

# NEUROLOGICAL DISEASES OF THE FACE

The work is supported by the **EFOP-3.4.3-16-2016-00021** „A Debreceni Egyetem fejlesztése a felsőfokú oktatás minőségének és hozzáférhetőségének együttes javítása érdekében” project. The project is co-financed by the European Union and the European Social Fund.



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# Classification

## ■ NEURITIS

- ## ■ NEURALGIAS
- **Typical neuralgias**
    - trigeminal neuralgia
    - glossopharyngeal neuralgia
  - **Atypical neuralgias**
    - pterigopalatine neuralgia
    - auriculotemporal neuralgia
    - posttraumatic trigeminal pain

- ## ■ NEUROPARALYSES
- **trigeminal**
  - **glossopharyngeal**
  - **hypoglossal**
  - **facial**



# Anatomy of the trigeminal nerve

The trigeminal nerve has three divisions

-  The ophthalmic nerve CN V1
-  The maxillary nerve CN V2
-  The mandibular nerve CN V3



# Trigeminal nerve

- Each division supplies not only the skin surface but the whole thickness of tissue from skin to mucous membrana
- Each of three divisions sends innervation to the dura mater
  - CN V1 to the tentorium cerebelli
  - CN V2 and V3 to the floor and side wall of the middle cranial fossa
- Each of the three divisions provides the sensory component to an autonomic ganglion



# Trigeminal nerve

- CN V1 to the **ciliary**
- CN V2 to the **pterygopalatine**
- CN V3 to the **submandibular and otic**






# Trigeminal nerve

In addition to its sensory component, the mandibular nerve is motor to four pairs of muscles.

- The two large elevators of the mandible-**temporal** and **masseteric**
- The two **pterygoid** muscle-**medial** and **lateral**
- The two **tensors**- **veli palatini** and **tympani**
- The two muscles of the floor of the mouth-**mylohyoid** and **anterior belly of the digastric**








# The ophthalmic nerve CN V1

-  **To the eyeball and cornea via the ciliary nerves, if paralyzed the ocular conjunctiva is insensible to touch**
-  **To the frontal, ethmoidal and sphenoidal sinuses via the supraorbital and ethmoidal nerves**
-  **To the skin and conjunctival surfaces of the upper eyelid and to the skin and mucous surfaces of the external nose**










# The maxillary nerve CN V2

-  To the upper teeth and gums
-  To the face, both surfaces of the lower lid, the skin of the side and vestibule of the nose and both surfaces of the upper lip
-  Via the pterygopalatine ganglion to the mucoperiosteum of the nasal cavity, palate and roof of the pharynx
-  To the maxillary, ethmoidal and sphenoidal sinuses; secretory fibers from the pterygopalatine ganglion pass with zygomatic nerve and then with the lacrimal nerve to the lacrimal gland
-  Foramen rotundum





# The mandibular nerve CN V3

-  Is motor to the four muscles of mastication but not the buccinator, the two tensors via the otic ganglion and the mylohyoid and the anterior belly of the digstric muscle
-  Sensory to the lower teeth and gums- inf. alv. nerve
-  Both surfaces of the lower lip by the mental nerve
-  The auricle and temporal region by the auriculotemporal nerve which also sends twigs to the external meatus and outer surface of the eardrum, and conveys secretory fibers from the otic ganglion to the parotid gland
-  The mucous membrane of the cheek by buccal nerve
-  The anterior two-thirds of the tounge, floor of the mouth and gums by the lingual nerve which also distributes the chorda tympani
-  Foramen ovale



# Trigeminal neuralgia

**Trigeminal neuralgia** is a disorder of the fifth cranial (trigeminal) nerve that causes episodes of intense, stabbing, electric shock pain in the areas of the face where the branches of the nerve are distributed. The disease is well known, but its cause is unknown.



# Trigeminal neuralgia




## Symptoms

- **Between the attacks there is a complete freedom of pain.**
- **It is main characteristics include a sudden onset of pain resembling an electric shock or a stroke of lightning which immediately of maximum strenght.**
- **Onset of symptoms occurs most often after age 50 in women**
- **The pain may be induced by external stimuli but also by stimuli such as speech, brushing teeth, putting on make up, or even a slight breeze**
- **During the time of attack the face contracts spasmodically and vegetative symptoms may appear: flushing of the face, watering of eyes, running of the nose**
- **The patients strives to avoid the induction of the painfull attack by not moving the facial muscles the face becomes un expressionless**



# Trigeminal neuralgia

## Symptoms

-  **The second and the third divisions together or separately are most often involved**
-  **The longer the disease has existed the more frequently the attacks occur and the more intolerable the pain.**
-  **It is called as a suicide disease as well, because it is universally considered to be the most painful affliction known to medical practice**



# Trigeminal neuralgia

## Diagnosis

- The attacks can most often be induced by contact with or pressure on the exit site of the supraorbital nerve, the infraorbital nerve and the mental nerve. The areas where the neuralgia can be induced are known as trigger zones.
- In every cases there is necessary above all to exclude any possible organic origin (most frequently inflammation of dental pulp) !!!
- Than we have to determine on which part of the face the neuralgia appers so which of the trigeminal branches are affected. Often it is very difficult.



# Trigeminal neuralgia Treatment

## Medical treatment

- **Carbamazepine 100-200 mg/day**
- **Gabapentin 300 mg/day**
- **Baclofen 5-10 mg up to 60 mg/day**



# Trigeminal neuralgia Treatment

## Surgical

### Percutaneous procedures

- less risk
- local or brief general anesthesia
- a needle or trocar is inserted on th cheek just lateral to the corner of the mouth under fluoroscopic guidance, introduced into ipsilateral foramen ovale
- gangliolysis is performed
- different type of procedure:
- percutaneous radiofrequency trigeminal gangliolysis PRTG
- percutaneous retrograserian glycerol rhizotomy PRGR
- percutaneous balloon microcompression PMB



# Trigeminal neuralgia Treatment

## Surgical

### Microvascular decompression

- requires general anesthesia
- 2,5-3 cm cranyectomy is performed, the dura is opened and the cerebellum is microsurgically retracted
- typically, an artery or other vascular cross-compression of the nerve is indentified, the vascular structure is padded away from the nerve with teflon felt
- this operation has a low mortality rate 0,1-0,5 %
- serious morbidity probably between 1-5%, numbness, hearing loss, dizziness, cerebellar syndrome, meningitis, diplopia





# Anatomy of the glossopharyngeal nerve

- It is motor to the stylopharyngeus muscle
- Its parasympathetic component supplies secretory fibers through the otic ganglion to the parotid gland
- It provides the special sense of taste to the posterior one-third of the tongue including the vallate papillae
- General sensory fibers supply almost the entire one-half of the pharyngeal wall, including the oropharyngeal isthmus
- It also supplies the dorsum of the soft palate, the auditory tube, tympanum, medial surface of the eardrum, mastoid antrum and mastoid cells.



# Glossopharyngeal neuralgia

- **Glossopharyngeal neuralgia closely resembles trigeminal neuralgia except for the location of the pain which follows the distribution of the ninth cranial nerve.**
- **This typical neuralgia has been reported to occur 40-70 times less frequently than trigeminal neuralgia.**



# Glossopharyngeal neuralgia


## Symptoms

- **Lightning-like attacks of pain lasting for a period of several seconds to several minutes are characteristic.**
- **The pain radiates from the lateral wall of the pharynx to the side of the neck and ear and may be induced by swallowing, yawning, extension of the tongue or speaking.**
- **In these patients the speech is very difficult to understand.**
- **Xerostomia, a dry cough or unilateral disturbance of taste sensation may develop.**





# Glossopharyngeal neuralgia

## Diagnosis

-  The trigger zones are primarily to be found at the site of appearance of the pain, in the regions of the tonsil, the posterior third of the tongue and the hypopharynx.




# Glossopharyngeal neuralgia Treatment

-  **Medical treatment** of this disease is the same as in the previous case.
-  **We can mention perineural Lidocaine infiltration of the glossopharyngeal nerve.**



# Atypical neuralgias

-  **Atypical neuralgia involve the following criteria**
- **the periods of pain are accompanied by vegetative phenomena**
  - **the neuralgia is constant in nature and not attack-like**
  - **the borders of the painful zone are not defined by the courses of the individual cranial nerves**
  - **there are no trigger zones**





# Atypical neuralgias

- **Extremely difficult to define this type of neuralgias**
- **Hard to diagnose.**
- **The treatment of is in part the same that for typical neuralgia but it is more difficult. The same surgical interventions may be performed but they should be considered even more carefully.**



# Pterygopalatine neuralgia

-  The syndrome was described by Sluder in 1908, the pain can extend to the root of the nose, the eye, the maxilla, the upper teeth, the zygomatic bone, the ear and possibly the mandible and even the nape of the neck, the neck itself and the shoulder.
-  This neuralgia is unilateral, the origin is unknown, and it has indefinite border on the central and possibly the lower part of the face





# Auriculotemporal neuralgia

- This clinical entity was described by Frey in 1923, it can develop as a consequence of an injury to or inflammation of this nerve.
- The essence of this disease not the pain itself but the typical combination of vegetative symptoms: the part of the face in front of the ear becomes red, while on eating or on the secretion of saliva perspiration begins on the skin (above the parotid in front of the ear)
- It may occur 10-15% of patients after parotid surgery
- Starch Iodine Test for Frey's Syndrome





# Posttraumatic trigeminal pain

- Also known as a trigeminal neuropathy, is a diffuse, burning pain causing a sensation of pressure which extends to one or more branches of the trigeminal nerve, it is not triggered by stimuli. It may last for hours, at an almost intolerable level.
- In the patient's history there is some external influence (trauma) in the region or along the course of the nerve.
- Direct irritation of the nerve is involved in this entity
- In posttraumatic trigeminal neuralgia this primary cause does not feature, even if it does exist the pain continues after the cause eliminated., for a permanent lesion has developed in the nerve tissue.
- Its treatment is very difficult, the principles are same that in trigeminal neuralgia, but it is more difficult to attain freedom from pain and this condition lasts for a shorter time.



# Neuroparalyses

-  If the neuroparalyses occur on the trigeminal or on the glossopharyngeal nerve these have the common symptoms of paresthesia, due to the paralysis of the sensory fibers , or a total loss of sensation.
-  In consequence of the paralysis of the motor fibers the affected muscle is partially or totally paralysed.



# Neuroparalyses

## Anatomy of the facial nerve

- **Motor** to the „muscles of expression” the superficial muscles around the eye, nose, mouth and ear; of the the scalp superiorly and the platysma inferiorly.
- It also supplies the stylohyoid and posterior belly of the digastric muscles, as well as the stapedius
- **Special sense** taste fibers with cell stations in the geniculate ganglion



# Neuroparalyses

## Anatomy of the facial nerve

### ■ Parasympathetic

- **Secretory via the greater petrosal nerve and the nerve of the pterygoid canal to the pterygopalatine ganglion, then relayed by postsynaptic fibers to the glands of the nose and palate and to the lacrimal gland**
- **via the chorda tympani to the submandibular ganglion from which postsynaptic fibers are relayed to the submandibular and sublingual salivary glands and via its connection with the otic ganglion it innervates the parotid gland**






# Neuroparalyses

## Anatomy of facial nerve

-  **Sensory** Supplies general sensation to a small area of the external acoustic meatus and the auricle



# Paralysis of the facial nerve

-  **The movement of the human face is both simple and complex. This simplicity lies in the movement of mimic muscles under the control of facial nerve.**
-  **The complexity lies in the richness and coordination of these movements. Beside the natural spontaneous movements humans have learnt how to influence their facial expression to reflect to their mood and frame of mind, how to control these expressions.**
-  **So it is understandable that one of the most unpleasant distortions is a paralysed face. It is not only deprives the human ability to express sadness, happiness, anger but also distorts the direct reflection of human emotion.**



# Facial paresis

- Injuries can occur in various sites in the facial nerve system.
- The facial paresis could be **central** or **peripheral**.
- Muscles around the eye and the wings of the nose receive innervation from the cortex both sides, whereas the muscles around the mouth are connected only to the contralateral cortical centre.
- In **peripheral facial paresis** all of the muscles relating to this nerve are paralysed. Lagophthalmus, inability to furrow the forehead, and loss of the angle of the mouth on the same side is developed.
- In **central facial paresis** paralysis can be observed only in the muscles around the mouth, the angle of the mouth on one side is lost, the nasolabial fold is smoother.





# Facial paresis

**Examination of these frequently allows an exact determination of the site of the damage to the nerve.**

- **If the lesion occurs after the stylomastoid foramen, towards the parotid, but still before the splitting into the individual branches then the whole of the mimic musculature is paralysed but without any disturbance of taste sensation, lacrimation, salivation or hearing**



# Facial paresis

- **If the retroauricular nerve is damaged hypaesthesia of the auricle may be observed**
- **If the lesion takes place in the facial canal before the outlet of the corda tympany but after the outlet of stapedius then the previous symptoms will be accompanied by disturbances of taste sensation and salivation**
- **If the conductance disturbance is situated even higher, the lesion of the major superficial petrosal nerve will mean that lacrimatory disturbances also arise as an additional symptoms.**



# Facial paresis

## Cause of the facial paresis

- Trauma
- During operation on the face
- During intervention of the parotid gland
- Malignant tumors



# Hypoglossal nerve palsy

- Hypoglossal nerve palsy is a rare condition, most commonly associated with impairment of other cranial nerves (IX-XI). The hypoglossal nerve is a motor nerve it innervates the muscle of the tongue.
- M.genioglossus
- M.hyoglossus
- M.styloglossus
- M.longitudinalis superior et inferior
- M.transversus
- M.verticalis
- It innervates also the infrahyoid muscles.
- M.sternohyoideus
- M.sternothyroideus
- M.thyrohyoideus
- M.omohyoideus
- M.geniohyoideus



# Anatomy

- **The nerve is involved in controlling tongue movements required for speech, food manipulation (i.e. formation of bolus), and swallowing.**
- **The hypoglossal nerve arises from the paired hypoglossal nucleus of the caudal brain stem emerging from the ventromedial medulla oblongata from a number of smaller rootlets.**
- **After passing through the subarachnoid space the nerve then exits the skull-base of the posterior fossa through the hypoglossal canal**



# Anatomy

- **Following in near proximity to the vagus and spinal division of the accessory nerve, it spirals behind the vagus nerve and passes between the internal carotid artery and internal jugular vein lying on the carotid sheath**
- **After passing deep to the posterior belly of the digastric muscle, it passes to the submandibular region, passes lateral to the hyoglossus muscle, and inferior to the lingual nerve to reach and efferently innervate the tongue**



# Aetiology

- Benign or malignant tumors
- Trauma
- Vascular lesions
- Infection
- Autoimmun diseases
- **Iatrogenic** (operations of neck, shoulders, cervical spine, carotid endarterectomy, general anaesthesia, radiation th, central venous catheter)
- Idiopathic



# Symptoms

## Unilateral

- Atrophy of half of the tongue, fasciculation
- Deviation to the affected side
- Dysphagia
- Dysarthria
- Minor forms may be unnoticeable
- Infrahyoid muscles could be affected

## Bilateral

- Hardly tolerable condition
- Difficulty in swallowing
- Disability in speech
- Bilateral atrophy, fasciculation
- Difficulty in breathing, airway obstruction
- Accumulation of saliva





# Treatment

- **No therapeutic guidelines**
- **Causal therapy**
- **Antibiotics**
- **NSAID**
- **Vitamin (B12, B vitamin complex)**
- **Systemic steroid**
- **Fluid replacement, nasogastric tube**
- **Tracheotomy if it necessary**
- **Speech- and swallowing therapy**



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