APPROACH TO THE PATIENT WITH SELECTED DISEASES OF THE ***CRANIAL NERVES***

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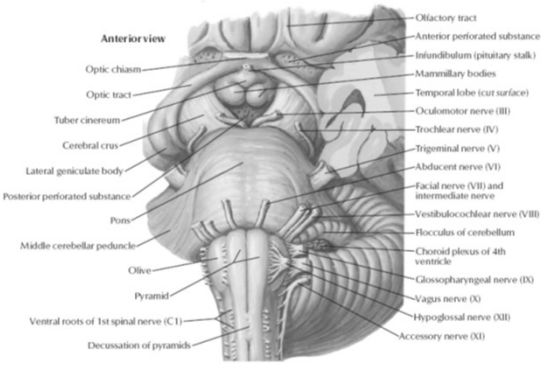
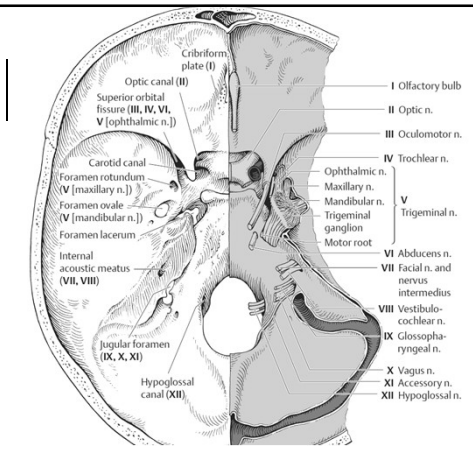
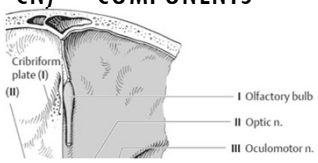




THE OLFACTORY (1ST CN) – COMPONENTS AND LESIONS



Function:
- olfaction or smell

Origin:
- Olfactory epithelium

Peripheral distribution:
- superior nasal concha and nasal septum

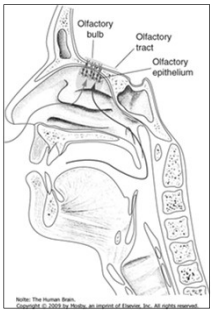
Central connections:
- Olfactory bulb

Signs:
- Loss of sense of smell

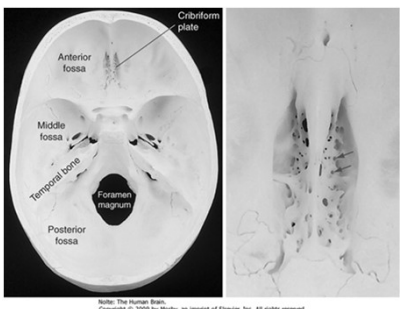
LOCATION OF THE OLFACTORY EPITHELIUM IN THE ROOF AND LATERAL WALL OF THE NASAL CAVITY

The olfactory epithelium continues across the roof of the cavity into a patch of similar size on the nasal septum. Odorants can reach this epithelium either through the nostrils (orthonasally) or by way of the oropharynx (retronasally).

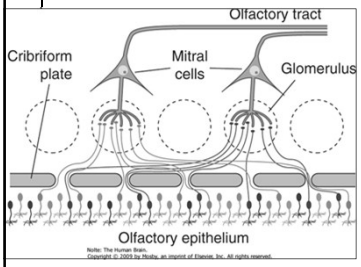
(Modified from Parker CA: A guide to diseases of the nose and throat and their treatment, New York, 1906, Longmans, Green.)



CRIBRIFORM PLATE OF THE ETHMOID BONE OLFACTORY FILA PASS THROUGH SMALL HOLES (ARROWS) IN THE PLATE TO REACH THE OLFACTORY BULB.



SORTING OF OLFACTORY NERVE FIBERS AMONG GLOMERULI OF THE OLFACTORY BULB.



Olfactory receptors of different types-each type characterized by its receptor protein and a restricted range of odor sensitivities (represented here by different colors)-are intermingled in a given area of olfactory epithelium. The axon terminals of any given type all converge on one or two glomeruli (which in reality would contain thousands of axon terminals and the dendrites of up to dozens of mitral and tufted cells).

ANOSMIA AND HYPOSMIA (QUANTITATIVE DISTURBANCES OF OLFACTION)

Craniocerebral injury

Subfrontal meningioma (olfactory groove menigioma) – unilateral anosmia

Upper respiratory tract infections (common cold)

SAH

Zinc and vitamin A deficiency

Inflammatory changes at the scull base

dulling of olfaction in

- Elderly people,
- Early feature of Parkinson's disease,
- Early feature of Alzheimer's disease.

PAROSMIAS - QUALITATIVE DISTURBANCES OF OLFACTION

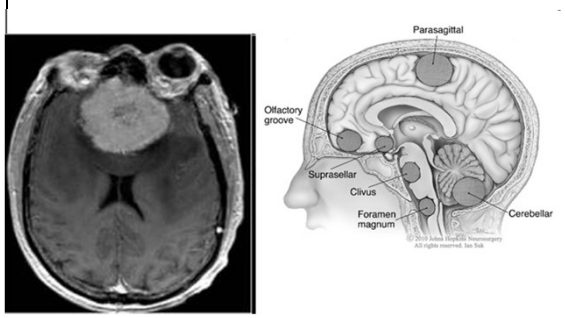
Hyperosmia – abnormally intensive

Cacosmia - spontaneous attack of an abnormal odour

- Olfactory hallucinations are rare form of aura in temporal lobe epilepsy (Also called uncinate gyrus seizure or hippocampal fit)
- typically unpleasant
- Amygdala are the most likely epileptogenic zone of olfactory auras
- Tumours and hippocampal sclerosis are common cause

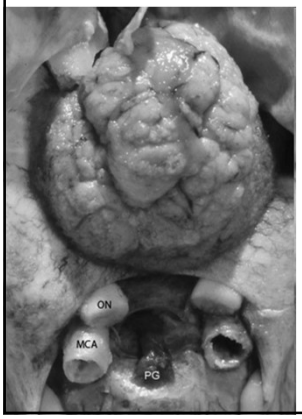
Always due to central dysfunction

OLFACTORY GROOVE MENINGIOMA



Hyposmia due to meningioma

SCULL BASE MENINGIOMA



Post-mortem view of the skull base, showing a large meningioma originating from the olfactory groove. The tumour compressed frontobasal brain structures. Note the smooth, slightly lobulated surface. ON, optic nerve; MCA, middle cerebral artery; PG, pituitary gland.

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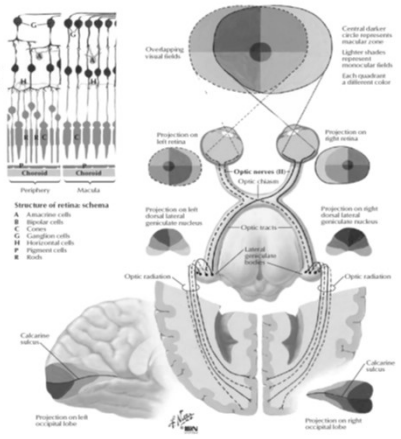
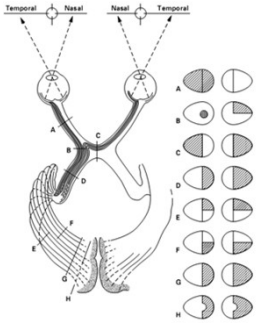
THE OPTIC NERVE (2ND CN)

Monocular disorder

Binocular disorders

2ND CN ANATOI





CASE: VISION PROBLEM

Transient vison loss in one eye yesterday

It lasted about 10 minutes

Past medical history: hypertension, diabetes, CHD

Exam: normal vision and normal rest of the neurological exam

Neck exam?

MONOCULAR LOSS OF VISION

- Transient monocular blindness (*Amaurosis fugax*) - an attack of transient painless loss of vision
- Optic neuritis
- Anterior Ischemic Optic Neuropathy (AION)
- Giant Cell (temporal) Arteritis
- Optic nerve compression (tumours)
- Retinal migraine (rare)

OPTIC NEURITIS

commonest presenting feature of MS

unilateral or bilateral

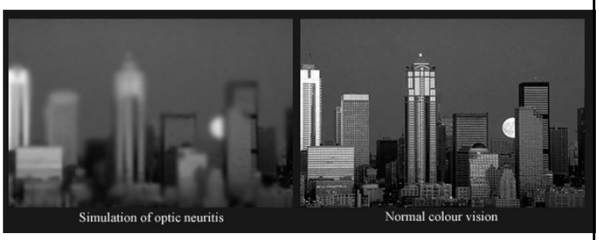
pain is common, followed by rapid visual failure

blurred vision and a distortion or lack of color vision

symptoms across the entire visual field

recovery in majority of patients

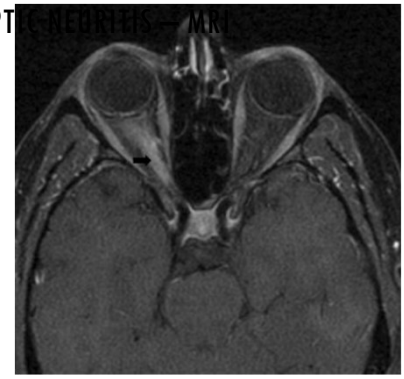
BLURRED VISION AND A DISTORTION OR LACK OF COLOR VISION IN ON



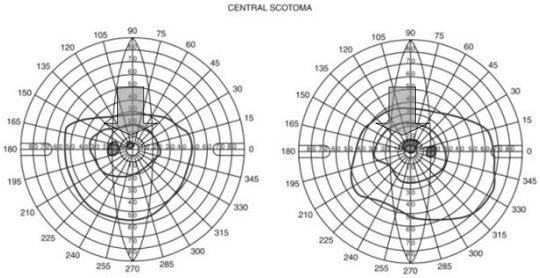
http://www.psych.ucalgary.ca/pace/va-lab/Brian/default.htm

BLURRED VISION AND A DISTORTION OR LACK OF COLOR VISION IN ON

OPTIC NEURITIS – MRI



CENTRAL SCOTOMA AFTER RETROBULBAR NEURITIS



ANTERIOR ISCHEMIC OPTIC NEUROPATHY (AION)

- in older patients (>40 yrs)
- Sudden (seconds/minutes) loss of vision in one eye with plateau, may improve after several weeks
- atheromatous occlusion of the short posterior cilliary arteries or in GCA
- altitudinal or arcuate field defect
- disc swelling then followed by optic atrophy

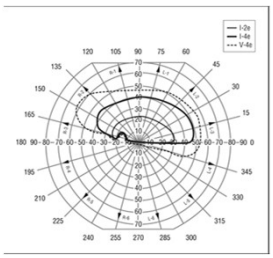


Figure Legend:

Visual field defects in nonarteritic anterior ischemic optic neuropathy, plotted with a Goldmann perimeter (using I-2e, I-4e, and V-4e targets), show absolute inferior altitudinal defect with I-2e, I-4e, and V-4e isopters. The visual acuity in the eye was 20/20.

OPTIC NERVE COMPRESSION

- insidious loss of vision
- marked impairment of colour vision
- afferent pupillary defect
- a central scotoma, later extending out to the periphery
- optic atrophy
- Due to tumours

ORBITAL TUMORS

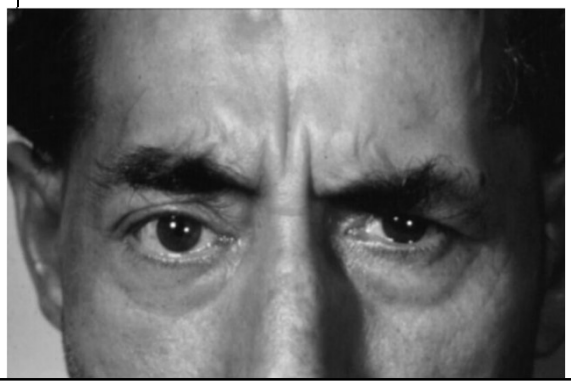
Meningiomas

- menigiomas of the optic nerve sheath - gradual visual failure with mild proptosis
- meningiomas within the optic canal (extended from the orbit or from the region of the anterior clinoid process)
- easily missed but easy to identify by neuro-imaging

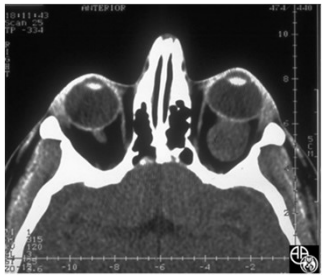
Optic nerve gliomas

- Benign in childhood, slow growing (pathology: the pilocytic astrocytoma), unilateral or bilateral enlargement of optic canals
- In adulthood – rare, but malignant with rapid visual loss with ocular pain and early involvement of the other eye

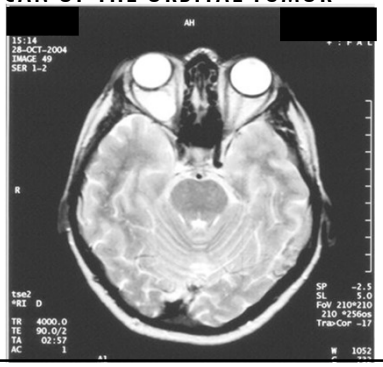
ORBITAL TUMOR

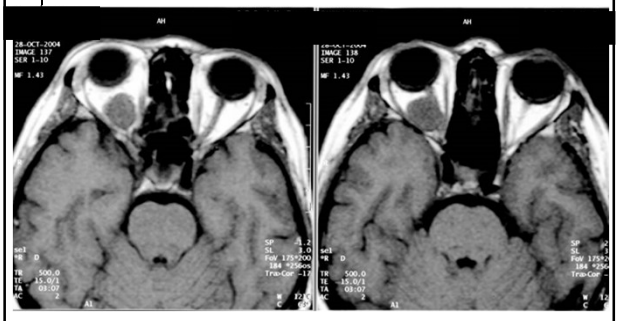


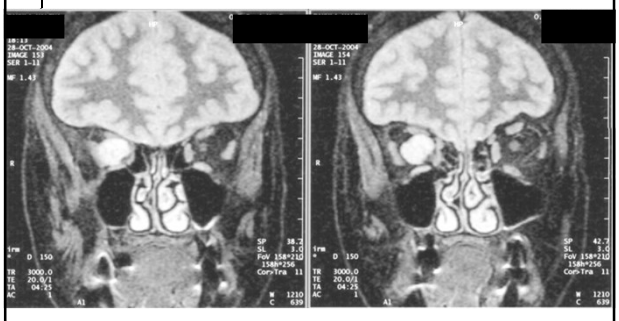
CT SCAN OF THE ORBITAL TUMOR

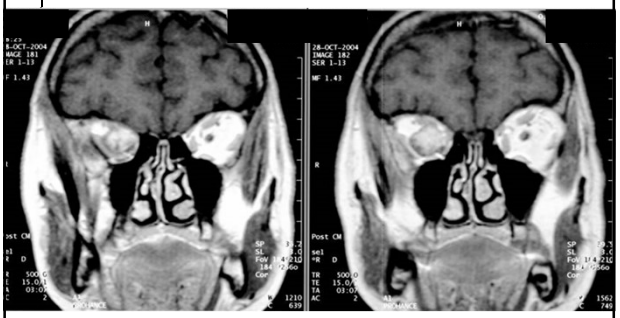


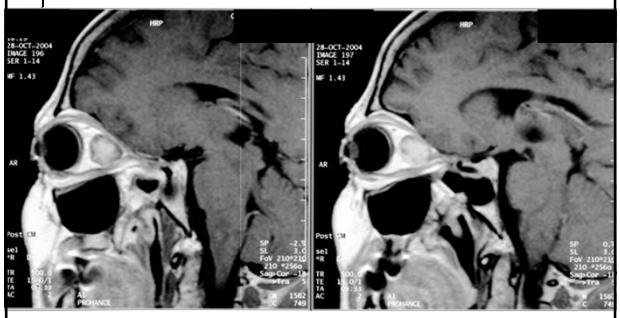
MR SCAN OF THE ORBITAL TUMOR



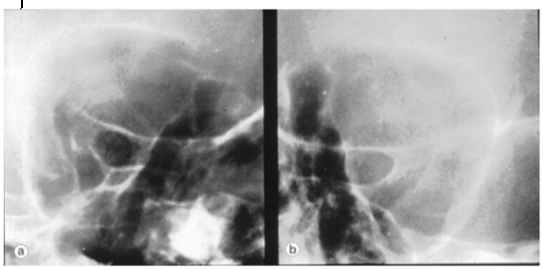






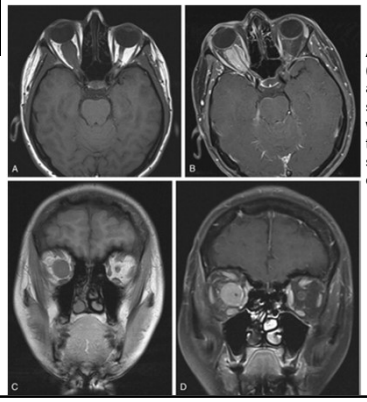


AN ORBIT X-RAY IN OPTIC NERVE GLIOMA



bilateral enlargement of optic canals

OPTIC NERVE SHEATH MENINGIOMA – MRI



Axial **(A, B)** and coronal **(C, D)** precontrast **(A, C)** and postcontrast with fat suppression **(B, D)** T1-weighted images showing the optic nerve surrounded by an enhancing meningioma.

OTHER DISEASES CAUSING VISION LOSS (AMBLYOPIA)

- Toxic amblyopia
  - **drugs** such as chloramphenicol, isoniazid, ethambutol and digoxin
  - **toxins** such as lead, ethylene glycol (antifreeze), or methanol (wood alcohol or methyl alcohol).
- Deficiency diseases
  - vitamins B1 and B12 or folate
- Sarcoidosis
  - optic nerve granuloma at optic nerve head or in its retrolaminar portion

BILATERAL DISORDERS

Papilledema

Chiasmal lesions

Retrochiasmal lesions

PAPILLEDEMA

Swelling of the optic disk due to increased intracranial pressure (more precisely called edema of the optic nerve head).

Gradual loss

Often asymptomatic, or complaints of transient visual obscurations triggered by eye movements.

With prolonged papilloedema the optic nerve sheath expands and nerve fibre atrophy appears leading to various visual field changes (arcuate defects, peripheral constriction)

PAPILLEDEMA CAUSES

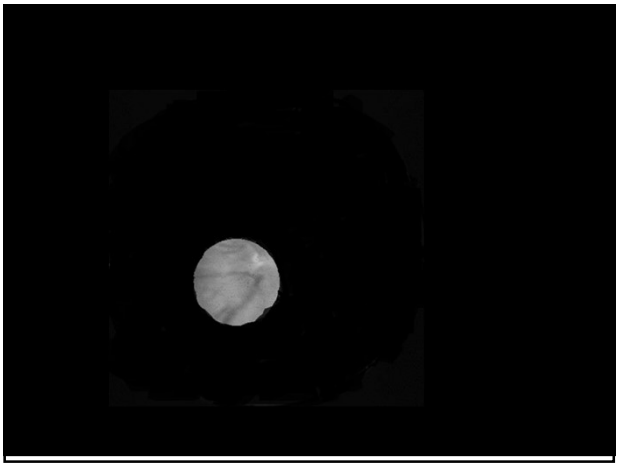
Increased intracranial pressure (brain tumours)

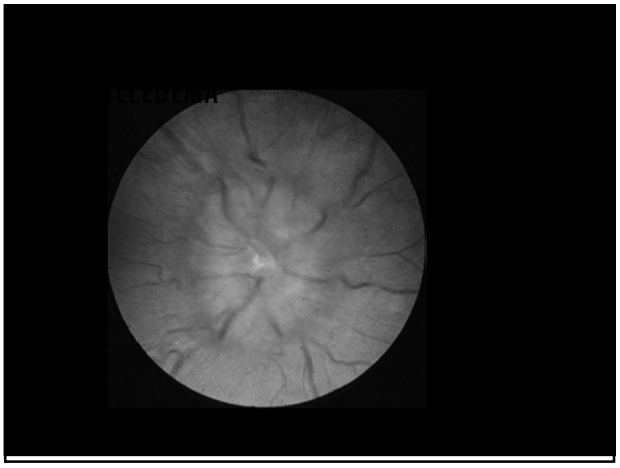
hypertensive retinopathy

central vein thrombosis

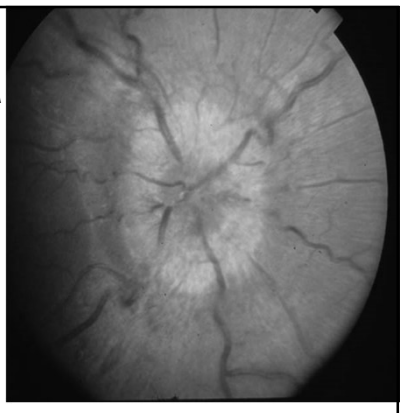
pappilitis – MS

retrobulbar retinitis – MS



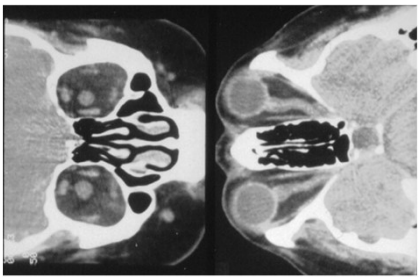


PAPILLEDEMA



PAPILLOEDEMA

PROLONGED PAPILLOEDEMA



CHIASMATIC LESIONS

Chiasmatic compression secondary to

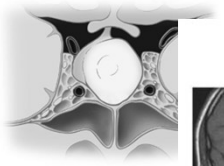
- pituitary tumour,
- craniopharyngioma,
- menigioma,
- aneurysm.

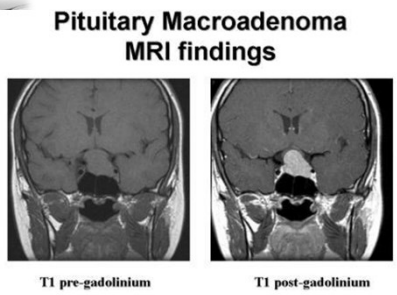
Other causes: trauma, demyelinating disease

Bitemporal hemianopia, typically asymmetrical;

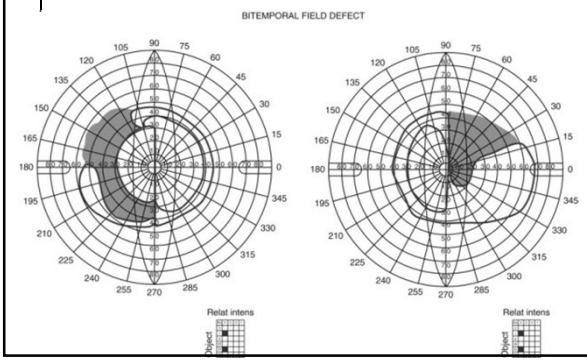
compression from below produces superior bitemporal hemianopia.

PITUITARY TUMOUR





BITEMPORAL VISUAL FIELD LOSS

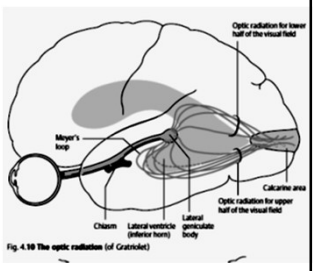


RETROCHIASMATIC LESIONS

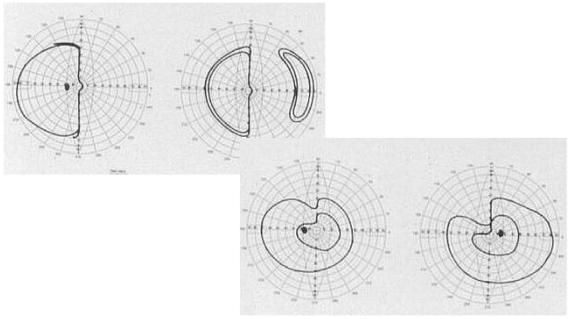
Homonymous hemianopia

Involvement of the optic radiation

- Within the temporal lobe produces a homonymous defect that predominates in the superior quadrants.
- Within parietal lobe produces a defect that is usually complete.
- Within occipital lobe – defects that can be quadrantic, scotomatous or complete.

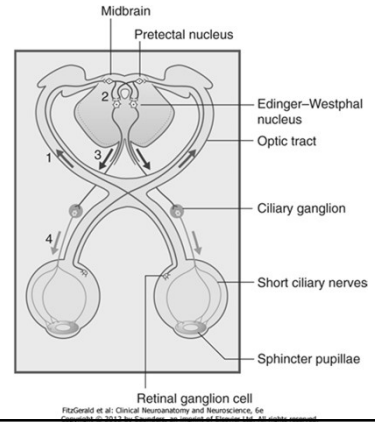


TYPES OF HOMONYMOUS FIELD DEFECT



APPROACH TO THE PATIENT WITH ABNORMAL PUPILS

PUPILLARY LIGHT REFLEX



EXAMINATION OF THE PUPILS

Pupil size

- Anisocoria greater in dark => smaller pupil abnormal
- Anisocoria greater in bright light => larger pupil abnormal
- Asymmetry of pupils diameter < 0.4 mm ➔ normal

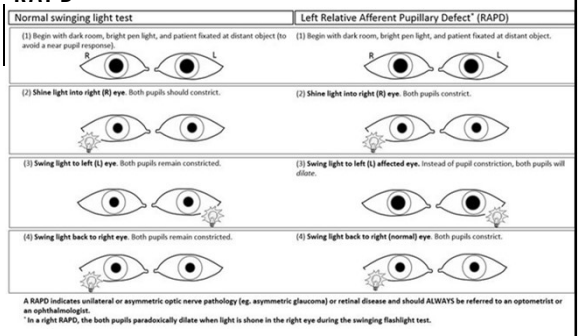
Pupillary response to light

Alternating light test for relative afferent pupillary defect or RAPD AKA Marcus Gunn pupil

PUPILLARY ABNORMALITIES

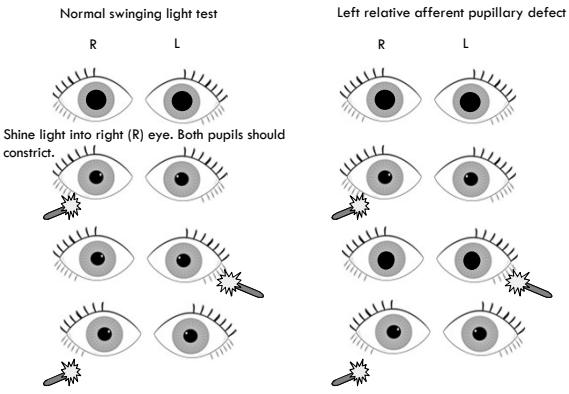
- RAPD
- Mydriasis
  - 3rd CN palsy
  - Tonic pupil syndrome (pupillotonia)
- Miosis
  - Horner' syndrome
- Light near dissociation
  - Argylle-Robertson pupil
    - responsible lesion in the upper midbrain rostral to the oculomotor nucleus and interrupts the light reflex pathway

RAPD



Alfred Basilious and Yvonne M. Buys: A woman with a family history of glaucoma *CMAJ cmaj.140685;*

RAPD



RAPD

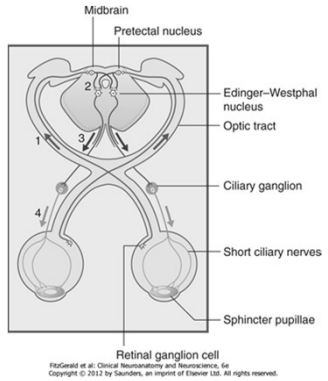
Ocular & retinal lesions

Optic nerve lesions

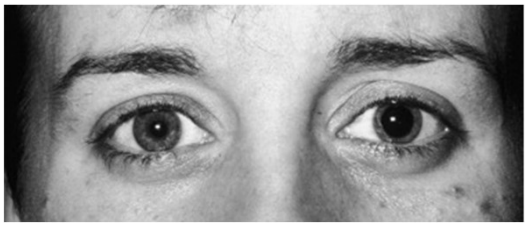
Optic chiasm

Optic tract lesions

Pretectal nucleus

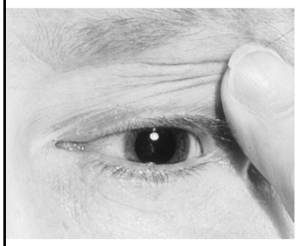


NON-REACTIVE PUPIL



- Local disease of iris (trauma, iritis, glaucoma)
- Oculomotor nerve compression
- Optic nerve disorders
- An isolated iridoplegia with fixed dilated pupil unresponsive to light and near effort can result from an organic insult to the cilliary ganglion (cilliary ganglionitis) without ptosis

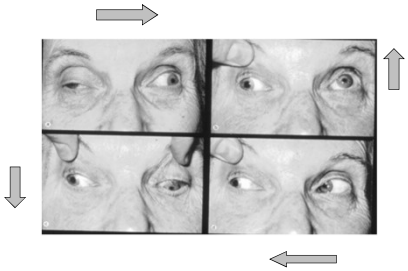
PREGANGLIONIC LESION - 3RD CN PARESIS



Clinical features

- Large poorly reactive pupil (direct & indirect), & near effort
- Ipsilateral ptosis
- Ocular motility disturbances
- Responsiveness to full– strength pilocarpine

3RD CN PARESIS



ETIOLOGY OF THE 3RD CN PALSY

Brainstem/fascicular

- Ischemia
- Hemorrhage
- Tumor, arteriovenous malformation

Interpeduncular fossa, subarachnoid space

- Interneural Ischemia – **vasculopathic: diabetes** - may be presenting sign of DM; hypertension; GCA
- posterior communicating artery aneurysm. The pupil is inevitably affected if the paresis is complete.

Sphenocavernous syndrome (IV CN, V1±V2 CN)

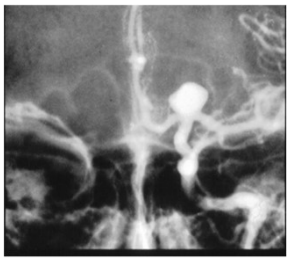
- Cavernous sinus thrombosis
- Fistula
- Tumor
- Tolosa-Hunt syndrome

POSTERIOR COMMUNICATING ARTERY (PCOA) ANEURYSM CAN CAUSE 3RD CN PALSY

9% to 36% of oculomotor nerve palsies are caused by an intracranial aneurysm.

PCA aneurysms present with III nerve palsy 30% to 60% of the times

Aneurysmal III nerve palsy typically presents with pain, *mid-dilated pupil* with poor or absent light reaction, and *complete or partial external paresis* including ptosis with supra, infra, and adduction deficits



PCA ANEURYSM ARE RARE

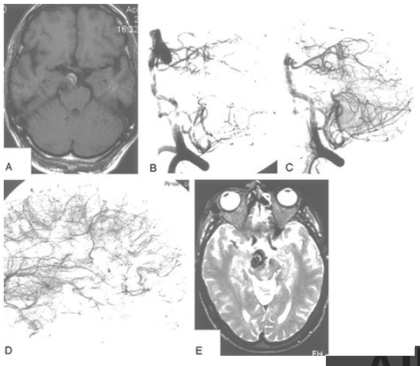
Among 1880 aneurysms treated between January 1995 and January 2005, 22 aneurysms (1.2%) in 22 patients were located on the PCA.

10 patients presented with subarachnoid hemorrhage (SAH) from the PCA aneurysm: 2 of these patients had additional visual field deficits and 2 had additional oculomotor palsy.

One patient presented with acute oculomotor palsy only.

van Rooij W et al. AJNR Am J Neuroradiol 2006;27:300-305

64-year-old man presenting with HH grade I SAH and right occulomotor palsy.
A and B, MR imaging and angiography show dissecting fusiform P2 aneurysm with intramural thrombus.
C and D, Vertebral (C) and right internal carotid (D) angiogram after the aneurysm occlusion



van Rooij W et al. AJNR Am J Neuroradiol 2006;27:300-305

TONIC PUPIL (PUPILOTONIA, ADIE'S)

Unilateral pupillary dilatation.

Sluggish reaction to light and accommodation

In the dark the affected pupil becomes the smaller one due to failure of reflex dilatation.

Predominates in women, between 20-50 yrs.

After an acute irydoplegia (ciliary ganglionitis), orbital trauma or viral infection (Herpes Zoster)

If associated with diminished deep tendon reflexes is called Holmes-Adie syndrome

OCULOSYMPATHETIC DEFECT -HORNER'S SYNDROME

Myosis – not marked at room light (~1.0 mm), apparent in dark

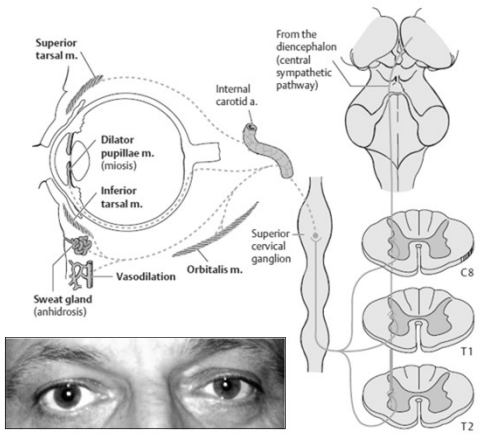
Pupillary dilation lag – slowly (20-30 s.) enlarges in dark

Ptosis- denervation of tarsal muscles, mild, absent in 12%

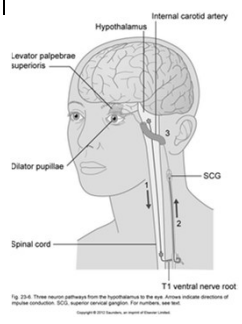
Enophtalmos - denervation of tarsal muscles

Anhidrosis – variably present

Heterochormia iridis – in congenital Horners



HORNERS SYNDROME CAUSES



- Stroke (in Wallenberg syndrome)
- Disc prolapse (C8-T2)
- Brachial plexus lesion – Pancoast tumor
- Surgical procedures
- Internal carotid dissection
- Cluster headache

HORNER'S SYNDROME.

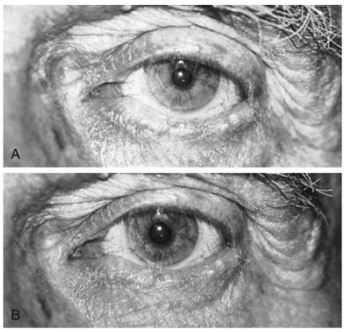


Fig. 15-14 Horner's syndrome. The normal left pupil before **(A)** and after **(B)** instillation of 4% cocaine.

HORNER'S SYNDROME

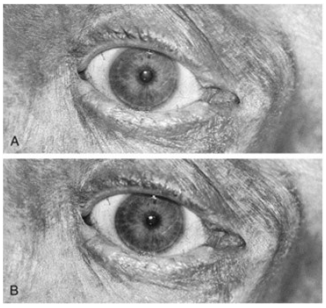
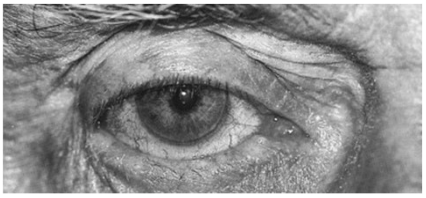


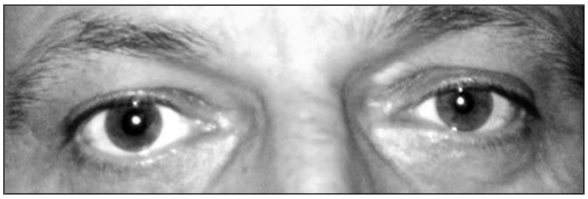
Fig. 15-15 Horner's syndrome. The affected right pupil before **(A)** and after **(B)** instillation of 4% cocaine; there is no response. The upper lid has been retracted

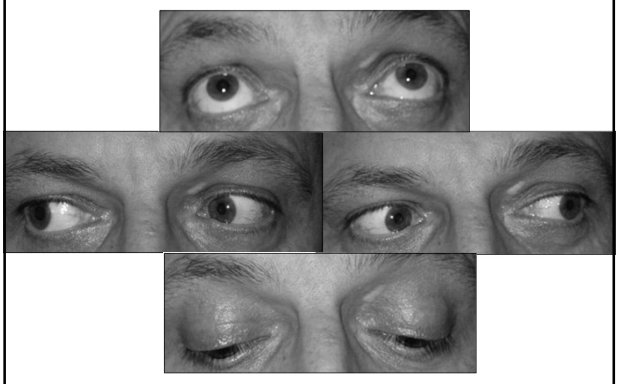
HORNER'S SYNDROME



Horner's syndrome. Dilatation of the pupil after instillation of 1% hydroxyamphetamine.

HORNER SYNDROME





LIGHT – NEAR DISSOCIATION ARGYLL-ROBERTSON PUPIL (ARP)

The pupil is

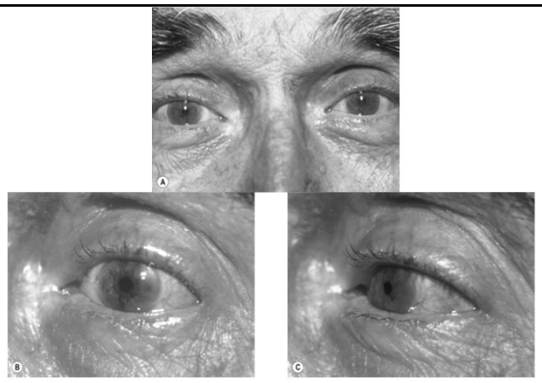
- constricted (miosis)
- irregular
- reactive to convergence but not to light (although not always) → light-near dissociation
- bilateral phenomenon (almost always)

in tertiary syphilis

dorsal midbrain lesion that interrupts the pupillary light reflex pathway but spares the more ventral pupillary near reflex pathway.

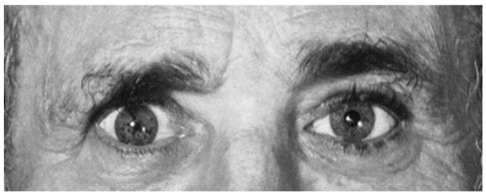
ARGYLL-ROBERTSON PUPIL ETIOLOGY

- neurosyphilis, mainly tabes dorsalis,
- multiple sclerosis,
- viral encephalitis,
- Lyme disease,
- sarcoidosis,
- alcoholic encephalopathy,
- midbrain hemorrhages and tumors involving the area of the colliculi



Argyll Robertson pupils in tabes dorsalis (absent deep tendon reflexes, loss of vibratory sense and proprioception in the lower extremities, and Charcot joints). The pupils are small (**A**), poorly reactive to light (**B**), but constrict to near stimuli (**C**). (The patient was seen courtesy of Dr. J. Lawton Smith.) (From Liu GT. Disorders of the eyes and eyelids: Disorders of the pupil. In Samuels MA, Feske S (eds): The Office Practice of Neurology, p 66. New York, Churchill Livingstone, 1996, with permission.)

ARGYLL-ROBERTSON PUPIL



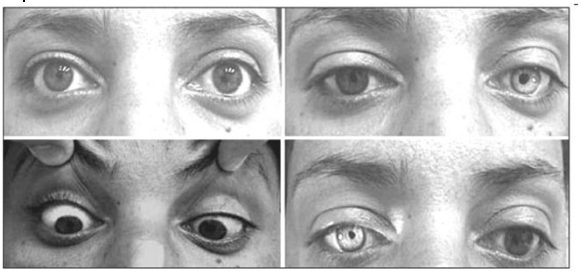


Figure. Argyll Robertson pupil.

BRAINSTEM ORIGINS OF THE OCULAR M

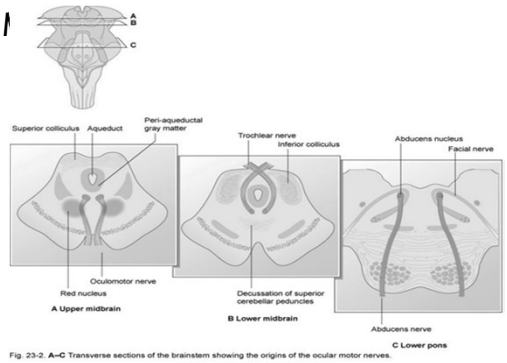


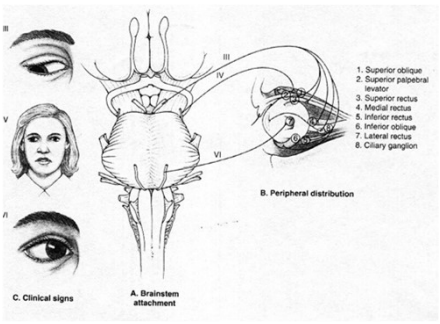
Fig. 23-2. **A–C** Transverse sections of the brainstem showing the origins of the ocular motor nerves.

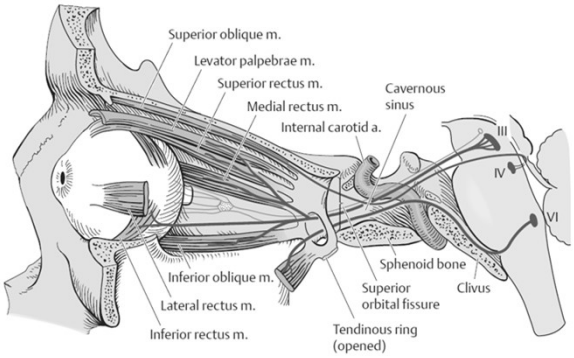
THE ANATOMY OF OCULAR MOTOR NERVES

III

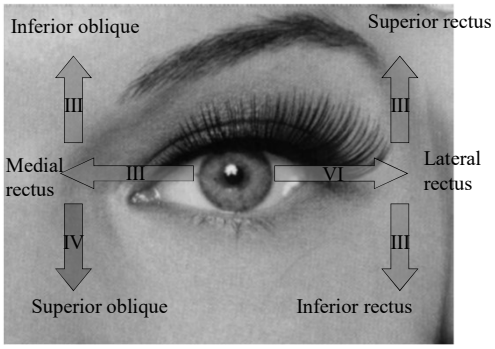
IV

VI





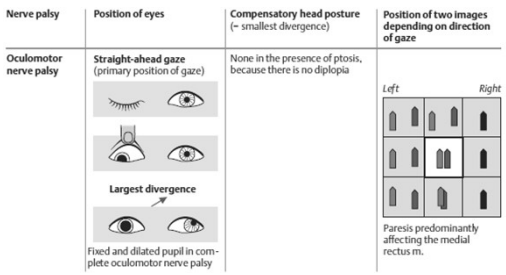
THE OCULAR MOTOR NERVES – EYE MOVEMENTS



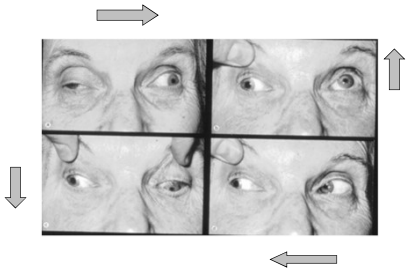
COMPONENTS AND LESIONS OF THE 3 CN

Function	Origin	Peripheral distribution	Signs
Eye movements	oculomotor nucleus	Medial, superior, inferior recti; inferior oblique muscles	Opthalmoplegia with eye turned down and out
Elevation of the eyelid	oculomotor nucleus	superior palberal levator	ptosis
Pupillary constriction and accommodation	Edinger-Westphal nucleus	cilliary ganglion; postganglionic fibers to sphincter of the pupil and cilliary muscle	Mydriasis; loss of accomodation

3RD CN PARESIS



3RD CN PARESIS



ETIOLOGY OF THE 3RD CN PALSY

Brainstem (nuclear & fascicular)

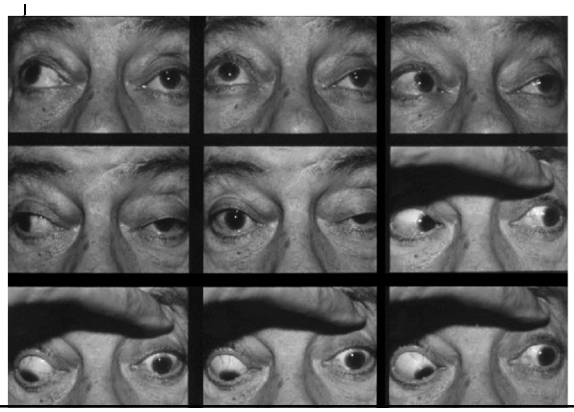
- Ischemia
- Hemorrhage
- Tumor, arteriovenous malformation

Interpeduncular fossa, subarachnoid space

- Interneural Ischemia – vasculopathic: diabetes - can be a presenting sign of DM; hypertension; GCA
- posterior communicating artery aneurysm. The pupil is inevitably affected if the paresis is complete.

Sphenocavernous syndrome (IV CN, V1±V2 CN)

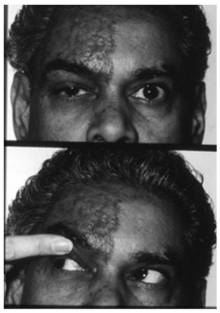
- Cavernous sinus thrombosis
- Fistula
- Tumor
- Tolosa-Hunt syndrome



HERPES OPTHALMICUS

Presents with different ocular motor palsies and often a partial or complete oculomotor palsy.

Aberrant regeneration of the 3rd n. can lead to different anomalous co-movements (synkinesia) – most often there is lid elevation on attempted down gaze.



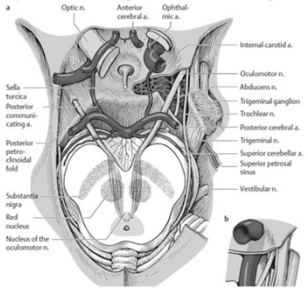
IV CN (TROCHLEAR)

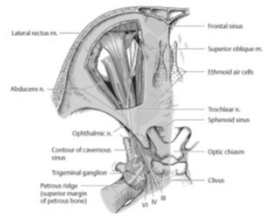
Lesions are uncommon

Causes

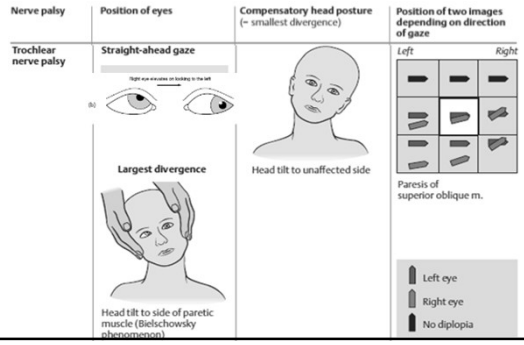
- usually due to trauma, often minor
- Vascular or DM
- often idiopathic

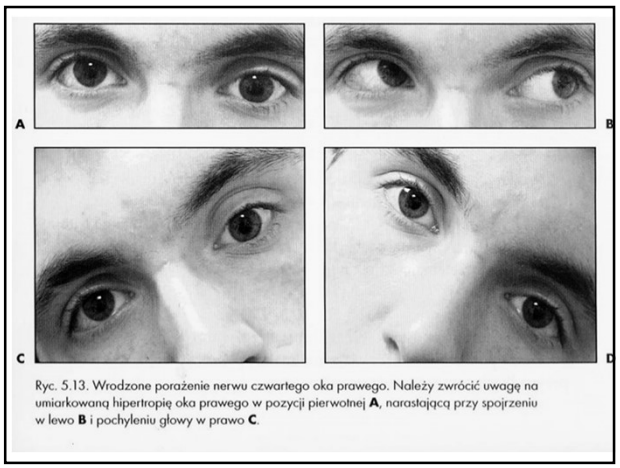
4TH CN (TROCHLEAR)



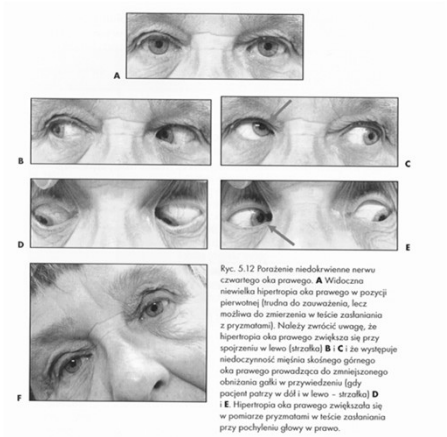


4TH CN PARESIS



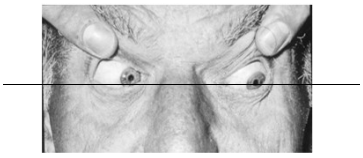


Ryc. 5.13. Wrodzone porażenie nerwu czwartego oka prawego. Należy zwrócić uwagę na umiarkowaną hipertropię oka prawego w pozycji pierwotnej **A**, narastającą przy spojrzeniu w lewo **B** i pochyleniu głowy w prawo **C**.



Ryc. 5.12 Porażenie niedokrwienne nerwu czwartego oka prawego. **A** Widoczna niewielka hipertropia oka prawego w pozycji pierwotnej (trudna do zauważenia, lecz możliwa do zmierzenia w teście zasłaniania z pryzmatami). Należy zwrócić uwagę, że hipertropia oka prawego zwiększa się przy spojrzeniu w lewo (strzałka) **B** i **C** i że występuje niedoczynność mięśnia skośnego górnego oka prawego prowadząca do zmniejszonego obniżania gałki w przywiedzeniu (gdy pacjent patrzy w dół i w lewo – strzałka) **D** i **E**. Hipertropia oka prawego zwiększała się w pomiarze pryzmatami w teście zasłaniania przy pochyleniu głowy w prawo.

IV CN PALSY



Lesion: diplopia, extortion of the eye; weakness in depression of adducted eye

CAUSES OF THE 6TH CN LESIONS:

- Idiopathic
- ischemic process in patients with hypertension or diabetes
- Giant cell arteritis (rare)
- Tumours (carcinoma of the meninges, nasopharyngeal carcinoma, chordoma)
- Increased intracranial pressure
- lesions within the cavernous sinus (idiopathic inflammation of cavenous sinus – Tolosa-Hunt s.)
- infection of the petrous part of temporal bone

BILATERAL VI CN PALSIES

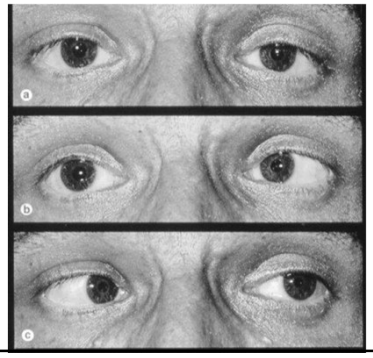
raised intracranial pressure

Guillain-Barré syndrome

carcinomatous meningitis

Sarcoidosis

BILATERAL 6TH CN PALSY



FOVILLE' SYNDROME

Nuclear lesion of VI CN

- pontine lesion (gliomas) that affects also 5th and 6th CN and the corticospinal tract producing lateral rectus palsy, facial paresis and diminished sensation with contrlateral hemiparesis

ORBITAL LESIONS CAUSING OCULAR MOTOR NERVES PALSIES

Tumours

Superior orbital fissure syndrome

Cavernous sinus syndrome

Tolosa-Hunt syndrome

ORBITAL TUMORS

Small tumours in the region of the optic nerve are likely to have produced substantial visual loss with only small proptosis.

Malignant melanoma is the most common primary intraocular neoplasm in adults.

Metastases – Ca of the lung or breast or leukemic or lymphomatous deposit.

Orbital angiomas (haemangiomas) – produce proptosis, diplopia with little visual impairment.

Orbital pseudo-tumour - combination of pain, proptosis and ophtalmoplegia – can be bilateral.

CAVERNOUS SINUS & SUPERIOR ORBITAL FISSURE SYNDROME (SPHENOCAVERNOUS SYNDROME)

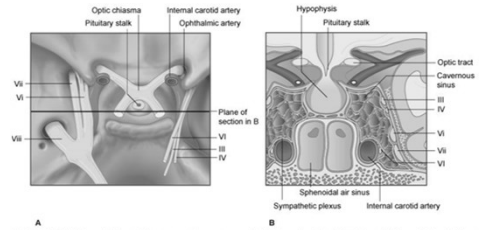


Fig. 23-3. **(A)** Middle cranial fossa with cavernous sinuses removed. **(B)** Coronal section in the plane of the hypophysis with the cavernous sinuses in place. III, Oculomotor nerve; IV, trochlear nerve; VI, abducens nerve; Vi, Vii, Viii, ophthalmic, maxillary, mandibular divisions of trigeminal nerve.

SUPERIOR ORBITAL FISSURE SYNDROME

3rd, 4th and 6th palsy and sensory loss in first div. of 5th nerve

due to meningioma, nasopharyngeal carcinoma, aneurysm and pituitary tumour.

associated with a mild degree of proptosis.

CAVERNOUS SINUS SYNDROME

3rd, 4th and 6th CN palsy and sensory loss in first div. of 5th CN; in addition the 2nd div. of the 5th CN nerve affected.

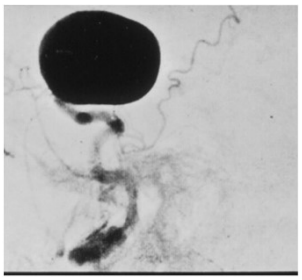
Giant internal carotid aneurysm within the cavernous sinus presents with a slowly progressive painful ophtalmoplegia

Caroticocavernous fistulas

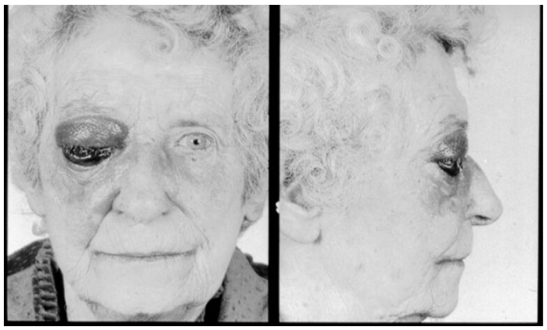
- abnormal communications between the carotid artery and the cavernous sinus
- usually traumatic or consequent to rupture of a cavernous aneurysm. Treated by a balloon introduced to the lumen of the fistula and then injection of the silicon into the balloon.

CAVERNOUS SINUS THROMBOSIS

GIANT INTERNAL CAROTID ARTERY ANEURYSM



CAROTICO-CAVERNOUS FISTULA



INTERNUCLEAR OPHTHALMOPLEGIA (INO)

Due to disruption of the medial longitudinal fasciculus (MLF).

A left INO is one in which there is an impairment of adduction of the left eye.

there is inadequate or slowed adduction of the ipsilateral eye with dissociated nystagmus of the abducted eye (typically)

May be partial or complete. INO is present whether the eye movement is volitional or reflexive.

Unilateral INO is caused by any brain stem lesion. Bilateral INO in younger subjects is usually due to MS.

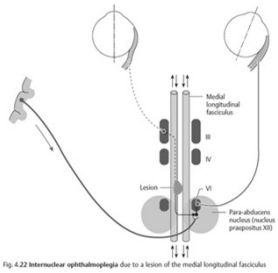


Fig. 4.22 **Internuclear ophthalmoplegia** due to a lesion of the medial longitudinal fasciculus

RIGHT INO (INTERNUCLEAR OPHTALMOPLEGIA)

INO

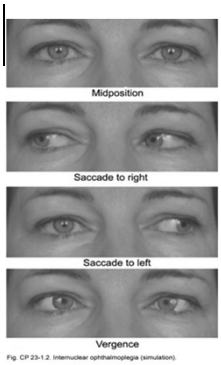


Fig. CP 23-1.2. Internuclear ophthalmoplegia (simulation).

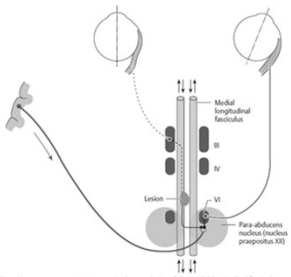
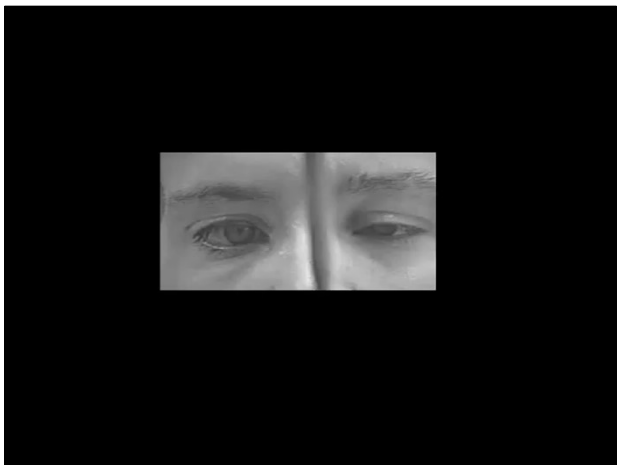


Fig. 4.22 **Internuclear ophthalmoplegia** due to a lesion of the medial longitudinal fasciculus



TOLOSA-HUNT SYNDROME

Painful ophtalmoplegia

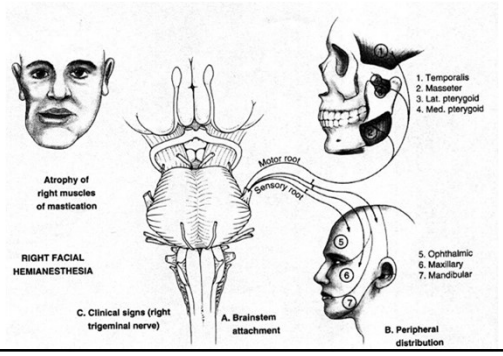
Inflammatory disorder of unknown etiology,

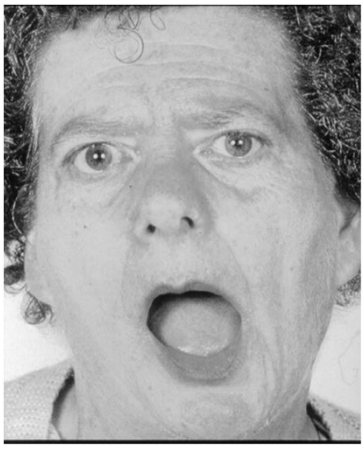
Involves the region of the anterior cavernous sinus or the superior orbital fissure.

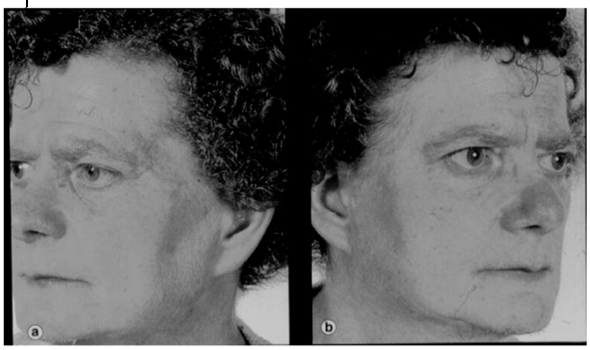
Acute onset of severe steady retroorbital or periorbital pain

Disruption of the function of the 3rd, 4th and 6th nerves alone or in combination, as well as with impairment of the sensation in the 1st div. of the trigeminal nerve.

THE TRIGEMINAL NERVE (5TH CN)







LESIONS OF THE TRIGEMINAL NERVE

The V CN can be affected in the brainstem or along its peripheral course.

Lesions affecting the trigeminal nerve or the Gasserian ganglion include

- aneurysm,
- menigioma,
- nasopharyngeal carcinoma,
- infections of the petrous temporal bone,
- any cerebellopontine angle tumour.

TRIGEMINAL NEUROPATHY

Rare condition

Progressive loss of facial sensation occurs without involvement of motor fibers.

Etiology is unknown

TRIGEMINAL NEURALGIA

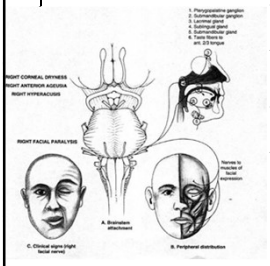
No loss of sensation, but there is pain in the region innervated by the 5th CN.

- Some patients report decreased sensation oh affected side when clinically teste

Trigeminal neuralgia may be a presenting sign of MS

Minute sensation loss can be estimated by checking the corneal reflexes.

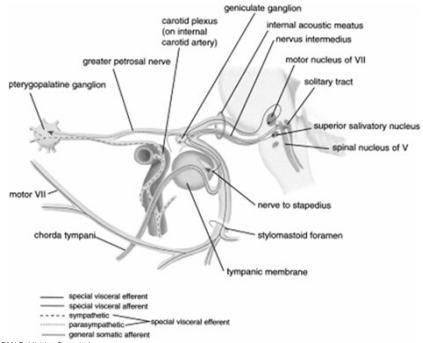
THE FACIAL NERVE (7TH CN)



This is the nerve that lets you cry
And wets your mouth when it is dry
Dampens noise when your are young
Tastes on two thirds of your tongue
And lastly – now, just let me think
Lets you give a smile – and wink!

Meredith Rose Golomb, MD

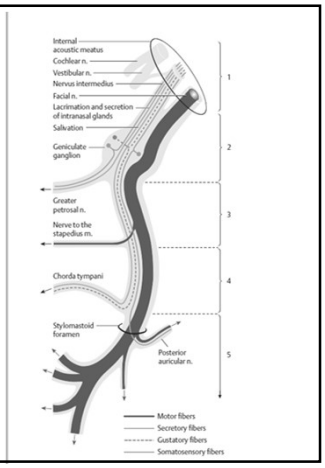
FUNCTIONAL ANATOMY OF THE FACIAL NERVE.

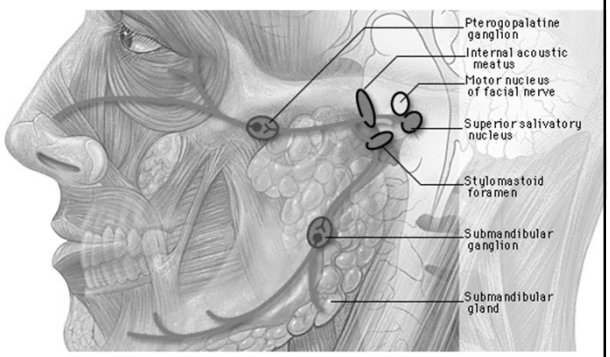


 Sweeney C J , and Gilden D H J Neurol Neurosurg Psychiatry 2001;71:149-154

JNNP

FUNCTIONAL ANATOMY OF THE FACIAL NERVE





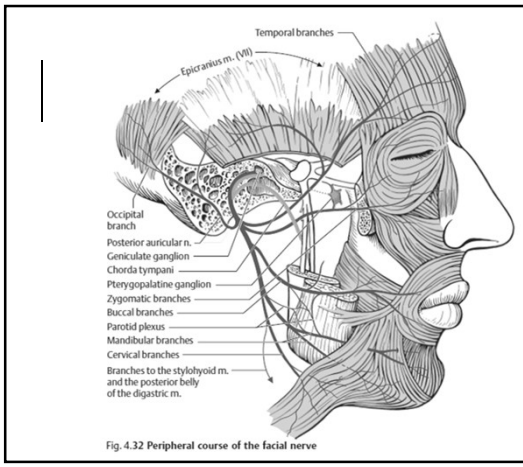
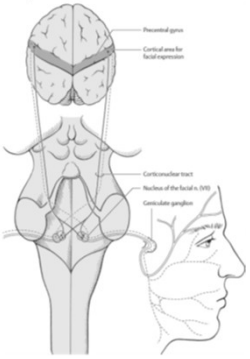


Fig. 4.32 **Peripheral course of the facial nerve**

CENTRAL FACIAL NERVE PARESIS

UPPER MOTOR NEURON 7TH CN LESION



Due to bilateral cortical representation of upper part of the facial nucleus (double corticobulbar innervation)

Sparing of the upper face often with a hemiplegia

DISSOCIATION OF VOLUNTARY AND EMOTIONAL INNERVATION AFTER STROKE

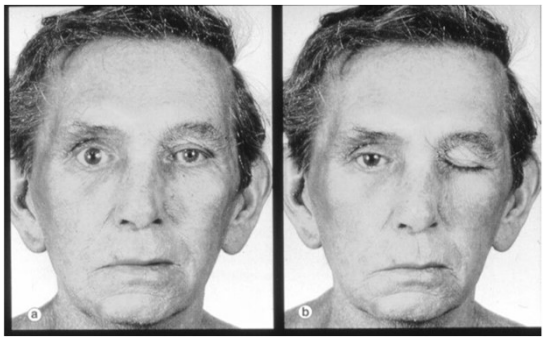
Ludwig Kappos, M.D. Matthias Mehling, M.D.
N Engl J Med 363;16 nejm.org october 14, 2010

LOWER MOTOR NEURON LESION

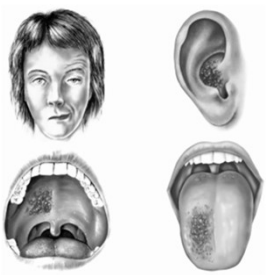
Causes:

- idiopathic - Bell's palsy – fairly abrupt onset (48hrs) proceeded by pain behind the ear (the small area of the 7th n. sensory innervation), taste sensation may be lost unilaterally, and hyperacusis may be present
- the Ramsay-Hunt syndrome – *geniculate zoster* – facial paralysis associated with a vesicular eruption in the pharynx, external auditory canal or other parts of the cranial skin; often 8th n. affected.
- acoustic neuromas and other tumors of the c-p angle
- Lyme disease
- trauma
- compression

BELL'S PALSY



CLINICAL FEATURES OF RAMSAY HUNT SYNDROME



Sweeney C J , and Gilden D H J Neurol Neurosurg Psychiatry 2001;71:149-154

JNNP

BILATERAL FACIAL PARALYSIS (FACIAL DIPLEGIA)

Bilateral Bell's palsy

Guillain-Barré syndrome

Lyme disease

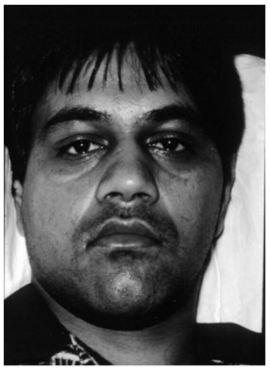
leprosy

Meningitis, cryptococcus or tubercoulus, and as a part of acquired immunodeficiency syndrome

Sarcoidosis, systemic lupus erythemathosus

Prepontine, intrapontine tumor

BILATERAL FACIAL PARALYSIS



NEW ONSET BELL'S PALSY TREATMENT

Steroids - highly likely effective in increasing the probability of complete facial functional recovery (NNT 6–8, 2 Class I studies).

- Start in first 72 hrs
- 60 mg/d for 5 days followed by a 5-day taper by 10 mg (other regime 25 mg BID for 10 days.)

Antivirals (in addition to steroids) might be offered (Level C). Patients offered antivirals should be counseled that a benefit from antivirals has not been established, and, if there is a benefit, it is likely that it is modest at best

- Acyclovir 1,0-2,0g / day per 10 dys; Valacyclovir 3,0 g/day000 7 days

EYE CARE IN FACIAL NERVE PALSY

Effective eye protection

- barrier protection (eg, wrapped sunglasses),
- lubrication (artificial tears during the day, ointment at night)
- taped closure at night.

FACIAL MOVEMENT DISORDERS

Facial myokimia – a fine rippling activity of the facial muscles; may be caused by a plaque of MS

Hemifacial spasm– contraction of muscles supplied by the 7 CN

THE VESTIBULOCOCHLEAR NERVE (8TH CN) AND CEREBELLOPONTINE ANGLE TUMOR

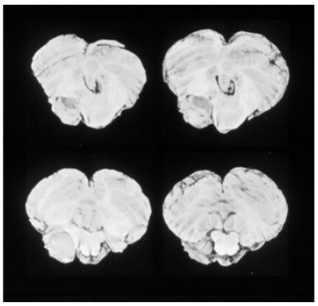
10% of cases of unilateral hearing loss are due to an acoustic neuroma.

unilateral hearing loss assoc. with tinnitus and vertigo (25%). Then facial pain and numbness, facial weakness and eventually ipsilateral cerebellar signs. Pain in the ear is seen and sometimes headache.

Audiometry shows a high tone loss, the caloric responses are depressed or absent

MRI is a technique of choice for establishing the diagnosis

CEREBELLOPONTINE TUMOR



THE GLOSSOPHARYNGEAL NERVE (9TH CN)

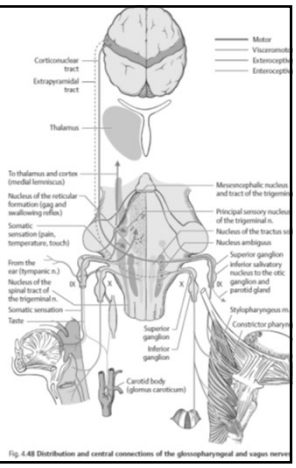


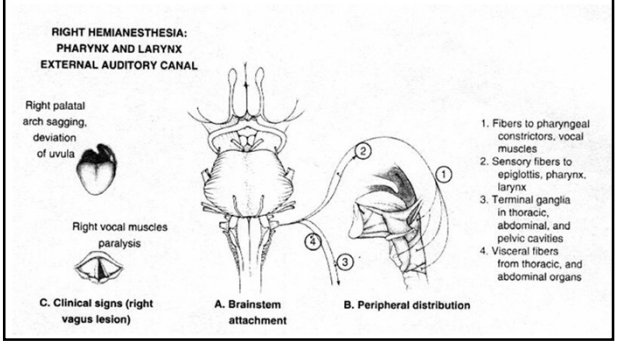
Fig. 4.48 Distribution and central connections of the glossopharyngeal and vagus nerves

THE GLOSSOPHARYNGEAL NERVE (9TH CN)

Isolated lesions of the nerve are rare.

In glossopharyngeal neuralgia paroxysm of pain in the tongue or throat occur and can be triggered by swallowing

VAGUS NERVE (10TH CN)



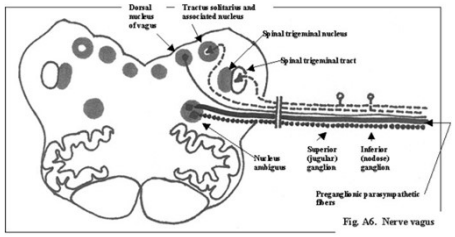


Fig. A6. Nerve vagus

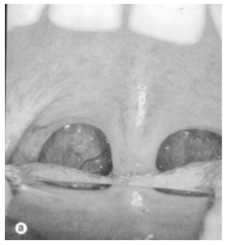
VAGUS NERVE

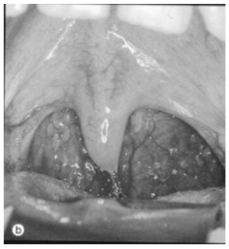
efferent component of the gag reflex.

In unilateral 10th CN palsy the soft palate lies lower on the affected side and along with the posterior pharyngeal wall moves toward the intact side during phonation.

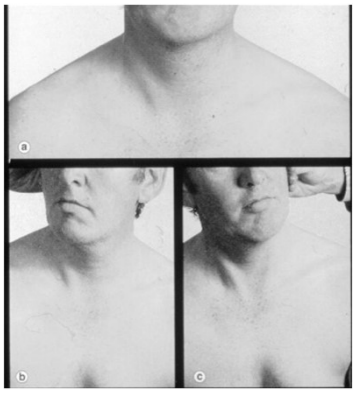
The vocal cord on the affected side lies fixed in a midposition resulting in slightly hoarse voice.

VAGUS NERVE PALSY



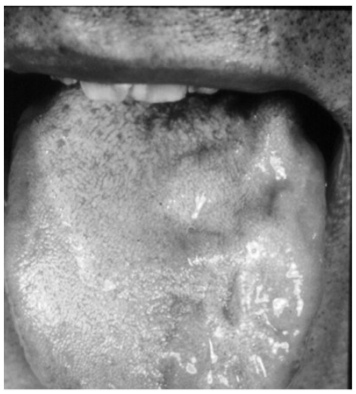


THE ACCESSORY NERVE (11TH CN)



HYPOGLOSSAL NERVE

A lesion of the hypoglossal nerve produces ipsilateral wasting and fasciculation of the tongue. Little effect on phonation or swallowing.



BULBAR AND PSEUDOBULBAR PALSY

Bulbar palsy - LMN

Vascular disorders, syringobulbia

lesion to IX, X, XII nuclei

- disturbances of swallowing,
- dysartria, palate paralysis,
- absent gag and palate reflexes
- atrophy and fasciculations of the tongue

Pseudobulbar palsy - UMN

bilateral lesion of corticospinal tracts (vascular disorders):

- dysartria,
- the tongue cannot be fully extruded,
- marked liability and „incontinence" of the affect,
- brisk jaw reflexes,
- UMN signs

COMBINED CRANIAL NERVE PALSIES

Multiple cranial nerves affected

- by result of trauma – sudden onset,
- localized infections (herpes zoster) – acute onset;
- Wegener's granulomatosis – subacute onset;
- tumours – chronic.

Jugular foramen syndrome (of Vernet)

- signs of damage to cranial nn. 9th, 10th and 11th (sometimes 12th) - causes: glomus jugular tumor

PROF. PETER GATES' RULE OF 4

"*Clinical Neurology: a primer*", 2010 Elsevier

'*The rule of 4 of the brainstem: a simplified method for understanding brainstem anatomy and brainstem vascular syndromes for the non-neurologist*'. Intern Med J. 2005 Apr;35(4):263-6.

THE 4 RULES OF THE 'RULE OF 4'

4 structures in the 'midline' beginning with M

4 structures to the side beginning with S

4 cranial nerves in the medulla, 4 in the pons and 4 above the pons (2 in the midbrain)

The 4 motor nuclei that are in the midline are those that divide equally into 12 except for 1 and 2 that is 3, 4, 6 and 12 (5, 7, 9 and 11 are in the lateral brainstem)

4 MEDIAL STRUCTURES AND THE ASSOCIATED DEFICITS

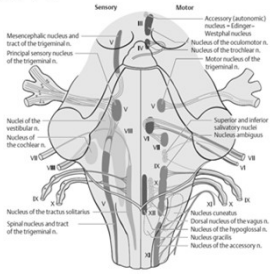
Structure	Deficit
Motor pathway (corticospinal tract)	Contralateral weakness of arm and leg
Median longitudinal fasciculus	Ipsilateral internuclear ophtalmoplegia
Medial lemniscus	Contralateral loss of vibration and proprioception affecting the arm and leg
Motor nuclei	Ipsilateral loss of the cranial nerve that is affected-3rd,4th,6th or 12th

4 LATERAL ('SIDE') STRUCTURES AND THE ASSOCIATED DEFICITS

Structure	Deficit
Spinocerebellar tract	Ipsilateral ataxia of the arm and leg
Spinothalamic pathway	Contralateral alteration of pain and temperature affecting the arm, leg and often the body
Sensory nucleus of the 5th CN	Ipsilateral alteration of pain and temperature sensation on the face
Sympathetic pathway	Ipsilateral Horner's syndrome

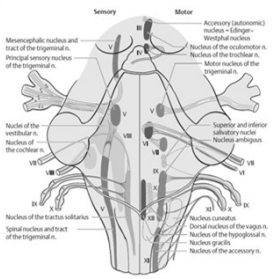
THE 4 CNS IN THE MEDULLA

Structure	Deficit
9th – glossopharyngeal	Ipsilateral loss of pharyngeal sensation
10th – vagus	Ipsilateral palatal weakness
11th – spinal accessory	Ipsilateral weakness of trapezius and sternocleidomastoid muscle
12th – hypoglossal	Ipsilateral weakness of the tongue



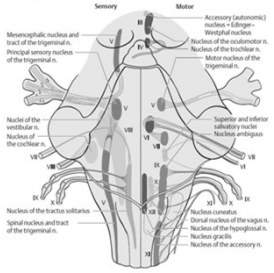
THE 4 CNS IN THE PONS

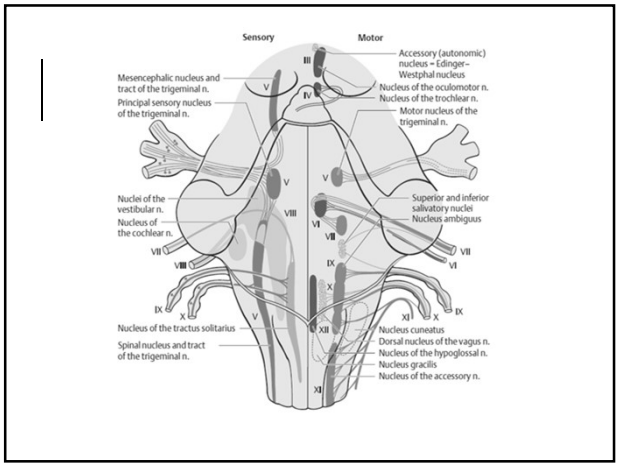
Structure	Deficit
5th – trigeminal	Ipsilateral loss of pain, temperature and light touch on the face
6th – abducent	Ipsilateral weakness of abduction of the eye
7th – facial	Ipsilateral facial weakness
8th – auditory	Ipsilateral deafness



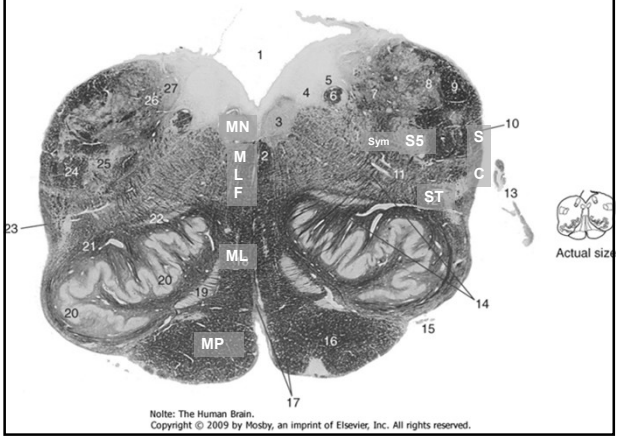
THE 4 CNS ABOVE THE PONS

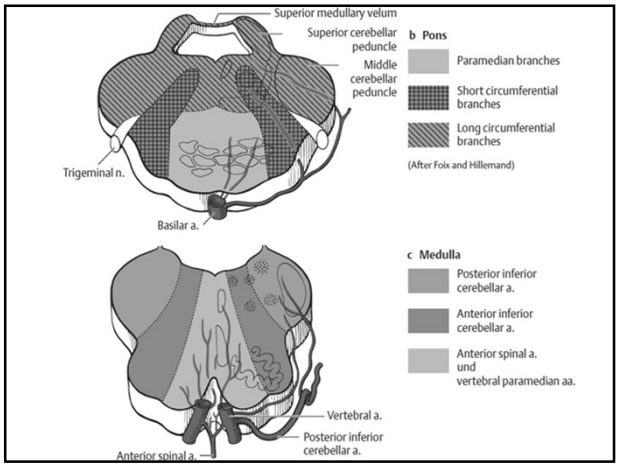
Structure	Deficit
1st – olfactory	Not in the midbrain
2nd – optic	Not in the midbrain
3rd – oculomotor	Impaired adduction, elevation and depression of the ipsilateral eye +/– dilated pupil
4th – trochlear	Impaired depression of the adducted eye

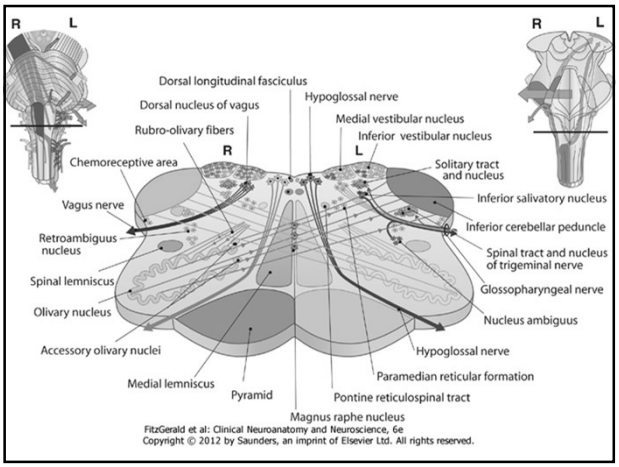




ROSTRAL MEDULLA

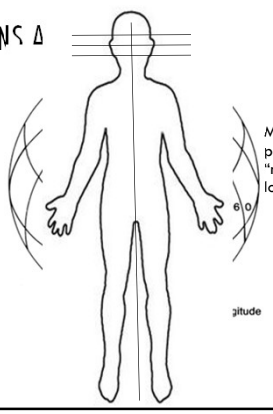






MERIDIANS Δ

Cranial nerves → "parallels of latitude"



Motor and sensory pathways → "meridians of longitude"

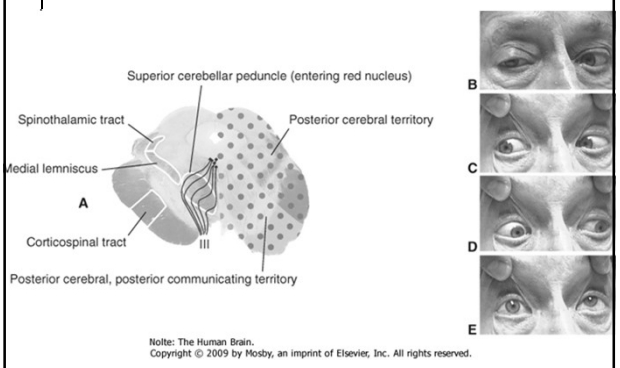
MEDIAN AND LATERAL BRAINSTEM SYNDROME

Median	Lateral
4Ms	4Ss
+	+
3rd, 4th, 6th, 12th	9th,10th and 11th (if in the medulla) Or 5th, 7th and 8th (if in the pons)

CASE - BRAINSTEM 3RD CN LESION

Signs
- ipsilateral 3rd paresis
  - Ptosis
  - Pupillary dilation
  - Lateral strabismus
- contralateral central 7th and hemiplegia

Localization
- middle cerebral peduncle affected
- Weber' syndrome

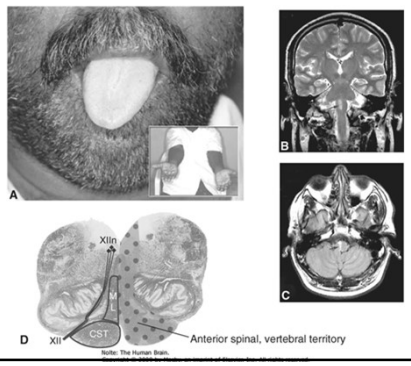


CASE

a 60 year old man … sought neurological consultation for sudden onset of numbness over the left side of the body.

Exam:
- weakness of the right side of his tongue
- tactile sensation on the left side of his body and left pyramidal weakness

MEDIAL MEDULLARY SYNDROME



CASE

50 years old male, non diabetic, non hypertensive but smoker presents with sudden onset of dysphagia, dysarthria and ataxia of the gait

Exam:
- loss of pain and temperature sensations over the contralateral body (with relative sparing of tactile sensation),
- loss of pain and temperature sensations over the ipsilateral face,
- hoarseness and difficulty in swallowing
- ipsilateral Horner's syndrome.

WALLENBERG SYNDROME – LATERAL MEDULLARY SYNDROME