Cranial Neuropathies

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Lecture Content: Neuropathies

- Olfactory
- Optic
- Oculomotor
- Trochlear
- Trigeminal
- Abducens
- Facial
- Vestibulocochlear
- Glossopharyngeal
- Spinal accessory
- Hypoglossal

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Question 1: A 27 year old female presents with six months of right shoulder weakness. She lost consciousness 6 months ago and landed on a radiator pipe, sustaining a burn injury to her right lateral neck. She can raise her arm to 90° but no higher. There is unilateral scapular winging with arms abducted. What is the affected muscle or muscles?

A. Serratus anterior
B. Trapezius
C. Deltoid
D. Rhomboid

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Question 2: A 37 year old male presents with left facial weakness involving the forehead and lower face and reduced taste sensation. Loud sounds are bothersome. What treatment should be offered?

A. Aspirin 81 mg daily
B. Acyclovir only
C. Acyclovir and prednisone
D. Prednisone only

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Question 3: A 43 year old male presents with left-sided weakness and neglect. His examination is notable for mild right ptosis. Eyes are midposition. The right pupil is reduced in diameter compared to the left. What muscle is most likely involved?

A. Levator palpebrae
B. Tarsal (Müller's muscle)
C. Orbicularis oris
D. Superior rectus

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Question 4: A 73 year old male presents with right shoulder droop and weakness of head turning, difficulty swallowing, and softening of speech. He reports decreased taste sensation. There is unilateral palatal droop noted on examination. Where is the most likely site of injury?

A. Median midbrain
B. Rostral pons
C. Lateral medulla
D. Jugular foramen

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Question 5: A 41 year old Caucasian male presents for acute onset hearing loss. Examination is notable for bilateral keratitis and an ataxic gait. What is the most likely diagnosis?

A. Ramsay-Hunt syndrome
B. Tolosa-Hunt syndrome
C. Multiple sclerosis
D. Cogan's syndrome

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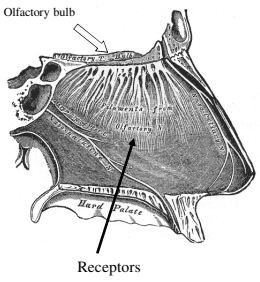
Olfactory Nerve

- Pathway
  - Olfactory receptors located in wall of the nasal cavity
  - Penetrate cribiform plate of ethmoid bone -> olfactory bulb
  - 2nd-order neurons course posteriorly as the olfactory tract
    - Crossed and uncrossed



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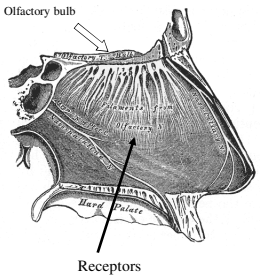
Olfactory Nerve *(cont.)*

- Pathway
  - Fibers go to
    - Frontal lobe
    - Temporal lobe
      - Terminate in the amygdala nucleus, hypothalamus, septal nuclei
  - Only sensory nerve that avoids the thalamus
  - Cortical representation is bilateral
    - Unilateral lesions distal to the decussation *do not* produce anosmia



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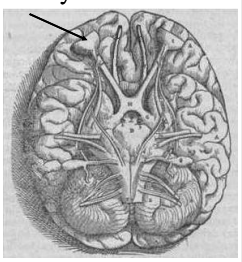
Olfactory Neuropathies

- Anosmia-lack of smell
  - Common cold
    - Most common cause of bilateral transient anosmia
  - Head trauma
    - Damage to fibers over cribriform plate
    - Back/side impact more damaging than frontal impact
    - Closed head injury can produce impairment of *recognition* despite preserved *detection*
  - Neurodegenerative disease
    - Sensitive as an initial deficit
      - Alzheimer's, Parkinson's, Huntington's disease
  - Other
    - Cystic fibrosis and adrenal insufficiency

Olfactory tract



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Olfactory Neuropathies

- Foster Kennedy syndrome
  - Noted with olfactory groove or sphenoid ridge masses
    - Commonly seen with meningiomas
  - Ipsilateral anosmia due to direct pressure on the olfactory bulb
  - Ipsilateral optic atrophy due to injury of the ipsilateral optic nerve
  - Contralateral papilledema due to raised ICP
  - *Do not confuse with….Pseudo Foster Kennedy syndrome*
    - May be noted when increased ICP of any cause occurs in a patient with previous unilateral optic atrophy
    - No anosmia!
    - Most often due to sequential anterior ischemic optic neuropathy

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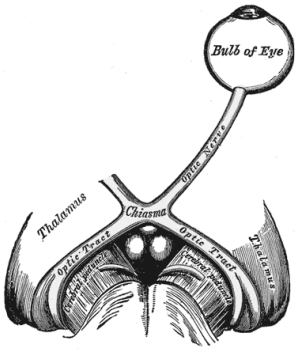
Optic Nerve

- Course
  - 50 mm long with 4 parts
    - Intraocular (nerve head)
    - Intraorbital
    - Intracanalicular
    - Intracranial
- Optic neuropathies
  - Anterior ischemic optic neuropathy (AION)
  - Posterior ischemic optic neuropathy (PION)
  - Optic neuritis
  - Leber's optic neuropathy



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Optic Neuropathy

- Anterior ischemic optic neuropathy (AION)
  - More likely in those >50, acute onset, minimal pain, limited recovery, altitudinal defect
  - Unilateral optic disc swelling
  - 2 types
    - Non-arteritic – Most common, painless, acute, altitudinal
    - Arteritic – Greater than 70 years of age
      - Usually giant cell arteritis
  - Associated with HTN, DM, OSA, hypercholesterolemia
- Posterior ischemic optic neuropathy (PION)
  - Similar to AION but ischemia *behind* the optic disc
  - Do not appreciate optic disc swelling
  - Bilateral PION commonly seen with cardiac and spinal surgeries
    - Surgeries greater than 6 hours
    - Patients with DM and carotid atherosclerosis

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Optic Neuropathy

- Optic neuritis
  - Demyelinating inflammatory condition
  - More likely in those <40, subacute onset, painful, good recovery
  - Four subtypes
    - Retrobulbar neuritis: optic nerve
    - Papillitis: optic disc
    - Perineuritis: optic nerve sheath (sparing the nerve)
      - Infection (syphilis)
      - Sarcoid
    - Neuroretinitis: swelling of the nerve & macula

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Optic Neuritis

- Clinical features
  - Vision loss
    - Gradual-occurs hours to days
    - Nadir within 1-2 weeks, recovery within 2-4 weeks, 6-12 months for nerve to fully heal
    - 2/3rds have 20/20 vision once recovered
  - Eye pain
    - 87% report pain, worse with movement
  - Loss of color vision
    - 88% with abnormal color vision (usually red and green)
  - Relative APD
    - Persists in >90% of cases
- Differential diagnosis
  - Commonly seen with inflammatory/autoimmune disease
  - Multiple sclerosis
  - Neuromyelitis optica
  - Syphilis
  - Cat scratch disease
  - Sarcoidosis
  - Lupus

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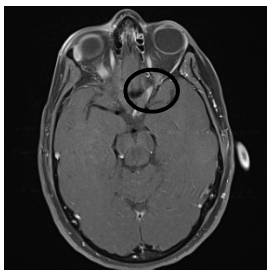
Optic Neuritis

- MRI
  - GdE fat saturated T1-weighted MRI of the orbits best sequence
  - GdE shows enhancement in 95% of cases
    - Rarely occurs in AION
  - Those without concomitant brain lesions have a 25% risk of MS vs. 72% with lesions



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Optic Neuropathies

- Leber optic neuropathy
  - Genetic cause of optic neuropathy
    - Point mutation in mitochondrial DNA
  - Adolescent males
  - Painless vision loss over weeks to months
  - Cardiac anomalies: atrioventricular conduction pathway defects (Wolf-Parkinson white)

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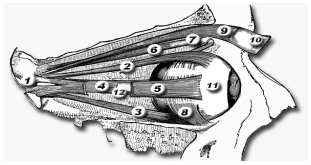
Oculomotor Nerve

- Function
  - Extraocular muscles
    - Superior rectus
    - Medial rectus
    - Inferior rectus
    - Inferior oblique
    - Levator palpebrae
  - Constricts the pupil
  - Accommodates
  - Converges



1. Annulus tendinous
2. Superior rectus
3. Inferior rectus
4. Medial rectus
5. Lateral rectus
6. Superior oblique
7. Trochlea of S.O.
8. Inferior oblique
9. Levator palpebrae
10. Eyelid
11. Eyeball
12. Optic nerve

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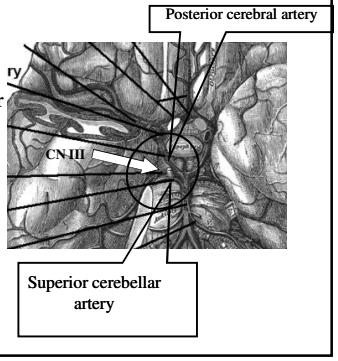
Oculomotor Nerve

- Pathway
  - Exits medial midbrain between midbrain & pons
  - Runs between the SCA and PCA
  - Then parallel to the posterior communicating artery
    - Parasympathetic fibers ride atop the nerve
  - Through cavernous sinus
  - Exits at superior orbital fissure
    - Splits into 2 divisions
      - Superior division
      - Inferior division



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Oculomotor Neuropathies

- Categorized by location
  - Nuclear lesions
    - Parinaud's syndrome
  - Fascicular lesions
    - Weber
    - Claude
    - Benedikt
  - Subarachnoid lesions
  - Carvernous sinus lesions
    - Tolosa Hunt syndrome

Classic 3rd nerve palsy
- Eye is "down & out"
- Dilated pupil
- Paralysis of accommodation (cycloplegia)
- Ptosis

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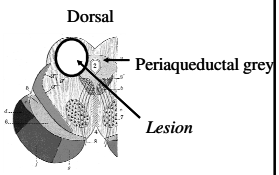
Oculomotor Neuropathies

- Nuclear lesions
  - Parinaud's syndrome
    - Lesion location
      - Dorsal midbrain
      - Periaqueductal grey
    - Clinical features
      - Supranuclear upgaze paralysis
      - Setting sun sign
        - Conjugate downgaze in primary position
      - Convergence and Eyelid retraction
        - Collier's sign
    - Causes
      - Pinealomas
      - Multiple sclerosis
      - Stroke
      - Hydrocephalus
        - VP shunt failure



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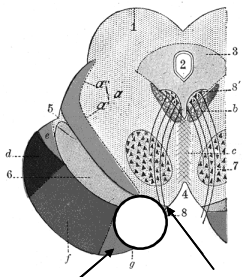
Oculomotor Neuropathies

- Fascicular lesions
  - Weber's syndrome
    - Lesion location
      - Base of midbrain
      - CNIII
      - Cortical spinal tracts
    - Clinical features
      - Ipsilateral 3rd nerve palsy
      - Contralateral hemiplegia



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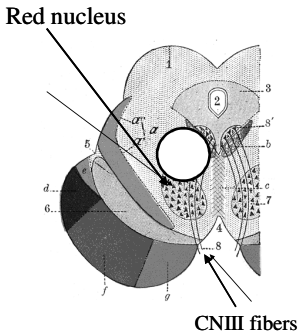
Oculomotor Neuropathies

- Fascicular lesions
  - Claude syndrome
    - Lesion location
      - CNIII
      - Red nucleus
      - Brachium conjunctivum
    - Clinical symptoms
      - Ipsilateral 3rd nerve palsy
      - Contralateral ataxia
      - Contralateral tremor



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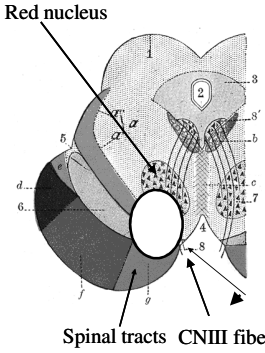
Oculomotor Neuropathies

- Fascicular lesions
  - Benedikt syndrome
    - Lesion Location
      - CNIII
      - Red nucleus
      - Cortical spinal tract
    - Clinical features
      - Ipsilateral 3rd nerve palsy
      - Contralateral tremor
      - Contralateral hemiplegia



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Oculomotor Neuropathies

- Subarachnoid lesions
  - Compressive lesions
    - Tumors, aneurysms
      - Dilated, unresponsive pupil
        - Absence of an affected pupil with complete motor paresis almost always excludes an aneurysm
      - Posterior communicating aneurysm the most common aneurysm to cause a CN IIIrd nerve palsy
    - Uncal herniation
      - Hutchinson pupil
      - Pupillary dilatation associated with poor response to light but preserved convergence
      - Ischemic lesions
    - Pupil sparing
      - Usually resolves in 3-6 months
    - Diabetes, giant cell arteritis

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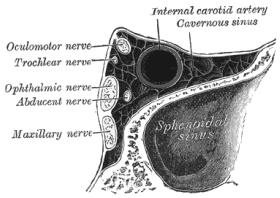
Oculomotor Neuropathies

- Cavernous sinus lesions
  - CN III, CNIV and CNVI
    - Resulting in oculomotor paresis
    - Fixed and dilated pupil
  - Trigeminal nerve V-1, V-2
    - Sensory loss over
      - Ophthalmic branch, maxillary branch
  - Postganglionic sympathetic fibers
    - Wrapped around internal carotid artery
    - Can result in a Horner's syndrome
      - Hard to note due to CNIII lesions
- Orbital apex syndrome
  - CNII, CNIII, CNIV, CNVI, CNV-1
    - Optic nerve is medial to the cavernous sinus
    - Results in cavernous syndrome with visual loss



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Oculomotor Neuropathies

- Cavernous sinus lesions
  - Tolosa-Hunt syndrome
    - Clinical Features
      - Episodic orbital pain
      - Episodic paralysis of either or all of CN 3, 4, 6
    - Diagnosis
      - Clinical history
      - Granuloma seen on MRI or biopsy
      - ESR/CRP elevated
      - CSF normal
      - Other causes excluded
    - Treatment
      - Sensitive to high dose steroids

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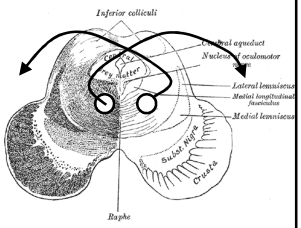
Trochlear Nerve

- Function
  - Innervates superior oblique muscle
    - Depresses, intorts and abducts the eye
- Pathway
  - Nucleus at level of the inferior colliculus
  - Exits midbrain dorsally & decussates
  - Runs along undersurface of tentorium
  - Along later wall of cavernous sinus
  - Enters orbit through superior orbital fissure
  - Cranial nerve with the longest course (75 mm)



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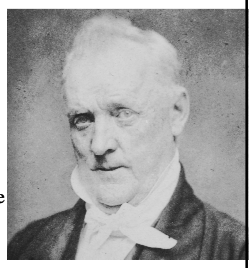
Trochlear Neuropathies

- Incomitant hypertropia
  - Clinical features
    - Vertical diplopia
      - Worse with adduction and downgaze
    - Head tilting
    - Hypertropia (elevated eye)
      - Occurs on side of the palsied nerve
      - Pts unconsciously tilt head away from the palsy
      - Worse
        - With lateral gaze to opposite side
        - Head tilt to same side (Bielschowsky test)
        - Downgaze
    - Weakness of down gaze



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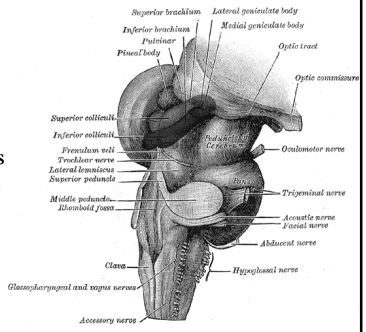
Trochlear Neuropathies

- Congenital palsy
  - Most common cause in children
  - Decompensation of congenital palsy should be suspected in all adults with new onset 4th nerve palsy
- Head trauma
  - Most common acquired cause



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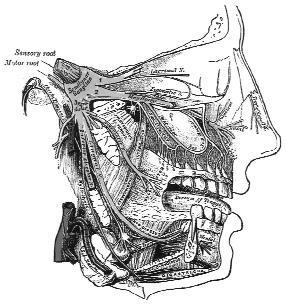
Trigeminal Nerve

- Function
  - Innervates muscles of mastication
    - Temporalis, masseter, lateral and medial pterygoids,
  - Other muscle groups
    - Tensor tympani, veli palatini, myohyoid and anterior belly of the digastric muscles
  - Sensation of face, eye, nasal and oral cavities
- Pathways
  - Motor nucleus
  - Medial to main sensory nucleus
  - Exits foramen ovale
    - →muscles of mastication



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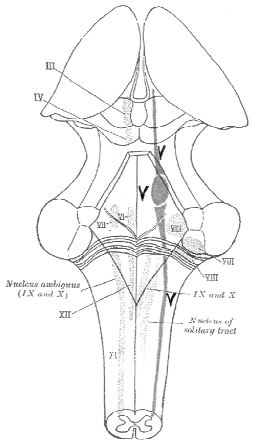
Trigeminal Nerve

- Pathways
  - Sensory bodies in trigeminal ganglion (in petrous bone, lateral to cavernous sinus)
    - 3 divisions
      - Ophthalmic (superior orbital fissure)
      - Maxillary (foramen rotundum)
      - Mandibular (foramen ovale)
    - Central processes
      - Synapse within main sensory nucleus
      - Synapse within spinal nucleus
        - Descend to different levels of the pons, medulla or cervical spine
    - Secondary neurons project to VPM-> cortex



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Trigeminal Neuropathies

- Trigeminal neuralgia (IHS criteria)
  - Paroxysmal brief attacks of pain involving one or more divisions of the trigeminal nerve
    - Pain has at least one characteristic
      - Intense sharp, superficial, stabbing
      - Precipitated from trigger zones or trigger factors
    - Attacks are stereotyped in the individual patient
  - Classical trigeminal neuralgia
    - No clinically evident neurological deficit
    - No other disorder to explain symptoms
  - Symptomatic trigeminal neuralgia
    - Causative lesion is found other than a vascular compression

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Trigeminal Neuralgia

- Basic facts
  - Maxillary branch most commonly affected
  - Female>male
  - Peak incidence ages 60-70
  - Unusual before age 40
  - Etiologies
    - Multiple sclerosis
    - Ectopic loop (SCA, ICA)
    - Meningioma
    - Bony deformity
    - Charcot-Marie-Tooth
    - Schwannoma
    - AV malformation
    - Tortuous basilar
    - Primitive trigeminal artery
    - Saccular aneurysm

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Trigeminal Neuralgia

- Treatments
  - Medicines
    - Carbamazepine (200 mg-1200 mg/qd)-established as effective
    - Oxycarbazepine (600 mg-1800 mg/qd)-probably effective
    - Baclofen, lamotrigine-possibly effective
    - Topical ophthalmic agents-probably ineffective
  - Surgical options
    - Percutaneous procedures on gasserian ganglion
      - Gamma knife and microvascular decompression probably effective

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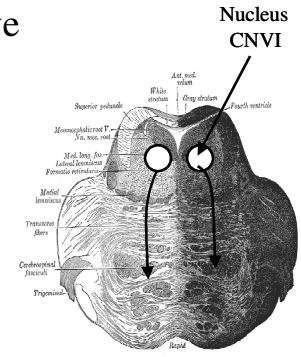
Abducens Nerve

- Function
  - Innervates lateral rectus muscle
    - Abducts the eye
- Pathway
  - Nucleus in lower dorsal pons
  - Emerges between pons & medulla
  - Lateral cavernous sinus
  - Exits out superior orbital fissure



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Abducens Neuropathies: Congenital

- Mobius syndrome
  - Horizontal gaze disturbance
  - Bilateral abducens palsies
  - Facial diplegia
  - Can be associated with
    - Limb abnormalities
    - Chest-wall abnormalities
    - Crossed eyes
    - Corneal erosions
- Duane's syndrome
  - Aplasia of one or both CNVI nuclei
  - Lateral rectus palsy
  - Some limitation of adduction
  - Retraction of eyeball into socket on adduction
  - Poor convergence
  - Face turns to affected side to compensate for limited movements
  - Can be associated with other ocular, ear and systemic malformations

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Abducens Neuropathies: Non-Congenita

- Millard-Gubler syndrome
  - Ipsilateral horizontal gaze palsy (CNVI)
  - Ipsilateral facial weakness (CNVII)
  - Contra lateral hemiparesis (cortical spinal tracts)
- Foville syndrome
  - Ipsilateral horizontal gaze palsy (CNVI)
  - Ipsilateral facial weakness
  - Contra lateral hemiparesis (cortical spinal tracts)
  - Contralateral sensory loss
  - Internuclear ophthalmoplegia
  - *Result of AICA infarct commonly*

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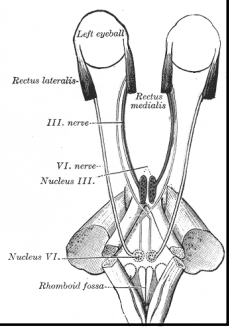
Abducens Neuropathies: Internuclear Ophthalmoplegia (INO)

- Anatomy
  - Internuclear neurons exit the abducens nucleus
  - Cross midline and arise in the MLF
  - Terminate in the MR nucleus
- Clinical features
  - Inability to adduct one eye with contralateral nystagmus
    - Adduction with convergence movements are intact
  - Common causes
    - Multiple sclerosis
    - Vascular disorders
    - Head trauma



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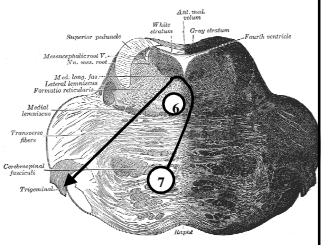
Facial Nerve

- Function
  - Muscles of facial movements
  - Other muscles
    - Stylohyoid muscle, posterior belly of the digastric, stapedius (dampens sounds)
  - Taste
    - Anterior 2/3rds of tongue
  - Salivation and lacrimation
    - Parasympathetic component that innervates lacrimal, submandibular and sublingual glands
  - Sensation
    - Posterior surface of the external ear and ear canal



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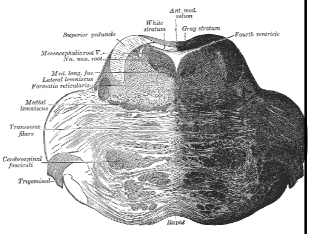
Facial Nerve

- Motor pathways
  - Nucleus in ventrolateral pons
  - Fibers swing around CNVI
  - Exits lateral caudal pons
- Sensory pathways
  - Superior salivatory nucleus
    - Reticular formation of lower pons
    - Innervates smooth muscle and glands for lacrimation
  - Tractus solitarius
    - Cell bodies in the geniculate ganglion
    - Taste for anterior 2/3rds tongue



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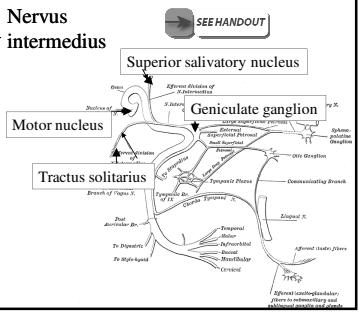
Facial Nerve

- Pathways
  - Superior salivatory nucleus
  - Tractus solitarius
  - Nervus intermedius + motor nucleus fibers
    - Join in the *internal auditory canal*→ Geniculate ganglion

Nervus intermedius



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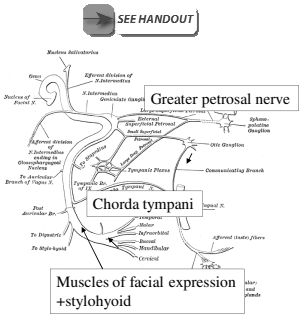
Facial Nerve

- Pathways
  - Geniculate ganglion (branches)
    - Greater petrosal nerve
      - Pterygopalatine ganglion
        - Lacrimal gland and glands of nose, palate and sinus
    - Stapedius
    - Chorda tympani
      - Lingual nerve-taste for anterior 2/3rds tongue
      - Submandibular ganglion
    - Stylomastoid foramen
      - Muscles of facial expression
      - Stylohyoid, posterior belly of the digastric, anterior/superior auricular muscle, occipitalis



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Facial Neuropathy: Bell's Palsy

- Incidence
  - Most common from ages 10-40yrs
- Symptoms
  - Pain behind ear
  - Hyperacusis
  - Facial weakness
  - Loss of taste
- Clinical course
  - 2/3rds recover spontaneously
  - 85% report improvement in about 3 weeks
    - 15% improvement within 3-6 months
- Treatment
  - N Engl J Med (2007)
    - Early treatment with prednisolone (25 mg BID) significantly improves the chances of recovery at 3 & 9 months
    - There is no evidence of a benefit of acyclovir in combination with prednisolone
    - Lancet neurology (2008)
      - Prednisolone (60 mg x5d then 10 mg x5) shortened the time to complete recovery in patients with Bell's
      - Valacyclovir had no affect on recovery

44

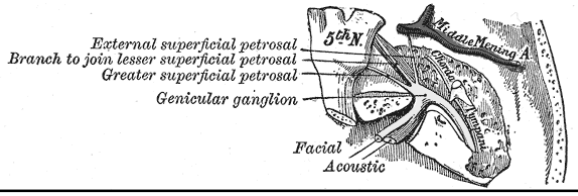
Facial Neuropathies

- Blepharospasm
  - Repeated involuntary bilateral contractures of the orbicularis oculi muscles
  - Common causes
    - Idiopathic
      - Meige syndrome
        - Idiopathic blepharospasm and oromandibular dystonia
        - Sustained grimacing around the mouth, platysma contraction and sustained neck flexion
    - Multiple sclerosis
    - Multi-system atrophy
- Hemifacial spasm
  - Unilateral involuntary hyperactive dysfunction
  - Insidious onset of painless, arrhythmic, tonic or clonic intermittent spasms
  - Lesions near CP angle are the most common cause

45

Vestibulocochlear Nerve

- Anatomy
  - Function and pathways
    - Auditory nerve
      - Receives information fro the cochlea (organ for hearing)
    - Vestibular nerve
      - Input from the saccular and utricle macules (linear acceleration)
      - Cristae of the semicircular canal (angular acceleration)



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Vestibulocochlear Neuropathies

- Vertigo
  - Subjective sense of movement by the patient that is false
  - Caused by imbalance of vestibular tone
    - Labyrinth disease
      - Associated with nausea and vomiting usually
- Ménière's disease
  - Episodic vertigo
  - Fluctuating sensorineural hearing loss
    - Low frequencies
  - Tinnitus
  - Usually unilateral

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Vestibulocochlear Neuropathies

- Neurofibromatosis I
  - AD
  - Chromosome 17
  - Protein: Neurofibromin
  - Tumors
    - Plexiform neurofibromas
    - Optic gliomas
    - High grade astrocytomas
  - Other
    - Café-au-lait
    - Axillary / inguinal freckling
    - Iris hamartomas (Lisch nodules)
- Neurofibromatosis II
  - AD
  - Chromosome 22
  - Protein: Merlin/Schwannomin
  - Tumors
    - Bilateral vestibular schwannomas
    - Meningiomas
    - Ependymomas
  - Other
    - Cataracts

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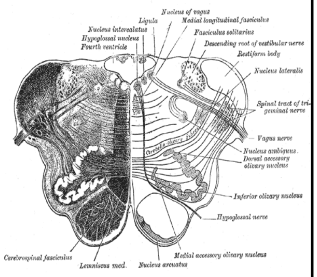
Vestibulocochlear Neuropathies

- Cogan's syndrome
  - A chronic inflammatory disease seen mostly in young white males
  - Symptoms
    - Bilateral sensorineural hearing loss
    - Progressive hearing loss with deafness in 2 years
    - Vestibular symptoms Ménière's like
    - Inflammatory ocular manifestations (keratitis)
  - Systemic symptoms
    - Seen in 30% of patients
    - Aortic, musculoskeletal complaints
  - Evaluation
    - Clinical presentation-imaging often normal
  - Treatment
    - Corticosteroids

49

Glossopharyngeal Nerve

- Anatomy
  - Shared by CN X also
- Function (categorized by nuclei)
  - Solitary nucleus
    - Taste and sensation in posterior 1/3rd of tongue
    - Carotid body (O_2 sensor)
    - Carotid sinus (baroreceptor Bp)
  - Spinal nucleus of V
    - Postauricular skin, inner tympanic membrane
  - Mesencephalic nucleus of V
    - Sensory (proprioception)-stylopharyngeus
  - Ambiguus nucleus
    - Motor- innervates stylopharyngeal muscle (elevates pharynx)
  - Inferior salivatory nucleus
    - Stimulates parotid gland to release saliva



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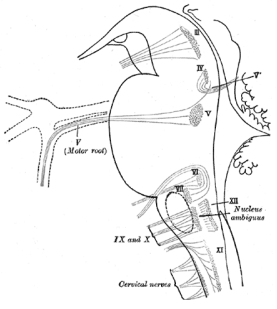
Glossopharyngeal Neuropathy

- Glossopharyngeal neuralgia
  - Clinical features
    - Unilateral stabbing, sharp paroxysmal pain
    - Abrupt severe pain in the throat, base of tongue or ear
    - Triggered by chewing, talking
      - May be associated with coughing, excessive salivation, hoarseness or syncope
    - Peak age 40-60
  - Treatment
    - Carbamazepine



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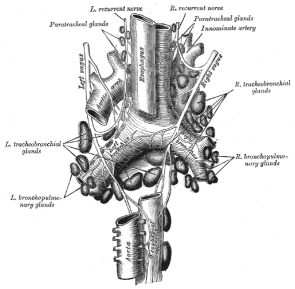
Vagus Nerve

- Anatomy
  - Shared also by CNIX
- Function (categorize based on nuclei)
  - Spinal nucleus of V
    - Sensation to external ear, auditory canal and external surface of tympanic membrane
  - Solitary nucleus
    - Visceral sensation
  - Nucleus ambiguus
    - Motor to striated muscles
    - Sensory fibers from below the vocal cords→recurrent laryngeal nerve
  - Dorsal motor nucleus of X
    - Motor to smooth muscles

52

Vagus Nerve

- Recurrent laryngeal nerve
  - Prone to injury throughout its course
    - Aneurysm of the aortic arch, subclavian artery, tracheal-bronchial lymph nodes, thyroidectomy
    - Rowland Payne syndrome: paralysis of the recurrent laryngeal, phrenic, vagal & Horners 2' breast CA
  - The left is longer & more likely to be injured



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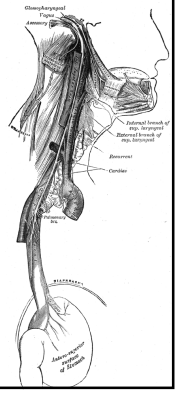
Glossopharyngeal and Vagus Nerve

- Syncope
  - Syncope may be the only symptom of metastatic involvement of CNs IX & X
    - Accompanies head and neck tumors, esp after recurrence
    - "Swallow syncope" assoc with esophageal CA
      - Pts report paroxysmal pain lasting seconds to 30 min



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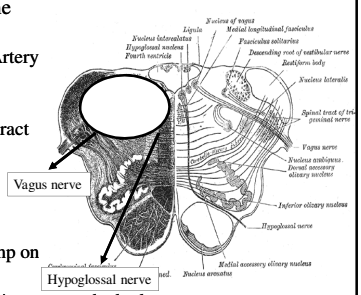
Glossopharyngeal and Vagus

- Lateral medullary syndrome "*Wallenberg Syndrome*"
  - Vessel: PICA/Vertebral Artery
- Location
  - Spinothalamic tract
  - Descending sympathetic tract
  - CN IX,X
  - Vestibular nuclei
- Clinical symptoms
  - Nystagmus, vertigo, N/V
  - Ipsilateral loss of pain/temp on face
  - Contralateral loss of pain./temp over the body
  - Ipsilateral horner's
  - Ipsilateral paralysis of palate/vocal cord
    - Diminished gag, hoarseness, dysphagia



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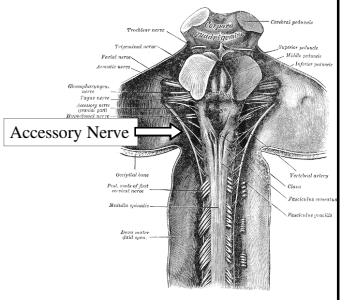
Spinal Accessory Nerve

- Function
  - Motor for sternocleidomastoid and trapezius muscles
- Pathway
  - Originates from medulla and spinal cord (C1-C6)
  - Fibers unite and ascend
    - Enter skull through foramen magnum
    - Exit skull through jugular foramen
      - Cranial portion joins the vagus to supply pharynx and larynx
      - Extra cranial portion supplies the sternocleidomastoid and trapezius



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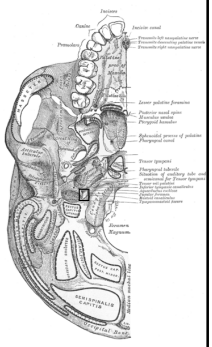
Spinal Accessory Neuropathies

- Jugular foramen syndrome
  - "Vernet's syndrome"
    - Cranial nerves IX, X, XI pass through the foramen
    - Obstruction leads to
      - Ipsilateral trapezius and sternocleidomastoid paresis
        - Weakness turning head away from lesion and ipsilateral weak shoulder shrug
      - Dysphonia with palatal droop
      - Dysphagia with absent gag reflex
      - Loss of taste over posterior 1/3rd of tongue
      - Depressed sensation over posterior 1/3rd of tongue, soft palate, uvula, pharynx and larynx



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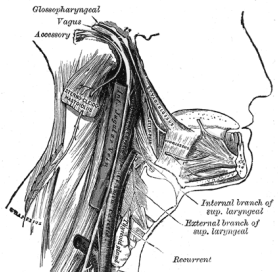
Spinal Accessory Neuropathies

- Winged scapula
  - Isolated spinal accessory neuropathy
    - Can occur with surgery, line placement (jugular cannulation), lymph node biopsy>tumor excision>trauma
  - The trapezius is required to rotate the scapula in order to elevate the arm above the horizontal
    - The arm can not be abducted above the horizontal
    - The upper portion of the scapula falls laterally, the inferior angle is drawn medially, and the vertebral border is flared
    - This is accentuated on attempted abduction



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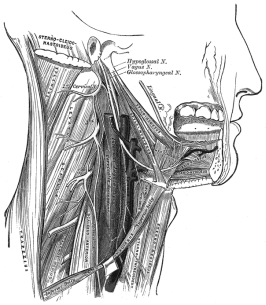
Hypoglossal Nerve

- Function
  - Movements of the tongue
- Pathways
  - Nucleus runs from pontine-medullary junction to caudal medulla
  - Rootlets unite and pass through hypoglossal canal



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Hypoglossal Neuropathy

- Dejerine's anterior bulbar syndrome
  - Occlusion of anterior spinal artery or its parent vertebral artery
  - 3 clinical features
    - Ipsilateral paresis, atrophy and fibrillations of the tongue
      - *Protruded tongue deviates toward the lesion*
    - Contralateral hemiplegia sparing the face
    - Contralateral loss of position and vibratory sensation (pain and temperature are spared)

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Hypoglossal Neuropathy

- Collet-Sicard syndrome
  - Lesion damaging hypoglossal nerve and jugular foramen
  - 5 clinical features
    - Ipsilateral trapezius and sternocleidomastoid paralysis
    - Vocal cord and pharynx weakness
    - Hemiparalysis of the tongue
    - Loss of taste on posterior 1/3rd of the tongue
    - Hemianesthesia of the palate, pharynx, larynx

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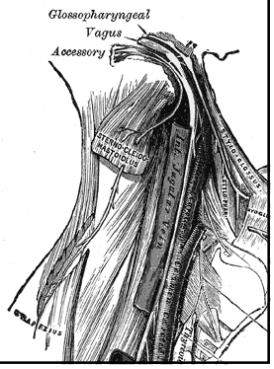
Question 1: A 27 year old female presents with six months of right shoulder weakness. She lost consciousness 6 months ago and landed on a radiator pipe, sustaining a burn injury to her right lateral neck. She can raise her arm to 90° but no higher. There is unilateral scapular winging with arms abducted. What is the affected muscle or muscles?

A. Serratus anterior
B. Trapezius
C. Deltoid
D. Rhomboid

62

Question 1: Explanation

- B. The trapezius. Scapular winging is a common question—and the long thoracic nerve/serratus anterior is the common answer. However, this patient sustained lateral neck trauma and cannot raise the arm greater than 90°. The spinal accessory nerve can be easily injured in the neck due to its superficial course. The trapezius helps stabilize & rotate the scapula



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Question 2: A 37 year old male presents with left facial weakness involving the forehead and lower face and reduced taste sensation. Loud sounds are bothersome. What treatment should be offered?

A. Aspirin 81 mg daily
B. Acyclovir only
C. Acyclovir and prednisone
D. Prednisone only

64

Question 2: Explanation

- D. The patient presents with typical features of Bell's palsy. Two recent studies indicated that prednisone alone is superior to acyclovir or acyclovir & prednisone. Aspirin is not indicated

Treatment

N Engl J Med (2007): In patients with Bell's palsy, early treatment with prednisolone (25mg BID) significantly improves the chances of recovery at 3 & 9 months. There is no evidence of a benefit of acyclovir in combination with prednisolone.

Lancet Neurology (2008): Prednisolone (60mg x5d then 10mg x5) shortened the time to complete recovery in patients with Bell's, whereas valcyclovir did not affect facial recovery.

65

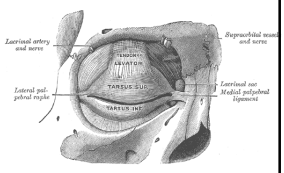
Question 3: A 43 year old male presents with left-sided weakness and neglect. His examination is notable for mild right ptosis. Eyes are midposition. The right pupil is reduced in diameter compared to the left. What muscle is most likely involved?

A. Levator palpebrae
B. Tarsal (Müller's muscle)
C. Orbicularis oris
D. Superior rectus

66

Question 3: Explanation

- B. The tarsal muscle has sympathetic innervation. It contributes mildly to lid elevation (far less than the levator palpebrae innervated by the cranial nerve III). It can be injured anywhere along its path. In this particular case, the mechanism of injury was a right carotid dissection. Fibers that are responsible for eyelid elevation and pupil size reside on the ICA; fibers responsible for sweating are on the ECA



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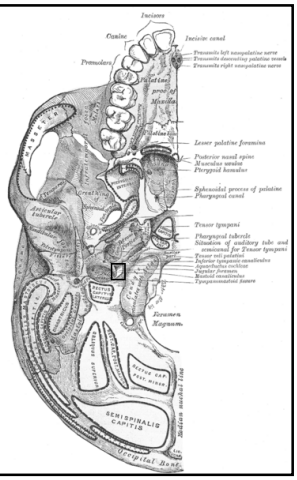
Question 4: A 73 year old male presents with right shoulder droop and weakness of head turning, difficulty swallowing, and softening of speech. He reports decreased taste sensation. There is unilateral palatal droop noted on examination. Where is the most likely site of injury?

A. Median midbrain
B. Rostral pons
C. Lateral medulla
D. Jugular foramen

68

Question 4: Explanation

- D. The IX, X and XIth cranial nerves run together through the Jugular Foramen. Glomus jugulare tumors and basal skull fractures can injure all three CNs at this site



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Question 5: A 41 year old Caucasian male presents for acute onset hearing loss. Examination is notable for bilateral keratitis and an ataxic gait. What is the most likely diagnosis?

A. Ramsay-Hunt syndrome
B. Tolosa-Hunt syndrome
C. Multiple sclerosis
D. Cogan's syndrome

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Question 5: Explanation

- D. Cogan's syndrome is a chronic inflammatory disease. It is common in young white males
- Symptoms include
  - Bilateral sensorineural hearing loss (acute onset)
    - Progressive hearing loss up to deafness within 2yrs
  - Vestibular symptoms
    - Ménière disease
  - Inflammatory ocular manifestations (non-syphilitic interstitial keratitis)

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Questions & Answers

The End