Neurology

Telencephalon – Language Centers, Structure of the Limbic System and Basal Ganglia

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The language centers of the telencephalon, i.e. the Broca area and the Wernicke area, are highly relevant as examination topics. The same is true for the structure of the limbic system and the basal ganglia.

Language Centers of the Telencephalon

The language centers of the telencephalon include the **Broca area** (Brodmann area 44) and the **Wernicke area** (Brodmann area 22). The Broca area is located in the area of the **inferior frontal gyrus** of the **frontal lobe**, and the Wernicke area is located in the center of the **temporal lobe**.

**Note:** Both language centers are located only on the language-dominant hemisphere.
Function of the Broca’s Area

The Broca area is also referred to as the motor speech center because it governs the articulation of words and sounds. Studies suggest that, apart from language production, it is also associated with action processing and the sequencing of motor actions associated with verbal and nonverbal communication. Syntax is formed in the Broca area. Furthermore, the Broca area also manages the repetition of words via afferents from the secondary auditory cortex, which is located in the area of the temporal lobe and runs around the primary auditory cortex in the form of a horseshoe. The fibers of the secondary auditory cortex and the fibers of the secondary visual cortex reach the Broca area.

Function of the Wernicke’s area

The Wernicke area is also called the sensory speech area because it is responsible for the understanding of words, sentences, and requests. This area controls the process of phonologic retrieval, which is the process of sequencing the sounds of words before they are uttered. The arcuate fasciculus provides a connection between the Wernicke area and the Broca area.

Note: The Wernicke area is essential for speech comprehension.

Speech Disorders due to the Failure of a Language Centre (Aphasia)

Depending on which language center is affected, the symptoms of aphasia may vary. A common cause for aphasia is stroke, which in most cases (approximately 80%) is caused by ischemia.
Aphasia is defined as an acquired language disorder, and the cause of aphasia is centrally located. Language disorders must be distinguished from speech disorders (dysarthria), which have a variety of causes, such as cerebellar, bulbar, or extrapyramidal damage. The main forms of aphasia include Broca, Wernicke, amnesic, and global aphasia.

**Definition and Symptoms of Broca’s Aphasia (Motor Aphasia)**

Broca aphasia is caused by damage to the Broca area in the language-dominant hemisphere; it is also known as motor aphasia. It is characterized by a disturbance of the fluency of speech, which is slowed down considerably.

Patients usually speak only on request, and their speech is fragmented or telegram-like. They often speak in short, fragmented sentences that make sense but are uttered with great strain. Small words are often omitted. Patients with Broca aphasia may have weakness or paralysis of the right side of the body. However, patients can easily comprehend others’ speech.

In addition, phonemic paraphasia (discussed in the next section) can occur. Comprehension is hardly affected, but patients might have difficulty understanding long sentences and subordinate clauses. In some cases, reading, writing, and arithmetic abilities are also affected (alexia, agraphia, and acalculia).

**Definition and Symptoms of Wernicke’s Aphasia (Sensory Aphasia)**

Wernicke aphasia is also known as sensory aphasia. It is caused by damage to the temporal lobe within the area supplied by the posterior temporal artery.

The main symptoms of Wernicke aphasia are an increased output of speech (logorrhea) and phonemic and semantic paraphasias. A patient with phonemic paraphasia has trouble with the phonemic retrieval process and confuses syllables, changes the order of syllables, or shortens words, whereas a patient with semantic paraphasia confuses entire words. The confused words often come from the same subject area—for example, substituting “arm” for “leg.”

The increased speech output involves, among other things, making up new words (neologisms). In contrast to Broca aphasia, speech comprehension is disturbed, which additionally complicates the patient’s ability to communicate. Writing and reading are usually disturbed as well.

**Definition and Symptoms of Global Aphasia**

In global aphasia, both the Wernicke and Broca areas are affected, as is the basal ganglia region. This results in a complete loss of speech. Patients express themselves only through sounds and, possibly, isolated words like “yes” or “no.” Patients can barely understand spoken language, and they cannot write or read. However, intellectual and cognitive abilities are preserved. This form of aphasia is mainly caused by an extensive media infarction of the language-dominant hemisphere due to stroke or brain trauma. If the basal ganglia are also affected, additional unilateral symptoms, such as a form of...
This is the most severe type of aphasia, and the chances of improvement depend on the damage caused to the brain. If the damage is not very extensive, improvement may occur in 5 to 6 months; otherwise, long-term disability may remain.

**Definition and Symptoms of Amnesic Aphasia**

In patients with amnesic aphasia, there is not one single damaged area but, rather, **multiple smaller lesions** in the area of the **temporal** and **parietal lobes** of the language-dominant hemisphere.

Clinical presentation of amnesic aphasia involves significant word-finding problems, for which patients may try to compensate with phrases and circumlocutions. The spontaneous flow of speech, however, is not affected.

**Other Symptoms Associated with Damage to the Dominant Hemisphere**

In addition to the forms of aphasia described, damage to the dominant hemisphere may lead to other clinical signs, such as apraxia or agnosia.

**Definition of Apraxia**

Apraxia describes the inability to adapt behavior to specific purposes and situations. Based on clinical appearance, multiple manifestations can be distinguished, including ideomotor and ideational apraxia. Individuals with **ideomotor apraxia** have damage in the **Wernicke area** and the **primary motor cortex**, whereas those with **ideational apraxia** have damage in the area of the **temporoparietal junction**.

Clinical presentation of ideomotor apraxia includes a distortion of aiming movements, movement sequences, facial expressions, and gestures. Patients with ideational apraxia are able to perform individual movements correctly but cannot implement them in a more complex sequence.

**Definition of Agnosia**

Agnosia makes it impossible to recognize certain things, despite intact senses. Different forms of agnosia are distinguished by what types of things cannot be perceived. One form of agnosia is **visual agnosia**, where patients cannot recognize an object despite intact vision. When touching the same object, however, they can easily name it.

Further examples include **prosopagnosia**, which describes the inability to recognize people and their faces; **autotopagnosia**, which is characterized by the inability to localize parts of the body; and **anosognosia**, which describes the inability to perceive one’s own body ailments or neurologic dysfunctions. Patients with anosognosia, for instance, cannot recognize hemiparesis.
Other Symptoms Associated with Damage to the Non-Dominant Hemisphere

Damages to the nondominant hemisphere can cause, among other things, the so-called neglect syndrome.

Definition of Neglect Syndrome

Neglect is a motor or sensory inattention to one half of the body. Unilateral neglect can be detected with the clock-drawing test, in which a patient is asked to draw a clock face with the clock hands pointing to a certain time. Most patients who have a unilateral neglect will draw all of the numbers in one half of the clock.

Structure of the Limbic System

The limbic system consists of several components. These are the hippocampus, parahippocampal gyrus with entorhinal cortex (see above), cingulate gyrus, amygdala, and mammillary bodies.

Function of the Limbic System

The limbic system serves a variety of functions. These include the following:

- Social cognition
- Control of emotions like fear, rage, and tranquility
- Memory
- Processing of olfactory senses
- Sexual behavior
- Control of appetite and eating patterns
- Sleep and dreams (processing unconscious emotions and conscious thoughts and forming the base of dreams)
- Control of addiction and motivation pathways
- Control of autonomic and endocrinal response to emotions

Among other functions, the limbic system is responsible for the development of affective and instinctive behavior, sexual functions, and the formation of memory contents. However, the limbic system is not the only system involved with these functions; rather, they result from an interaction of several brain areas.

The individual structures of the limbic system can be assigned to specific functions. The hippocampus, for instance, is the primary site of memory formation; in addition, behavior, awareness, and motivation originate here.

Alongside the hippocampus, the other components of the Papez circuit are involved in memory formation. These include, for example, the mammillary bodies of the limbic system, which also mediate affective and sexual behavior.

Modulation of the autonomic system and development of motivation, among other things, take place in the cingulate gyrus.

The amygdala, which is located in the temporal lobe below the caudate nucleus, plays an important role in the development of behaviors and the storage of memory contents associated with emotions. The most relevant emotion here is the feeling of fear. Therefore, the amygdala is sometimes referred to as the ‘fear center.’ If there is damage in the area of the amygdala, it may cause misjudgment of a dangerous situation and, as a result, risky behavior.

In addition, the amygdala is involved in the modulation of autonomic hypothalamic areas, which may lead to, for example, an accelerated heart rate in scary situations.

**Structure of the Basal Ganglia**

The basal ganglia, which are also known as basal nuclei, include the caudate nucleus, putamen, and globus pallidus. They are present in pairs, one on each of the two hemispheres. An evolutionary particularity of the globus pallidus is that it developed in the diencephalon but shifted to the telencephalon.
The basal ganglia are part of the gray matter of the telencephalon and are located inside it.

On a cross section of the brain, the head of the caudate nucleus (caput nuclei caudati) is located laterally on both sides of the anterior cornu of the lateral ventricles. The tail of the caudate nucleus (cauda nuclei caudati) lies above the posterior cornu of the lateral ventricle. The putamen and globus pallidus are located lateral to the thalamus, with the globus pallidus being more medial.
The **internal capsule** runs between the thalamus and the caput nuclei caudati as the medial border and the globus pallidus and putamen as the lateral border (see “Structure of the Internal Capsule” section). The white matter of the internal capsule separates the nucleus caudatus from the putamen and creates a striped appearance, which led to the name ‘**corpus striatum**’ for the caudate nucleus and putamen.

The corpus striatum is especially well defined in the frontal section. Here, the close relationship of the nucleus caudatus and the ventriculus lateralis becomes apparent. Cranially, the lateral ventricle is adjacent to the corpus callosum. Lateral to the putamen is the **capsula externa** and the **claustrum**, which is believed to be involved in sexual arousal.

**Function of the Basal Ganglia**

The basal ganglia are part of the **extrapyramidal motor system**, modulating the effect of voluntary fine motor movements.

**Projections of the Basal Ganglia**

The projections of the basal ganglia that mediate motor information run in a loop, which is called a **basal ganglia circuit**, or motor circuit. Within this circuit, information is transported from the basal ganglia to the **premotor cortex** of the telencephalon. This information then returns from the **primary motor cortex** of the telencephalon to the basal ganglia.
The feedback of the information is carried out by the **thalamus**. In addition to the thalamus and the basal ganglia, the **subthalamic nucleus** and **substantia nigra** also form part of the basal ganglia circuit.

In the basal ganglia circuit, there is a direct pathway and an indirect pathway:

- In the **direct pathway**, the *putamen* projects to the *globus pallidus internus* and to the reticular part of the *substantia nigra*. Because of the transmitter γ-aminobutyric acid (*GABA*), the putamen has an inhibiting effect on the 2 mentioned projection areas. This eliminates the inhibitory effects of the globus pallidus and substantia nigra to the thalamus, and therefore the latter can act in an excitatory manner on the motor cortex.

- In the **indirect pathway**, the putamen has an inhibitory effect on the *globus pallidus externus*, which usually has an inhibitory effect on the *subthalamic nucleus*. By eliminating this inhibitory effect, the subthalamic nucleus acts with the transmitter *glutamate* in an excitatory manner on the globus pallidus externus, the globus pallidus internus, and the reticular part of the substantia nigra. The **reticular part** of the *substantia nigra* and the *globus pallidus internus* act as inhibitors on the thalamus so that it does not promote any motor movement.

In summary, the direct pathway acts in an excitatory manner and the indirect pathway in an inhibitory manner on the execution of movements.
Diseases Associated with the Basal Ganglia

Based on their symptoms, diseases of the basal ganglia can be differentiated into hyperkinetic and hypokinetic disorders. The hyperkinetic disorders include Huntington disease and hemiballismus; the best known of the hypokinetic forms is Parkinson disease.

Parkinson’s Disease

Parkinson disease shows damage in the area of the pars compacta of the substantia nigra. Death of dopamine-generating neurons occurs, which leads to an interruption of the inhibitory effect of the substantia nigra on the direct pathway of the basal ganglia circuit. Because of this disinhibition, the indirect pathway becomes more active and inhibits the thalamus, which leads to a reduction in movement.

Definition

Parkinson disease is a movement disorder caused by an insufficient amount of dopamine, a chemical produced by the neurons.

Clinical features

Parkinson disease is characterized by a typical clinical triad consisting of akinesia,
rigidity, and tremor. Sometimes, postural instability is included with the main symptoms as well. The tremor is typically a resting tremor that begins mostly on one side (i.e. it is asymmetric). Poor balance and coordination are marked.

The patient’s gait pattern provides a first indication of the disease. As part of hypokinesia or akinesia, the gait is characterized by short steps and a forward inflection (propulsion) with a reduced swinging of the arms, which is often more pronounced on one side as well. As the disease progresses, other symptoms appear, such as sleep disorders; depression; and difficulty chewing, swallowing, and speaking.

Symptoms are:
- Bradykinesia or akinesia
- Rigidity
- Resting tremor of the hands and fingers
- Difficulty initiating movements
- Movements that are abnormally slow
- Decreased facial expression

Structural changes and causes:
- Depigmentation of the substantia nigra
- Lewy bodies
- Causes include genetics, environment, certain medications, 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine (MPTP), and vascular insult

Diagnosis

Diagnosis is made on the basis of clinical presentation, history, and neurologic examination.

Treatment

The treatment of Parkinson disease involves different classes of drugs. These include anticholinergics, levodopa, N-methyl-D-aspartate (NMDA) receptor antagonists, dopamine agonists, monoamine oxidase B (MAO-B) inhibitors, or catechol-O-methyltransferase (COMT) inhibitors.

Recommendations for the initiation of therapy depend on the age of the patient. For patients younger than 70 years, initiation of therapy with a dopamine agonist is recommended; for patients 70 years and older, L-DOPA (levodopa) is recommended (Parkinson guideline of the German Society of Neurology, last updated September 2012). However, the treatment regimen has to be individually adjusted for each patient in accordance with his or her dominant symptoms.

Huntington’s Disease (Chorea)

Definition

Huntington disease involves atrophic lesions in the area of the basal ganglia, corpus striatum, and, in a later stage of the disease, cerebral cortex. The disease shows an autosomal dominant inheritance pattern and generally manifests in the third to fifth decades of life.

Pathophysiology/Clinical Features
In Huntington disease, there is a decrease in GABA concentration due to atrophy of the corpus striatum, and this leads to an imbalance between GABA and dopamine. Dopamine acts in support of the direct pathway and inhibits the indirect pathway of the basal ganglia circuit, which leads to increased activation of the thalamus and the motor cortex.

The resulting spontaneous movements are noticeable, primarily in the face and distal extremities. These repeated movements lead to increased caloric consumption, and patients often present as cachectic. Food intake is even more difficult when the caudal cranial nerve is affected and the chewing and tongue muscles are impaired.

Extrapyramidal movement disorder often follows previous psychopathologic changes, which can manifest 10–15 years before in the form of disturbed motivation and emotion or psychosis. In the course of Huntington disease, the development of dementia is rather common.

**Symptoms are:**
- Chorea
- Athetosis
- Personality changes
- Dementia

**Structural changes and causes:**
- Loss of GABAergic, medium size, spiny neurons from striatum
- Ventricular enlargement
- Genetic cause - autosomal dominant
- Excessive CGA repeats

**Treatment**

The progression of the disease cannot be influenced therapeutically; that is, there is no way to treat the cause. However, symptoms can be treated with medication, physiotherapy, and ergotherapy. For example, sulpiride can be used to treat hyperkinesia.

**Hemiballismus**

Hemiballismus is usually caused by damage in the area of the subthalamic nucleus. Clinical signs of hemiballismus include suddenly emerging proximal flinging movements.

There is no known cure. Treatment approaches include drugs such as neuroleptics, valproic acid, and, sometimes, benzodiazepines.

**Fibre System of the Telencephalon**
The fibers of the telencephalon are divided into 3 classes, depending on their function: the **commissural**, **association**, and **projection fibers**.

The **commissural fibers** connect certain areas of both hemispheres with one another, thus enabling an exchange between them. The majority of commissural fibers are located in the **corpus callosum**, which also connects the 2 hemispheres anatomically.

In contrast to the commissural fibers, the **association fibers** connect individual parts of the same hemisphere.

Projection fibers connect the cortex with the **subcortical areas**, including the thalamus and the basal ganglia. The majority of these fibers run through the **internal capsule** and the **external capsule**.

**Structure of the Internal Capsule**

The internal capsule is divided into 3 sections: the **crus anterius** (anterior limb), the **crus posterius** (posterior limb), and the **genu** (bend). Specific tracts project through each section, which are also organized somatotopically.

The **frontopontine tract** and the **anterior thalamic peduncle** run through the crus
anterius. In the genu of the internal capsule, there are parts of the tracts of the **dorsal thalamic peduncle**; the remaining parts of these tracts pass through the crus posterius. In addition, the crus posterius contains the **fibrae corticonuclearis**, **fibrae corticospinalis**, **tractus temporopontinus**, and **radiatio optica**.

**Somatotopy of the Internal Capsule**

**Note:** The corticospinal fibers are arranged somatotopically in the crus posterius of the internal capsule.

**Function of a Commissurotomy (Split-Brain)**

Separation of the commissural fibers within the **corpus callosum** is called commissurotomy. This interrupts the communication between the 2 hemispheres. This surgical intervention is used in **epilepsy treatment** as a last resort. The complete sectioning of the corpus callosum prevents generalization, that is, the propagation of an epileptic seizure to both hemispheres.

**References**


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