Medical Academy named after S.I. Georgievsky of V.I. Vernadsky CFU
Department of Neurology and Neurosurgery

Class 13

Epilepsy and other paroxysmal conditions. Headache. Major

Epilepsy and Its Differential Diagnosis. Types of Epilepsy
Episodic Neurological Disturbances of Nonepileptic Origin
Cerebral Palsy

1. **Goals:**
   - *1.1. To study the Neurological fundamentals of the Epilepsy.*
   - *1.2. To study the Neurological fundamentals of the Episodic Neurological Disturbances of Nonepileptic Origin.*

2. **Basic questions:**

2.1. Epilepsy:

2.1.1. *Etiology. Pathogenesis. Classifications. Clinical Features. Diagnostic evaluation. Treatment. Prophylaxis. Prognosis.*

2.1.2. *Generalized Epilepsy. Clinical Peculiarities. Diagnosis. Treatment.*

2.1.3. *Partial Epilepsy. Clinical Peculiarities. Diagnosis. Treatment.*

2.1.4. *Epileptic Status. Diagnostic Approaches. Treatment.*

2.2: Episodic Neurological Disturbances of Nonepileptic Origin:

2.2.1. *Syncope. Etiology. Pathogenesis. Clinical Features. Diagnostic evaluation. Treatment. Prophylaxis. Prognosis.*

2.2.2. *Drop Attacks. Etiology. Clinical Features. Diagnostic evaluation. Treatment. Prophylaxis.*

2.3. Cerebral Palsy

Literature:

Mark Mumenthaler, M.D., Heinrich Mattle, M.D. Fundamentals of Neurology. – P.161-172.

Headache. Vertigo.
Major Neurological Syndromes.

1. **Goals:**
   - *1.1. To study the different types of Headache and Facial Pain, their clinical peculiarities and treatment strategies.*
   - *1.2. To study the different types of Vestibular Dysfunction.*
   - *1.3. To study the Neurological fundamentals of the Major Neurological Syndromes.*

2. **Basic questions:**

2.1. Primary Headache:

2.1.1. *Migraine .*

2.1.2. *Cluster Headache.*

2.1.3. *Tension-type Headache.*

2.2. Secondary Headache:

2.2.1. *Intracranial Haemorrhage.*

2.2.2. *Temporal Arteritis.*

2.2.3. *Occlusions of Cerebral Vessels..*

2.3. Facial Pain:

2.3.1. *Trigeminal Neuralgia.*

2.3.2. *Glosso-pharyngeal Neuralgia .*

2.3.3. *Auriculotemporal Neuralgia*

2.4. Vertigo:

2.4.1. *Vestibular Vertigo.*

2.4.2. *Nonvestibular Vertigo.*

Literature:

Mathias Baehr, M.D., Michael Frotscher, M.D. Duus' Topical Diagnosis in Neurology. – 2005 – P.184-194
Mark Mumenthaler, M.D., Heinrich Mattle, M.D. Fundamentals of Neurology. – 2005 – P.243-255, 199-204

EPILEPSY

The physiological definition of epilepsy is unchanged from that provided by Hughlings Jackson in the nineteenth century: ***Epilepsy is the name for occasional, sudden, excessive, rapid and local discharges of grey matter.***

A distinction must be drawn between an isolated seizure and the recurring tendency to seizures which is epilepsy.

Epilepsy is a disorder characterized by recurrent (>2) unprovoked seizures.

A single seizure does not lead to a diagnosis of epilepsy.

An epileptic seizure is produced by a **temporally limited**, **synchronous electrical discharge of neurons in the brain**. It presents as a variable combination of motor, somatosensory, special sensory, autonomic, and/or behavioral disturbances, which arises suddenly and may last for a few seconds or a few minutes.

On rare occasions, seizure activity persists for more than 30 minutes and may go on for hours, or even longer, without interruption (**status epilepticus**).

The epileptic event may affect a circumscribed area of the brain (partial or **focal seizures**), or both cerebral hemispheres at the same time (**generalized seizures**).

An impairment of consciousness is found in generalized seizures and in so-called complex focal seizures.

In their differential diagnosis, epileptic seizures must be carefully distinguished from other sudden events involving neurological deficits and disturbances of consciousness.

Epidemiology. It has been calculated that 1% of all individuals suffer from epileptic seizures. The child of a parent with idiopathic epilepsy has a 4% likelihood of suffering from it.

Pathophysiology. Epileptic seizures are due to dysfunction of neurons in the brain, which expresses itself electrophysiologically as an abnormality of the fluctuations of electrical potential that are seen in an electroencephalogram.

If the surface EEG is normal, such abnormalities can be revealed by recording with depth electrodes.

The underlying cause is an *imbalance of excitatory and inhibitory potentials*, with predominance of excitatory neurotransmitters such as glutamate and aspartate, or diminished activity of inhibitory neurotransmitters such as GABA. The synchronous discharge of neurons in a particular area of the brain is accompanied by a local increase in blood flow.

Etiology. Epileptic seizures can be produced by structural lesions in the brain (so-called epileptic foci: scar, tumor, congenital malformation), by metabolic disturbances (e. g., hypoglycemia), or by toxic influences (e. g., alcohol). These are all types of **symptomatic epilepsy**.

In contrast, the **idiopathic epilepsies** involve a genetic predisposition to epileptic seizures, in the absence of a structural lesion.

The **cryptogenic epilepsies** are presumed to be of symptomatic origin, although their cause cannot (yet) be demonstrated. Molecular genetic techniques have made it possible to trace certain forms of focal epilepsy back to abnormalities of specific gene loci. Not uncommonly, more than one etiologic factor is at work: thus, diseases of the brain are more likely to produce epileptic seizures in persons with an inherited predisposition to seizures than in other, normal individuals.

General characteristics of epileptic disorders are the following:

- Epileptic seizures are *events of sudden onset*, which occur with *variable frequency* (generally in the range of a few seizures per year to several per day).
- They often present with *motor phenomena* (in particular, repetitive, clonic twitching or changes of muscle tone) and sometimes with *somatosensory*, *special sensory*, and/or *autonomic manifestations.*
- Depending on their type, they may involve an *impairment or loss* of consciousness, or consciousness may be *preserved* during the seizure.
- The seizure may be preceded by premonitory symptoms of various kinds (*auras*, e. g., nausea, ascending warmth, or a feeling or unreality).
- In some patients, seizures occur in response to specific *provocative* and *precipitating factors* (sleep deprivation, alcohol withdrawal, medications, strobe lighting, hyperventilation, fever).

Classification of the Epilepsies

Epilepsy can be **classified** according to a number of criteria, including:

1) **Etiology**, e. g.:
- "genuine/idiopathic," genetic,
- symptomatic,
- cryptogenic.
2) **Age of onset**, e. g.:
- epilepsy of childhood or adolescence,
- epilepsy of adulthood,
- late epilepsy (age 30 and up; <u>always suspect a primary organic disease</u>).
3) **Setting in which seizures are most frequent**, e. g.:
- sleep epilepsy,
- epilepsy on awakening.
4) **EEG correlate** and corresponding topographical localization, e. g.:
- generalized epilepsy,
- focal (partial) epilepsy.
5) **clinical manifestations** of each seizure.

Clinical classification of seizures.

The nomenclature for the different clinical types of epileptic seizure proposed by the International League Against Epilepsy is reproduced in Table below, with the addition of a few further designations that are currently in general use.

I. **Partial seizures** (attributed to seizure activity in one hemisphere or part of one hemisphere at the onset).

A. <u>Simple partial seizures</u>

1. With motor signs
    a. Focal motor without march
    b. Focal motor with march (Jacksonian)
    c. Versive
    d. Postural
    e. Phonatory
2. With somatosensory or special-sensory symptoms
    a. Somatosensory
    b. Visual
    c. Auditory
    d. Olfactory
    e. Gustatory
    f. Vertiginous
3. With autonomic symptoms or signs
4. With psychic symptoms
    a. Dysphasia
    b. Dysmnesic

c. Cognitive
d. Affective
e. Illusions
f. Structured hallucinations

B. Complex partial seizures
1. Simple partial seizures at onset, followed by impairment of consciousness
a. With simple partial features
b. With automatisms
2. With impairment of consciousness at onset
a. With impairment of consciousness only
b. With automatisms

C. Partial seizures evolving to secondarily generalized seizures
1. Simple partial seizures evolving to generalized seizures
2. Complex partial seizures evolving to generalized seizures
3. Simple partial seizures evolving to complex partial seizures evolving to generalized seizures

II. **Generalized seizures** (bilateral hemispheric symmetrical and synchronous discharge associated with loss of consciousness from the onset).

A. Absence seizures
1. Typical absence seizures
a. Impairment of consciousness only
b. With mild clonic components
c. With atonic components
d. With tonic components
e. With automatisms
f. With autonomic components
2. Atypical absence seizures

B. Myoclonic seizures
C. Clonic seizures
D. Tonic seizures
E. Tonic–clonic seizures
F. Atonic seizures

Generalized Seizures

Generalized seizures involve both cerebral hemispheres, either from the outset of seizure activity, or when an initially focal seizure becomes secondarily generalized (see below). They typically involve an obvious **impairment of consciousness. Abnormalities of muscle tone** are always present and there are often **involuntary**, **repetitive motor phenomena** involving both sides of the body.

Tonic–Clonic Seizures (Grand Mal Epilepsy)

Pathogenesis and etiology. A grand mal seizure may be **idiopathic**; in such patients, it is usually primarily generalized ("centrencephalic").

It may also be due to a circumscribed brain lesion (**secondary generalization**). The cause can sometimes be inferred from the findings of the clinical history, imaging studies, and EEG, though it often remains obscure.

Clinical manifestations. Grand mal seizures are the most common and most impressive type of epileptic seizure and also the most familiar to nonprofessionals. Such seizures are sometimes heralded by a loud cry or shout. Next, the patient *acutely loses consciousness* and falls to the ground, and the muscles are *tonically contracted*: the limbs are extended, and the trunk and neck are hyperextended. About 10 seconds later, there follows a *rhythmic*, *clonic*, *generalized twitching of all muscles of the body*, accompanied by cyanosis of the face, frothing at the mouth, and possibly by a tongue bite and urinary or fecal incontinence. The twitching persists for a minute or a little longer and is followed by a period of initially deep unconsciousness. Within a few minutes, a gradual transition begins to a phase of confusion and somnolence (postictal twilight state) and

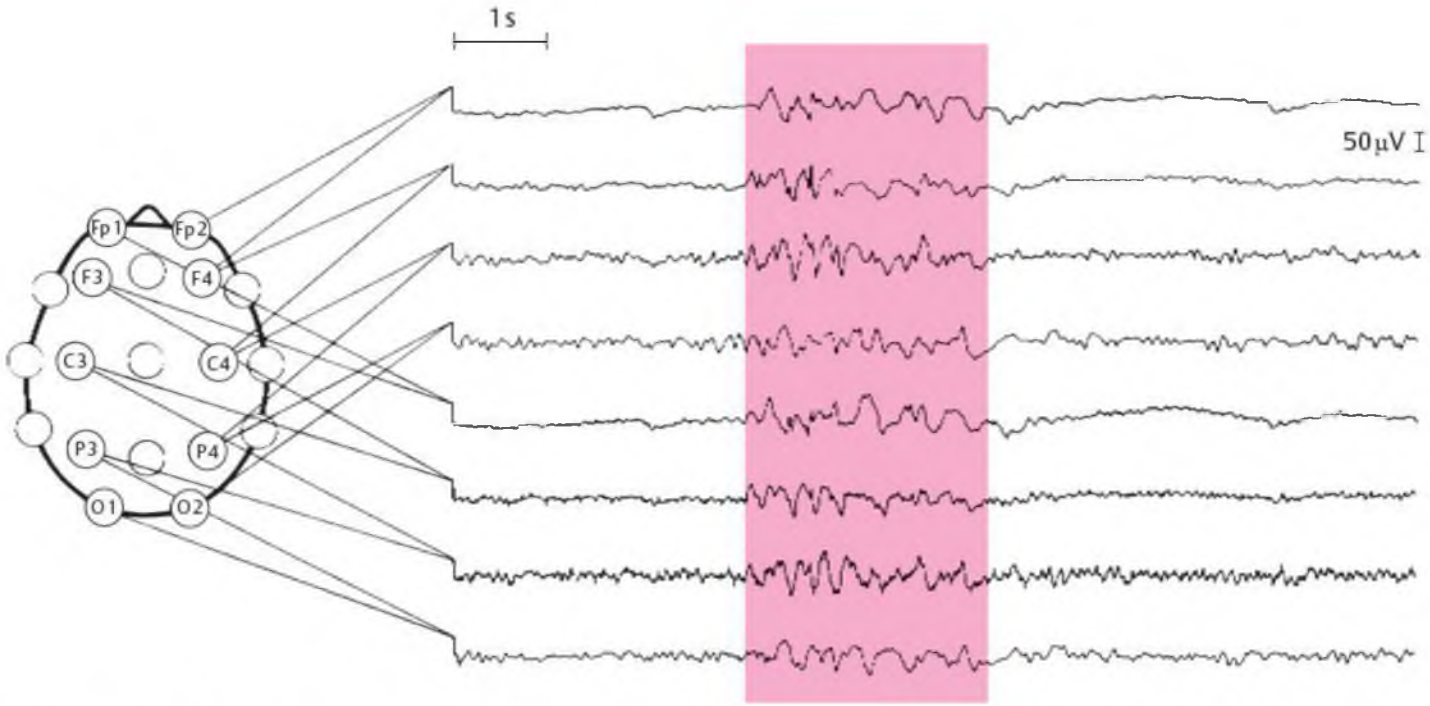
then to the return of normal consciousness. The entire seizure typically lasts about 10 minutes. The patient may remember the aura but is otherwise entirely amnestic for the seizure event. Afterward, the patient is tired and may complain of myalgia. A tongue bite, urinary or fecal incontinence, and fall related injuries may be evident. Shoulder dislocation and vertebral or other fractures are rare.

Diagnostic evaluation (<u>for all types</u>).

In an adult presenting with a first seizure, direct tests towards establishing a possible underlying cause, particularly if no provoking factors are obvious, for example alcohol withdrawal. Test should include the following:

- <u>Blood tests</u>: FBC, U & E, serum glucose level, calcium, magnesium, LFT and blood and urine toxin levels if suspected drug or alcohol abuse.
- <u>Imaging studies</u>: Perform a CT with or without contrast to look for a structural lesion. This may be followed up with an MRI with or without gadolinium to assess for structural lesions (neoplasm, arteriovenous malformations or cavernomas). Repeat imaging studies may be needed. Remember: In an adult presenting with a first seizure, imaging studies starting with a CT of brain with or without contrast is an important part of diagnostic work up to exclude a structural lesion like neoplasm, abscess or vascular malformations.
- <u>EEG</u>: Is performed to help in classifying seizure type. EEG is often diagnostic; careful scrutiny should be made to distinguish between focal and generalized interictal activity. Video-EEG monitoring is the gold-standard test to diagnose and classify seizures if the diagnosis is unclear and also if the seizures are refractory to initial treatment.

Even in the interictal period, the electroencephalogram may reveal the typical picture of synchronous, generalized spikes and waves in all electrodes (Fig. below).



Interictal EEG in a patient with grand mal seizures, showing a synchronous paroxysm of generalized, partly atypical spikes and waves.

Differential diagnosis

- Psychogenic nonepileptic seizures can be confused with epileptic seizures. Video-EEG monitoring can help distinguish psychogenic spells from epileptic seizures.
- Tic disorders and other movement disorders may be confused for myoclonic or other seizures

Treatment. The medication of first choice for the treatment of grand mal epilepsy is **valproate**.

Absences (Petit Mal)

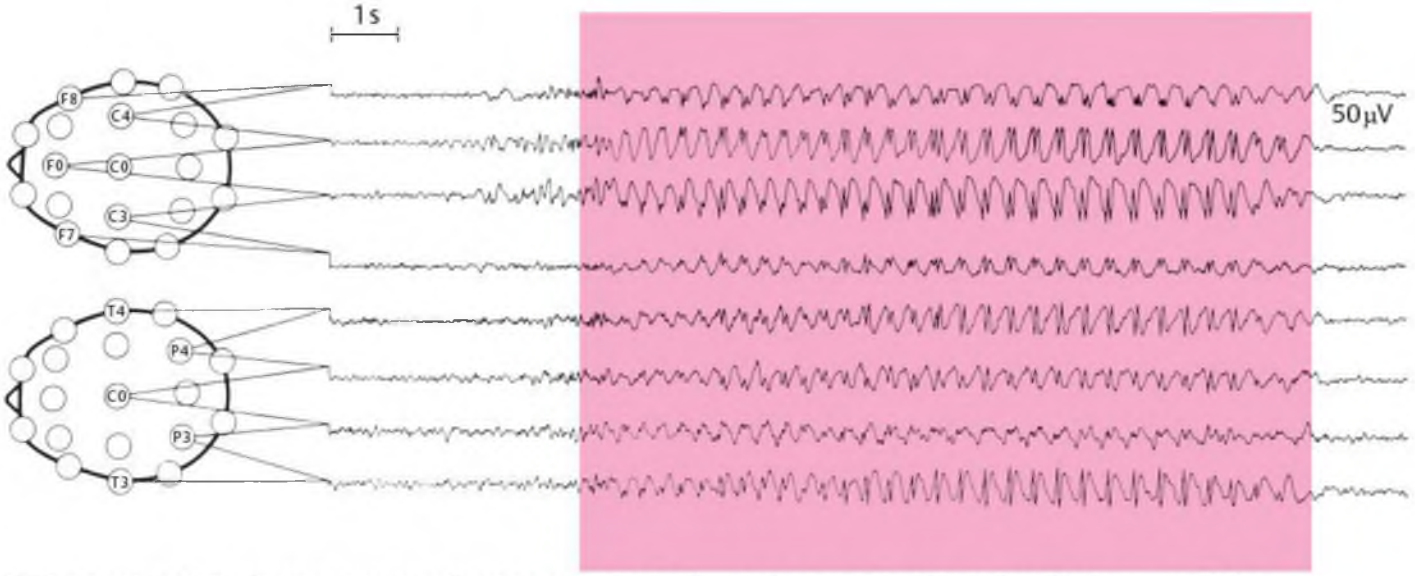
Absences are very brief seizures involving a momentary diminution of consciousness, rather than a complete loss of consciousness. They most commonly occur in children and adolescents.

Etiology. Like other types of childhood epilepsy, absences are **idiopathic**.

Clinical manifestations. Motor phenomena are not always seen; if present, they are only mild (blinking, automatisms, loss of muscle tone, brief clonus).

In the simplest type of absence epilepsy, **petit mal epilepsy of school-aged children**, the seizures often seem to be no more than brief periods of "*absent-mindedness*": the child stares fixedly with eyes turned upward, blinks, and may make movements of the tongue or mouth, or pick at his or her clothes. These types of movements are called *petit mal automatisms.* The entire event lasts no more than a few seconds. Absences usually occur multiple times per day. The examining physician may be able to provoke an absence by having the patient hyperventilate.

Diagnostic evaluation. The **EEG** reveals a pathognomonic pattern of bursts of synchronous, generalized spike-and-wave activity at a frequency of about 3 Hz. These can be provoked by hyperventilation (Fig. below).



EEG in a patient with absence seizures, showing generalized spikes and waves at 3–4 Hz, induced by hyperventilation.

Treatment. The medications of first choice for the treatment of absences are **valproate** and **ethosuximide**.

Prognosis. About one in four affected children become free of seizures during puberty; in the remainder, seizures persist. One half of these patients with persistent absence seizures will go on to develop grand mal seizures as well.

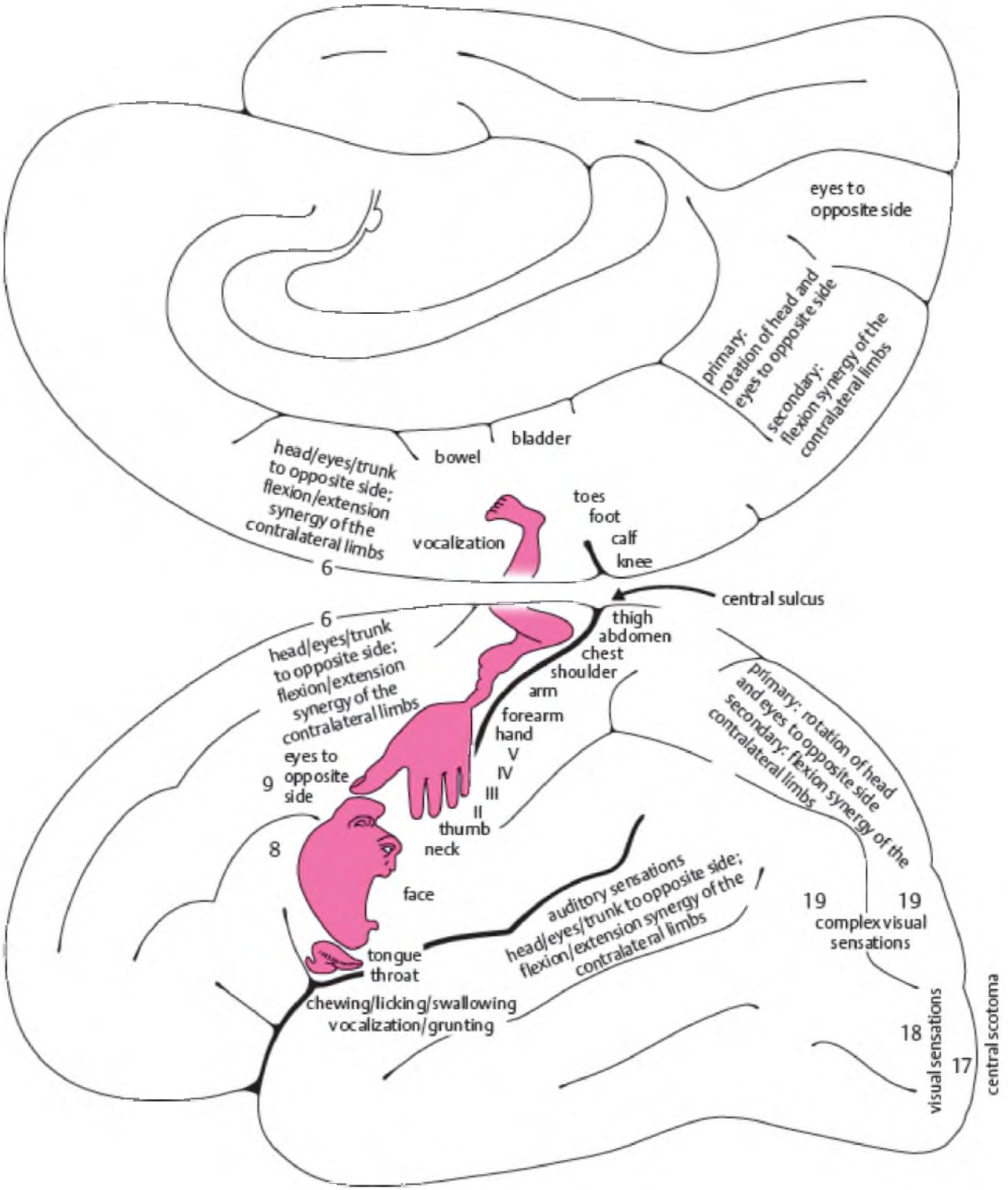
Partial (Focal) Seizures

Focal seizures are always due to a **circumscribed lesion in the brain**.

The specific manifestations of the seizure correspond to the site of the lesion. Unlike generalized seizures, which always involve an impairment of consciousness, focal seizures may occur with the patient remaining fully conscious (**simple partial seizures**).

They can, however, involve an impairment of consciousness, in which case they are called **complex partial seizures**. The excitation arising in the epileptic focus may spread to the entire brain and thereby provoke a **secondarily generalized grand mal seizure**. In such patients, the initial focal phase may be very brief and is not always clinically recognizable.

Figure below schematically represents the clinical manifestations that can be expected in focal seizures arising from various brain areas. The type of attack depends on the site of the focal lesion



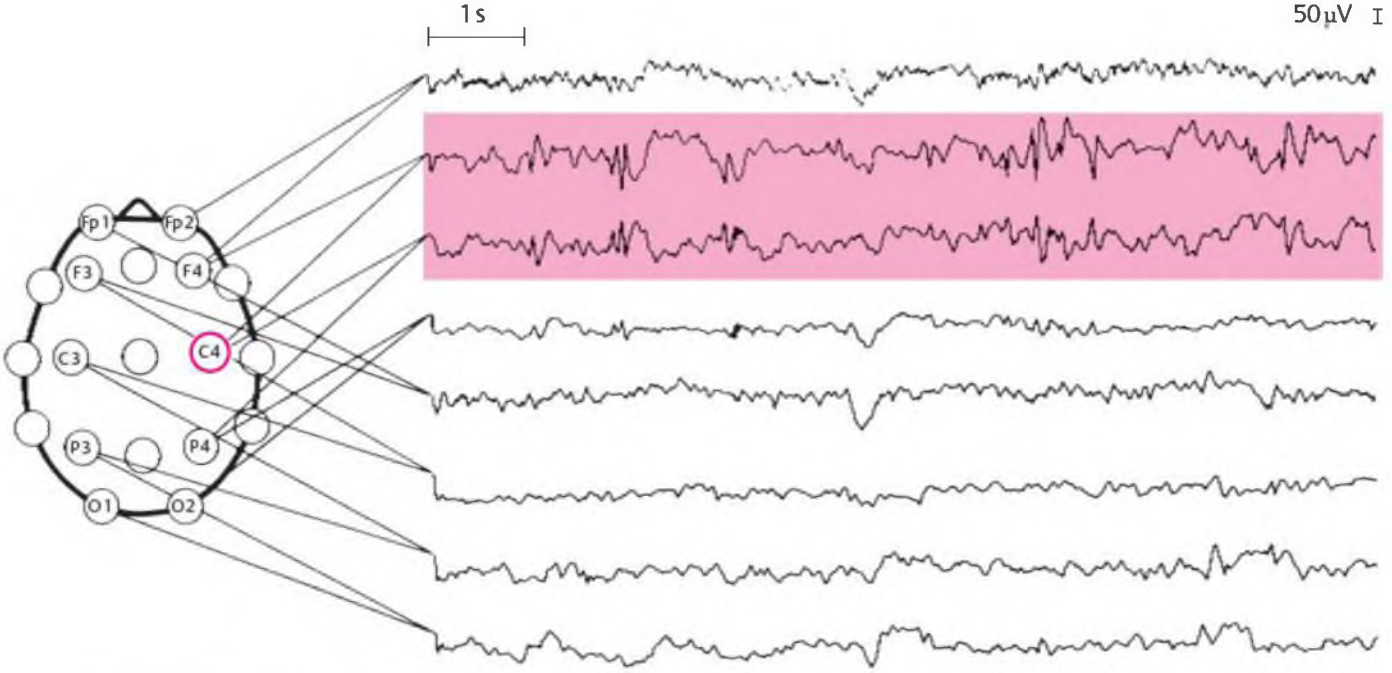
Simple Partial Seizures

Simple partial seizures can be purely motor, mixed sensory and motor, or purely sensory (either Somatosensory or special sensory). They are, by definition, not accompanied by an impairment of consciousness, though they may undergo secondary generalization.

Individual types of simple partial seizure. A simple partial seizure may involve **focal motor twitching** on one side of the body, or sensory disturbances that suddenly arise in a circumscribed area of the body. Focal twitching confined to a very small area (e. g., a hand) and lasting for a very long time (hours or more) is called **epilepsia partialis continua** (of Kozhevnikov). In a **Jacksonian seizure**, the motor (or sensory) phenomena rapidly spread to the entire ipsilateral half of the body ("*Jacksonian march*"). If the seizure focus lies in the

precentral or the supplementary motor area, the seizure will be of **adversive** type: the patient's head and eyes turn tonically to the opposite side, while the contralateral arm is abducted and elevated. If the seizure focus lies in the visual or auditory cortex or the neighboring association areas, the seizure may consist of, or begin with, auditory or visual sensations, or even scenic images.

Diagnostic evaluation. The focal nature of the seizure, or its focal origin (in the case of secondarily generalized seizures), is demonstrated not only by the clinical manifestations, but also by the **electroencephalogram**, which displays localized epileptic activity over the seizure focus (Fig. below).



EEG during a focal epileptic seizure. Right central epileptogenic focus with spikes, sharp waves, and slow waves. Phase reversal at electrode C4.

Treatment. The medications of first choice for the treatment of focal seizures are **carbamazepine** and **oxcarbazepine**.

Complex Partial Seizures

This type of seizure was previously known as *psychomotor epilepsy* or, alternatively, *temporal lobe epilepsy*. It is due to a lesion in the limbic system, usually in the temporal lobe, but sometimes in the frontal lobe.

Etiology. The most common cause of complex partial seizures is a **perinatal lesion** of hypoxic origin (mesial temporal sclerosis or hippocampal sclerosis). Other causes include **congenital developmental anomalies** (e. g., disorders of neuroblast migration), trauma, tumors.

Clinical manifestations. Complex partial seizures have *sensory*, *behavioral*, *psychomotor*, and *autonomic manifestations*, which are described in further detail in Table 9.**8**. Which of these manifestations will be present in the individual patient depends on the precise location of the epileptic focus. The manifestations may vary to a mild extent from seizure to seizure, though an unvarying, stereotypical course is more common. In addition to the manifestations mentioned above, many patients report experiencing *deja vu* and related phenomena, i. e., the strong, but nonetheless inaccurate, feeling of having already seen or experienced what one is seeing or experiencing at the moment. Patients whose seizure focus lies in the uncinate gyrus also have *olfactory hallucinations*, or, as they are called, *uncinate fits*. These are often produced by a mass in the temporal lobe.

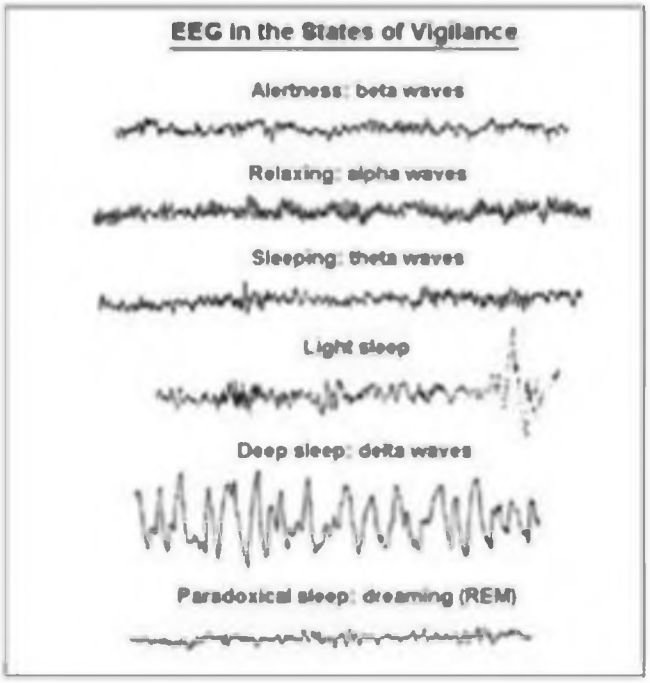
Clinical manifestations of partial complex seizures (temporal lobe epilepsy)

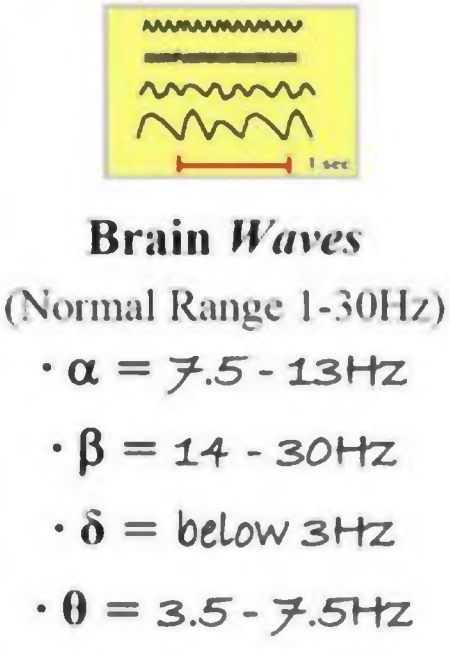
Category	Manifestations	Remarks
Sensory disturbances	Dizziness, dysmorphopsia (macropsia, micropsia, everything seems to be far away), gustatory sensations, unpleasant olfactory sensations	Uncinate fits
Autonomic phenomena	Shortness of breath, palpitations, nausea, salivation, dry mouth, hunger, urge to urinate, abdominal sensations	Often, ascending sensation from stomach to throat
Behavioral and psychomotor manifestations	Traumatic experience, feeling of unreality, feeling of unfamiliarity (jamais vu), forced thoughts, déjà vu, déjà vécu, unfounded anxiety or rage, hallucinations, twilight states	
Twilight states	Automatic, semiorganized, but inappropriate behavior, e.g., picking at clothes, senseless moving around of objects, etc. (twilight attacks); long-lasting, semiorganized complex behaviors that may even involve travel over a long distance (twilight state, fugue épileptique)	Amnesia for these states
Temporal syncope	Collapse, usually immediately following one of the above phenomena, typically with only brief unconsciousness	No sudden falling
Psychomotor status epilepticus	Very long persistence of the above phenomena, or repeated occurrence with less than full recovery in between	Rare

Illustrative case description: The patient's seizures begin suddenly, with a peculiar feeling of distance from his surroundings. Everything seems to be far away, unreal, and like a dream. At the same time, he notices a strange sensation in the pit of his stomach, ascending to his neck. He may also have palpitations or shortness of breath. On some occasions, his consciousness is more severely affected: he stares blankly ahead, makes chewing and swallowing movements, produces gagging noises, and fails to respond to questions. He picks at his clothes, makes purposeless hand movements, and sometimes falls over. Rarely, when he is in this "twilight state," he carries out complex activities, perhaps even a "*fugue epileptique.*" The entire episode usually lasts one or a few minutes, but may last much longer.

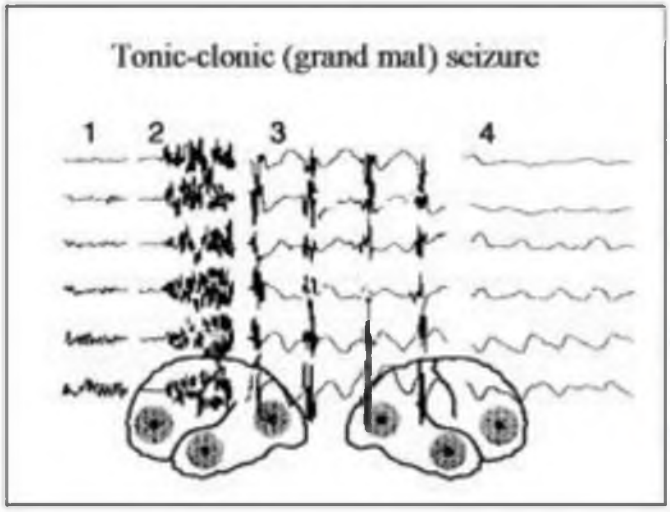
Diagnostic evaluation. Complex partial seizure activity can generally be diagnosed from its typical clinical picture. The **EEG** reveals temporal slow waves or spikes. In the interictal period, however, it is *usually normal*.

Treatment. The medications of choice for complex partial seizures, as for simple partial seizures, are **carbamazepine** and **oxcarbazepine**. Alternatively, **valproate** can be used.



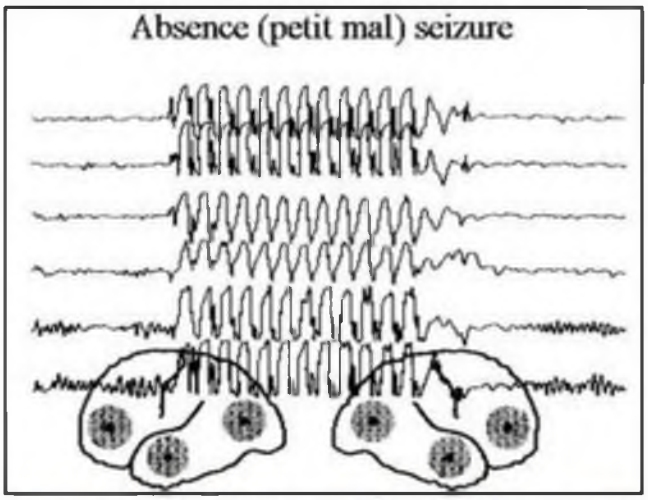


EEG during generalized tonic-clonic seizure:



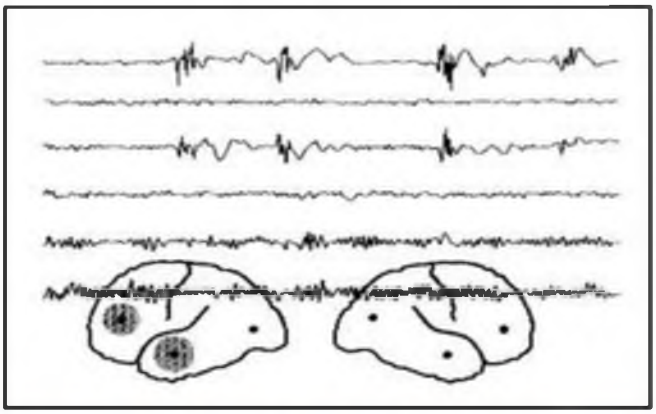
1. Normal EEG
2. Tonic Phase
3. Clonic Phase
4. Coma Phase

EEG during an absence seizure:



3 Hertz Spike & Wave Discharge all channels

EEG during a focal (simple or complex) seizure:



Spike & wave discharge in focussed region of brain

Antiepileptic drugs of choice depending on the type of seizure

	Partial seizures with or without generalization	Absences	Primary generalized tonic–clonic seizures	Myoclonic seizures
1st choice	carbamazepine valproate	valproate ethosuximide	valproate	valproate
2nd choice	gabapentin lamotrigine oxcarbazepine phenytoin tiagabine topiramate levetiracetam	lamotrigine	lamotrigine	clonazepam ethosuximide lamotrigine
3rd choice	vigabatrin clonazepam phenobarbital primidone	clonazepam	phenobarbital primidone	primidone

Status Epilepticus

Status epilepticus refers to a prolonged, uninterrupted epileptic seizure, or multiple seizures occurring in rapid succession without recovery in between.

! Generalized (grand mal) status epilepticus, in which the patient does not regain consciousness between seizures, is a life-threatening emergency because of the danger of respiratory complications and ensuing cerebral ischemia.

In **absence status** the patient is not unconscious, but rather confused or mildly dazed, with inappropriate behavior. **Complex partial status epilepti**cus can be mistaken for an acute psychotic episode.

The EEG is diagnostic.

Treatment. Grand mal status epilepticus should be treated with a bolus dose of a **benzodiazepine**, e. g., diazepam 10–20 mg i. v., followed by **intravenous phenytoin** (by slow push or drip) or **valproate** (IV push followed by drip). If seizure activity does not stop within 40 minutes, the patient must be *intubated and ventilated* and put in artificial coma with a barbiturate such as *thiopental* or *propofol.* Petit mal status epilepticus and psychomotor status epilepticus respond to *clonazepam* 2–4 mg i. v.

Episodic Neurological Disturbances of Nonepileptic Origin

Because the clinical presentations of epileptic seizures are so highly varied, their differential diagnosis necessarily includes a wide variety of conditions. Any episodic loss of consciousness, impaired motor function, or fall might be due either to an epileptic seizure or to a ***nonepileptic*** event of another etiology.

Episodic neurological disturbances of nonepileptic origin can be classified into four major types:

1. Transient loss of consciousness and falling;
2. Falling without loss of consciousness;
3. Loss of consciousness without falling;
4. Episodic movement disorders without loss of consciousness.

Episodic Disturbances with Transient Loss of Consciousness and Falling. Syncope

Syncope is a *very brief loss of consciousness* during which the affected individual falls to the ground.

It is due to a very brief loss of function of the brainstem reticular formation, which, in turn, is usually caused by temporary ischemia and tissue hypoxia.

Syncope can be of **vasomotor** or **cardiogenic** origin. Reflex circulatory syncope, the commonest kind of syncope, can be precipitated by intense emotion (e. g., the sight of blood, anticipatory anxiety), heat, prolonged standing, or physical pain. The affected person becomes dizzy, sees black spots before his or her eyes, turns pale, breaks out in a sweat, and then collapses to the ground. Wakefulness and full orientation are regained at once in most patients.

Etiologic subtypes of reflex circulatory syncope include
- *idiopathic vasomotor collapse in adolescents*,
- *pressor syncope* after prolonged coughing,
- *micturition syncope*,
- *swallowing syncope*,
- *extension syncope* (mainly seen in younger patients who stand up too quickly from a squatting position).

Orthostatic syncope is a feature of many neurological diseases (e. g., multisystem atrophy),

Cardiogenic syncope is especially common in older patients. Its causes include cardiac arrhythmias (third degree AV block, sick sinus syndrome, tachycardias) and other types of heart disease (e. g., valvular aortic stenosis, atrial myxoma, and chronic pulmonary hypertension with cor pulmonale).
"Convulsive syncope": syncopal episodes are sometimes accompanied by brief, clonic muscle twitching. This may make a syncopal episode even harder to distinguish from an epileptic seizure.

Episodic Falling without Loss of Consciousness. Drop Attacks

In a so-called ***drop attack***, the patient suddenly falls to the ground without braking the fall. Consciousness is apparently preserved during the event; in some patients, however, the patient may, in fact, lose consciousness without realizing it afterward, and too briefly for others to observe.

Some drop attacks are due to ***atonic epilepsy***, others to ***basilar ischemia. Cryptogenic drop attacks*** have been described in older women ("climacteric drop syncope"). Finally, drop attacks can be caused by basilar impression and other structural abnormalities of the craniocervical junction.

CEREBRAL PALSY

Cerebral palsy is the term used to describe the neurological disorder of motor dysfunction that occurs as a direct result of injury to the developing brain. The insult is nonprogressive and occurs before the age of 3–5 years and manifests as abnormalities of tone, posture, or motion. Although the insult is nonprogressive, the manifestations of motor dysfunction may subtly change with time, as the injured brain matures. However, by definition, this condition does not involve true neurological regression.

Prevalence. The prevalence of cerebral palsy among children at school entry is about 2 per 1,000 live births.

Risk factors. The most common risk factors for cerebral palsy have varied over time because of advances in prenatal and neonatal care. Prematurity is a risk factor for cerebral palsy. The risk of cerebral palsy rises steadily as birth weight declines. The risk is approximately 3.4 per 1,000 in infants weighing 2,500 g and over, 13.9 per 1,000 in infants weighing 1,501–2,500 g, and 90.4 per 1,000 in infants less than or equal to 1,500 g. Infants of normal birth weight with a 5-minute Apgar score of 3 or less had a 5% probability of developing cerebral palsy. Similar scores at 10 minutes increased the risk to 17%, and scores of 3 or less at 20 minutes were associated with a 57% risk of cerebral palsy. In a term infant, risk factors include in-utero infections, maternal chorioamnionitis, genetic thrombophilic tendencies, meconium aspiration, breech presentation, placental abnormalities that include placental abruption and maternal factors such as hypertension.

Etiology. It is known that many conditions can injure the developing brain and lead to cerebral palsy. Yet, approximately one quarter of all cases have no definable cause.

Causes of cerebral palsy.

1. ***Prenatal***
   - First trimester (44%): Teratogens, genetic syndromes, brain malformations, chromosomal abnormalities
   - Second and third trimesters: Intrauterine infections, fetal/placental dysfunction
2. ***Labor and delivery*** (19%): Pre-eclampsia/eclampsia, complications of labor and delivery
3. ***Perinatal*** (8%): Hypoxic-ischemia, sepsis/CNS infections, prematurity, stroke, traumatic brain injury
4. ***Childhood*** (5%): Meningitis/encephalitis, traumatic brain injury, toxins
5. ***No obvious cause*** (24%)

Note: Cerebral palsy occurring repeatedly in a family that is not due to a definable genetic syndrome or chromosomal abnormality should raise the concern that the diagnosis of cerebral palsy is inaccurate. In these cases, an underlying neurometabolic or neurodegenerative disorder should be sought.

Commonly associated conditions.

Many children with cerebral palsy have at least 1 additional disability associated with damage to the CNS. The most common associated deficits are:

- Cognitive impairment
- Sensory deficits
- Communication disorders
- Seizures
- Feeding problems
- Behavioral and emotional problems

The most important cerebral movement disorders

Name of disorder	Clinical features	Pathoanatomical substrate	Causes
infantile spastic diplegia (Little disease)	spasticity, predominantly in the legs; pes equinus, scissor gait, mentally often normal	pachymicrogyria (abnormally hard, small gyri)	perinatal injury (disturbance of cerebral development, embryopathy, severe neonatal jaundice)
congenital cerebral monoparesis	usually, paresis of arm and face	porencephaly (cavities in the brain parenchyma), localized atrophy	birth trauma (asphyxia, hemorrhage)
congenital hemiparesis	arms more severely affected than legs, seizures in ca. 50%, usually mentally impaired	porencephaly	birth trauma (asphyxia, hemorrhage)
congenital quadriparesis (bilateral hemiparesis)	arms more severely affected than legs, occasionally bulbar signs, seizures; severe mental impairment	porencephaly, bilateral; often hydrocephalus	birth trauma (asphyxia, hemorrhage), also prenatal injury
congenital pseudobulbar palsy	dysphagia to liquids, dysarthria, usually not mentally impaired	bilateral lesions of the corticobulbar pathways	prenatal injury or birth trauma, congenital malformation (syringobulbia)
atonic–astatic syndrome (Foerster)	generalized flaccid weakness, inability to stand, impaired coordination, severe mental impairment	frontal lobe atrophy cerebellar defects	
bilateral athetosis (athétose double) **and congenital chorea** (choreoathetosis)	athetotic or other involuntary movements, often combined with spastic paresis	basal ganglionic defects, status marmoratus (multiple confluent gliotic areas in the basal ganglia); status dysmyelinisatus (Vogt) in cases of later onset	disturbances of cerebral development, perinatal injury, esp. severe neonatal jaundice
congenital rigor	rigor without involuntary movements, postural abnormalities, no pyramidal tract signs, severe mental impairment, seizures	status marmoratus	disturbances of cerebral development, perinatal injury, esp. severe neonatal jaundice
congenital cerebellar ataxia	gait ataxia, intention tremor and impaired coordination, motor developmental retardation, dysarthria, possibly in combination with other motor syndromes	cerebellar developmental anomalies	disturbances of cerebellar development

Diagnosis.

Cerebral palsy is a clinical diagnosis. To make the diagnosis, there has to be motor dysfunction that localizes to the brain as opposed to the peripheral nervous system. Motor dysfunction can manifest as failure to attain motor milestones at the appropriate age or abnormalities in tone. Clinical examination should localize the lesion to the brain. Clues on physical examination, which raise the suspicion of peripheral nervous system dysfunction include difficult-to-elicit or absent reflexes. Neurological regression or loss of neurological skills either in the
area of motor dysfunction or in other areas of development makes the diagnosis of cerebral palsy suspect.

Classification of cerebral palsy:
◦ ***Spastic***: Quadriparesis, hemiparesis, diparesis
◦ ***Dyskinetic***: Choreoathetosis, dystonia
◦ ***Ataxic***
◦ ***Mixed type***
◦ ***Spastic cerebral palsy***: Abnormalities of the pyramidal tract, increased tendon reflexes, increased muscle tone
◦ ***Dyskinetic***: Choreoathetosis or dystonia with variable tone and rigidity
◦ ***Ataxic cerebral palsy***: Truncal ataxia, limb dysmetria, and tremor

Diagnostic tests and interpretation.
Laboratory

Laboratory testing in patients suspected of having cerebral palsy is undertaken to delineate the extent of neurological impairment and the presence of other associated deficits, as well as in selected cases a thorough evaluation for progressive disorders mimicking cerebral palsy is undertaken. Testing that may be helpful includes:

- Hearing evaluation
- Eye examination including dilated eye examination
- Swallowing evaluation
- X-rays when scoliosis or dislocation of hips are suspected
- Serial developmental assessments
- EEG when spells suspicious of seizures are present

Imaging

- Imaging studies are helpful with regard to pattern recognition. Certain patterns are recognized as occurring in static disorders:
    a) Developmental abnormalities such as migrational disorders.
    b) Patterns of previous insult such as periventricular leukomalacia, multicystic encephalomalacia, and porencephalic cysts.
- Certain imaging abnormalities are specific in pointing away from cerebral palsy to a neurodegenerative disorder such as white matter changes indicative of leukodystrophy. In a portion of children with cerebral palsy, the MRI of the brain reveals no radiographic abnormality.
- In general, the MRI is a better tool for assessing brain parenchyma, whereas the CT is a better test for evaluation of the size of the ventricles.

Diagnostic Procedures/Other

Evaluation for a neurometabolic or neurodegenerative disease should be undertaken in any child with motor dysfunction and neurological regression.

Treatment. There are no specific medications for cerebral palsy.

Management should involve a multi-disciplinary approach aimed at addressing symptoms of spasticity, adventitious movements, and bulbar dysfunction; medications used for spasticity include baclofen, diazepam, or tizanidine. Dosages depend on age and body weight.

Early and aggressive physical and occupational therapy is recommended, with enrollment in an early intervention program. Speech therapy should be instituted if speech is delayed as well. Treatment, by

medications and surgical measures, is aimed at maximizing motor function, treatment of associated conditions, and monitoring and treatment of complications.

Patient education. Educating parents of children with cerebral palsy, demonstrating how positioning can be an effective way of helping the child with mobility, encouraging the parent–child interaction, and muscle stretching should be part of the information given to parents. Counseling on the need to monitor for associated conditions and complications is also an important aspect of treatment. Management should include education of all caretakers including teachers and therapists involved. Attempts should be targeted at maximizing function, including routes of communication, prevention of complications, and treatment of associated medical symptoms.

Prognosis. Although cerebral palsy is a static condition, the clinical symptoms can subtly change with time. Usually infants who are hypotonic with increased reflexes eventually become hypertonic within a few years. Athetosis or chorea in dyskinetic cerebral palsy may gradually appear toward the end of the first year of life. Ataxia may be noted only when the child begins to sit or reach for objects. Contractures tend to develop over time in patients with spasticity. In general, a number of factors affect prognosis: The type of cerebral palsy, the degree of delay in motor milestones, and the degree of associated deficits in intelligence, sensation, and emotional adjustment. The following are general guidelines:

- Children with hemiplegia and no other problems have a good chance of walking at about the age of 2 years.
- More than 50% of children with spastic diplegia learn to walk by about the age of 3 years.
- Of children with quadriplegia, 25% require total care, approximately 33% walk – usually after the age of 3 years.
- Few children who do not sit by the age of 4 years learn to walk.
- Seizure disorder is seen in up to a third of individuals who have cerebral palsy.
- Cognitive deficits are seen in about 50% of people with cerebral palsy.
- Complications of motor dysfunction include muscle spasms, orthopedic issues, undetected dental caries, skin breakdown, constipation, and gastric reflux, which is commonly seen in nonambulatory patients.

QUESTIONS FOR SELF-EDUCATION

1. ***How is epilepsy diagnosed (select applicable)?***
    1. Occurrence of 1 seizure
    2. Occurrence of more than 1 seizure
    3. Occurrence of more than 2 seizures
    4. Occurrence of more than 3 seizures
    5. Occurrence of more than 5 seizures
    6. Occurrence of less than 2 seizures
    7. Occurrence of less than 1 seizure

2. ***Indicate main types of epilepsy (select applicable):***
    1. Generalized
    2. Seasonal
    3. Syncope
    4. Focal
    5. Epileptic status
    6. Stroke
    7. Spinal
    8. Peripheral
    9. Drop-attack
    10. Acute

3. ***What is a seizure (select applicable)?***
    1. Clinical syndrome of rapid onset of focal or global cerebral deficit, lasting more than 24 hours, with no apparent cause other than a vascular one

2. Clinical syndrome doe to acute and severe inflammation of the brain parenchyma
3. Clinical expression of a sudden excessive discharge of neurons
4. Is a disease caused by the inflammation of the meninges covering the brain and spinal cord
5. Condition in which headache is a primary manifestation and no underlying disease is present

4. ***Indicate types of generalized seizures (select applicable):***
   1. Clonic seizures
   2. Simple partial seizures
   3. Absence seizures
   4. Tonic seizures
   5. Syncope
   6. Complex partial seizures
   7. Myoclonic seizures
   8. Partial seizures evolving to secondarily generalized seizures
   9. Tonic–clonic seizures
   10. Atonic seizures

5. ***Indicate types of partial seizures (select applicable):***
   1. Clonic seizures
   2. Simple partial seizures
   3. Absence seizures
   4. Tonic seizures
   5. Syncope
   6. Complex partial seizures
   7. Myoclonic seizures
   8. Partial seizures evolving to secondarily generalized seizures
   9. Tonic–clonic seizures
   10. Atonic seizures

6. ***Which type of seizures is characterized with presence of the following manifestations: sudden onset and offset of impaired responsiveness, staring, no aura, no postictal state, at times associated with automatic behaviors, EEG: generalized 3 Hz spike and wave pattern (select applicable)?***
   1. Clonic seizures
   2. Simple partial seizures
   3. Absence seizures
   4. Tonic seizures
   5. Syncope
   6. Complex partial seizures
   7. Myoclonic seizures
   8. Partial seizures evolving to secondarily generalized seizures
   9. Tonic–clonic seizures
   10. Atonic seizures

7. ***Which type of seizures is characterized with presence of the following manifestations: loss of consciousness and falling, body stiffens with subsequent jerking of extremities, cyanosis, excessive salivation, tongue or cheek biting, uncontrolled urination, retrograde amnesia (select applicable)?***
   1. Simple partial seizures
   2. Absence seizures
   3. Syncope
   4. Complex partial seizures
   5. Myoclonic seizures
   6. Partial seizures evolving to secondarily generalized seizures
   7. Tonic–clonic seizures
   8. Atonic seizures

8. ***Which type of seizures is characterized with presence of the following manifestations: sudden and brief contraction of a muscle or muscle group or several muscle triggered by cortical discharge, can be single or repetitive, recovery is almost immediate and patient often claims no loss of consciousness***
    1. Clonic seizures
    2. Simple partial seizures
    3. Absence seizures
    4. Tonic seizures
    5. Syncope
    6. Complex partial seizures
    7. Myoclonic seizures
    8. Partial seizures evolving to secondarily generalized seizures
    9. Tonic–clonic seizures
    10. Atonic seizures

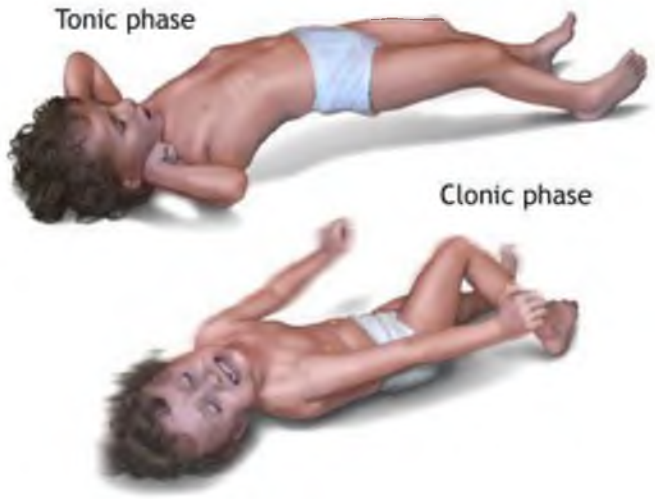
9. ***Indicate type of seizures with the following characteristics: seizure begins in specific region of cortex, produces signs and symptoms related to the function of the area of the brain involved (select applicable):***
    1. Clonic seizures
    2. Simple partial seizures
    3. Absence seizures
    4. Tonic seizures
    5. Myoclonic seizures
    6. Tonic–clonic seizures
    7. Atonic seizures

10. ***Which type of seizures is characterized with presence of the following manifestations: person experience unusual feelings or sensations that can take many forms, sudden unexplained feelings of joy, anger, sadness, or nausea, may hear smell taste see or feel things that are not real (select applicable)?***
    1. Clonic seizures
    2. Simple partial seizures
    3. Absence seizures
    4. Tonic seizures
    5. Syncope
    6. Complex partial seizures
    7. Myoclonic seizures
    8. Partial seizures evolving to secondarily generalized seizures
    9. Tonic–clonic seizures
    10. Atonic seizures

11. ***Metabolic derangements that can cause seizures (select applicable)?***
    1. Hyponatremia
    2. Hypernatremia
    3. Hypoglycemia
    4. Hyperglycemia
    5. Hypocalcemia
    6. Hypercalcemia
    7. Hypomagnesia
    8. Hypermagnesia
    9. Hypoosmolar state
    10. Hyperosmolar state

12. ***What does the work up for epilepsy include (select applicable)?***
    1. Labs: CMP, CBC with differential, toxicology screens, drug levels
    2. ECG, 24 hours holter monitor if indicated

3. CT scan/MRI
4. EEG
5. Neurological examination
6. Physical examination



13. Using the Figure from the left indicate type of seizure (select applicable):

1. Simple partial seizures
2. Absence seizures
3. Complex partial seizures
4. Myoclonic seizures
5. Tonic–clonic seizures
6. Atonic seizures

14. Indicate correct answers regarding state of consciousness in simple and complex partial seizure (select applicable):

1. Simple partial = Consciousness preserved
2. Simple partial = Consciousness impaired
3. Simple partial = Consciousness fluctuated
4. Complex partial = Consciousness preserved
5. Complex partial = Consciousness impaired
6. Complex partial = Consciousness fluctuated

15. Indicate common characteristics of simple partial seizures (select applicable):

1. Short in duration, impaired consciousness, tongue biting due to uncontrolled muscle contraction, incontinence, postictal state with confusion and exhaustion
2. Brief episodes of loss of attention or awareness lasting between 3 to 15 seconds without an aura or postictal state, may include chewing, lip smacking, swallowing, or facial twisting
3. Start with presentation of a motor, sensory, automatic, or psychic disturbance that manifests on the basis of the area of the origin in one cerebral hemisphere, involuntary motor activity of the face, limbs, or head; sensory symptoms of tingling, numbness, or pins and needles
4. Loss of consciousness may occur alone or in association with integrated purposeful movements like getting dressed or buttoning/unbuttoning or operating with a remote control

16. Indicate common characteristics of complex partial seizures (select applicable):

1. Short in duration, impaired consciousness, tongue biting due to uncontrolled muscle contraction, incontinence, postictal state with confusion and exhaustion
2. Brief episodes of loss of attention or awareness lasting between 3 to 15 seconds without an aura or postictal state, may include chewing, lip smacking, swallowing, or facial twisting
3. Start with presentation of a motor, sensory, automatic, or psychic disturbance that manifests on the basis of the area of the origin in one cerebral hemisphere, involuntary motor activity of the face, limbs, or head; sensory symptoms of tingling, numbness, or pins and needles; consciousness not impaired
4. Loss of consciousness may occur alone or in association with integrated purposeful movements like getting dressed or buttoning/unbuttoning or operating with a remote control

17. A 9-year-old boy is brought to your clinic by his parents because he has begun to have episodes of eye fluttering lasting several seconds. Sometimes he loses track of his thoughts in the middle of a sentence. There was one fall off a bicycle that may have been related to one of these events. There are no other associated symptoms, and the episodes may occur up to 20 or more times per day. The most likely patient's diagnosis is:

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Myoclonic seizures
6. Partial seizures evolving to secondarily generalized seizures
7. Tonic–clonic seizures
8. Atonic seizures

18. A 44-year-old man presents with left arm shaking. Two days ago, the patient noted left arm paresthesias along the lateral aspect of his left arm and left fourth and fifth fingers while he was reading. He thinks he may have been leaning on his left arm at the time; the symptoms resolved after 30 s. This morning, he noted the same feelings, lasting a few seconds, but then his 4th and 5th fingers started shaking rhythmically, and the shaking then migrated to all his fingers, his hand, and then his arm up to his elbow. This episode lasted a total of 30 s. The patient has never lost his consciousness. The most likely patient's diagnosis is:

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Myoclonic seizures
6. Partial seizures evolving to secondarily generalized seizures
7. Tonic–clonic seizures
8. Atonic seizures

19. A 31-year-old woman awoke feeling well. She was speaking with her bridegroom, went to the bathroom, and got back into bed. She had no headache, fever, chills, nausea, vomiting, or pain. Suddenly her body became stiff with arms flexed for a few seconds, followed by rhythmic jerking of both arms. Her legs were shaking, but less so. Her eyes were open, and she was foaming at the mouth. After 1 min, this stopped, and she initially did not recognize her bridegroom or his sister. She slowly returned to a normal level of consciousness over a 10-min period. She remembers events just prior to the episode, and she remembers being in the car on the way to the hospital. Her only medication is a multivitamin. Her examination is entirely normal. Routine labs and a brain MRI are normal. The most likely patient's diagnosis is:

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Syncope
6. Complex partial seizures
7. Myoclonic seizures
8. Partial seizures evolving to secondarily generalized seizures
9. Tonic–clonic seizures
10. Atonic seizures

20. Which type of seizures is characterized with presence of the following manifestations: jerking may begin in one area of the body, arm, leg, face, them move in steps "Jacksonian March"; no impairment of consciousness; may have unexplained sadness, fear, anger, joy; may have nausea, experience odd smells and ' funny" feeling in the stomach (select applicable):

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Syncope
6. Complex partial seizures
7. Myoclonic seizures
8. Partial seizures evolving to secondarily generalized seizures
9. Tonic–clonic seizures
10. Atonic seizures

21. Which type of seizures is characterized with presence of the following manifestations: starts with blank stare then by chewing, random activity like picking at clothes or objects; patient unaware of surrounding, dazed & mumbling; impairment of consciousness; unresponsive with actions that are clumsy, not directed; patient may experience structured hallucination (patient can see a scenery picture):

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Syncope
6. Complex partial seizures
7. Myoclonic seizures
8. Partial seizures evolving to secondarily generalized seizures
9. Tonic–clonic seizures
10. Atonic seizures

22. Which type of seizures is characterized with presence of the following manifestations: sudden cry, fall, stiffness followed by muscle jerks, cyanosis, loss of bladder or bowel control, impaired consciousness (select applicable):

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Complex partial seizures
6. Myoclonic seizures
7. Tonic–clonic seizures
8. Atonic seizures

23. Which type of seizures is characterized with presence of the following manifestations: patient has blank stare for a few seconds; brief Impairment or loss of consciousness; may be accompanied by rapid blinking; awareness returns after seizure; may result in learning difficulties if not treated (select applicable):

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Syncope
6. Complex partial seizures
7. Myoclonic seizures
8. Partial seizures evolving to secondarily generalized seizures

9. Tonic–clonic seizures
10. Atonic seizures

24. Which type of seizures is characterized with presence of the following manifestations: sudden brief massive muscle jerks; involve a part of the body or whole body; may cause patient to spill what they are holding or fall off chair (select applicable):

1. Clonic seizures
2. Simple partial seizures
3. Absence seizures
4. Tonic seizures
5. Syncope
6. Complex partial seizures
7. Myoclonic seizures
8. Partial seizures evolving to secondarily generalized seizures
9. Tonic–clonic seizures
10. Atonic seizures

25. Focal/partial seizures involving temporal lobe brain areas cause the following (select applicable):

1. Flushing and sweating
2. Arms and legs become stiff and move upwards
3. Cycling movements of the legs
4. Hearing, seeing, tasting or smelling things that aren't there
5. Rhythmic jerking of arms and legs
6. Feeling sad, happy, frightened or panicked
7. Swearing, shouting, screaming
8. Losing control of bladder/bowel
9. Feelings of detachment and deja vu

26. What is preferred choice of drugs for treatment of primary generalized tonic-clonic seizures (select applicable):

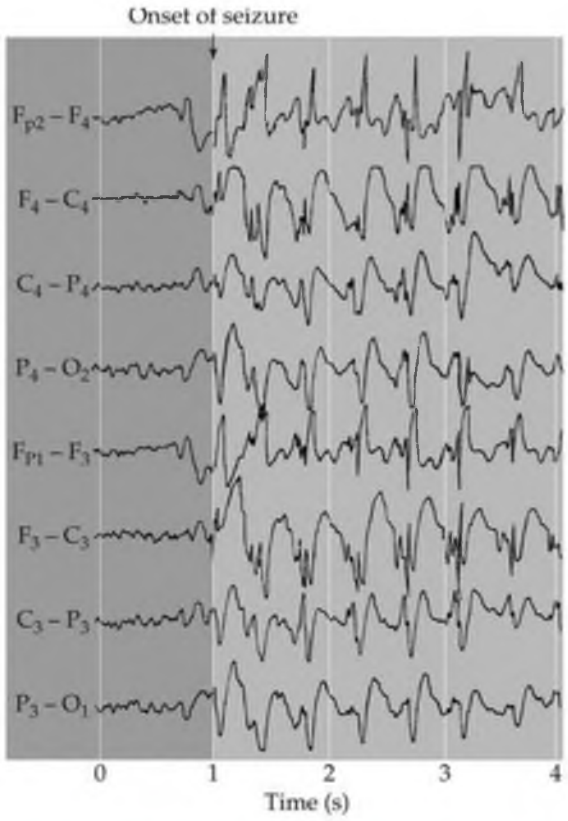
1. Topiramate
2. Carbamazepine
3. Clonazepam
4. Lamotrigine
5. Valproate
6. Levetiracetam
7. Oxcarbazepine
8. Phenobarbital
9. Ethosuximide
10. Primidone
11. Phenytoin

27. What is preferred choice of drugs for treatment of partial seizures (select applicable):

1. Topiramate
2. Carbamazepine
3. Clonazepam
4. Lamotrigine
5. Valproate
6. Levetiracetam
7. Oxcarbazepine
8. Phenobarbital
9. Ethosuximide
10. Primidone
11. Phenytoin

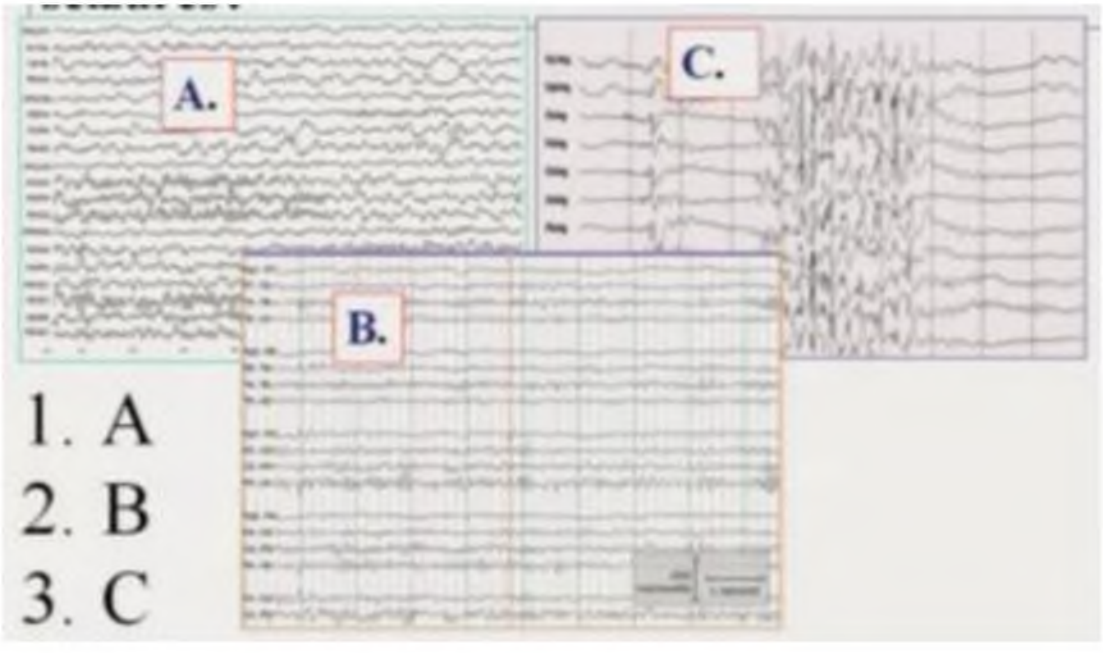
28. What is the 2nd choice of drugs for treatment of primary generalized tonic-clonic seizures (select applicable):

1. Topiramate
2. Carbamazepine
3. Clonazepam
4. Lamotrigine
5. Valproate
6. Levetiracetam
7. Oxcarbazepine
8. Phenobarbital
9. Ethosuximide
10. Primidone
11. Phenytoin



29. Using the Figure from the left indicate type of seizure (select applicable):

1. Seasonal
2. Syncope
3. Focal
4. Stroke
5. Generalized
6. Spinal
7. Peripheral
8. Drop-attack



30. Which one of the ECG traces from the left shows generalized tonic-clonic seizures?

1. A
2. B
3. C

Painful Syndromes of the Head And Neck

Headache can be either idiopathic or symptomatic. The most common ***idiopathic*** or "***primary***" types of headache are ***tension-type headache***, ***migraine***, and ***cluster*** headache. These three types of headache were once collectively designated "vasomotor headache." While migraine and cluster headache are typified by highly characteristic, usually unilateral attacks of pain, tension-type headache more commonly assumes the form of a diffuse, continuous headache of lesser intensity. ***Symptomatic*** headaches are, by definition, a manifestation of some other underlying condition. The possible causes include many types of neurological disease, as well as diseases of the eyes, teeth, jaw, ear, nose, and throat. Spondylogenic headache is caused by pathological processes in the cervical spine. Headache can also include a variably significant component of ***facial pain***—a typical example is cluster headache, in which the pain is felt mainly in the forehead, eye, and temple.

IHS Classification of Headache

The classification of headache syndromes proposed by the *International Headache Society* (IHS) has won general. For the beginning student of neurology, however, it is more useful to gain a descriptive overview of the more common, "classic" types of headache. In particular, he or she should learn to distinguish the common *idiopathic* types of headache, i. e., those not due to any demonstrable structural lesion in the head, from *symptomatic* types. The latter are caused by organic disease of the cranial vessels or other structures in the head. 90% of all cases of headache are idiopathic.

Approach to the Patient with Headache

The patient who goes to the doctor because of headache is suffering from pain and, often, anxiety. He or she therefore expects

_ to be taken seriously,

_ to be examined carefully,

_ to have the cause of the problem identified and clearly explained.

The physician must take the time needed to meet these expectations fully. The headache history. The clinical history is a vital step in the evaluation of headache (as of any other physical complaint). Some important points to be covered in the *systematic interview of the headache patient* are listed in Table 13.3. A carefully elicited history usually yields a clear-cut diagnosis. Nonetheless, the neurological and general physical examination (Table 13.4) should never be omitted, not least because these steps help the physician win the patient's confidence—an important factor for the success of treatment.

Table 13.3 **Headache history**

- **Family history** of headache?
- **How long** have headaches been present?
- **Nature of headache:**
  - site?
  - continuous or episodic?
  - usual or strange quality of pain?
  - timing of onset?
  - speed of development?
  - nature of pain?
  - precipitating factors?
  - duration of episodes?
  - accompanying signs?
- **Frequency?**
- **Headache-free intervals?**
- **Intensity,** impairment of activities at home and at work?
- **Medications and other counteractive measures:**
  - frequency
  - dose
  - efficacy
- **Other symptoms besides headache:**
  - ENT, eye, or dental disease?
  - memory?
  - neurological/neuropsychological deficits?
  - epileptic seizures?
  - general symptoms (fatigue, weight loss, circulatory problems, etc.)?
- **Personality:**
  - character?
  - occupation?
  - private life?
  - conflicts?
  - alcohol, tobacco, caffeine, drugs of abuse?
  - medications?

Table 13.4 **Examination of patients with headache**

General medical examination	**Neurological examination, with particular attention to:**
blood pressure	meningism
circulatory function	signs of intracranial hypertension
renal function	focal neurological signs
signs of infection	cranial nerve deficits
signs of meningitis	**Mental status, with particular attention to:**
signs of malignancy	psycho-organic syndrome
ENT diseases	neuropsychological deficit
eye diseases	impairment of consciousness
dental diseases, jaw diseases	psychological conflicts
cervical spondylosis	depression
	neurotic personality traits

Migraine

Migraine, a type of idiopathic headache, is the second most common type of headache overall, after tensiontype headache (see below). ***Migraine without aura*** (formerly called ***simple*** or ***common migraine***), whose sole neurological manifestation is headache, is distinguished from ***complicated migraine***, in which additional neurological manifestations are present.

Pathogenesis. Multiple factors contribute to the generation of a migraine attack:

_ ***Genetic factors*** play a role; in some patients, for example, there are well-documented ion channel abnormalities. Many patients report a history of migraine in their relatives, particularly on the maternal side.

_ ***Abnormal neural excitation in the diencephalon****, particularly in the* ***thalamic zone representing the trigeminal area***, also plays a role in the pathogenesis of migraine. Events occurring in this nuclear area are responsible for the triggering of (unilateral) migraine attacks by peripheral stimuli or emotional factors.

_ The pathogenetic role of so-called "***spreading depression***" is unclear. This is a phenomenon, known from animal research, in which a stimulus delivered locally to the occipital cortex induces a wave of excitation that spreads toward the frontal lobe. The excitation is then followed by reduced excitability ("depression"). It is an established fact that the speed of this disturbance, as it moves from back to front, correlates precisely with the speed of a scintillating *scotoma* moving across the visual field in an attack of ophthalmic migraine.

_ Finally, a number of ***biochemical processes*** in peripheral blood vessels, modulated by the trigeminovascular reflex, are another contributing factor. These include the *secretion of serotonin and histamine* by platelets and mast cells. A rise in serotonin concentration initially induces contraction of the cerebral vessels. At the same time, serotonin and histamine act together to increase capillary permeability. Plasma kinins penetrate the vessel wall and lower the pain threshold in the periarterial tissue. The vessels then expand again and, at the moment of vasodilation, the typical throbbing pain begins. Different types of serotonin receptors in the periphery and in the brain play a role in the generation of migraine. Many of the modern pharmacologic treatments for migraine exert their effects by influencing these processes.

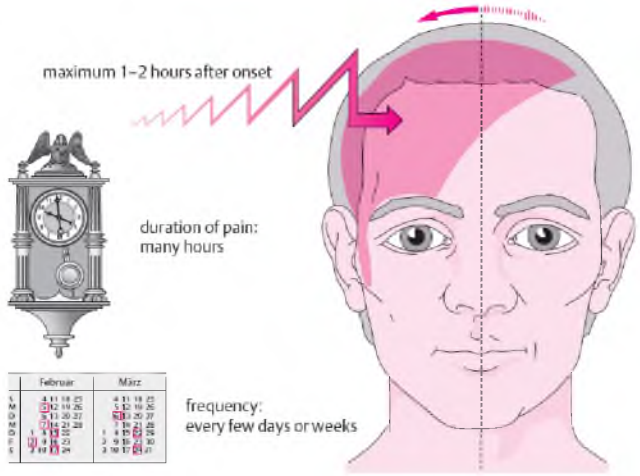
Migraine without Aura

An attack of simple (common) migraine develops without any premonitory symptoms (aura) and is characterized by headache and accompanying autonomic manifestations. About 70% of all migraine attacks are of this type.

Epidemiology. Women suffer from migraine more commonly than men. The initial attacks usually occur in the first or second decade of life; about 5% of all schoolchildren already suffer from true migraine. The overall prevalence of migraine in the population is estimated at 10%.

Clinical manifestations. The *headache* is unilateral (*hemicranial*) in two-thirds of patients and is usually located in the temporal and parietal areas. (The word "migraine" comes from the Latin "hemicrania.") In most patients, the attacks tend to affect one side much more often than the other, though contralateral headaches do sometimes occur. The pain occasionally migrates from one side to the other during an attack; it is primarily bilateral in about one third of all attacks. It rises to a maximum in one hour or a few hours and then generally persists for many hours after that. The *quality of the pain* is usually described as *pulsating and throbbing*; it worsens with any kind of physical exertion, even as mild as climbing a staircase. *Accompanying symptoms* are usually present: 60% of patients complain of nausea, anorexia, and intolerance of light, sound, and (frequently) odors. Patients are often irritable and in a bad mood during the attack. The *objective findings* may include pallor, diaphoresis (common), and sometimes tachycardia, vomiting, and/or diarrhea. The *frequency* of attacks varies from a few per year to several per week. The frequency and intensity of attacks determine the degree to which migraine affects the individual patient in his or her everyday activities; the impairment may be severe.

Fig. 13.1 **Migraine attack:** schematic diagram.



The IHS has promulgated the following defining criteria for simple migraine:

_ A: At least five episodes fulfilling criteria B through D, below.

_ B: The headache episodes last four to 72 hours (or, in children under 15 years of age, two to 48 hours), either when untreated or when treated unsuccessfully.

_ C: The headache has at least two of the following features:

_ 1. unilateral localization,

_ 2. pulsating character,

_ 3. moderate or marked intensity (makes everyday activities difficult or impossible),

_ 4. exacerbation by climbing stairs or other habitual physical activities.

_ D: At least one of the following symptoms is present during the headache:
_ 1. nausea and/or vomiting,
_ 2. abnormal sensitivity to light and noise.
The typical features of a migraine attack are shown schematically in Fig. 13.**1**.

Treatment. The treatment of common migraine depends on the frequency and severity of the attacks. If the attacks are rare and mild, treatment is generally not necessary. Headaches of intermediate severity, not very prolonged duration, and low frequency (less than once per week) can be managed by ***treating the individual attacks*** with a*nalgesic medications* in adequately high doses in the early stages of each attack, e. g., 1000 mg of acetylsalicylic acid. An antiemetic should be prescribed as well, e. g., 20 mg of metoclopramide, by mouth or, if necessary, as a suppository. If these measures do not adequately treat the headache, *triptanes* are given by mouth, or, if necessary, as a nasal spray or by injection. Some patients obtain relief with *ergotamines.* If the attacks occur more than once per week and/or severely hamper the patient's everyday activities by their severity or duration, ***attack prophylaxis*** can be initiated. Once begun, this must usually be maintained continuously for months or years afterward. Medications for attack prophylaxis include the beta-blocker propranolol and the tricyclic antidepressant amitriptyline, as well as valproate, dihydroergotamine, and flunarizine. These recommendations also apply to the various types of complicated migraine described below.

Types of Complicated Migraine

About one-third of all persons with migraine have additional neurological manifestations besides the headache itself, e. g., visual disturbances, paralysis, sensory abnormalities, vertigo, or abdominal or cardiac symptoms. These manifestations may be very dramatic, sometimes overshadowing the headache to such an extent that the patient's illness is not immediately recognizable as migraine.

Ophthalmic migraine, the most common and probably best-known type of migraine, is often called "classic migraine" in the English-speaking world. A scintillating *scotoma* appears 10 to 20 minutes before the headache. It begins in the center of the visual field, impairing the patient's ability to read (for example). A bright, zigzag line then travels from the center of the visual field outward in one-half of the visual field, until it "falls off the visual field" at the periphery, leaving behind a transient blurriness of vision in the corresponding hemifield. A typical scintillating scotoma or "fortification specter" (think of a crenellated medieval fortress) is shown in the diagram in Fig. 13.2.

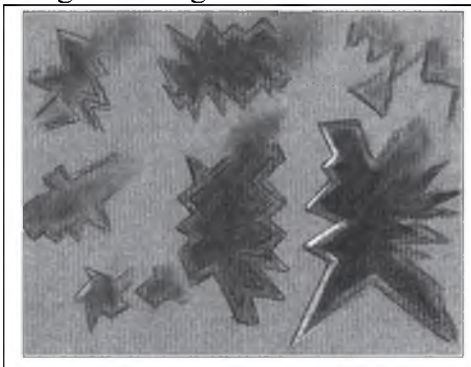


Fig. 13.2 **Scintillating scotoma** during an attack of ophthalmic migraine: typical fortification figures.

The scintillating scotoma is usually followed by an attack of *hemicranial headache.*

Occasionally, a scintillating scotoma may occur without subsequent headache; such events are called "*migraine sans migraine.*"

So-called ***retinal migraine*** is a very rare variant involving a vertically demarcated scintillating scotoma or a monocular visual disturbance lasting a few minutes.

Migraine accompagnee is characterized by the combination of headache with hemiparesis, a hemisensory deficit, or some other type of focal neurological or neuropsychological deficit. This variant of migraine usually has its onset in childhood or adolescence. In each attack, the affected individual progressively develops, for example, weakness or numbness and paresthesia on one half of the body, over the course of a few minutes. When the migrainous process is located in the left hemisphere, the patient can often become aphasic as well. The hemiparesis is never complete (i. e., never a hemiplegia) and the patient remains conscious. The headache usually comes after the neurological disturbances, but may also accompany or precede them, and it may be on the same or the opposite side. All manifestations resolve within a few hours, or in one or two days at most. Low-grade CSF pleocytosis and a focus of δ-activity in the EEG can often be demonstrated during an attack. SPECT, too, reveals focal changes. If there is no headache, one speaks of "migraine sans migraine" here aswell (just as in ophthalmic migraine without headache). With regard to *differential diagnosis*, an attack of migraine accompagnee can usually be distinguished from *an acute ischemic stroke* by the slow development of the neurological deficit and by the accompanying headache. In patients without headache, however, this distinction is not always easy to make, particularly in a first attack, and a thorough diagnostic evaluation is required.

Basilar migraine. In this type of migraine, the pathological process takes place in the *structures of the posterior fossa.* Basilar migraine mainly affects girls and young women. Its main manifestations are listed in Table 13.**5**: visual disturbances, symmetrical paresthesiae in the perioral region and the limbs, and impairment of consciousness. The accompanying headache is commonly felt at the back of the head.

Table 13.5 **Manifestations of basilar migraine (according to frequency)**, after Sturzenegger and Meienberg

Bilateral visual disturbance
• scintillating scotoma or elementary hallucinations
• diffuse loss of visual acuity
• transient amaurosis
• visual field defects
• dysmorphopsias
Nausea
Impairments of consciousness
• syncope
• confusion
• stupor
• amnesia
• coma
Paresthesiae (bilateral)
Vomiting
Dizziness
Gait ataxia
Dysarthria
Limb weakness

Other types. Two special types of migraine deserve mention. ***Familial hemiplegic migraine,*** caused by a hereditary disease of ion channels, generally appears in childhood and may be accompanied by cerebellar ataxia. Rarely, an attack of this type of migraine can be followed by a permanent neurological deficit. The rare ***alternating migraine of childhood*** begins in the first year of life and is associated with progressive psychomotor retardation. Attacks of hemiparesis on alternating sides of the body last from a quarter of an hour to several hours or even days. There may also be dystonic movements, nystagmus, or tonic crises. Naloxone and flunarizine are effective in treating this disorder.

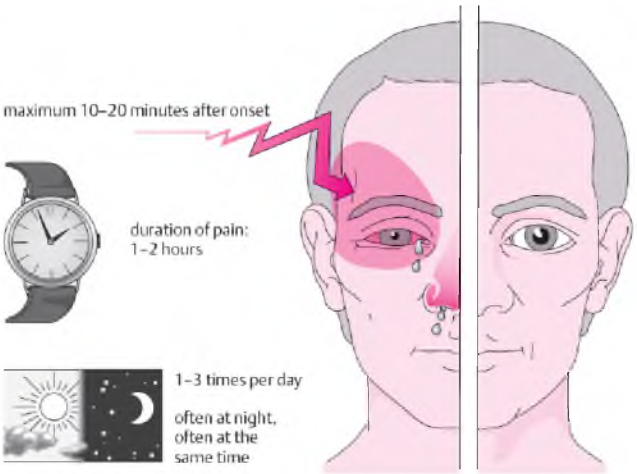
Cluster Headache

Alternative names for this disorder include "Bing–Horton neuralgia" and "erythroprosopalgia."

Etiology and pathogenesis. The appearance of cluster headache attacks in a circadian rhythm seems to imply that this disorder is due to a functional disturbance of the diencephalon. It shares a number of pathophysiological and biochemical features with migraine. In addition to idiopathic cluster headache, there are also symptomatic forms, caused, e. g., by mass lesions.

Clinical manifestations. These are highly typical and are illustrated in Fig. 13.**3**.

Fig. 13.3 **Cluster headache:** schematic diagram of an attack.



_ The *headache attacks* always occur on the same side of the head. The pain is mainly felt in the temple, eye, and forehead.

_ The pain reaches its maximum in 10–20 minutes. Each attack lasts from half an hour to two hours; attacks may occur "*on schedule*" at the same time of day every day (during a cluster), particularly at night.

_ *A number of attacks can occur "in series"* in a 24-hour period.

_ The *pain* is very intense, often *pulsating and throbbing.* The patient is agitated and paces around restlessly.

_ Attacks are usually accompanied by the following *objective findings*:

1) *Horner syndrome* of the ipsilateral eye.
2) A *red, teary eye* and periorbital erythema.
3) A stuffed nose and/or increased nasal secretion.

4) The attacks appear during periods called "clusters," lasting weeks or months; the clusters alternate with attackfree intervals lasting months or years.

Besides the ***episodic form*** just described, there is also a ***chronic form*** of cluster headache, in which the headache attacks occur every day and there are no attack-free intervals. It is not uncommon for typical migraine to be replaced by typical cluster headache (or vice versa) at some point in a patient's life. Many patients, too, have headaches bearing some of the features of both migraine and cluster headache.

Diagnostic evaluation. The acute attacks, because they are brief, are only rarely observed by the physician. The diagnosis thus depends on a *precise clinical history* (as it does in nearly all types of headache).

Treatment. ***Treatment of an acute attack*** is difficult. *Triptanes* can be given by subcutaneous injection, or the patient can be given *pure oxygen* to breathe (7 liters per minute for 15 minutes). Medications for the ***reduction of attack frequency*** include *verapamil* and *indomethacin*, sometimes in combination with a *tricyclic antidepressant.* A brief course of cortisone treatment is often effective. The chronic form responds to lithium.

Tension-type Headache

This is probably the most common type of headache. It was previously known as "vasomotor headache." Its clinical features resemble those of postconcussive headache.

Etiology. The cause of tension-type headache is unknown. It is no longer thought to be due to muscle tension. A current hypothesis attributes it to *abnormal sensitivity to pain in the trigeminal nuclear complex.* This complex, in turn, receives input from other structures in the brain, including the limbic system.

Clinical manifestations. Patients complain of a pressing, aching pain in the head, which is usually *diffuse*, i. e., there is no particular location at which it is most intense. It is of no more than *intermediate severity* and does not worsen with physical activity. It is not associated with nausea, photophobia, or phonophobia, and usually does not keep patients from going about their everyday activities.

There are ***episodic and chronic forms*** of tension-type headache. Patients with the episodic form suffer from headache on fewer than 15 days per month (180 days per year). The individual episodes of headache may last from 30 minutes to several days. Patients with the chronic form suffer from headache on more than 15 days per month (180 days per year).

Diagnostic evaluation. The *history* is crucial, as the neurological examination and ancillary tests do not reveal any abnormalities.

Treatment. Life-style readjustment is the recommended first line of therapy: removal of headache-producing substances such as alcohol and nicotine, regular physical exercise, regular sleep, stress reduction, and, if necessary, other changes in the patient's living situation and mode of living. If medications are needed, tricyclic antidepressants are the agents of choice, followed by beta-blockers or tizanidine.

Symptomatic Headache

Symptomatic headache is due to a ***structural lesion, infection, or inflammation of intra- and/or extracranial tissue***. Its direct cause is often a *pathological alteration of intracranial pressure (ICP)*, which excites nociceptive nerve endings in the meninges. The ICP may be either too high (particularly because of space-occupying lesions such as hematomas, tumors, and hydrocephalus) or too low (e. g., in the intracranial hypotension syndrome after a lumbar puncture). Symptomatic headache, however, is not necessarily due to neurological disease. ***Pathological conditions of the ears, nose, throat, eyes, teeth, or jaw*** can also cause head and/or facial pain, which may be quite severe. Even ***diseases of the cervical spine*** can, rarely, produce headache (*spondylogenic headache*). Table 13.7 below provides an overview of the major causes of symptomatic headache. We will describe a few of the
causative neurological illnesses and spondylogenic headache, in detail in the following paragraphs.

Occlusions and Dissections of Cranial Vessels

Arterial occlusion only rarely causes headache, but, if it does, the site of the headache may be a clue to the identity of the occluded vessel: occlusion of the internal carotid a. produces temporal headache, while occlusion of the basilar a. produces headache in a ringlike distribution around the head. Very intense pain on one side of the neck and face accompanies acute ***dissection*** of the internal carotid a. Vertebral artery dissection produces pain in the ipsilateral occiput and nuchal region. Dissections can arise spontaneously or after trauma to the head or neck.

Intracranial Hemorrhage

Subarachnoid hemorrhage (SAH) due to a ruptured saccular aneurysm at the base of the brain produces an extremely intense, diffuse headache of lightning-like onset ("*the worst headache of my life*"), possibly accompanied by meningismus and an impairment of consciousness. If the patient is comatose, of course, the headache and meningismus may be masked or absent.

Table 13.7 **Important types of symptomatic headache**

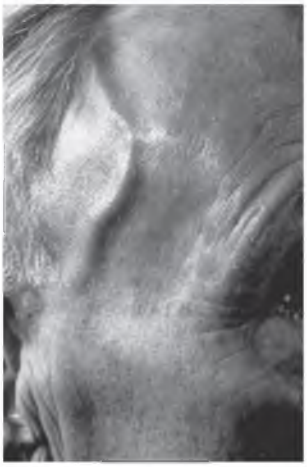
Type	Cause	Features
Headache of subarachnoid hemorrhage	usually rupture of a saccular aneurysm at the base of the brain	sudden, extremely severe headache, usually diffuse, accompanied by vomiting, drowsiness, and meningism
Headache due to intracranial mass	brain tumor, chronic subdural hematoma, brain abscess	permanent headache of increasing severity; nausea, bradycardia, papilledema, focal neurological deficits
Headache due to occlusive hydrocephalus	aqueductal stenosis, intraventricular mass, mass in posterior cranial fossa	manifestations like those of a brain tumor
Headache due to malresorptive hydrocephalus	prior subarachnoid hemorrhage or meningitis, venous sinus thrombosis	diffuse, increasingly severe headache, gait ataxia, incontinence
Intracranial hypotension	prior lumbar puncture, (rarely) spontaneous	orthostatic headache that improves or resolves when the patient lies down; normal neurological examination, CSF not obtainable by lumbar puncture (or only with aspiration); elevated CSF protein concentration
Pseudotumor cerebri	often seen in overweight young women; may be secondary to prior head trauma, anovulatory drugs, steroid withdrawal, tetracycline, etc.	chronic headache without any other detectable cause; often papilledema; CT or MRI reveals slit ventricles; elevated pressure on LP
Headache in meningitis	bacterial or viral meningitis	hyperacute in purulent meningitis; very severe headache, meningism, drowsiness, vomiting
Headache in carcinomatous or leukemic meningitis	primary tumors of various types, e. g., breast carcinoma	chronic, diffuse headache, cranial nerve deficits or spinal radicular deficits; LP and CSF cytology are essential
Postinfectious headache	after recovery from a (viral) infection	diffuse, often intractable headache without other neurological abnormalities, resembling tension headache; mild elevation of CSF cell count
Headache in ENT disease	chronic sinusitis, neoplasia in the pharyngeal cavity	headache or facial pain depending on the site of the disease process; no neurological deficit
Headache in eye disease	e. g., heterophorias, acute glaucoma, iritis, infectious/inflammatory processes in the orbit	usually frontal and temporal headache
Headache due to dental conditions	pulpitis, periodontitis, retained teeth, and myofascial pain syndrome due to malocclusion	severe, acute facial pain or chronic facial pain, depending on cause

Intracerebral hemorrhage (e. g., spontaneous hemorrhage due to hydrocephalus, into a tumor, or from a ruptured arteriovenous malformation) causes rapidly progressive headache in addition to hemiparesis and progressive impairment of consciousness. A more slowly progressive headache, perhaps accompanied by a fluctuating level of consciousness, is typical of ***chronic subdural hematoma***. Neurological deficits in these patients are surprisingly rare.

Cranial Arteritis

Cranial (also called temporal) arteritis is an autoimmune disease of blood vessels that almost exclusively affects persons over age 60. Its most prominent, though by no means only, clinical manifestations are in the large and middle-sized extracranial arteries.

Clinical manifestations. The leading symptom is an atypical headache that rapidly worsens over a few days or weeks and then becomes *constant*, *often in the temporal region*. The affected arteries (particularly the superficial temporal a.) are tender, tortuous, and swollen (see the image below).



They may occlude by thrombosis, in which case they cease to pulsate. The headache is often accompanied by further manifestations: pain in the shoulder and pelvic girdle muscles, fatigue, subfebrile temperature, weight loss, and nocturnal diaphoresis—a systemic syndrome known as *polymyalgia rheumatica.* The *complication* to be most feared is involvement of the ophthalmic a. causing occlusion of the central retinal a. and sudden blindness.

Painful Syndromes of the Face

Facial pain is often due to a ***lesion of a sensory nerve in the face,*** most commonly the trigeminal n. It typically presents with ***very brief, but very intense attacks of pain*** ("classic" or "genuine" neuralgia in the face). There are also a variety of other kinds of facial pain with other pathogenetic mechanisms, e. g., a structural anomaly of the jaw. The pain may resemble neuralgia in these other conditions as well; thus, patients with any kind of facial pain always require careful evaluation to establish the differential diagnosis.

"Genuine" Neuralgias in the Face

Typical manifestations of "genuine" neuralgia in the face are the following:

_ ***pain*** is located in the face or the mucous membranes of the head,
_ usually comes in brief attacks lasting a few seconds to a few minutes at most,
_ is usually very intense;
_ is described as electrical, knifelike, cutting, stabbing, or lightninglike;
_ arises either spontaneously or on provocation by touch or other mechanical or thermal stimuli;
_ is always on the same side of the face (in most patients);
_ and is always in the same location.
_ In addition, the ***attacks*** are very frequent, up to several times a day,
_ with no pain in between attacks, except, possibly, for a dull background pain.
_ Finally, there are ***no objective neurological abnormalities***, except in the rare forms of symptomatic neuralgia.

The localization and radiation of pain in the various types of neuralgia are depicted in Fig. 13.7.

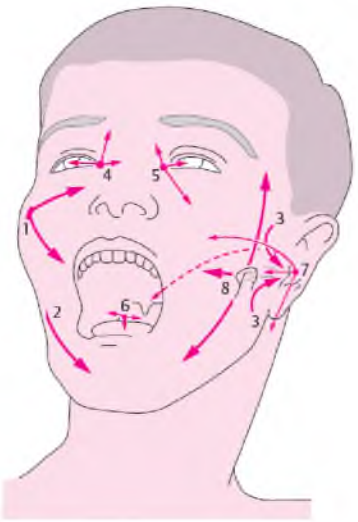


Fig. 13.7 **Localization of various types of facial pain and neuralgia.** 1 Trigeminal neuralgia in the distribution of the maxillary nerve (V_2). 2 Trigeminal neuralgia in the distribution of the mandibular nerve (V_3). 3 Auriculotemporal neuralgia. 4 Nasociliary neuralgia. 5 Sluder's neuralgia. 6 Glossopharyngeal neuralgia. 7 Neuralgia of the geniculate ganglion. 8 Temporomandibular joint "neuralgia" (myofacial pain syndrome).

Trigeminal Neuralgia

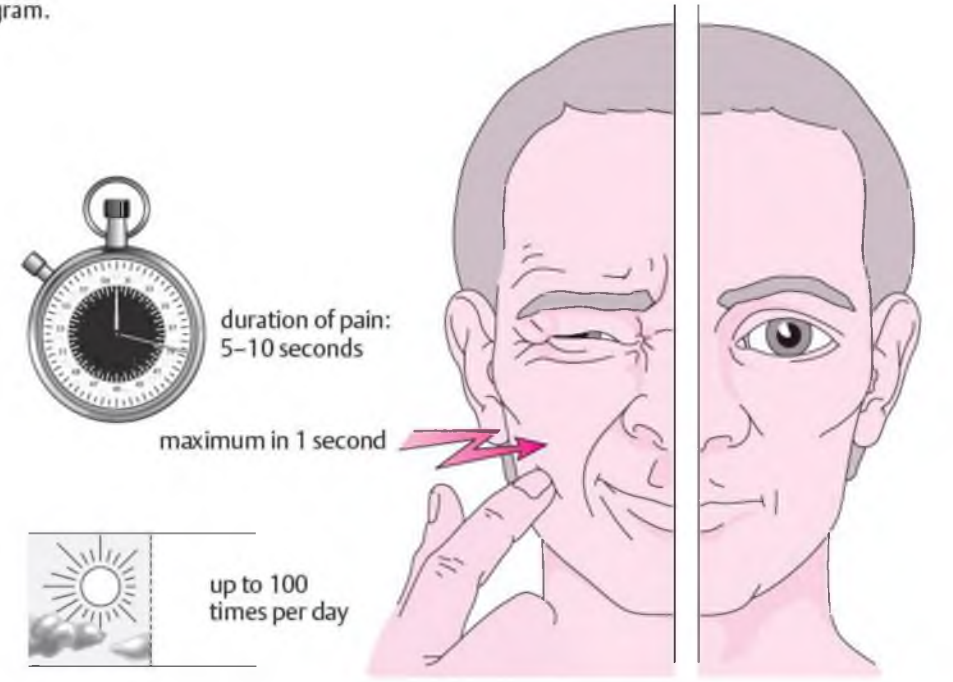
Idiopathic trigeminal neuralgia, which is far more common than the ***symptomatic*** type, only affects individuals over age 50.

Pathogenesis. Idiopathic trigeminal neuralgia appears to have more than one possible cause. In many patients, neuroimaging studies reveal a looping blood vessel that makes contact with the trigeminal n. at its zone

of entry into the pons. In other patients, defective myelin sheaths are found in the Gasserian ganglion (naturally only at postmortem examination). In symptomatic trigeminal neuralgia, on the other hand, the pain is due to an underlying neurological disease, e. g., multiple sclerosis.

Clinical manifestations. The painful attacks of idiopathic trigeminal neuralgia are usually located in the distribution of the *second trigeminal division* (the maxillary n.), less commonly in that of the first or third divisions. The pain is nearly always unilateral; it is felt on both sides simultaneously, or in alternation, in only 3% of patients. The individual attacks last only a few seconds and cause a reflexive grimace or pulling of the face ("*tic douloureux*").

Fig. 13.8 **Trigeminal neuralgia:** schematic diagram.



The pain is *unbearably intense*. The attacks occur spontaneously or on provocation by eating, speaking, tooth brushing, or touch; they may come dozens of times per day. Some patients eat and speak so little to avoid the pain that they lose weight, even to the point of cachexia. Attacks generally do not occur during sleep. The typical clinical manifestations of trigeminal neuralgia are depicted in Fig. 13.**8**. Sometimes, a long period with frequent attacks months until the attacks return.

In the rarer ***symptomatic*** cases of trigeminal neuralgia, the clinical manifestations are slightly different. The attacks are more commonly bilateral (either simultaneously or in alternation) and a neurological deficit may be found, depending on the underlying etiology, e. g., multiple sclerosis or a tumor compressing the trigeminal n.

Diagnostic evaluation. As stated above, the neurological examination generally reveals no abnormality. About one-quarter of patients who have suffered from idiopathic trigeminal neuralgia for a long time have a mild sensory deficit in the affected area of the face.

Treatment. About 80% of patients initially respond to correctly dosed treatment with *carbamazepine* or *gabapentin*. The medication must be taken every day and the dose must be steadily increased until the pain is relieved.

! The pharmacological treatment of trigeminal neuralgia often fails because of underdosing or irregular consumption of the prescribed drug.

If pharmacological treatment does not eliminate the pain despite a high dose of medication (in some patients, just below the threshold for intolerable side effects, which varies greatly from one patient to the next), neurosurgical treatment is indicated. The available, effective procedures include open microvascular decompression of the trigeminal n. (requires craniotomy) and percutaneous techniques such as selective radiofrequency coagulation of the Gasserian ganglion, balloon compression of the ganglion, and glycerol injection into Meckel's cave.

Auriculotemporal Neuralgia

In this disorder, the pain is located *in the temple and in front of the ear*. It is usually a sequela of disease of the ipsilateral parotid gland, appearing a few days or months after the parotid condition resolves. The attacks of pain can be provoked by chewing or by chemical stimuli, particularly sour (acidic) food. The pain is of a burning quality. It is often accompanied by erythema and increased sweating in the preauricular area. The differential diagnosis of this condition includes temporomandibular joint syndrome.

Glossopharyngeal Neuralgia

This condition is usually seen in the elderly. Its typical manifestations are lightninglike pain in the *base of the tongue*, the *hypopharynx*, and the *tonsillar fossa*, radiating toward the ear. The pain can be provoked by swallowing (especially of cold liquids), speaking, or sticking out the tongue. The painful attacks are, on rare occasions, accompanied by syncope. The pharmacological treatment of this disorder is like that of trigeminal neuralgia. If surgical treatment is required, resection of the glossopharyngeal n. and the upper root of the vagus n. has a good chance of success.

Vertigo

The vestibular organ (semicircular canals, saccule, and utricle) plays a central role in the regulation of balance. Disturbances of the vestibular apparatus (composed of the vestibular organ, the vestibulocochlear n., and the vestibular nuclei of the brainstem) cause dysequilibrium, the main symptom of which is vertigo.

It must be emphasized, however, that vestibular disturbances are just *one* cause of vertigo (see below) and not even the most common one.

Types of vertigo. **Directional vertigo** (vestibular vertigo) is characteristic of lesions of the peripheral portion of the vestibular apparatus, i. e., the vestibular organ and/or the vestibulocochlear n. The patient *perceives the environment as if it were in motion* (= *oscillopsia*), e. g., rotating or heaving up and down like the deck of a boat. Vestibular vertigo is often accompanied by *autonomic manifestations*, such as nausea and vomiting, and by *nystagmus*. Central vestibular lesions (i. e., lesions of the vestibular nuclei in the brainstem) also cause directional vertigo, which is generally less intense than that due to peripheral lesions. The autonomic manifestations, too, tend to be milder or absent.

Nonvestibular vertigo is nondirectional and often difficult for the patient to describe. The patient may report a woozy feeling, emptiness in the head, or darkness before the eyes. Oscillopsia is absent and there are usually no autonomic manifestations. Central nervous lesions can cause pathological nystagmus. Nonvestibular vertigo is caused either by a lesion of the nonvestibular parts of the regulatory system for balance, or else by faulty information processing within the central nervous system (e. g., because of a cerebellar lesion). Pathological processes outside the central nervous system, such as orthostatic hypotension or aortic stenosis, can also cause nonvestibular vertigo. The characteristic features of peripheral and central vestibular vertigo and of nonvestibular vertigo are summarized in Table 11.10 below.

Table 11.10 **Differentiation of peripheral vestibular, central vestibular, and nonvestibular vertigo**

	Type of vertigo		
Signs and symptoms	peripheral vestibular (labyrinth, nerve)	central vestibular	nonvestibular
Nausea, vomiting, diaphoresis	severe	moderate	mild
Vertigo—intensity	marked	moderate	mild
Vertigo—type	in a specific direction	directional to some degree	not in any specific direction
Nystagmus	spontaneous nystagmus of vestibular type	spontaneous nystagmus of vestibular type	nonvestibular nystagmus, or no pathological nystagmus
Hearing loss, tinnitus	usual	unusual	absent
Other neurological deficits	unusual	usually present	the neurological examination may or may not yield positive findings

Special aspects of history taking and diagnostic evaluation. The clinician should be able to tell whether the patient is suffering from *vestibular* or *nonvestibular vertigo* based on a meticulously elicited clinical history alone. It is also important to determine whether the vertigo is *episodic* or *continuous* and to ask about any *precipitating factors* (e. g., changes of position or particular situations that make the vertigo worse). If the vertigo worsens in the dark or when the patient's eyes are closed, the cause is likely to be a disturbance of proprioception (polyneuropathy, posterior column disease) or a bilateral vestibulopathy. The examiner should also always ask about *accompanying symptoms* (in particular, autonomic symptoms, tinnitus, hearing loss, and prior illnesses and infections). The history combined with the physical findings (nystagmus, results of balance tests, any other neurological abnormalities) usually allows localization of the functional disturbance. Further testing (e. g., neuroimaging of the head) mainly serves to determine the etiology.

Vestibular Vertigo

Acute loss of vestibular function is also called vestibular neuritis, acute vestibular neuropathy, or an acute vestibular crisis. It can be produced by a variety of pathogenetic mechanisms, the most common of which is a viral infection. The patient suddenly experiences *acute rotatory vertigo with nausea*, *vomiting*, and *falling to the*

side of the diseased vestibular organ. Every movement of the head makes the vertigoworse; therefore the patient, noting this, lies perfectly still. Examination reveals *horizontally beating, spontaneous nystagmus* in the direction opposite the side of the lesion, with a rotator component. The nystagmus is more intense when the patient lies on the affected side; it can be diminished by visual fixation. The affected vestibular organ is *less responsive than normal to caloric stimulation.* Vertigo usually resolves fully within a few days, rarely within a few hours. Often a so-called "trigger labyrinth" remains as a residual phenomenon, i. e., vertigo on acceleration or rapid movements of the head. The condition may relapse.

Positional and positioning vertigo. These types of vertigo arise only with certain positions or positioning movements of the head and manifest themselves as *brief attacks of vertigo* that diminish in intensity if they are provoked in rapid succession. These conditions have a number of different causes.

Benign paroxysmal positioning vertigo is the most common type of positioning vertigo. It is provoked by *changes in the position of the head*, usually involving lying down rapidly, bending forward, turning in bed, or rapidly sitting up. It manifests itself as very brief (15–30 seconds) and very severe *attacks of rotatory vertigo and nausea.* With respect to the *pathogenesis* of this condition, it is thought that small pieces of the otolith membranes of the saccule and utricle can break off and float freely in the endolymph—usually in the posterior semicircular canal, less commonly in the horizontal one. When the head is moved, these free particles move together with the endolymph and slide over the hair cells of the cupula, even after the movement is completed. The abnormally prolonged activation of the hair cells induces acute rotatory vertigo. The condition is also termed *cupulolithiasis* or *canalolithiasis.*

Meniere disease is a common cause of acute vestibular vertigo. It is caused by *endolymphatic hydrops* and manifests itself clinically in *episodes of acute rotatory vertigo*, a tendency to fall to the affected side, and horizontal, directional, *spontaneous nystagmus*, accompanied by *nausea*, *vomiting*, and *tinnitus.* Slowly progressive *hearing loss* is worse after each attack.

Bilateral vestibular deficits. While *unilateral* dysfunction of the vestibular apparatus can either recover or be compensated for by the intact opposite side within a matter of weeks, *bilateral* dysfunction deprives the regulatory system for balance of all incoming vestibular information. Consequently, the patient's gait becomes very unsteady in the dark (i. e., when visual input, too, is inoperative), or when the patient must walk on an uneven or soft surface (i. e., when the incoming proprioceptive information is difficult to interpret). Subjectively, the patient suffers from oscillopsia (apparent movement of the external world), particularly when walking.

Nonvestibular Vertigo

Dysfunction of the nonvestibular components of the regulatory system for balance can also cause vertigo.

_ ***Visually induced vertigo*** occurs, e. g., when an individual looks down from a great height, or when the incoming proprioceptive information is inconsistent with the visual information (*polysensory mismatch*). The vertigo of seasickness is a type of visually induced vertigo.

_ ***Impaired proprioception***, e. g., in polyneuropathy or posterior column disease, can also cause vertigo.

_ ***Cervical vertigo*** is thought to be due to faulty proprioceptive information arising in diseased cervical intervertebral joints or the adjacent soft tissues, which is then transmitted to the integrating apparatus for balance in the brainstem. This type of vertigo worsens in the dark. Its existence is debated.

_***Pathological processes affecting the central motor structures*** (e. g., paralysis, cerebellar or extrapyramidal disease, brainstem disorders) impair the patient's motor adaptation to changes in position, or cause oculomotor disturbances that can give rise to "dizziness."

_ ***Partial impairment of consciousness***, e. g., in presyncope or certain types of epilepsy (particularly temporal lobe epilepsy and absence seizures), is often experienced by the patient as "dizziness."

_ Another frequent occurrence is ***psychogenic vertigo***, particularly due to phobias, in the setting of depression, neurotic conflict situations, and panic attacks.

_ Finally, any ***general medical conditions*** that can temporarily diminish blood flow to the brain must be included in the differential diagnosis of "dizziness" and vertigo, e. g., arterial hypotension and heart disease.