Facial Nerve Disorders

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FIACAL PALSY

Etiology


2. Ear pathology (ear infection, cholesteatoma).


4. Reinnervation (any lesion that can cause unilateral palsy).

5. Trauma (fracture of temporal bone, birth trauma) improvement is rare (when palsy is associated with head trauma), but recovery may not be complete!

6. Surgery:

a) Palsy immediately after surgery - nerve transaction - prompt surgical evaluation

b) Delayed onset - nerve edema - improves with time.

7. Tumors (of nerve, of temporal bone, of parotid gland) facial schwannoma (5% of CN7 palsies) see p. Onc62>> palpate parotid bimanually in all patients! thorough neck examination for adrenopathy!

8. Toxins (arsenic, CO, ethylene glycol).

9. Part of polyequilitidis (leprosy, Guillain-Barré syndrome, diphtheritic; only seldom in diabetic or alcoholic neuropathy?)

10. Idiopathic - Bell’s palsy.

Etiology of NUCLEUS LESION - vascular lesions, tumors, inflammatory lesions, acute polymyelitis, MS.

Bilateral facial palsy (any lesion that can cause unilateral palsy) - most often sarcoidosis, Guillain-Barré syndrome, leprosy, leukemia, meningococcal meningitis.

Positive FAMILY HISTORY:

1. MEIKERSON-Rosenenthal syndrome.

2. MöBIus syndrome.

3. BELL’s palsy (occasionally).

Perinatal facial palsy:

1. Maternal infections (e.g. rubella).

2. Drugs used during pregnancy (e.g. thalidomide).

3. Difficult delivery (esp. forceps) - recovers spontaneously in few days or weeks.

4. CT7 (11)
GUIDE TO LESION SITE LOCALIZATION

1. Other neurological symptoms (e.g. hemiplegia, loss of sensation, cranial nerve dysfunction) - ipsilateral hemiparesis - cortical / subcortical lesion; contralateral hemiparesis - pontine lesion near facial motor nucleus.

2. Otologic signs and symptoms

3. What other functions of CN7 are involved:
   a) change in taste (quantified by Schirmer test) see p. D1 >>
   b) lesion distal to geniculate ganglion → tear production)
   c) change in tear production (e.g. irritation from lagophthalmos); atomic epiphora (ectropion from orbicularis oculi weakness → tears pool in lower conjunctival sac and spill over lower lid margin).

   2) reduced taste (quantified by electrogustometry - taste threshold in response to small electric current applied to anterior tongue).

   3) reduced salivation (quantified by cannulating Wharton duct or by checking saliva pH - pH rises with increased salivation).

   4) HYPERACUSIS (confirmed by acoustic reflex testing).

CLINICAL GRADING OF SEVERITY

HOUSE-BRACKMANN GRADING SCALE

<table>
<thead>
<tr>
<th>Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Normal symmetrical function in all areas</td>
</tr>
<tr>
<td>II</td>
<td>Slight weakness noticeable only on close inspection</td>
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<tr>
<td></td>
<td>Complete eye closure with minimal effort</td>
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<tr>
<td></td>
<td>Slight asymmetry of smile with maximal effort</td>
</tr>
<tr>
<td></td>
<td>Synkinesis barely noticeable, contracture, or spasm absent</td>
</tr>
<tr>
<td>III</td>
<td>Obvious weakness, but not disfiguring</td>
</tr>
<tr>
<td></td>
<td>May not be able to lift eyebrow</td>
</tr>
<tr>
<td></td>
<td>Complete eye closure and strong but asymmetrical mouth movement with maximal effort</td>
</tr>
<tr>
<td></td>
<td>Obvious, but not disfiguring synkinesis, mass movement or spasm</td>
</tr>
<tr>
<td>IV</td>
<td>Obvious disfiguring weakness</td>
</tr>
<tr>
<td></td>
<td>Inability to lift brow</td>
</tr>
<tr>
<td></td>
<td>Incomplete eye closure and asymmetry of mouth with maximal effort</td>
</tr>
<tr>
<td></td>
<td>Severe synkinesis, mass movement, spasm</td>
</tr>
<tr>
<td>V</td>
<td>Motion barely perceptible</td>
</tr>
<tr>
<td></td>
<td>Incomplete eye closure, slight movement corner mouth</td>
</tr>
<tr>
<td></td>
<td>Synkinesis, contracture, and spasm usually absent</td>
</tr>
<tr>
<td>VI</td>
<td>No movement, loss of tone, no synkinesis, contracture, or spasm</td>
</tr>
</tbody>
</table>

CLINICO-ANATOMICAL SYNDROMES

<table>
<thead>
<tr>
<th>Lesion Site</th>
<th>CN7 Findings</th>
<th>Possible Other Findings</th>
<th>Common Etiologies</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Motor cortex, internal capsule</td>
<td>Contralateral central* palsy of lower face (upper face spared**); tearing, salivation, taste intact</td>
<td>Hemiparesis (ipsilateral); tongue weakness (ipsilateral); frontal lobe signs</td>
<td>Vascular</td>
<td></td>
</tr>
<tr>
<td>Ponto-medullary junction</td>
<td>Ipsilateral full* palsy; tearing, salivation, taste intact</td>
<td>CN7; CN8 lesion; cerebellar ataxia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cerebellopontine angle</td>
<td></td>
<td>Tumors (acoustic / facial neuralgia, meningioma, cholesteatoma)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Internal auditory meatus (MESTAL SEGMENT)</td>
<td>Ipsilateral full palsy; [tearing], [salivation], [taste]</td>
<td>CN8 lesion</td>
<td>Intracranialulceromas (acoustic or facial)</td>
<td>CN8 is usually involved first</td>
</tr>
<tr>
<td>Facial canal: Labyrinthine segment</td>
<td>Ipsilateral full palsy; [tearing], [salivation], [taste]</td>
<td>Inner, middle ear dysfunction</td>
<td>Bell’s palsy; Ramsay Hunt syndrome, temporal fracture, hemangiomata</td>
<td>Cochlea / vestibular organs may be involved.</td>
</tr>
<tr>
<td>Facial canal: Tympanic segment</td>
<td>Ipsilateral full palsy; [tearing], [salivation], [taste]</td>
<td></td>
<td>Bell’s palsy; otitis media, cholesteatoma, temporal bone fracture</td>
<td>With lesion distal to chorda tympani, salivation &amp; taste are spared.</td>
</tr>
<tr>
<td>Facial canal: Mastoid segment</td>
<td>Ipsilateral full palsy; [tearing]</td>
<td>Inner, middle ear dysfunction</td>
<td>Facial trauma or surgery; parotid tumors; sarcoidosis</td>
<td></td>
</tr>
<tr>
<td>Stylo-mastoid foramen</td>
<td>Ipsilateral full* palsy; salivation**, taste, tearing intact</td>
<td></td>
<td></td>
<td>Staring of peripheral branches possible; drooling</td>
</tr>
</tbody>
</table>

NONSENSE/CLEAR (CENTRAL PAIN)

- only contralateral lower face is paralyzed (esp. perioral muscle); mild upper face weakness is possible.
- see table above
- paralysed m. buccinator → chewing difficulty (food pools in vestibulum oris)
- paralyzed m. orbicularis oris → flattened nasolabial fold, drooping mouth corner, drooling
- when patient attempts to smile, lower face is pulled to opposite side (this distortion may give false appearance of deviation of protruded tongue or open jaw)

- emotional control of facial motion is provided by EXTRAPYRAMIDAL INPUT (via RF - from frontal cortex, thalamus, globus pallidus) – voluntary (pyramidal) and emotional (extrapyramidal) components may be affected separately.
**NIGHT**

**guide as to severity of corneal epithelial disruption**

**Eye protection**

All complete

4.

3.

2.

a) "preservative free" artificial tears

b) contact lenses

c) punctal plug insertion

d) room humidifiers

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**SCHIRMER**

- **only motor facial palsy.**
- **some motor branches may be spared.**

**DIAGNOSIS**

Schirmer test (lacrimal secretion) ⇒ see p: D1 >>

1. Check cornea for exposure keratitis!

2. **AUDIOMETRY** should be performed in all patients.

3. **NEUROIMAGING** (MRI, CT) indications:
   - 1) unmyelinated (nerve bundles)
   - 2) recurrent / progressive / atypical paralysis
   - 3) trauma.

4. **ELECTROPHYSIOLOGY**:
   - 1) **ENoG** (electronystagmography) - in acute unilateral complete paralysis (if threshold is 3.5 mA greater than normal side or if ≥ 90% axons are lost, there is poor prognosis for spontaneous recovery) during days 4-21.
   - even in complete transaction, nerve conduct impulses distal to injury for up to 72 hours (until Wallerian degeneration occurs) - **ENoG** is not performed until more than 3 days after total paralysis!
   - **ENoG** is also not of benefit in long-standing or congenital facial paralysis.
   - 2) **voluntary EMG** is helpful in long-standing or congenital facial paralysis.
   - 3) **trigeminal reflex** (electrodes record blink reflex after percutaneous stimulation of supraorbital nerve) - conduction of reflex arc between CN5 (afferent) and CN7 (efferent) - the only direct test able to measure intracranial CN7 pathology!!!

5. If facial reanimation procedure is considered (in long-standing or congenital facial paralysis) ⇒ facial **MUSCLE HOPS** (whether viable muscle fibers are present).

**GENERAL MANAGEMENT**

All complete traumatic cases must be considered for surgical repair!

Good guide (also for patients) - http://www.bellspalsy.ws

Lacrimal protection from exposure keratitis (esp. if ipsilateral CN5 deficit co-exists) - patient has no reliable guide as to severity of corneal epithelial disruption:

**DAY:**

- **artificial tears** frequently (q15-30min); best are "for sensitive eyes", "non-allergic", or "preservative free" artificial tears, **manuka honey** is preservative that can be particularly irritating!
- room humidifiers
- soft **contact lenses** are hydrophilic (hold water) - can provide source of moisture directly over cornea + protect eye from debris.
- **eyelids** help to hold contact lens in place - if kids become limp, contacts cannot be worn until function begins to return.
- **wearing lens is not complete replacement for your tears - use plenty of saline or eye drops designed for use with contacts.**
- **at acute stage; eye can be so dry that it is impossible to keep contact lens moist for more than few minutes - contacts can not be worn!!**
- **Do not let the lens dry out**!
- **sunglasses** while outside (avoid contaminated air!)

**NIGHT:**

- **ophthalmic ointment** (e.g. with dexpanthenol) - **thicker than artificial tears due to addition of mineral oil.

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**SUPERFICIAL_OOPHARYNGEALANEUROSIS**

- ipsilateral full (upper + lower) face paralysis.

**STYLOMASTOID_FACIAL_PARALYSIS**

- only motor facial palsy.
- some motor branches may be spared.
2) eye patch or taping closed affected eyelid*

*if not done correctly eyelid can easily pop open, exposing eye to worse damage than without tape; make sure to use gentle, non-abrasive and easily removed tape (such as paper surgical tape), and learn correct way to apply it.

Any corneal abrasion / infection should be treated immediately.

REHABILITATION

For early period (when muscles are completely flaccid) - limit therapy to:

1) moist heat (to ease soreness and reduce swelling)
2) massage (to ease soreness, to provide degree of motion & stimulation to muscles and increase circulation)
3) mental exercises (to retain “memory” of facial motions).

Focus your exercise energy on maintaining brain-to-nerve-to-muscle connection - this is more important than physical motions your muscles did before, and will do again.

- when laughing / talking support face muscles with hand – to counteract pulling by muscles of nonparalysed side.
- some advocate during recovery period – electrophoresis with PROSERINE.
- ELECTRICAL STIMULATION continues to be widely used in treatment of facial paralysis although there is mounting evidence that it may be contraindicated:
  - facial muscles resist degeneration post denervation for longer periods of time than other skeletal muscle - facial muscles may remain viable for 3 or more years, - use of electrical stimulation to maintain viability of facial muscle is unfounded!
  - electrical stimulation may interfere with neural regeneration!
  - patients who undergo electrical stimulation acutely may demonstrate more synkinesis and mass action than those who do not (it is difficult to produce isolated contraction of facial muscles using electrical stimulation due to their small size and close proximity to each other - contraction produced causes mass action which reinforces abnormal motor patterns and can be painful)!
  - avoid gross maximum effort exercises (close your eyes as hard as you can, “smile broadly”, “pucker your lips”, etc) - recruit excessive motor units (producing patterns that differ from typical facial expressions which are gentle and fluid) + reinforce abnormal movement patterns.

N.B. one of first signs of regenerating nerve may be pain; in case of CN7 - pain in ear canal!

For permanent facial paralysis:

- massage / electric stimulation of paralyzed muscles (no proven benefit!) – to prevent atrophy, to relieve strain on relaxed muscles and to preserve tone.
- wait minimum 6 months after subtotal nerve stump formation (return function (exception - nerve cut during surgery - restore nerve continuity ASAP!)

A. STATIC PROCEDURES:

1) gold weights, weight-adjustable magnet, palpebral springs, silicone-encircling bands can be inserted into eyelids.
2) lateral tarsorrhaphy (reversible), canthoplasty (irreversible), lower lid shortening.
3) sling suspension (with fascia lata or alloplastic strips) – for very old / very ill patients.

B. DYNAMIC PROCEDURES:

1) extracranial CN12 –CN7 or CN11 –CN7 anastomosis; sooner or later, new motor pattern develops in cerebral cortex and movements of facial muscles are dissociated from those of tongue / shoulder.
2) cross-facial nerve grafting (part of proximal nerve on intact side is anastomosed to distal nerve on paralyzed side with use of nerve grafts)
3) muscle transfer procedures are preferred in patients > 50 years (have relatively small chance of success with nerve grafting): – temporalis or masseter transposition; – free muscle grafts; – microvascular free nerve-muscle grafts.

SPECIFIC DISORDERS

BELL’S PALSY (s. idiopathic acute facial palsy)

Epidemiology, Etiology

- 60-75% of all cases of facial paralysis.
- INCIDENCE: 20-30 cases per 100,000 population per year (increases with age).
- male = female, right = left.
70-100% cases are recurrent (ipsilateral or contralateral).

*consider tumor as etiology

4-10% have positive family history.

** RISK FACTORS: pregnancy (triple risk), diabetes mellitus (quadruple risk), immunosuppression.

60-70% had viral prodrome: 7-10 days before.

most commonly accepted cause is **HERPES SIMPLEX-1** infection (reactivation of HSV genomes from gieniculate ganglia); other suggested causes – other viruses, vascular ischemia.

**mechanism** – inflammatory nerve edema within rigid facial (falloppian) canal.

alternative cause/ mechanism – body arteres elongate and become tortuous (may do so even in third decade of life) --> **sudden shift of arterial loop** in cerebellepontine angle:

- rapid stretch of N7-8 bundle between brain stem and internal auditory meatus from anterior aspect (facial nerve side), actively compressing nerve against petrous bone.

- b) direct compression of intrapontine facial nerve.

**CLINICAL CHARACTERISTICS**:

- **proptosis preceding symptoms** - facial paralysis may be heralded/ accompanied by pain behind ear - sense of mastoid fullness (pain at stylo mastoid foramen), periauricular paresthesias or even otalgia (without vesicles/!)

- **sudden onset**; maximal facial weakness is reached within 48-72 hours (vs. CN7 schwannoma – progresses over > 3 weeks).

- peripheral CN7 dysfunction may involve all branches (LMN palsy, dyguesia, hyperacusis).

- may complain of numb / heavy feeling in face, but no sensory loss is demonstrable.

- pain is absent (if present – consider Ramsay-Hunt syndrome).

- at onset, and for variable time thereafter, ipsilateral ocipital headaches and frequently tenderness over ipsilateral occipital bone is present.

- no other symptoms & signs!!!

- may develop ipsilateral CN8 dysfunction (tinnitus, vertigo, disequilibrium, sensorineural hearing loss).

**DIAGNOSIS**:

- **clinical**:

- diagnosis of exclusion!!!

- no imaging is required! (MRI may show contrast enhancement of nerve, but CT and X-rays are typically negative) – imaging results do not change initial management.

N.B. perform MRI if tumor is suspected!

- in endemic areas, Lyme disease is consideration.

**ENG (electroneurography)** during days 4-21 – for prognostic purposes. see below

**PROGNOSIS**:

- spontaneous improvement within 6 months.

- without treatment, improvement is noted in 3-12 weeks in 85%; remainder show improvement by 3-6 months (= 70% regain normal function; vs. 92% with treatment).

N.B. all patients should show some improvement by 6 months! (vs. CN7 schwannoma – gradually worsening paralysis, steroid responsiveness eventually disappears)

- denervation (after 10 days from onset) indicates axonal degeneration - will be long delay (3 months, as rule) before regeneration occurs (may be incomplete).

- when recovery is partial, “contractures” may develop on paralyzed side (400 mg ×5 /d) is not helpful; adding acyclovir is **PREDNISONE** on day 1 of protocol.

- only may improve outcome!!! (must be used only in COMBINATION WITH PREDNISONE).

- **ACYCLOVIR alone is not effective in facial recovery!!!** (must be used only in COMBINATION WITH PREDNISONE).

- **ACYCLOVIR** alone is not effective in facial recovery!!!

- in Lithuania, diuretics are also administered.

- those who will not recover conservatively must be identified* early – within 2 weeks (to prevent permanent paralysis).

- 2) **degeneration of ≥ 90% axons on electroneurography** – **SURGICAL Decompression (NEUROLYSIS)** of entire infratemporal facial nerve; – because presumed site of entrapment is meatal foramen (beginning of labyrinthine segment).

- alternate approach is to decompress this region; remainder of nerve can then be decompressed via transmastoid approach.

- irradiation of decompression may be guided by MRI.

N.B. neurolysis after 2 weeks (from paralysis onset) does not improve outcome!! (MRI may show contrast enhancement of nerve, but CT and X-rays are typically negative)

- N.B. neurolysis after 2 weeks (from paralysis onset) does not improve outcome!!

- according to newest studies surgical decompression of CN7 is not beneficial in Bell palsy!

**TREATMENT**:

- see also - **GENERAL MANAGEMENT (above)**

- **mainstay of treatment** - 10-day course (must be started early!) of **PREDNISONE** (60 mg/d for 5 days – tapered down by 10 mg/day for 5 days) – level A recommendation!

- **ACYCLOVIR** (400 mg ×5 /d) is not helpful; adding acyclovir is level C recommendation; – **ACYCLOVIR** alone is not effective in facial recovery!!!

- when recovery is partial, “contractures” may develop on paralyzed side (continuous diffuse contraction); H: botulinum toxin.

**Acute Paralysis**:

A) patient first seen on days 0-14:

- **ACYCLOVIR + PREDNISONE** – follow-up in 5 days:

a) parotidectomy – follow-up in 1 month

b) paralysis – paralysis protocol

B) patient first seen after 14 days – observation, follow-up in 6 months.

**Acute Paralysis**:

A) patient first seen on days 0-14:

- **ACYCLOVIR + PREDNISONE**; **ENG** on 3rd paralysis day and repeated every other day until 2 weeks have elapsed from onset of total paralysis:

a) < 90% degeneration – continue **ACYCLOVIR + PREDNISONE** until full course.

b) > 90% degeneration – surgical NEUROLYSIS.

c) 100% degeneration – voluntary **ENG** to confirm complete degeneration.

B) patient first seen after 14 days – **EMG** – follow-up in 5 months.

**RAMSAY-HUNT syndrome (s. geniculate zoster, herpes zoster oticus)**:

- herpes zoster of geniculate ganglion: see p. 256 (7.8) >>

1) **very painful** vesicular eruption on pinna, external auditory canal, pharynx

2) severe CN7 palsy

3) often CN8 is affected (vertigo, high-tone deafness)

**Treatment**:

- **VALACYCLOVIR**

**HEERFORDT syndrome (s. uveoparotid fever)**:

- form of sarcoidosis:

1) chronic parotid enlargement – CN7 palsy

2) uveitis

**FACIAL NERVE DISORDERS**

C7N (5)
MELKERSSON-ROSENTHAL syndrome

1) recurrent CN7 paralysis
2) recurrent and eventually permanent facial (particularly labial) edema [cheilitis granulomatosa]
3) fissured tongue

MOBIUS syndrome
- failure of motoneuron development in bilateral CN6 & 7 nuclei → CONGENITAL facial diplegia with abnormalities of horizontal gaze (convergent squint).
- most cases occur sporadically due to destructive (frequently vascular) lesions.
- 50% patients have limb anomalies (talipes, hypoplasia of digits, transverse retrograde defects, syndactyly).
- Möbius' syndrome is found more frequently among children with congenital heart malformations than in isolation.
- association with Poland anomaly may be familial (POLAND-MOBIUS syndrome).
- facial immobility can cause significant social handicap (immobile, dull facies may give incorrect impression of mental retardation).
- disabilities can initially be very significant (incl. feeding difficulties due to poor suck and speech impairments), but tend to improve with time.
- no treatment other than possible cosmetic surgery.

FACIAL OVERACTIVATION SYNDROMES

HEMIFACIAL SPASM
- involuntary, unilateral, painless, episodic tonic & clonic contraction of CN7 muscles.
- most often in middle-aged women (esp. Asian).
- begins as twitches around eye.
- progresses (during few months) to involve remaining ipsilateral facial muscles.
- even during sleep!
- stapedius contraction may produce tinnitus.
- muscles are normally relaxed between twitches.
- provoked by voluntary facial motion, emotional stress.
- idiopathic (generally, only cosmetic problem?)

a) compression of motor nerve root at brain stem by aberrant / ectopic vascular loop e.g.
- AICA loop in internal auditory canal (→ focal demyelination → ectopic excitation / ephaptic transmission → impulses conduct centrifugally to CN7 nucleus → nucleus hyperexcitability → bursts of hemifacial spasm).
- recovery from Bell's palsy.
- tumor
d) bony abnormalities.
- differentiates from blepharospasm (involuntary spasm of both orbicularis oculi muscles).
- diagnosis:
- MEG (vascular loop).
- EMG – 5-20 Hz bursts of muscle action potentials.
- treatment:
  1) BUTYLAMINOTONIN – treatment of choice!!!
  2) CARBAMAZEPINE, CLONAZEPAM
  3) nerve (or its branches) can be injected with alcohol or partially sectioned → relief from spasms until nerve regenerates.
  4) PERMANENT RELIEF:
     a) Janetta procedure - decompressing vessels at root entry zone – see p. 350
     b) anastomosing CN7 with CN11 or CN12.

MYOKYMIA
- progressive, irregular fibrillation of individual facial muscle fibers.
- begins in frontalis → extends to involve all ipsilateral facial muscles.
- EMG – spontaneous regular 30-70 Hz bursts of motor units.
- tinnitus (grave! vs. hemifacial spasm)
- sensations:
  a) Jannetta procedure - decompressing vessels at root entry zone – see p. 350
  b) anastomosing CN7 with CN11 or CN12.

FACIAL NERVE TRAUMA

CN7 injuries should be repaired surgically if they occur posterior to vertical line drawn from lateral canthus of eye.

- injuries anterior to this line do not need to be repaired (with microsurgical techniques, some branches can be rejoined).
- if nerve is cut by sharp object → repair primarily if wound site is clean.
- destructive injuries → nerve repair is usually secondary, following replacement of missing nerve tissue by graft.

RE-INNERRVATION SYNDROMES
a) incorrect postparalytic re-innervation (axons regrow to nonoriginal structures).
- spread of impulses between fibers within nerve (epiphaptic conduction).

POSTPARALYTIC SYNKINESIS
Synkinesis - unintentional muscle motion when another muscle is voluntarily contracted.

- example - jaw opening causes closure of eyelids (jaw-winking).
- mechanism - re-innervation of different muscles by axons from same motoneuron.

**CROCODILE TEARS**
- unilateral lacrimation during meals.
- mechanism - axons from superior salivatory nucleus (intended for chorda tympani → submandibular ganglion) enter greater superficial petrosal nerve (end up in pterygopalatine ganglion).

**GUSTATORY SWEATING (FREY syndrome)**
- facial sweating during meals.
- mechanism - cholinergic fibers originally intended for parotid gland end up in sweat glands.

**BIBLIOGRAPHY** for ch. “Cranial Neuropathies” — follow this [LINK >>](#)