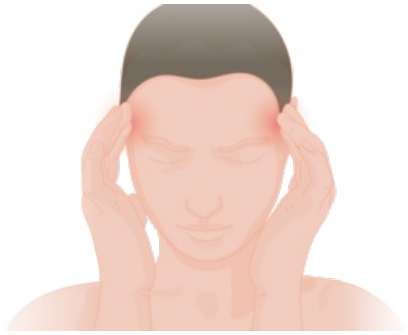
Cluster headache/PGS

Introduction to headache

Headache (cephalalgia) is a symptom that could involve sudden and intense pain, which could signal a serious disease threatening the patient's life.

Headaches are divided into:

- primary (pain is one of the manifestations of non-life-threatening, chronic, paroxysmal disease),
- secondary (arises as a symptom of another disease).



Headache

The most important task in examining a patient with a headache is to **identify a serious secondary headache that can be life-threatening.** The following are always alarming:

- first headache in a patient over 40 years of age,
- sudden and intense headache,
- progressively developing atypical headache that is unresponsive to standard treatment,
- headache in a patient with cancer or HIV infection,
- the presence of any focal neurological finding or impairment of consciousness.

As a secondary symptom, headaches are accompanied by the following serious diseases and conditions:

- subarachnoid hemorrhage
- meningitis and meningoencephalitis
- intracranial expansion
- strokes, especially hemorrhagic strokes
- carotid artery dissection
- cerebral venous thrombosis
- cranial neuralgia
  - trigeminal neuralgia
- sinusitis
- glaucoma attack
- hypertension
- temporal arteritis
- head injury
- stp. regime error, including alcohol excess
- sleep apnea syndrome (SAS)
- cervicocranial syndrome

The reason for a visit to the doctor is most often a sudden and/or severe headache.

The primary headaches are in particular:

- migraine
- tension headache
- **cluster headache**
- headache caused by physical and sexual activity.

Conducting careful medical history and neurological examination are most important in the differential diagnosis of headaches. We determine the **nature of pain** (dull, pulsating, whipping), the **location of pain** (hemicrania, diffuse, behind the eye), its **intensity**, **duration** (seconds, hours, days, daily pain), **frequency of pain** (several times a day, monthly), **other accompanying symptoms** (phonophobia, photophobia, odorophobia, lacrimation, nasal secretion, cerebral nerve palsy, hemiparesis, cerebellar symptoms, impaired consciousness) and **triggering factors** (physical activity, defecation, head tilt, stress, menstruation, drugs, alcohol). We also ask about the current treatment and its effects.

Past medical history

The diseases that the patient have along with the headaches are investigated (such as circulatory system disorders, kidney disease, general infections, cancer, ENT affections, eye diseases, dental and jaw affections).

Diagnosis

Primary headaches

Primary headaches except from the migraines have a normal neurological finding.

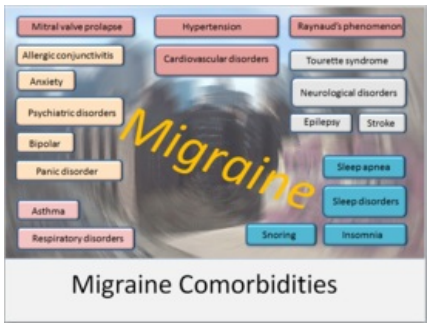
Migraine

The pain may be affecting one side or both sides of the head with moderate to high intensity, accompanied by nausea or vomiting, photophobia and phonophobia. The pain lasts 4–72 hours, a longer duration is called migraine status. Diagnostic criteria is as shown below:

Diagnostic criteria for migraine without aura

A. At least 5 attacks must meet point B-D
B. The headache lasts 4-72 hours without treatment
C. Pain can be characterized by at least two of the following symptoms: ▪ unilateral localization ▪ pulsating character ▪ medium to severe intensity ▪ aggravation during physical exertion
D. The pain is accompanied by at least one of the following symptoms: ▪ nausea and/or vomiting ▪ photophobia and/or phonophobia

In 20% of patients, the pain precedes up to 1 hour of the **aura** phase, which is any focal symptom that usually disappears with the onset of pain. The most common is the visual aura - scintillating patterns or negative phenomena of the nature of scotomas or hemianopsia. Furthermore, the aura can be sensitive in the form of paresthesias of the face or limbs. Rarely, the aura appears in the form of an incomplete expressive fatal disorder or paresis.



Migraine comorbidities

The diagnosis is determined on the basis of a typical clinical picture.

A **differential diagnosis** especially during the first migraine attack is **subarachnoid hemorrhage** - CT scan of the brain is performed and cerebrospinal fluid is examined.

The diagnosis is also confirmed by an evident therapeutic effect of 5-HT-1B/D receptor agonists - triptans:

- sumatriptan (Sumigra, Rosemig, Imigran, Cinie): 50-100 mg per seizure
- zolmitriptan (Zomig): 2.5-5 mg per seizure
- eletriptan (Relpax): 40-80 mg per seizure
- naratriptan (Naramig): 2.5-5 mg per seizure
- frovatriptan (Fromen): 2.5 mg per seizure

Tension headaches

Tension headaches are dull or astringent, mostly bilateral, mild to moderate in intensity, and are not accompanied by significant accompanying symptoms (vomiting, visual disturbances, focal neurological symptoms). The main diagnostic criteria are shown in the table:

Criteria for tension headaches

A. The patient must meet the criteria in at least two points:
B. Compressive or astringent pain
C. Pain of bilateral localization
D. Mild or moderate pain
E. Pain does not worsen with physical activity, is not accompanied by nausea or vomiting; mild photophobia or phonophobia may occur

According to the frequency of occurrence, tension pain is divided into **episodic** (less than 15 days per month) and **chronic** (more than 15 days per month).

Tension headaches can also occur in individuals with other primary or secondary headaches. In some individuals, tension pain is associated with increased pericranial and neck muscle tone.

The basis of successful therapy of tension headaches is a thorough psychoeducation (identification of adverse factors, adverse effects of mental imbalance, the art of physical and mental relaxation, life priorities).

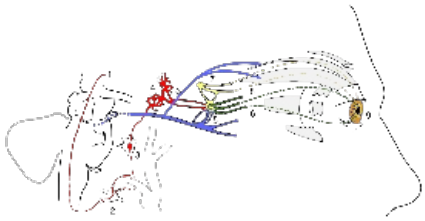
In episodic tension headaches, *acetylsalicylic acid derivatives (Aspegic plv 500 mg) and peripheral analgesics* - paracetamol (Paralen 500 mg) are the drugs of choice. Another group of medication is *non-steroidal anti-inflammatory drugs*: ibuprofen (Ibalgin, Dolgit) in a dose of 400–800 mg, indomethacin (Indomethacin supp) in a

dose of 50–100 mg, naproxen (Naproxen tbl, supp) in a dose of 500 mg and diclofenac (Diclofenac tbl) in a dose 50-100 mg.

For chronic tension pain, the drug of first choice is *amitriptyllin* at a dose of 25-50 mg per night while drugs from the SSRI group (such as citalopram, fluoxetine) tend to be less effective.

Cluster headache

The pain is whipped with very strong intensity. The patient may find that lying down worsens the pain and has to pace in order to alleviate the pain. The symptoms often appear at night: the pain is usually located temporally or periorbital, unilaterally and is accompanied by lacrimation, nasal secretions and Horner's syndrome. Symptoms of cluster headaches tend to manifest within a few days of the year, often in the spring and fall seasons. The diagnostic criteria are shown in the table:



Horner's syndrome

Cluster headache diagnostic criteria

A. At least 5 attacks meeting criteria B-D
B. Intense unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes
C. At least one of the following pain symptoms: ▪ conjunctival congestion ▪ lacrimation ▪ rhinorrhea ▪ forehead or facial hyperhydrosis ▪ miosis ▪ ptosis ▪ eyelid edema ▪ nasal congestion
D. Attack frequency 1-8 times daily
E. Medical history, neurological examination, or ancillary examination methods do not indicate secondary headache, or secondary headache is present, but the cluster headache did not occur for the first time in connection with this disease

- *Oxygen inhalation, sumatriptan, and in severe cases corticoids* are used therapeutically
- The **differential diagnosis** include **dissection of the carotid artery** and **rupture of aneurysm** - MRI, angiography and ultrasound could be performed

Headaches during sexual and physical activity

Pain occurs in connection with physical activity (for instance exercise) or during or just before orgasm, more often in men. It is typically very strong, localized occipitally or frontally and lasts several minutes to hours. This pain can be partially prevented by improving physical condition and reducing weight.

Conventional analgesics are used in the treatment.

A **differential diagnosis** especially for the first time occurance is **subarachnoid hemorrhage** - CT scan of the brain is performed and cerebrospinal fluid is examined.

Secondary headaches

Headache is often associated with other neurological symptoms.

Subarachnoid hemorrhage

Bleeding occurs during the rupture of an aneurysm, most often in the area of Circle of Willis especially on the anterior or posterior arteries. This often happens when blood pressure increases (such as during physical exertion, sexual intercourse, agitation, defecation, etc.).

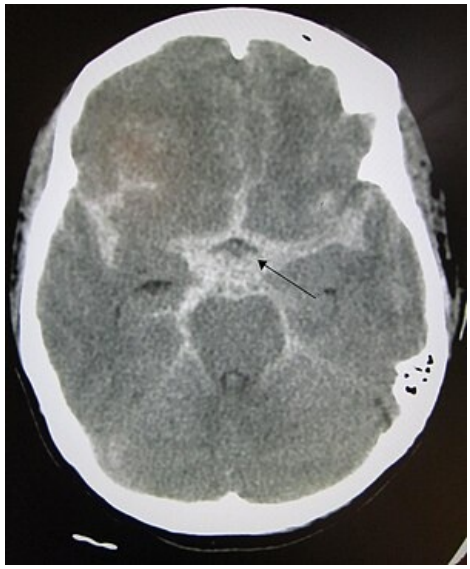
The headache appears within seconds and can be very intense, located on both sides. Initially, there may be a brief disturbance of consciousness. The pain is further accompanied by nausea, vomiting, photophobia and phonophobia.

Meningeal syndrome develops within minutes to hours. Patients are often disoriented and confused. Some patients are somnolent, sometimes with psychomotor restlessness, aggression and negativism.

The condition of the patient's subarachnoid hemorrhage is assessed based on the **Hunt and Hess grading system**. In some cases, the symptoms may not be overtly expressed and *mimic cervicocranial syndrome*, hence, in unclear cases CT examination (and cerebrospinal fluid examination) are especially necessary.

The diagnosis can be made with **CT examination**. However, subarachnoid hemorrhage is undetectable in about 5% of CT scans in the first 24 hours, so if suspicion of subarachnoid hemorrhage persists, **cerebrospinal fluid examination** is indicated.

A typical cerebrospinal fluid finding is oxyhemoglobin in spectrophotometric examination. The cerebrospinal fluid must be processed within 1 hour of collection. Typical findings include *increased protein* and in cytological examination, thousands to hundreds of thousands of *erythrocytes* and the presence of *bilirubin* in cerebrospinal fluid.


Subarachnoid hemorrhage

If subarachnoid hemorrhage is detected, the patient is referred to neurosurgery for brain panangiography, which should be performed within 72 hours of the onset of symptoms due to the risk of vasospasm. When an aneurysm is found and with an H+H score of up to 3, an operation is indicated - either clipping of the aneurysm's neck or filling the aneurysm's cavity with a detachable spiral - coiling. If the aneurysm is not detected, the patient is treated conservatively - opiates for pain, mucolytics and laxatives, and after 3-6 weeks, control panangiography is indicated.

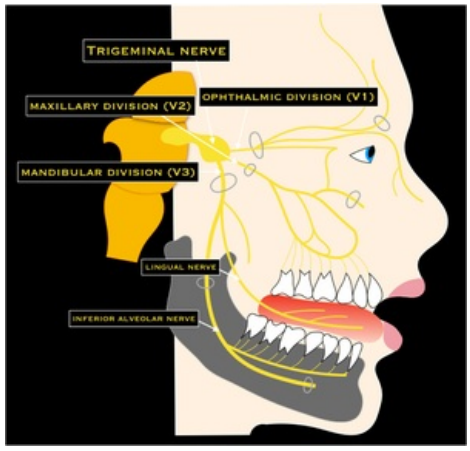
Cervicocranial syndrome

Cervicocranial syndrome is associated with occipital headache, which is accentuated by head and cervical spine movement. The pain may spread to the frontal or retrobulbar area, most often bilaterally. The pain can be dull, sharp or stabbing. The cause of pain is functional disorders of the dynamics of the atlantooccipital junction or the cervical spine, which may be caused by morphological changes and excessive load on the musculoskeletal system. Pain is sometimes combined with vertigo and nausea, vomiting and nystagmus (cervicovestibular syndrome).

X- ray examination of the cervical spine and possibly the skull may be required. In the acute stage, analgesics, non-steroidal anti-inflammatory drugs, short-term muscle relaxants and resting regime are indicated. In the chronic stage and as a recurrence prevention, rehabilitation is essential.

Primary neuralgia of trigeminal nerve

This is paroxysmal pain frequently in the regions of the 2nd or 3rd branches of the trigeminal nerve, mostly in patients over 40 years of age. The pain is localized unilaterally (especially in the initial phase) with the attacks occuring suddenly as very intense "whipping" pain, that last for a maximum of a few seconds and often recur. Triggering mechanisms such as chewing, brushing teeth, speaking, cold or tactile stimuli of the face are often associated.



Divisions of the trigeminal nerve

The neurological findings are normal.

Treatment

- *Carbamazepines* (initial dose 100 mg, gradually increase up to 800–1200 mg), *amitriptyline* from the dose 25 mg in the evening and increase slowly according to tolerance up to the effective dose (200–300 mg), sometimes also *phenytoin* (initial dose 100 mg and ascend to 300 mg) and *baclofen* (initial dose 10 mg and gradually increase according to tolerance up to 100 mg), *gabapentin* (initial dose 100 mg, gradually increase up to 2400 mg) may also be effective. The effect of this treatment may be exhausted over time.
- If conservative treatment fails, neurosurgical procedures are indicated (especially: glycerol instillation into the Meckeli cavum, Gasser ganglion microcompression) or gamma irradiation.
- In the case of proven neurovascular conflict (contact of the vascular loop with the trigeminal nerve root at the exit of the strain) microvascular decompression is performed.

Diagnostic criteria of primary neuralgia of trigeminal nerve

A. Paroxysmal attacks of facial pain lasting a few seconds and less than 2 minutes
B. Pain has at least 4 of the following characteristics: ▪ localization on some branch of trigeminal nerve ▪ sudden, intense, sharp, superficial, stabbing, burning pain ▪ high intensity pain ▪ pain can be triggered by trigger zones *or* by *certain* activities such as brushing teeth, chewing ▪ the patient is asymptomatic between paroxysms
C. Normal neurological findings
D. Attacks on individual patients are repeated stereotypically
E. Other causes of facial pain have been ruled out by all available methods

Secondary neuralgia of trigeminal nerve

It arises as a result of infections in the ENT area (chronic sinusitis), infections of the teeth, temporomandibular joint and after herpes zoster infection in the face. The pain tends to be more long-lasting and less intense than in primary neuralgia. Neurological findings often include sensitivity disorder in some branch of the nerve or an alteration of the corneal and masseter reflex.

- Diagnosis: ENT examination (including X-ray of the paranasal sinuses), dental examination (including a panoramic image of the jaw).
- Treatment: elimination of the cause if possible, medications and their dosing are similar to those of primary neuralgia.

Venous thrombosis

They arise in hypercoagulable conditions of various etiologies (such as bacteremia, sepsis, cancer, pregnancy and the puerperium, DIC (disseminated intravascular coagulopathy), collagenoses, traumas) or may also follow a local infection (for instance sinusitis, otitis media). The most commonly affected are the superior sagital sinus, the transverse sinus and the cavernosus sinus.

Headache is the most common symptom, typically accentuated by an abdominal press. Intracranial hypertension syndrome often develops, and focal ischemia, which arises from venous drainage disorders, results in a number of extinction and irritation symptoms.

Sinus cavernosus thrombosis - severe pain behind the eyes and in the eyes, or around it, which may protrude in front of the bulbus, papillary edema, hemorrhage on the back of the eye. This could result in impaired vision and even blindness.

Transverse sinus thrombosis - pain often has a maximum intensity in the area of the mastoid process. Intracranial hypertension syndrome and temporal symptoms dominate clinically.

Superior sagital sinus thrombosis - focal motor epileptic seizures often occur, later paresis and subsequently the development of intracranial hypertension.

Diagnostic methods: MRI AG, angiography.

It is a serious condition with a high risk of death or permanent consequences.

Therapy: heparinization followed by warfarinization. In the case of neurosurgery, local thrombolysis via the jugular vein to the transverus sinus and the superior sagitalis sinus could be performed.

Carotid artery dissection

Dissection most often affects young and middle-aged people. Clinically, unilateral headaches, neck pain, Horner's syndrome, cerebral nerve palsy or hemispheric symptoms may be present. Some patients could experience tinnitus.

Diagnostic methods: ultrasound, angiography, MRI (a wall hematoma is displayed on a classic MRI).

A serious condition with a high risk of death or permanent consequences.

Therapy: thrombolysis, anticoagulation, angioplasty, stents.

Sleep Apnea Syndrome (SAS)

Headaches are much more common in SAS than in other sleep disorders; it reported by about 20% of patients. It typically appears in the morning after waking up, are mild, dull, non-pulsating, diffuse, and usually subside within an hour. However, the intensity of the headache does not correlate with the severity of the SAS.

Chronic daily headache

It is a relatively common complication of primary headaches, especially tension pain and migraines. At various times (1-10 years), between the onset of the classic primary headache, lower-intensity headaches begin to appear, which occurs with increasing frequency until it later become permanent. The disease loses its original paroxysmal character and classic features. The risk factors for the progression are in particular:

- overuse of analgesics (especially combined with codeine, a maximum of 5 tablets per month is considered a safe limit),
- neurotic personality (tendency to anxiety, depression),
- external stressful conditions (employment, family),
- menopause.

Diagnosis: medical history, normal neurological findings, normal findings on auxiliary examination methods.

Therapy: omission of an overused drug, treatment of comorbidities - especially depression, psychotherapy.

References and sources

- [ws: Cluster headache/PGS]]
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