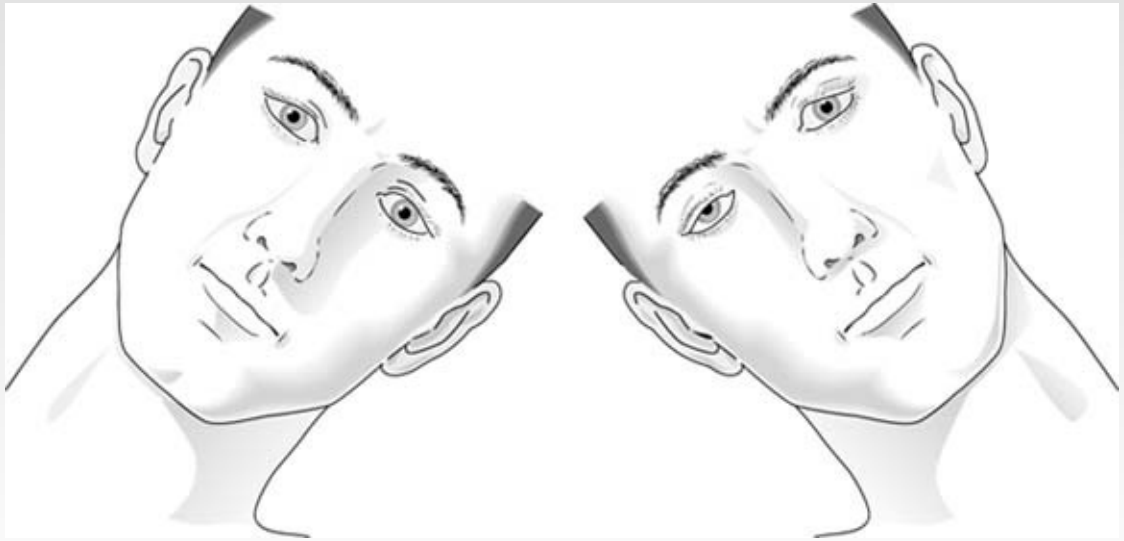
- **Upper eyelid ptosis**
- **Convergence insufficiency**
- **Eye movement disorder** – in multiple sizes of gaze (nasal, up, down)
- **Diplopia** (mixed – horizontal and vertical)
- **Anisocoria** (mydriasis on affected size)

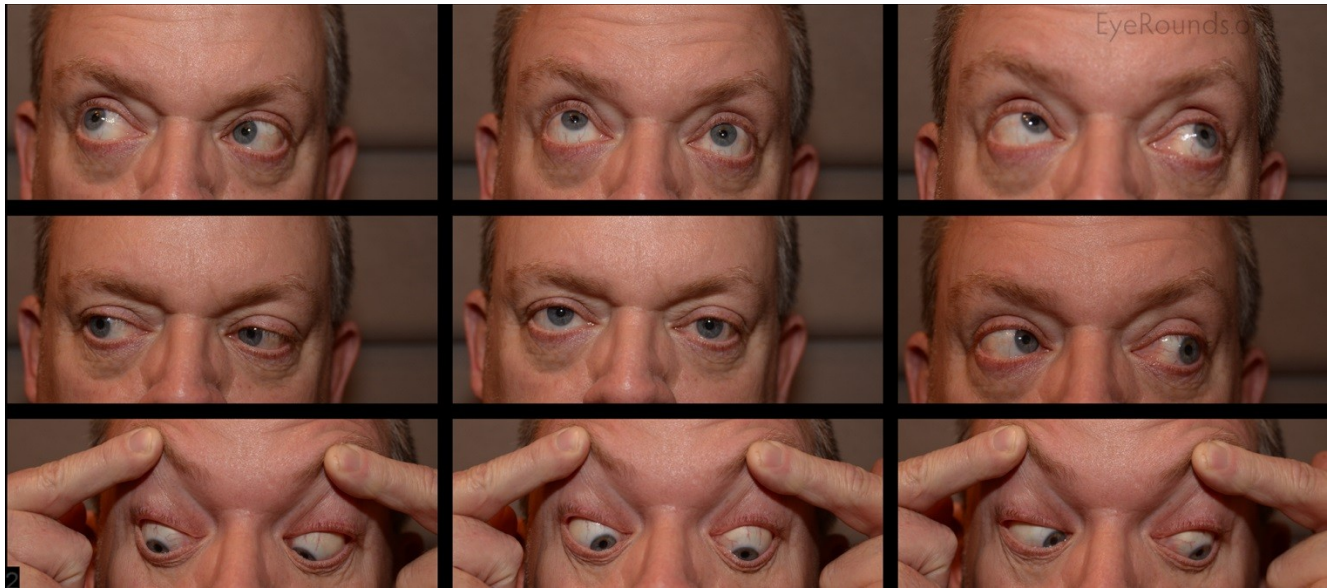




# Trochlear nerve palsy

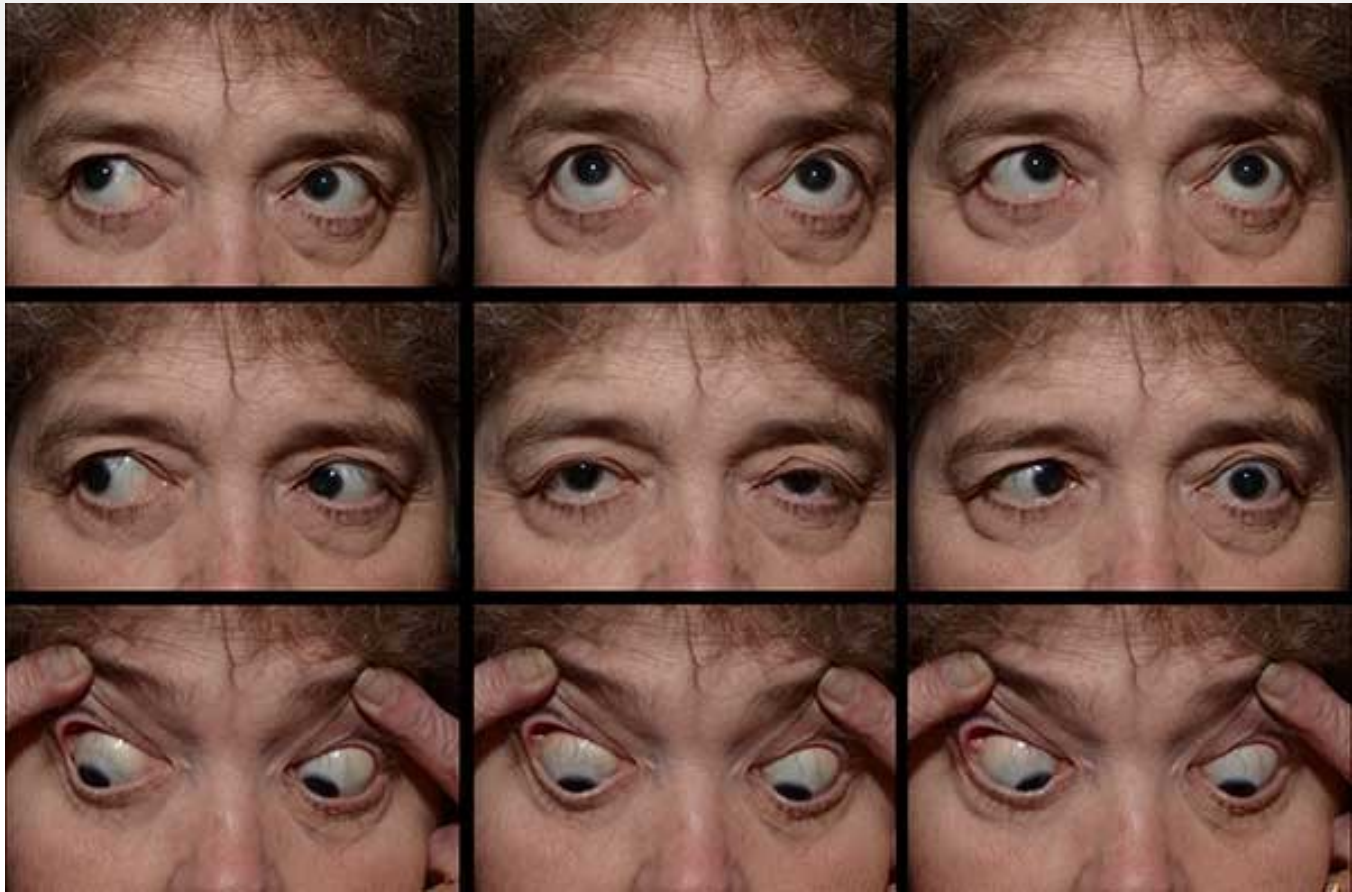
- **Diplopia** – vertical, major manifestation in downgaze or walking downstairs)
- **Eye movement disorder** – affected eye with small hypertropia, not necessary visible!
- **Compensation head posture** (Torticollis) – chin turning down, head posture at non affected size



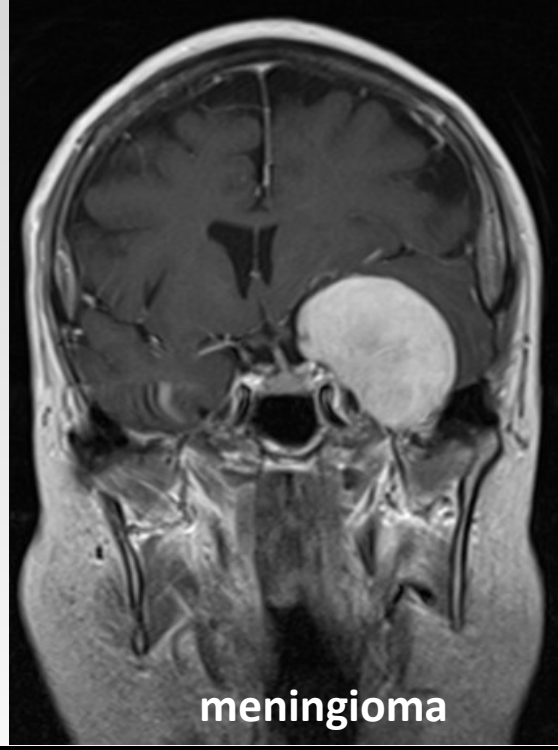


# Abducens nerve palsy

- **Diplopia** – typically horizontal, major manifestation in gaze to affected size
- **Eye movement disorder** – insufficiency of movement laterally (insufficiency of abduction)
- **Compensation head posture** – head turned laterally on affected size



# Cavernous sinus syndrome

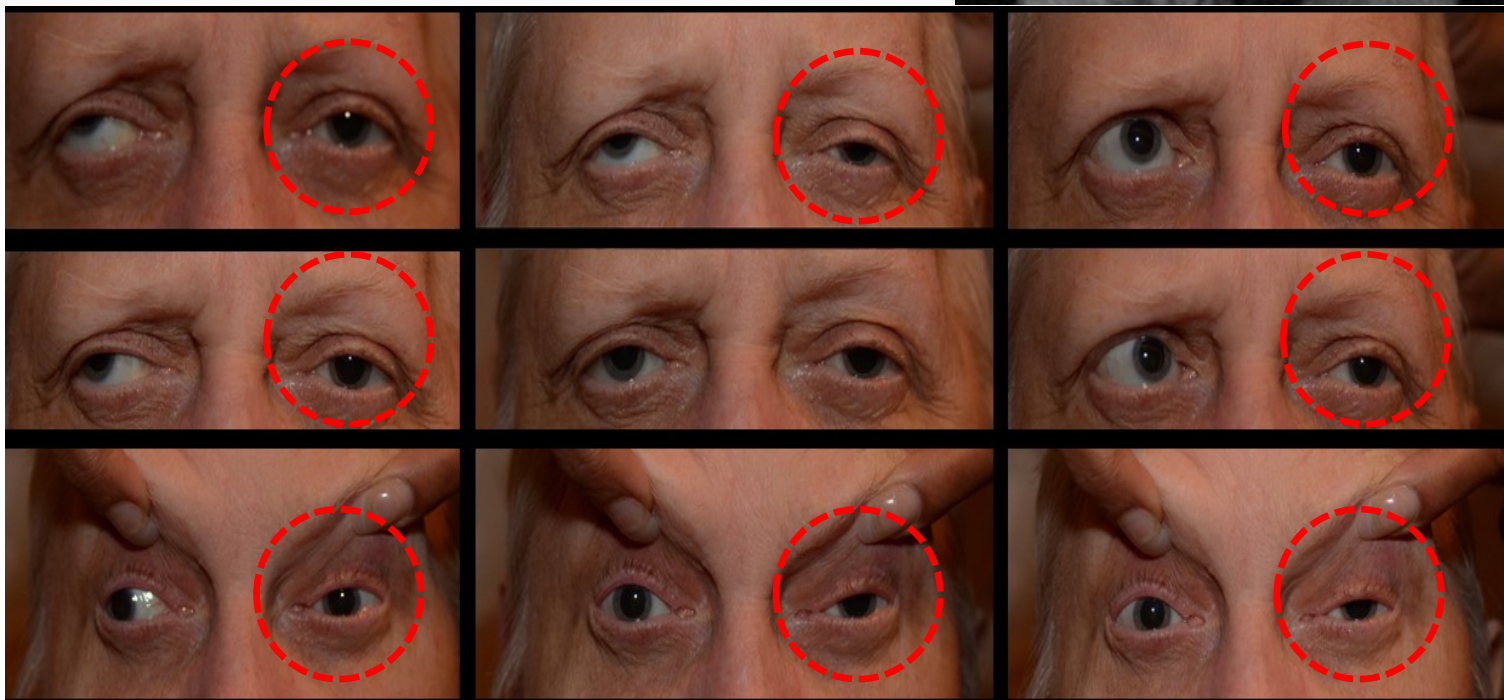



## Etiology

- Compressive / infiltrative lesion in cavernous sinus (thrombosis, tumor, metastasis, aneurysm)
- Lesion of multiple oculomotor nerves (oculomotor, trochlear, abducens) and trigeminal
- ***Absence of optic nerve lesion***, therefore the onset of ***diplopia***

## Clinical picture

- **Upper eyelid ptosis** (oculomotor nerve palsy)
- **ophthalmoplegia** (oculomotor nerves palsy)
- **diplopia** (no affection of optic nerve)
- **Mydriasis** (oculomotor nerve palsy)
- **exophthalmus** (oculomotor nerves palsy)
- Trigeminal pain



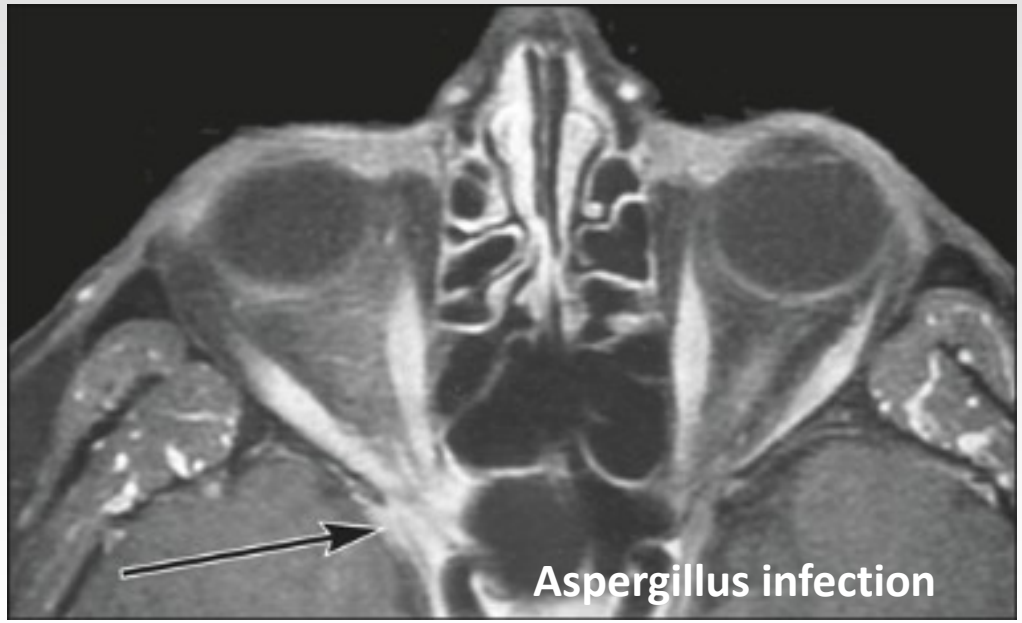
# Orbital apex syndrome

## Etiology

- Compressive / infiltrative lesion in the orbital apex (tumor, metastasis, inflammation)
- **Lesion of oculomotor, trochlear, abducens nerve and <u>also optic nerve</u>**

## Clinical picture

- **Upper eyelid ptosis** (oculomotor nerve palsy)
- **Ophthalmoplegia** (multiple oculomotor nerves palsy)
- **Decrease of visual acuity** (affection of optic nerve)
- **Exophthalmus** (multiple oculomotor nerves palsy)
- **Absence of diplopia** (the more severe lesion of optic nerve, the lower presentation of diplopia)
- Trigeminal pain



# Pupillary reactions

## Physiological appearance of pupils

- **Shape**
  - Always ***round and regular pupil*** even in dark or light
- **Size**
  - **isocoria** – equal size of pupils
    - *under fotopic conditions* - **miosis**
    - *Under photopic conditions* - **mydriasis**
  - **physiological anisocoria** – inequal size of pupils up to 1mm; cca 20% of population
    - Difference in size is usually preserved even in dark or light
  - Dependence of size of pupils:
    - Autonomous nervous sytem (i.e. anxiety, fear, stress, rest, sexual excitation…)
    - Medication, drugs

# Pupillary reactions

## Physiological reactions of pupil

- **Mydriasis** (wide pupil)
  - ***Sympathetic part*** (innervation - dilatator muscle)
- **miosis** (narrow pupil)
  - ***Parasympathetic part*** (innervation - sphincter muscle)
- **Consensual reaction** of both pupil even in the case of enlightenment of one pupil only

# Pupillary reaction

## diagnostic testing

### Photoreaction testing

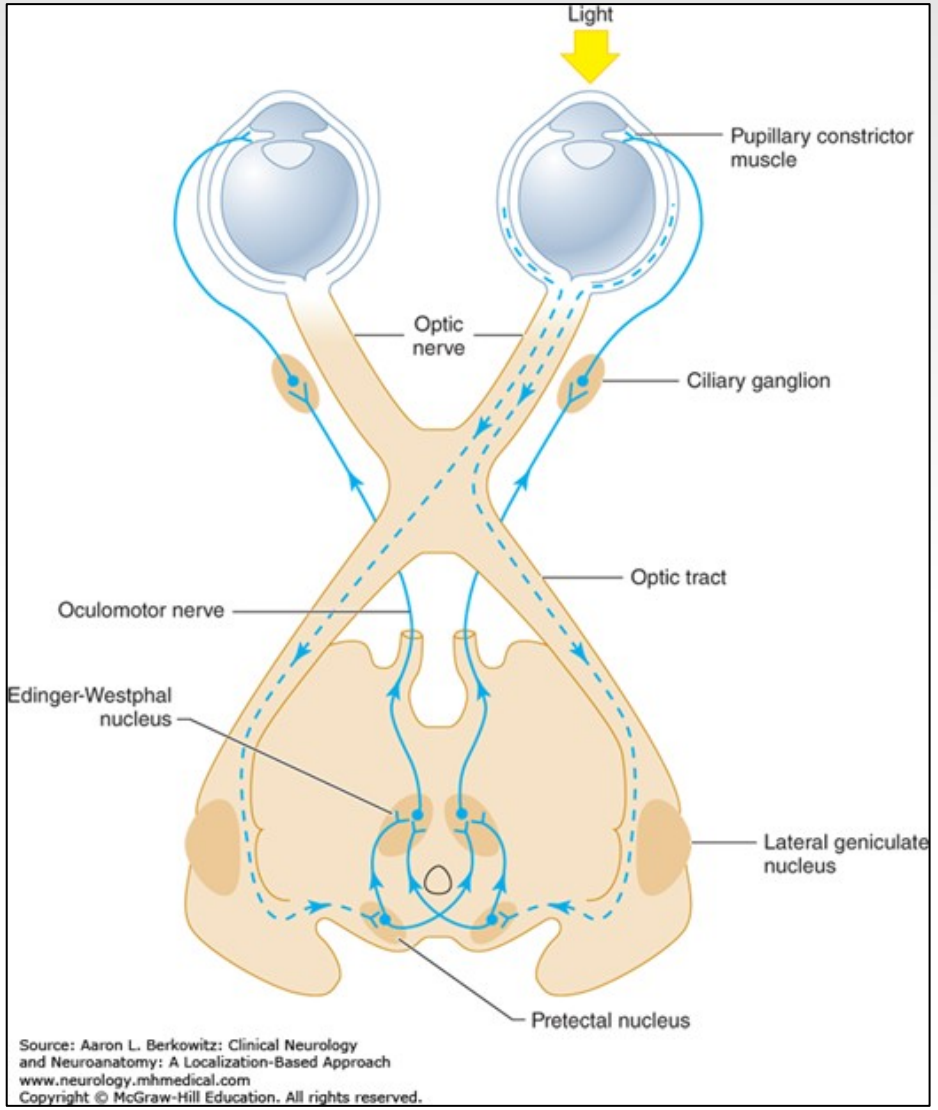
- Reaction to direct and indirect enlightenment – physiological miosis of both pupils at same time

### Test near response

- Testing of accomodation-convergence reflex
- Physiological miosis associated with convergence and accomodation (fixation to object moving closer to eyes)

### Pharmacological testing

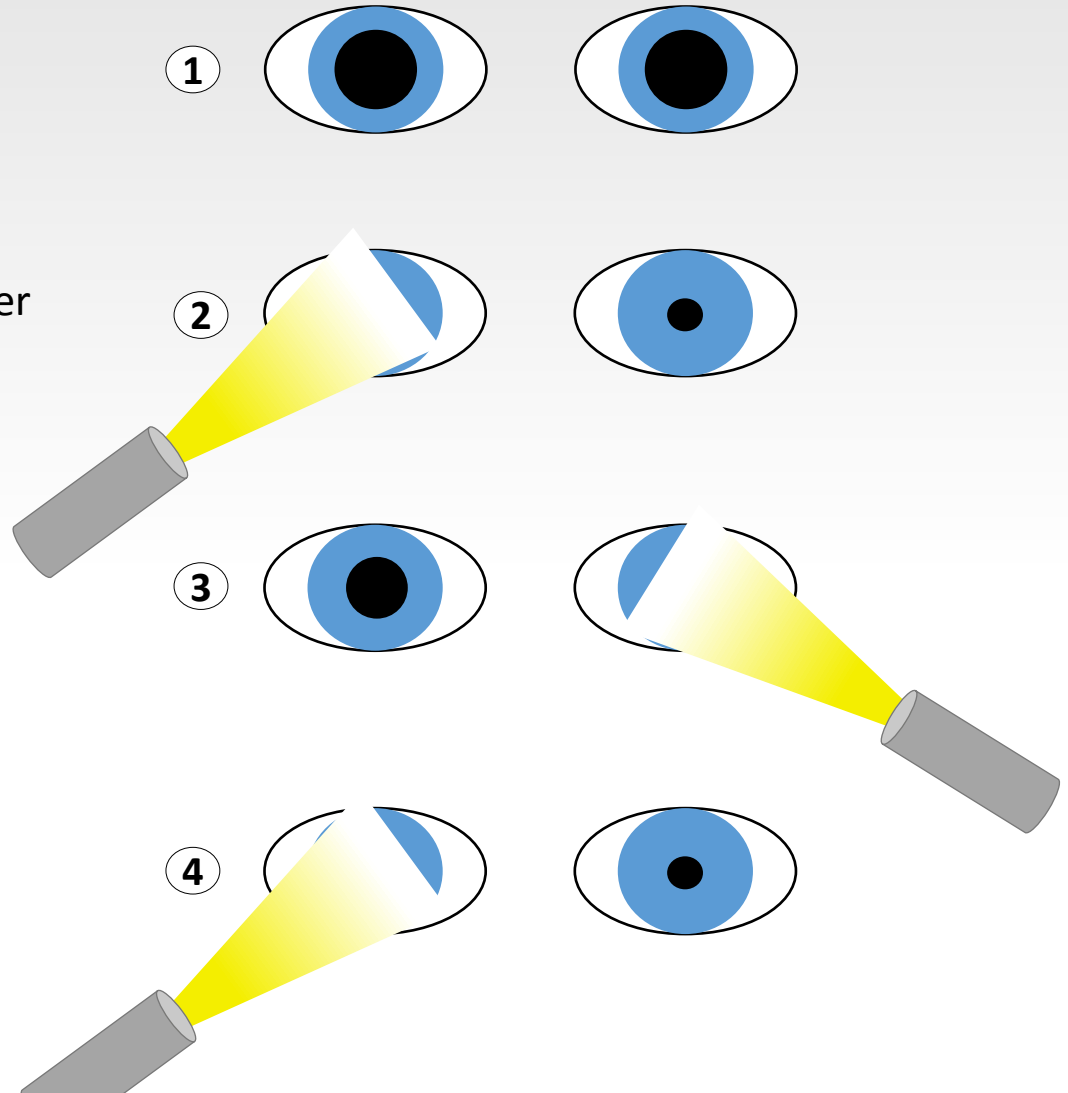
- Diagnosis of anisocoria



# Pupillary reactions
## Diagnostic testing

### Test swinging flashlight

- diagnosticic testing for presence of **relative afferent pupillary defect (RAPD)** – lesion of visual filed is shared with afferent part of pupilomotoric pathway; typical for unilateral lesion of optic nerve
- Principle: change of enlightenment of one pupil, followed by the other pupil with latency (circa 2 seconds) – this is necessary for mydriasis restoration and observation of reactions:
  - 1) both pupils with mydriasis in dark
  - 2) enlightenment of one (presumed normal) pupil only – fast miosis of both pupils
  - 3) enlightenment of fellow (presumed pathological) pupil – slower / abnormal reaction of both pupils (presence of **RAPD**)
  - 4) repeated enlightenment of normal pupil – normal reaction with fast miosis of both pupils



# Reactions of pupil

## Atypical size or reactions of pupil

- **Pupilotonia (Adie´s pupil)**
  - Wider (mydriasis) pupil with small or no reaction to light
  - Worm-like movement of pupillary margin (using slit lamp)
  - Accomodation failure (VA decrease to near distance)
  - Decreased / diminished tendon reflexes (patellar, Achilles)
- **Argyll-Robertson pupil**
  - Narrow pupil with no additional rection to light
  - Preservation of accomodation
  - Typically for patients with neurosyphillis, neuropathies, diabetic polyneuropathies
- **Anisocoria**
  - Inequal size of pupils (usually more than 1mm)

# Anisocoria

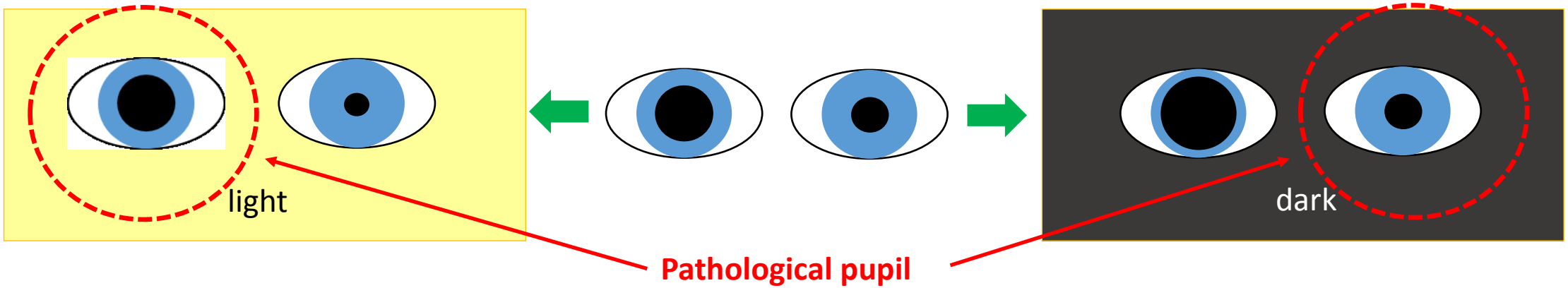
## diagnostic method

1) Exclusion of various possible ophthalmological causes:

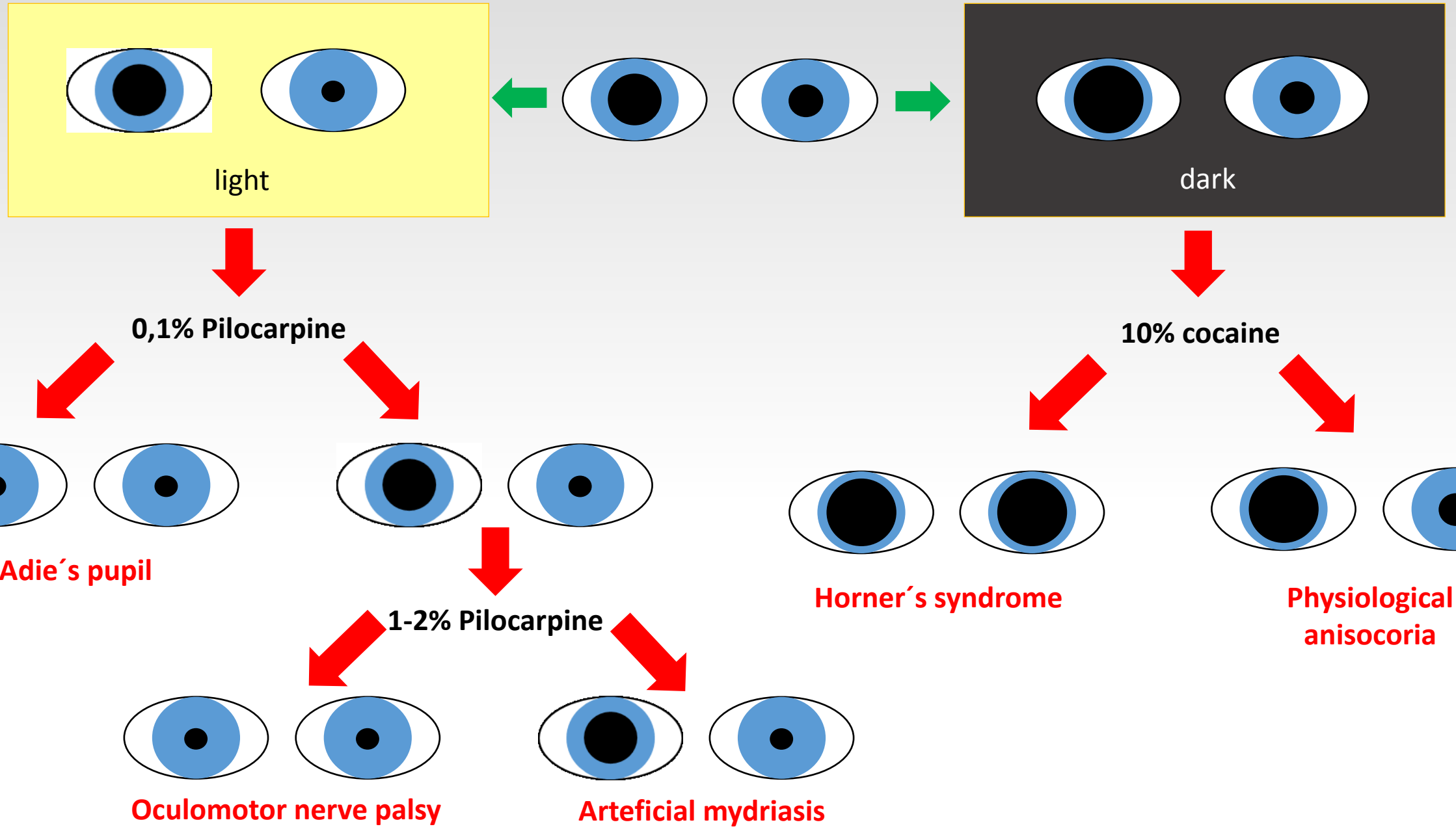
- **Plegia of pupil** after ocular trauma (contusion, perforating trauma)
- **Anomalies of shape** (i.e. congenital or acquired coloboma or anomaly, recurrent iridocyclitis – posterior synechiae)
- **Complications after intraocular surgery** (mostly cataract surgery) or **intraocular inflammation** (endophthalmitis)

2) Assessment of pathological pupil

- Size of pupil in light (wider pupil is pathological)
- Size of pupil in dark (narrowet pupil is pathological)



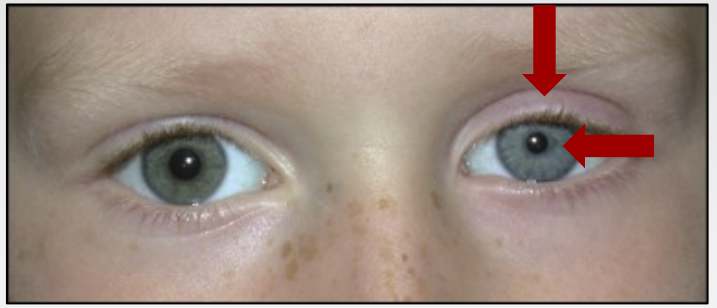
# Anisocoria – diagnostic method



# Horner´s syndrome

## Etiopathogenesis

- Lesion of sympathetic pupilomotorpathway



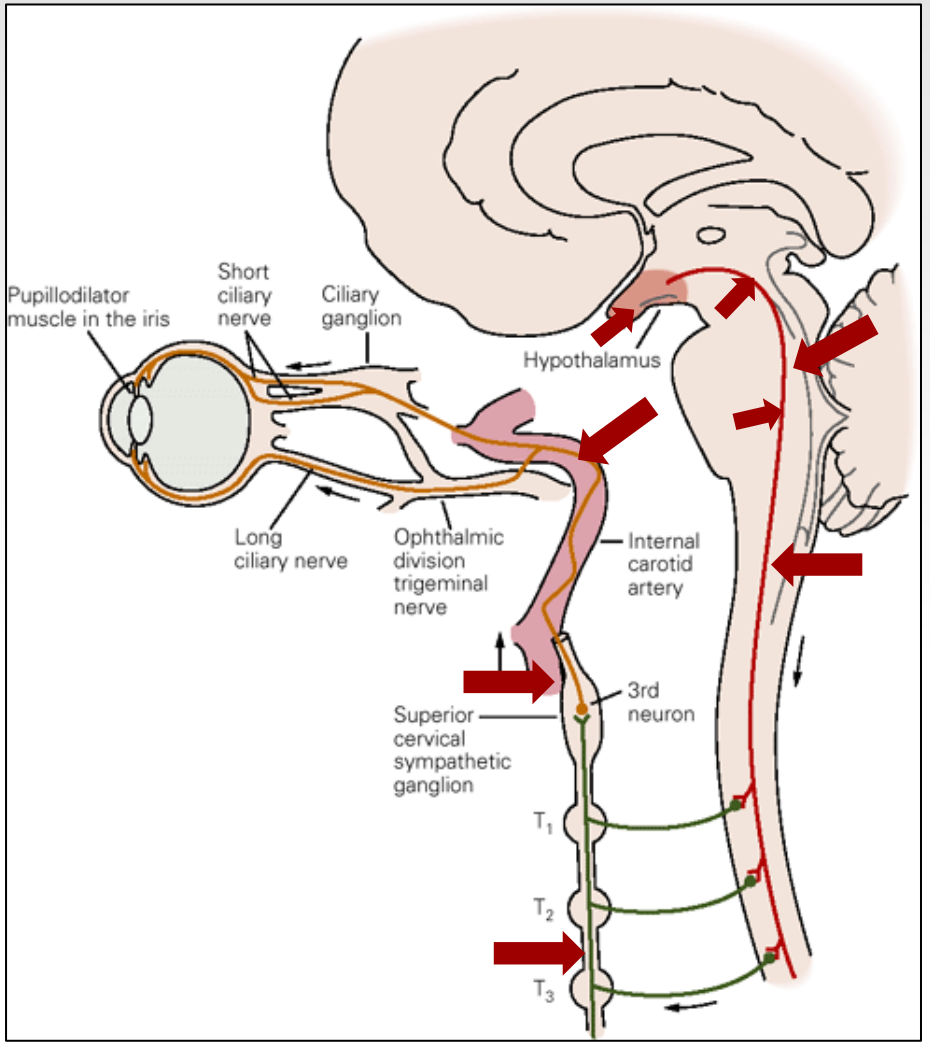
## Signs

- **Miosis of pupil** (no mydriasis in dark) – dilatator muscle palsy
- **Upper eyelid ptosis** (usually mild) and **pseudoenophthalmus** – tarsal muscle palsy
- **anhidrosis** (diminished sweating on affected size / half of face)
- **heterochromia** (congenital form only) – failure of chromatophores creation

# Horner´s syndrome

## Etiology

- 50% idiopathic
- 50% various causes
  - Trauma /surgery in cranial /cervical / upper thoracal area
  - Dissection of internal carotid artery
  - Brainstem ischemia
  - Multiple sclerosis
  - intracranial tumors (various locations)
  - Spinal cord (syringomyelia)
  - Pancoast tumor (lung apex tumor)
  - Thyroid diseases (goiter, carcinoma)



# Thank you for your attention!