Diseases of the Cranial Nerves

Diseases of the cranial nerves often lead to severe burdens on patients and to diverse functional disorders and neurological deficits. Due to the diverse functions of the cranial nerves, the exact topographic localization of the clinical symptoms is very important. Diseases of the cranial nerves are a crucial part of every medical exam. In the following article you will get an overview and a short summary of the most important diseases of all twelve cranial nerves. You can learn how to correctly examine pairs of cranial nerves here.

Peripheral and Central Facial Nerve Palsy

Facial nerve palsy is paresis of the muscles supplied by the facial nerve (VII) on one side of the face due to a lesion of the facial nerve. The paresis generally only occurs on one side, but it may also occur on both sides. Usually is temporary. Initially, the patient suffers from non-specific dragging pain in the region of the ear before the paresis develops over the course of a number of hours or days.

Occurs in 10 to 35 cases per 100,000 habitants. During pregnancy the prevalence increase three time, especially in the first trimester. This condition has a universal distribution, has no predilection for ethnicity or age.
Possible causes for facial nerve paresis:

- Viral and bacterial infections
- Stroke/Ischemic lesions
- Basal skull fracture
- Tumors in the petrous bone or the parotid gland
- Intracranial injury
- Toxic causes
- Idiopathic causes
- Chromosomal damage

Depending on the location of the lesion, one distinguishes between central and peripheral facial nerve palsy.

In both, rehabilitation must be start as zoom as possible to avoid complications.
Peripheral facial nerve paresis involves lesions of the second motor neuron, the anterior motor horn, the peripheral nerves, or the muscles outside the central nervous system. Peripheral facial nerve palsy is characterized by a weakened myotatic reflex, negative pyramidal tract signs, and a slack tone and atrophy of the affected muscles.

Idiopathic facial nerve palsy (Bell's palsy) is the most frequent peripheral cranial nerve lesion, and it is accompanied by a single-sided and acute occurrence of peripheral facial nerve palsy. This disease can occur at any age, often between the ages of 10-20 and 30-40 years. Women seem to be affected more frequently than men.

Bell's palsy heals in approx. 70% of cases without any consequences, but persistent defects after re-innervation may remain.

As a consequence of one-sided peripheral facial palsy, the following actions are no longer possible:

- Frowning
- Raising eyebrows
- Closing the eyes
- Puffing out the cheeks
- Whistling
- Showing the teeth

These symptoms also suggest the failure of the nerve, and weakness or complete paresis of the mimic muscles may result. Symptoms include:

- Bell’s phenomenon (incomplete closure of the eyelid)
- Upward rotation of the eyeball becomes visible
- Drooping of the labial angle and the lower eyelid
- Elapsed nasolabial fold
- Slackened platysma

Disorders of lacrimation, headache, gustatory disturbances in the anterior third of the tongue, and ear pain and increased hearing sensation are other accompanying symptoms.

Causes of Bell’s palsy are:

1. Herpes zoster infections
2. Otitis media
3. Guillain-Barré syndrome
4. HIV infection tumor
5. Ischemic stroke
6. Autoimmune disease
7. Lyme disease
8. Among others.

Central facial palsy

Central facial palsy is due to a lesion of the first motor neuron in the region of the brain or its descending projections to the spinal cord.

Central facial palsy involves increased myotatic reflexes, weakened multisynaptic
reflexes, and positive pyramidal tract signs; a cramp-like increase in tone of the affected muscles also occurs, without any relevant atrophy. **Frequently, central facial palsies are caused by cerebral circulatory impairments or brain tumors.**

In contrast to peripheral facial palsy, a patient with central facial palsy can frown and close his eyelid since the peripheral nuclear areas (facial nucleus) lead to the fibers of the facial nerve and are interconnected to finally reach the forehead and the eye, which also receive fibers from the other side.

The musculature is no longer mobile – especially in the area of the mouth – and it is flaccid, as with peripheral facial palsy. Also, the labial angle droops, is immobile, and is partially open on the affected side.

*Image: “Man suffering from right facial palsy” by St. Bartholomew’s Hospital Society. License: CC BY 4.0*

**Note:** A distinguishing feature of peripheral and central facial palsy: with central facial palsy, the patient can frown and close his eyelid.

**Trigeminal Neuralgia**

Trigeminal neuralgia occurs in the innervation area of the trigeminal nerve in the form of a severe, acute and recurrent attack-like facial pain that is generally single-sided. **Three forms** are distinguished:

- **Classic trigeminal neuralgia:** In patients with compression of the trigeminal nerve by a presumed or demonstrated vascular loop.
- **Secondary trigeminal neuralgia:** Associated to another disease such as multiple sclerosis and tumors.
- **Idiopathic trigeminal neuralgia:** When the cause is unknown.

Others categories include several causes of facial pain such as painful trigeminal neuropathy due to herpes zoster virus, post-traumatic trigeminal neuropathy, painful trigeminal neuropathy attributed to other disorder and idiopathic painful trigeminal neuropathy.

**Classic trigeminal neuralgia**
Previously called tic doloureux. Principal cause of this pain is the compression or mechanically irritation of the trigeminal nerve by blood vessel loops at the point where it exits the brainstem. The average age of onset is between 50 and 79 years.

**Secondary trigeminal neuralgia**

This form of trigeminal neuralgia accompanies demyelination diseases such as multiple sclerosis, occurs as a consequence of either tumors or Costen’s syndrome (a facial pain which originates from the facial muscles due to malfunction of the mandibular joint), for example. Inflammatory processes and, in rare cases, medical interventions can result in trigeminal neuralgia.

Symptomatic trigeminal neuralgia may be accompanied by hypoesthesia, i.e. reduced sensitivity towards tactile stimuli, in the form of numbness or tingling, in the region of the first trigeminal branch. Furthermore, the corneal reflex is weakened.

Patients with symptomatic trigeminal neuralgia are, on average, younger than patients with the classical form of the disease. Double-sided facial pain often also occurs in such patients. The goal of therapy is to treat the underlying cause.

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**Symptoms of trigeminal neuralgia:**

- Attacks of shooting, severe pain, which occur repeatedly, up to 100 times a day.
- Mostly, the supply area of the second branch of the trigeminal nerve is affected.
- It can be triggered by touch, coldness, speaking, swallowing, chewing, combing hair, touching or washing the face, spicy food, vibration at walking etc.
- Due to the pain, patients are sad, powerless, anorexic, they sleep badly, and they feel weak.

The patients try to avoid the trigger by reducing their mimic movement, not speaking, and not eating. Often, only fluid foods are taken in with a straw. In the cold seasons, most patients protect themselves with a scarf from coldness and wind.
Eye Muscle Paresis

Oculomotor Nerve Palsy

The oculomotor nerve (the eye movement nerve) innervates several eye muscles and – along with the trochlear nerve (IV) and the abducens nerve (VI) – is responsible for the movement of the eyeball.

**Roughly one third of all eye muscle pareses are caused by oculomotor nerve palsy**, which is overall slightly rarer than abducens nerve palsy. In 60-70 % of cases, oculomotor nerve palsy occurs as an isolated loss.

Lesions of this nerve can result in various types of paresis: **Complete (inner and outer) oculomotor nerve palsy**

Complete loss of the function of the nerve leads to the **following clinical picture:**

- Ptosis (drooping of the upper eyelid)
- The eyeball deviates to the outside and downwards
- Widened pupil (mydriasis) and pupils unresponsive to light (totally unresponsive pupil)
- Diplopia (double vision)

In cases of complete oculomotor nerve palsy, **the consensual reaction of the opposite eye remains**, i.e. a reflex-triggered concordant reaction occurs on the opposite side of the body. Opening of the eyelid may be possible by means of contraction of the frontal muscle, since double vision only occurs following elevation of the eyelid.
Causes of Oculomotor Nerve Palsy are:

1. Tumor
2. Stroke
3. Infections of the central nervous system (meningitis, encephalitis)
4. Aneurysm
5. Local lesion in base of the eye.

Ophthalmoplegia interna

Ophthalmoplegia interna involves completely unresponsive pupils accompanied by free movement of the eyeball; the pupil does not react to either direct or indirect light nor to convergence. As a result, the patient does not have clear vision in the affected eye when looking at close objects. There is also paresis of accommodation.

Ophthalmoplegia externa

In cases of ophthalmoplegia externa, however, the motility of the eyeball is impaired, yet autonomous innervation of the pupil and the ciliary muscle is intact. If pupil function has been preserved, complete paresis of all the muscles innervated by the oculomotor nerve is quite rare.

Anisocoria

Another disease or deficit of the oculomotor nerve would be pupils of unequal width, which is referred to as anisocoria. Anisocoria is present in Claude-Bernard-Horner syndrome, which is characterized by constriction of the pupil (miosis), a drooping eyelid (ptosis), anhidrosis (decrease sweating) and a posterior displacement of the eyeball (enophthalmus).

Anisocoria can also occur alongside intracranial pressure involving compression of the oculomotor nerve.

Trochlear Nerve Palsy

Trochlear nerve palsies are rarer than oculomotor or abducens nerve palsies. The most frequent cause of monosymptomatic trochlear nerve palsy is a traumatic brain injury.

Trochlear nerve palsy is characterized by isolated paresis of the superior oblique muscle. The function of this muscle is to depress the eyeball. In cases of paresis, the symptom increases during adduction and is virtually absent during abduction, i.e. the eye...
of the patient faces towards the nose and upwards, and the patient experiences double vision (diplopia), just as in cases of oculomotor nerve palsy.

Causes of trochlear nerve palsy are tumor, demyelination, meningitis and other.

A distinction is made between double and single-sided trochlear nerve palsy.

**Double-sided trochlear nerve palsy**

In cases of double-sided trochlear nerve palsy, the Bielschowsky phenomenon is often positive on both sides. In this case, the diseased eye stands higher, is rotated outwards to the temple and has a squint deviation to the nose, which creates oblique double vision. To compensate, the patient tries to rotate and lower the chin, and to tilt the head to the healthy side.

Compensatory head turning and tilting is usually not present, in contrast to one-sided trochlear nerve palsy.

**One-sided trochlear nerve palsy**

One-sided trochlear nerve palsy is accompanied by a compensatory head posture with turning and tilting towards the healthy shoulder and lowering of the chin. The affected eye is in an abduction position and is rotated outwards. An annoying pathological rolling image is avoided since the slackened internal rotator is not utilized this way.

**Abduccens Nerve Palsy**

The abduccens nerve palsy is characterized by an isolated paresis of the rectus lateralis muscle (an externally turning muscle), which often occurs without identifiable
intracranial lesions. In a high percentage of cases, the cause of these palsies remains idiopathic. However, trauma, a diabetic metabolic state, and increased intracranial pressure due to tumor or meningitis are some causes of abducens nerve palsy.

In cases of abducens nerve palsy, **convergent paralytic strabismus occurs even in the primary position**, i.e. the affected eye deviates towards the inside and the paralyzed eye is impaired or inhibited if it tries to turn to look to the side or to look up.

Furthermore, a slight adduction position can occur when looking up or down. Undisturbed binocular movement is, however, observed when looking to the healthy side.

**Horizontally parallel double images (i.e. double vision) are usually perceived even in the primary position.** The deviation of the images of objects on the retina increases on the paralyzed side, but when the affected eye is covered, the image corresponding to the respective side disappears.

However, the annoying phenomenon of double vision causes a compensatory head posture, which leads to the head being turned towards the side of the paralyzed muscle – a position that does not require the rectus lateralis muscle.

**Lesions of the Visual Pathway**

Each optic tract consists of ‘two half, former’ optical nerves. They conduct the lateral part of the visual information coming from the same side and also the medial sensations of the opposite side.
The visual pathway crosses the whole brain. **Very differing deficits in the visual field may arise depending on the location of the lesion.** Therefore, when there is a lesion in the visual pathway, the visual field is examined, and pupil reaction and the appearance of the papilla are examined.

The following deficits in the visual field can occur as a result of nerve lesions:

- If the optic nerve is severely damaged on one side, the patient is **blind on the affected eye**, and their sight is not impaired on the other side.
- When there is a lesion of the medial part of the optic chiasm, it is mainly the fibers which cross to the other side that are damaged. The patient can suffer from a bitemporal (heteronymous) hemianopsia, i.e. the patient does not get any visual information relating to what happens in his lateral field of vision. This is referred to as hemianopsia, or 'blinker vision.' The lateral section, which does not cross and leads to the optic tract, remains intact, meaning sight in this part of the visual field is not impaired.
- Visual loss can also affect the optic tract. For example, the patient may have a lesion in the right optic tract which **leads to a deficit in the left half of the visual field of both eyes**. This failure is referred to as bilateral homonymous hemianopsia (left). A lesion of the left optic tract would lead to a corresponding opposite deficit in the right half of the visual field. Depending on where the optic tract is damaged, this may lead to a complete or an incomplete deficit. Once such deficits have arisen, they do not usually disappear.

Further lesions can arise in the realm of visual radiation, and such lesions have diverse consequences since visual radiation spreads a fan-like manner. Tumors and strokes are the most frequent triggers for this visual disorder.

**Deficits in the visual field can be observed in the following form:**

- Upper part of the visual radiation (right) – homonymous hemianopsia (quadrant anopsia upper left)
- Lower part of the visual radiation (right) – homonymous hemianopsia (quadrant anopsia upper left)
- Lower part of the visual radiation (right) – homonymous hemianopsia (quadrant anopsia lower left)

*[Image: "Homonyme Hemianopsie" by Nunh-huh. License: CC BY-SA 3.0]*
Deficits in the upper part of the visual radiation are more severe than in the lower part. Deficits in the area of the visual radiation likewise do not regress either.

**Amaurosis**

This term refers to a vision complete loss without apparent lesion in eye, can be occur in one or two eyes. Amaurosis can be congenital (Leber’s congenital amaurosis) or secondary. In case of optic nerve is produce to the injury or compression of nerve by a tumor (adenohypofisis), trauma or stroke over the optic chiasm.

**Summary of the Important Diseases of All Twelve Pairs of Cranial Nerves**

**Olfactory nerve (I)**
- Anosmia (inability to smell)
- Hyposmia (weakened ability to smell)

**Optical nerve (II)**
- Anopsia or amaurosis (blindness in one or both eyes)
- Hemianopsia
- Quadrant anopsia
- Blinker-phenomenon
- Papilledema

**Oculomotor nerve (III)**
- Anisocoria (unequally wide pupils)
- Miosis (narrow pupils)
- Mydriasis (wide pupils)
- Gaze palsy
- Diplopia (double vision)
- Ptosis (drooping upper eyelid)

**Trochlear nerve (IV)**
- Strabismus
- Diplopia

**Trigeminal nerve (V)**
- Trigeminal neuralgia / tic douloureux
- Paresis of the muscles of mastication
- Loss of the sensation of touch and temperature

**Abducens nerve (VI)**
- Diplopia

**Facial nerve (VII)**
- Bell’s palsy (paralysis of the fascial muscles)
- Hyperacusis (sounds are perceived too loud)
- Loss of gustatory sensation in the anterior tongue
- Burning eye sensation due to dehydration of the conjunctiva/cornea
Vestibulocochlear nerve (VIII)
- Hypacusis (hearing loss)
- Deafness
- Tinnitus (permanent aural noises)
- Ataxia (instability with respect to movement)
- Rotatory vertigo
- Nystagmus (eye twitching)

Glossopharyngeal nerve (IX)
- Difficulty swallowing
- Diminished salivation
- Loss of gustatory sensation in the posterior part of the tongue
- Loss of sensation in the throat

Vagus nerve (X)
- Hoarseness
- Difficulties with swallowing and at phonation
- Posticus paralysis (severe respiratory distress when a particular muscle of the larynx fails)
- Changes in heart rate (quicker or slower)
- Less gastric acid and intestinal peristalsis

Accessory nerve (XI)
- Inability to lift the shoulder
- Weakness in turning the head

Hypoglossal nerve (XII)
- Speech disorders
- Difficulty swallowing

Review Questions
The answers can be found below the references.

1. Which fascial appearance is observable in cases of ocular nerve palsy that are accompanied by dysfunction?
   A. Ptosis, miosis, slightly unreactive pupils, diplopia, and an eyeball that deviates outwards and downwards
   B. Ptosis miosis, totally unreactive pupils, diplopia, and the Bielschowsky phenomenon
   C. Ptosis, mydriasis, totally unreactive pupils, diplopia, and an eyeball that deviates outwards and downwards
   D. Ptosis, mydriasis, the Bielschowsky phenomenon, diplopia, and an eyeball that deviates outwards and downwards
   E. Ptosis, miosis, totally unreactive pupils, diplopia, and strabismus

2. Which feature correctly distinguishes between peripheral and central facial nerve palsy?
   A. With peripheral facial nerve palsy, the patient is able to both frown and close their
   B. With central facial nerve palsy, the patient is unable to frown, but is able to close
their eyelid.
C. With central facial nerve palsy, the patient is unable to frown or close their eyelid.
D. With peripheral facial nerve palsy, the patient is able to frown, but unable to close their eyelid.
E. With central facial nerve palsy, the patient is able to both frown and close their eyelid.

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Correct answers: 1C, 2E, 3B

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