MINISTRY OF HEALTH OF UKRAINE

ODESA NATIONAL MEDICAL UNIVERSITY

Faculty of Medicine №2

Department of Neurology and Neurosurgery

APPROVED BY
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TEACHING MATERIAL FOR PRACTICAL CLASSES ON THE ACADEMIC SUBJECT

Faculty, Course: Stomatological, 3d year
Academic Discipline: Neurology

Approved by:
Meeting of the Department of Neurology and Neurosurgery
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PRACTICAL CLASSES

Practical Class No. 1

Theme: The principles of the structure and functioning of the nervous system. Voluntary movements and their disorders. Pyramidal system. Symptoms of central and peripheral paresis.

Actuality of theme:
Paralyses (disorders of movement functions) are the common signs of the lesion of the nervous system, which need the correct diagnosis and treatment.

Aims of the class:
Educational aims:
- to acquaint students with the defeat of movement functions, create imagination about forming of different types of paralyses.
- **a student must know:**
1) Movement pathways
2) Clinical signs of paralyses
3) Terminology of violations of movements
- **to give possibility to capture skills to the students:**
1) To explore derivative patient.
2) To explore the volume of self-willed motions.
3) To explore muscular force
4) To explore trophic of muscules
5) To explore muscular tone
- **to give to the students** of ability clinically to explore functions of movement spheres in patients and to diagnose central and peripheral paralyses

Educational aims connected with:
- forming of professional approach to diagnosis of various violations of motion
- the actual aspects of deontological, patriotic, professional, psychological, legal, ecological responsibility and others like that.

Table of contents of the class:

Neurology is a science about the development and functioning of the nervous system (NS) in norm and pathology. The problem of research of human brain, the problem of the ratio between the brain and mentality are the most important tasks which were considered by the science.

Modern neurology (neuropathology) as an independent science was established in the second half of the 19th century and it is a result of thousand-year work of many talented observers of antiquity; namely doctors, biologists, physiologists, morphologists, who studied the role of the NS in normal and unhealthy conditions of an organism.

Early history.

An ancient Egyptian treatise concerning trauma surgery, the Edwin Smith papyrus, contains descriptions and suggests treatments for various injuries, including some of neurological nature. Specifically, there are descriptions of the meninges, the external surface of the brain, the cerebrospinal fluid and the intracranial pulsations. Not only are these neurological features mentioned, but it is also noticed that some bodily functions can be impaired by brain injuries or injuries to the cervical spine.

There are many other examples of observations of neurological phenomena throughout history. The Sumerians illustrated paraplegia caused by physical trauma in a bas relief of a lion

with an arrow in its back. Neurological disorders not caused by physical disorder were also investigated. For example in the medicine of the Vedic period of ancient India, the Ayurvedic text Charaka Samhita discusses epilepsy, with a discussion of both symptoms and of possible treatments. Slightly later, the ancient Greek physician Hippocrates was convinced that epilepsy has a natural cause, not a sacred one.

The ancient Greeks also dissected the nervous system. For example, Aristotle (although he misunderstood the function of the brain) describes the meninges and also distinguishes between the cerebrum and the cerebellum. Slightly later, in Rome, Galen performed many dissections of the nervous system in a variety of species, including the ape. One particular discovery he made was of the importance of the recurrent laryngeal nerves. Originally, he cut through them accidentally while performing an experiment on the nerves that control breathing by vivisection of a strapped-down, squealing pig. The pig immediately stopped squealing, but continued struggling. Galen then performed the same experiment on a variety of animals, including dogs, goats, bears, lions, cows and monkeys, finding similar results each time. Finally, to publicise this new result, Galen demonstrated the experiment on a pair of pigs to a large audience in Rome, telling them: "there is a hairlike pair [of nerves] in the muscles of the larynx on both left and right, which if ligated or cut render the animal speechless without damaging either its life or functional activity"

Anatomy and physiology.

Along with most other sciences, the first real advances in neurology after the Greeks occur in the Renaissance. The invention of the printing press allowed the publication of anatomical textbooks,pages, allowing the dissemination of knowledge. An early example is Johann Peyligk's Compendium philosophiae naturalis, published in Leipzig, Germany in 1499. This work contained 11 woodcuts, depicting the dura mater and pia mater as well as the ventricles.

A revolution took place in both neurology in particular and in anatomy in general when Andreas Vesalius published his De humani corporis fabrica in 1543. It includes detailed images depicting the ventricles, cranial nerves, pituitary gland, meninges, structures of the eye, the vascular supply to the brain and spinal cord, and an image of the peripheral nerves. Vesalius also exposed the non-existence structures that had been believed to be in the brain since Galen's (revered) work, such as the rete mirabile. Galen's dissections were all on animals – in particular, the rete mirabile is only well developed in ungulates. Vesalius, unlike many of his contemporaries, did not subscribe to the then common belief that the ventricles were responsible for brain function, arguing that many animals have similar systems of ventricles to those of humans, but had no true intelligence. It appears that he rarely removed the brain from the skull before cutting it, most of his diagrams showing the brain sitting inside a severed head.

Thomas Willis in 1664, published his Anatomy of the Brain, followed by Cerebral Pathology in 1676. He removed the brain from the cranium, and was able to describe it more clearly, setting forth the circle of Willis – the circle of vessels that enables arterial supply of the brain. He had some notions as to brain function, including a vague idea as to localization and reflexes, and described epilepsy, apoplexy and paralysis. As already mentioned, he used the word neurology.

A beginning of the understanding of disease came with the first morbid anatomists, morbid anatomical illustration, and the development of effective colour printing. Matthew Baillie (1761–1823) and Jean Cruveilher (1791–1874) illustrated the lesions in stroke, in 1799 and 1829 respectively.

Microscopy.

Only when cells were identified microscopically was it possible to progress beyond the crudest anatomical notion. J.E. Purkinje (1787–1869) in 1837 gave the first description of neurones, indeed a very early description of cells of any kind. Later Golgi and Cajal stained the ramifying branches of nerve cells; these could only touch, or synapse. The brain now had demonstrated form, without localised function. The famous philosopher René Descartes (1596–1650) speculated that every activity of an animal was a necessary reaction to some external

stimulus; the connection between the stimulus and the response was made through a definite nervous path. Luigi Galvani (1737–1798) demonstrated that electrical stimulation of nerve produced muscle contraction, and the competing work of Charles Bell (1774–1842) and Francois Magendie (1783–1855) led to the view that the ventral horns of the spinal cord were motor and the dorsal horns sensory. A hemiplegic patient who could not speak led Paul Broca (1824–1880) to the view that functions in the cerebral cortex were anatomically localised. Ivan Pavlov (1849–1936) realised as his dogs dribbled that a simple reflex could be modified by higher brain functions. These neurological ideas were coordinated and integrated by the neurophysiologist Charles Scott Sherrington (1857–1952).

Diagnostics.

Physicians could use the ideas of neurology in practice only if they developed proper tools and procedures for clinical investigation. This happened step by step in the 19th century – tendon hammer, ophthalmoscope, pin and tuning fork, syringe and lumbar puncture. X rays, the electro-encephalography, angiography, and CAT scans were to follow. The clinical neurologists correlated their findings after death with those of the neuropathologist. The best known was W.R. Gowers (1845–1915) who owned a major text in two volumes, of a cerebrospinal tract. By the end of the nineteenth century, the connection was established between stroke and hemiplegia, between trauma and paraplegia, between the spirochaete and the paralysed demency people who filled the mental hospitals. The first chemotherapeutic cure of a serious infection was salvarsan for syphilis, followed by the induction of fever in neurosyphilis. The treatment of neurosyphilis became highly effective when antibiotics were introduced.

Neuropathology (neurology) as an independent clinical science appeared in 1862 when the department for patients with diseases of the NS in Salpetrier hospital near Paris was opened. Jean-Martin Sharko (1835-1893) whom we quite often we name the father of neuropathology headed it.

The first Kiev Department of neurology was founded in 1884 by M.M. Lapinsky. The first Kharkiv Department of neurology was founded in 1884 by P.I. Kovalevsky (S.M. Davidenkov, K.I. Platonov, O.M. Grinshtein, G.D. Leshchenko their mantle had fallen). In 1922 Ukrainian Research Institute of psychiatry and neurology was founded in Kharkiv by G.I. Geimanovitch. In 1926 the Ukrainian Institute of Clinical Psychiatry was founded by V.P. Protopopov. Now its headed by professor P.V. Voloshin.

The new stage of the development of neurology was opened by Marushi. He described the reaction of the activation of the brainstem through the implanted electrodes.

The newest period has been since the 60s-70s of the 20th century. The ultra structure of synapses has been discovered, the theory of ligand-synaptic communications has been specified. In peripheral NS albuminous or axonal transport has been found.

The elements of the nervous system (NS) of the person develop from embryonal ectoderm (neurons and neuroglia) and mesoderm (meninges, vessels, mesoglia). By the end of the 3rd week of the development of human embryo the neural plate, which is located longitudinal on the back side of embryo, is formed from ectoderm. On the neural plate a neural groove appears which turns to the neural tube then. In the generated neural tube three layers are distinguished: the inner ependymal layer; the middle layer of the mantle and the outer layer. The cells of the inner layer turn into glia cells, the cells of the mantle layer give neuroblasts, which turn to mature nervous cells and spongioblasts, giving rise to various kinds of neuroglia (astrocyte, oligodendrocyte).

The NS develops from the (initially) longitudinally oriented neural tube, which consists of a solid wall and a central fluid-filled cavity. The cranial portion of the neural tube grows more extensively than the rest to form three distinct brain vesicles, the rhombencephalon (hindbrain), the mesencephalon (midbrain), and the prosencephalon (forebrain).

The NS is composed of cells, called neurons that are specialized for information processing and transmission. Neurons make contact with each other at junctions called synapses, at which information is transferred from one neuron to the next by means of chemical messenger substances called neurotransmitters. In general, neurons can be divided into two classes: excitatory and

inhibitory. The structure of neurons is various. There are numerous classifications of nervous cells based on the form of their body, extent and the form of dendrite and other features. On functional value these cells are subdivided into motor, sensory and interneuron.

A nervous cell carries out two basic functions: 1) specific – processing the information coming to the neuron and transferring of a nerve impulse; 2) biosynthetic, directed on the maintenance of the ability to live. It is reflected in the ultra structure of the nervous cell.

The structure of a nervous cell includes: mitochondrion, determining its energy metabolism; nucleus, nucleolus, granular and not granular endoplasmic reticulum, Golgi complex, polysomes and ribosomes, basically providing synthesis of protein; lysosomes and phagosomes (the basic organelles of «endocellular digestive system»); axons, dendrites, synapses, providing morphofunctional connection of separate cells. The polymorphism of the cell structure is due to the various role of separate neurons in system activity of the brain as a whole. The axons are exposed to myelinization and in such a way myelinated nerve fibers are formed. The fascicles of nerve fibers which can include separate non-myelinated fibers form the white substance of the brain, the cranial and peripheral nerves. The dendrites and the processes of glia cells form complex and unique pictures of the neuropile. However it is the allocation of the axons and the dendrites, their interposition, afferent-efferent interrelation and the regularity of architectonics of the synapses that are determining in mechanisms of the integral function of the brain.

The synapses are divided into axosomatic, axoaxonic and axodendritic synapses; dendrodendritic synapses are observed less often. In the synapses they distinguish presynaptic part, which contains the vesicles and postsynaptic part. The active zone of synaptic contact where the release of mediator and nerve impulse transmission take place is characterized by the increase of the electronic density of presynaptic and postsynaptic membranes. Different systems of interneuronic connections use various mediators. It is the essential moment in the synaptic transmission. Now about 30 chemically active substances (acetylcholine, dopamine, noradrenaline, serotonin, GABA (gamma aminobutyric acid) and etc.), which take part in synaptic transmission from one cell to another are known. Recently numerous neuropeptides such as enkephalins, endorphines and P-substance are actively studied as mediator in synaptic transmission.

The reflex principle of the work of the NS underlies the functioning of the motor system of the person. The complex of nervous mechanisms takes part in the realization of motor reflex activity: peripheral motor neuron (the cells of anterior horns of the spinal cord and nuclei of the cranial nerves), central motor neuron - or the pyramidal tract, and also numerous structures of the brainstem and the extrapyramidal system, which provide the automatism of stereotyped movements, smooth regulation of muscular tone, the realization of consensual movements. All these movements give individual character to the motor activity of a person. The cerebellum with its system of afferent and efferent tracts plays an important role in the development of automatic coordination of movements, the maintenance of equilibrium of a body.

Thus motor activity is the result of integral activity of the NS. They distinguish voluntary movement which is realized with the help of the cerebral cortex, and involuntary, which is constructed as simple and complex reflex acts. Voluntary movements are controlled by the pyramidal tract, involuntary – the extrapyramidal system, the reticular formation and the segmental apparatus of the spinal cord.

The NS is divided into central and peripheral. Roots, plexuses and nerves belong to peripheral NS. The central nervous system consists of the brain and the spinal cord.

Central Nervous System (CNS)

The forebrain or prosencephalon (supratentorial portion of the brain) comprises the telencephalon (the two cerebral hemispheres and the midline structures connecting them) and the diencephalon. The midbrain or mesencephalon lies between the fore brain and the hind brain. It passes through the tentorium cerebelli. The hindbrain or rhombencephalon (infratentorial portion

of the brain) comprises the pons, the medulla oblongata (almost always called “medulla” for short), and the cerebellum. The mid brain, pons, and medulla together make up the brain stem.

Spinal Cord

The spinal cord is approximately 45 cm long in adults. Its upper end is continuous with the medulla; the transition is defined to occur just above the level of exit of the first pair of cervical nerves. Its tapering lower end, the conus medullaris, terminates at the level of the L3 vertebra in neonates, and at the level of the L1–2 intervertebral disk in adults. Thus, lumbar puncture should always be performed at or below L3–4. The conus medullaris is continuous at its lower end with the threadlike filum terminale, composed mainly of glial and connective tissue, which, in turn, runs through the lumbar sac amidst the dorsal and ventral roots of the spinal nerves, collectively called the cauda equine (“horse’s tail”), and then attaches to the dorsal surface of the coccyx. The cervical, thoracic, lumbar, and sacral portions of the spinal cord are defined according to the segmental division of the vertebral column and spinal nerves.

Peripheral Nervous System (PNS)

The PNS connects the central nervous system with the rest of the body. All motor, sensory and autonomic nerve cells and fibers outside the CNS are generally considered part of the PNS. Specifically, the PNS comprises the ventral (motor) nerve roots, dorsal (sensory) nerve roots, spinal ganglia, and spinal and peripheral nerves, and their endings, as well as a major portion of the autonomic nervous system (sympathetic trunk). Peripheral nerves may be purely motor or sensory but are usually mixed, containing variable fractions of motor, sensory, and autonomic nerve fibers (axons). A peripheral nerve is made up of multiple bundles of axons, called fascicles, each of which is covered by a connective tissue sheath (perineurium). The connective tissue lying between axons within a fascicle is called endoneurium, and that between fascicles is called epineurium. Fascicles contain myelinated and unmyelinated axons, endoneurium, and capillaries. Individual axons are surrounded by supportive cells called Schwann cells. Tight winding of the Schwann cell membrane around the axon produces the myelin sheath that covers myelinated axons. The Schwann cells of a myelinated axon are spaced a small distance from one another; the intervals between them are called nodes of Ranvier. The nerve conduction velocity increases with the thickness of the myelin sheath.Nokia 225 Dual SIM

Autonomic Nervous System (ANS)

The autonomic nervous system regulates the function of the internal organs in response to the changing internal and external environment. ANS realize its functions independently of consciousness. It consists in two major parts: the sympathetic and parasympathetic systems. According to the anatomy it divided into central part, which include suprasegmental level (limbic system, hypothalamus, reticular formation) and segmental level and peripheral part (all the other structures).

Cerebral Ventricles and Cisterns

The fluid-filled cerebral ventricles constitute the inner CSF space. Each of the two lateral ventricles communicates with the third ventricle through the interventricular foramen of Monro (one on each side). Fluid passes from the third ventricle through the cerebral aqueduct (of Sylvius) into the fourth ventricle, and thence through the single midline foramen (of Magendie) and paired lateral foramina (of Luschka) into the subarachnoid space (outer CSF space). Dilatations of the subarachnoid space are called cisterns. The cerebellomedullary cistern (cisterna magna) lies between the posterior surface of the medulla and the undersurface of the cerebellum.The cerebellopontine cistern occupies the cerebellopontine angle. The ambient cistern lies lateral to the cerebral peduncle and contains the posterior cerebral and superior cerebellar arteries, the basal vein, and the trochlear nerve. The interpeduncular cistern lies in the midline betweenthe cerebral peduncles and contains the oculomotor nerves, the bifurcation of the basilarartery,andtheoriginsof

thesuperior cerebellar and posterior cerebral arteries; anterior to it is the chiasmatic cistern, which surrounds the optic chiasm and the pituitary stalk. The portion of the subarachnoid space extending from the foramen magnum to the dorsum sellae is collectively termed the posterior cistern.

Cerebrospinal Fluid (CSF)

The CSF, a clear and colorless ultrafiltrate of blood plasma, is mainly produced in the choroid plexus of the cerebral ventricles and in the capillaries of the brain. It normally contains no red blood cells and at most 4 white blood cells/μl. Its functions are both physical (compensation for volume changes, buffering and equal distribution of intracranial pressure despite variation in venous andarterialblood pressure)andmetabolic (transport of nutrients and hormones into the brain, and of waste products out of it). The total CSF volume in the adult is ca. 150ml, of which ca. 30 ml is in the spinal subarachnoid space. Some 500ml of cerebrospinal fluid is produced per day, corresponding to a flow of ca. 20 ml/h. The normal pulsation of CSF reflects brain pulsation due to changes in cerebral venous and arterial volume, respiration, and headmovements. AValsalva maneuver increases the CSF pressure.

CSF circulation

CSF formed in the choroid plexus flows through the ventricular system and through the foramina of Magendie and Luschka into the

basal cisterns. It then circulates further into the spinal subarachnoid space, over the surfaces of the cerebellum and cerebrum, eventually reaching the sites of CSF absorption. It is mainly absorbed through the arachnoid villi (arachnoid granulations, pacchionian corpuscles), which are most abundant along the superior sagittal sinus but are also found at spinal levels. CSF drains through the arachnoid villi in one direction, from the subarachnoid space to the venous compartment, by a valve mechanism. This so-called bulk flow is apparently achieved witht heaid ofpinocytotic vacuoles that transport the CSF, and all substances dissolved in it, in ladlelike fashion. At the same time, CSF diffuses into the brain tissue adjacent to the CSF space and is absorbed by the capillaries.

Blood is pumped from the left ventricle of the heart to the aortic arch and thence to the common carotid arteries and anterior circulation of the brain (internal carotid, middle cerebral, and anterior cerebral arteries), and to the subclavian arteries and posterior circulation of the brain (vertebral, basilar, and posterior cerebral arteries). The anterior circulation supplies the eyes, basal ganglia, part of the hypothalamus, the frontal and parietal lobes, and a large portion of the temporal lobes, while the posterior circulation supplies the brain stem, cerebellum, inner ear, occipital lobes, the thalamus, part of the hypothalamus, and a smaller portion of the temporal lobes. Venous blood from the superficial and deep cerebral veins drains via the dural venous sinuses into the internal jugular veins and thence into the the superior vena cava and right atrium.

The Reflexes

The investigation of the reflexes is often considered to be the most important part of the neurologic examination. The testing of the reflexes is the most objective procedure of the neurologic examination. Reflex activity is essential to the normal functions of the human body. All involuntary and many voluntary acts are reflexes in nature. A reflex is an invariable adaptive response to the stimulation of a sense organ, which involves the use of a center of adjustment and of the conductors necessary to connect this center with the appropriate receptor and effector apparatus. By neurologycal examination mainly involuntary reflexes are investigated. An intact sensory system and an intact motor system are needed for a normal reflex response, and knowledge of both sensory and motor functions is necessary to understanding of reflex action. The stimulus is received by the receptor, which may be a sensory ending in the skin, mucous membranes, muscle, tendon, or periosteum etc. The stimulation of the receptor initiates an impulse that is carried along the afferent (sensory) nervous fibers, and then is transmitted to the CNS. There a synapse takes place with the intercalated neuron, which relays the impulse to the center of adjustment, the cell body of the efferent neuron. The neuraxis of the efferent neuron transmits the

impulse to the effectors (the cells, muscles, glands, or blood vessels that then respond). A disturbance in function of any of the above parts of the reflex arc will cause a break in the reflex arc and a consequent decrease or loss of the reflex. Some hundreds of reflexes have been identified; only the more important ones will be described.

The Muscle Stretch (Proprioceptive or Deep) Reflexes

The muscle stretch reflexes are those that are elicited in response to application of the stimulus to either tendons or periosteum, or occasionally to bones, joints, fascia, or aponeurotic structures. Because the stimulus is mediated through the deeper sense organs such as the neuromuscular and neurotendinous spindles, they may be referred to as the proprioceptive (or deep) reflexes. The proprioceptive reflexes are best tested by the use of a rubber percussion hammer.

The stimulus should be quick and direct, and should be a threshold one, and no greater than necessary. The patient should be comfortable and relaxed. The part of the body to be tested should be in a position for optimal muscular response. In order to compare the reflexes on the two sides of the body, the position of the extremities should be symmetric. Reflexes may be classified as normal, absent, sluggish (diminished), and exaggerated. The response should always be compared on the two sides of the body; unequal reflexes may be as significant as either increased or diminished reflexes.

The Orbicularis Oculi (blinking) Reflex. Percussion at the outer aspect of the supraorbital area and other kinds of irritation are followed by a reflex contraction of this muscle, with resulting closing of the eye. The response is usually bilateral. The afferent portion of the arc may be carried through the trigeminal nerve; the efferent impulses pass through the facial nerve, and the reflex center is in the pons.

The Jaw (Masseter or Mandibular Reflex). To elicit the jaw reflex the examiner places his index finger over the middle of the patient's chin, holding the mouth slightly opened and the jaw relaxed. Then he taps his finger with the reflex hammer. The response is a contraction of the masseter and temporal muscles, causing a sudden closing of the mouth. The afferent impulses of this reflex are carried through the sensory portion of the trigeminal nerve, and the efferent impulses through its motor portion; the reflex center is in the pons.

The upper extremities. The Biceps Reflex. The arm is held in a relaxed position, with the forearm midway between flexion and extension and in slight pronation. The examiner places his thumb or finger over the biceps tendon and taps the thumb with a reflex hammer. The major response is a contraction of the biceps muscle with flexion of the forearm. The sensory supply of this reflex is through the midcervical nerves, and the motor supply to the biceps is through the musculocutaneous nerve. The reflex center is at the CV-CVI segments.

The Triceps Reflex is elicited by tapping the triceps tendon just above its insertion on the olecranon process of the ulna. The arm is held midway between flexion and extension, and it may be rested on the examiner's hand or on the patient's thigh. The response is one of contraction of the triceps muscle, with extension of the forearm. The sensory and motor innervations are through the radial nerve, and the center is in the lower cervical portion of the spinal cord (CVI-CVIII).

The Brachioradialis (Radial Periosteal or Supinator) Reflex. If the styloid process of the radius is tapped while the forearm is in semi-flexion and semipronation, there will be flexion of the forearm, together with supination. The supination is more marked if the forearm has been extended and pronated, but there is less flexion. If the reflex is exaggerated there is associated flexion of the wrist and fingers, with adduction of the forearm. The innervation of this reflex is through the radial nerve and spinal segments CV-CVI.

The lower extremities. The Patellar (Quadriceps) Reflex. The patellar, or quadriceps, reflex, usually called the knee jerk, is characterized by contraction of the quadriceps femoris muscle, with resulting extension of the leg, in response to a stimulus directed toward the patellar tendon. The patellar reflex is innervated by the femoral nerve and spinal segments LII-LIV.

This reflex may be elicited with the patient seated in a chair with his feet resting on the floor or with the patient lying in bed or by having the patient sit with one leg crossed over the other and tapping the patellar tendon of the superior leg. Reinforcement of the patellar reflexes may be carried out according to the method of Jendrassik: on testing reflex the patient is asked to hook the flexed fingers of the two hands together, placing the palmar surfaces of the fingers of one hand against the palmar surfaces of the other, and to attempt to pull them apart at the time the reflex is being stimulated.

The Achilles (Triceps Surae) Reflex. The Achilles, or triceps surae, reflex, or the ankle jerk, is obtained by tapping the Achilles tendon just above its innervation on the posterior surface of the calcaneus. This is followed by contraction of the posterior crural muscles, the gastrocnemius, soleus, and plantaris, with resulting plantar flexion of the foot at the ankle. If the patient is seated or is lying in bed, the thigh should be moderately abducted and rotated externally, the knee should be flexed, and the foot should be in moderate inversion; the examiner should place one hand under the foot to produce moderate dorsiflexion at the ankle. If it cannot be elicited in this manner, the patient should be asked to kneel on his knees on a chair, while the feet project at right angles; the Achilles tendons are percussed while the patient is in this position. The Achilles reflex is innervated by the tibial nerve and LV and SI-SII spinal segments.

The muscle stretch reflexes are increased with lesions of the corticospinal or pyramidal system. These changes are due to involvement of a variety of structures in the descending motor pathways at cortical, subcortical, midbrain and brain stem levels as well as in the spinal cord. The flexor reflexes are exaggerated to a greater degree in the upper extremities, and the extensor reflexes in the lower.

The superficial (cutaneous) reflexes

The superficial reflexes are those that are elicited in response to the application of a stimulus to either the skin or mucous membrane, they are sometimes known as exteroceptive reflexes.

The Corneal Reflex. To elicit the corneal reflex, the examiner touches the cornea lightly with a wisp of cotton or a piece of a thin paper to avoid irritating the cornea. In response to this stimulus there is a blinking, or closing of the ipsilateral eye, the direct corneal reflex, and also a closing of the opposite eye, the consensual corneal reflex. The afferent portion of the reflex arc is mediated by the ophthalmic division of the trigeminal nerve, whereas the efferent or motor response is a function of the facial nerve. The reflex center is in the pons.

The pharyngeal, or gag, reflex is elicited by applying a stimulus, such as a tongue blade or an applicator, to the posterior pharyngeal wall, tonsillar regions or even the base of the tongue. If the reflex is present, there will be elevation and constriction of the pharyngeal musculature. The afferent impulses of the reflex arc are primarily carried through the glossopharyngeal, the efferent elements primarily through the glossopharyngeal and vagus nerve. The reflex center is in the medulla.

The palatal or uvular reflex is tested by stimulating the lateral and inferior surface of the uvula, or soft palate, with a tongue blade or a cotton applicator. Elevation of the soft palate and retraction of the uvula occur simultaneously. The center for this reflex is also in the medulla. Both the sensory and the motor portions of the reflex arc are carried through the vagus and glossopharyngeal nerves.

The superficial abdominal reflexes. Gentle stroking of the abdomen or scratching it with a blunt object is followed by homolateral contraction of the abdominal muscles and retraction or deviation of the linea alba and umbilicus toward the area stimulated. These reflexes should be tested with the patient recumbent and the abdominal wall thoroughly relaxed.

The Upper Abdominal Reflex is elicited by stimulating the skin of the upper abdominal quadrants, usually in a diagonal fashion, downward and outward from the tip of the sternum. This reflex is innervated by the intercostal nerves from ThVI-ThVIII.

The Middle Abdominal Reflex. Stimulation of the skin of the abdomen at the level of the umbilicus, either by a horizontal stimulus, starting externally and proceeding medially is followed by a lateral deviation of the linea alba and umbilicus. This reflex is innervated by the intercostal nerves from ThIX-ThX.

The Lower Abdominal Reflex. This is elicited by stimulating the skin of the lower abdominal quadrants, either diagonally in an upward and outward direction from the region of the symphysis pubis. There is a contraction of the abdominal muscles and a diagonal deviation of the umbilicus toward the site of the stimulation. This reflex is innervated by the lower intercostal and the iliohypogastric and ilioinguinal nerves ThXI-ThXII. These reflexes may be difficult to obtain or absent in obese individuals and those with relaxed abdominal walls, and in women who have borne children. In pathology the absence of superficial abdominal reflexes is a significant finding.

The Cremasteric Reflex is elicited by stroking the skin on the upper, inner aspect of the thigh, from above downward, with a blunt point, or by pricking or lightly pinching the skin in this area. The response consists of a contraction of the cremasteric muscle with homolateral elevation of the testicle. This reflex may be absent in elderly males, in individuals who have a hydrocele or varicocele, and in those who have had orchitis or epididymitis. The innervation is through ilioinguinal and genitofemoral nerves LI-LII.

The Plantar Reflex. In the normal individual, stimulation of the plantar surface of the foot is followed by plantar flexion of the toes. It innervated by the LV-SII segments by means of the tibial nerve.

The Superficial Anal Reflex consists of a contraction of the external sphincter in response to stroking or pricking the skin or mucous membrane in the perianal region. This reflex is innervated by the inferior hemorrhoidal nerve SIV-SV.

Abnormalities of the superficial reflexes

The superficial reflexes are either diminished or absent in the event of a disturbance in the continuity of the reflex arc: in the afferent nerve, motor center, or efferent nerve. The superficial reflexes, however, especially the abdominal and cremasteric reflexes, have a special significance when their absence is associated with an exaggeration of the deep reflexes (dissociation of reflexes) or when they are absent in instances where signs of corticospinal tract involvement are elicited (since the superficial reflexes have, in addition to a spinal reflex arc, a superimposed cortical pathway).

All movements are effected by contractions of striated muscles through the control of the NS. Thus motor activity is the result of integral activity of the NS. They distinguish voluntary movement which is realized with the help of the cerebral cortex, and involuntary, which is constructed as simple and complex reflex acts. Voluntary movements are controlled by the pyramidal tract, involuntary – the extrapyramidal system, the reticular formation and the segmental apparatus of the spinal cord. By means of our motor system we move our bodies in space. The motor system controls the timing, direction, amplitude, and force of movement.

The examination of motor functions includes the determination of muscle power, an evaluation of muscle tone and muscle bulk and the observation of abnormalities of movement.

The motor impulses for voluntary movement are mainly generated in the precentral gyrus of the frontal lobe (first motor neuron). They pass in the long fiber pathways (mainly the corticonuclear and corticospinal tracts (pyramidal pathway) through the brainstem and down the spinal cord to the anterior horn, where they make synaptic contact with the second motor neuron—usually by way of one or more intervening interneurons.

Pyramidal System.

The pyramidal system appears only in mammals (in the elephant – 4,8 %, in the primates - 20,1 %, in the person - 30 %). Pyramidal system consists from two neurons - central (the first) and peripheral (the second) ones. The pyramidal tract (the one forming a pair) basically begins from Betz (large pyramidal) cells, in the 3rd and 5th layers of the precentral gyrus (the 4th Brodmann's

area). Some part of the fibers begins from the neurons of the 1st somatosensory area (the postcentral gyrus; the 3rd, 1st, 2nd Brodmann's areas); the part - from the neurons of superior and middle frontal gyri (the 6th and the 8th areas).

In the precentral gyrus there is a determined somatotopical distribution of the central motoneurons (homunculus). The motoneurons are located both on convex, and on the internal surface of hemispheres. In the brain stem, the pyramidal tract gives off fibers to the motor nuclei of the cranial nerves (corticopontine and corticobulbar tracts). Its function is the realization of voluntary movements of muscles of face, pharynx and larynx. The corticospinal tract begins from the top and average third of the precentral gyrus and ends along the whole length of the spinal cord at the motor neurons of the anterior horns. This tract realizes voluntary movements of the muscles of the limbs and trunk. The axons of the pyramidal tract pass through the radiate crown to the anterior two thirds of the posterior limb of the internal capsule (the posterior third is occupied by sensory and cerebellar tracts; while the anterior limb of the internal capsule – by frontopontine tracts).

Further the pyramidal tract passes into the basis of the mesencephalon, goes into the base of the brain and the medulla oblongata where it forms "pyramids". After that the corticonuclear tract branches off, the majority of the remaining fibers of the corticospinal tract (85 %) at the level of the great occipital foramen cross over to the other side and descend in the lateral white column of the spinal cord and end at the level of the anterior horns: for hands - at the level of the cervical intumescence, for legs -lumbosacral intumescence.

About 15 % of fibers do not cross and don’t reach the opposite side, providing bilateral innervation of the muscles of the trunk and the sphincters of the urinary bladder. The peripheral motor neurons are located in the anterior horns. Among them we distinguish big alpha motoneurons, (whose axons end in muscle fibers which can make fast contraction) and small α-motoneurons (whose axons end in muscle fibers, capable to support long tonic contraction). Besides in the pyramidal tract there are γ-motoneurons with thin and slow-conducting axons, which provide the innervation of the proprioceptors of the muscular spindle and provide the maintenance of the muscular tone. Among intercalary neurons of the pyramidal tract it is necessary to distinguish Renschaw cells braking the action of big alpha motoneurons. The axons of the neurons of the pyramidal tract form the anterior motor root.

The distal fibers making these root, are located next to the spinal ganglion which contains the sensory cells - the first sensory neurons. The dendrites of the neurons of the spinal ganglion form the spinal nerve. The spinal nerves from several segments form the plexus. Large nerve trunks are formed from the plexus: ulnar, radial and median nerves – for a hand; femoral and sciatic nerves - for a leg from which peripheral nerves reach to separate muscles. The zone of the contact of the ending of the motor nerve and the muscle is called neuromuscular synapse. The acetylcholine is the mediator of synaptic transmission in it. The part of the dendrites of the nerve cells of the spinal ganglion ends in the receptors of the tendons (Golgi’s organs). They are receptors for the conduction of the impulses, braking activity of the alpha motoneurons. The axons of these sensory cells end at intercalary neurons which contact with alpha motoneurons.

Lesions of the first motor neuron in the brain or spinal cord usually produce spastic paresis, while lesions of the second motor neuron in the anterior horn, anterior root, peripheral nerve, or motor end plate usually produce flaccid paresis. Motor deficits rarely appear in isolation as the result of a lesion of the NS; they are usually accompanied by sensory, autonomic, cognitive, and/or neuropsychological deficits of various kinds, depending on the site and nature of the causative lesion.

The flaccid paralysis is characterized by a loss of tone and atrophy of the involved muscles; this is called flaccidity, or hypotonicity. Denervated muscle fibers undergo spontaneous contractions, known as fibrillations or fasciculations; these are too fine and rapid to be seen with the naked eye but can be demonstrated electromyographically. No pathologic reflexes are found.

The spastic paralysis is characterized by diminished muscular strength and impaired fine motor control, spastic increased tone, abnormally brisk stretch reflexes, possibly with clonus,

hypoactivity or absence of exteroceptive reflexes (abdominal, plantar, and cremasteric reflexes), pathological reflexes (Babinski, Oppenheim, Gordon, and Bekhterev reflexes, preserved muscle bulk. If the anterior horn are affected it may provoke fibrillation.

The Examination of Motor Functions

The examination of motor function consist in:

Muscle volume and contour. The volume and contour of the muscles give information about the presence of either atrophy or hypertrophy. Muscle atrophy, or amyotrophy, may be defined as the wasting or diminution in size of a muscle part. It is usually accompanied by changes in shape or contour. Its results from disorders affecting the anterior horn cell, the nerve root, the peripheral nerve, or the muscle itself. Muscle hypertrophy is an increase in the bulk, or volume, of muscle tissues. It may be the result of excessive use of the muscles, or it may occur on a pathologic basis. Muscle volume and contour are examined and atrophy or hypertrophy by inspection, palpation, and measurement. By means of inspection the general muscular development of the size of the muscles are noted, and special attention is paid to abnormalities in volume and contour and to evidence of atrophy and hypertrophy. Symmetric parts of the two sides of the body should be compared. The muscle masses should also be carefully palpated, and their volume, contour, and consistency noted. To determine the degree of atrophy or hypertrophy, measurements may be essential. The appraisal of muscle bulk and contour should be correlated with the other items of the motor examination, especially with the evaluations of strength and tone.

If muscular changes in the form of atrophy or hypertrophy are present, and if we need to refine the etiology of lesion a muscle biopsy should be considered. Electromyography aids in the differential diagnosis of muscular atrophy. It helps to decide the level of lower motor neuron and its peripheral axons lesion.

Motor strength and power. Motor strength and power indicate the capacity to exert and release force. It’s necessary to examine both the power of movement and the strength of contraction. In examining strength and power we are interested especially in voluntary, or active, motility. This is tested by having the patient carry out movements against the resistance of the examiner, and by having him resist active attempts on the part of the examiner to move fixed parts. Impairment of strength and power results in weakness, or paresis, absence of strength. Associated functions and abnormalities must be also noted. Abnormal fatigability may precede other objective manifestations of some neuromuscular disorders (such as myasthenia). There is a marked individual variation in muscle strength.

The paralysis may involve one muscle, a group of muscles, certain movements, or one or more extremities. A monoplegia is the paralysis of one extremity; diplegia- is the paralysis of extremities on the two sides of the body; hemiplegia, of one half of the body; paraplegia, the legs or the lower parts of the body; quadriplegia (tetraplegia) – the paralysis of all extremities. Hemiplegia alternans affects the upper extremity on one side of the body and the lower extremity on the other side.

When a muscle is maintained in a position of contraction or shortening for a period of time, a contracture may develop: the muscle cannot be stretched to normal limits without considerable pressure and the production of pain. Contractures may develop following prolonged spasm of muscles, in association with spastic paralysis.

To test motor power, the various movements at each joint and the strength of each important muscle should be examined individually. He is instructed to either resist active attempts by the examiner to move fixed parts or to initiate and carry out movements that are resisted by the examiner. Corresponding muscles on the two sides of the body should always be compared. Both active and passive movements should be tested and if limitation of movement is accompanied by discomfort or pain, this should be noted. In the presence of coma assessment of motor function may have to depend upon the presence of spontaneous movements, the position of the extremities, asymmetries of voluntary movements on the two sides, or withdrawal of an extremity in response to painful stimulation. Hemiplegia may be diagnosed if the contraction of the facial muscles on

one side is absent and the similar extension and external rotation of the thigh and leg is present. It is usually possible to evaluate muscle strength and power sufficiently well without recourse to special instruments (e.g. dynamometer).

Muscle function may be graded in various ways, but the following is an acceptable classification:

0. No muscular contraction occurs;
1. A flicker, or trace, of contraction occurs without actual movement, or contraction may be palpated in the absence of apparent movement; there is minimal or no motion of joints (0% - 10% of normal movement);
2. The muscle moves the part through a partial arc of movement with gravity eliminated (11% - 25% of normal movement);
3. The muscle completes the whole arc of movement against gravity (26% - 50% of normal movement);
4. The muscle completes the whole arc of movement against gravity together with variable amounts of resistance (51% - 75% of normal movement);
5. The muscle completes the whole arc of movement against gravity and maximum amounts of resistance several times without signs of fatigue. This is normal muscular power (76% - 100% of normal movement).

Examination of muscle tone. Tone, or tonus, has been defined as the tension of the muscles when they are relaxed, or as their resistance to passive movement when voluntary control is absent. In testing tone, the examiner should attempt to secure the complete cooperation of the patient, who should be comfortable and relaxed. Palpation of the muscles reveales their consistency, passive elasticity, firmness, or turgor. The most important criterion in the examination of tone is the resistance of muscles to passive manipulation when they are relaxed and when voluntary control is absent. Loss or diminution of tone is classified as hypotonicity, and pathologic increase, as hypertonicity. Hypotonicity or flaccidity results from involvement of the spinomuscular level or interference with the proprioceptive pathways, but may also be present with cerebellar lesions. It is characterized by a decrease or loss of normal tone. The muscle is flaccid and flabby, or soft to palpation. The excursion at the joint may be normal, but is usually increased. The decrease in tone appears in the anterior horn cell damage, abnormalities of the muscle itself or of the myoneural junction.

Hypertonicity is usually caused by lesions central to the anterior horn cells, or by interruption of impulses from supraspinal regions. It is seen most frequently with dysfunction of the so-called extrapyramidal and corticospinal levels, and is caused by either interruption of impulses that normally inhibit lower centers or imbalance of facilitatory and inhibitory centers, with consequent alteration of the motor neuron balance or lowering of the threshold of the spinal reflexes.

Spasticity. This occurs in association with lesions of the pyramidal or corticospinal level of function. There may be an elastic, springlike resistance to stretching at the beginning of movement, especially if the part is moved abruptly or suddenly, following which the muscle resists to a certain point and then suddenly relaxes – the phenomenon sometimes referred to as the clasp-knife type of resistance.

Extrapyramidal rigidity occurs with lesions of the basal ganglia or of some other portion of the extrapyramidal level of motor function or its connections with the brain stem reticular formations. This phenomenon is called “cogged-wheel symptom”. Tonus is expressed in the same manner in the group of flexors and extensors.

Corticospinal (Pyramidal) Tract Responses

With disease of the corticospinal or pyramidal system, certain abnormalities are found in the reflex pattern. This is true whether the disorder is in the motor cortex itself or anywhere along the descending tracts. The superficial reflexes may be decreased or absent, and deep reflexes are exaggerated.

Clonus. If the muscle tonus is markedly increased, there is also a pathologic response in the form of clonus – a series of rhythmic involuntary muscular contractions induced by the sudden passive stretching of a muscle or tendon.

The most frequently are occurred ankle clonus, patellar clonus.

Corticospinal tract responses in the upper extremities. The corticospinal tract responses in the upper extremities occur more rarely than those found in the lower extremities.

In the Rossolimo sign, flexion of the fingers and supination of the forearm follow either percussion of the palmar aspect of the metacarpophalangeal joints or tapping the volar surface of the patient's fingertips. Flexion of the fingers and hand may follow not only stimulation of the flexor tendons on the velar surface of the forearm, but also percussion of the dorsal aspect of the carpal and metacarpal areas (the Mendel-Bechterev sign). Jukovski sign caused by hammer impact on a sole on a palm under fingers; response is flexing of II-V fingers. Jackobson-Laske reflex. It’s caused by hammer impact on processus styloideus. The thumb is flexing.

Corticospinal tract responses in the lower extremities. The corticospinal tract responses in the lower extremities are more constant and more clearly defined than those in the upper limbs and may be elicited with more ease. They may be classified as those characterized in the main by dorsiflexion of the toes, and those characterized by plantar flexion of the toes.

Corticospinal responses characterized in the main by extension (dorsiflexion) of the toes. The Babinski Sign. In disease of the corticospinal system there is an inversion of the plantar reflex, the Babinski sign or extensor plantar response. Stimulation of the plantar surface of the foot is followed by dorsiflexion of the toes, especially of the great toe, together with a separation or fanning of the toes. The Babinski sign has been called the most important sign in clinical neurology. It is considered to be one of the most significant indications of disease of the corticospinal system at any level from the motor cortex through the descending pathways. It is the most delicate, the first to be evident in the presence of disease, and the one that occurs most frequently.

The Oppenheim sign is elicited by applying heavy pressure with the thumb and index finger to the anterior surface of the tibia, mainly on its medial aspect, and stroking down from the infrapatellar region to the ankle. The response is a slow one and usually occurs toward the end of stimulation.

The Gordon sign is obtained by squeezing or applying deep pressure to the calf muscles. The Schaefer sign is produced by deep pressure on the Achilles tendon.

Corticospinal tract responses characterized by plantar flexion of the toes. There is a group of reflexes in which the pathologic response is one of plantar flexion of the toes.

The Rossolimo sign is elicited by tapping to the tips of the toes.

The Mendel - Bechterev sign is elicited by tapping or stroking the outer aspect of the dorsum of the foot in the region of the cuboid bone. Plantar flexion of the toes may also be elicited by application of the stimulus to other portions of the foot and ankle. Bechterew found that percussion of the middle of the sole or of the heel was followed by a plantar flexion response.

Reflexes of spinal automatism. The reflexes of spinal automatism are also termed defense reflexes. Like the corticospinal tract signs, they become manifest when the inhibiting action of the higher centers has been removed, and thus indicate, in part at least, a release from such inhibition.

The Flexion Spinal Defense Reflex (or the Marie–Foix–Bechterev) sign most frequently may be evoked by an uncomfortable or nociceptive irritation. Pricking, pinching the skin on the

dorsal aspect of the foot or sharp flexion of the foot may initiate the response, as may squeezing the toes or extreme passive plantar flexion of the toes or foot.

Associated Movements (Pathologic Synkinesis)

Certain voluntary movements have a tendency to be accompanied by other involuntary responses called the associated, or synkinetic, movements. These are defined as automatic modifications of the attitude of certain parts of the body as a reflex response to the volitional motion of some other portion. Associated movements may be either physiological or pathological phenomenon. Pathologic associated movements are usually expressions of activity in paretic groups of muscles that are stimulated by active innervation of other groups.

There may be generalized (global) associated movements, symmetric (imitative, mirror) associated movements and coordinated associated movements. Most often they can be seen in patients with hemiparesis or hemiplegia of cerebral origin. Generalized associated movements tend to produce the characteristic position of the extremities in patient with hemipareis: the upper limb is held in a position of flexion of the fingers and wrist, and the elbow, and flexion and adduction at the shoulder (the paralysis of the extensors is more marked than that of the flexors). The lower extremity is held with extension at the hip and knee and plantar flexion at the ankle and toes, and with more marked paralysis of the flexors. These characteristics are increased with exertion. Straining and attempts to grip with the paretic hand may cause an increase in the spasticity, with increased flexion of the wrist, elbow, and shoulder. Involuntary movements such as yawning, coughing, and stretching may also increase the tonus and cause the affected arm to extend at the elbow, wrist, and fingers.

Symmetric associated movements are usually seen in the paretic limb when the opposite healthy one is forcibly moved. Thus, in squeezing the examiner's hand with the healthy hand the paretic hand is seen to flex.

Coordinated associated movements are characterized by a spread of response from one muscle or group of muscles to others. They alter the position of the part and lead to the adoption of new postures.

Methods of detecting of signs of central and peripheral paralysis (conclusion)

Signs of central paralysis:

Impairment of fine motor function. Voluntary movement of paretic limbs requires greater effort than normal and causes greater muscular fatigue. Moreover, rapid alternating movements are slowed by hypertonia in the opposing agonist and antagonist muscles of paretic limbs. There may be synkinesia (involuntary movement of paretic limbs associated with other movements, e. g., yawning), undifferentiated accessory movements (mass movements), or spinal automatisms (involuntary movements triggered by somatosensory stimuli).

Paralysis affects multiple (but not all) muscle groups on one side of the body. Bilaterally innervated movements (e. g., of eyes, jaw, pharynx, neck) may be only mildly paretic, or not at all. Paralysis that is initially total usually improves with time, but recovery may be accompanied by other motor disturbances such as tremor, hemiataxia, hemichorea, and hemiballism. Fine motor control is usually more severely impaired than strength. Neurogenic muscular atrophy does not occur in paralysis of central type.

Spasticity. The defining feature of spasticity is a velocity-dependent increase of muscle tone in response to passive stretch. Spasticity is usually, but not always, accompanied by hypertonia. The “clasp-knife phenomenon” (sudden slackening of muscle tone on rapid passive extension) is rare. Spasticity mainly affects the antigravity muscles (arm flexors and leg extensors).

Reflex abnormalities. The intrinsic muscle reflexes are enhanced (enlargement of reflex zones, clonus) and the extrinsic reflexes are diminished or absent. Pathological reflexes such as the Babinski reflex can be elicited.

Signs of peripheral paralysis:

Paralysis of peripheral origin can be caused by lesions of the anterior horn, nerve root, peripheral nerve, or motor end plate and must be distinguished from weakness due to disease of the muscle itself (myopathy). Apparent weakness can also be produced by tendon rupture or injury to bones and joints.

Paralysis. Paralysis is accompanied by diminution of muscle tone (flaccidity). The extent of weakness depends on the type, severity, and distribution of peripheral neuron or myopathic involvement.

Reflex abnormalities. The intrinsic muscle reflexes are diminished or absent to a degree that may be disproportionate to the degree of weakness: in peripheral-type paralysis, loss of reflexes is independent from the loss of strength; in myopathy, it parallels the weakness. Extrinsic reflexes are unaffected unless the effector muscle is atrophic. Pathological reflexes are absent.

Muscle atrophy. Muscle atrophy due to peripheral lesion may be disproportionate to the degree of weakness (either greater or less). Progressive atrophy of paralyzed muscles begins ca. 3 weeks after a peripheral nerve injury. The distribution and severity of muscle atrophy in myopathy depends on the etiology.

Spontaneous movements. Spontaneous movements are seen in affected muscles. Fasciculations are involuntary, nonrhythmic contractions of motor units in a relaxed muscle. They are not exclusively caused by anterior horn lesions. Myokymia is rhythmic contraction of muscle fibers; if the affected muscle is superficial the orbicularis oculi), waves of muscle contraction are visible

The subcortical structures include the basal ganglia, thalamus, subthalamic nucleus, hypothalamus, red nucleus, substantia nigra, cerebellum, and brain stem, and their nerve pathways.

The extrapyramidal system consists of the following gray structures: caudate nucleus, putamen, pallidum, subthalamic nucleus, substantia nigra and red nucleus.

Corpus striatum consists of two parts, the caudate and lentiform nuclei, which are incompletely separated one from the other. The caudate nucleus (nucleus caudatus) is located above and medial to the lentiform nucleus and is separated from it by a layer of white matter called the internal capsule (capsula interna). The thickened anterior part of the nucleus, its head (caput nuclei caudati) forms the lateral wall of the anterior horn of the lateral ventricle, the body forms the inferior wall of the central part of the lateral ventricle; the cauda forms the superior wall of the inferior horn. The caudate nucleus is separated from thalamus by the stria semicircularis (stria terminalis). Anteriad and deeper head of the nucleus approaches the anterior perforated substance where it is united with the lentiform nucleus (with the part called the putamen). The lentiform nucleus (nucleus lentiformis) is located laterally to the caudate nucleus and the thalamus and is separated from them by the internal capsule. On a frontal section the lentiform nucleus is wedge-shaped, the apex of the wedge is directed medially, and the base laterally. Two parallel white layers called the medullary laminae (laminae medullares) divide the lentiform nucleus into three segments, one lateral grey segment called the putamen and two medial lighter coloured segments united under the term globus pallidus.

The globus pallidus has a distinctive macroscopic appearance and also differs from the other parts of the corpus striatum histologically. It is phylogenetically older (palaeostriatum) than the putamen of caudate nucleus (neostriatum).

The claustrum is a thin sheet of grey matter in the region of the insula between it and the putamen. It is separated from the putamen by a thin layer of white matter, the external capsule (capsula externa) and from the cortex of the insula by a similar layer called the capsula extrema.

The amygdaloid nucleus (corpus amygdaloideum), or the epistriatum is located under the putamen in the anterior end of the temporal lobe. It does not reach the temporal pole, but lies in front of the apex of the inferior horn of the lateral ventricle. Morphologically, the amygdaloid nucleus is a posteroventral continuation of the claustrum.

As the formation of the cerebral cortex expands, the phylogenetically older motor centers (paleostriatum and neostriatum) become increasingly controlled by the new motor system, the system of the pyramidal tracts.

Major afferent and efferent connections of the striatal system. The striatum receives major input from three sources: the thalamus, the neocortex, and the substantia nigra. The striatum projects to the globus pallidus and the substantia nigra. The globus pallidus is the effector nucleus of the striatal system; it projects to the thalamus and to the subthalamic nucleus. The substantia nigra also projects to the thalamus. The striatal motor system expresses itself via the corticobulbar and corticospinal tracts. Major neurotransmitters of the striatal motor system. Within the striatum, globus pallidus, and pars reticularis of the substantia nigra, γ-aminobutyric acid (GABA) is the predominant neurotransmitter. GABA may coexist in the same neuron with enkephalin or substance P. Dopamine-containing neurons are found in the pars compacta of the substantia nigra. Acetylcholine is found in local circuit neurons of the striatum. The subthalamic nucleus projects excitatory glutaminergic fibers to the globus pallidus.

Clinical Features of Extrapyramidal Disease

The main signs of extrapyramidal lesions are disorders of muscle tone (dystonia) and involuntary movement disorders (hyperkinesia, hypokinesia, akinesia) absent during sleep. Two clinical syndromes can be differentiated. One is characterized by a combination or hyperkinesia and hypotonia and is caused by a disease of the neostriatum. The other presents as a combination of hypokinesia and hypertonia or rigidity and stems from a disease of the substantia nigra.

The Hypokinetic-Hypertonc Syndrome

It may be primary Parkinson's disease. In this situation, the pigmented neurons of the substantia nigra, locus caeruleus, and other brain stem dopaminergic cell groups are lost. The cause is not known. The loss of substantia nigra neurons, which project to the caudate nucleus and putamen, results in depletion of the neurotransmitter dopamine in these areas. Onset is generally after age 40, with increasing incidence in older age groups.

Secondary parkinsonism results from loss of or interference with the action of dopamine in the basal ganglia due to other idiopathic degenerative diseases, drugs, or exogenous toxins. The most common cause is ingestion of antipsychotic drugs or reserpine, which produce parkinsonism by blocking dopamine receptors. Less common causes include carbon monoxide or manganese poisoning, hydrocephalus, structural lesions (tumors, infarcts affecting the midbrain or basal ganglia), subdural hematoma, and degenerative disorders, including striatonigral degeneration and multiple systems atrophy. In postencephalitic parkinsonism, which is rare now (it occurred after the epidemic of von Economo's encephalitis in 1918 to 1924), an inflammatory process destroys the region of the midbrain containing the substantia nigra.

Manifestations

Bradykinesia, hypokinesia, and akinesia. Motor disturbances include slow initiation of movement (akinesia), sluggishness of movement (bradykinesia) and diminished spontaneous movement (hypokinesia); these terms are often used nearly interchangeably, as these disturbances all tend to occur together. Spontaneous fluctuations of mobility are not uncommon. The motor disturbances are often more pronounced on one side of the body, especially in the early stages of disease. They affect the craniofacial musculature to produce a masklike facies (hypomimia), defective mouth closure, reduced blinking, dysphagia, salivation (drooling), and speech that is diminished in volume (hypophonia), hoarse, poorly enunciated, and monotonous in pitch (dysarthrophonia). It may be hard to initiate speech, or repeat syllables. Postural changes include stooped posture, a mildly flexed and adducted posture of the arms, and postural instability. Gait disturbances appear in the early stages of disease and typically consist of a small-stepped gait, shuffling, and limping, with reduced arm swing. Difficulty initiating gait comes about in the later stages of disease, along with episodes of “freezing” - complete arrest of gait when the patient is confronted by doorway or a narrow path between pieces of furniture. It becomes difficult for the patient to stand up from a seated position, or to turn over in bed. Festinating gate - an involuntary

quickening of gait, as in some persons with Parkinson's disease. Impairment of fine motor control impairs activities of daily living such as fastening buttons, writing (micrographia), eating with knife and fork, shaving, and hair-combing. It becomes difficult to perform two activities simultaneously, such as walking and talking.

Behavioral Changes: Depression. The range of depressive manifestations includes worry, anxiety, avoidance of social contact, general unhappiness, listlessness, querulous, brooding, somatoform disturbances, and (rarely) suicidal ideation. Anxiety, tension, worry, mental agitation, lack of concentration, and dizziness are relatively common complaints.

In 50 to 80% of patients, the disease begins with a resting 4- to 8-Hz pill-rolling tremor of one hand. The tremor is maximal at rest, diminishes during movement, and is absent during sleep; it is enhanced by emotional tension or fatigue. Usually, the hands, arms, and legs are most affected, in that order. Jaw, tongue, forehead, and eyelids may also be affected, but the voice escapes the tremor.

Rigidity progresses, and movement becomes slow (bradykinesia), decreased (hypokinesia), and difficult to initiate (akinesia). Rigidity and hypokinesia may contribute to muscular aches and sensations of fatigue. In contrast to spastic elevation of muscle tone, rigor can be felt in extensors as a sticky, waxy resistance to all passive movements. The muscles cannot be relaxed. In passive movements one can feel that the tone of the antagonist muscles decreases in steps and not in an even, continuous fashion (cog-wheel phenomenon). The lifted head of a lying person, when suddenly released, does not fall down as usual but sinks gradually back onto the pillow (head-dropping test).

In contrast to their behavior in a spastic condition, the proprioceptive reflexes are not increased, and no pathologic reflexes can be observed. Paresis is absent. If it is too difficult to elicit reflexes, it is not possible to intensify the patellar reflex by Jendrassik's maneuver. (The patient hooks his hands together by the flexed fingers and tries to pull them apart as hard as he can while the patellar reflexes are checked). The result is an increase in the tonic stretching reflex, that is, an activated rigidity. The face becomes masklike, with mouth open and diminished blinking, which may be confused with depression. The posture becomes stooped. Patients find it difficult to start walking; the gait becomes shuffling with short steps, and the arms are held flexed to the waist and do not swing with the stride. Steps may inadvertently quicken, and the patient may break into a run to keep from falling (festination). The tendency to fall forward (propulsion) or backward (retropulsion) when the center of gravity is displaced results from loss of postural reflexes.

Speech becomes hypophonic, with a characteristic monotonous, stuttering dysarthria. Hypokinesia and impaired control of distal musculature are results in micrographia and increasing difficulty with activities of daily living. Dementia affects about 50% of patients, and depression is common.

During examination, passive movement of the limbs is met with a plastic, unvarying lead-pipe rigidity; superimposed tremor bursts may have a ratchet-like cogwheel quality. The sensory examination is usually normal. Signs of autonomic nervous system dysfunction (e.g., seborrhea, constipation, urinary hesitancy, orthostatic hypotension) may be found. Muscle strength is usually normal, although useful power may be diminished and the ability to perform rapid successive movements is impaired. Reflexes remain normal but may be difficult to elicit in the presence of marked tremor or rigidity.

The Hyperkinetic-Hypotonic Syndrome

This syndrome develops if the neostriatum is damaged. Occasionally, such lesions are accompanied by others in the globus pallidus, thalamus, or cerebral cortex; in such cases the hyperkinesia is possibly caused by a loss of inhibitory neurons of the neostriatum that descend to pallidum and substantia nigra. In other words, a loss of a neuronal system of higher order has occurred, producing excessive excitation of the neurons of the next lower system. The resulting hyperkinesias are of different kinds: athetosis, chorea, spasmodic torticollis, torsion dystonia, ballism, and other conditions .

Clinical Features of the Hyperkinesia-Hypotonia Syndrome.

Athetosis

This kinetic disorder is usually caused by perinatal damage to the striate bodies. Involuntary movements are slow and wormlike, with a tendency to overextend the peripheral portions of the extremities. In addition, there are irregular, spasmodic increases in muscle tensions between agonists and antagonists. As a result, postures and movements are rather bizarre. Voluntary movements are severely distorted by the spontaneous appearance of hyperkinetic movements that may include face and tongue and thus cause grimacing with abnormal tongue movements like laughing or crying. The athetosis may be combined with a contralateral paresis; it may also be bilateral and is then called double athetosis, which usually occurs in association with spastic paraplegia (Little's disease).

Chorea

The chorea syndrome is characterized by short, fast, involuntary jerks occurring in single muscles, random and producing various patterns of movements. At first the peripheral portions of the extremities are involved and the proximal portions follow. Involuntary jerks of the facial muscles produce grimacing. A combination of choreiform and (distal) dystonic movements is termed choreoathetosis.

Huntington disease, an autosomal dominant disorder, is the best-known cause of chorea

Others include hereditary diseases (e. g., neuroacanthocytosis, and benign hereditary chorea) and neurodegenerative diseases (e. g., Alzheimer disease, multisystem atrophy). Secondary chorea may be caused by infections (e. g., Sydenham’s chorea due to streptococcal infection; herpes encephalitis, toxoplasmosis), vascular disease (e. g., lupus erythematosus, stroke), brain tumor, drug therapy (e. g., estrogen, neuroleptic drugs), or old age (senile chorea).

Spasmodic torticollis and torsion dystonia

These are the most important types of dystonia syndromes. In both diseases there are usually alterations within the putamen and the centromedian nucleus of the thalamus and in other extrapyramidal nuclei (pallidum, substantia nigra, and others).

Blepharospasm

Spasmodic contraction of the orbicularis oculi muscle causes excessive blinking and involuntary eye closure. It can often be accompanied by ocular foreign-body sensation and be ameliorated by distracting maneuvers, and is worse at rest or in bright light. There may be involuntary clonic eye closure, tonic narrowing of the palpebral fissure, or difficulty opening the eyes. Blepharospasm may be so severe as to leave the patient no useful vision.

Cervical dystonia

Cervical dystonia may involve head rotation (torticollis), head tilt to one side (laterocollis), or flexion or extension of the neck (anterocollis, retrocollis), often accompanied by tonic shoulder elevation or head tremor. It may be difficult to distinguish nondystonic from dystonic head tremor; only the latter can be improved by antagonistic maneuvers. Dystonia often causes pain, usually in the neck and shoulder.

Torsion dystonia

Torsion dystonia is characterized by rather extensive turning and twisting movements of trunk and proximal extremities. They can be so severe that the patient can neither stand nor walk without support. The disease may be idiopathic or symptomatic; in the latter case the cause may be birth injury, kernicterus, former encephalitis, early Huntington's chorea, Hallervorden-Spatz disease, or a hepatocerebral degeneration (Wilson's diease, Westphal-Strumpell disease).

Tics

Tics are rapid, irregular, involuntary movements (motor tics) or utterances (vocal tics) that interrupt normal voluntary motor activity. They are triggered by stress, anxiety, and fatigue but may also occur at rest; they can be suppressed by a voluntary effort, but tend to re-emerge with greater intensity once the effort is relaxed. Tics are often preceded by a feeling of inner tension. They may be transient or chronic.

Simple tics. Simple motor tics involve isolated movements, e. g., blinking, twitching of abdominal muscles, or shrugging of the shoulders. Simple vocal tics may involve moaning, grunting, hissing, clicking, shouting, throat clearing, sniffing, or coughing.

Complex tics. Complex motor tics consist of stereotyped movements that may resemble voluntary movements, e. g., handshaking, scratching, kicking, touching, or mimicking another person's movements (echopraxia). Complex vocal tics may involve obscene language (coprolalia) or the repetition of another person's words or sentences (echolalia).

Gilles de la Tourette syndrome (often abbreviated to Tourette syndrome) is a chronic disease in which multiple motor and vocal tics begin in adolescence and progress over time. Other features of the disease are personality disturbances, obsessive-compulsive phenomena, and an attention deficit.

Hemiballism (Ballism)

Ballism consists of violent flinging movements of the limbs due to involuntary contraction of the proximal limb muscles, and usually affects only one side of the body (hemiballism). It may be continuous or occur in attacks lasting several minutes. The most common cause is an infarction or other destructive lesion of the subthalamic nucleus (STN). Diminished neural outflow from the STN leads to increased activity in the thalamocortical motor projection.

Myoclonus

Myoclonus consists of involuntary, brief, sudden, shocklike muscle contractions producing visible movement. It has a variety of causes and may be focal, segmental, multifocal, or generalized. It's cortical, subcortical, or spinal origin can be determined by neurophysiological testing. Attacks of myoclonus may be spontaneous or may be evoked by visual, auditory, or somatosensory stimuli (reflex myoclonus) or by voluntary movement (postural myoclonus, action myoclonus).

Clinical features of the internal capsule lesions. As a result of compact location of the different passways, damages of the capsulae internae include disorders of the movement, decrease in the threshold for perceiving touch, pain, and temperature and deep sensations, and the visual problems. The complete syndrome is called "the syndrome of three hemi". Signs that may be found as a result of unilateral or bilateral involvement of the capsulae internae:

Unilateral paralysis (hemiplegia) of the opposite side of the body and contralateral III nerve palsy.

Contralateral hemihypesthesia, usually involving trunk and extremities more severely than the face.

Visual field deficit (contralateral homonymous hemianopia).

Fragmentary lesions of the anterior part of the posterior limb result in abnormalities only of the movement (contralateral hemiplegia). If damage is localized in the posterior part of the posterior limb capsulae internae the "syndrome of three hemi" is formed (contralateral homonymous hemianopia, hemihypesthesia and hemiataxia).

Method of determination of extrapyramidal disorders

Chorea (hyperkinetic-hypotonic syndrome) is an abnormal involuntary movement usually distal in location, brief, nonrhythmic, abrupt, and irregular, that seems to flow from one body part

to another. The movements are random, unpredictable in timing, direction, and distribution. Chorea can be partially suppressed; some patients can incorporate these into semipurposeful movements called parakinesia. Motor impersistence, the inability to maintain a sustained contraction, is a typical feature of chorea. Choreic gait has a stuttering or dance-like quality that reflects superimposed choreic and choreoathetotic movements. Stride length and cadence are irregular and random. The base is variable. Steps often deviate from the direction of travel, giving the gait a somewhat ataxic quality. Choreic gait can be seen in patients with Huntington disease and in patients with PD who experience medication-induced dyskinesias.

Athetosis and ballism are sometimes confused with chorea. Athetosis is a slow, writhing, continuous set of involuntary movements, usually affecting limbs distally, but it can involve the axial musculature (neck, face, and tongue). If athetosis becomes faster, it sometimes blends with chorea, that is, choreoathetosis. Ballism is large-amplitude, involuntary movements affecting the proximal limbs, causing flinging and flailing limb movements.

Patients with chorea are often initially unaware of these involuntary movements. The chorea is often first interpreted by observers as fidgetiness. The patients are usually frustrated by their own incoordination or clumsiness.

Key diagnostic elements of parkinsonism (hypokinetic-hypertonic syndrome) are the presence of two of the four cardinal signs: bradykinesia, tremor, rigidity, and gait problems. Hypokinetic-rigid gait is also known as akinetic-rigid gait or parkinsonian gait and is seen in any of the various parkinsonian syndromes. The gait is characterized by flexed posture, reduced arm swing and stride length, shuffled steps, turning en bloc, and postural instability. Patients frequently exhibit festination, an acceleration of gait in which the steps get shorter and faster as the patient attempts to keep pace with his or her displaced.

Methods of examination of muscle tone and definition of spasticity and rigidity

The patient's body tone is best evaluated when the individual is fully relaxed. Sometimes, it is useful to check tone more than once during the examination. Tone is described as the patient's primary level of muscular tension. To become comfortable with this part of the examination, it is important, as with other portions of the neurologic evaluation, to routinely check these parameters in healthy individuals to establish one's normal base of observations.

The increase in tone is referred to as spasticity. This has an interesting paradox in that at rest the spastic muscle has limited tone, but if there is a sudden attempt by the examiner to change the posture the limb is easily moved for a very short distance and then the degree of resistance immediately and rapidly increases up to a maximum and then dissipates. This resembles a "clasp knife," i.e., pocket knife, resistance/relaxation.

Four primary types of changes in tone are found in patients with primary CNS disease: hypotonia, spasticity, flaccidity, and rigidity. It is important to place these observed changes in motor tone within the context of the complete neurologic examination rather than in isolation.

Hypotonia. This is occasionally demonstrable in patients with cerebellar hemispheric lesions. For example, the distal part of the ipsilateral extremity may not be able to perform rapid alternate movements (called dysdiadochokinesia) because of the inability to maintain a stable posture. Similarly, the smooth, straight pursuit seen when one elicits the knee MSR loses the out-and-back motion that typically has an inhibitory cerebellar check. Instead, on return, there is overshoot with no check, leading to repetitive pendular response. This classic hypotonic cerebellar tone is a relatively uncommon finding. A more generalized loss of normal tone is most commonly seen among infants with either central or peripheral motor unit disorders, classically spinal muscular atrophy (Werdnig–Hoffmann disease, Dubowitz disease, Kugelberg–Welander disease) or the various congenital myopathies. Although a similar example is seen in adults, rarely, a floppy head syndrome develops in an older patient.

Flaccidity. This is the term for a total loss of tone and is seen in various disease processes affecting the upper motor neurons. Most commonly, this occurs in acute settings such as with a recent stroke or a sudden spinal cord injury, that is, spinal shock. However, with both of these, the

flaccidity is temporary and tone increases later to present in the form of varying degrees of spasticity.

Rigidity. Increasing tone from basal ganglia disorders, as may occur with Parkinson disease, is known as rigidity. Rigidity creates a continuous sense of tightness in the attempt to move the joint through a full excursion from extension to flexion.

Control materials to the preparatory stage of the class:

Questions:

What parts the central nervous system consist of?
What anatomic structures includes in the peripheral part of the nervous system?
What parts of the vegetative nervous system do you know?

Standards of answers:
The central nervous system consists of spinal cord and brain.
In the peripheral part of the nervous system includes: roots of cranial nerves and spine, cranial and spinal ganglia, nervous plexuses, peripheral nerves.
Segmental and over segmental parts.

Tests:

1 level (with the single chosen answer)
Subcortical nodes are:
a. Anterior horns
b. Anterior funiculi
c. Thalamus
d. Pons

Answer: C.

The spinal cord has:
a. lateral columns
b. neural plexuses
c. medulla oblongata

Answer: A.

Tests of the II level (with a few chosen answers)

1. Which structures are the parts of the brain?
2. Which structures are the parts of the spinal cord?

a. cerebellum
b. anterior horns
c. spinal roots
d. pons
e. lateral column

Answer:
1. a,d.
2. b,c,e.

Tasks for control with answers:
1. A 15 years old boy paid attention, that he can not recognize the touched object in his right hand. Define localization of process.
2. A child does not know objects which are shown to her. Define what structures suffered.
Answer:

1. Left parietal lobe of the brain
2. Occipital lobe of the brain.

Questions (right answer in bold):
Cortico-spinal (or cortico-nuclear) pathway decussates in: a) cerebral cortex; **b) brainstem;** c) capsula interna; d) corona radiate; e) all above mentioned

Clinical manifestation of diseases of the spinomuscular level includes in: a) hypotonic and atrophy of involved muscles, b) muscles stretch reflexes are exaggerated, hypertonic; **c) all above mentioned**

Stimulation of pyramidal cortex may causes: **a) Jacksonian convulsive seizures;** b) fibrillation; c) hypertrophy; d) all above mentioned

Paralysis is: **a) absence of strength;** b) contraction of single muscle fibers; c) excessive movement; d) Jacksonian convulsive seizures

Hemiplegia is: a) the paralysis of one extremity; b) the paralysis of four extremities; **c) the paralysis of half of the body;** d) the paralysis of three extremities

Spinomuscular level has got the motor nuclei in: a) cerebral cortex; **b) anterior horns;** c) capsula interna; d) corona radiate; e) brainstem

Monoplegia is: **a) the paralysis of one extremity;** b) the paralysis of four extremities; c) the paralysis of half of the body; d) the paralysis of three extremities

Clinical manifestation of diseases of the corticomuscular level includes in: a) hypotonic and atrophy of involved muscles, b) muscles stretch reflexes are exaggerated, hypertonic; **c) all above mentioned**

Spasticity is: **a) an elastic, springlike resistance to stretching at the beginning of movement;** b) loss of muscle tone; c) contraction limited to a single muscle fiber or to a group of fibers; d) seizures

Cerebellum has nucleus: a) fastigii; b) emboliformis; c) globosus; d) dentatus; e) rubber; f) all above mentioned; **g) all above mentioned except "e)"**

The cerebellum has peduncles: a) superior; b) inferior, c) middle; d) anterior; e) superior, inferior, middle, anterior; f**) superior, inferior, middle**

Main symptoms of the cerebellum dysfunctions are: a) tremor; b) scaning speech; c) nistagmus; d) dismetria; e) ataxia; f) muscles hypotonia; **g) all above mentioned**

Muscle tonus results from cerebellar disease: a) hypertonic; **b) hypotonic**; c) distonic

Dysdiadochokinesia means: **a) inability to perform rapid alternating movements;** b) inability to control range of movements

Materials for self-control of quality of preparation:

A. Questions for self-control:

1. Anatomy of the central nervous system.
2. Anatomy of the peripheral nervous system.
3. Types of neurons, their functional value.
4. Basic stages of phylo- and ontogenesis of the nervous system.
5. Anatomy of the vegetative nervous system.
6. Structural and functional unit of the nervous system.
7. Method of research of the nervous system.

Questions (right answer in bold):

Spinomuscular level has got the motor nuclei in: a) cerebral cortex; **b) anterior horns;** c) capsula interna; d) corona radiate; e) brainstem

Monoplegia is: **a) the paralysis of one extremity;** b) the paralysis of four extremities; c) the paralysis of half of the body; d) the paralysis of three extremities

Clinical manifestation of diseases of the corticomuscular level includes in: a) hypotonic and atrophy of involved muscles, b) muscles stretch reflexes are exaggerated, hypertonic; **c) all above mentioned**

Spasticity is: **a) an elastic, springlike resistance to stretching at the beginning of movement;** b) loss of muscle tone; c) contraction limited to a single muscle fiber or to a group of fibers; d) seizures

B. Tests for self-control with the standards of answers:

1. At a patient a pathological process arranged the back horns of neck bulge. Name affected segments of spinal cord?
2. To name vegetative reflexes?

Answers:

1. C5 - C8 segments.
2. Dermographysm, pulomotoric reflexes, Stcherbak's reflex, oculo-cardial reflex of Ashner-Danini, neck reflex, epigastral reflex.

B. Tasks for self-control with answers:

1. A girl of 7 years has the states of a different going pale of skin, which is accompanied by tachcardia, falling of the arterial pressure, fever, hyperhydrosis . Define localization of process.
2. A 15 years old boy paid attention, that does not feel the details of object a right hand. Define localization of process.
3. A child does not know objects which are shown to her. Define what structures suffered.

Answer:

1. Area of hypothalamus.
2. Parietal lobe of the brain
3. Cervical lobe of the brain.

Questions (right answer in bold):

25-years-old woman had a clonic convulsion attack in the right leg, which extended on the right hand and right facial muscles. It continued for 3 minutes. Point the syndrome.
Answer: **Jacksonian convulsive seizures**

A 42-years-old patient complains of weakness in his right hand. During the examination right hand muscles atony and atrophy, diminution of strength were detected. It is registered fibrillation in damaged muscles. Deep tendon reflexes (biceps, triceps, carporadial are absent to the right. Point the lesion locus.
Answer: **Anterior horn at the level C 5-8**

Atrophy in the head of the caudate nucleus in patients with Huntington's disease affects the shape of which of the following?
a. Cerebellum
b. Lateral ventricle
c. Third ventricle
d. Lenticular nuclei
e. Temporal lobe

A 65-years-old patient has got festinating gait, semiflexed position and half-bent extremities, masklike face and low monotonous speech. Pill-rolling tremor of one hand is observed. The muscule tone decrease with "cog-wheel phenomenon". Ground the topical diagnosis. Point the syndrome.
Answer: **parkinsonism**

9-years-old girl observed rapid, arrythmic, involuntary movements of trunk and limbs. She frequently grimaces, extrude her tongue. Muscle tone is decreased. Ground the topical diagnosis. Point the syndrome.
Answer: **chorea minor**

A 61-year-old right-handed man presents with involuntary twitches of his left hand. He first noticed between 6 months and 1 year ago that when he is at rest, his left hand shakes. He can stop the shaking by looking at his hand and concentrating. The shaking does not impair his activities in any way. He has no trouble holding a glass of water. There is no tremor in his right hand, and his lower extremities are not affected. He has had no trouble walking, and there have been no falls. There have been no behavioral or language changes. On examination, a tremor of the left hand is evident when the man is distracted. His handwriting is mildly tremulous. He has bilateral cogwheel rigidity with contralateral activation, which is worse on the left. His rapid alternating movements are bradykinetic on the left. Which of the following is the most likely diagnosis in this case?
a. Epilepsy
b. Guillain-Barré syndrome
c. Multiple sclerosis
d. Parkinson's disease
e. Stroke

An 85-year-old man is being evaluated for gait difficulties. On examination it is found that joint proprioception is absent in his toes. People with impaired position sense will usually fall if they stand with their feet together and do which of the following?
a. Flex the neck
b. Extend their arms in front of them
c. Flex the knees
d. Turn the head
e. Close their eyes

Tremor in the hands that is most obvious when the patient is awake and trying to perform an action is most likely due to disease in which of the following structures?
a. Thalamus
b. Cerebellum
c. Substantia nigra
d. Spinal cord
e. Internal capsule

A 42-year-old woman is being evaluated for gait difficulties. On examination, it is found that her ability to walk along a straight line touching the heel of one foot to the toe of the other is impaired. This finding is most common with which of the following?
a. Cerebellar dysfunction
b. Parietal lobe damage
c. Temporal lobe damage
d. Ocular motor disturbances
e. Dysesthesias in the feet

A 24-years-old male complains of unsteadiness. During the examination unsteady wide-based gait, horizontal double-acting nystagmus was detected. Romberg test is positive. The patient falls on the right. The finger nose and heel-knee test are positive on the right. Muscle tone is decreased on the right. Deep sensation is present. Palsies are absent. Ground topical diagnosis.

Answer**: right cerebellar hemisphere**

A 30-years-old male has got decrease of muscle tone in left extremity, dysdiadochokinesia, intentional tremor on the left and decreased tendon reflexes. Ground the topical diagnosis.

Answer**: left cerebellar hemisphere**

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources

1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical Class No. 2.

Theme: Sensitive system and symptoms of injury. Kinds and types of sensory loss - 2 hours.

Actuality of theme.

Disorders of sensation are the common sign of the nervous diseases. Investigation of sensation helps to make the topical diagnosis.

Aims of the class:

Educational aims:

	Even mastering for Bezpalko
- to acquaint students with the variety kinds of sensation and with the importance of the normal functioning of sensation in human life.	1 level
- a student must to know: 1) Classification of types of superficial sensation. 2) Types of deep sensation. 3) Pathways of pain and temperature sensation. 4) Pathways of deep sensation. 5) Location of cortical parts of general sensation.	2 level
- to give possibility to capture research skills to the students: 1) Pain sensation. 2) Temperature sensation. 3) Touch sensation. 4) Muscular-joint sensation. 5) Sensation of vibration. 6) Feeling of stamping and weight. 7) Stereognosis	3 level
- to give to the students of ability to explore the types of sensation disorders (central and peripheral).	4 level

Educating whole related to:

- by forming at the students of ability to put the topical diagnosis of defeat of cerebrum.
- by the actual aspects of deontological, patriotic, professional, psychological, legal, ecological responsibility and others like that.

Table of contents of the class:

There are two functionally and anatomically distinct types of somatic sensation and pain. The spatially and temporally precise perception of light tactile, noxious, and temperature stimuli is called *epicritic sensation*, and the more diffuse perception of stronger tactile, noxious, and temperature stimuli is called *protopathic sensation*.

Sensation in the deep tissues (muscles, viscera) is predominantly protopathic.

Receptors

Sensory stimuli affect the nervous system by physically interacting with *receptors*. *Exteroceptors* respond to external stimuli (mechanical, thermal, optic, acoustic, olfactory, gustatory); *interoceptors* respond to internal stimuli (stretch, pressure, chemical irritation of internal organs). A stimulus activates a receptor only if it is sufficiently intense (above threshold). Receptors are classified according to their activating stimuli: *mechanoreceptors* (pressure, touch; proprioceptive sensations such as joint postion, muscle contraction, muscle stretch; hearing, sense of balance), *thermoreceptors* (heat, cold), *chemoreceptors* (pain, smell, itch, taste), and *photoreceptors* (light). *Cutaneous receptors* include both "free" nerve endings and specially adapted receptors (e. g., corpuscles of Meissner and Vater-Pacini). The former type mainly subserve pain and temperature sense, the latter tactile sensation (touch, pressure, vibration). In hair-covered skin there are tactile receptors around the hair roots.

Nerve Pathways

From the receptor, information is transmitted to the afferent fibers of the pseudounipolar spinal ganglion cells, whose efferent fibers reach the spinal cord by way of the dorsal root. A synapse onto a second neuron in the sensory pathway is made either immediately, in the posterior horn of the spinal cord (protopathic system), or more rostrally, in the brain stem (epicritic/lemniscal system). The highest level of the somatosensory pathway is the contralateral primary somatosensory cortex. The somatotopic organization of the somatosensory pathway is preserved at all levels.

Posterior column (epicritic/lemniscal system).

Fibers mediating sensation in the legs are in the fasciculus gracilis (medial), while those for the arms are in the fasciculus cuneatus (lateral). These fibers synapse onto the second sensory neuron in the corresponding somatosensory nuclei of the lower medulla (nucleus gracilis, nucleus cuneatus), which emit fibers that decussate and ascend in the contralateral medial lemniscus to the thalamus (ventral posterolateral nucleus, VPL). VPL projects to the postcentral gyrus by way of the internal capsule.

Anterolateral column (protopathic system).

Fibers of the protopathic pathway for *somatic sensation* (strong pressure, coarse touch) enter the spinal cord through the dorsal root and then ascend two or more segments before making a synapse in the ipsilateral posterior horn. Fibers originating in the posterior horn decussate in the anterior commissure of the spinal cord and enter the *anterior spinothalamic tract*, which is somatotopically arranged: fibers for the legs are anterolateral, fibers for the arms are posteromedial. The anterior spinothalamic tract traverses the brain stem adjacent to the medial lemniscus and terminates in VPL, which, in turn, projects to the postcentral gyrus. The protopathic

pathway for *pain* (aswell as tickle, itch, and temperature sensation) is organized in similar fashion: Central fibers of the first sensory neuron ascend 1 or 2 segments before making a synapse in the substantia gelatinosa of the posterior horn. Fibers from the posterior horn decussate and enter the lateral spinothalamic tract, which, like the anterior spinothalamic tract, projects to VPL; VPL projects in turn to the postcentral gyrus.

Methods of assessment of superficial sensitivity

A carefully designed sensory system evaluation is essential to define the presence or absence of normal sensation and, if abnormal, to define the specific anatomic patterns of loss for the affected modalities. Because part of the sensory examination is fairly subjective, the examiner should analyze the consistency of responses. Additionally, the relevance of sensory changes to the patient's complaints and other findings needs to be carefully evaluated. Initially, the examination needs to focus on defining the presence or loss of sensation. One must avoid having the patient be overly zealous trying to define the most subtle differences in sensory appreciation. This often leads to an exhausted patient and a frustrated clinician.

In most clinical settings, it is best to separate the sensory examination into two major categories, that is, those derived from superficial skin receptors or deeper mechanoreceptors. The former are small, unmyelinated, slowly conducting type C fibers or larger, slightly myelinated, somewhat more rapidly conducting type A-delta fibers. These small fibers primarily subserve pain and temperature (respectively tested using a pin point or a cold object such as the handle of a tuning fork) and gross touch modalities. Fine tactile discrimination is evaluated by using a pair of calipers to check their ability to recognize whether one or two points are applied to the digit.

Methods of assessment of deep sensitivity

The large, well-myelinated type A-alpha and A-beta fibers carry the kinesthetic modalities of position sense studied by the examiner's passively moving the finger or toe in the vertical plane and asking the patient which direction the digit was moved, either up or down.

Vibratory sensation depends on both deep afferent and cutaneous sensory modalities subserved by type A-alpha fibers. It is best tested by a 128-Hz tuning fork that typically has a low frequency rate and longer duration of action. This modality is the one that most commonly diminishes in sensitivity with aging.

Methods of assessment of complex sensitivity

Two-point discrimination: The ability to distinguish two pointsfrom one is tested by using a compass, the points of which should be blunt and applied simultaneously and painlessly. The distance at which such stimuli can be recognized as a distinct pair varies but is roughly 1 mm at the tip of the tongue, 2 to 3 mm on the lips, 3 to 5 mm at the fingertips, 8 to 15 mm on the palm, 20 to 30 mm on the dorsa of the hands and feet, and 4 to 7 cm on the body surface. It is characteristic of the patient with a lesion of the sensory cortex to mistake two points for one, although occasionally the opposite occurs.

Cutaneous localization and figure writing (graphesthesia): Localization of cutaneous tactile or painful stimuli is tested by touching various points on the body and asking the patient to place the tip of his index finger on the point stimulated or on the corresponding point of the examiner's limb. Recognition of numbers or letters traced on the skin (these should be larger than 4 cm on the palm) with a pencil or similar object or the direction of a line drawn across the skin also depends on localization of tactile stimuli. Normally, traced numbers as small as 1 cm can be detected on the pulp of the finger if drawn with a sharp pencil. Futhermore, these are also the most useful and simple tests of posterior column function.

Appreciation of texture, size, and shape: Appreciation of texture depends mainly on cutaneous impressions, but recognition of the shapes and sizes of objects is based on impressions from deeper receptors as well. Inability to recognize shape and form is frequently a manifestation of cortical disease, but a similar clinical defect will occur if tracts that transmit proprioceptive and

tactile sensation are interrupted by lesions of the spinal cord and brainstem (and, of course, of the peripheral nerves). This type of sensory defect is called stereoanesthesia and is distinguished from ***astereognosis,*** which connotes an inability to identify an object by palpation, even though the primary sense data (touch, pain, temperature, and vibration) are intact. In practice, a pure astereognosis is rarely encountered, and the term is employed when the impairment of superficial and vibratory sensation in the hands seems to be of insufficient severity to account for the defect in tactile object identification. Defined in this way, astereognosis is either right- or leftsided and, with the qualifications mentioned below, is the product of a lesion in the opposite hemisphere, involving the sensory cortex, particularly S2 or the thalamoparietal projections.

Interpretation of findings. There is a wide range of normal findings. Apparent abnormalities should be interpreted in conjunction with findings of other types, such as abnormal reflexes or paresis.

Sensory dysfunction may involve not only a diminution or absence of sensation (hypesthesia, anesthesia), but also sensations of abnormal type (paresthesia, such as prickling or formication) or spontaneous pain (dysesthesia, often of burning type). Patients often use the colloquial term “numbness” to mean hypesthesia, anesthesia, or paresthesia; the physician should ask specific questions to determine what is meant.

Types of the sensory disorders.

1. Peripheral type: polyneuritic, neural, plexal.
2. Segmentary type: ganglionary, radicular and segmental-dissociated.
3. Central: conduction, central, cortical.
4. Symptoms of loss and irritating signs (sensitive "Jackson").

Syndromes of Lesion of Sensory System on Different Levels

Peripheral Nerve Lesions

Mononeuropathy. A mononeuropathy is a lesion of one nerve by a local process, usually compression, trauma or a vascular cause. Clinical examination typically demonstrates negative and positive sensory disturbances restricted to the territory of the nerve involved.

Polyneuropathy. In patients with polyneuropathies, sensory loss is symmetric and more frequently distally than proximally (stocking-and-glove sensory loss). The peripheral nervous system is involved diffusely. Sensory loss may be accompanied by a motor deficit.

Plexus lesion. It is appeared by hypo- or anesthesia of all sorts of sensation in the region of plexal innervation with pain, paresthesias and vegetative disorders

Root lesion. Pain due to root compression is generally accompanied by sensory loss in appropriate dermatom, muscle weakness, and decreased or absent tendon reflexes. If the spinal ganglion is involved, the herpes zoster is appeared.

Cord lesion. In patients with a cord lesion, there may be a transverse sensory level. Physiologic areas of increased sensitivity do occur, however, at the costal margin, over the breasts, and in the groin, and these must not be taken as abnormal. Therefore, the level of a sensory deficit affecting the trunk is best determined by careful sensory testing over the back rather than the chest and abdomen.

Central cord lesion. Central cord lesion (such as syringomyelia, following trauma, certain cord tumors) may cause a loss of pain and temperature appreciation with sparing of other modalities. This loss is due to the interruption of fibers conveying pain and temperature that cross from one side of the cord to the spinothalamic tract on the other. Such a loss is usually bilateral, may be asymmetric. It may be accompanied by lower motor neuron weakness in the muscles supplied by the affected segments.

The level of the posterior horn. Disorders in the posterior horn are characterized by pain and temperature sensivity disorders.

Anterolateral cord lesion. Lesions involving the anterolateral portion of the spinal cord (lateral spinothalamic tract) can cause contralateral abnormality of pain and temperature sensation in segments below the level of the lesion.

Anterior grey commissure. Lesion in its region cause dissociated disorders – pain and temperature sensations disorders in symmetrical parts of body like a “butterfly” or “jacket”.

Anterior cord lesion. With destructive lesions involving the anterior portion of the spinal cord, pain and temperature appreciation are impaired below the level of the lesion from lateral spinothalamic tract involvement. Weakness or paralysis of muscles supplied by the involved segments of the cord results from damage to motor neurons in the anterior horn.

Posterior column lesion. There is loss of vibration and joint position sense below the level of the lesion, with preservation of other sensory modalities.

Cord hemisection. Lateral hemisection of the cord leads to Brown-Sequard's syndrome. Below the lesion, there is an ipsilateral pyramidal deficit and disturbed appreciation of vibration and joint position sense, with contralateral loss of pain and temperature appreciation that begins two or three segments below the lesion.

Brainstem lesion. Sensory disturbances may be accompanied by a motor deficit, cerebellar signs, and cranial nerve palsies when the lesion is in the brainstem. In patients with lesions involving the spinothalamic tract in the dorsolateral medulla and pons, pain and temperature appreciation are lost in the limbs and trunk on the opposite side of the body. When such a lesion is located in the medulla, it also typically involves the spinal trigeminal nucleus, impairing pain and temperature sensation on the same side of the face as the lesion. The result is a crossed sensory deficit that affects the ipsilateral face and contralateral limbs. In contrast, spinothalamic lesions above the spinal trigeminal nucleus affect the face, limbs, and trunk contralateral to the lesion. With lesions affecting the medial lemniscus, there is loss of touch and proprioception on the opposite side of the body. In the upper brainstem, the spinothalamic tract and medial lemniscus run together so that a single lesion may cause loss of all superficial and deep sensation over the contralateral side of the body.

Medial lemniscus lesion cause hemianesthesia and sensitive hemiataxia (anesthesia of deep sorts of sensation).

Thalamic Lesions. Thalamic lesions may lead to loss or abnormality of all forms of sensation on the contralateral side of the body and is appeared by hemianesthesia, sensitive ataxia, hemianopsia, hemialgia. Spontaneous pain, hyperpathia may occur on the affected side. Patients may describe it as burning, tearing, knifelike, or stabbing. Any form of cutaneous stimulation can lead to painful or unpleasant sensations. Some later develop persistent severe pain and choreoathetoid movements on the affected side, mild hemiataxia, and astereognosis. Called also Dejerine-Roussy syndrome.

Lesion of internal capsule. Involvement of the sensory radiations in the internal capsule causes variable and sometimes extensive diminution of all types of sensation on the opposite side of the body. The changes are similar to those which follow a thalamic lesion, and it may be difficult to differentiate between the two. Pain, however, is rarely experienced. There are hemianesthesia, hemiataxia, hemiplegia.

Lesions of the sensory cortex (postcentral gyrus). Cortical sensory disturbances are usually appeared by monoanesthesia in the opposite side of body. Stimulation of postcentral gyrus cause sensitive "Jackson"-the feeling of tingling or numbness in the opposite side. Lesion of upper parietal lobule is characterized by astereognosis.

Aphasia is an acquired disturbance of language.

Lesions at various sites produce different types of aphasia; focal lesions do not cause total loss of all language functions simultaneously. The side of cerebral dominance for language can be determined by the Wada test (intracarotid amobarbital procedure, IAP), in which amobarbital is injected first into one internal carotid artery and then into the other, under angiographic control, to

selectively anesthetize each hemisphere (this is done, for example, before cortical resections for epilepsy). *Crossed aphasia*, i.e., aphasia due to a right hemispheric lesion in a right-handed patient, is rare. Aphasia usually improves markedly within a few weeks of onset and may continue to improve gradually over the first year, even if the symptoms temporarily appear to have stabilized. Improvement beyond one year is rare and usually minor.

Aphasia in bilingual and multilingual persons (usually) affects all of the languages spoken. The severity of involvement of each language depends on the age at which it was acquired, premorbid language ability, and whether the languages were learned simultaneously or sequentiallly. Aphasia is most commonly due to stroke or head trauma and may be accompanied by apraxia.

Global aphasia involves all aspects of language and severely impairs spoken communication.

The patient cannot speak spontaneously or can only do so with great effort, producing no more than fragments of words. Speech comprehension is usually absent; at best, patients may recognize a few words, including their own name. Perseveration (persistent repetition of a single word/subject) and neologisms are prominent, and the ability to repeat heard words is markedly impaired. Patients have great difficulty naming objects, reading, writing, and copying letters or words. Their ability to name objects, read, and write, except for the ability to copy letters of the alphabet or isolated words, is greatly impaired.

Language automatism (repetition of gibberish) is a characteristic feature. *Site of lesion:* Entire distribution of the middle cerebral artery, including both Broca's and Wernicke's areas.

Broca's aphasia (also called anterior, motor, or expressive aphasia) is characterized by the absence or severe impairment of spontaneous speech, while comprehension is only mildly impaired. The patient can speak only with great effort, producing only faltering, nonfluent, garbled words. Phonemic paraphasic errors are made, and sentences are of simple construction, often with isolated words that are not grammatically linked (agrammatism, "telegraphic" speech).

Naming, repetition, reading out loud, and writing are also impaired. *Site of lesion:* Broca area; may be due to infarction in the distribution of the prerolandic artery (artery of the precentral sulcus).

Wernicke's aphasia (also called posterior, sensory, or receptive aphasia) is characterized by severe impairment of comprehension. Spontaneous speech remains fluent and normally paced, but paragrammatism, paraphasia, and neologisms make the patient's speech partially or totally incomprehensible (word salad, jargon aphasia). Naming, repetition of heard words, reading, and writing are also markedly impaired. *Site of lesion:* Wernicke's area (area 22). May be due to infarction in the distribution of the posterior temporal artery.

Transcortical aphasia. Heard words can be repeated, but other linguistic functions are impaired: spontaneous speech in transcortical *motor* aphasia (syndrome similar to Broca's aphasia), language comprehension in transcortical *sensory* aphasia (syndrome similar to Wernicke's aphasia). *Site of lesion:* Motor type, left frontal lobe bordering on Broca's area; sensory type, left temporo-occipital junction dorsal to Wernicke's area. Watershed infarction is the most common cause.

Amnestic (anomic) aphasia. This type of aphasia is characterized by impaired naming and wordfinding. Spontaneous speech is fluent but permeated with word-finding difficulty and paraphrasing. The ability to repeat, comprehend, and write words is essentially normal. *Site of lesion:* Temporoparietal cortex or subcortical white matter.

Conduction aphasia. Repetition is severely impaired; fluent, spontaneous speech is interrupted by pauses to search for words and by phonemic paraphasia. Language comprehension is only mildly impaired. *Site of lesion:* Arcuate fasciculus or insular region.

Subcortical aphasia. Types of aphasia similar to those described may be produced by subcortical lesions at various sites (thalamus, internal capsule, anterior striatum).

Agraphia.

Agraphia is the acquired inability to write. Agraphia may be isolated (due to a lesion located in area 6, the superior parietal lobule, or elsewhere) or accompanied by other disturbances: aphasic agraphia is fluent or nonfluent, depending on the accompanying aphasia; apraxic agraphia is due to a lesion of the dominant parietal lobe; spatial agraphia, in which the patient has difficulty writing on a line and only writes on the right side of the paper, is due to a lesion of the nondominant parietal lobe; alexia with agraphia may be seen in the absence of aphasia. Micrographia (abnormally small handwriting) is found in Parkinson disease and is not pathogenetically related to agraphia. Various forms of agraphia are common in Alzheimer disease.

Examination: The patient is asked to write sentences, long words, or series of numbers to dictation, to spell words, and to copy written words.

Alexia.

Alexia is the acquired inability to read. In isolated alexia (alexia without agraphia), thepatient cannot recognize entire words or read them quickly, but can decipher them letter by letter, and can understand verbally spelled words. The ability to write is unaffected. The responsible lesion is typically in the left temporooccipital region with involvement of the visual pathway and of callosal fibers. Anterior alexia (difficulty and errors in reading aloud; impaired ability to write, spell, and copy words) is usually associated with Broca's aphasia. Central alexia (combination of alexia and agraphia) is usually accompanied by right-left disorientation, finger agnosia, agraphia, and acalculia (**Gerstmann syndrome; lesions of the angular and supramarginal gyri**), or by Wernicke's aphasia. Other features include the inability to understand written language or to spell, write, or copy words.

Examination: The patient is asked to read aloud and to read individual words, letters, and numbers; the understanding of spelled words and instructions is tested.

Acalculia.

Acalculia is an acquired inability to use numbers or perform simple arithmetical calculations. Patients have difficulty counting change, using a thermometer, or filling out a check. Lesions of various types may cause acalculia.

Examination: The patient is asked to perform simple arithmetical calculations and to read numbers.

Apraxia.

There are several kinds of apraxia; in general, the term refers to the inability to carry out learned motor tasks or purposeful movements. Apraxia is often accompanied by aphasia.

Ideomotor apraxia involves the faulty execution (parapraxia) of acquired voluntary and complex movement sequences; it can be demonstrated most clearly by asking the patient to perform pantomimic gestures. It can involve the face (buccofacial apraxia) or the limbs (limb apraxia). It is due to a lesion in the association fiber pathways connecting the language, visual, and motor areas to each other and to the two hemispheres (disconnection syndrome).

Examination

(pantomimic gestures on command): face (open eyes, stick out tongue, lick lips, blow out a match, pucker, suck on a straw); arms (turn a screw, cut paper, throw ball, comb hair, brush teeth, snap fingers); legs (kick ball, stamp out cigarette, climb stairs). The patient may perform the

movement in incorrect sequence, or may carry out a movement of the wrong type (e. g., puffing instead of sucking).

Ideational apraxia is impairment of the ability to carry out complex, learned, goal-directed activities in proper logical sequence. A temporal or parietal lesion may be responsible.

Examination: The patient is asked to carry out pantomimic gestures such as opening a letter, making a sandwich, or preparing a cup of tea.

Apraxia-like syndromes. The following disturbances are termed "apraxia" even though actual parapraxia is absent: Lid-opening apraxia is difficulty opening the eyes on command. Gait apraxia is characterized by difficulty initiating gait and by short steps. Dressing apraxia is often seen in patients with nondominant parietal lobe lesions. They cannot dress themselves and do not know how to position a shirt, shoes, trousers, or other items of clothing to put them on correctly. An underlying impairment of spatial orientation is responsible.

Agnosia is defined as a disturbance of recognition in which perception, attention, and general intelligence are (largely) unimpaired.

Disturbances of body image perception

Autotopagnosia (body-image agnosia) is the inability to correctly orient or perceive different body parts; patients cannot obey commands to point to parts of their own or the examiner's body (e. g., foot, hand, nose). The responsible lesion is usually, though not always, in the temporoparietal region (angular and supramarginal gyri). An aphasic patient may appear to have autotopagnosia because he cannot understand verbal instructions, but aphasia may also coexist with true autotopagnosia.

Finger agnosia is the inability to identify, name, or point to fingers. These patients can not mimic the examiner's finger movements or copy finger movements of their own contralateral hidden hand with the affected hand.

Right–left disorientation is the inability to distinguish the right and left sides of one's own or another's body; these patients cannot obey a command to raise their left hand or touch it to their right ear. This type of disorientation can cause *dressing apraxia* and similar problems.

Anosognosia is the unawareness or denial of a neurological deficit, such as hemiplegia. Patients may claim that they only want to give the paralyzed side a rest, or attempt to demonstrate that their condition has improved without realizing that they are moving the limb on the unaffected side. Most such patients have extensive lesions of the nondominant hemisphere. Anosognosia may also accompany visual field defects due to unilateral or bilateral lesions of the visual cortex (homonymous hemianopsia, cortical blindness). The most striking example of this is *Anton syndrome*, in which cortically blind patients act as if they could see, and will even "describe" details of their surroundings (incorrectly) without hesitation.

Disturbances of Spatial Orientation

A number of different types of agnosia impair the awareness of one's position relative to the surroundings, i.e., *spatial orientation*. Parietooccipital lesions are commonly responsible.

Constructional apraxia is characterized by the inability to represent spatial relationships in drawings, or with building blocks. Affected patients cannot copy a picture of a bicycle or clock. Everyday activities are impaired by the inability to draw diagrams, read (analog) clocks, assemble pieces of equipment or tools, or write words in the correct order (*spatial agraphia*).

Hemineglect is the inability to consciously perceive, react to, or classify stimuli on one side in the absence of a sensorimotor deficit or exceeding what one would expect from the severity of the sensorimotor deficit present. Hemineglect may involve unawareness of one side of the body (one-sided tooth brushing, shaving, etc.) or of one side of an object (food may be eaten from only

one side of the plate, eyeglasses may be looked for on only one side of the room). When addressed, the patient always turns to the healthy side. Neurological examination reveals that double simultaneous stimulation (touch, finger movement) of homologous body parts (same site, e. g., face or arm) is not felt on the affected side (*extinction phenomenon*). In addition, perception of stimuli on the affected side is quantitatively lower than on the healthy side, there is limb akinesia despite normal strength on the side of the lesion, and spatial orientation is impaired (e. g., the patient copies only half of a clock-face).

Methods of diagnosis of aphasias

Spontaneous speech. The aphasia examination starts by the examiner listening to the quality and quantity of spontaneous speech. No aphasia is pure; all have elements of expressive and receptive deficits. Difficulty with prosody and fluency argue strongly for an anterior aphasia while normal fluency and prosody is characteristic of a posterior aphasia. Anterior aphasias are accompanied by hemiparesis and subtle ideomotor apraxias as well as agrammatisms. Posterior aphasias have a pressure of speech, the rhythm and prosody are normal, but speech is often contaminated by translittoral aphasic errors (two consonants used together), word substitutions (green for red) and neologisms ("garunch" for garage). The patient often uses a long sentence to describe a noun to overcome failure to find the correct word ("it is used to write" for "pen").

Comprehension. Patients can utilize both hemispheres to perform midline commands such as "Stick your tongue out," "Get up" and "Close your eyes." Comprehension needs to be tested by commands that are off the midline. Severely affected patients should be able to answer simple questions with a yes or no ("Is it a sunny day?"). A patient is asked to point up or down with the thumb or to point out objects in the room. The patient may then be asked to place specific objects in specific places.

Naming objects. All aphasias have a degree of nominal aphasia. Lesions of the supramarginal gyrus may give a rather pure nominal aphasia. The patient is asked to name small parts of common objects such as a watch or pen. The examiner notes the speed of the patient's response, paraphasias, neologisms and perseveration. A patient who names small parts of common objects quickly and correctly is not aphasic. The test can be sharpened by asking the patient to pick out objects in front of him or her by name.

Repetition

The patient is asked to repeat a simple sentence and then a complex word. This test evaluates Wernicke's area (decodes language), the arcuate fasciculus that connects Wernicke's and Broca's areas which initiates expressive speech. Patients with a conduction aphasia, a disconnection between Broca's and Wernicke's areas, often do better with a complicated word than a simple one. If asked to repeat "Today is a sunny day," they may be hesitant and stammer "Today sunny." They may easily enunciate "presidential address."

Reading. The posterior parietal areas 39–41 as well as Wernicke's area are critically important for reading and decoding language. The patient is asked to read individual words, sentences and then instructions to perform specific actions.

Writing. Errors in grammar are most characteristic of anterior aphasias. All posterior aphasic patients have writing disabilities. Exner's area, immediately anterior to Broca's area, in addition to Wernicke's and areas 39–41 seems to be important for writing. If this area is damaged, patient's have more difficulty writing than their degree of weakness would suggest. Patients are asked to write their name and address, to take dictation and to write a few sentences about the weather.

Calculation. This is not strictly a part of the examination for aphasia, but is important as a component of posterior aphasia pathology and particularly von Gerstmann's syndrome. Dyscalculia is frequently seen with posterior parietal deficits.

Method of diagnosis of apraxias

Methods of testing

The examiner must be certain that the patient has no deficits of strength sensation or coordination that would interfere with his or her ability to perform the required tasks. The examiner asks the patient to perform a series of tasks to test specific forms of apraxia. The major apraxias tested are the following:

1 visual;
2 oral buccal lingual;
3 gait;
4 ideomotor;
5 ideational;
6 constructional;
7 callosal.

Visual praxis. The patient is asked to copy the position of the examiner's hand.

The examiner demonstrates the posture to be copied and then withdraws the hand. The patient must be able to see the requested hand position, hear and understand the task, and then form the motor program to perform it. The latter requires the patient to form an "engram" of the desired movement and then relay this to the primary motor cortex. Lesions in the prefrontal cortex and supplementary motor areas preclude the patient from making the engram and he or she will make a fist or place the fingers in an incorrect position.

Oral buccal lingual apraxia.

The patient is asked to touch a tongue blade with the tongue and to touch it above the horizontal. It is striking that most patients with this form of apraxia are unable to lift their tongue above the horizontal to touch the blade. They have difficulty moving the tongue side-to-side outside of the mouth or to touch the tongue blade as requested. This often reflects degeneration of the frontal operculum which is the cortical region for all activities requiring cranial nerves involved with tongue movements, swallowing and breathing. Coughing, sneezing and involuntary movements are preserved.

Gait apraxia. The patient may have two forms of gait apraxia. The first is a "magnetic gait" in which the foot seems to be stuck to the floor and the patient can only move a few inches before it becomes stuck again. This appears to be a foot grasp. "Egg-walking" is striking. The patient gently picks the feet up as if walking on eggs, but does not advance. These gaits are caused by failure to activate various brainstem locomotor centers, particularly the nucleus cuneiformis of the midbrain. In clinical practice, it is most commonly seen with normal pressure hydrocephalus in which the descending motor fibers to the legs are closest to the dilating lateral ventricles and are compromised. These gaits are frequently seen in patients with degenerative processes and lacunar strokes that affect the descending corticospinal pathways as well as basal ganglion diseases.

Ideomotor apraxia. This form of apraxia refers to the inability of a patient to carry out a single purposeful movement such as a salute or to demonstrate how to turn a key in a lock or to comb one's hair. Patients understand the task and are able to formulate how it should be performed, but cannot perform it on command. Patients often substitute a body part for the object, such as using the index finger as a comb rather than demonstrating how to hold a comb. Patients may perform better with an object (transitive) than without (intransitive). Ideomotor apraxias may be seen with conductive aphasias. Limb-kinetic apraxia is a judgment call. The patient has a certain clumsiness when performing a simple task that is out of proportion to his or her weakness. The lack of ability to form an engram prevents the patient from imitating an act that utilizes objects.

Ideational apraxia. The patient is unable to correctly carry out the sequence of a common task although each of its component parts can be successfully performed. If given a package of cigarettes and a matchbox and asked how he or she would light a cigarette and smoke it, the patient could remove the cigarette from the package, but would strike the match against the package and fail to put the cigarette in the mouth. Lesions causing ideational apraxia are primarily in the posterior parietal areas 5 and 7. Patients may have an inability to handle real objects even though they can mimic the use of an object (the opposite of ideomotor apraxia).

Constructional apraxia. This is the inability of a patient to construct or copy a visually presented object with blocks or by drawing. The examiner may utilize four matches to construct a box and then ask the patient to copy the construction. The angles are not placed correctly and often the heads of the matches do not correspond to the examiner's construct. The examiner must be sure that the patient can perceive the elements of the object, their spatial relationships and has the strength and coordination to perform the task. Left and right parietal lesions may cause a constructional apraxia. Right-sided lesions are more commonly associated with constructional apraxia than left and are often associated with some degree of neglect of the left side. Patients with left parietal lesions and constructional apraxia may have a concomitant fluent aphasia. A deficit of visual constructive ability is hard evidence of a parietal lesion. The supramarginal gyrus projections to the motor cortex are affected.

Callosal apraxia. Patients with callosal apraxia are unable to perform a simple task on command with the left hand. To perform a task with the left hand the patient must hear and understand the test which is accomplished by Wernicke's area (posterior one-third of the superior temporal gyrus). The information must cross the midline anteriorly in the corpus callosum to synapse in the right prefrontal area which in turn relays the information to the right primary motor cortex for execution. Lesions of both prefrontal cortices and the anterior corpus callosum can interrupt this distributed system and cause inability to utilize the left hand on command. The usual lesions that cause this deficit are prefrontal branch occlusions of the superior division of the middle cerebral artery, strokes of the anterior cerebral artery, callosotomy for epilepsy and frontal lobe gliomas.

Apraxia of eyelid opening. Patients are unable to open their eyes to command. They frequently tape the lids to their glasses. They often think they are blind but pain and startle responses open their eyes. The lesion is in the second frontal convolution of the frontal eye fields.

Dressing apraxia. The patient has inordinate difficulty in dressing or undressing themselves. When given a garment, they may attempt to put it on backwards or upside down. Some just stand and appear blank as they have no concept of how to start the dressing process. This apraxia is most often noted with right occipitoparietal or bilateral occipitoparietal lesions. It is usually not noted in isolation and has elements of neglect and ideational apraxia.

Movement and task-specific praxis. Conceptual praxis is the associative knowledge of tool action while mechanical knowledge refers to the advantage tools offer. Frontal lobe degenerations cause conceptual praxis.

Method of diagnosis of agnosias

Examining for agnosia. The patient is shown an array of common small objects and is asked to name them, describe their use and to pick out specific ones named by the examiner. If the patient is unable to do this visually, he or she is allowed to palpate the object and is asked the same questions. The patient is shown several different colors and asked to name them, match them with duplicates and then to arrange them in shades of increasing darkness or lightness. The patient is asked to walk to a specific location in a room when by doing so he or she would have to circumvent objects.

Visual agnosia. This is a disorder of higher cortical function in which an alert, intelligent, non-aphasic patient with normal visual perception cannot recognize a visual stimulus. The patient is unable to name or describe the function of objects shown, but immediately identifies them by touch or noise (bell) or smell (rose). A patient with a nominal aphasia cannot name the object by any modality of presentation (visual object agnosia). The usual lesion for this deficit is the second and third gyri of the dominant occipital lobe and its adjacent white matter outflow tracts. The patient may also be unable to identify or match colors which is visual agnosia for colors. There are several rather specific constellations of visual agnosia, which can be easily recognized and are quite striking.

Prosopagnosia. This is a visual agnosia in which the patient cannot recognize previously known faces and learn new ones but can do so through other modalities. The most striking example

of this is a husband who cannot recognize his wife by sight, but does so immediately when she speaks. These patients are unable to recognize visual stimuli of a group that has subcomponents. Often, they can identify a class of a visual stimulus but are unable to identify a specific member within the generic class. The usual lesions associated with prosopagnosia are in the inferior or mesial visual association cortices of the lingual and fusiform gyri of the temporal lobe or their adjacent white matter. The problem lies in the patient's inability to access associated information from contextual memory banks.

Visual object agnosia. These patients are unable to recognize the generic class of an object. The finding is frequently clouded by both a nominal aphasia and alexia. Some patients complain that their vision is unclear when scanning the static object, but can recognize it when it is moved or rotated. This may be a defect of interpreting static low-contrast stimuli, which is overcome by movement which evokes high-contrast stimulus interpretations. The usual lesions identified are bilateral in the ventral and mesial part of the occipital second and third gyri.

Disorder of color perceptions. These are defects of color perception in all or part of the visual field with preservation of formed vision. Most often, the patient reports dull or washed out colors in the affected visual field, but when severe, he or she sees objects only as black or white. They have normal vision in the colorless portion of the visual field deficit. The usual lesion is in the left occipitotemporal cortex which may be associated with alexia. Rare patients with this deficit have lesions in the occipitotemporal white matter or superior occipital lobe.

Disorders of color naming. These patients can match colors, but are unable to name them. They have a concomitant right homonymous hemianopia, pure alexia, but intact color perception in the left visual field. The usual defect is between the occipital and temporal lobe of the dominant hemisphere. Some patients perform better when given the color's name and pointing them out rather than naming them on demand.

Visual agnosia for spaces. Patients with this deficit are incapable of maneuvering around obstructions or to go from one point to another. The usual lesions are bilateral in the posterior inferior parietal lobes (area 7). It may occur with unilateral lesions in which the patient will always turn to the ipsilateral side and will return to the starting point. This is usually a non-dominant parietal lesion. In its mildest form, migraine patients complain of visual disorientation when spreading depression affects the parietal lobe.

Tactile recognition. The patient must have normal sensation in both hands. He or she is then asked to close the eyes and several common objects are placed first in one hand and then the other. The patient is asked to describe their texture, size, shape and use. If he or she cannot accomplish this, the patient is allowed to look at, hear or smell the object. Complete absence of the ability to describe details of the object is usually astereognosis. If the patient can describe its size, shape and texture, but is unable to name it or describe its use by touch alone but does so with vision, the defect is tactile agnosia. It frequently coexists with visual object agnosia and is secondary to a lesion of the contralateral supramarginal gyrus. In a left-handed person, the defect could be in the corpus callosum (sensory information decoded in the right hemisphere cannot be transferred to the left supramarginal gyrus).

Auditory recognition. The patient has to have normal hearing. The examiner asks the patient to close the eyes and then uses a bell, rattles coins or whistles and the patient is asked to identify the sounds. If a patient cannot recognize the sounds made by objects, but does so by sight or palpation he or she has auditory agnosia. These patients may have word deafness, so the instructions for the examination may have to be presented in written form. The usual lesion is in the posterior one-third of the dominant superior and medial temporal lobe.

The parietal lobe and disorders of the body scheme.

Normally, a person knows at any given point in time the position of their body in space and its functional capacity. There is also a sense of the relation of the body in horizontal and vertical space. In the upright position, this sense of the midline of the body to a vertical axis is called the subjective visual vertical. The right parietal lobe is specialized for this body awareness, as it is for

the patient's perception of near space (that which they can touch) and for space beyond their grasp. These abilities can be affected together or dissociated one from the other with lesions of the right parietal lobe.

Examination. The patient is asked to move the right and left hands. The examiner may then place his or her hands crossed behind the back and face the patient with the back turned and ask the patient to identify the right hand. The patient is asked to point out parts of his or her own body with each hand, with particular attention to fingers. The examiner may interlock the patient's fingers with his or her own and then ask the patient to identify individual digits. The examiner draws attention to the paralyzed or weak extremity. The examiner inquires if there is anything wrong with it and if it is part of the body. The patient is then asked to move it. Many patients will have to think for a second or two to identify the right and left part of the body. If this is a problem, the examiner asks the patient to take his or her right hand and touch the left ear. Failing to cross the midline and to recognize the right from the left side of the body is usually a right parietal lesion.

Failure to identify a part of the body is autotopagnosia. Failure to recognize a side of the body is asomatotopagnosia. Failure to identify specific fingers is striking and is finger agnosia. The easiest finger to recognize is the thumb and the hardest is the fourth finger.

In non-dominant right parietal lobe lesions, the patient often neglects the left side of the body, is unaware of the deficit and may deny body parts. This often takes some bizarre turns as the patient may ascribe the hemiplegia to fatigue: "I used my arm yesterday and now it is resting." It may assume a negative quality: "Get this other arm out of my bed." Parts of an extremity may be denied. The patient agrees that the upper arm belongs to them, but the associated connected hand does not. As the stroke clears, patients recognize more of the affected body part. Anosognosia is denial of illness or dysfunction, which may also occur with deafness and blindness as well as motor function. The opposite of denial of a body part is the phantom limb phenomenon of amputees. The patient imagines and feels the existence of the phantom. It is often exaggerated or distorted in size. It may appear on the stump of the extremity with the proximal component of the amputated part missing. It is frequently distorted, an example of which is one finger being elongated. Stroking the stump may evoke the phantom. The phantom is often painful and feels as if it is being twisted or crushed or stuffed into a shoe that is too small. There is clear physiologic reorganization of the sensory cortex in the absence of an extremity. Intact adjacent cortical areas innervate the prior territory of the amputated part. Phantoms shrink in size with time.

Cerebrospinal fluid (CSF) is a clear, colorless body fluid found in the brain and spine. It is produced in the choroid plexuses of the ventricles of the brain. It acts as a cushion or buffer for the brain's cortex, providing basic mechanical and immunological protection to the brain inside the skull. The CSF also serves a vital function in cerebral autoregulation of cerebral blood flow.

The CSF occupies the subarachnoid space (between the arachnoid mater and the pia mater) and the ventricular system around and inside the brain and spinal cord. It constitutes the content of the ventricles, cisterns, and sulci of the brain, as well as the central canal of the spinal cord.

The brain produces roughly 500 mL of cerebrospinal fluid per day. This fluid is constantly reabsorbed, so that only 100-160 mL is present at any one time.

Ependymal cells of the choroid plexus produce more than two thirds of CSF. The choroid plexus is a venous plexus contained within the four ventricles of the brain, hollow structures inside the brain filled with CSF. The remainder of the CSF is produced by the surfaces of the ventricles and by the lining surrounding the subarachnoid space.

Ependymal cells actively secrete sodium into the lateral ventricles. This creates osmotic pressure and draws water into the CSF space. Chloride, with a negative charge, moves with the positively charged sodium and a neutral charge is maintained. As a result, CSF contains a higher concentration of sodium and chloride than blood plasma, but less potassium, calcium and glucose and protein.

CSF circulates within the ventricular system of the brain. The ventricles are a series of cavities filled with CSF, inside the brain. The majority of CSF is produced from within the two

lateral ventricles. From here, the CSF passes through the interventricular foramina to the third ventricle, then the cerebral aqueduct to the fourth ventricle. The fourth ventricle is an outpouching on the posterior part of the brainstem. From the fourth ventricle, the fluid passes through three openings to enter the subarachnoid space – these are the median aperture, and the lateral apertures. The subarachnoid space covers the brain and spinal cord.

The CSF moves in a pulsatile manner throughout the CSF system with a nearly zero net flow, as shown on an MRI.

It had been thought that CSF returns to the vascular system by entering the dural venous sinuses via the arachnoid granulations (or villi). However, some have suggested that CSF flow along the cranial nerves and spinal nerve roots allow it into the lymphatic channels; this flow may play a substantial role in CSF reabsorbtion, in particular in the neonate, in which arachnoid granulations are sparsely distributed. The flow of CSF to the nasal submucosal lymphatic channels through the cribriform plate seems to be especially important.

The CSF contains approximately 0.3% plasma proteins, or approximately 15 to 40 mg/dL, depending on sampling site, and it is produced at a rate of 500 ml/day. Since the subarachnoid space around the brain and spinal cord can contain only 135 to 150 ml, large amounts are drained primarily into the blood through arachnoid granulations in the superior sagittal sinus. Thus the CSF turns over about 3.7 times a day. This continuous flow into the venous system dilutes the concentration of larger, lipid-insoluble molecules penetrating the brain and CSF.

The meninges lie immediately deep to the inner surface of the skull and constitute the membranous covering of the brain. The pericranium of the inner surface of the skull and the dura mater are collectively termed the pachymeninges, while the pia mater and arachnoid membrane are the leptomeninges. Between the dura mater and the arachnoid is the (normally only virtual) subdural space; between the arachnoid and the pia mater is the subarachnoid space. The subarachnoid space contains the cerebrospinal fluid (CSF). The functions of CSF are physical (compensation for volume changes, buffering and equal distribution of intracranial pressure despite variation in venous andarterial blood pressure) and metabolic (transport of nutrients and hormones into the brain). The cerebrospinal fluid is formed in the choroid plexuses of the four cere- bral ventricles (right and left lateral ventricles, third ventricle, and fourth ventricle). It flows through the ventricular system, then enters the subarachnoid space surrounding the brain and spinal cord (external CSF space). It is resorbed in the arachnoid granulations of the superior sagittal sinus and in the perineural sheaths of the spinal cord.

Meningeal syndrome include in neck stiffness (passive flexion of the neck is restricted and painful), Kernig sign (resistance to passive extension of the knee while the hip is flexed), attempts at neck flexion, pressing the pubis may induce flexion of the hip or knee Brudzinski signs (upper, middle). Passive extension of the knee may induce flexion of the opposite hip or knee. and general cerebral signs (headache, vomiting, nausea, general hyperesthesia, seizures and changes of consciousness).

What is meningism? It’s a condition in which the symptoms simulate meningitis, but in which no actual inflammation of these membranes is present.

Meningeal signs, method of lumbar puncture and assesment of cerebrospinal fluid in normal and in meningitis of different etiologies.

A stiff neck suggests blood, pus or chemical irritation of the meninges.

The patient’s neck should be completely supple and on flexion the chin should touch the chest wall. The examiner must be careful not to flex the neck until it has been cleared by X-ray in any suspected trauma (concomitant fractured spine). A child may demonstrate classic flexion withdrawal of the lower extremities.

The Kernig sign is elicited by flexing the patient’s hip to a 90-degree angle and then attempting to passively straighten the leg at the knee; pain and tightness in the hamstring muscles

prevent completion of this maneuver. This sign should be present bilaterally to support a diagnosis of meningitis.

Flexion at the hip and knee in response to forward flexion of the neck is the upper Brudzinski sign.

Flexion at the hip and knee in response to the pressure on the pubic symphysis is the middle Brudzinski sign.

Flexion at the hip and knee of contralateral leg in response to assessment of Kernig's sign is the lower Brudzinski sign.

Lumbar puncture:

Contraindications

- Raised intracranial pressure (falling level of consciousness with falling pulse, rising bp, vomiting, focal signs, papilloedema). In general a CT scan should always be carried out prior to LP to exclude an obstructed CSF system
- Coagulopathy or low lewel of platelets (<50 Г— 109/L).
- Infection of the skin at the site of lumbar puncture.

Before performing a lumbar puncture is necessary to do a skin test for sensitivity to anesthetic.

You will need the following:

- Spinal needles.
- Dressing pack (sterile gauze, drapes, antiseptic, gloves, plaster).
- Local anaesthetic (e.g. 2% lignocaine), 5ml syringe.
- 3 sterile bottles for collecting cerebrospinal fluid (CSF) and glucose bottle.
- Manometer and 3-way tap for measuring the opening CSF pressure.

Procedure

For suspected meningitis, antibiotics should be given first.

- Explain the procedure to the patient.
- Spend time positioning the patient, this is crucial to success. Lie patient on their left side (or R side if you are left handed), with back on edge of bed, fully flexed (knees to chin), with a folded pillow between their legs, keeping the back perpendicular to the bed (Fig 27). Flexion separates the interspaces between the vertebrae.
- The safest site for LP is the L3-L4 interspace (the spinal cord ends at L1-L2). An imaginary line drawn between the iliac crests intersects the spine at the L4 process or L3-L4 space exactly. Mark the L 3-4 intervertebral space (e.g. with a ballpoint pen).
- Clean the skin widely and place the sterile drapes over the patient.
- Inject Local anaesthetic 0.25-0.5ml 2% lignocaine under skin at pen mark. Anaesthetize the deeper structures. Use the anaesthetic sparingly: this may distort the anatomy making the procedure difficult and unnecessarily longer.
- Insert the spinal needle (stilette in place) in the mid-line, aiming slightly cranially (towards umbilicus), horizontal to the bed. Do not advance the needle without the stylet in place.
- With experience, you will feel the resistance of the spinal ligaments, and then the dura, alternatively, periodically remove the stylet and look for escape of CSF. Replace the stylet before advancing.
- Measure CSF pressure with manometer and 3-way tap. CSF pressure is increased with anxiety, SAH, infection, space occupying lesion, benign intracranial hypertension, CCF.
- Collect 0.5-1.5ml fluid in 3 serially numbered bottles and remember to fill the glucose bottle.
- Send specimens promptly for microscopy, culture, protein, glucose (with a simultaneous plasma sample for comparison), and where appropriate, virology, syphilis serology, cytology for malignancy, oligoclonal bands (multiple sclerosis), cryptococcal antigen testing, India ink stains, and fungal culture.

- Remove needle and place a plaster over the site.
- Patient should lie flat for at least 6h and have hourly neurological observation and blood pressure measurement. Encourage fluid intake.

The initial CSF analysis needs to include measurement of the opening pressure, color (clear, turbid, or purulent), WBC count and differential, and levels of glucose and protein. Typically in bacterial meningitis, the CSF opening pressure is increased (>200 mm of CSF lying down and >35 mm Hg upright). The fluid is usually turbid or frankly purulent and contains predominantly (>90%) polymorphonuclear leukocytes. The CSF glucose level is very low, usually less than 50% of that found with measurement of concomitant serum glucose. A low glucose level (<40 mg/100 mL) is also found in some other types of microbial meningitides including L. monocytogenes, Mycobacterium tuberculosis, and Cryptococcus neoformans. Normal glucose levels are common in viral meningitis. Usually, CSF protein levels are increased, often greater than 100 mg/dL (reference range, <45 mg/dL). In patients with parameningeal foci, such as a brain or epidural spinal abscess, or with multiple septic emboli, CSF glucose may not be as low as with typical bacterial meningitis, even though in these instances the CSF protein level is significantly increased.

Control materials to the preparatory stage of the class:

Questions (right answer in bold):

Where are third neurons of all kinds of sensation? a) spinal ganglion; b) brain cortex; **c) thalamus;** d) spinal cord

Stocking-and-gloves sensory loss is typical for: a) mononeuropathy; **b) polyneuropathy**; c) cord lesion; d) brain lesion; e) all above mentioned

Light touch is examined with: a) a pin; b) a tuning fork; c) Romberg's test; **d) wisp of cotton wool;** e) test-tubes with cold or hot water

Where are first neurons of all kinds of sensation? **a) spinal ganglion;** b) brain cortex; c) thalamus; d) spinal cord

Where is second neurons of deep sensation?: a) posterior horn; b) anterior horn; c) thalamus; d) brain cortex; **e) medulla oblongata**

Vibration is examined with: a) a pin; **b) a tuning fork**; c) Romberg's test; d) a phonendoscope: e) all above mentioned

Loss of the power of expression by speech or writing is called: **a) motor (Broca's) aphasia;** b) sensory (Wernicke's) aphasia; c) agnosia; d) disarthria; e) apraxia

Stimulation of occipital lobe may cause: **a) visual hallucinations;** b) auditory hallucinations; c) visual agnosia; d) blindness

Can the destruction of frontal lobes cause ataxia?: **a) yes**; b) no; c) I don't know

Enumerate the signs of lesion of frontal lobe: **a) paresis, seizures, motor aphasia;** b) auditory hallucinations, sensory aphasia; c) blindness, visual hallucinations; d) astereognosis, loss of sensory; e) all above mentioned

Difficulty in evoking the names for object, conditions or qualities is named: a) motor (Broca's) aphasia; b) sensory (Wernicke's) aphasia; **c) amnestic aphasia;** d) disarthria; e) semantic aphasia

Point the main method for liquor research: a) EEG; b) computed tomography; c) magnetic resonance imaging; **d) lumbar puncture**

At which level is lumbar puncture usually performed?: a) L1-L2; b) L2-L3; **c) L3-L4**

Point the contraindications to lumbar puncture: a) meningitis; b) spinal cord tumors; **c) brain oedema;** d) subarachnoid hemorrhage

Describe the pathological changes in CSF test:
Turbid
Pressure: 250 mm H_2O
Protein: 3,5 g/L
Glucose: 1,6 mmol/L
RW: "negative"
Predominant cell type is 850 of neutrophils*10^6
Answer: **bacterial meningitis**

Describe the pathological changes in CSF test.
Transparent, saturated color
Pressure: 300 mm H_2O
Protein: 1,3 g/L
Glucose: 2,6 mmol/L
RW: "negative"
fibrin film is absent
Predominant cell type is 40*10^6 of lymphocytes
Answer: **viral meningitis**

Materials for self-control of quality of preparation:

1. The patient is asked to stay with feet together and arms out stretched. It's: **a) Romberg's test:** b) examination of two-point discrimination; c) examination of vibration

2. A 56-years-old male has suffered from diabetes mellitus for more than 8 years. 3 month ago mild ache, numbness and creeping sensation in lower legs and feet began. Now such sensation is in the upper limb. During examination it was detected that cranial nerves were without peculiarity, gait was ordinary, muscle strength was decreased Tendon reflexes from upper limbs were sluggish. Knee reflexes were sluggish, Achilles' reflexes are absent. Deep and superficial sensation was decreased down elbow joint and down lower legs.
What type of sensory disorders was described?
Answer: polyneural type.

3. A 59-years-old female complains of cramping pain in the left forehead region and left eye. During the examination loss of all kind of sense in forehead region, dorsum of nose and superior eyelid on the left were detected. Supraorbital point is sharply painful. Corneal and blinking reflexes are absent on the left. Describe the type of sensory disorders
Answer: mononeural type.

4. A 68-years-old patient was taken to the clinic after brain stroke. He complains of absence of movement in left limbs. During the examination lack of all kinds of sensitivity in right half of the body and face was detected. Describe the type of sensory disorders.
Answer: hemitype.

5. A 45-years-old female complains of acute belting pain in the right half of the chest. On neurological assessment vesicular appearance of skin rash at left half of thoracic cage in Th6-Th7 zone was detected. There is hyperestesia in this zone. Describe the type of sensory disorders and ground the lesion locus.
Answer: root type.

Betz cells are in the: a) 2nd layer; b) 3d layer; c) 4th layer; **d) 5th layer;** e) 6th layer

Destruction of frontal motor eye field causes: a) paresis of conjugate gaze to the opposite side; **b) paresis of the opposite side of body;** c) visual agnosia; d) hemianopia; e) blindness

What kind of ataxia is caused by lesion of frontal lobe?: **a) motor;** b) sensory; d) amnesic; e) semantic; f) all above mentioned

What signs of precentral gyrus lesion do you know?: a) paresthesias of opposite side of the body; b) paresthesias of ipsilateral side of the body; **c) paresis of opposite side of the body;** d) paresis of ipsilateral side of the body

Stimulation of the limbic lobe may cause: a) visual hallucination; b) auditory hallucinations; c) paresthesias; d) paresis; **e) olfactory hallucinations**

The patient has a tumor in the left frontal-parietal-temporal region. The abnormality is due to a glioblastoma multiforme. Which of the following was most likely to be her presenting symptom?

a. Aphasia
b. Neglect
c. Left hemiparesis
d. Left homonymous hemianopia
e. Alexia without agraphia

A 73-years-old female one year ago was treated for brain stroke at the left middle cerebral artery one year ago. Right-side hemiparesis was succsessfully rehabilitated. But the patient cannot speak and cannot understand people around her. What speech disorders does our patient have?
Answer: **Wernike´s aphasia**

Routine spinal fluid examination in a patient with spongiform encephalopathy would be expected to show which of the following?
a. No abnormalities on routine studies
b. Elevated protein
c. More than 100 lymphocytes
d. More than 1000 red blood cells
e. Decreased glucose

List Contraindications to lumbar puncture:
1. Suspected intracranial mass lesion.
2. Local infection overlying the site of puncture.
3. Coagulopathy.

4. Suspected spinal cord mass lesion.
5. All above mentioned

Lumbar puncture is indicated for the following purposes:
1. Diagnosis of meningitis and other infective or inflammatory disorders, subarachnoid hemorrhage, hepatic encephalopathy, meningeal malignancies, paraneoplastic disorders, or suspected abnormalities of intracranial pressure.
2. Assessment of the response to therapy in meningitis and other infective or inflammatory disorders.
3. Administration of intrathecal medications or radiologic contrast media.
4. Rarely, to reduce cerebrospinal fluid (CSF) pressure.
5. All above mentioned

What is the most common complication of lumbar puncture?
Meningitis
Encephalitis
Stroke
Post-lumbar-puncture headache

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources
1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical Class No. 3.

Theme: Localization of functions in the cerebral cortex. Syndromes of affection.

Actuality of theme.
The lesions of cortical areas are observed in patients with trauma, strokes e.t.c. The correct treatment depends on the right topical diagnosis and help achieve the better results of the treatment.

Aims of the class:
Educational aims:

- to acquaint students with importance of the normal functioning of cerebral cortex for normal patient's life.

- a student must know:Structure of cortex.
1. Types of aphasias.
2. Localization of centers of speech in a cortex.
3. Symptoms of lesion of angular gyrus.
4. Topography of motor, sensory, visual, taste, smell, auditory areas in cortex.
5. Meningeal symptoms

- to give possibility to capture skills to the students:
1. To make a diagnosis of a motor aphasia.
2. To make diagnosis of a sensory aphasia.
3. To make a diagnosis of an amnestic aphasia.
4. To learn disorders of speech, reading, writing, counting, right-left orientation
5. To make a diagnosis of menigitis

- to give to the students of ability clinically to expose patients with disorders of speech, reading, writing, counting, right-left orientation, with meningitis.

Educational aims are connected with:

- forming professionally of meaningful substructures of personality;
- the actual aspects of deontological, patriotic, professional, psychological, legal, ecological responsibility and others like that.

Educating whole related to:
- by forming at the students of ability to put the topical diagnosis of defeat of cerebrum.
- by the actual aspects of deontological, patriotic, professional, psychological, legal, ecological responsibility and others like that.

Table of contents of the class:

Aphasia is an acquired disturbance of language.

Lesions at various sites produce different types of aphasia; focal lesions do not cause total loss of all language functions simultaneously. The side of cerebral dominance for language can be determined by the Wada test (intracarotid amobarbital procedure, IAP), in which amobarbital is injected first into one internal carotid artery and then into the other, under angiographic control, to selectively anesthetize each hemisphere (this is done, for example, before cortical resections for epilepsy). *Crossed aphasia*, i.e., aphasia due to a right hemispheric lesion in a right-handed patient, is rare. Aphasia usually improves markedly within a few weeks of onset and may continue to improve gradually over the first year, even if the symptoms temporarily appear to have stabilized. Improvement beyond one year is rare and usually minor.

Aphasia in bilingual and multilingual persons (usually) affects all of the languages spoken. The severity of involvement of each language depends on the age at which it was acquired, premorbid language ability, and whether the languages were learned simultaneously or sequentiallly. Aphasia is most commonly due to stroke or head trauma and may be accompanied by apraxia.

Global aphasia involves all aspects of language and severely impairs spoken communication.

The patient cannot speak spontaneously or can only do so with great effort, producing no more than fragments of words. Speech comprehension is usually absent; at best, patients may recognize a few words, including their own name. Perseveration (persistent repetition of a single word/subject) and neologisms are prominent, and the ability to repeat heard words is markedly impaired. Patients have great difficulty naming objects, reading, writing, and copying letters or words. Their ability to name objects, read, and write, except for the ability to copy letters of the alphabet or isolated words, is greatly impaired.

Language automatism (repetition of gibberish) is a characteristic feature. *Site of lesion:* Entire distribution of the middle cerebral artery, including both Broca's and Wernicke's areas.

Broca's aphasia (also called anterior, motor, or expressive aphasia) is characterized by the absence or severe impairment of spontaneous speech, while comprehension is only mildly impaired. The patient can speak only with great effort, producing only faltering, nonfluent, garbled words. Phonemic paraphasic errors are made, and sentences are of simple construction, often with isolated words that are not grammatically linked (agrammatism, "telegraphic" speech).

Naming, repetition, reading out loud, and writing are also impaired. *Site of lesion:* Broca area; may be due to infarction in the distribution of the prerolandic artery (artery of the precentral sulcus).

Wernicke's aphasia (also called posterior, sensory, or receptive aphasia) is characterized by severe impairment of comprehension. Spontaneous speech remains fluent and normally paced, but paragrammatism, paraphasia, and neologisms make the patient's speech partially or totally incomprehensible (word salad, jargon aphasia). Naming, repetition of heard words, reading, and writing are also markedly impaired. *Site of lesion:* Wernicke's area (area 22). May be due to infarction in the distribution of the posterior temporal artery.

Transcortical aphasia. Heard words can be repeated, but other linguistic functions are impaired: spontaneous speech in transcortical *motor* aphasia (syndrome similar to Broca's aphasia), language comprehension in transcortical *sensory* aphasia (syndrome similar to Wernicke's aphasia). *Site of lesion:* Motor type, left frontal lobe bordering on Broca's area; sensory type, left temporo-occipital junction dorsal to Wernicke's area. Watershed infarction is the most common cause.

Amnestic (anomic) aphasia. This type of aphasia is characterized by impaired naming and wordfinding. Spontaneous speech is fluent but permeated with word-finding difficulty and paraphrasing. The ability to repeat, comprehend, and write words is essentially normal. *Site of lesion:* Temporoparietal cortex or subcortical white matter.

Conduction aphasia. Repetition is severely impaired; fluent, spontaneous speech is interrupted by pauses to search for words and by phonemic paraphasia. Language comprehension is only mildly impaired. *Site of lesion:* Arcuate fasciculus or insular region.

Subcortical aphasia. Types of aphasia similar to those described may be produced by subcortical lesions at various sites (thalamus, internal capsule, anterior striatum).

Agraphia.

Agraphia is the acquired inability to write. Agraphia may be isolated (due to a lesion located in area 6, the superior parietal lobule, or elsewhere) or accompanied by other disturbances: aphasic agraphia is fluent or nonfluent, depending on the accompanying aphasia; apraxic agraphia is due to a lesion of the dominant parietal lobe; spatial agraphia, in which the patient has difficulty writing on a line and only writes on the right side of the paper, is due to a lesion of the nondominant parietal lobe; alexia with agraphia may be seen in the absence of aphasia. Micrographia (abnormally small handwriting) is found in Parkinson disease and is not pathogenetically related to agraphia. Various forms of agraphia are common in Alzheimer disease.

Examination: The patient is asked to write sentences, long words, or series of numbers to dictation, to spell words, and to copy written words.

Alexia.

Alexia is the acquired inability to read. In isolated alexia (alexia without agraphia), thepatient cannot recognize entire words or read them quickly, but can decipher them letter by letter, and can understand verbally spelled words. The ability to write is unaffected. The responsible lesion is typically in the left temporooccipital region with involvement of the visual pathway and of callosal fibers. Anterior alexia (difficulty and errors in reading aloud; impaired ability to write, spell, and copy words) is usually associated with Broca's aphasia. Central alexia (combination of alexia and agraphia) is usually accompanied by right-left disorientation, finger agnosia, agraphia, and acalculia (**Gerstmann syndrome; lesions of the angular and supramarginal gyri**), or by Wernicke's aphasia. Other features include the inability to understand written language or to spell, write, or copy words.

Examination: The patient is asked to read aloud and to read individual words, letters, and numbers; the understanding of spelled words and instructions is tested.

Acalculia.

Acalculia is an acquired inability to use numbers or perform simple arithmetical calculations. Patients have difficulty counting change, using a thermometer, or filling out a check. Lesions of various types may cause acalculia.

Examination: The patient is asked to perform simple arithmetical calculations and to read numbers.

Apraxia.

There are several kinds of apraxia; in general, the term refers to the inability to carry out learned motor tasks or purposeful movements. Apraxia is often accompanied by aphasia.

Ideomotor apraxia involves the faulty execution (parapraxia) of acquired voluntary and complex movement sequences; it can be demonstrated most clearly by asking the patient to perform pantomimic gestures. It can involve the face (buccofacial apraxia) or the limbs (limb

apraxia). It is due to a lesion in the association fiber pathways connecting the language, visual, and motor areas to each other and to the two hemispheres (disconnection syndrome).

Examination

(pantomimic gestures on command): face (open eyes, stick out tongue, lick lips, blow out a match, pucker, suck on a straw); arms (turn a screw, cut paper, throw ball, comb hair, brush teeth, snap fingers); legs (kick ball, stamp out cigarette, climb stairs). The patient may perform the movement in incorrect sequence, or may carry out a movement of the wrong type (e. g., puffing instead of sucking).

Ideational apraxia is impairment of the ability to carry out complex, learned, goal-directed activities in proper logical sequence. A temporal or parietal lesion may be responsible.

Examination: The patient is asked to carry out pantomimic gestures such as opening a letter, making a sandwich, or preparing a cup of tea.

Apraxia-like syndromes. The following disturbances are termed "apraxia" even though actual parapraxia is absent: Lid-opening apraxia is difficulty opening the eyes on command. Gait apraxia is characterized by difficulty initiating gait and by short steps. Dressing apraxia is often seen in patients with nondominant parietal lobe lesions. They cannot dress themselves and do not know how to position a shirt, shoes, trousers, or other items of clothing to put them on correctly. An underlying impairment of spatial orientation is responsible.

Agnosia is defined as a disturbance of recognition in which perception, attention, and general intelligence are (largely) unimpaired.

Disturbances of body image perception

Autotopagnosia (body-image agnosia) is the inability to correctly orient or perceive different body parts; patients cannot obey commands to point to parts of their own or the examiner's body (e. g., foot, hand, nose). The responsible lesion is usually, though not always, in the temporoparietal region (angular and supramarginal gyri). An aphasic patient may appear to have autotopagnosia because he cannot understand verbal instructions, but aphasia may also coexist with true autotopagnosia.

Finger agnosia is the inability to identify, name, or point to fingers. These patients can not mimic the examiner's finger movements or copy finger movements of their own contralateral hidden hand with the affected hand.

Right–left disorientation is the inability to distinguish the right and left sides of one's own or another's body; these patients cannot obey a command to raise their left hand or touch it to their right ear. This type of disorientation can cause *dressing apraxia* and similar problems.

Anosognosia is the unawareness or denial of a neurological deficit, such as hemiplegia. Patients may claim that they only want to give the paralyzed side a rest, or attempt to demonstrate that their condition has improved without realizing that they are moving the limb on the unaffected side. Most such patients have extensive lesions of the nondominant hemisphere. Anosognosia may also accompany visual field defects due to unilateral or bilateral lesions of the visual cortex (homonymous hemianopsia, cortical blindness). The most striking example of this is *Anton syndrome*, in which cortically blind patients act as if they could see, and will even "describe" details of their surroundings (incorrectly) without hesitation.

Disturbances of Spatial Orientation

A number of different types of agnosia impair the awareness of one's position relative to the surroundings, i.e., *spatial orientation*. Parietooccipital lesions are commonly responsible.

Constructional apraxia is characterized by the inability to represent spatial relationships in drawings, or with building blocks. Affected patients cannot copy a picture of a bicycle or clock.

Everyday activities are impaired by the inability to draw diagrams, read (analog) clocks, assemble pieces of equipment or tools, or write words in the correct order (*spatial agraphia*).

Hemineglect is the inability to consciously perceive, react to, or classify stimuli on one side in the absence of a sensorimotor deficit or exceeding what one would expect from the severity of the sensorimotor deficit present. Hemineglect may involve unawareness of one side of the body (one-sided tooth brushing, shaving, etc.) or of one side of an object (food may be eaten from only one side of the plate, eyeglasses may be looked for on only one side of the room). When addressed, the patient always turns to the healthy side. Neurological examination reveals that double simultaneous stimulation (touch, finger movement) of homologous body parts (same site, e. g., face or arm) is not felt on the affected side (*extinction phenomenon*). In addition, perception of stimuli on the affected side is quantitatively lower than on the healthy side, there is limb akinesia despite normal strength on the side of the lesion, and spatial orientation is impaired (e. g., the patient copies only half of a clock-face).

Methods of diagnosis of aphasias:

Spontaneous speech. The aphasia examination starts by the examiner listening to the quality and quantity of spontaneous speech. No aphasia is pure; all have elements of expressive and receptive deficits. Difficulty with prosody and fluency argue strongly for an anterior aphasia while normal fluency and prosody is characteristic of a posterior aphasia. Anterior aphasias are accompanied by hemiparesis and subtle ideomotor apraxias as well as agrammatisms. Posterior aphasias have a pressure of speech, the rhythm and prosody are normal, but speech is often contaminated by translittoral aphasic errors (two consonants used together), word substitutions (green for red) and neologisms ("garunch" for garage). The patient often uses a long sentence to describe a noun to overcome failure to find the correct word ("it is used to write" for "pen").

Comprehension. Patients can utilize both hemispheres to perform midline commands such as "Stick your tongue out," "Get up" and "Close your eyes." Comprehension needs to be tested by commands that are off the midline. Severely affected patients should be able to answer simple questions with a yes or no ("Is it a sunny day?"). A patient is asked to point up or down with the thumb or to point out objects in the room. The patient may then be asked to place specific objects in specific places.

Naming objects. All aphasias have a degree of nominal aphasia. Lesions of the supramarginal gyrus may give a rather pure nominal aphasia. The patient is asked to name small parts of common objects such as a watch or pen. The examiner notes the speed of the patient's response, paraphasias, neologisms and perseveration. A patient who names small parts of common objects quickly and correctly is not aphasic. The test can be sharpened by asking the patient to pick out objects in front of him or her by name.

Repetition

The patient is asked to repeat a simple sentence and then a complex word. This test evaluates Wernicke's area (decodes language), the arcuate fasciculus that connects Wernicke's and Broca's areas which initiates expressive speech. Patients with a conduction aphasia, a disconnection between Broca's and Wernicke's areas, often do better with a complicated word than a simple one. If asked to repeat "Today is a sunny day," they may be hesitant and stammer "Today sunny." They may easily enunciate "presidential address."

Reading. The posterior parietal areas 39–41 as well as Wernicke's area are critically important for reading and decoding language. The patient is asked to read individual words, sentences and then instructions to perform specific actions.

Writing. Errors in grammar are most characteristic of anterior aphasias. All posterior aphasic patients have writing disabilities. Exner's area, immediately anterior to Broca's area, in addition to Wernicke's and areas 39–41 seems to be important for writing. If this area is damaged, patient's have more difficulty writing than their degree of weakness would suggest. Patients are asked to write their name and address, to take dictation and to write a few sentences about the

weather.

Calculation. This is not strictly a part of the examination for aphasia, but is important as a component of posterior aphasia pathology and particularly von Gerstmann's syndrome. Dyscalculia is frequently seen with posterior parietal deficits.

Method of diagnosis of apraxias

Methods of testing

The examiner must be certain that the patient has no deficits of strength sensation or coordination that would interfere with his or her ability to perform the required tasks. The examiner asks the patient to perform a series of tasks to test specific forms of apraxia. The major apraxias tested are the following:

1 visual;
2 oral buccal lingual;
3 gait;
4 ideomotor;
5 ideational;
6 constructional;
7 callosal.

Visual praxis. The patient is asked to copy the position of the examiner's hand.

The examiner demonstrates the posture to be copied and then withdraws the hand. The patient must be able to see the requested hand position, hear and understand the task, and then form the motor program to perform it. The latter requires the patient to form an "engram" of the desired movement and then relay this to the primary motor cortex. Lesions in the prefrontal cortex and supplementary motor areas preclude the patient from making the engram and he or she will make a fist or place the fingers in an incorrect position.

Oral buccal lingual apraxia.

The patient is asked to touch a tongue blade with the tongue and to touch it above the horizontal. It is striking that most patients with this form of apraxia are unable to lift their tongue above the horizontal to touch the blade. They have difficulty moving the tongue side-to-side outside of the mouth or to touch the tongue blade as requested. This often reflects degeneration of the frontal operculum which is the cortical region for all activities requiring cranial nerves involved with tongue movements, swallowing and breathing. Coughing, sneezing and involuntary movements are preserved.

Gait apraxia. The patient may have two forms of gait apraxia. The first is a "magnetic gait" in which the foot seems to be stuck to the floor and the patient can only move a few inches before it becomes stuck again. This appears to be a foot grasp. "Egg-walking" is striking. The patient gently picks the feet up as if walking on eggs, but does not advance. These gaits are caused by failure to activate various brainstem locomotor centers, particularly the nucleus cuneiformis of the midbrain. In clinical practice, it is most commonly seen with normal pressure hydrocephalus in which the descending motor fibers to the legs are closest to the dilating lateral ventricles and are compromised. These gaits are frequently seen in patients with degenerative processes and lacunar strokes that affect the descending corticospinal pathways as well as basal ganglion diseases.

Ideomotor apraxia. This form of apraxia refers to the inability of a patient to carry out a single purposeful movement such as a salute or to demonstrate how to turn a key in a lock or to comb one's hair. Patients understand the task and are able to formulate how it should be performed, but cannot perform it on command. Patients often substitute a body part for the object, such as using the index finger as a comb rather than demonstrating how to hold a comb. Patients may perform better with an object (transitive) than without (intransitive). Ideomotor apraxias may be seen with conductive aphasias. Limb-kinetic apraxia is a judgment call. The patient has a certain clumsiness when performing a simple task that is out of proportion to his or her weakness. The lack of ability to form an engram prevents the patient from imitating an act that utilizes objects.

Ideational apraxia. The patient is unable to correctly carry out the sequence of a common

task although each of its component parts can be successfully performed. If given a package of cigarettes and a matchbox and asked how he or she would light a cigarette and smoke it, the patient could remove the cigarette from the package, but would strike the match against the package and fail to put the cigarette in the mouth. Lesions causing ideational apraxia are primarily in the posterior parietal areas 5 and 7. Patients may have an inability to handle real objects even though they can mimic the use of an object (the opposite of ideomotor apraxia).

Constructional apraxia. This is the inability of a patient to construct or copy a visually presented object with blocks or by drawing. The examiner may utilize four matches to construct a box and then ask the patient to copy the construction. The angles are not placed correctly and often the heads of the matches do not correspond to the examiner's construct. The examiner must be sure that the patient can perceive the elements of the object, their spatial relationships and has the strength and coordination to perform the task. Left and right parietal lesions may cause a constructional apraxia. Right-sided lesions are more commonly associated with constructional apraxia than left and are often associated with some degree of neglect of the left side. Patients with left parietal lesions and constructional apraxia may have a concomitant fluent aphasia. A deficit of visual constructive ability is hard evidence of a parietal lesion. The supramarginal gyrus projections to the motor cortex are affected.

Callosal apraxia. Patients with callosal apraxia are unable to perform a simple task on command with the left hand. To perform a task with the left hand the patient must hear and understand the test which is accomplished by Wernicke's area (posterior one-third of the superior temporal gyrus). The information must cross the midline anteriorly in the corpus callosum to synapse in the right prefrontal area which in turn relays the information to the right primary motor cortex for execution. Lesions of both prefrontal cortices and the anterior corpus callosum can interrupt this distributed system and cause inability to utilize the left hand on command. The usual lesions that cause this deficit are prefrontal branch occlusions of the superior division of the middle cerebral artery, strokes of the anterior cerebral artery, callosotomy for epilepsy and frontal lobe gliomas.

Apraxia of eyelid opening. Patients are unable to open their eyes to command. They frequently tape the lids to their glasses. They often think they are blind but pain and startle responses open their eyes. The lesion is in the second frontal convolution of the frontal eye fields.

Dressing apraxia. The patient has inordinate difficulty in dressing or undressing themselves. When given a garment, they may attempt to put it on backwards or upside down. Some just stand and appear blank as they have no concept of how to start the dressing process. This apraxia is most often noted with right occipitoparietal or bilateral occipitoparietal lesions. It is usually not noted in isolation and has elements of neglect and ideational apraxia.

Movement and task-specific praxis. Conceptual praxis is the associative knowledge of tool action while mechanical knowledge refers to the advantage tools offer. Frontal lobe degenerations cause conceptual praxis.

Method of diagnosis of agnosias

Examining for agnosia. The patient is shown an array of common small objects and is asked to name them, describe their use and to pick out specific ones named by the examiner. If the patient is unable to do this visually, he or she is allowed to palpate the object and is asked the same questions. The patient is shown several different colors and asked to name them, match them with duplicates and then to arrange them in shades of increasing darkness or lightness. The patient is asked to walk to a specific location in a room when by doing so he or she would have to circumvent objects.

Visual agnosia. This is a disorder of higher cortical function in which an alert, intelligent, non-aphasic patient with normal visual perception cannot recognize a visual stimulus. The patient is unable to name or describe the function of objects shown, but immediately identifies them by touch or noise (bell) or smell (rose). A patient with a nominal aphasia cannot name the object by any modality of presentation (visual object agnosia). The usual lesion for this deficit is the second

and third gyri of the dominant occipital lobe and its adjacent white matter outflow tracts. The patient may also be unable to identify or match colors which is visual agnosia for colors. There are several rather specific constellations of visual agnosia, which can be easily recognized and are quite striking.

Prosopagnosia. This is a visual agnosia in which the patient cannot recognize previously known faces and learn new ones but can do so through other modalities. The most striking example of this is a husband who cannot recognize his wife by sight, but does so immediately when she speaks. These patients are unable to recognize visual stimuli of a group that has subcomponents. Often, they can identify a class of a visual stimulus but are unable to identify a specific member within the generic class. The usual lesions associated with prosopagnosia are in the inferior or mesial visual association cortices of the lingual and fusiform gyri of the temporal lobe or their adjacent white matter. The problem lies in the patient's inability to access associated information from contextual memory banks.

Visual object agnosia. These patients are unable to recognize the generic class of an object. The finding is frequently clouded by both a nominal aphasia and alexia. Some patients complain that their vision is unclear when scanning the static object, but can recognize it when it is moved or rotated. This may be a defect of interpreting static low-contrast stimuli, which is overcome by movement which evokes high-contrast stimulus interpretations. The usual lesions identified are bilateral in the ventral and mesial part of the occipital second and third gyri.

Disorder of color perceptions. These are defects of color perception in all or part of the visual field with preservation of formed vision. Most often, the patient reports dull or washed out colors in the affected visual field, but when severe, he or she sees objects only as black or white. They have normal vision in the colorless portion of the visual field deficit. The usual lesion is in the left occipitotemporal cortex which may be associated with alexia. Rare patients with this deficit have lesions in the occipitotemporal white matter or superior occipital lobe.

Disorders of color naming. These patients can match colors, but are unable to name them. They have a concomitant right homonymous hemianopia, pure alexia, but intact color perception in the left visual field. The usual defect is between the occipital and temporal lobe of the dominant hemisphere. Some patients perform better when given the color's name and pointing them out rather than naming them on demand.

Visual agnosia for spaces. Patients with this deficit are incapable of maneuvering around obstructions or to go from one point to another. The usual lesions are bilateral in the posterior inferior parietal lobes (area 7). It may occur with unilateral lesions in which the patient will always turn to the ipsilateral side and will return to the starting point. This is usually a non-dominant parietal lesion. In its mildest form, migraine patients complain of visual disorientation when spreading depression affects the parietal lobe.

Tactile recognition. The patient must have normal sensation in both hands. He or she is then asked to close the eyes and several common objects are placed first in one hand and then the other. The patient is asked to describe their texture, size, shape and use. If he or she cannot accomplish this, the patient is allowed to look at, hear or smell the object. Complete absence of the ability to describe details of the object is usually astereognosis. If the patient can describe its size, shape and texture, but is unable to name it or describe its use by touch alone but does so with vision, the defect is tactile agnosia. It frequently coexists with visual object agnosia and is secondary to a lesion of the contralateral supramarginal gyrus. In a left-handed person, the defect could be in the corpus callosum (sensory information decoded in the right hemisphere cannot be transferred to the left supramarginal gyrus).

Auditory recognition. The patient has to have normal hearing. The examiner asks the patient to close the eyes and then uses a bell, rattles coins or whistles and the patient is asked to identify the sounds. If a patient cannot recognize the sounds made by objects, but does so by sight or palpation he or she has auditory agnosia. These patients may have word deafness, so the instructions for the examination may have to be presented in written form. The usual lesion is in the posterior one-third of the dominant superior and medial temporal lobe.

The parietal lobe and disorders of the body scheme.

Normally, a person knows at any given point in time the position of their body in space and its functional capacity. There is also a sense of the relation of the body in horizontal and vertical space. In the upright position, this sense of the midline of the body to a vertical axis is called the subjective visual vertical. The right parietal lobe is specialized for this body awareness, as it is for the patient's perception of near space (that which they can touch) and for space beyond their grasp. These abilities can be affected together or dissociated one from the other with lesions of the right parietal lobe.

Examination. The patient is asked to move the right and left hands. The examiner may then place his or her hands crossed behind the back and face the patient with the back turned and ask the patient to identify the right hand. The patient is asked to point out parts of his or her own body with each hand, with particular attention to fingers. The examiner may interlock the patient's fingers with his or her own and then ask the patient to identify individual digits. The examiner draws attention to the paralyzed or weak extremity. The examiner inquires if there is anything wrong with it and if it is part of the body. The patient is then asked to move it. Many patients will have to think for a second or two to identify the right and left part of the body. If this is a problem, the examiner asks the patient to take his or her right hand and touch the left ear. Failing to cross the midline and to recognize the right from the left side of the body is usually a right parietal lesion.

Failure to identify a part of the body is ***autotopagnosia.*** Failure to recognize a side of the body is ***asomatotopagnosia.*** Failure to identify specific fingers is striking and is ***finger agnosia.*** The easiest finger to recognize is the thumb and the hardest is the fourth finger.

In non-dominant right parietal lobe lesions, the patient often neglects the left side of the body, is unaware of the deficit and may deny body parts. This often takes some bizarre turns as the patient may ascribe the hemiplegia to fatigue: "I used my arm yesterday and now it is resting." It may assume a negative quality: "Get this other arm out of my bed." Parts of an extremity may be denied. The patient agrees that the upper arm belongs to them, but the associated connected hand does not. As the stroke clears, patients recognize more of the affected body part***. Anosognosia*** is denial of illness or dysfunction, which may also occur with deafness and blindness as well as motor function. The opposite of denial of a body part is the ***phantom limb phenomenon of amputees***. The patient imagines and feels the existence of the phantom. It is often exaggerated or distorted in size. It may appear on the stump of the extremity with the proximal component of the amputated part missing. It is frequently distorted, an example of which is one finger being elongated. Stroking the stump may evoke the phantom. The phantom is often painful and feels as if it is being twisted or crushed or stuffed into a shoe that is too small. There is clear physiologic reorganization of the sensory cortex in the absence of an extremity. Intact adjacent cortical areas innervate the prior territory of the amputated part. Phantoms shrink in size with time.

Mini mental state examination (MMSE)

This test examines the patients' cognitive function. They are awarded a score out of 30. The scores available for each question are shown in brackets.

(1) Orientation: 'What is today's day? date? month? year? season?' (5)

'Where are we – country? county? city? hospital? ward/clinic?' (5)

(2) Memory (registration): 'I am going to name three objects. I want you to repeat them after me and then remember them, because I will ask you to name these objects in a few minutes – APPLE, BOOK, COAT'. Give one point for each one that they can repeat immediately. (3)

(3) Attention and concentration: 'Subtract 7 from 100. Keep subtracting 7 from each answer until I tell you to stop.' Maximum 5 answers (93, 86, 79, 72, 65). (5) or 'Spell WORLD backwards.' Score 1 point for each correctly placed letter. (5)

(4) Memory (recall): Ask the patient to repeat the objects named above. (3)

(5) Language:

Naming – Show the patient a pen and a watch, ask to name them. (2)

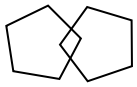
Repetition – Ask the patient to repeat 'No ifs, ands or buts'. (1)

Three-stage command – Ask the patient to take a piece of paper in the right hand, fold it in half and put it on the table. (3)

Reading – Ask the patient to read and obey a command written on paper, e.g. 'Close your eyes'. (1)

Writing – 'Write a sentence.' The sentence should have a verb and a subject. 'Go away' is not allowed! (1)

Copying – Ask the patient to copy a design, e.g. intersecting pentagons. (1)



TOTAL /30

Control materials to the preparatory stage of the class:

Questions (right answer in bold):

Loss of the power of expression by speech or writing is called: **a) motor (Broca's) aphasia;** b) sensory (Wernicke's) aphasia; c) agnosia; d) disarthria; e) apraxia

Stimulation of occipital lobe may cause: **a) visual hallucinations;** b) auditory hallucinations; c) visual agnosia; d) blindness

Can the destruction of frontal lobes cause ataxia?: **a) yes**; b) no; c) I don't know

Enumerate the signs of lesion of frontal lobe: **a) paresis, seizures, motor aphasia;** b) auditory hallucinations, sensory aphasia; c) blindness, visual hallucinations; d) astereognosis, loss of sensory; e) all above mentioned

Difficulty in evoking the names for object, conditions or qualities is named: a) motor (Broca's) aphasia; b) sensory (Wernicke's) aphasia; **c) amnestic aphasia;** d) disarthria; e) semantic aphasia

Materials for self-control of quality of preparation:

Questions (right answer in bold):

Betz cells are in the: a) 2nd layer; b) 3d layer; c) 4th layer; **d) 5th layer;** e) 6th layer

Destruction of frontal motor eye field causes: a) paresis of conjugate gaze to the opposite side; **b) paresis of the opposite side of body;** c) visual agnosia; d) hemianopia; e) blindness

What kind of ataxia is caused by lesion of frontal lobe?: **a) motor;** b) sensory; d) amnesic; e) semantic; f) all above mentioned

What signs of precentral gyrus lesion do you know?: a) paresthesias of opposite side of the body; b) paresthesias of ipsilateral side of the body; **c) paresis of opposite side of the body;** d) paresis of ipsilateral side of the body

Stimulation of the limbic lobe may cause: a) visual hallucination; b) auditory hallucinations; c) paresthesias; d) paresis; **e) olfactory hallucinations**

The patient has a tumor in the left frontal-parietal-temporal region. The abnormality is due to a glioblastoma multiforme. Which of the following was most likely to be her presenting symptom?

a. Aphasia

b. Neglect
c. Left hemiparesis
d. Left homonymous hemianopia
e. Alexia without agraphia

A 73-years-old female one year ago was treated for brain stroke at the left middle cerebral artery one year ago. Right-side hemiparesis was succsessfully rehabilitated. But the patient cannot speak and cannot understand people around her. What speech disorders does our patient have?
Answer: **Wernike´s aphasia**

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources

1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical Class No. 4

Theme: Vascular diseases of the brain.

Actuality of theme:
The vascular diseases of brain take leading seat among the nervous diseases and make the frequent reason of death, temporal or permanent loss or decline of capacity, and also acute states which need urgent help from the doctor of some speciality.

Aims of the class:
To know:
- Classification of acute and chronic disorders of cerebral blood circulation.

2) Etiologic factors and pathogenesis of acute disorders of cerebral blood circulation in patients with haemorrhage strokes.
3) Syndromes of lesion of anterior cerebral artery, middle cerebral artery and posterior cerebral artery.
4) Syndrome of occlusion and stenosis of main vessels of brain.

To be able:
to explore patients:
- With acute disorders of cerebral blood circulation and to set the type of stroke.
- To find the site of lesion.
- To give an appropriate treatment

Questions for discussion:
Classification of ischemic stroke
Treatment of ischemic stroke
Secondary prevention of stroke
Surgical treatment

Theme of reports, abstracts, reviews:
- Lacunar Syndromes
- Etiologic factors and pathogenesis of acute disorders of cerebral blood circulation in patients with haemorhrage strokes.
- Causes of intracranial hemorrhage (including intracerebral, subarachnoid, ventricular, and subdural)

Control questions:
A 67-year-old woman with a history of type 2 diabetes mellitus and atrial fibrillation presents to the emergency room with left body weakness and slurred speech. The onset was sudden while she was brushing her teeth 1 h ago, and she was brought immediately to the emergency room.
She denies word-finding difficulties, dysesthesia, and headache. She is taking warfarin. Physical exam findings include blood pressure of 205/90 and irregularly irregular heartbeat. There is left side neglect with slurred speech. There is a corticospinal pattern of weakness of the left body, with the face and upper extremity being worse than the lower extremity. Routine chemistries and cell counts are normal. Her INR is 1.8. Which of the following is the most appropriate first step in management?
a. Administer tissue plasminogen activator
b. Call a vascular surgery consult for possible endarterectomy
c. Order a brain CT
d. Order a cerebral angiogram
e. Start heparin

A pure motor stroke is most likely with damage to which of the following?
a. Internal capsule
b. Cerebellum
c. Putamen
d. Caudate
e. Amygdala

A pure sensory stroke is most likely with damage to which of the following?
a. Internal capsule
b. Thalamus
c. Hippocampus
d. Globus pallidus
e. Pons

A 61-year-old man with a history of hypertension has been in excellent health until he presents with vertigo and unsteadiness lasting for 2 days. He then develops nausea, vomiting, dysphagia, hoarseness, ataxia, left facial pain, and right-sided sensory loss. There is no weakness. On examination, he is alert, with a normal mental status. He vomits with head movement. There is skew deviation of the eyes, left ptosis, clumsiness of the left arm, and titubation. He has loss of pin and temperature sensation on the right arm and leg and decreased joint position sensation in the left foot. He is unable to walk. Magnetic resonance imaging (MRI) in this patient might be expected to show which of the following?
a. Basilar artery tip aneurysm
b. Right lateral medullary infarction
c. Left lateral medullary infarction
d. Left medial medullary infarction
e. Right medial medullary infarction

A 75-year-old man with a history of recent memory impairment is admitted with headache, confusion, and a left homonymous hemianopsia. He has recently had two episodes of brief unresponsiveness. There is no history of hypertension. Computed tomography (CT) scan shows a right occipital lobe hemorrhage with some subarachnoid extension of the blood. An MRI scan with gradient echo (susceptibility) sequences reveals foci of hemosiderin in the right temporal and left frontal cortex. Which of the following is the most likely cause of this patient's symptoms and signs?
a. Gliomatosis cerebri
b. Multi-infarct dementia
c. Mycotic aneurysm
d. Amyloid angiopathy
e. Undiagnosed hypertension

A thorough evaluation reveals that a 69-year-old patient has a symptomatic 90% stenosis of the right internal carotid artery at the bifurcation. Which of the following management options is most likely to prevent a future stroke?
a. Warfarin
b. Carotid artery angioplasty
c. Carotid endarterectomy
d. Extracranial-intracranial bypass
e. Aspirin

Focal weakness lasting for 24 h following a motor seizure is most likely attributable to which of the following?
a. Intracerebral hemorrhage
b. Subarachnoid hemorrhage
c. Encephalitis
d. Todd's paralysis
e. Hyponatremia

A 16-year-old girl with complex partial seizures and mild mental retardation has an area of deep red discoloration (port-wine nevus) extending over her forehead and left upper eyelid. A CT scan of her brain would be likely to reveal which of the following?
a. A hemangioblastoma
b. A Charcot-Bouchard aneurysm
c. An arteriovenous malformation
d. A leptomeningeal angioma
e. A fusiform aneurysm

A 72-year-old woman has the abrupt onset of right face and hand weakness, disturbed speech production, and a right homonymous hemianopsia. This is most likely attributable to occlusion of which of the following arteries?
a. Left middle cerebral artery
b. Left anterior cerebral artery
c. Left vertebrobasilar artery
d. Right anterior choroidal artery
e. Left posterior inferior cerebellar artery (PICA)

A 73-year-old man with a history of hypertension has a 10-min episode of left-sided weakness and slurred speech. On further questioning, he relates three brief episodes in the past month of sudden impairment of vision affecting the right eye. His examination now is normal. Which of the following is the most appropriate next diagnostic test?
a. Creatine phosphokinase (CPK)
b. Holter monitor
c. Visual evoked responses
d. Carotid artery Doppler ultrasound
e. Conventional cerebral angiography

Episodes of visual loss known as amaurosis fugax are most likely related to which of the following?
a. Retinal vein thrombosis
b. Central retinal artery ischemia
c. Posterior cerebral artery ischemia
d. Middle cerebral artery ischemia
e. Posterior ciliary artery ischemia

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b. Multi-infarct dementia
c. Mycotic aneurysm
d. Amyloid angiopathy
e. Undiagnosed hypertension

A 39-year-old woman has diplopia several times a day for 6 weeks. She consults a physician when the double vision becomes unremitting, and also mentions a dull pain behind her right eye. When a red glass is placed over her right eye and she is asked to look at a flashlight off to her left, she reports seeing a white light and a red light. The red light appears to her to be more to the left than the white light. Her right pupil is more dilated than her left pupil and responds less briskly to a bright light directed at it than does the left pupil. Before any further investigations can be performed, the woman develops the worst headache of her life and becomes stuporous. Her physician discovers that she has marked neck stiffness and photophobia. The physician performs a transfemoral angiogram. This radiologic study is expected to reveal that the woman has which of the following?

a. An arteriovenous malformation
b. An occipital astrocytoma
c. A sphenoidal meningioma
d. A pituitary adenoma
e. A saccular aneurysm

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d. Todd's paralysis
e. Hyponatremia

Three days after a subarachnoid hemorrhage, a patient begins to develop neck stiffness and photophobia. This is followed by left-sided weakness and hyperreflexia. Her left plantar response is upgoing. Her physician presumes that these deficits are a delayed effect of the subarachnoid blood. Which of the following is the most appropriate treatment?

a. Heparin
b. Warfarin
c. Nimodipine
d. Phenytoin
e. Carbamazepine

A 43 year-old man presents with a left CN III deficit and headache. Which of the following is the most likely site of the lesion responsible for this presentation?

a. Anterior communicating artery

b. Posterior communicating artery
c. Anterior cerebral artery
d. Middle cerebral artery
e. Posterior cerebral artery

A 67-year-old woman with a history of type 2 diabetes mellitus and atrial fibrillation presents to the emergency room with left body weakness and slurred speech. The onset was sudden while she was brushing her teeth 1 h ago, and she was brought immediately to the emergency room. She denies word-finding difficulties, dysesthesia, and headache. She is taking warfarin. Physical exam findings include blood pressure of 205/90 and irregularly irregular heartbeat. There is left side neglect with slurred speech. There is a corticospinal pattern of weakness of the left body, with the face and upper extremity being worse than the lower extremity. Routine chemistries and cell counts are normal. Her INR is 1.8. Which of the following is the most appropriate first step in management?

a. Administer tissue plasminogen activator
b. Call a vascular surgery consult for possible endarterectomy
c. Order a brain CT
d. Order a cerebral angiogram
e. Start heparin

A 61-year-old man with a history of hypertension has been in excellent health until he presents with vertigo and unsteadiness lasting for 2 days. He then develops nausea, vomiting, dysphagia, hoarseness, ataxia, left facial pain, and right-sided sensory loss. There is no weakness. On examination, he is alert, with a normal mental status. He vomits with head movement. There is skew deviation of the eyes, left ptosis, clumsiness of the left arm, and titubation. He has loss of pin and temperature sensation on the right arm and leg and decreased joint position sensation in the left foot. He is unable to walk. Magnetic resonance imaging (MRI) in this patient might be expected to show which of the following?

a. Basilar artery tip aneurysm
b. Right lateral medullary infarction
c. Left lateral medullary infarction
d. Left medial medullary infarction
e. Right medial medullary infarction

Graph of logical structure of class

Syndromes of Cerebral Infarction	
Artery Occluded	**Syndrome**
Common carotid	Asymptomatic
Internal carotid	Ipsilateral blindness
	Contralateral hemiparesis and hemianesthesia
	Hemianopia
	Aphasia or anosognosia and hemineglect
Middle cerebral	
Main trunk	Hemiplegia
	Hemianesthesia
	Hemianopia
	Aphasia or anosognosia and hemineglect
Upper division	Hemiparesis and sensory loss (arm and face more affected than leg)
	Broca aphasia or anosognosia and hemineglect
Lower division	Wernicke aphasia or nondominant behavior disorder without hemiparesis
Penetrating artery	Pure motor hemiparesis
Anterior cerebral	Hemiparesis and sensory loss affect leg more than arm

	Impaired responsiveness (“abulia” or “akinetic mutism”), especially if bilateral infarction
	Left-sided ideomotor apraxia or tactile anomia
Posterior cerebral	Cortical, unilateral: isolated hemianopia (or quadrantic field cut); alexia or color anomia
	Cortical, bilateral: cerebral blindness, with or without macular sparing
	Thalamic: pure sensory stroke; may leave anesthesia dolorosa with “spontaneous pain”
	Subthalamic nucleus: hemiballism
	Bilateral inferior temporal lobe: amnesia
	Midbrain: oculomotor palsy and other eye-movement abnormalities

Signs that Indicate the Level of Brain Stem Vascular Syndromes

Syndrome	Artery Affected	Structure Involved	Manifestations
Medial Syndromes			
Medulla	Paramedian branches	Emerging fibers of 12th nerve	Ipsilateral hemiparalysis of tongue
Inferior pons	Paramedian branches	Pontine gaze center, near or in nucleus of 6th nerve	Paralysis of gaze to side of lesion
		Emerging fibers of 6th nerve	Ipsilateral abduction paralysis
Superior pons	Paramedian branches	Medial longitudinal fasciculus	Internuclear ophthalmoplegia
Lateral Syndromes			
Medulla	Posterior inferior cerebellar or vertebral artery branches	Emerging fibers of 9th and 10th nerves	Dysphagia, hoarseness, ipsilateral paralysis of vocal cord; ipsilateral loss of pharyngeal reflex
		Vestibular nuclei	Vertigo, nystagmus
		Descending tract and nucleus of 5th nerve	Ipsilateral facial analgesia
		Solitary nucleus and tract	Taste loss on ipsilateral half of tongue posteriorly
Inferior pons	Anterior inferior cerebellar	Emerging fibers of 7th nerve	Ipsilateral facial paralysis
		Solitary nucleus and tract	Taste loss on ipsilateral half of tongue anteriorly
		Cochlear nuclei	Deafness, tinnitus
Mid-pons		Motor nucleus of 5th nerve	Ipsilateral jaw weakness
		Emerging sensory fibers of 5th nerve	Ipsilateral facial numbness

Modified from Rowland LP. In: Kandel ER, Schwartz JH, eds. *Principles of Neural Science.* New York: Elsevier; 1991.

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources

1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical Class No. 5.

Theme: Infectious diseases of the nervous system.

Actuality of theme: Neuroinfections make the most widespread group of organic nervous diseases. Level of them in general pathology of the nervous system is 35-37%. Consequently all doctors must know this pathology and diagnose it on time.

Particular goals:
To know:
1) Etiology and pathogenesis of primary and secondary meningitis;
2) Patomorphology of infection diseases;
3) Clinical course and clinical forms of:
a) primary meningitis;
b) secondary meningitis.
4) Liquorodiagnosis of meningitis.

Table of contents of the class:

Localization

CNS infection may involve the leptomeninges and CSF spaces (*meningitis*), the ventricular system (*ventriculitis*), the gray and white matter of the brain (*encephalitis*), or the spinal cord (*myelitis*).

A focus of bacterial infection of the brain is called a *brain abscess*, or *cerebritis* in the early stage before a frank abscess is formed. Pus located between the dura mater and the arachnoid membrane is called a *subdural empyema*, while pus outside the dura is called an *epidural abscess*.

Course

The clinical manifestations may be *acute* (purulent meningitis, CNS listeriosis, herpes simplex encephalitis), *subacute* (cerebral abscess, focal encephalitis, neuroborreliosis, neurosyphilis, tuberculous meningitis, actinomycosis, nocardiosis, rickettsiosis, neurobrucellosis), or *chronic* (tuberculous meningitis, neurosyphilis, neuroborreliosis, Whipple encephalitis, Creutzfeldt–Jakob disease). The epidemiological pattern of infection may be *sporadic*, *endemic* or *epidemic*, depending on the pathogen.

Clinical Manifestations

Meningitis and encephalitis rarely occur as entirely distinct syndromes; they usually present in mixed form (meningoencephalitis, encephalomyelitis). CSF examination establishes the diagnosis. These disorders may present in specific ways in certain patient groups. *Neonates and children* commonly manifest failure to thrive, fever or hypothermia, restlessness, breathing disorders, epileptic seizures, and a bulging fontanelle. *The elderly* may lack fever but frequently have behavioral abnormalities, confusion, epileptic seizures, generalized weakness, and impairment of consciousness ranging to coma. *Immunodeficient patients* commonly have fever, headache, stiff neck, and drowsiness in addition to the manifestations of their primary illness.

Meningitic syndrome is characterized by fever, severe, intractable headache and backache, photophobia and phonophobia, nausea, vomiting, impairment of consciousness, stiff neck, and hyperextended posture, with opisthotonus or neck pain on flexion.

Kernig's sign (resistance to passive raising of leg with extended knee) and *Brudzinski's sign* (involuntary leg flexion on passive flexion of the neck) are signs of meningeal involvement. Painful neck stiffness is due to (lepto)meningeal irritation by infectious meningitis, septicemia, subarachnoid hemorrhage, neoplastic meningitis, or other causes.

Isolated neck stiffness not caused by meningitis (meningism) may be due to cervical disorders such as arthrosis, fracture, intervertebral disk herniation, tumor, or extrapyramidal rigidity. Papilledema is usually absent; when present, it indicates intracranial hypertension.

Encephalitic syndrome is characterized by headache and fever, sometimes accompanied by epileptic seizures (often focal), focal signs (cranial nerves deficits, especially of CN III, IV, VI, and VII; aphasia, hemiparesis, hemianopsia, ataxia, choreoathetosis), behavioral changes, and impairment of consciousness (restlessness, irritability, confusion, lethargy, drowsiness, coma). The neurological signs may be preceded by limb pain (myalgia, arthralgia), a slight increase in body temperature, and malaise. For *acute cerebellitis* (ataxia). *Brain stem encephalitis* produces ophthalmoplegia, facial paresis, dysarthria, dysphagia, ataxia, and hearing loss.

Myelitic syndrome. Myelitis presents with severe local pain, paraparesis, paresthesiae, or some combination of these. Incomplete or complete paraplegia or quadriplegia develops within a few hours (acute) or days (subacute). The differential diagnosis may be difficult.

Clinical Features of bacterial meningitis

Adults and Children The early clinical effects of acute bacterial meningitis are fever, severe headache, and stiffness of the neck (resistance to passive movement on forward bending), sometimes with generalized convulsions and a disorder of consciousness (i.e., drowsiness, confusion, stupor, and coma). Flexion at the hip and knee in response to forward flexion of the neck (Brudzinski sign) and inability to completely extend the legs (Kernig sign) have the same significance as stiff neck but are less consistently elicitable. Basically, all of these signs are part of a flexor protective reflex (one of the "nocifensive" responses in Fulton's terms). Stiffness of the neck that is part of paratonic or extrapyramidal rigidity should not be mistaken for that of meningeal irritation. The former is more or less equal in all directions of movement, in distinction to that of meningitis, which is present only or predominantly on forward flexion. Whether it is stiffness in the initial few degrees of flexion of the neck or in the subsequent part of the movement that is more specific for meningitis has been debated; our experience has been that the latter is more sensitive but also proves to be mistaken for other disorders; therefore the first may be more

specific for meningitis. Diagnosis of meningitis may be difficult when the initial manifestations consist only of fever and headache, when stiffness of the neck has not yet developed, or when there is only pain in the neck or abdomen or a febrile confusional state or delirium. Also, stiffness of the neck may not be apparent in the deeply stuporous or comatose patient or in the infant or the elderly, as indicated further on. The symptoms comprised by the meningitic syndrome are common to the three main types of bacterial meningitis, but certain clinical features and the setting in which each of them occurs correlate more closely with one type than another.

Meningococcal meningitis should be suspected when the evolution is extremely rapid (delirium and stupor may supervene in a matter of hours), when the onset is attended by a ***petechial or purpuric rash*** or by large ecchymoses and lividity of the skin of the lower parts of the body, when there is ***circulatory shock***, and especially during local outbreaks of meningitis. Since a petechial rash accompanies approximately 50 percent of meningococcal infections, its presence dictates immediate institution of antibiotic therapy, even though a similar rash may be observed with certain viral (echovirus serotype 9 and some other enteroviruses) as well as *Staph. aureus* infections and rarely with other bacterial meningitides.

Pneumococcal meningitis is often preceded by an infection in the lungs, ears, sinuses, or heart valves. In addition, a pneumococcal etiology should be suspected in alcoholics, in splenectomized patients, in the very elderly, and in those with recurrent bacterial meningitis, dermal sinus tracts, sickle cell anemia ("autosplenectomized"), and basilar skull fracture.

On the other hand, *H. influenzae* meningitis usually follows upper respiratory and ear infections in the child.

Other specific bacterial etiologies are suggested by particular clinical settings. Meningitis in the presence of furunculosis or following a neurosurgical procedure should direct attention to the possibility of a coagulase-positive staphylococcal infection. Ventriculovenous shunts, inserted for the control of hydrocephalus, are particularly prone to infection with coagulase-negative staphylococci. HIV infection, myeloproliferative or lymphoproliferative disorders, defects in cranial bones (tumor, osteomyelitis), collagen diseases, metastatic cancer, and therapy with immunosuppressive agents are clinical conditions that favor invasion by such pathogens as Enterobacteriaceae, *Listeria*, *A. calcoaceticus*, *Pseudomonas*, and occasionally by parasites. Focal cerebral signs in the early stages of the disease, although seldom prominent, are most frequent in pneumococcal and *H. influenzae* meningitides. Some of the transitory focal cerebral signs may represent postictal phenomena (Todd's paralysis); others may be related to an unusually intense focal meningitis-for example, purulent material collected in one sylvian fissure. Seizures are encountered most often with *H. influenzae* meningitis. Although this has happened most often in infants and children, it is difficult to judge the significance, since young children may convulse with fever of any cause. Persistent focal cerebral lesions or intractable seizures usually develop in the second week of the meningeal infection and are caused by an infectious vasculitis, as described earlier-usually with occlusion of surface cerebral veins and consequent infarction of cerebral tissue. Cranial nerve abnormalities are particularly frequent with pneumococcal meningitis, the result of invasion of the nerve by purulent exudate and possibly ischemic damage as the nerve traverses the subarachnoid space.

Infants and Newborns Acute bacterial meningitis during the first month of life is said to be more frequent than in any subsequent 30-day period of life. It poses a number of special problems. Infants, of course, cannot complain of headache, stiff neck may be absent, and one has only the nonspecific signs of a systemic illness-fever, irritability, drowsiness, vomiting, convulsions-and a bulging fontanel to suggest the presence of meningeal infection. Signs of meningeal irritation do occur, but only late in the course of the illness. A high index of suspicion and liberal use of the lumbar puncture needle are the keys to early diagnosis. Lumbar puncture is ideally performed before any antibiotics are administered for other neonatal infections. An

antibiotic regimen sufficient to control a septicemia may allow a meningeal infection to smolder and to flare up after antibiotic therapy for the systemic infection has been discontinued.

A number of other facts about the natural history of neonatal meningitis are noteworthy. It is more common in males than in females, in a ratio of about 3:1. Obstetric abnormalities in the third trimester (premature birth, prolonged labor, premature rupture of fetal membranes) occur frequently in mothers of infants who develop meningitis in the first weeks of life. The most significant factor in the pathogenesis of the meningitis is maternal infection (usually a urinary tract infection or puerperal fever of unknown cause). The infection in both mother and infant is most often due to gram-negative enterobacteria, particularly *E. coli*, and group B streptococci and less often to *Pseudomonas*, *Listeria*, *Staph. Aureus* or *epidermidis* (formerly *albus*), and group A streptococci. Analysis of postmortem material indicates that in most cases infection occurs at or near the time of birth, although clinical signs of meningitis may not become evident until several days or a week later. In infants with meningitis, one should be prepared to find a unilateral or bilateral sympathetic *subdural effusion* regardless of bacterial type. Young age, rapid evolution of the illness, low polymorphonuclear cell count, and markedly elevated protein in the CSF correlate to some extent with the formation of effusions, according to Snedeker and coworkers. Also, these attributes greatly increase the likelihood of the meningitis being associated with neurologic signs. Transillumination of the skull is the simplest method of demonstrating the presence of an effusion, but computed tomography (CT) and magnetic resonance imaging (MRI) are the definitive diagnostic tests. When aspirated, most of the effusions prove to be sterile. If recovery is delayed and neurologic signs persist, a succession of aspirations is required. In our experience and that of others, patients in whom meningitis is complicated by subdural effusions are no more likely to have residual neurologic signs and seizures than are those without effusions.

Treatment

Bacterial meningitis is a medical emergency. The first therapeutic measures are directed to sustaining blood pressure and treating septic shock (volume replacement, pressor therapy). A premium is then placed on choosing an antibiotic that is known both to be bactericidal for the suspected organism and is able to enter the CSF in effective amounts. Treatment should begin while awaiting the results of diagnostic tests and may be altered later in accordance with the laboratory findings.

THE SYNDROME OF ACUTE ASEPTIC MENINGITIS

The term *aseptic meningitis* was first introduced to designate what was thought to be a specific disease-“aseptic” because bacterial cultures were negative. The term is now applied to a symptom complex that is produced by any one of numerous infective agents, the majority of which are viral (but a few of which are bacterial-mycoplasma, Q fever, other rickettsial infections, etc.). Since aseptic meningitis is rarely fatal, the precise pathologic changes are uncertain but are presumably limited to the meninges. Conceivably, there may be some minor changes in the brain itself, but these are of insufficient severity to cause neurologic symptoms and signs or to alter the results of computed tomography (CT) or magnetic resonance imaging (MRI). In outline, the clinical syndrome of aseptic meningitis consists of fever, headache, signs of meningeal irritation, and a predominantly lymphocytic pleocytosis with normal cerebrospinal fluid (CSF)glucose. Usually the onset is acute and the temperature is elevated, from 38 to 40_C (100.4 to 104_F). Headache, perhaps more severe than that associated with other febrile states, is the most frequent symptom. A variable degree of lethargy, irritability, and drowsiness may occur; confusion, stupor, and coma mark the case as an encephalitis rather than a meningitis. Photophobia and pain on movement of the eyes are common additional complaints. Stiffness of the neck and spine on forward bending attests to the presence of meningeal irritation (meningismus), but at first it may be so slight as to pass unnoticed. Here the Kernig and Brudzinski signs help very little, for they are often absent in the presence of manifest viral meningitis. When there are accompanying neurologic signs, they too tend to be mild or fleeting: paresthesias in an extremity, isolated strabismus and diplopia, a slight inequality of reflexes, or wavering Babinski signs. Other

symptoms and signs are infrequent and depend mainly on the systemic effects of the invading virus; these include sore throat, nausea and vomiting, vague weakness, pain in the back and neck, conjunctivitis, cough, diarrhea, vomiting, rash, adenopathy, etc. The childhood exanthems associated with meningitis and encephalitis (varicella, rubella, mumps) produce well-known eruptions and other characteristic signs. An erythematous papulomacular, nonpruritic rash, confined to the head and neck or generalized, may also be a prominent feature (particularly in children)of certain echoviruses and Coxsackie viruses. An enanthem (herpangina), taking the form of a vesiculoulcerative eruption of the buccal mucosa, may also occur with these viral infections.

The CSF findings consist of a pleocytosis (mainly mononuclear except in the first days of the illness, when more than half the cells may be neutrophils), and a small and variable increase in protein. In milder cases, in the first hours or day of the illness, there may be no abnormalities of the spinal fluid, and the patient may erroneously be thought to have migraine or a headache induced by a systemic infectious illness. Micro-organisms cannot be demonstrated by conventional smear or bacterial culture. As a rule, the glucose content of the CSF is normal; this is important because a low glucose concentration in conjunction with a lymphocytic or mononuclear pleocytosis usually signifies tuberculous or fungal meningitis or certain noninfectious disorders such as carcinomatous or lymphomatous invasion, or sarcoid of the meninges. Infrequently, a mild depression of the CSF glucose (never below 25 mg/dL) has been reported with the meningitis caused by mumps, HSV-2, lymphocytic choriomeningitis, or VZV.

THE SYNDROME OF ACUTE ENCEPHALITIS

From the foregoing discussion it is evident that the separation of the clinical syndromes of aseptic meningitis and encephalitis is not always easy. In some patients with aseptic meningitis, mild drowsiness or confusion may be present, suggesting cerebral involvement. Conversely, in some patients with encephalitis, the cerebral symptoms may be mild or inapparent, and meningeal symptoms and CSF abnormalities predominate. These facts make it difficult to place complete reliance on statistical data from various virus laboratories about the relative incidence of meningitis and encephalitis. The common practice is to assume that viral meningitis causes only fever, headache, stiff neck, and photophobia;if any other CNS symptoms are added, the condition is generally called meningoencephalitis. As has been emphasized, it appears that the same spectrum of viruses gives rise to both meningitis and encephalitis. It is our impression that many cases of enteroviral and practically all cases of mumps and LCM encephalopathy are little more than examples of intense meningitis. Rarely have they caused death with postmortem demonstration of cerebral lesions, and surviving patients seldom have residual neurologic signs. Conversely, several agents, notably the arboviruses, may cause encephalitic lesions with only mild meningeal symptoms.

The core of the *encephalitis syndrome* consists of an acute febrile illness with evidence of meningeal involvement (sometimes only headache), added to which are various combinations of the following symptoms and signs: convulsions, delirium, confusion, stupor, or coma;aphasia;hemiparesis with asymmetry of tendon reflexes and Babinski signs;involuntary movements, ataxia, and myoclonic jerks;nystagmus, ocular palsies, and facial weakness. The spinal fluid invariably shows a cellular reaction and the protein is slightly elevated. Imaging studies of the brain are most often normal but may show diffuse edema or enhancement of the cortex and, in certain infections, subcortical and deep nuclear involvement as well as, in the special case of HSV encephalitis, selective damage of the inferomedial temporal and frontal lobes. One or another of these findings predominates in certain types of encephalitis, but the clinical diagnosis of encephalitis in the setting of a febrile aseptic meningitis always rests on the demonstration of derangement of the function of the cerebrum, brainstem, or cerebellum.

Differentiation of Viral from Postinfectious Encephalitis The acute encephalitis syndrome described above may take two forms: the more common direct invasion of brain and meninges (true viral encephalitis) and a postinfectious encephalomyelitis that is based on an auto-immune

reaction to the systemic viral infection but in which virus is not present in neural tissue. The distinction between postinfectious encephalomyelitis and infectious encephalitis may be difficult, especially in younger patients who have a proclivity to develop the postinfectious variety. The latter, termed acute disseminated encephalomyelitis (ADEM), occurs after a latency of several days, as the infectious illness is subsiding. It is expressed by a low-grade fever and cerebral symptoms such as confusion, seizures, coma, ataxia, etc. The spinal fluid shows slight inflammation and elevation of protein-sometimes a more intense reaction, and there are usually characteristic confluent bilateral lesions in the white matter in imaging studies, findings that differ from those of viral encephalitis. When there is no coexistent epidemic of encephalitis to suggest the diagnosis or the systemic illness is absent or obscure, a differentiation between the two may not be possible on clinical grounds alone. The fever is generally higher in the infectious type but even this difference does not always hold in young children with ADEM.

Localization

CNS infection may involve the leptomeninges and CSF spaces (*meningitis*), the ventricular system (*ventriculitis*), the gray and white matter of the brain (*encephalitis*), or the spinal cord (*myelitis*).

A focus of bacterial infection of the brain is called a *brain abscess*, or *cerebritis* in the early stage before a frank abscess is formed. Pus located between the dura mater and the arachnoid membrane is called a *subdural empyema*, while pus outside the dura is called an *epidural abscess*.

Course

The clinical manifestations may be *acute* (purulent meningitis, CNS listeriosis, herpes simplex encephalitis), *subacute* (cerebral abscess, focal encephalitis, neuroborreliosis, neurosyphilis, tuberculous meningitis, actinomycosis, nocardiosis, rickettsiosis, neurobrucellosis), or *chronic* (tuberculous meningitis, neurosyphilis, neuroborreliosis, Whipple encephalitis, Creutzfeldt–Jakob disease). The epidemiological pattern of infection may be *sporadic*, *endemic* or *epidemic*, depending on the pathogen.

Clinical Manifestations

Meningitis and encephalitis rarely occur as entirely distinct syndromes; they usually present in mixed form (meningoencephalitis, encephalomyelitis). CSF examination establishes the diagnosis. These disorders may present in specific ways in certain patient groups. *Neonates and children* commonly manifest failure to thrive, fever or hypothermia, restlessness, breathing disorders, epileptic seizures, and a bulging fontanelle. *The elderly* may lack fever but frequently have behavioral abnormalities, confusion, epileptic seizures, generalized weakness, and impairment of consciousness ranging to coma. *Immunodeficient patients* commonly have fever, headache, stiff neck, and drowsiness in addition to the manifestations of their primary illness.

Encephalitic syndrome is characterized by headache and fever, sometimes accompanied by epileptic seizures (often focal), focal signs (cranial nerves deficits, especially of CN III, IV, VI, and VII; aphasia, hemiparesis, hemianopsia, ataxia, choreoathetosis), behavioral changes, and impairment of consciousness (restlessness, irritability, confusion, lethargy, drowsiness, coma). The neurological signs may be preceded by limb pain (myalgia, arthralgia), a slight increase in body temperature, and malaise. For *acute cerebellitis* (ataxia). *Brain stem encephalitis* produces ophthalmoplegia, facial paresis, dysarthria, dysphagia, ataxia, and hearing loss.

Myelitic syndrome. Myelitis presents with severe local pain, paraparesis, paresthesiae, or some combination of these. Incomplete or complete paraplegia or quadriplegia develops within a few hours (acute) or days (subacute). The differential diagnosis may be difficult.

Pathogenesis. Pathogens usually reach the CNS by local extension froma nearby infectious focus (e. g. sinusitis, mastoiditis) or by hematogenous spread from a distant focus. The ability of pathogens to spread by way of the bloodstream depends on their virulence and on the immune status of the host. They use special mechanisms to cross or circumvent the *blood–brain barrier*. Some pathogens enter the CNS by centripetal travel along peripheral nerves (herpes simplex virus type I, varicella-zoster virus, rabies virus), others by endocytosis (*Neisseria meningitidis*),

intracellular transport (*Plasmodium falciparum* via erythrocytes, *Toxoplasma gondii* via macrophages), or intracellular invasion (*Haemophilus influenzae*). Those that enter the *subarachnoid space* probably do so by way of the choroid plexus, venous sinuses, or cribriform plate. Having entered the CSF spaces, pathogens trigger an *inflammatory response* characterized by the release of complement factors and cytokines, the influx of leukocytes and macrophages, and the activation of microglia and astrocytes. Disruption of the blood–brain barrier results in an influx of fluids and proteins across the vascular endothelium and into the CNS, causing vasogenic cerebral edema (p. 162), which is accompanied by both cytotoxic cellular edema and interstitial edema due to impaired CSF circulation. Cerebral edema causes *intracranial hypertension*. These processes, in conjunctionwith vasculitis, impairment of vascular autoregulatory mechanisms, and/or fluctuations of systemic blood pressure, lead to the development of ischemic, metabolic, and hypoxic cerebral lesions (focal necrosis, territorial infarction). The immune system is generally no longer able to hold pathogens in check once they have spread to the CNS, as the immune response in the subarachnoid space and the neural tissue itself is less effective than elsewhere in the body. Having gained access to the CNS, pathogens meet with favorable conditions for further spread within it.

Prophylaxis. The occurrence and spread of CNS infection can be prevented by *mandatory reporting* (as specified by local law), *prevention of exposure* (isolation of sources of infection, disinfection, sterilization), and *prophylaxis in persons at risk* (active and passive immunization, chemoprophylaxis).

Treatment. Patients with bacterial or viral meningoencephalitis must be treated at once. The treatment strategy is initially based on the clinical and additional findings. Antimicrobial therapy is first given empirically in a broadspectrum combination, then specifically tailored in accordance with the species and drug sensitivity pattern of the pathogen(s) identified. Causative organisms may be found in the CSF, blood, or other bodily fluids (e. g., throat smear, urine or stool samples, bronchial secretions, gastric juice, abscess aspirate).

Encephalitis Lethargica (Von Economo Disease, Sleeping Sickness) Although examples of a somnolent-ophthalmoplegic encephalitis dot the early medical literature (e.g., nona, fe´bre lethargica, Schlafkrankheit), it was in the wake of the influenza pandemic of World War I that this disease appeared prominently and continued to reappear for about 10 years. The viral agent was never identified, but the clinical and pathologic features were typical of viral infection.

The importance of encephalitis lethargica relates to its unique clinical syndromes and sequelae and to its place as the first recognized “slow virus infection” of the nervous system in human beings. The unique symptoms were ophthalmoplegia and pronounced somnolence, from which the disease took its name. Some patients were overly active, and a third group manifested a disorder of movement in the form of bradykinesia, catalepsy, mutism, chorea, or myoclonus. Lymphocytic pleocytosis was found in the spinal fluid of half the patients, together with variable elevation of the CSF protein content. More than 20 percent of the victims died within a few weeks, and many survivors were left with varying degrees of impairment of mental function. However, the most extraordinary feature was the appearance of a Parkinsonian syndrome, after an interval of weeks or months (occasionally years), in a high proportion of survivors. This is the only form of encephalitis known to cause a delayed extrapyramidal syndrome of this type (a similar though not identical syndrome with a much shorter latency may follow Japanese B encephalitis and occasionally other arboviral encephalitis, as occurred in one of our cases after Eastern equine encephalitis). Myoclonus, dystonia, oculogyric crises and other muscle spasms, bulimia, obesity, reversal of the sleep pattern, and, in children, a change in personality with compulsive behavior were other distressing sequelae. The pathology was typical of a viral infection, localized principally to the midbrain, subthalamus, and hypothalamus. In the patients who died years later with a Parkinson syndrome, the main findings were depigmentation of the substantia nigra and locus ceruleus due to nerve cell destruction. Neurofibrillary changes in the surviving nerve cells of the substantia nigra and the oculomotor and adjacent nuclei were also described,

indistinguishable from those of progressive supranuclear palsy. Lewy bodies were not seen, in contrast to idiopathic Parkinson disease (paralysis agitans), where they are consistently present. Only a few new questionable cases of postencephalitic type have been seen in the United States and western Europe since 1930. Sporadic cases, such as the four reported by Howard and Lees, may be examples of this disease, but there is no way of proving their identity.

Rabies

Pathogenesis. Rabies virus is a rhabdovirus that is mainly transmitted by the bite of a rabid animal. The reservoirs of infection are wild animals in Europe and America (foxes, wild boar, deer, martens, raccoons, badgers, bats; *sylvatic rabies*) and dogs in Asia (*urban rabies*). The virus replicates in muscles cells near the site of entry and then spreads via muscle spindles and motor end plates to the peripheral nerves, as far as the spinal ganglia and spinal motor neurons, where secondary replication takes place. It subsequently spreads to the CNS and other organs (salivary glands, cornea, kidneys, lungs) by way of the fiber pathways of the autonomic nervous system. The limbic system is usually also involved. The mean incubation time is 2–3 months (range: 1 week to 1 year). Proof that the biting animal was rabid is essential for diagnosis, as rabies is otherwise very difficult to diagnose until its late clinical manifestations appear. The virus can be isolated from the patient's sputum, urine or CSF in the first week after infection.

Symptoms and signs. The course of rabies can be divided into three stages. The *prodromal stage* (2–4 days) is characterized by paresthesia, hyperesthesia, and pain at the site of the bite and the entire ipsilateral side of the body. The patient suffers from nausea, malaise, fever, and headache and, within a few days, also from anxiety, irritability, insomnia, motor hyperactivity, and depression.

Hyperexcitability stage. In the ensuing days, the patient typically develops increasing restlessness, incoherent speech, and painful spasms of the limbs and muscles of deglutition, reflecting involvement of the midbrain tegmentum. *Hydrophobia*, as this stage of the disease is called, is characterized by painful laryngospasms, respiratory muscle spasms, and opisthotonus, with tonic-clonic spasms throughout the body that are initially triggered by attempts to drink but later even by the mere sight of water, unexpected noises, breezes, or bright light. There may be alternating periods of extreme agitation (screaming, spitting, and/or scratching fits) and relative calm. The patient dies within a few days if untreated, or else progresses to the next stage after a brief clinical improvement.

Paralytic stage (*paralytic rabies*). The patient's mood and hydrophobic manifestations improve, but spinal involvement produces an ascending flaccid paralysis with myalgia and fasciculations.Weakness may appear in all limbs at once, or else in an initially asymmetrical pattern, beginning in the bitten limb and then spreading. In some cases, the clinical picture is dominatedby cranial nerve palsies (oculomotor disturbances, dysphagia, drooling, dysarthrophonia) and autonomic dysfunction (cardiac arrhythmia, pulmonary edema, diabetes insipidus, hyperhidrosis). **Rabies prophylaxis.** *Preexposure prophylaxis:*

Vaccination of persons at risk (veterinarians, laboratory personnel, travelers to endemic areas).

Local wound treatment: Thorough washing of the bite wound with soap and water.

Postexposure prophylaxis: Vaccination and rabies immunoglobulin.

Herpes Simplex Virus Infection

Pathogenesis. *Herpes simplex virus type 1* (HSV-1) is usually transmitted in childhood through lesions of the oral mucosa (gingival stomatitis, pharyngitis). The virus travels centripetally by way of nerve processes toward the sensory ganglia (e. g., the trigeminal ganglion), where it remains dormant for a variable period of time until reactivated by a trigger such as ultraviolet radiation, dental procedures, immunosuppression, or a febrile illness. It then travels centrifugally, again over nerve processes, back to the periphery, producing blisterlike vesicles (herpes labialis). HSV-1 also causes eye infection (keratoconjunctivitis), as well as (meningo)encephalitis when it

spreads via CN I and leptomeningeal fibers of CN V. There is no association between herpes labialis and HSV-1 encephalitis. *Herpes simplex virus type 2* (HSV-2) reaches the lumbosacral ganglia by axonal transport from a site of (asymptomatic) urogenital infection. Its reactivation causes genital herpes. In adults, HSV-2 infection can cause (aseptic) *meningitis* and, occasionally, *polyradiculitis* or *myelitis*. HSV-2 virus can be transmitted to the newborn during the birth process, causing encephalitis. HSV-1 encephalitis is very rare in neonates, and HSV-2 encephalitis is very rare in adults.

Symptoms and signs. *Herpes simplex encephalitis* (HSE) in adults begins with local inflammation of the caudal and medial parts of the frontal and temporal lobes. Uncharacteristic prodromal signs such as fever, headache, nausea, anorexia, and lethargy last a few days at the most. Focal symptoms including olfactory and gustatory hallucinations, aphasia, and behavioral disturbances (confusion, psychosis) then appear, along with focal or complex partial seizures with secondary generalization. There is usually repeated seizure activity, but status epilepticus is rare. Intracranial hypertension causes impairment of consciousness or coma within a few hours. In neonates, the inflammation spreads throughout the CNS. The diagnosis of HSE can be difficult, especially at first. The *clinical findings* include neck stiffness, hemiparesis, and mental disturbances. *CSF examination* reveals a lymphomonocytic pleocytosis (granulocytes may predominate initially) with an elevated protein concentration; low glucose and high lactate concentrations are only rarely found. Xanthochromia and erythrocytes may be present (hemorrhagic necrotizing encephalitis). In the first 3 weeks, the virus can almost always be detected in the CSF by polymerase chain reaction; brain biopsy is only rarely needed for identification of the viral pathogen. Lumbar puncture carries a risk if intracranial hypertension is present (p. 162). EEG reveals periodic high-voltage sharp waves and 2–3 Hz slow wave complexes as a focal or diffuse finding in one or both temporal lobes. In the acute stage of HSE, CT is normal or reveals only mild temporobasal hypointensity without contrast enhancement. Hemorrhage may appear as a hyperdense area. Sharply defined areas of hypodensity appear on CT only in the later stages of HSE. T2-weighted MRI, however, already reveals inflammatory lesions in early HSE. Thus, MRI is used for early diagnosis, CT for the monitoring of encephalitic foci and cerebral edema over the course of the disease. *Meningitis.* The clinical manifestations are those of aseptic meningitis. *Myelitis.* Low back pain, fever, sensory deficit with spinal level, flaccid or spastic paraparesis, bladder and bowel dysfunction. These manifestations usually regress within 2 weeks. *Radiculitis*. Inflammation of the lumbosacral nerve roots produces a sensory deficit and bladder and bowel dysfunction.

Virustatic agents. HSV infection of the CNS is treated with acyclovir 10 mg/kg (i. v.) q8h for 14–21 days. Particularly in HSE, it is important to begin treatment as soon as possible.

Lyme Disease (Neuroborreliosis)

Pathogenesis. The spirochete *Borrelia burgdorferi* sensu lato (Europe: *B. garinii*, *B. afzelii*; North America: *B. burgdorferi* sensu stricto) is transmitted to man by ticks (Europe: *Ixodes ricinus*; North America: *Ixodes pacificus*, *I. scapularis*). The probability of infection is low unless the infected tick remains attached to the skin for at least 24–48 hours. Only 1–2% of individuals bitten by ticks become infected. The incubationtime ranges from 3–30 days. The disease occurs in three stages, as described below.

Clinical manifestations.

Stage I (localized infection). Up to 90% of all patients develop a painless, erythematous macule or papule that gradually spreads outward from the site of the tick bite in a ringlike or homogeneous fashion (*erythema chronicum migrans*). This is commonly accompanied by symptoms due to hematogenous spread of the pathogen, such as fever, fatigue, arthralgia, myalgia, or other types of pain, which may be the chief complaint, rather than the skin rash. Regional or generalized lymphadenopathy (*lymphadenosis benigna cutis*) is a less common presentation. All of these findings may resolve spontaneously.

Stage II (disseminated infection). Generalized symptoms such as fatigue, anorexia, muscle and joint pain, and headache develop in 10–15% of patients within ca. 3–6 weeks, sometimes accompanied by mild fever and neck stiffness.
Cardiac manifestations: Myocarditis or pericarditis with AV block.
Neurological manifestations: Cranial nerve palsies, painful polyradiculitis and lymphocytic meningitis (*Bannwarth syndrome*, meningopolyneuritis) are commonly seen in combination. One or more cranial nervesmay be affected; the most common finding is unilateral or bilateral facial palsy of peripheral type. Neuroborreliosis-related *polyradiculoneuropathy* (which may bemistaken for lumbar disk herniation) is characterized by intense pain in a radicular distribution, most severe at night, with accompanying neurological deficits (motor, sensory, and reflex abnormalities, focal muscle atrophy). Borrelia-related *meningitis* (Lyme meningitis) usually causes alternating headache and neck pain, but the headache is mild or absent in some cases. It may be worst at certain times of day. CSF studies reveal a mononuclear pleocytosis with a high plasma cell count and an elevated protein concentration, while the glucose concentration is normal. *Encephalitis* occurs relatively rarely and may cause focal neurological deficits as well as behavioral changes (impaired concentration, personality changes, depression). MRI reveals cerebral white-matter lesions, and the CSF findings are consistent with meningitis. *Myelitis*, when it occurs, often affects the spinal cord at the level of a radicular lesion.
Stage III (persistent infection). The latency from clinical presentation to the onset of stage III disease varies from 1 to 17 years (chronic Lyme neuroborreliosis). Few patients ever reach this stage, characterized by neurological deficits such as ataxia, cranial nerve palsies, paraparesis or quadriparesis, and bladder dysfunction (*Lyme encephalomyelitis*). *Encephalopathy* causing impairment of concentration and memory, insomnia, fatigue, personality changes, and depression has also been described. *Myositis* and *cerebral vasculitis* may also occur. In stage III of Lyme disease, *acrodermatitis chronica atrophicans* of the extensor surface of the limbs may be seen along with a type of polyneuropathy specific to *Borrelia afzelii*.
Diagnosis. Many patients have no memory of a tick bite. The diagnosis of Lyme disease is based on the presence of erythema chronicum migrans, the immunological confirmation of *Borrelia* infection (e. g., by ELISA, indirect immunofluorescence assay, Western blot, or specific IgG antibody–CSF-serum index) and/or the identification of the causative organism (e. g., by culture, histology, or polymerase chain reaction). By definition, the diagnosis also requires the presence of lymphocytic meningitis (with or without cranial nerve involvement or painful polyradiculoneuritis), encephalomyelitis, or encephalopathy.
Treatment. *Local symptoms:* Antibiotic such as doxycycline or amoxicillin (p.o.) for 3 weeks.
Neuroborreliosis: Ceftriaxone or cefotaxime (i. v.) for 2–3 weeks. A vaccine has been approved for use in the United States, and another is being developed for use in Europe.

Other Forms of Subacute Encephalitis A number of uncommon conditions not covered above are characterized by regional inflammation in the cerebrum. Among these, *Rasmussen encephalitis*, which causes intractable focal seizures and progressive hemiparesis, has been connected to infections by CMV and HSV-I in various studies that used PCR techniques. However, a specific immune reaction consisting of antibodies to glutamate receptors has been implicated more consistently and immunosupressive treatments may be effective. It is not clear whether this process can be classed with the infectious encephalitides. Similarly, the restricted inflammatory conditions called *limbic encephalitis* and "brainstem encephalitis"-most often a distant effect of lung cancer-have some characteristics of a subacute viral encephalitis, but no agent has been consistently isolated and they are also best considered immunologic reactions.

Poliomyelitis is a word derived from the Greek polio (gray) and myelin (marrow), indicating the spinal cord. Spinal cord infection with poliomyelitis virus leads to the classic paralysis secondary to destruction of the anterior horn cells. The incidence of polio peaked in the United States in 1952 with more than 21,000 cases but rapidly decreased after introduction of effective killed parenteral Salk vaccines in 1954 and the live Sabin vaccine a few years later. The

last case of wild-virus polio acquired in the United States was in 1979, and the Global Polio Eradication Program dramatically reduced transmission elsewhere. Polio is eradicated from most of the world but still circulates in many developing countries, particularly in Africa and the Indian subcontinent, and to a lesser degree in Indonesia, a few remote parts of Russia, as well as China and the Arabian Peninsula. Travelers to areas where naturally occurring poliovirus still circulates need to be vaccinated as follows: persons who completed an adequate primary series during childhood should have a one-time booster dose of inactivated poliovirus vaccine (IPV); those who have not received a primary series should receive it although even a single dose prior to travel is of benefit. Humans are the only known reservoir. Transmission occurs most frequently with an unapparent infection. An asymptomatic carrier state occurs only in those with immunodeficiency. Person-to-person spread occurs predominantly via the fecal– oral route. Infection typically peaks in summer in temperate particularly refused immunizations, reported headache, fever, nausea, and general malaise 1 week after camping. Two days later, he felt better, but 48 hours after that, the general symptoms returned, with more headache, generalized muscle aching and pain, and some drowsiness. When weakness supervened a week after illness onset, he was brought to the emergency department. The patient's temperature was 39.5° C (103.1° F), his pulse was 100 beats/min, and his blood pressure 130/70 mm Hg. He had a stiff neck, generalized muscle tenderness, asymmetric weakness (right arm and left leg more than elsewhere), preserved though hypoactive muscle stretch reflexes, flexor plantar responses, normal sensation, and intact cranial nerves. His cough was weak, with a vital capacity of barely 1 L. His WBC count was 15,000/mm3 (40% lymphocytes). Lumbar puncture revealed somewhat cloudy fluid under increased pressure (220 mm Hg), 170/mm3 nucleated cells (60% poly- morphonuclear leukocytes), 150 mg/dL protein, and 80 mg/ dL glucose. Spinal MRI showed enhancement of the cord interiorly, especially the right cervical region. Comment: this is a classic case of infantile poliomyelitis as one would have experienced prior to the widespread utilization of oral and parenteral polio vaccines. In this instance, the patient was at high risk of developing polio either from exposure to a baby recently immunized with live vaccines or the more remote setting here wherein this young man was inadvertently exposed to wild-type polio virus.

Poliovirus is highly infectious and may be present in stool up to 6 weeks; serocon- version in susceptible household contacts of children is nearly 100%, and that of adults is greater than 90%. Persons are most infectious from 7 to 10 days before and after symptom onset. IPV, an inactive, killed vaccine, was licensed in 1955 and used until the early 1960s, when trivalent oral poliovirus vaccine (OPV), containing attenuated strains of all three serotypes of poliovirus in 10 : 1 : 3 ratios, largely replaced it. Enhanced potency trivalent poliovirus vaccine (IPV) was introduced in 1988. The viruses are grown in monkey kidney (Vero) cells and are inactivated with formaldehyde. An occasional live vaccine– associated case of paralytic polio continued to occur in infants after their first immunization at approximately 3 months of age until the CDC mandated in the late 1990s that initial vaccinations must be with the Salk IPV. Since then, no such incidents have been reported. Between 1980 and 1999, a total of 152 confirmed cases of paralytic polio occurred in the United States. Of these, 145 (95%) were vaccine-associated. For this reason, in 2000, the recommendation was made to use IPV exclusively in the United States. Vaccine-associated paralytic polio is thought to occur from a reversion or mutation of the vaccine virus to a more neurotropic form. Live attenuated polioviruses replicate in the intestinal mucosa and lymphoid cells and draining lymph nodes. Vaccine viruses are excreted in stool for up to 6 weeks, with maximal shedding in the first 1–2 weeks after vaccination. IPV is highly effective in producing immunity (99% after three doses) and protection from paralytic poliomyelitis. IPV seems to produce less local gastrointestinal immunity than OPV. Thus, persons immunized with IPV could still become infected with wild-type poliovirus and shed it on return to the United States, with subsequent potential spread. Although most individuals in economically privileged countries are immunized, occasionally, an instance such as described in the vignette in this chapter is seen. Asymmetric weakness distribution and CSF findings help to differentiate it from Guillain–Barré syndrome.

Pathogenesis. Poliovirus is a member of the family Picornaviridae, enterovirus subgroup. Enteroviruses are transient inhabitants of the gastro- intestinal tract and are stable at acid pH. Picornaviruses have an RNA genome; the three poliovirus serotypes (P1, P2, and P3) have minimal heterotypic immunity among them. The virus enters through the mouth and multiplies primarily at the implantation site in the pharynx and gastrointestinal tract; usually, it is present in the throat and stool before clinical onset. Within 1 week of clinical onset, little virus exists in the throat, but it continues to be excreted in the stool for several weeks. The virus invades local lymphoid tissue, enters the bloodstream, and then may infect CNS cells. Viral replication in anterior horn and brainstem motor neuron cells results in cell destruction and paralysis.

Clinical presentation. The incubation period for poliomyelitis is usually 6–20 days, with a range of 3–35 days. Clinical response to poliovirus infection varies. Up to 95% of all polio infections are asymptomatic even though infected persons shed virus in stool and are contagious. Abortive poliomyelitis occurs in 4–8% of infections. It causes a minor illness, without evidence of CNS infection. Complete recovery characteristically occurs within 1 week. Upper respiratory infection (sore throat and fever), gastrointestinal disturbances (nausea, vomiting, abdominal pain, constipation, or rarely diarrhea), and influenza like illness can all occur and are indistinguishable from other enteric viral illnesses. Nonparalytic aseptic meningitis, usually occurring several days after a prodrome similar to the minor illness, occurs in a small percentage of infections. Increased or abnormal sensations may occur with stiffness in the neck, back, leg, or a combination of those areas, typically last 2–10 days, and are then followed by complete recovery. Flaccid paralysis occurs in less than 1% of polio infections. Paralytic symptoms typically begin 1–10 days after the prodromal symptoms and evolve for 2–3 days. Paralysis does not usually progress after defervescence. In children, the prodrome may be biphasic, with initial minor symptoms separated by 1–7 days from major symptoms. Initially, severe muscle aches and spasms are typically seen with significant meningismus and a Kernig sign. The illness evolves into asymmetric flaccid paralysis with diminished muscle stretch reflexes, typically reaching a plateau within days or weeks. Some strength gradually returns. No sensory or cognitive loss occurs. Most patients recover some function, and many recover completely; however, weakness or paralysis that is still discernible 12 months after onset is usually permanent. Three types of paralytic polio are described. Most common is spinal polio (approximately 79% of cases in the 1970s), characterized by asymmetric paralysis usually involving the legs (Fig. 49-8). Bulbar polio (2%) causes weakness of muscles innervated by cranial nerves. Bulbospinal polio (19%) is a combination of the two. Mortality in paralytic polio cases is lower in children (2–5%) than in adults (15–30%) and highest (25–75%) with bulbar involvement.

Postpolio syndrome. This clinical picture usually presents 30–40 years after paralytic poliomyelitis early in life, especially during childhood; in these later, and often more advanced years of life, 25–40% of previous acute polio patients note a seeming increased weakness. This, per se, does not constitute recurrence of a dormant infectious process likened to herpes zoster. Rather, it is thought to involve failure of oversized previously reinnervated motor units that developed during the recovery process from the initial paralytic syndrome.

Diagnostic approach. Poliovirus can be isolated from the pharynx or stool; however, paradoxically this is rarely isolated from CSF. Sequencing can distinguish wild-type from vaccine-type virus in acute flaccid paralysis. Neutralizing antibodies are often present early and at high levels. CSF usually shows an increased WBC count (10– 200 cells/mm3, primarily lymphocytes) and a mildly increased protein level (generally 40–50 mg/dL).

Prognosis. At its most severe bulbospinal form, poliomyelitis often can be fatal. Today with very much enhanced intensive care support, the fatality rate more than likely would be significantly lessened if poliomyelitis reoccurred with the same incidence so typical of 50 years ago. Fortunately, this disease is now so rare that it is difficult to begin to predict what the outcome

might be today. When West Nile virus first appeared about 10 years ago, with its similar predilection for the anterior horn cell, those of us who lived through poliomyelitis as children and adolescents paused to wonder whether this terrible clinical disorder might once again appear in the mask of this virus previously unknown to the western hemisphere. Very fortunately, our fears were not correct.

ARACHNOIDITIS is chronic productive inflammatory process, or reactive local, or more spread change of arachnoidal and pia mater of brain, molecular cortex layer, sometimes in association with inflammation of brain ventricle ependyma and sunependymal layer.

Etiology of cerebral arachnoiditises (CA):

1. Infectious period (grippe) - 49%
2. Infection-allergic (chronical tonsillitis, sinusitis, measles) -26%
3. Traumatic - 17%
4. Uncertain etiology - 12%
5. In specific infections (syphilis, brucellosis, tuberculosis)
6. Otogenic.

CA qualification according to:

I. course:

- acute-7%
- subacute - 10%
- chronic -84%

(Pia mater, arachnoidea, periventricular zone (periventricular encephalitis), walls of cerebrum ventricles and vascular plexuses (chorioependimatitis) are damaged.

II. character; as a result of zone changes in subarachnoidal space, arachnoiditises are divided into:

1. adhesive
2. cystic
3. fixed

III. CA form according to process location:

2. Convexital - a process is localized in frontal, occipital, temporal, parietal cerebrum lobes, and in central gyri.
3. Basal - opticochiasms, interpeduncular zone and transversal cistern are danaged.
4. Posterior cranial fossa - cerebellopontine angel, lateral cerebrum cistern, large cerebrum cistern, cranio-spinal localization.
4. Diffusive

Clinical course:

More often disease starts gradually, subacutely with subfebrile temperature (37, 5 -38 C). Headache, vertigo, nausea, vomiting are gradually increased, consciousness may faint (depending on condition of intracranial hypertension), convulsive and various psychopathological syndromes may developed. Arterial pressure is increased, eye fundus changes are observed (congestion effect), astenization and vegetative dysfunction increase.

Neurological status depends on process location. Arachnoiditisis of the posterior cranial fossa: (PCF) -this is mainly a damage of pia mater of brain in a zone of lateral and large cistern as well as in craniospinal zone with possible disturbance of intraspinal fluid circulation (IFC) in posterior cranial fossa.

General cerebrum effects are marked - occipital headache (with irradiation into the eye fundus) at first persistent, then of paroxysmal character (associated with PCF circulation damages), headache is accompanied with forced head position. Eye fundus changes occur (from a light dilatation retina veins to the congenital effects or visual nerve secondary atrophy. Psychic disturbances are joined. The main focus signs are cerebellum symptoms, HMH lesions of V, VI, VII, VIII pairs and mild pyramidal insufficiency (asymmetry of normal and pathological reflexes). In patients with occlusion forms of arachnoiditisis static and walking severe lesions are observed. Dissociation between significant disturbances of statics and insignificant expression of coordinator damages is characteristics. Occlusion is more often occurred in a big brain cistern (arachnoidal cysts) with following development of hypertension-hydrocéphalie syndrome. It is must be differentiated with brain tumors.

Basal arachnoiditisis. Congestion of optic papilla nerves, decrease of visual acuity, concentric narrowing of visual field, hemianopsia, tendinous hyperreflexia, central paralysis of VII, separate pathological reflexes, retrobulbar neuritis, increased intracranial pressure occur in damage of opticochiasm zone. In damage of interpeduncular cistern headache, total III paralysis and alternating paralyses are observed. In cross cistern arachnoiditisis the main symptom is affection of acoustic nerve with vestibular and acoustic lesions, headache, and insignificant affections of V and VII, mainly on the one side.

Convexital arachnoiditises. Signs of function affection of frontal, occipital, temporal lobes and a zone of central gyri are leading clinical symptoms.

Headache of hypertensive character, increase of a pain is often associated with mental overload and physical overstrains. On percussion cranial bone pain occurs. Epileptic attacks are characteristics. General weakness, vertigo, instability of arterial pressure, signs of vegetative dysfunction. Affections of pyramidal system in form of anisoreflexia, individual pathological reflexes and decrease abdominal reflexes are observed. Transitory visual lesions, supnuclear affections of VII and XII pairs are developed. Dilatation of retina veins is on the eye fundus. In CA the protein blood fractions (index of organism allergisation) are appeared. A/P becomes lower of norm.

Autoantibodies to cerebrum antigens, which emphasize the immunological disease genesis, are determined in blood serum.

During the therapy administration it should be taken into account:

1. Morphological changes in cerebrum membranes.
2. Condition of hemo- and liquorodynamics.

Treatment must be complex. From the first week of a disease antibiotics, sulfa drugs, corticosteroids, desensitizing, spasmodic preparations, Vitamins of B and C group, preparations of calcium and nicotinic acid, cerebrum metabolites are administered. Anticholinesterase and resorption drugs are involved from the second week. Iodine preparation may be added from the third week.

Treatment must be carried no systematically, during some years, till a constant remission.

Neurosyphilis

Pathogenesis. Syphilis is caused by the spirochete (bacterium) Treponema pallidum (TP) ssp. pallidum and is transmitted by direct exposure to infected lesions, usually on the skin or mucous membranes, during sexual contact. Other routes of transmission, such as the sharing of needles by intravenous drug users, are much less common.

The disease has three clinical stages. In the primary and secondary stages, nonspecific tests (VDRL and RPR) and specific tests (TPHA, FTA-ABS, and 19S-(IgM-) TA-ABS tests) yield positive results. Tertiary stage (currently rare): after an asymptomatic period of a few months to years (latent syphilis), organ manifestations develop, such as gummata (skin, bone, kidney, liver) and cardiovascular lesions (aortic aneurysm). The first year of the tertiary stage is designated the

early latency period and is characterized by a high likelihood of recurrence and, thus, recurrent infectivity.

Serologic Diagnosis of Syphilis This depends on the demonstration of one of two types of antibodies-nonspecific or nontreponemal (reagin) antibodies and specific treponemal antibodies. The common tests for reagin are the Kolmer, which uses a complement fixation technique, and the Venereal Disease Research Laboratory (VDRL) slide test, which uses a flocculation technique. These reagin tests, if positive in the CSF, are diagnostic of neurosyphilis. Serum reactivity alone demonstrates exposure to the organism in the past but does not imply the presence of neurosyphilis. However, serum reagin tests are negative in a significant proportion of patients with late syphilis and in those with neurosyphilis in particular (seronegative syphilis). In such patients (and **in patients with suspected false-positive reagin tests**), it is essential to employ tests for antibodies that are directed specifically against treponemal antigens. The latter are positive in the serum of practically every instance of neurosyphilis. The fluorescent treponemal antibody absorption (FTA-ABS) test is the one in common use and is more than adequate for the majority of clinical situations. The T. pallidum immobilization (TPI) test is the most reliable, but it is expensive, difficult to perform, and available in only a few laboratories.

Asymptomatic Neurosyphilis. In this condition, there are no symptoms or physical signs except, in rare cases, abnormal pupils which are light-unreactive but accommodate Argyll-Robertson pupils. The diagnosis is based entirely on the CSF findings, which vary, as mentioned above.

Meningeal Syphilis. Symptoms of meningeal involvement may occur at any time after inoculation but most often do so within the first 2 years. The most common symptoms are headache, stiff neck, cranial nerve palsies, convulsions, and mental confusion. Occasionally headache, papilledema, nausea, and vomiting-due to the presence of increased intracranial pressure-are added to the clinical picture. The patient is afebrile, unlike the case in tuberculous meningitis. The CSF is always abnormal, more so than in asymptomatic syphilitic meningitis. Obviously the meningitis is more intense in the symptomatic type and may be associated with hydrocephalus. With adequate treatment, the prognosis is good. The symptoms usually disappear within days to weeks, but if the CSF remains abnormal, it is likely that some other form of neurosyphilis will subsequently develop if treatment is not continued.

Meningovascular Syphilis This form of neurosyphilis should always be considered when a young person has one or several cerebrovascular accidents, i.e., the sudden development of hemiplegia, aphasia, sensory loss, visual disturbance, or mental confusion. As indicated earlier, this clinical syndrome is now probably the most common form of neurosyphilis. Whereas in the past strokes accounted for only 10 percent of neurosyphilitic syndromes, their frequency is now estimated to be 35 percent. The most common time of occurrence of meningovascular syphilis is 6 to 7 years after the original infection, but it may be as early as 9 months or as late as 10 to 12 years. The CSF almost always shows some abnormality, usually an increase in cells, protein content, and gamma globulin as well as a positive serologic test. However, most patients in middle or late life with stroke and only a positive serologic test will be found at autopsy to have nonsyphilitic atherothrombotic or embolic infarction rather than meningovascular syphilis. The changes in the latter disorder consist not only of meningeal infiltrates but also of inflammation and fibrosis of small arteries (Heubner arteritis), which lead to narrowing and finally occlusion. Most of the infarctions occur in the distal territories of medium- and small-caliber lenticulostriate branches that arise from the stems of the middle and anterior cerebral arteries. Most characteristic perhaps is an internal capsular lesion, extending to the adjacent basal ganglia. The presence of multiple small but not contiguous lesions adjacent to the lateral ventricles is another common pattern. Small, asymptomatic lesions are often seen in the caudate and lenticular nuclei. Several of our patients have had transient prodromal neurologic symptoms. The neurologic signs that remain

after 6 months will usually be permanent, but adequate treatment will prevent further vascular episodes. If repeated cerebrovascular accidents occur despite adequate therapy, one must always consider the possibility of nonsyphilitic vascular disease of the brain.

Paretic Neurosyphilis (General Paresis, Dementia Paralytica, Syphilitic Meningoencephalitis). The general setting of this form of cerebral syphilis is a long-standing meningitis; as remarked above, some 15 to 20 years usually separate the onset of general paresis from the original infection. The history of the disease is entwined with some of the major historical events in neuropsychiatry. Haslam in 1798 and Esquirol at about the same time first delineated the clinical state. Bayle in 1822 commented on the arachnoiditis and meningitis, and Calmeil, on the encephalitic lesion. Nissl and Alzheimer added details to the pathologic descriptions. The syphilitic nature of the disease was suspected by Lasegue and others long before Schaudinn's discovery of the spirochete; it was finally confirmed by Noguchi in 1913. Kraepelin's monograph General Paresis (1913) is one of the classic reviews (see Merritt, Adams, and Solomon for these and other historical references). Once a major cause of insanity, accounting for some 4 to 10 percent of admissions to asylums (hence the term "general paresis of the insane," or GPI), general paresis is now a rarity. Since syphilis is acquired mainly in late adolescence and early adult life, the middle years (35 to 50) are the usual time of onset of the paretic symptoms. Possibly the shortened life span of patients with AIDS has limited the emergence of general paresis, or the immunodeficiency has altered the biologic reaction to the organism. Congenital syphilitic paresis blights early mental development and results in late childhood and adolescent regression in both normal and mentally retarded children. The clinical picture in its fully developed form includes dementia, dysarthria, myoclonic jerks, action tremor, seizures, hyperreflexia, Babinski signs, and Argyll-Robertson pupils. However, more importance attaches to diagnosis at an earlier stage, when few of these manifestations are conspicuous. The insidious onset of memory defect, impairment of reasoning, and reduction in critical faculties-along with minor oddities of deportment and conduct, irritability, and lack of interest in personal appearance-are not too different from the general syndrome of dementia outlined in Chap. 21. One can appreciate how elusive the disease may be at any one point in its early evolution. Indeed, with the currently low index of suspicion of the disease, diagnosis at this preparalytic stage is more often accidental than deliberate. Although classic writings have stressed the development of delusional systems, most dramatically in the direction of megalomania, such symptoms are exceptional in the early or preparalytic phase. More usual has been a simple dementia with weakening of intellectual capacities, forgetfulness, disorders of speaking and writing, and vague concerns about health. In a few patients the first hint of a syphilitic encephalitis, as mentioned earlier, may be facial quivering; tremulousness of the hands; indistinct, hurried speech; myoclonus; and seizures-reminiscent of delirium or acute viral encephalitis. As the deterioration continues into the paralytic stage, intellectual function progressively declines, and aphasias, agnosias, and apraxias intrude themselves. Physical dissolution progresses concomitantly-impaired station and gait, debility, unsteadiness, dysarthria, and tremor of the tongue and hands. All these disabilities lead eventually to a bedridden state; hence the term paretic. Other symptoms are hemiplegia, hemianopia, aphasia, cranial nerve palsies, and seizures with prominent focal signs of unilateral frontal or temporal lobe disease-a syndrome known pathologically as Lissauer's cerebral sclerosis. Normal-pressure hydrocephalus may be the basis of some of the cerebral symptoms. Agitated, delirious, depressive, and schizoid psychoses are special psychiatric syndromes that can be differentiated from general paresis by the lack of mental decline, neurologic signs, and CSF findings.

The blood serology is positive in nearly all cases. The CSF is invariably abnormal, usually with 10 to 200 lymphocytes, plasma cells, and other mononuclear cells per cubic millimeter; a total protein of 40 to 200 mg/dL; an elevated gamma globulin; and strongly positive serologic tests. The elevated gamma globulin in the CSF is produced intrathecally and has been shown to be adsorbed to T. pallidum (Vartdal et al). Hence the gamma globulin (oligoclonal IgG) represents a specific antibody response to this organism. The pathologic changes consist of meningeal

thickening, brain atrophy, ventricular enlargement, and granular ependymitis. Microscopically, the perivascular spaces are filled with lymphocytes, plasma cells, and mononuclear cells; nerve cells have disappeared; there are numerous rod-shaped microgliacytes and plump astrocytes in parts of the cortex devastated by neuronal loss; iron is deposited in mononuclear cells; and, with special stains, spirochetes are visible in the cortex. The changes are most pronounced in the frontal and temporal lobes. The ependymal surfaces of the ventricles are studded with granular elevations protruding between ependymal cells (granular ependymitis). Meningeal fibrosis with obstructive hydrocephalus is present in many cases. The prognosis in early treated cases has in the past been fairly good; 35 to 40 percent of patients made some occupational readjustment; in another 40 to 50 percent, the disease was arrested but left the patient dependent. Without treatment there is progressive mental decline, and death occurs within 3 to 4 years.

Tabetic Neurosyphilis (Tabes Dorsalis) This type of neurosyphilis, described by Duchenne in his classic monograph L'ataxie locomotrice progressive (1858), usually develops 15 to 20 years after the onset of the infection. The major symptoms are lightning pains, ataxia, and urinary incontinence; the chief signs are absent tendon reflexes at knee and ankle, impaired vibratory and position sense in feet and legs, and a Romberg sign. The ataxia is due purely to the sensory defect. Muscular power, by contrast, is fully retained in most cases. The pupils are abnormal in over 90 percent of cases, usually Argyll-Robertson in type (see page 242), and the majority of patients show ptosis or some degree of ophthalmoplegia. Optic atrophy is frequent. The lancinating or lightning pains (present in over 90 percent of cases) are, as their name implies, sharp, stabbing, and brief, like a flash of lightning. They are more frequent in the legs than elsewhere but roam over the body from face to feet, sometimes playing persistently on one spot "like the repeated twanging of a fiddle string," as Wilson remarked. They may come in bouts lasting several hours or days. "Pins and needles" feelings, coldness, numbness, tingling, and other paresthesias are also present and are associated invariably with impairment of tactile, pain, and thermal sensation. The bladder is insensitive and hypotonic, resulting in unpredictable overflow incontinence. Constipation and megacolon as well as impotence are other expressions of dysfunction of the sacral roots and ganglia. In the established phase of the disease, now seldom seen, ataxia is the most prominent feature. The patient totters and staggers while standing and walking. In mild form it is best seen as the patient walks between obstacles or along a straight line, turns suddenly, or halts. To correct the instability, the patient places his feet and legs wide apart, flexes his body slightly, and repeatedly contracts the extensor muscles of his feet as he sways (la danse des tendons). In moving forward, the patient flings his stiffened leg abruptly, and the foot strikes the floor with a resounding thump in a manner quite unlike that seen in the ataxia of cerebellar disease. The patient clatters along in this way with eyes glued to the floor. If his vision is blocked, he is rendered helpless. When the ataxia is severe, walking becomes impossible despite relatively normal strength of the leg muscles. Trophic lesions, perforating ulcers of the feet, and Charcot joints are characteristic complications of the tabetic state. The deformity of deafferented Charcot joints occurs in less than 10 percent of tabetics (the most common cause nowadays is diabetic neuropathy). Most often the hips, knees, and ankles are affected but occasionally also the lumbar spine or upper limbs. The process generally begins as an osteoarthritis, which, with repeated injury to the insensitive joint, progresses to destruction of the articular surfaces. Osseous architecture disintegrates, with fractures, dislocations, and subluxations, only some of which occasion discomfort. The arthropathy has been observed to occur as frequently in the burned-out as in the active phase of tabes; hence it is only indirectly related to the syphilitic process. Although the basic abnormality appears to be repeated injury to an anesthetic joint, the process need not be painless. Presumably a deep and incomplete hypalgesia and loss of autonomic function are enough to interfere with protective mechanisms. Sherrington reproduced the joint change in animals deprived of pain sensation.

Visceral crises represent another interesting manifestation of this disease, now rarely seen. The gastric ones are the best known. The patient is seized abruptly with epigastric pain that spreads around the body or up over the chest. There may be a sense of thoracic constriction as well as

nausea and vomiting-the latter repeated until nothing but blood-tinged mucus and bile are raised. The symptoms may last for several days; a barium swallow sometimes demonstrates pylorospasm. The attack subsides as quickly as it came, leaving the patient exhausted, with a soreness of the epigastric skin. Intestinal crises with colic and diarrhea, pharyngeal and laryngeal crises with gulping movements and dyspneic attacks, rectal crises with painful tenesmus, and genitourinary crises with strangury and dysuria are all less frequent but well-documented types. In most cases now being seen, the CSF is normal when the patient is first examined (so-called burned-out tabes). In the remainder it is abnormal, but less often than in general paresis. Pathologic study reveals a striking thinness and grayness of the posterior roots, principally lumbosacral, and a thinness of the spinal cord due mainly to the degeneration of the posterior columns. Only a slight outfall of neurons is observed in the dorsal root ganglia; the peripheral nerves are therefore essentially normal. For many years there was an argument as to whether the spirochete first attacked the posterior columns of the spinal cord, the posterior root as it pierced the pia, the more distal part of the radicular nerve where it acquires its arachnoid and dural sheaths, or the dorsal root ganglion cell. The observations of our colleagues of rare active cases have shown the inflammation to be all along the posterior root; the slight dorsal ganglion cell loss and posterior column degeneration were found to be secondary. The hypotonia, areflexia, and ataxia relate to destruction of proprioceptive fibers in the sensory roots. The hypotonia and insensitivity of the bladder are due to deafferentation at the S2 and S3 levels; the same is true of the impotence and obstipation. Lightning pains and visceral crises cannot be fully explained but are probably attributable to incomplete posterior root lesions at different levels. Analgesia and joint insensitivity relate to the partial loss of A and C fibers in the roots. If there is no pleocytosis, the CSF protein content is normal, and there is no evidence of cardiovascular or other types of syphilis, no further antisyphilitic treatment is necessary. If the CSF is positive, the patient should be treated with penicillin, as described below. Residual symptoms in the form of lightning pains, gastric crises, Charcot joints, or urinary incontinence frequently continue long after all signs of active neurosyphilitic infection have disappeared. These should be treated symptomatically rather than by antisyphilitic drugs.

Syphilitic Optic Atrophy This takes the form of progressive blindness beginning in one eye and then involving the other. The usual finding is a constriction of the visual fields, but scotomata may occur in rare cases. The optic discs are gray-white. Other forms of neurosyphilis, particularly tabes dorsalis, not infrequently coexist. The CSF is almost invariably abnormal, though the degree of abnormality may be slight in some cases. The prognosis is poor if vision in both eyes is greatly reduced. If only one eye is badly affected, sight in the other eye can usually be saved. In exceptional cases, visual impairment may progress, even after the CSF becomes negative. The pathologic changes consist of a perioptic meningitis, with subpial gliosis and fibrosis replacing degenerated optic nerve fibers. Exceptionally there are vascular lesions with infarction of central parts of the nerve.

Spinal Syphilis There are several types of spinal syphilis other than tabes. Two of them, syphilitic meningomyelitis (sometimes called Erb's spastic paraplegia because of the predominance of bilateral corticospinal tract signs) and spinal meningovascular syphilis, are observed from time to time, though less often than tabes. Spinal meningovascular syphilis may occasionally take the form of an anterior spinal artery syndrome. In meningomyelitis, there occurs a subpial loss of myelinated fibers and gliosis as a direct result of the chronic fibrosing meningitis. Gumma of the spinal meninges and cord seldom is found. It was not present in a single case in Merritt and Adams' study of spinal syphilis. Progressive muscular atrophy (syphilitic amyotrophy) is a very rare disease of questionable syphilitic etiology; most cases are degenerative. Also rare is syphilitic hypertrophic pachymeningitis or arachnoiditis, which allegedly gives rise to radicular pain, amyotrophy of the hands, and signs of long tract involvement in the legs (syphilitic

amyotrophy with spastic-ataxic paraparesis). In all these syndromes there is an abnormal CSF unless, of course, the

neurosyphilitic infection is burned out. The prognosis in spinal neurosyphilis is uncertain. There is improvement or at least an arrest of the disease process in mostinstances, though a few patients may progress slightly after treatment is begun. A steady advance of the disease in the face of a negative CSF usually means that there has been a secondary constrictive myelopathy or that the original diagnosis was incorrect and the patient suffers from some other disease, e.g., a spinal form of multiple sclerosis as a degenerative disease.

Syphilitic Nerve Deafness. This may occur in either early or late syphilitic meningitis and may be combined with other syphilitic syndromes. We and our colleagues have had little clinical experience with this disorder and have no pathologic material.

Treatment of Neurosyphilis The treatment of these tertiary forms of neurosyphilis consists of the administration of penicillin G, given intravenously in a dosage of 18 to 24 million units daily (3 to 4 million units every 4 h) for 14 days. Erythromycin and tetracycline, in doses of 0.5 g every 6 h for 20 to 30 days, are suitable substitutes in patients who are sensitive to penicillin. The so-called Jarisch-Herxheimer reaction, which occurs after the first dose of penicillin and is a matter of concern in the treatment of primary syphilis, is of little consequence in neurosyphilis; it usually consists of no more than a mild temperature elevation and leukocytosis. The effects of treatment on certain symptoms of neurosyphilis, especially of tabetic neurosyphilis, are unpredictable and often little influenced by treatment with penicillin; they require other measures. Lightning pains may respond to gabapentin or carbamazepine. Analgesics may be helpful, but opiates should generally be avoided. Neuropathic (Charcot) joints require bracing or fusion. Atropine and phenothiazine derivatives are said to be useful in the treatment of visceral crises. In all forms of neurosyphilis, the patient should be re-examined every 3 to 6 months and the CSF should be retested after a 6-month interval. If after 6 months the patient is free of symptoms and the CSF abnormalities have been reversed (disappearance of cells as well as reduction in protein, gamma globulin, and serology titers), no further treatment is indicated. Follow-up should include clinical examinations at 9 and 12 months and another lumbar puncture as part of the latter examination. Satisfactory progress is judged by absence of symptoms and further improvement in the CSF. These procedures should be repeated every 6 months until the CSF becomes completely negative. In the opinion of most experts, a persistent weakly positive serologic (VDRL) test after the cells and protein levels have returned to normal does not constitute an indication for additional treatment. Such a CSF formula assures that the disease is quiescent or arrested. Others are not convinced of the reliability of this concept and prefer to give more penicillin. If at the end of 6 months there are still an increased number of cells and an elevated protein in the fluid, another full course of penicillin should be given. Clinical relapse is almost invariably attended by recurrence of cells and increase in protein levels. Rapid clinical progression in the face of a negative CSF suggests the presence of a nonsyphilitic disease of the brain or cord. A proven maxim for treatment was that a syphilitic patient who has neither abnormal CSF nor aortitis needs no therapy. (In the days of arsenical treatment, the therapy put patients at greater risk than did the disease). Finally, it may be said that even with the mild resurgence of the disease due to AIDS, the neurologist finds the various forms of neurosyphilis of more theoretical than practical importance. But no other disease has portrayed more vividly the effects of a chronic, continuously active cerebrospinal meningitis on the entire neuraxis.

Primary HIV infection. Early manifestations at the time of seroconversion are rare; these include acute reversible encephalitis or aseptic meningitis, cranial nerve deficits (especially facial nerve palsy), radiculitis, or myelitis.

Neurological signs are usually late manifestations of HIV infection. HIV encephalopathy progresses over several months and is characterized by lethargy, headache, increasing social withdrawal, insomnia, forgetfulness, lack of concentration, and apathy. Advanced AIDS is

accompanied by bradyphrenia, impaired ocular pursuit, dysarthrophonia, incoordination, myoclonus, rigidity, and postural tremor. Incontinence and central paresis develop in the final stages of the disease. CT of the brain reveals generalized atrophy, and MRI reveals multifocal or diffuse white-matter lesions. The CSF examination may be normal or reveal a low-grade pleocytosis and an elevated protein concentration. EEG reveals increased slow-wave activity. Other neurological manifestations include HIV myelopathy (vacuolar myelopathy), distal symmetrical polyradiculoneuropathies, mononeuritis, multiplex, and polymyositis.

Virustatic treatment. Antiretroviral combination therapy (HAART: highly active antiretroviral therapy).

Secondary complications of HIV infection include opportunistic CNS infections (toxoplasmosis, cryptococcal meningitis, aspergillosis, progressive multifocal leukoencephalopathy, cytomegalovirus encephalitis, herpes simplex encephalitis, herpes zoster, tuberculosis, syphilis), tumors (primary CNS lymphoma), stroke (infarction, hemorrhage), and metabolic disturbances (iatrogenic or secondary to vitamin deficiency).

Toxoplasmosis

Of the focal complications, cerebral toxoplasmosis is the most frequent (and treatable; see page 623). In the autopsy series of AIDS reported by Navia and colleagues, areas of inflammatory necrosis due to Toxoplasma were found in approximately 13 percent. Lumbar puncture, contrastenhanced CT scanning, and MRI are useful in diagnosis. The spinal fluid usually shows an elevation of protein in the range of 50 to 200 mg/dL, and one-third of patients have a lymphocytic pleocytosis. Since the disease represents reactivation of a prior Toxoplasma infection, it is important to identify Toxoplasmaseropositive patients early in the course of AIDS and to treat them vigorously with oral pyrimethamine (100 mg initially and then 25 mg daily)and a sulfonamide (4 to 6 g daily in four divided doses). Curiously, the toxoplasmosis infection, so common in the brains of AIDS patients, is not a frequent cause of the typical infestation, namely, myositis. The main clinical problem in reference to toxoplasmosis in AIDS is its differentiation from cerebral lymphoma.

CNS Lymphoma In the Johns Hopkins study (see Johnson), about 11 percent of AIDS patients developed a primary CNS lymphoma, which may in some cases be difficult to distinguish from toxoplasmosis clinically and radiologically. If the cytologic study of the CSF is negative and there has been no response to antibiotics, stereotaxic brain biopsy may be necessary for diagnosis. The prognosis in such patients is considerably less favorable than in non-AIDS patients; the response to radiation therapy, methotrexate, and corticosteroids is short-lived, and survival is usually measured in months. In the face of enhancing focal brain lesions in AIDS, the current approach is to assume initially the presence of toxoplasmosis, which is treatable. Antibody tests for toxoplasmosis should be obtained; the absence of IgG antibodies mandates that treatment be changed in order to address the problem of brain lymphoma. Also, if antitoxoplasmal therapy with pyrimethamine and sulfadiazine fails to reduce the size of the lesions within several weeks, another cause should be sought, again mainly lymphoma. In those patients who cannot tolerate the frequent side effects of pyrimethamine or sulfonamides (rash or thrombocytopenia), clindamycin may be of value. Recently it has been suggested that thallium isotope singlephoton emission computed tomography (SPECT)and positron emission tomography (PET)can reliably exclude lymphoma as the cause of a mass lesion in the AIDS patient. The less frequent possibilities of tuberculous or bacterial brain abscess should be kept in mind if none of the other avenues allow a confident diagnosis.

Cytomegalovirus Among the nonfocal neurologic complications of AIDS, the most common are CMV and cryptococcal infections. At autopsy, about one-third of AIDS patients are found to be infected with CMV. However, the contribution of this infection to the total clinical

picture is often uncertain. Despite this ambiguity, certain features have emerged as typical of CMV encephalitis in the AIDS patient. According to Holland and colleagues, late in the course of AIDS and usually concurrent with CMV retinitis, the encephalopathy evolves over 3 to 4 weeks. Its clinical features include an acute confusional state or delirium combined, in a small proportion of cases, with cranial nerve signs including ophthalmoparesis, nystagmus, ptosis, facial nerve palsy, or deafness. In one of our patients, there were progressive oculomotor palsies that began with light-fixed pupils. Pathologic specimens and MRI show the process to be concentrated in the ventricular borders, especially evident as T2 signal hyperintensity in these regions. It may be seen to extend more diffusely through the adjacent white matter and be accompanied by meningeal enhancement by gadolinium in a few cases. Extensive destructive lesions have also been reported; this has been true in two of our own cases. Such lesions may be associated with hemorrhagic changes in the CSF in addition to showing an inflammatory response. CMV may also produce a painful lumbosacral polyradiculitis in AIDS. The diagnosis of CMV infection during life is often difficult. Cultures of the CSF are usually negative and IgG antibody titers are nonspecifically elevated. Newer PCR methods may prove useful here. Where the diagnosis is strongly suspected, treatment with the antiviral agents ganciclovir and foscarnet has been recommended; but, as pointed out by Kalayjian and colleagues, the CMV disease may develop and progress while patients are taking these medications as a form of maintenance therapy.

Cryptococcal Infection Meningitis with this organism and less often solitary cryptococcoma are the most frequent fungal complications of HIV infection. Flagrant symptoms of meningitis or meningoencephalitis may be lacking, however, and the CSF may show little abnormality with respect to cells, protein, and glucose. For these reasons, evidence of cryptococcal infection of the spinal fluid should be actively sought with India ink preparations, antigen testing, and fungal cultures. Treatment is along the lines outlined.

Varicella Zoster Cerebral infections with this virus are less common complications of AIDS, but when they do occur, they tend to be severe. They take the form of multifocal lesions of the cerebral white matter, somewhat like those of progressive multifocal leukoencephalopathy, a cerebral vasculitis with hemiplegia (usually in association with ophthalmic zoster), or rarely a myelitis. Encephalitis due to HSV-1 and HSV-2 has also been identified in the brains of AIDS patients, but the clinical correlates are unclear. There is no evidence that acyclovir or other antiviral agents are effective in any of these viral infections. Shingles involving several contiguous dermatomes is known to occur in AIDS, as in other immunosuppressed conditions.

Progressive Multifocal Leukoencephalopathy (PML)

Pathogenesis. The causative organism, JC virus, is a ubiquitous papovavirus that usually stays dormant within the body. It is reactivated in persons with impaired cellular immunity and spreads through the bloodstream to the CNS, where it induces multiple white-matter lesions.

Symptoms and signs. PM appears as a complication of cancer (chronic lymphatic leukemia,

Hodgkin lymphoma), tuberculosis, sarcoidosis, immune suppression, and AIDS, producing variable symptoms and signs. The major manifestations in patients without AIDS are visual disturbances (visual field defects, cortical blindness), hemiparesis, and neuropsychological disturbances (impairment of memory and cognitive functions, dysphasia, behavioral abnormalities). The major manifestations of PML as a complication of AIDS are (from most to least frequent): central paresis, cognitive impairment, visual disturbances, gait impairment, ataxia, dysarthria, dysphasia, and headache. PML usually progresses rapidly, causing death in 4–6 months. The definitive diagnosis is by histological examination of brain tissue obtained by biopsy or necropsy. CT reveals asymmetrically distributed, hypodense white-matter lesions without mass effect or contrast enhancement; these lesions are hyperintense on T2-weighted MRI, which also

demonstrates involvement of the subcortical white matter ("U fibers"). The CSF findings are usually normal, but oligoclonal bands may be found in AIDS patients.

Virustatic therapy. There is as yet no validated treatment regimen.

Cytomegalovirus (CMV) Infection

Pathogenesis. CMV, a member of the herpesvirus family, is transmitted through respiratory droplets, sexual intercourse, and contact with contaminated blood, blood products, or transplanted organs. It is widely distributed throughout the world, with a regional and agedependent prevalence of up to 100%. CMV virions are thought to replicate initially in oropharyngeal epithelial cells (salivary glands) and then disseminate to the organs of the body, including the nervous system, through the bloodstream. The virus remains dormant in monocytes and lymphocytes as long as the immune system keeps it in check. Reactivation of the virus is almost always asymptomatic in healthy individuals, but severe generalized disease can develop in persons with immune compromise due to AIDS, organ transplantation, immunosuppressant drugs, or a primary malignancy.

Symptoms and signs. The primary infection is usually clinically silent. Intrauterine fetal infection leads to generalized fetopathies in fewer than 5% of neonates. In immunocompromised patients, particularly those with AIDS, (reactivated) CMV infection presents a variable combination of manifestations, including retinitis (partial or total loss of vision), pneumonia, and enteritis (colitis, esophagitis, proctitis). The neurological manifestations of CMV infection are manifold. PNS involvement is reflected as Guillain–Barre syndrome or lumbosacral polyradiculopathy (subacute paraparesis with or without back pain or radicular pain). CNS involvement produces encephalitis, meningitis, ventriculitis (inflammatory changes in the ependyma) and/or myelitis. Symptoms and signs may be absent, minor, or progressively severe, as in HIV-related encephalopathy. CMV vasculitis may lead to ischemic stroke. The diagnosis usually cannot be made from the clinical findings alone (except in the case of CMV retinitis). MRI reveals periventricular contrast enhancement in CMV vasculitis; other MRI and CT findings are nonspecific. There may be CSF pleocytosis with an elevated protein concentration. The diagnosis can be established by culture or identification by polymerase chain reaction of CMV in tissue, CSF, or urine, or by serological detection of CMV-specific antibodies.

Virustatic therapy. Gancyclovir, foscarnet, or cidofovir are given for initial treatment and secondary prophylaxis.

Tuberculosis Two particular types of mycobacterial infection tend to complicate AIDS-Mycobacterium tuberculosis and Mycobacterium avium-intracellulare. Tuberculosis predominates among drug abusers and AIDS patients in underdeveloped countries, and a higher than usual proportion of these immunosuppressed individuals develop tuberculous meningitis. Diagnosis and treatment are along the same lines as in non-AIDS patients. Atypical mycobacterial infections are usually associated with other destructive cerebral lesions and respond poorly to therapy.

Tuberculous Meningitis

Pathogenesis. Mycobacterium tuberculosis transmission in man is usually by transfer of droplets fromand to the respiratory tract (rarely orally or through skin lesions). The pathogen replicates in the lungs (primary infection), either in the lung tissue itself or within alveolar macrophages. Macrophages can only destroy tubercle bacilli after they have been activated by T cells; the course of the infection thus depends on the state of the immune system, i.e., on the ability of activated macrophages to hold the bacilli in check. The stage of primary infection lasts 2–4 weeks, is not necessarily symptomatic (if it is, then with nonspecific symptoms such as fever, anorexia, and lethargy), and cannot be detected by immune tests performed on the skin. The inflammatory process may also involve the regional (hilar) lymph nodes (primary complex). Calcified foci in the primary complex are easily seen on plain radiographs of the chest. The bacilli may remain dormant for years or may be reactivated when the patient's immune defenses are lowered by HIV infection, alcoholism, diabetes mellitus, corticosteroid therapy, or other factors

(reactivated tuberculosis). Spread from the primary focus to other organs (organ tuberculosis) can occur during primary infection in immunocompromised patients, but only after reactivation in other patients. The bacilli presumably reach the CNS by hematogenous dissemination; local extension to the CNS from tuberculous bone (spinal cord, base of skull) is rare. **Symptoms and signs.** The type and focus of CNS involvement (neurotuberculosis) vary, depending mainly on the age and immune status of the host.

Tuberculous meningoencephalitis. The prodromal stage lasts 2–3 weeks and is characterized by behavioral changes (apathy, depression, irritability, confusion, delirium, lack of concentration), anorexia, weight loss, malaise, nausea, and fever. Headache and neck stiffness reflect meningeal involvement. Finally, cerebral involvementmanifests itself in focal signs (deficits of CN II, III, VI, VII, and VIII; aphasia, apraxia, central paresis, focal epileptic seizures, SIADH) and/or general signs (signs of intracranial hypertension, hydrocephalus). The focal signs are caused by leptomeningeal adhesions, cerebral ischemia due to vasculitis, or mass lesions (tuberculoma). Chronic meningitis most likely reflects inadequate treatment, or resistance of the pathogen, rather than being a distinct form of the disease. Diagnosis: CSF examination for initial diagnosis and monitoring of disease course. The diagnosis of tuberculous meningitis can only be confirmed by detection of mycobacteria in the CSF with direct microscopic visualization, culture, or molecular biological techniques. As the prognosis of untreated tuberculous meningitis is poor, treatment for presumed disease should be initiated as soon as the diagnosis is suspected from the clinical examination and CSF findings; the latter typically include high concentrations of protein (several grams/liter) and lactate, a low glucose concentration (<50% of blood glucose), a high cell count (over several hundred), and a mixed pleocytosis (lymphocytes, monocytes, granulocytes).

Tuberculoma is a tumorlike mass with a caseous or calcified core surrounded by granulation tissue (giant cells, lymphocytes). Tuberculomas may be solitary or multiple and are to be differentiated from tuberculous abscesses, which are full of mycobacteria and lack the surrounding granulation tissue. Diagnosis: CT or MRI.

Spinal tuberculosis. Transverse spinal cord syndrome can arise because of tuberculous myelomeningoradiculitis, epidural tuberculous abscess associated with tuberculous spondylitis/discitis, or tuberculoma. Diagnosis: MRI.

Antibiotic treatment. One treatment protocol specifies a combination of isoniazid (with vitamin B6), rifampicin (initially i. v., then p.o.), and pyrazinamide (p.o.). After 3 months, pyrazinamide is discontinued, and treatment with isoniazid and rifampicin is continued for a further 6–9 months. The treatment for HIVpositive patients includes up to five different antibiotics.

Materials for self-control of quality of preparation:

A 35-year-old woman who has received a liver transplant develops meningeal signs and fever. Cerebrospinal fluid testing reveals a fungal infection. Which of the following is the most common cause of fungal meningitis?

a. Aspergillus
b. Candida
c. Mucor
d. Cryptococcus
e. Rhizopus

A 17-year-old right-handed boy has had infectious meningitis 8 times over the past 3 years. He has otherwise been generally healthy and developed normally. Recurrent meningitis often develops in persons with which of the following?

a. Otitis media
b. Epilepsy
c. Multiple sclerosis

d. Whipple's disease
e. Cerebrospinal fluid (CSF) leaks

A 27-year-old man presents to his primary care doctor with a lowgrade fever, headache, and neck stiffness, which have become more bothersome over the past 1 to 2 weeks. CSF and serological testing for Lyme is positive and antibiotic treatment is initiated. The cranial neuropathy most commonly found with Lyme disease is that associated with damage to which cranial nerve?

a. III
b. V
c. VII
d. IX
e. XII

A 13-year-old boy is brought into the emergency room lethargic with a stiff neck and fever. Despite aggressive therapy, the child dies. Postmortem evaluation reveals that the child had primary amebic meningoencephalitis. This condition is usually acquired through which of the following means?

a. Freshwater swimming
b. Eating contaminated meat
c. Eating calves' brains
d. Anal intercourse
e. Animal bites

Following several days of low-grade fever and mild neck and head pain, a 10-year-old boy develops bilateral face drooping and difficulty fully closing his eyes. Serum is positive for Borrelia burgdorferi IgM. CSF PCR is also positive for this organism's DNA. After B. burgdorferi is introduced by the tick that carries it, the skin around the bite develops which of the following?

a. An exfoliative dermatitis
b. Purpura
c. Localized edema
d. Erythema chronicum migrans
e. Vesicular lesions

The most striking neurologic complication of von Economo's encephalitis (encephalitis lethargica), a type of encephalitis that occurred in epidemic proportions along with viral influenza between 1917 and 1928, was which of the following?

a. Blindness
b. Hearing loss
c. Paraplegia
d. Parkinsonism
e. Incontinence

A 17-year-old female presents initially with fever and progressive weakness. An extensive neurological evaluation including EMG/NCS suggests a motor neuron disease. The motor neuron disease most certainly traced to a virus is which of the following?

a. Poliomyelitis
b. Subacute sclerosing panencephalitis (SSPE)
c. Progressive multifocal leukoencephalopathy (PML)
d. Subacute HIV encephalomyelitis
e. Kuru

A 72-year-old right-handed woman has 2 days of headache and fever, followed by worsening confusion. She is taken to the hospital after having a generalized seizure. A head CT is consistent with left temporal hemorrhage and swelling. Localization of an encephalitis to the medial temporal or orbital frontal regions of the brain is most consistent with which of the following?

a. Treponema pallidum
b. Varicella zoster virus
c. Herpes simplex virus
d. Cryptococcus neoformans
e. Toxoplasma gondii

A 21-year-old college student was found walking around his dormitory naked. He is disoriented, inattentive, and shows poor comprehension. In the emergency room he is found to have a fever of 102°F . There are no apparent motor, sensory, or coordination abnormalities. The emergency room physician orders a brain MRI and then decides to perform a lumbar puncture. Neuroimaging of the brain before attempting a lumbar puncture is advisable in cases of acute encephalitis for which one of the following reasons?

a. The diagnosis may be evident on the basis of magnetic resonance imaging (MRI) alone
b. Massive edema in the temporal lobe may make herniation imminent
c. The computed tomography (CT) picture may determine whether a brain biopsy should be obtained
d. Shunting of the ventricles is usually indicated, and the imaging studies are needed to direct the placement of the shunt
e. It may establish which pathology is responsible

A 38-year-old man who is immunocompromised because of HIV presents with 1 month of worsening right headache, ear pain, and fever. He is determined to have malignant external otitis and osteomyelitis of the base of the skull. The etiology is fungal. Fungal malignant external otitis and osteomyelitis of the base of the skull in HIV patients is most commonly caused by which of the following?

a. Nocardia
b. Cryptococcus neoformans
c. Actinomyces
d. Aspergillus
e. Candida

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources

1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical Class No. 6.

Theme: Cranial nerves I - XII and syndromes of damage

Actuality of theme.

The brainstem is a very important part of the CNS, which includes 10 CN, a lot of pathways and provide all vital functions. The early diagnosis of the affection of the brainstem can prevent the dangerous complications.

Whole employments:

Educational aims:

- to acquaint students with the variety of the special sense and importance of the normal functioning of vision, smell. with the structure and functions of the brainstem, create the picture of importance of the normal functioning of this structure.

a student must know:

- Structure of visual analyzer.
- Terminology of the lesion of visual pathways.
- Structure of smell analyzer.
- Functions of the groups of CNs.
- Symptoms of lesion of CNs.

Table of contents of the class:

Olfactory epithelium. The olfactory mucosa on either side of the nasal cavity occupies an area of approximately 2.5 cm2 on the roof of the superior nasal concha, extending to the nasal septum.

The mucus covering the olfactory epithelium is necessary for olfactory function, because molecules interact with olfactory receptors only when they are dissolved in the mucus.

Olfactory cells are bipolar sensory cells with a mean lifespan of about 4 weeks. Fine bundles of cilia project from one end of each olfactory cell into the mucus. *Olfactory receptors* located on the cilia are composed of specific receptor proteins that bind particular odorant molecules. Each olfactory cell produces only one type of receptor protein; the cells are thus *chemotopic*, i.e., each responds to only one type of olfactory stimulus.

Olfactory cells are uniformly distributed throughout the olfactory mucosa of the nasal conchae.

Olfactory pathway. The unmyelinated axons of all olfactory cells converge in bundles of up to 20 *fila olfactoria* on each side of the nose (these bundles are the true olfactory nerves), which pass through the cribriform plate to the olfactory bulb. Hundreds of olfactory cell axons converge on the dendrites of the mitral cells of the olfactory bulb, forming the *olfactory glomeruli*. Other types of neurons that modulate the olfactory input (e. g., granular cells) are found among the mitral cells. Neural impulses are relayed through the projection fibers of the olfactory tract to other areas of the brain including the prepiriform cortex, limbic system, thalamus (medial nucleus), hypothalamus, and brain stem reticular formation. This complex interconnected network is responsible for the important role of smell in eating behavior, affective behavior, sexual behavior, and reflexes such as salivation. The *trigeminal nerve* supplies the mucous membranes of the nasal, oral, and pharyngeal cavities. Trigeminal receptor cells are also stimulated by odorant molecules, but at a higher threshold than the olfactory receptor cells.

Olfactory Disturbances (Dysosmia)

Olfactory disturbances can be classified as either *quantitative* (anosmia, hyposmia, hyperosmia) or *qualitative* (parosmia, cacosmia). Congenital olfactory disturbances manifest themselves as *partial anosmia* ("olfactory blindness").

The perceived intensity of a persistent odor decreases or disappears with time (*olfactory adaptation*). External factors such as an arid environment, cold, or cigarette smoke impair the ability to smell; diseases affecting the nasopharyngeal cavity impair both smell and taste. Odors and emotions are closely linked and can influence each other. The perception of smell may be qualitatively changed (*parosmia*) because of autonomic (hunger, stress) and hormonal changes (pregnancy) or disturbances such as ozena, depression, traumatic lesions, or nasopharyngeal empyema. *Olfactory hallucinations* can be caused by mediobasal and temporal tumors (focal epilepsy), drug or alcohol withdrawal, and psychiatric illnesses such as schizophrenia or depression.

Tests of smell. One nostril is held closed, and a bottle containing a test substance is held in front of the other. The patient is then asked to inhale and report any odor perceived. In this subjective test, odor perception per se is more important than odor recognition. Odor perception indicates that the peripheral part of the olfactory tract is intact; odor recognition indicates that the cortical portion of the olfactory pathway is also intact. More sophisticated tests may be required in some cases. Because there is bilateral innervation, unilateral lesions proximal to the anterior commissure and cortical lesions may not cause anosmia.

Anosmia/hyposmia. Unilateral anosmia may be caused by a tumor (meningioma). *Korsakoff syndrome* can render the patient unable to identify odors. Viral infections (influenza), heavy smoking, and toxic substances can damage the olfactory epithelium; trauma (disruption of olfactory nerves, frontal hemorrhage), tumors, meningitis, or radiotherapy may damage the olfactory pathway. Parkinson disease, multiple sclerosis, Kallmann syndrome (congenital anosmia with hypogonadism), meningoencephalocele, albinism, hepatic cirrhosis, and renal failure can also cause olfactory disturbances.

Retina. Visible light is electromagnetic radiation at wavelengths of 400–750 nanometers. The *dioptric system* (cornea, aqueous humor of the anterior and posterior ocular chambers, pupil, lens, vitreous body) produces a miniature, upside-down mirror image of the visual field on the retina. The fovea, located in the center of the macula at the posterior pole of the eyeball, is the area of sharpest vision in daylight. Blood is supplied to the eye by the ophthalmic artery via the ciliary arteries (supplies the choroid) and the central retinal artery (supplies the retina). The optic disk, the central retinal artery that branches from it, and the central retinal vein can be examined by *ophthalmoscopy*.

Visual pathway. The visual pathway begins in the retina (first three neurons) and continues through the optic nerve to the optic chiasm, from which it continues as the optic tract to the lateral geniculate body. The *optic radiation* arises at the lateral geniculate body and terminates in the *primary* (area 17) and *secondary visual areas* (areas 18, 19) of the occipital lobe. The fibers of the retinal neuronal network converge at the *optic disk* before continuing via the optic nerve to the *optic chiasm*, in which the medial (nasal) fibers cross to the opposite side. The right optic tract thus contains fibers from the temporal half of the right retina and the nasal half of the left retina. The *lateral geniculate body* is the site of the fourth neuron of the optic pathway. Its efferent fibers form the optic radiation, which terminates in the visual cortex (striate cortex) of the occipital lobe. The central foveal area has the largest cortical representation. The visual pathway is interconnected with midbrain nuclei (medial, lateral, and dorsal terminal nuclei of the pretectal region; superior colliculus), nonvisual cortical areas (somatosensory, premotor, and auditory), the cerebellum, and the pulvinar (posterior part of thalamus).

Visual field. The *monocular visual field* is the portion of the external world seen with one eye, and the *binocular visual field* is that seen by both eyes. The visual fields of the two eyes overlap; the overall visual field therefore consists of a central zone of clear binocular vision produced by the left and right central foveae, a peripheral binocular zone, and a monocular zone. Partial decussation at the optic chiasm brings visual information from the right (left) side of the world to the left (right) side of the brain. The visual field is topographically represented at all levels of the visual pathway from retina to cortex; lesions at any level of the pathway cause visual field defects of characteristic types. If the images on the two retinas are displaced by more than a certain threshold distance, double vision (diplopia) results. This is most commonly due to disturbances of the extraocular muscles, e. g., paralysis of one or more of these muscles.

Color vision. Testing of color vision requires standard definition of the colors red, blue, and green. The visual threshold for various colors, each defined as a specific mixture of the three primary colors, is determined with a standardized color perception chart. *Disturbances of color vision* may be due to disturbances of the dioptric system, the retina, or the visual pathway. Cortical lesions cause various kinds of visual agnosia. Lesions of area 18 may make it impossible for patients to recognize colors despite intact color vision (color agnosia), or to recognize familiar objects (object agnosia) or faces (prosopagnosia). Patients with lesions of area 19 have intact vision but cannot recognize or describe the objects that they see. Spatial orientation may be impaired (visuospatial agnosia), as may the inability to draw pictures. Persons with visual agnosia may need to touch objects to identify them.

Limbic system. Connections with the limbic system(hippocampus, amygdala, parahippocampal gyrus;) account for the ability of visual input to evoke an emotional response.

Examination. The visual fields of both eyes should always be jointly assessed. The *confrontation test*, in which the examiner "confronts" the patient's visual field with his or her own, intact, contralateral visual field, is used to check for visual field defects. For the test to be

performed correctly, the patient and the examinermust first fixate along the same line. The examiner then slowly moves a white or red object (at least 1 cm in diameter) from the periphery of the visual field toward the center in a number of different directions, and determines where the patient can and cannot see it. Alternatively, the examiner may raise one or more fingers and ask the patient to count them (a useful test for small children, and for persons whose vision is so poor that it cannot be tested by the first method). The perceived *brightness* (unequal in patients with hemianopsia) of the hand in the nasal and temporal portions of the visual field is also determined. The *red vision test* enables the detection of a central scotoma as an area in which the red color is perceived as less intense. More detailed information can be obtained by further ophthalmological testing (Goldmann perimetry, automatic perimetry).

Visual field defect (scotoma). The thin myelinated fibers in the center of the optic nerve, which are derived from the papillomacular bundle, are usually the first to be affected by optic neuropathy (*central scotoma*). From the optic chiasm onward, the right and left visual fields are segregated into the left and right sides of the brain. Unilateral lesions of the retina and optic nerve cause monocular deficits, while retrochiasmatic lesions cause homonymous defects (quadrantanopsia, hemianopsia) that do not cross the vertical meridian, i.e., affect one side of the visual field only. Anterior retrochiasmatic lesions cause incongruent visual field defects, while posterior retrochiasmatic lesions lead to congruent visual field defects. Temporal lobe lesions cause mildly incongruent, contralateral, superior homonymous quadrantanopsia. Bitemporal visual field defects (heteronymous hemianopsia) have their origin in the chiasm. Unilateral retrochiasmatic lesions cause visual field defects but do not impair visual acuity. Organic visual field defects widen pregressively with the distance of test objects from the eye, whereas psychogenic ones are constant (“tubular fields”).

Prechiasmatic lesions may affect the retina, papilla (= optic disk), or optic nerve. Transient episodes of monocular blindness (amaurosis fugax). Acute or subacute unilateral blindness may be caused by optic or retrobulbar neuritis, papilledema (intracranial mass, pseudotumor cerebri), cranial arteritis, toxic and metabolic disorders, local tumors, central retinal artery occlusion, or central retinal vein occlusion.

Chiasmatic lesions. Lesions of the optic chiasm usually produce bitemporal visual field defects. Yet, because the medial portion of the chiasm contains decussating fibers while its lateral portions contain uncrossed fibers, the type of visual field defect produced varies depending on the exact location of the lesion. As a rule, anterior chiasmatic lesions that also involve the optic nerve cause a central scotoma in the eye on the side of the lesion and a superior temporal visual field defect (junction scotoma) in the contralateral eye. Lateral chiasmatic lesions produce nasal hemianopsia of the ipsilateral eye; those that impinge on the chiasm from both sides produce binasal defects. Dorsal chiasmatic lesions produce bitemporal hemianopic paracentral scotomata. Double vision may be the chief complaint of patients with bitemporal scotomata.

Retrochiasmatic lesions. Depending on their location, retrochiasmatic lesions produce different types of *homonymous unilateral scotoma*: the defect may be congruent or incongruent, quadrantanopsia or hemianopsia. As a rule, temporal lesions cause contralateral superior quadrantanopsia, while parietal lesions cause contralateral inferior quadrantanopsia. Complete hemianopsia may be caused by a relatively small lesion of the optic tract or lateral geniculate body, or by a more extensive lesion more distally along the visual pathway. Sparing of the temporal sickle indicates that the lesion is located in the occipital interhemispheric fissure. *Bilateral homonymous scotoma* is caused by bilateral optic tract damage. The patient suffers from “tunnel vision” but the central visual field remains intact (sparing of macular fibers). Cortical blindness refers to subnormal visual acuity due to bilateral retrogeniculate lesions. Bilateral *altitudinal* homonymous hemianopsia (i.e., exclusively above or exclusively belowthe visual equator) is due

to extensive bilateral damage to the temporal lobe (superior scotoma) or parietal lobe (inferior scotoma).

Perception of Sound

Sound waves enter the ear through the external acoustic meatus and travel through the ear canal to the tympanic membrane (eardrum), setting it into vibration. Vibrations in the 20–16 000 Hz range (most sensitive range, 2000–5000 Hz) are transmitted to the auditory ossicles (malleus, incus, stapes). The base of the stapes vibrates against the oval window, creating waves in the perilymph in the vestibular canal (scala vestibuli) of the cochlea; these waves are then transmitted through the connecting passage at the cochlear apex (helicotrema) to the perilymph of the tympanic canal (scala tympani). (Oscillations of the round window compensate for volume changes caused by oscillations of the oval window. Sound waves can also reach the cochlea by direct conduction through the skull bone.) Migrating waves are set in motion along the basilar membrane of the cochlear duct; they travel from the stapes to the helicotrema at decreasing speed, partly because the basilar membrane is less tense as it nears the cochlear apex. Thesewaves have their amplitude maxima at different sites along the basilar membrane, depending on frequency (tonotopicity): there results a frequency-specific excitation of the receptor cells for hearing—the hair cells of the organ of Corti, which is adjacent to the basilar membrane as it winds through the cochlea.

Cochlear Nerve

The tonotopicity of the basilar membrane causes each hair cell to be tuned to a specific sound frequency (*spectral analysis*). Each hair cell is connected to an afferent fiber of the cochlear nerve inside the organ of Corti. The cochlear nerve is formed by the central processes of the bipolar neurons of the cochlear ganglion (the first neurons of the auditory pathway); it exits from the petrous bone at the internal acoustic meatus, travels a short distance in the subarachnoid space, and enters the brain stem in the cerebellopontine angle. Central auditory processing involves interpretation of the pattern and temporal sequence of the action potentials carried in the cochlear nerve.

Auditory Pathway

As it ascends from the cochlea to the auditory cortex, the auditory pathway gives off collateral projections to the cerebellum, the oculomotor and facial nuclei, cervical motor neurons, and the reticular activating system, which form the afferent arm of the acoustically mediated reflexes. Axons of the cochlear nerve originating in the cochlear apex and base terminate in the anterior and posterior cochlear nuclei, respectively. These nuclei contain the second neurons of the auditory pathway. Fibers from the *posterior cochlear nucleus* decussate in the floor of the fourth ventricle, then ascend to enter the lateral lemniscus and synapse in the inferior colliculus (third neuron). The inferior colliculus projects to the medial geniculate body (fourth neuron), which, in turn, projects via the acoustic radiation to the auditory cortex. The acoustic radiation passes below the thalamus and runs in the posterior limb of the internal capsule. Fibers from the *anterior cochlear nucleus* also decussate, mainly in the trapezoid body, and synapse onto the next (third) neuron in the olivary nucleus or the nucleus of the lateral lemniscus. This branch of the auditory pathway then continues through the lateral lemniscus to the inferior colliculus and onward through the acoustic radiation to the auditory cortex.

The *primary auditory cortex* (area 41: Heschl's gyrus, transverse temporal gyri) is located in the temporal operculum (i.e., the portion of the temporal lobe overlying the insula and separated from it by the sylvian cistern). Areas 42 and 22 make up the *secondary auditory cortex,* in which auditory signals are further processed, recognized, and compared with auditory memories. The auditory cortex of each side of the brain receives information from both ears (contralateral more than ipsilateral); unilateral lesions of the central auditory pathway or auditory cortex do not cause clinically relevant hearing loss.

Methods of assessment of olfactory function

One nostril is held closed, and a bottle containing a test substance is held in front of the other. The patient is then asked to inhale and report any odor perceived. In this subjective test, odor perception per se is more important than odor recognition. Odor perception indicates that the peripheral part of the olfactory tract is intact; odor recognition indicates that the cortical portion of the olfactory pathway is also intact. More sophisticated tests may be required in some cases. Because there is bilateral innervation, unilateral lesions proximal to the anterior commissure and cortical lesions may not cause anosmia.

Methods of assessment of the visual analyzer functions (acuity, visual fields, color perception)

In the measurement of distance ***visual acuity*** the Snellen chart, which contains letters (or numbers or pictures) arranged in rows of decreasing size, is used. Each eye is tested separately and, if glasses are required, glasses for distance, not reading glasses, should be used. The letter at the top of the chart subtends 5 min of an arc at a distance of 200 ft (or roughly 60 m). The patient follows rows of letters that can normally be read at lesser distances. Acuity is reported as a nonmathematical fraction that represents the patient's ability compared to that of a person with normal distance vision. Thus, if the patient can read only the top letter at 20 rather than the normal 200 ft, the acuity is expressed as 20/200 or, if the distance is measured in meters rather than feet, 6/60. If the patient's eyesight is normal, the visual acuity will equal 20/20, or 6/6 using the metric scale. Many persons, especially during youth, can read at 20 ft the line that can normally be read at 15 ft from the chart (20/15) and hence have better than normal vision. Patients with a corrected refractive error should wear their glasses for the test. For bedside testing, a "near card" or newsprint held 14 in. from the eyes can be used and the results expressed in a distance equivalent as if a distance chart had been used. Here, the Jaeger system is sometimes used also (J1 is "normal" vision, corresponding to 20/25, J5 to 20/50, J10 to 20/100, J16 to 20/200, and so on). In young children, acuity can be estimated by having them mimic the examiner's finger movements at varying distances or having them recognize and pick up objects of different sizes from varying distances.

The visual fields of both eyes should always be jointly assessed. The *confrontation test*, in which the examiner "confronts" the patient's visual field with his or her own, intact, contralateral visual field, is used to check for visual field defects. For the test to be performed correctly, the patient and the examinermust first fixate along the same line (Fig. 16). The examiner then slowly moves a white or red object (at least 1 cm in diameter) from the periphery of the visual field toward the center in a number of different directions, and determines where the patient can and cannot see it. Alternatively, the examiner may raise one or more fingers and ask the patient to count them (a useful test for small children, and for persons whose vision is so poor that it cannot be tested by the first method). The perceived *brightness* (unequal in patients with hemianopsia) of the hand in the nasal and temporal portions of the visual field is also determined. The *red vision test* enables the detection of a central scotoma as an area in which the red color is perceived as less intense. More detailed information can be obtained by further ophthalmological testing (Goldmann perimetry, automatic perimetry).

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Oculomotor nerve (III cranial nerve)

Oculomotor nerve's (III cranial nerve pair) motoneurons located on either side of the midline in the rostral part of the midbrain. These nuclei of the oculomotor nerve (III pair of cranial nerves) innervate five extraocular muscles of the eyeball, including the muscle that elevate upper eyelid. The nucleus of the oculomotor nerve also contain parasympathetic neurons (Edinger-Westphal and Perlea nucleus) involved in the processes of pupil constriction and accommodation.

Oculomotor nerve fibers that innervate the medial rectus, inferior oblique and inferior rectus muscles are located on the same side. Subnucleus of oculomotor nerve for upper rectus muscle is located on the contralateral side. Elevate upper eyelid muscle is innervated by the oculomotor nerve's central group of cells.

Trochlear nerve (IV cranial nerve)

Trochlear nerve (IV cranial nerve pair) motoneurons closely adjacent to the main part of the complex nuclei of the oculomotor nerve. Left trochlear nerve nucleus innervate the upper right oblique muscle of the eye, right nucleus - innervate the eyeball's upper-left oblique muscle.

Abducens nerve (VI cranial nerve)

Motoneurons of the abducent nerve (VI cranial nerve) innervating the lateral (outer) rectus muscle of the eye on the same side and located in the nucleus abducens in the caudal part of the pons (part of the brainstem). All three oculomotor nerves, coming from the brain stem, pass through the cavernous sinus and enter the orbit through the upper orbital fissure.

Clear binocular vision provided by joint activity of individual eye muscles (oculomotor muscles). Friendly eye movements controlled by supranuclear sight centers and their connections. Functionally, there are five different supranuclear systems. These systems provide various kinds of movements of the eyeballs. Among them there are centers that control:

- saccadic (rapid) eye movements;
- focused eye movements;
- convergent eye movement;
- retention of eye in a certain position;
- vestibular centers.

Saccadic (rapid) eye movements

Saccadic (fast) movements of the eyeball appear as a command in the opposite visual field of the cortex of the brains frontal region (field 8). Exceptions are the fast (saccadic) movement arising during stimulation fovea of the retina, which come from the occipital-parietal region of the brain. These frontal and occipital control centers in the brain have projections in the supranuclear stem centers on both sides. The activity of these stem supranuclear visual centers exposed by influences from the cerebellum and the vestibular nuclei complex. Paracentral sections of the reticular formation of the pons are the stem center that providing friendly fast (saccadic) eye movements. Simultaneous innervation of internal (medial) rectus and opposite the rectus (lateral) rectus muscle while moving eyeballs horizontally provided by medial longitudinal fasciculus. This medial longitudinal fasciculus connects the abducent nerve nucleus with subnucleus complex of oculomotor nuclei, which are responsible for the innervation of the opposite internal (medial) rectus muscle of the eye. For the beginning of vertical rapid (saccadic) eye movements requires bilateral stimulation paracentral departments of the reticular formation in the pons from the cortical brain structures. Paracentral sections of the pons reticular formation transmit signals from the brain stem to the supranuclear centers that controlling eye movement vertically. Such supranuclear eye movement center is relates an intermediate rostral nucleus of the medial longitudinal fasciculus, located in the midbrain.

Focused eye movements

Cortical center for smooth tracking or focused movements of the eyeballs is located in the occipital-parietal region of the brain. Control performed with same side, i.e., the right occipital-parietal area of the brain controls the smooth eye movement focused to the right.

Convergent eye movement

Control mechanisms of convergent movements are less well understood, however, as is known, the neurons responsible for converging eye movement are arranged in the reticular formation of the midbrain, surrounding the complex of the oculomotor nerve nuclei. They provide projections in the motor neurons of the internal (medial) rectus muscle of the eye.

Retention of eye in a specific position

Stem centers of eye movements called neural integrators. They are responsible for holding the gaze in a certain position. These centers are changing the signals of the eyeballs movement speed in information about their location. Neurons possess their property, located in the pons below (caudal) the abducens nucleus.

Eye movements in acceleration and gravity changing

Coordination of the eyeballs movements in response to changes in gravity and acceleration carried out by vestibular system (vestibulo-ocular reflex). In case of lesion, coherence movements of both eyeballs will be developed double vision, because the images projected onto the disparate (inappropriate) portions of the retina. In congenital strabismus or squint, impaired muscles balance, leading to the wrong eyeballs location (non parethic strabismus), can help to ensure that the brain will suppress one of the images. Such decrease in visual acuity in non-locating eyeball called amblyopic without anopsia. In paralytic strabismus, double vision occurs in resulting of the eyeball muscles paralysis, usually due to lesions of the oculomotor (III), trochlear (IV) or abducens (VI) cranial nerves.

Extraocular muscles and gaze palsy

There are three types of the eyeball extraocular muscles paralysis:

individual eye muscles paralysis;

friendly movements (gaze) paralysis;

mixed paralysis.

Paralysis of individual eye muscles

Typical clinical manifestations occur with isolated lesions of the oculomotor (III), trochlear (IV) or abducens (VI) nerve.

Total damage to the oculomotor (III) nerve leads to the appearance of ptosis. Ptosis manifested in the form of weakening (paresis) muscles that lifting the upper eyelid and the impairment of any movements of the eyeball upward, downward and inward, and exotropia due to safety features lateral rectus muscle. In case of damage of the oculomotor (III) nerve there are as pupil dilation (mydriasis) and the absence of his reaction to light (iridoplegiya) and paralysis of accommodation (cycloplegia). Isolated paralysis of the muscles of the iris and ciliary body called internal ophthalmoplegia.

Damage of trochlear (IV) nerve cause paralysis of the eyeballs superior oblique muscle. Similar damages of trochlear (IV) nerve lead to a deviation of the eyeball outwards and gaze downward movement difficulty (paresis). Gaze downward paresis mostly clearly manifested in turning the eye inward. Diplopia (double vision) disappears when the head is tilted to the opposite shoulder, at which the compensatory deviation of intact eyeball medially.

Damage of abducens (VI) nerve leads to paralysis of the muscles, which move the eyeball sideward. Damage of abducens (VI) nerve develops esotropia due to the predominance of eyeball's normally working medial rectus muscle’s tone exposure. With a partial paralysis of the abducens (VI) nerve patient can turn his head to the side of affected eye abductor to eliminate double vision through a compensatory effect on the eyeball's weakened lateral rectus muscle.

Intensity of listed above symptoms with damage oculomotor (III), trochlear (IV) or abducens (VI) nerve will depend on the lesions severity and its localization in patient.

Friendly gaze paralysis

Friendly gaze called simultaneous movement of both eyes in the same direction. Acute lesions one of the frontal lobes, such as cerebral infarction (ischemic stroke), can lead to a transient

paralysis random friendly movements of the eyeballs in a horizontal direction. At the same time, independent of eyeballs movements in all directions fully preserved. Paralysis friendly random movements of the eyeballs in the horizontal direction revealed in the dolls eye phenomenon during passive head rotation horizontally lying person or by caloric reflex test (vestibular caloric stimulation - infusion into the ear canal with cold water).

Unilateral damage of pons disposed downwardly of paracentral reticular formation at the abducens nerve nucleus level causes persistent gaze palsy toward destruction side and oculocephalic reflex loss. Oculocephalic reflex (or vestibulo-ocular reflex) - is a eyeballs motor reaction on vestibular apparatus mechanical or thermal irritation, as in the phenomenon of the dolls head and eyes or caloric stimulation of the external auditory canal walls with cold water. Optical image stabilization (OIS) systems in digital cameras work at the same principles.

The damage of the rostral interstitial nucleus of the medial longitudinal fasciculus in front of the midbrain and/or damage of posterior commissure cause gaze supranuclear upward palsy to this focal neurologic symptom added of pupil's dissociated reaction on light:

- pupils on light weak reaction;
- pupils fast reaction to the accommodation (eyeballs focal length changing) and view on close objects.

In some cases in a patient is also being developed paralysis of convergence (eyeballs toward each other movement, when gaze will focus on the nose). This symptom called Parinaud syndrome. Parinaud syndrome common in tumors of the pineal gland, in some cases with cerebral infarction (ischemic stroke), multiple sclerosis and hydrocephalus.

An isolated downward gaze palsy occurs rarely in patients. When this occurs, the most commonly cause is a blockage of the lumen (occlusion) of penetrating artery and bilateral midline infarction (ischemic stroke) of the midbrain. Some hereditary extrapyramidal disease (Huntington's chorea, progressive supranuclear palsy) can cause restriction of movements of the eyeballs in all directions, especially upwards.

Mixed gaze and individual eyeball muscles palsy

Simultaneous combination of the patient gaze palsy and paralysis of certain oculomotor muscles, usually a sign of midbrain or pons lesions. Lower pons parts lesions with destruction of located there abducens nucleus can lead to paralysis of fast (saccadic) movements of the eyeballs and horizontal abducens (VI) nerve paralysis on affected side.

Lesions of medial longitudinal fasciculus lead to various eye disorders in the horizontal direction (internuclear ophthalmoplegia).

Unilateral damage of medial longitudinal fasciculus caused by infarction (ischemic stroke), or demyelination process, lead to the eyeball toward movement weakness (to the nose). This may manifest clinically as complete paralysis, eyeball inwards abduction inability from the midline, or form of moderate paresis, which will manifest itself in lower speed abduction fast (saccadic) eye movements to the nose (adduction delay). Lesions on the opposite side of the medial longitudinal fasciculus, usually observed abduction nystagmus: nystagmus arising in eyeballs abduction outwards with a slow phase directed to the midline, and fast horizontal saccadic movements. Asymmetrical arrangement of eyeballs to the vertical line often develops in unilateral internuclear ophthalmoplegia. The eye on the affected side will be located higher (gipertropiya).

Bilateral internuclear ophthalmoplegia occurs in demyelinating processes, tumors, infarcts, or arteriovenous malformations. Bilateral internuclear ophthalmoplegia leads to a more complete eyeballs movement's syndrome disorder that manifest bilateral paresis of the muscles, eyeballs toward movement, a impairment of the vertical movements, tracking purposeful movements and movements, caused by the vestibular system. Noted vertical line gaze disorders, upward nystagmus during looking up and downward nystagmus when looking down. Lesion of medial longitudinal fasciculus in the midbrain overlying (rostral) sections accompanied by convergence damage (convergent eye movements to each other in the direction of the nose).

Assesment of oculomotor nerves

Third (Oculomotor) nerve A complete third nerve lesion causes ptosis, or drooping of the upper eyelid (since the levator palpebrae is supplied mainly by the third nerve), and an inability to rotate the eye upward, downward, or inward. This corresponds to a combined weakness of the medial, superior, and inferior recti and the inferior oblique muscles. The remaining actions of the fourth and sixth nerves give rise to the mnemonic "down and out" to describe the position of the eye in third nerve palsy. When the lid is passively elevated, the eye is found to be deviated outward and slightly downward because of the unopposed actions of the intact lateral rectus and superior oblique muscles. In addition, one finds a dilated nonreactive pupil (iridoplegia) and paralysis of accommodation (cycloplegia) due to interruption of the parasympathetic fibers in the third nerve. However, the extrinsic and intrinsic eye muscles may be affected separately. For example, infarction of the central portion of the oculomotor nerve, as occurs in diabetic ophthalmoplegia, typically spares the pupil, since the parasympathetic preganglionic pupilloconstrictor fibers lie near the surface. Conversely, compressive lesions of the nerve usually dilate the pupil as an early manifestation. After injury, regeneration of the third nerve fibers may be aberrant, in which case some of the fibers that originally moved the eye in a particular direction now reach another muscle or the iris; in the latter instance the pupil, which is unreactive to light, may constrict when the eye is turned up and in.

Fourth (Trochlear) nerve. A lesion of the fourth nerve, which innervates the superior oblique muscle, is the most common cause of isolated symptomatic vertical diplopia. Although oculomotor palsy was a more common cause of vertical diplopia in Keane's series, as stated earlier, in instances where this is the sole complaint, trochlear palsy (and brainstem lesions) predominate. Paralysis of the superior oblique muscle results in weakness of downward movement of the affected eye, most marked when the eye is turned inward, so that the patient complains of special difficulty in reading or going down stairs. The affected eye tends to deviate slightly upward. This defect may be overlooked in the presence of a third nerve palsy if the examiner fails to note the absence of an expected intorsion as the patient tries to move the paretic eye downward. Head tilting to the opposite shoulder (Bielschowsky sign) is especially characteristic of fourth nerve lesions; this maneuver causes a compensatory intorsion of the unaffected eye and ameliorates the double vision. Bilateral trochlear palsies, as may occur rarely after head trauma, give a characteristic alternating hyperdeviation depending on the direction of gaze (unilateral traumatic trochlear paresis is more common). A detailed review of the clinical approach to vertical diplopia is given by Palla and Straumann.

Sixth (Abducens) nerve. Lesions of the sixth nerve result in a paralysis of the abducens muscle and a resultant weakness of lateral or outward movement as well as a crossing of the visual axes. The affected eye deviates medially, i.e., in the direction of the opposing muscle. With incomplete sixth nerve palsies, turning the head toward the side of the paretic muscle overcomes the diplopia.

The pupils

Examination

The pupillomotor examination is an assessment of two of the three internal motor functions of the eye, pupil constriction and dilation (the third is lens accommodation). The motor assessment is often supplemented by slit-lamp observation of the iris, which may reveal abnormalities of iris structure. Such iris defects can cause abnormalities of pupillary function that are unrelated to any neuropathy.

To examine pupillomotor functions, seat the patient comfortably with his or her gaze directed at a distance (12–20 ft forward). The examiner should position in front, and slightly to one side, so that the pupils may be observed without interrupting the patient's fixation. For examination of the light reflex, the room should be dim, and the examination light bright. The traditional stimulus is the ophthalmologic Finoff scleral transilluminator ("muscle light"), which features a shielded,

directed beam of variable brightness, making it ideal for isolated illumination of one eye with minimal "scatter" illumination to the fellow eye. Any nonmedical light source with similar features will do as well.

Ideally, the examination of the light reflex requires the patient to be wearing their distance correction, as an unfocused distance fixation-target may stimulate pupillary contraction via the near triad—convergence, lens accommodation, and miosis—should the patient attempt to focus. This consideration is perhaps most important in young patients, whose ability to accommodate is significant. However, especially if the lenses are thick, the patient's use of spectacles may obscure the pupil to the examiner.

In dim illumination, the pupil's shape (degree of roundness) is noted, its size is measured, and both are recorded. A "pupil gauge" (a printed card with full- or half-circles of given sizes, usually in 1-mm increments) is helpful, but a simple ruler may also be used. A dimmest-visible slit-beam directed from 45° degrees temporally can also be used to measure the pupil at the on the slit-lamp or examiner. Alternatively, the pupil can be measured in dark conditions using a quantitative (scaled) infra-red pupillometer; this device has become much more readily available in recent years because of the need to assess maximum pupil dilation at night in patients considering refractive corneal surgery.

Bright-light stimulus is then applied to one eye, and the pupil response of that eye (*direct response*) is observed. The final size of the pupil in response to light and the speed or briskness of that response is recorded. The normal pupillary light reaction is a brisk, uniform concentric constriction; when the light stimulus is removed, an equally brisk redilation is seen.

When light stimulation is prolonged, normal constriction is followed after about a second by minimal redilation. In some patients, cycles of small-amplitude redilation and reconstriction are seen, and are termed *hippus*. Hippus can be quantified clinically: it can often be induced or emphasized with special lighting conditions ("edge lighting" at the slit-lamp), so that the frequency of the redilation/reconstruction cycles can be measured to give a "pupil cycle time" value. A prolonged pupil cycle time may suggest disease of the optic nerve or the pupilloconstrictive neural efferents. Light stimulus is then applied to one eye while assessing the *indirect pupillary response* in the fellow eye. In healthy individuals, the indirect pupillary response should be clinically equivalent to the direct response. However, in many clinical settings, actual observation of an unilluminated pupil in a dim room is impractical. Instead, the *swinging flashlight test* is generally utilized to better document the indirect response by comparing it to the same eye's direct response. The swinging flashlight test begins with the light directed to one eye; the direct response is observed. The flashlight is then quickly swung over to the other eye. Normally, the second pupil begins to dilate during the short time that neither is illuminated, but once it is directly stimulated a slight, brisk contraction is seen. If a large contraction or a continued dilation is seen after the flashlight has been swung, a relative afferent pupillary defect is suggested (see below). Next, the reaction to near is checked; often in the clinical setting, the near reaction is only checked if the light reaction is found to be abnormal. The patient is asked to shift visual attention from the far fixation target to a minimally illuminated near fixation target, perhaps 6–10 inches away. The normal reaction of the pupil to near stimulus is a brisk, uniform constriction that may be of slightly greater amplitude than the light response. When gaze is redirected to the distant target, brisk redilation is normally observed. It should be noted that although the light reaction is involuntary, the near reaction requires voluntary triggering of the near triad and so is dependent on patient alertness, attention, and cooperation. Notation of the complete clinical pupillary exam then consists of a description of the shape and measurement of the size of the pupil in dim lighting with distance fixation; speed of the reaction (both constriction and redilation) and final size of the light-stimulated pupil; presence (and severity) or absence of a RAPD; and reaction speed and final size of the near-stimulated pupil, especially if the light reaction is abnormal in any way.

Control materials to the preparatory stage of the class:
Questions (right answer in bold):

The homonymous hemianopia may be: a) bitemporal; b) binasal; **c) right side; d) left side;** e) all above mentioned

Visual hallucinations can be the signs of lesions of: a) the retina; b) the optic nerve; c) corpus callosum; **d) the occipital cortex**

Chiasm lesion produces: a) homonymous hemianopia; **b) heteronymous hemianopia**

Can you test olfactory system with ammonia?: a)Yes; **b) No**; c) I don't know

Optic hallucination is caused by: a) lesion of temporal lobe; **b) lesion of occipital lobe;** c) temporal lobe; d) parietal lobe

Trochlear nerve innervates: a) m. levator palpebrae superioris; b) m. rectus lateralis; c) m. superior rectus; **d) m. oblique superior;** e) all above mentioned

If the lesion of the third nerve occurs, the eyeball is turned: a) medially; d) laterally; c) upward; **d) laterally and downward**

A 72-year-old woman presents with the acute onset of double vision.

The second image disappears if she covers either eye. Which of the following ocular motor nerves is most likely to be impaired in this patient?

a. Oculomotor
b. Trochlear
c. Abducens
d. Ciliary
e. Müller's

A 33-years-old patient complains of double vision when gazing on the left and straight. During examination ptosis on the right, mydriasis were detected. Accommodation response and light reaction of the right pupil are absent. Strabismus divergens. Movement of the eyeball downward and outward on the right side is limited. Which cranial nerve is affected?

Answer: **oculomotor.**

Questions (right answer in bold):

The olfactory cortex in humans is located in which of the following locations?
a. Anterior perforated substance
b. Lateral olfactory gyrus (prepiriform area)
c. Posterior third of the first temporal gyrus
d. Angular gyrus
e. Calcarine cortex

Injuries to the macula or fovea centralis typically affect vision by producing which of the following?
a. Bitemporal hemianopsia
b. Nyctalopia (night blindness)
c. Scintillating scotomas
d. Mild loss of visual acuity
e. Severe loss of visual acuity

A 19-year-old man is hit in the face with a lead pipe. The ocular motor muscle most likely to be injured in this case is that innervated by which of the following?
a. Superior division of the third cranial nerve
b. Inferior division of the third cranial nerve
c. Fourth (trochlear) cranial nerve

d. Sixth (abducens) cranial nerve
e. Long ciliary nerve

A 17-year-old girl develops a painful vesicular rash around her left eye. This is followed by blurry vision that occurs only when both eyes are open. She is diagnosed with varicella zoster ophthalmicus. Which ocular motor nerve is most likely to be affected?
a. Superior division of the third
b. Inferior division of the third
c. Fourth (trochlear)
d. Sixth (abducens)
e. Long ciliary

A 32-year-old woman has an MRI done because of a first seizure. No etiology for the seizure is found, but there is the incidental finding of an aneurysm. The aneurysm is 5 mm and affects the posterior communicating artery. It is very close to the third cranial nerve. The initial sign of pressure on the third nerve is usually which of the following?
a. Impaired adduction
b. Impaired abduction
c. Impaired depression
d. Impaired elevation
e. Impaired pupillary constriction

A 58-year-old man with type 2 diabetes presents with the acute onset of double vision. Examination reveals a deficit of the third cranial nerve. A third-nerve palsy associated with diabetes mellitus is usually characterized by which of the following?
a. Poor pupillodilation
b. Poor pupilloconstriction
c. Sparing of pupillary function
d. Inversion of the affected eye
e. Upward deviation of the affected eye

A young man with multiple sclerosis (MS) exhibits paradoxical dilation of the right pupil when a flashlight is redirected from the left eye into the right eye. Swinging the flashlight back to the left eye produces constriction of the right pupil. Which of the following is the most likely diagnosis?
a. Early cataract formation in the right eye
b. Occipital lobe damage on the left
c. Oscillopsia
d. Hippus
e. Optic atrophy

The fifth nerve is a mixed sensory and motor nerve. It conducts sensory impulses from the greater part of the face and head; from the mucous membranes of the nose, mouth, and paranasal sinuses; and from the cornea and conjunctiva. It also innervates the dura of the anterior and middle cranial fossae. The cell bodies of the sensory part of the nerve lie in the gasserian, or semilunar, ganglion. This, the largest sensory ganglion in humans, lies in the medial part of the middle cranial fossa at the base of the cranium. The central axons of the ganglion cells form the sensory root. These fibers, on entering the mid pons, divide into short ascending and long descending branches. The former are concerned mainly with tactile and light pressure sense and synapse with second-order neurons in the principal sensory nucleus. Proprioceptive afferents from facial muscles and the masseter terminate in the mesencephalic nucleus. The fibers that mediate pain and temperature

sensation do not end in these nuclei but form the unique anatomy of the long descending branches of the spinal trigeminal tract. The latter pathway, which contains both facilitatory and inhibitory fibers, together with its nucleus, extends from the junction of the pons and medulla to the uppermost segments (C2 or C3) of the spinal cord (as evidenced by the relief of facial pain after medullary trigeminal tractotomy). The spinal nucleus is a continuation of the spinal tract of Lissauer and substantia gelatinosa; the main sensory nucleus is a continuation of the nucleus of the medial lemniscus. From all parts of the principal trigeminal sensory and spinal nuclei, second order fibers cross to the opposite side and ascend to the thalamus. They come to lie in the most medial part of the spinothalamic tract and lateral part of the medial lemniscus. These systems of fibers are called the trigeminothalamic tract. In addition, the secondary trigeminal neurons project to the facial and hypoglossal nuclei bilaterally, the saliva- tory nuclei, the cuneate nuclei of the upper cervical segments, and other cranial nerve nuclei. The main sensory and spinal trigeminal nuclei receive fibers from the reticular formation, the thalamus, the nucleus solitarius, and the sensory cortex. The peripheral branches of the gasserian ganglion form the three sensory divisions of the nerve. The first (ophthalmic) division passes through the superior orbital fissure; the second (maxillary) division leaves the middle fossa through the foramen rotundum; and the third (mandibular), through the foramen ovale.

The motor portion of the fifth nerve, which supplies the masseter and pterygoid muscles, has its origin in the trigeminal motor nucleus in the midpons; the exiting fibers pass under neath the gasserian ganglion and become incorporated into the mandibular nerve. The masseter and pterygoid muscles are utilized in chewing and are implicated in a number of brainstem reflexes, the best known of which is the jaw jerk. Tapping the chin with the jaw muscles relaxed stimulates proprioceptive afferents that terminate in the mesencephalic nucleus of the midbrain, which sends collaterals to the motor nucleus of the fifth nerve and causes the masseters to contract. This reflex is enhanced in spastic bulbar (pseudobulbar) palsy. Another pontine reflex that utilizes afferent trigeminal sensory nerves is the blink reflex. Tapping of the brow or bridge of the nose evokes bilateral blink through activation of the orbicularis oculi muscles (facial nerve efferents). Touching the eyelids and cornea (corneal reflex) does the same. Because of their wide anatomic distribution, complete interruption of both the motor and sensory fibers of the trigeminal nerve is rarely observed. In contrast, partial affection of the trigeminal nerve, particularly of the sensory part, is not uncommon, the main symptoms being numbness and pain. The various cranial nerve and brainstem syndromes in which the fifth nerve is involved.

Methods of examination of the trigeminal nerve.

To determine precisely which portion of the trigeminal nerve complex is affected, the examiner needs to initially test touch, temperature, and pain sensation within the distribution of each of the three major divisions.

Examining the corneal reflex provides another useful clinical tool. Application of a wisp of cotton to the cornea normally leads to an eye blink, provided the facial nerve is intact, permitting a blink to occur when the sensory portion of this reflex arc is preserved. When there is a significantly asymmetrical corneal response or actual unilateral loss of this reflex, there is evidence of ophthalmic division CN-V neuropathy.

The muscles of mastication are the temporal, masseter and pterygoids. The pterygoids move the jaws from side to side while chewing and the masseter and temporal muscles clench the jaws. Placing the fingers lightly on the masseter muscle and then asking the patient to bite down tests the latency of contraction and its force. As the jaws are opened they are pushed forward. The opposing pterygoids are balanced. If one is weak, the jaw deviates to the side of the weak muscle. If the lesion is supranuclear (corticobulbar fibers are lesioned), the jaw will deviate to the opposite side. The symmetry of the temporal fossa and the angles of the jaw should be noted. The examiner places his or her hand against the side of the jaw and instructs the patient to push against it. Ipsilateral weakness or deviation occurs from a contralateral supranuclear lesion or an ipsilateral nuclear lesion. The ability to swallow without choking is accomplished by the tensor veli palatini

muscle which closes off the nasopharynx. Lower motor pathologies such as motor neuron disease or tumor are associated with hollowing at the temple and flattening of the angle of the jaw.

Masticatory weakness is often very difficult to test when the change is subtle. Normally both lateral pterygoid muscles pull the jaw anteriorly or forward. When there is a unilateral motor fifth nerve lesion, the healthy unopposed pterygoid pulls the jaw across the midlines, thus leading to a deviation of the jaw to the side of the paretic motor fifth. This occurs when the motor nucleus, root, or mandibular division of the fifth cranial nerve is damaged.

Jaw jerk: The jaw jerk is helpful in localization in the brainstem and, in conjunction with other reflexes, gives information in regard to the patient's reflex status. It is dependent on the mesencephalic tract of the fifth cranial nerve which mediates proprioceptive information from jaw muscles. The patient is instructed to open the jaw slightly and the examiner places a forefinger below the lower lip and gently taps it downward with the reflex hammer. There is a slightly palpable upward movement. It is frequently unobtainable in normal people.

Trigeminal Neuralgia

Trigeminal neuralgia (tic douloureux) is characterized by the sudden onset of excruciating, intense stabbing pain (during waking hours). Several brief attacks (<2 minutes each) generally occur in succession. The pain is almost always precipitated by a trigger stimulus or activity (e. g., chewing, speaking, swallowing, touching the face, cold air, tooth brushing, shaving) and is located in the distribution of one or two branches of the trigeminal nerve, usually V/2 and/or V/3. Involvement of V/1, all three branches, or both sides of the face is uncommon. The attacks may persist for weeks to months or may spontaneously remit for weeks, or even years, before another attack occurs. Trigeminal neuralgia in the V/3 distribution is often mistaken for odontogenic pain, sometimes resulting in unnecessary tooth extraction. Typical (idiopathic) trigeminal neuralgia must be distinguished from secondary forms of the syndrome.

Pathogenesis. Idiopathic trigeminal neuralgia much evidence points to microvascular compression of the trigeminal nerve root (usually by a branch of the superior cerebellar artery) where it enters the brain stem, leading to the development of ephapses or suppression of central inhibitory mechanisms. Symptomatic trigeminal neuralgia cerebellopontine angle tumors, multiple sclerosis, vascular malformations.

To determine precisely which portion of the trigeminal nerve complex is affected, the examiner needs to initially test touch, temperature, and pain sensation within the distribution of each of the three major divisions. Examining the corneal reflex provides another useful clinical tool. Application of a wisp of cotton to the cornea normally leads to an eye blink, provided the facial nerve is intact, permitting a blink to occur when the sensory portion of this reflex arc is preserved. When there is a significantly asymmetrical corneal response or actual unilateral loss of this reflex, there is evidence of ophthalmic division CN-V neuropathy.

Both general and special sensation to the tongue and palate are necessary for fully functional taste. Impairment of general sensation from the tongue and palate carried by CN-V may also cause taste disturbances, even though the special sensory fibers providing primary taste sensation are supplied by the facial and glossopharyngeal nerves. Masticatory weakness is often very difficult to test when the change is subtle. Normally both lateral pterygoid muscles pull the jaw anteriorly or forward. When there is a unilateral motor fifth nerve lesion, the healthy unopposed pterygoid pulls the jaw across the midlines, thus leading to a deviation of the jaw to the side of the paretic motor fifth. This occurs when the motor nucleus, root, or mandibular division of the fifth cranial nerve is damaged.

Differential Diagnosis

Facial pain, numbness, or both are the hallmarks of most CN-V lesions. **Facial trauma**, particularly secondary to vehicular accidents or rarely invasive **dental treatments**, accounts for the majority of trigeminal nerve injuries. CN-V divisions and branches are exposed to **trauma** especially from **fractures of facial bones** within the face and neck and other clinically invasive procedures, particularly from dental work. Typically, these injuries lead to anesthesia within the specific distribution of whichever trigeminal branch is compromised.

In the Western world, **Herpes zoster** is the most common infectious cause of a trigeminal neuropathy. **Herpes zoster ophthalmicus** occurs when latent varicella zoster virus infection within the trigeminal ganglion becomes reactivated, almost always affecting the ophthalmic division. However, very rarely, the maxillary or mandibular divisions may be primarily affected. Most patients present with a characteristic periorbital vesicular rash and severe neuralgic pain within the ophthalmic division. Similar to other herpes zoster syndromes, the pain may precede the eruption of cutaneous lesions. Permanent visual impairment is the most serious outcome of ophthalmic zoster infection. Antiviral agents such as acyclovir are the main therapy. Corticosteroid eyedrops are shown to decrease pain and hasten corneal healing and are sometimes prescribed by ophthalmologists if there are no other ocular or systemic diseases that might contraindicate their use. Rarely, an ipsilateral middle cerebral artery infarction may subsequently develop in these patients as the virus travels within the trigeminal nucleus and affects the adjacent intracavernous portion of the carotid artery or its branches with a granulomatous angiitis of the brain. CSF varicella zoster virus antibodies are frequently found, and antiviral treatment is felt to be helpful.

Worldwide, leprosy or **Hansen disease** is a more common cause of CN-V neuropathy. This primarily occurs in economically depressed countries. It generally affects the coolest areas of the skin. Thus, if sensory loss is confined to the tip of the nose or the pinna of the ear, Hansen disease is a primary consideration.

Trigeminal sensory neuropathy occurs when the trigeminal ganglion cell bodies are the primary pathologic target. Although the pathogenesis of this ganglionopathy is frequently enigmatic, an association with connective tissue disorders, particularly **scleroderma or Sjogren** syndrome, is recognized. Patients with these disorders typically have numbness that begins around the mouth and spreads slowly over months involving all CN-V divisions, unilaterally or bilaterally, with the ophthalmic division at times spared. In Sjogren syndrome, trigeminal neuropathy is common and is usually representative of a more widespread sensory ganglionopathy. Presumably, blood–brain barrier is more permeable to large molecules than is the blood–brain barrier elsewhere in the peripheral nervous system.

A specific medication reaction is another exceedingly uncommon cause of a trigeminal neuropathy. The two best-recognized offenders are the industrial solvent trichloroethylene and the biological tracer hydroxystilbamidine.

The possibility of a **metastatic neoplasm** infiltrating a fifth nerve branch must always be included in the differential diagnosis of a persistent, but not always severe, facial numbness or pain. This is very well illustrated in this chapter's vignette. Tumors involving the face, such as **squamous cell carcinoma**, microcystic adnexal (sweat gland) carcinoma, and keratoacanthoma, have a proclivity for invading cutaneous nerves because of their innate neurotropism.

Primary trigeminal neurinomas are rare, usually benign, well-demarcated, and slowly growing neoplasms. These comprise 0.2% of intracranial tumors and 2–3% of intracranial neurinomas. Most frequently, these tumors arise near the trigeminal ganglion, usually extending into the middle and posterior cranial fossae. Rarely, they arise exclusively from one of the sensory divisions and spread extracranially. Rare instances of malignant schwannomas originating within the trigeminal ganglion also occur. Most neurinomas have very slow growths, and reach a 2.5 cm diameter before a specific diagnosis is made. Initially, these patients typically report developing numbness and paresthesiae within the CN-V distribution. It is very rare for typical trigeminal neuralgia pain or even intermittent burning pain to be the presenting sign of a primary trigeminal tumor. Rarely, these sensory findings are accompanied by other neurologic symptoms resulting from damage to adjacent structures. For example, tumors growing downward into the posterior fossa may lead to cerebellar ataxia and lesions of CN-VII and CN-VIII, manifesting with facial palsy, tinnitus, or hearing loss.

In contrast, neurinomas exert pressure upward on the lateral wall of the cavernous sinus, leading to CN-II, -III, -IV, and -VI lesions.

Cerebellopontine angle tumors, typically acoustic neurinomas arising form CN-VIII, may enlarge and compress the trigeminal sensory root and lead to facial numbness or pain with

subsequent ipsilateral loss of the corneal reflex. Other neoplasms include meningiomas, epidermoids, lymphomas, hemangioblastomas, gangliocytomas, chondromas, and sarcomas. Similarly, certain skull base tumors, such as **nasopharyngeal carcinoma**, **salivary gland adenocarcinoma**, and **metastatic disease**, may invade various trigeminal divisions. The numb chin syndrome consists of unilateral numbness of the chin and adjacent lower lip. Although seemingly harmless, it is usually an ominous sign of primary or metastatic cancer involving the mandible, skull base, or leptomeninges. **Lymphoproliferative malignancies** and **metastatic breast cancer** are the most common etiologies.

Lastly one may find that some primary trigeminal neuropathies defy specific definition and thus are labeled as **idiopathic**. However, one must emphasize the need for a vigilant approach with frequent follow-up as illustrated in the above vignette, especially with patients with prior known facial malignancies such as SCC or other masses.

Cases of paroxysmal pain behind the eye or nose or in the upper jaw or temple—associated with blocking of the nostril or lacrimation and described under the titles of **sphenopalatine (Sluder), petrosal, vidian, and ciliary neuralgia (Charlin or Harris, "lower half" headache)—probably represent variants of cluster headache.** A similar head pain may occasionally be confined to the lower facial, postauricular, or occipital areas. Ekbom distinguished yet another "lower cluster headache" syndrome with infraorbital radiation of the pain, an ipsilateral partial Horner syndrome, and ipsilateral hyperhydrosis. There is no evidence to support the separation of these neuralgias as distinct entities.

Chronic paroxysmal hemicrania is the name given by Sjaastad and Dale to a unilateral form of headache that resembles cluster headache in some respects but has several distinctive features. Like cluster headaches, these are of short duration (2 to 45 min) and usually affect the temporo-orbital region of one side, accompanied by conjunctival hyperemia, rhinorrhea, and in some cases a partial Horner syndrome. The acronym SUNCT has been applied to the condition (short-lasting unilateral neuralgiform attacks with conjunctival injection and tearing). Unlike cluster headache, however, the paroxysms occur many times each day, recur daily for long periods (the patient of Price and Posner had an average of 16 attacks daily for more than 40 years), and, most importantly, respond dramatically to the administration of indomethacin, 25 to 50 mg tid. Unlike the usual form of cluster headache, chronic paroxysmal hemicrania is more common in women than in men, in a ratio of 3:1.

Geniculate neuralgia.

Geniculate neuralgia - is episodes of severe lancinating pain occurring in the region of the pinna and external auditory canal.

The etiology of this condition is unknown. Neuralgia affecting the nervus intermedius (the bipolar neurons of the n. intermedius are located in the geniculate ganglion and the afferent axons enter the spinal tract of the trigeminal nerve. The peripheral fibers are distributed to the external auditory canal and the pinna. There may also be some distribution to deeper structures of the face and hard palate.

Clinical features. Typical spasmodic attacks of severe pain in the region of the pinna and external auditory canal. The pain is occasionally felt in the throat, deep in the face and in the orbit. Treatment same as for trigeminal neuralgia. Surgical excision of the geniculate ganglion has been performed in some cases.

Facial sympathalgias is the group of the similar clinical states. As a rule they have paroxysmal courses and normal state between attacks. Attacks last from ten minutes to days (rarely). Typical sign is the acute, often unendurable pain in face, sometimes pulsating character.

The important clinical sign are autonomic signs on the side of pain: lacrymation, conjunctival injection, rhinorrhoea from one half of nose and nasal obstruction, forehead and facial sweating. Syndrome meets more frequent among men. Attacks happen up acutely, mainly at nights, the sharpest pain does make a patient to move, because at rest pain becomes yet sharper.

Facial sympathalgias are expression two on principle different forms of pathology:
Sympathalgic syndromes (lesion of autonomic peripheral ganglions and nerves)
1) Nasal-ciliary neuralgia – Charlen syndrome.
2) Pterygopalatal neuralgia – Sluder syndrome.
3) Neuralgia of large petrosal superficial nerve – Gartner syndrome.

Vascular syndromes, as migraine:
1) Cluster headache.
2) Cluster-effect (Horton histamine migraine, Garris migrainous neuralgia).
3) Glyazer Carotid syndrome.

Sympathalgic syndromes, caused the lesion of autonomic peripheral ganglions and nerves.

Sympathalgic syndromes (Charlen and Sluder) are caused by pathological process of peripheral autonomic ganglions or their irritation. The etiology is not enough clear. Herpetic rashes in case of Charlen syndrome tells about herpetic ganglionitis of nasal-ciliary ganglion. Etiology of pterygopalatine sympathalgy is nasal sinus infection (in particular, in maxillar cavity) and lesion of ganglion pterygopalatine.

Charlen syndrome (Ciliary neuralgia). Typical attack of one-sided pain in internal corner of eye and base of the nose, in an eyeball, irradiation in a nose, rhinorrhoea and nasal obstruction, conjunctival injection, blepharospasm. Pain is typical in internal corner of eye. Usually an attack is associated with acute lacrymation, changes in the front parts of eyeball, cornea (phenomena of keratitis or iritis), herpetic exanthema at skin of the nose

The Charlen syndrome should be differentiated with herpetic ganglionitis of trigeminal ganglion. It manifests as symptoms in zone of 1 branche of trigeminal nerve innervation. Bright autonomic supplement is not typical.

Sluder syndrome (neuralgia of ganglion pterygopalatal). Pain is localized in base of the nose, maxilla, teeth, tongue, soft palate, ear, neck-shoulder area, then with irradiation in an eye and temple. Sometimes there is reduction of muscles of soft palate (typical click). The attack of pains begins and finishes gradually. Pain of middle cruelty with out motor anxiety After an attack - paresthesias in face and noise in ear.

Differential diagnosis Typical signs of neuralgia: one-sided autonomic features, one-sided oedema of face in the attack period. Differential diseases: Kvinke angiotrophoneurotic oedema: typical localization in lips, cheeks, bilateral oedema does not cause diagnostic difficulties. Recurrence of Facial neuropathy in combination with a plicate tongue, heilitis is determined as disease of **Rossolimo-Melkerson-Rozental.**

Treatment. Application of autonomic tropic drugs (ganglioblockercs – Pyrroxanum) – uninnervation effect on the ganglion, antiepileptic drugs - carbamazepinum (Tegretol, Finlepsin), psychotropic medications - tranquilizers and antidepressants. Effectively in acute period greasing a cocaine or lidocaine of middle nasal cavity, Novocaine or Lidocaine blockade of autonomic ganglions.

Meige Syndrome. This condition is the result of dystonic stimulation mediated through the facial nerve. Occur in middle-aged or elderly individuals. The signs of orofacial mandibular dystonia and blepharospasm are characteristic. The changes, which resemble tardive dyskinesia are not induced by neuroleptic drugs and are unlike the open mouth dystonia of Bruegel syndrome, lower pontine lesions can be identified in some cases There may be temporary improvement with haloperidol, tetrabenazine, or intravenous Cogentin. Botulinum toxin injection into the affected facial muscles is the treatment of choice.

The Seventh, or Facial, Nerve. The seventh cranial nerve is mainly a motor nerve supplying all the muscles concerned with facial expression on one side. The sensory component is small (the nervus intermedius of Wrisberg); it conveys taste sensation from the anterior two thirds of the tongue and, variably, cutaneous sensation from the anterior wall of the external auditory canal. The taste fibers at first traverse the lingual nerve (a branch of the trigeminal mandibular) and then join the chorda tympani, which conveys taste sensation via the facial nerve to the nucleus of the tractus solitarius. Secretomotorfibers innervate the lacrimal gland through the greater superficial petrosal nerve and the sublingual and submaxillary glands through the chorda tympani. Several other anatomic facts are worth remembering. The motor nucleus of the seventh nerve lies ventral and lateral to the abducens nucleus, and the intrapontine fibers of the facial nerve partly encircle and pass ventrolaterally to the abducens nucleus before emerging from the pons, just lateral to the corticospinal tract. At their juxtaposition in the floor of the upper fourth ventricle, the sixth and seventh nerves may be affected simultaneously by a vascular or infiltrative lesion. The facial nerve enters the internal auditory meatus with the acoustic nerve and then bends sharply forward and downward around the anterior boundary of the vestibule of the inner ear. At this angle (genu) lies the sensory ganglion (named geniculate because of its proximity to the genu). The nerve continues in its own bony channel, the facial canal, within which, just distal to the geniculate ganglion, it provides a branch to the pterygopalatine ganglion, i.e., the greater superficial petrosal nerve; somewhat more distally, it gives off a small branch to the stapedius muscle and is joined by the chorda tympani. It makes its exit from the skull at the stylomastoid foramen, then passes through the parotid gland and subdivides into five branches that supply the facial muscles, the stylomastoid muscle, the platysma, and the posterior belly of the digastric muscle. A complete interruption of the facial nerve at the stylomastoid foramen paralyzes all muscles of facial expression. The corner of the mouth droops, the creases and skin folds are effaced, the forehead is unfurrowed, the palpebral fissure is widened, and the eye- lids will not close. Upon attempted closure of the lids, both eyes roll upward (Bell'sphenomenon), but the one on the paralyzed side remains visible. The lower lids ags also, and the punctum falls away from the conjunctiva, permitting tears to spill over the cheek. Food collects between the teeth and cheek, and saliva may dribble from the corner of the mouth. The patient complains of a heaviness or numbness and sometimes an aching pain in the face, but sensory loss can usually not be demonstrated. Taste, however, is intact because the lesion is beyond the site where the chorda tympani has separated from the main trunk of the facial nerve. If the lesion is in the facial canal above the junction with the chorda tympani but below the geniculate ganglion, all the preceding symptoms occur; in addition, taste is lost over the anterior two-thirds of the tongue on the same side. If the nerve to the stapedius muscle is involved, there is hyperacusis (painful sensitivity to loud sounds). With a stethoscope in the patient's ears, a tuning fork at the bell is louder on the side of the paralyzed stapedius muscle. If the geniculate ganglion or the motor root proximal to it is in- volved, lacrimation and salivation may be reduced. Lesions at this point may also affect the adjacent eighth nerve, causing deafness, tinnitus, or dizziness.

Methods of examination of the facial nerve

Inspection

1. The symmetry of the face.

2. The wrinkles of the forehead and the prominence or flatness of the nasolabial folds.

3. The symmetry of blinking; the eyeballs should turn up and out with each blink. This is a normal phenomenon but is not seen because of full closure of the lids if there is no weakness.

4. Spontaneous movements of the mouth. Note abnormal movements such as twitching, tremor, involuntary movements and myokymia. This is a vermicular (worm-like) undulating movement around the eye which may be associated with a pontine glioma or demyelinating disease. Hemifacial spasm and oculopalatal myoclonus (rhythmic movement of the platysma muscle in conjunction with the palate) are quite specific. In the former, there is an abnormal branch of the anterior inferior cerebellar artery impinging on the seventh nerve and in the latter a lesion

(usually vascular) in Mollaret's triangle. These are connections between the red nucleus, dentate and inferior olivary nucleus.

Motor functions

The examiner asks the patient to mimic him or her in baring the teeth. Note the symmetry of the movement with particular attention to the nasolabial folds. Ask the patient to open the mouth and note the symmetry of the nasolabial folds. The upper facial muscles are tested by asking the patient to close the eyes tightly and to resist the examiner opening them. The patient is then asked to frown, wrinkle the forehead and raise the eyebrows.

Weakness of facial muscles is apparent as asymmetry. The platysma is tested by having the patient bare the teeth and open the mouth simultaneously. Blowing out the cheeks and pursing the lips tests the lower facial muscles.

Examination of taste

The patient is asked to protrude the tongue to the side. The tip is gently held with a piece of gauze. The patient points to a card on which the four possible tastes are written. The side of the tongue is moistened with the test substances 2 cm from its tip. The patient swishes out the mouth with water between applications of the test substances which are sugar, salt, vinegar and quinine. A qualitative assessment of taste is now performed with electrical depolarization of taste buds (produces a metallic taste) in taste and smell centers.

Secretory functions.

Schirmer's test is performed by gently applying a strip of filter paper from the lower eyelids and measuring the length of moistening on each side after the patient is stimulated to tear by inhaling ammonia. This is helpful in Sjögren's syndrome. Saliva flow is rarely tested. If the examiner stimulates its flow by placing a spicy substance on the tongue, which is slightly raised, the examiner can compare submaxillary flow from side to side.

The facial nerve can be damaged at any level along its complex course. Paralysis of the facial musculature is the hallmark of seventh cranial nerve lesions no matter what the lesion's anatomic site. The clinical presence or absence of symptoms related to the various other components of the facial nerve is very important in identifying the lesion site.

The patient with a **peripheral seventh (facial) nerve** palsy in most instances, with the exception of an early very distal branch lesion within the parotid gland, loses function of the entire ipsilateral side of their face and cannot smile, close their eyelid (orbicularis oculi), or wrinkle (frontalis) their forehead.

When **intrapontine** lesions affect the facial motor nucleus per se, as well as its exiting fibers, involvement of neighboring brainstem structures is typically seen. The association of peripheral facial paralysis with ipsilateral conjugate gaze palsy (paramedian pontine reticular formation lesion), ipsilateral lateral rectus palsy (sixth cranial nerve lesion), or paresis of the opposite arm and leg (corticospinal tract lesion) usually indicates a pontine localization.

Extramedullary lesions affecting the seventh nerve as it enters its intracranial course primarily occur within the cerebellopontine (CP) angle. Most commonly these are benign, relatively large acoustic neuromas that initially involve the eighth nerve and later extend to produce a seventh-nerve lesion. Thus, diminished hearing, sometimes initially presenting with tinnitus, usually precedes the onset of this type of peripheral facial paresis (see Fig. 7-4). Occasionally, with very large CP angle tumors there is concomitant involvement of the ipsilateral fifth cranial nerve (trigeminal nerve V) with unilateral facial numbness or initially only loss of the corneal reflex.

A relatively **proximal pregeniculate, intracanicular facial nerve** lesion characteristically leads to diminished lacrimation from greater petrosal nerve involvement as well as hyperacusis (an increased sensitivity to sound that is particularly noticeable while using a telephone), due to associated stapedius muscle paresis. These lesions also lead to diminished salivation, absent or altered taste sensation for the anterior two thirds of the tongue, and affected somatic sensation for the external auditory canal.

When a facial nerve lesion is more distally situated, **between the geniculate ganglion and the stapedius nerve** all of the above findings occur, but lacrimation is spared as the greater petrosal nerve has already exited the geniculate ganglion. If damage occurs in the **facial canal**, involvement of the **stapedius nerve and the chorda tympani** leads to hyperacusis and impaired salivation and taste but no change in lacrimation. When the seventh-nerve lesion is **distal to the chorda tympani**, it is characterized by a pure ipsilateral facial weakness. Very rarely, a lesion of this type occurs after the facial nerve exits the skull through the stylomastoid foramen. On occasion, this can cause diagnostic difficulty early on as it may initially involve just individual motor branches, with limited weakness of individual facial muscles before a complete palsy develops. Facial trauma is the most common cause for acute pure motor CN-VII lesions; however, an insidious progressive course suggests that a parotid adenocarcinoma, as illustrated in the vignette on p. 98, is the most likely cause.

Idiopathic Facial Palsy (Bell Palsy)

This vignette describes a benign, idiopathic facial palsy. The lesion had a proximal location, denoted by the loss of total motor function on one side of the patient's face involving the frontalis, orbicularis oculi, and the lower facial muscles, as well as loss of stapedius muscle action, taste, and lacrimal gland function.

Bell palsy is one of the most common and distinctive entities in clinical neurology. Typically, patients present with acute unilateral partial weakness of all mimetic muscles that evolves over several hours to no more than a few days, at times to complete facial paralysis. Although Bell palsy is usually benign, its

dramatic appearance initially creates in many a major concern that they may have had a stroke and that permanent facial disfigurement will result.

Rare instances of direct examination of the facial nerve in the setting of Bell palsy have shown signs of edema with subsequent nerve compression within the facial canal with resultant ischemia and nerve fiber degeneration. There is evidence to support reactivation of latent herpes simplex or varicella-zoster virus infection arising within the geniculate ganglion as the cause in a large proportion of common idiopathic Bell palsy.

Clinical Presentation

In retrospect, a preceding dull ache behind the ipsilateral ear is a common initial sign. Patients usually first become aware of weakness per se when a family member points out facial asymmetry, or when the individual personally notes an inability to close an eye, or experiences difficulty holding saliva, food, and fluids in the affected side of the mouth. Less commonly, decreased taste, or hyperacusis, is the first symptom.

Facial asymmetry is unequivocally present; the affected frontalis is smooth and cannot be normally corrugated, whereas the angle of the mouth appears depressed even in repose. Inability to completely close the eyelids (lagophthalmos) results from orbicularis oculi weakness. The Bell phenomenon refers to the eyeball turning up without eyelid closure despite attempted contraction of the orbicularis oculi. Facial palsy accompanied by taste disturbances may help to distinguish whether the lesion is proximal or distal to the chorda tympani branch. For example, a pure motor lesion suggests a lesion at the distal part of the facial canal or within the parotid gland, whereas when all four primary functions are affected, an unusually proximal lesion is deduced.

Differential Diagnosis

The examiner needs to first differentiate between an upper, central, or a lower motor neuron peripheral facial paralysis. Patients with upper motor neuron paralysis primarily have lower facial weakness with an asymmetric smile or unilateral drooling, while the upper face is relatively spared. In peripheral facial palsy, all musculature innervated by CN-VII is affected.

Lyme disease is the primary identifiable infectious etiology that may present with an acute facial palsy; later on, a contralateral lesion may develop. Typically, there are other neurological signs such as headache or radiculitis and cerebrospinal fluid pleocytosis. In the uncommon circumstance of a Bell palsy associated with herpes zoster infection (Ramsay–Hunt syndrome),

facial paralysis often precedes the appearance of typical herpetic vesicles within the external auditory canal. Middle-ear infection can rarely damage the facial nerve as it travels through the petrous bone. In regions endemic to tuberculosis, facial nerve palsies in association with petrous bone or mastoid process infections have been described.

Bilateral sequential Bell palsies are the most common neurologic manifestation of sarcoidosis. Frequently, associated hypothalamic–pituitary axis dysfunction (particularly impotence in men) and other cranial neuropathies are also present. Simultaneous bilateral facial weakness is an initial presentation of

Guillain–Barre syndrome that is soon followed by the more classic rapidly progressive polyradiculoneuropathy. Leprosy may lead to bilateral facial nerve lesions but with a unique patchy distribution.

A slowly progressive evolution of a unilateral facial palsy most typically suggests the presence of a neoplasm. Pontine lesions, especially **brainstem gliomas**, are the most proximal cause for a peripheral facial weakness. These tumors usually present in conjunction with other signs such as a lateral rectus palsy. Extramedullary tumors originating near the brainstem are often associated with facial nerve lesions and other cranial neuropathies, as with eighth-nerve acoustic neuromas or other **cerebellopontine angle tumors**. When there is diffuse **leptomeningeal** involvement, such as with metastatic carcinoma or lymphoma, the facial nerves may be part of the initial clinical profile of infiltration with these malignancies. Eventually other and often multiple cranial nerves become involved, particularly the trigeminal, oculomotor, and optic nerves. As noted in vignette on p. 98, evolving, progressive, and purely motor facial palsies presenting with varying degrees of individual facial muscle involvement are classic for a **parotid malignancy**.

Treatment

Corticosteroids reduce the duration of paralysis and risk of permanent impairment. The typical regimen is 1 mg/kg oral

only if it can be initiated within the first 3 days. Treatment is continued for 5 days and then tapered by 10-mg decrements over each of the next 5 days. This leads to much earlier recovery presumably by decreasing nerve swelling within the tight facial canal and thus diminishing nerve injury. There is no consistent evidence that antiviral medications, such as acyclovir or valacyclovir, shorten the course or improve outcome in Bell palsy and are no longer part of the routine management of idiopathic facial palsy. Although occasionally advocated, there is insufficient evidence to suggest that surgical CN-VII decompression is effective.

During the period of facial paralysis with incomplete eye closure, great care is required to protect the exposed cornea, which is subject to trauma from simple things such as turning over in bed and dryness. Eye patching and artificial tears during the day and a lubricant eye gel at night are usually sufficient to prevent corneal abrasions.

Prognosis

The severity of the underlying facial nerve injury determines how quickly and completely recovery from Bell palsy occurs. The degree of injury ranges from mild, with pure demyelinating conduction block, to severe, with axon loss and resulting wallerian degeneration. Up to 90% of Bell palsy cases are caused by a demyelinating conduction block with little or no associated axon loss, and therefore, recovery is prompt, complete, and without synkinesis. The remaining patients have axonal damage with wallerian degeneration, and improvement requires regenerating axons to reinnervate paralyzed muscles, resulting in slow and incomplete recovery.

The recovery rate from Bell palsy follows two patterns: most patients begin to regain facial strength within 3 weeks after onset, but in some, the initiation of recovery is delayed for at least 3–6 months. The overall prognosis is good; most patients (80–85%) recover completely, but the rest may have various residual effects. These include synkinesis, residual weakness, tearing, or contracture. Synkinesis, the most frequent permanent sequel, clinically manifests as synchronized movement of different muscles that normally do not contract together. Typically, there is subtle eye closure with smiling, or a lip or chin twitch with blinking. Synkinesis occurs when there is a misdirection of regenerating axons into muscles that they originally did not innervate.

The Ramsay–Hunt syndrome, caused by reactivation of the varicella-zoster virus (VZV) within the geniculate ganglion, is the second most common cause of atraumatic facial palsy. Clinically, it is characterized by the triad of acute facial palsy, neuralgic pain, and eruption of herpetic vesicles within the external auditory canal, ipsilateral palate, and anterior two thirds of the tongue. The areas of pain and rash are appropriate to the general sensory innervation of the afferent facial nerve branches. The geniculate ganglion cell bodies host the latent varicella-zoster virus infection. The close proximity of the geniculate ganglion to the vestibulocochlear nerve in the bony facial canal explains the concomitant otologic symptoms such as tinnitus, vertigo, and hearing loss in some patients. The detection of VZV IgM antibody in blood and cerebrospinal fluid (CSF), or VZV DNA in CSF, saliva, or blood is often helpful in assigning a viral etiology.

The prognosis for Ramsay–Hunt syndrome is worse than that of idiopathic Bell palsy, with frequent complete paralysis,

Methods of examination of the vestibular-cochlear nerve

Examination of hearing.

Place a finger in the external auditory meatus opposite the ear to be tested and move it continuously to produce a masking sound. Ask the patient to repeat “26” or “68” whispered into the tested ear to test high tones and “42” or “100” to test low tones. The examiner must use the otoscope to be certain there is no pathology of the middle ear and eardrum that would block sound transmission.

Rinne’s test: the tuning fork is gently struck, mask the contralateral ear and hold it near the external auditory meatus of the tested ear. If the patient can hear it, move it to the mastoid bone. The second the patient can no longer hear it ask him or her to say “now.” Then hold it near the external auditory meatus and ask if he or she can still hear it. In a normal patient the sound is still audible. In deafness, from middle ear disease, the patient will not be able to hear the vibration. In incomplete nerve deafness both air and bone conduction are decreased, but air conduction may still be perceived.

Weber’s test: the vibrating tuning fork is placed in the middle of the forehead, and then ask the patient where he or she hears it. In nerve deafness, the patient perceives the sound in the normal ear, whereas in chronic middle ear disease, it is heard in the affected ear.

Assesment of vestibular functions

Disturbance of vestibular function causes falling, past pointing, vertigo and nystagmus. As with tests of auditory functions, the examiner can make an educated guess as to the side of the difficulty and the circumstances of its occurrence and thus narrow the diagnostic possibilities. The easiest way to evaluate vestibular function is to utilize the hands as for conjugate gaze centers. Flex the hands so that the fingers of the left hand are pointing to the right hand. Thus, if there is a lesion of the right vestibular system the left will be predominant and the patient will past point to the left, fall to the left and drift on pendular walking to the left. The cortex will correct for the eyes being driven to the left and the fast component of corrective nystagmus will be to the right.

Pendular walking. The patient is instructed to go to the corner of the room, close the eyes and walk in a straight line, back and forth. The examiner explains to the patient that he or she will be stopped before crashing into an object. The second the patient closes the eyes he or she broadens their stance. During the maneuver the patient drifts to the side of the lesion as the opposite vestibular complex predominates.

Past pointing. The patient is seated in front of the examiner and asked to close the hand except for the index finger. The patient is instructed to raise and lower the finger to touch the outstretched finger of the examiner. The patient is then instructed to perform the maneuver rapidly. The patient will past point with both hands to the side of the lesion. If the patient has a cerebellar lesion, he or she will past point with the hand on the side of the cerebellar lesion while the other hand remains on target. The examiner will not see dramatic changes in chronic disease.

Acutely, the patient is often too ill to perform the maneuver. Patients with vestibular disease feel as if they are pushed to the affected side. Patients with cerebellar lesions fall to the affected side as they are clumsy with the affected foot. Utricular and saccular disease may cause the patient to be pushed in a linear direction. Vestibular sensations of dramatic displacement are termed pulsions. Thus, a patient who has acute loss of vestibular function on the left side may feel as if he or she is pushed to the left and would have lateral pulsion. This is not uncommon in a posterior inferior cerebellar artery stroke.

Caloric tests

This test is used in comatose patients. Because there are no corrective responses (the cortex is non-functional), the eyes will deviate to the side of the lesion. The patient is placed 30° above horizontal. Cold water at 30° (100 mL; some say 1 mL) is irrigated through a soft rubber catheter for 40 seconds into the right external auditory meatus. The eyes deviate to the side of the irrigated ear. If the right vestibular system is working, the eyes stay deviated for 30 seconds to 1 minute and then return to the midline. The test is repeated in the other ear and if the eyes conjugately deviate, the vestibular system, the extraocular muscles and the MLF are intact. It is best to think of the cold water as shutting off the stimulated vestibular system. In fact, raising the head 30° from the horizontal position places the ampulla that will be stimulated in the vertical plane with the ampulla being at the highest point. Warm water will cause the endolymph to rise and stimulate the canal to drive the eyes to the right. Cold water will cause the endolymph to flow in the downward direction, which would generate less stimulation to the ipsilateral ear and the eyes would be driven to the side of the infused cold water stimulation because the endolymph would flow away from the cold stimulated ear. It is easier to think of the test as cold water shutting down the irrigated ear. Canal paresis is the term used to describe an absent or diminished caloric test. This is usually caused by a lesion of the labyrinth such as Méniére's disease, an acoustic tumor, vestibular neuronitis or autoimmune etiologies. MRI with gadolinium enhancement demonstrates tumors (often the specific type) and inflammatory conditions. Thin cut CT radiography demonstrates bony erosion and fractures. It is rare to need caloric testing in the modern age of neuroimaging. Magnetic resonance angiography can demonstrate an aberrant branch of the anterior inferior cerebellar artery (brought to neurologic attention by Dr. Janetta’s observations of its occurrence with tic douloureux), which impinges upon the eighth nerve and diminishes its function.

Positional vertigo and nystagmus

The patient lies on the examination table with the shoulders at the cephalad edge of the table. The examiner lowers the head 30° and turns it to the side. Patients with positional vertigo will develop vertigo, usually within 10–15 seconds, associated with nystagmus, the fast component of which beats toward the down ear. Adaptation occurs and the symptoms and signs cannot be reproduced for 10–15 minutes. This may be because of utricular lesions although it has recently been reported from posterior semicircular canal disease. It is seen from degeneration of the cupula (cupulolithiasis) vascular lesions and head trauma. It occurs most frequently when arising from sleep.

Central positional vertigo is most often caused by a tumor of the posterior fossa and is characterized by:

1 no latency;

2 no adaptation;

3 the nystagmus appears immediately when the former provoking position is resumed.

The nystagmus will change direction with different head positions. Patients with positional nystagmus from central lesions often have positional vertigo with movement, but it is more prolonged and severe. It usually is associated with other long tract or cerebellar signs.

Control materials to the preparatory stage of the class:

Questions (right answer in bold):

What are the signs of peripheral paralysis of facial nerve?: **a) peripheral flaccid paralysis of facial expression muscles;** b) peripheral flaccid paralysis of eyeball muscles; c) peripheral flaccid paralysis of chewing muscles d) disorders of sensation on the face
What nerve innervates the expressive musculature of the face?: **a) VII**; b) V; c) IX; d) XII; e) XI

A 44-years-old male is detected: expression muscles palsy on the right: down turning mouth, nasolabial fold is smoothed, palpebral fissure is dilated. Patient cannot knit one's brow on this side; Bell's symptome is positive. There is lacrimation from the right eye. Hearing and sense of taste are preserved. Ground the topical diagnosis.
Answer: **lesion of the right facial nerve at the level of the foramen stylomastoideum**

A 48-year-old left-handed man develops increased sensitivity to sound in his left ear. A brain MRI reveals a posterior fossa mass. This symptom may develop in one ear with damage to which of the following ipsilateral cranial nerves?
a. V
b. VII
c. VIII
d. IX
e. X

Glossopharyngeal nerve (IX)

The glossopharyngeal nerve is part of the group of cranial nerves responsible for innervation of structures derived from the branchial arches. This nerve innervates structures related to the third branchial arch. It is also part of a group together with the vagus and accessory nerves that passes through the jugular foramen which is termed the vagus group. The glossopharyngeal nerve has cell bodies that are referred to as nucleus ambiguus. The glossopharyngeal nerve originates from the medulla oblongata and has several branches including the pharyngeal nerve, the lingual nerve and the tympanic branches. The glossopharyngeal nerve is composed of many fibre types including general somatic efferent fibres that innervate the stylopharyngeus muscle; the general visceral afferent fibres that provide sensory information from the carotid body, the pharynx and the middle ear; the general visceral efferent fibres that provide parasympathetic innervation to the parotid and zygomatic salivary glands; the special visceral afferent fibres that provide taste caudal to the tongue and finally the general somatic afferent fibres that provide sensory information from the external ear. The lingual branch of the glossopharyngeal nerve provides general somatic afferent fibres and special visceral afferent fibres to the caudal 1/3 of the tongue. On clinical examination, choking or dysphagia as a result of malfunctioning or absent pharyngeal reflexes would indicate a problem with the glossopharyngeal nerve.

Vagus nerve (X)

The vagus nerve is part of the group of cranial nerves responsible for innervation of structures derived from the branchial arches. It is also part of a group together with the glossopharyngeal and accessory nerves that passes through the jugular foramen which is termed the vagus group. The vagus nerve innervates structures related to the fourth branchial arch. The vagus nerve has cell bodies that are referred to as nucleus ambiguus. The vagus nerve is composed of many different types of nerve fibre including general somatic efferent fibres supplying motor function to the muscles of the larynx, pharynx, palate and oesophagus; general visceral afferent fibres to the base of the tongue, pharynx and larynx; general visceral efferent fibres for parasympathetic supply of the thoracic and abdominal viscera; special visceral afferent fibres supplying taste to regions of the epiglottis and palate and finally general somatic afferent fibres to the external ear and the dura mater. The vagus nerve also supplies general somatic afferent fibres and special visceral afferent fibres to the root of the tongue. There are many functional components of the vagus nerve including the heart, larynx, pharynx and many other viscera. On clinical

examination any changes related to gag reflexes, blood pressure or heart rate, changes in 'voice' or inspiratory dyspnoea may indicate a problem with the vagus nerve.

Accessory nerve (XI)

The accessory nerve is part of the group of cranial nerves responsible for innervation of structures derived from the branchial arches. It is also part of a group together with the glossopharyngeal and vagus nerves that passes through the jugular foramen which is termed the vagus group. The accessory nerve supplies structures related to the fourth branchial arch. The accessory nerve has cell bodies that are referred to as nucleus ambiguus and originate in the medulla oblongata. The cranial root of the accessory nerve actually contributes to the vagus nerve and to the striated muscles of the pharynx, larynx, palate and oesophagus.

However, the accessory nerve also contributes to the cervical spinal cord and spinal root through the foramen magnum providing innervation to muscles of the neck. The spinal root of the accessory nerve branches into the dorsal branch and the ventral branch. The dorsal branch innervates the brachiocephalicus, trapezius and omotransversarius muscles of the dorsal neck. The ventral branch innervates the sternocephalicus muscle.

During clinical examination any difficulties in turning the neck or muscle atrophy around the dorsal and ventral neck may indicate a problem with the accessory nerve.

Hypoglossal nerve (XII)

The hypoglossal nerve is part of the group of cranial nerves responsible for the control of muscles of the head. It is in part a cervical nerve due to its caudal position on the brain stem. The nerve is composed of general somatic efferent fibres which control the intrinsic and extrinsic muscles of the tongue (together with other nerves including the lingual nerve, facial nerve, lingual branch of the glossopharyngeal nerve and the vagus nerve). The nucleus of the nerve is located within the medulla oblongata of the brain stem and it passes through the hypoglossal canal.

Bulbar palsy. When there is bilateral impairment of function in the 9th, 10th and 12th cranial nerves, the clinical syndrome of bulbar palsy evolves. The features of bulbar palsy are:

dysarthria;
dysphagia, often with choking episodes and/or nasal regurgi- tation of fluids;
dysphonia and poor cough, because of weak vocal cords;
susceptibility to aspiration pneumonia.

Dysarthria The speech disturbance in patients with dysarthria is a purely mechanical one caused by defective movement of the lips, tongue, palate, pharynx and larynx. Clear pronunciation of words is impaired due to the presence of a neuromuscular lesion.

Dysphagia, or difficulty swallowing, can result from many causes, including neurologic disorders, both peripheral and central; viral, bacterial, or fungal infections of the upper airway; surgeries or disease processes that directly involve the oral, pharyngeal, or laryngeal structures; and psychogenic mechanisms. Aging and medications may exacerbate dysphagia. A number of antidepressant medications cause xerostomia (reduced salivary flow), affecting bolus formation. Sedative medications may significantly affect swallowing by reducing oropharyngeal coordi nation for bolus formation and airway protection. Common signs of dysphagia include pocketing of food in the oral cavity, drooling, wet vocal quality during meals, episodes of coughing and throat clearing, and shortness of breath during meals. Aspiration, the primary concern when dealing with dysphagia, is technically defined as the entry of a foreign substance below the level of the vocal cords into the trachea. Aspiration is a dangerous precursor of aspiration pneumonia. Risk factors include chronic obstructive pulmonary disorders, congestive heart failure, feeding tubes, dependence for oral care and feeding, decreased laryngopharyngeal sensation, medications,

and a reduced level of alertness. Any patient with aspiration risk needs to be placed on aspiration precautions and referred for swallowing evaluation before oral intake is initiated.

Upper motor neurone lesions. The upper motor neurones involved in speech have their cell bodies at the lower end of the precentral (motor) gyrus in each cerebral hemisphere. From the motor cortex, the axons of these cells descend via the internal capsule to the contralateral cranial nerve nuclei 5, 7, 9, 10 and 12. A unilateral lesion does not usually produce a major problem of speech pronunciation. There is some slurring of speech due to facial weakness in the presence of a hemiparesis. Bilateral upper motor neurone lesions, on the other hand, nearly always produce a significant speech disturbance. Weakness of the muscles supplied by cranial nerves 5–12 is known as bulbar palsy if the lesion is lower motor neurone in type. It is known as pseudobulbar palsy if the weakness is upper motor neurone in type. Patients who have bilateral upper motor neurone weakness of their lips, jaw, tongue, palate, pharynx and larynx, i.e. patients with pseudo- bulbar palsy, have a characteristic speech disturbance, known as a spastic dysarthria. The speech is slow, indistinct, laboured and stiff. Muscle wasting is not present, the jaw-jerk is in- creased, and there may be associated emotional lability. The patient is likely to be suffering from bilateral cerebral hemisphere cerebrovascular disease, motor neurone disease or serious multiple sclerosis.

Methods of detection of bulbar and pseudobulbar disorders

Initial observation. The examiner notes the pitch and quality of the patient's voice, spontaneous cough and the ability to swallow. Regurgitation of fluids is sought by history. A high-pitched hoarse voice suggests vocal cord paralysis, whereas a nasal tone that increases with forward flexion is characteristic of palate weakness. This improves with the head extended. If the patient chokes on saliva while speaking this is characteristic of both palatal and pharyngeal weakness.

Motor function. The examiner asks the patient to open the mouth wide. The examiner inspects the uvula and the tongue while it is at rest on the floor of the mouth. A fibrillating tongue with scalloping at its edges (twelfth nerve) is most often motor neuron disease. The patient is asked to say "Ah" while exhaling and "Ugh" while inhaling. The palate should elevate and move backwards, the uvula remains in the midline while the posterior pharyngeal muscles contract. A deep breath alone elevates the palate and is easy for the patient to perform.

Sensory functions. Touch sensation. A throat swab is used to stimulate the back of the throat while the tongue is gently depressed. The sensory component of the gag reflex is elicited from any part of the palate, tonsil or posterior part of the tongue that is touched and initiates contraction of the pharynx (posterior constrictor muscles) elevation of the palate and tongue retraction. The threshold of the reflex varies from patient to patient.

Taste sensation. Testing taste in the posterior tongue is now primarily carried out in taste and smell centers. It is rarely necessary as pathology never affects this component of the nerve alone.

On phonation. If there is lower motor neuron weakness of the vagus, the palate elevates and moves to the normal side. This pulling movement resembles cloth being pulled over a table. Bilateral absence of movement of the palate and pharynx is accompanied by dysphagia, nasal regurgitation and nasal speech. This is usually secondary to bilateral medullary lesions or a bilateral upper motor lesion from pseudobulbar palsy. ***Pseudobulbar palsy is accompanied by a hyperactive gag reflex in association with an abnormal affect (inappropriate crying or laughter).*** It is most commonly seen with severe vascular disease, demyelinating disease and head trauma. A combination of both upper and lower motor neuron disease may be seen in amyotrophic lateral sclerosis (ALS). Repeated phonation causes fatigueability in myasthenia gravis.

Sensory loss. Unilateral absence of the gag reflex is seen with isolated lesions of the ninth nerve that occur with schwannomas, glomus jugulare tumors, lymphoma or a surgical procedure. The normal side will trigger a full reflex. Phonation and direct inspection of the pharynx demonstrates unilateral muscle weakness as a cause of unilateral decrease of a gag reflex. A

combination of glossopharyngeal and vagus lesions causes the palate and posterior pharynx to pull to the normal side when stimulated. A bilateral motor and sensory deficit of the pharynx is most often secondary to a severe medullary vascular lesion or tumor.

An upper motor neuron lesion from vascular disease causes inability to elevate the palate and constrict the pharynx on the side contralateral to the lesion (the pharynx pulls and the palate elevates to the normal side).

Examining the vocal cords. This examination is required in any patient who is hoarse or whenever it is suspected that there is a neurologic lesion responsible for palatopharyngeal weakness. The examination is performed by an otolaryngologist. The movements of the vocal cord during inspiration, expiration and phonation (the patient saying "Ah") are recorded. The cords abduct during inspiration and adduct during phonation.

The hypoglossal nerve (twelfth) is a purely motor nerve and controls all movements of the tongue. It is also involved with specific movements of the hyoid bone and larynx during and following swallowing. Method of examination. The patient is instructed to open the mouth and the tongue is evaluated for bulk, shape, position and its surfaces. At complete rest, if it is denervated, fibrillations will be noted. These are the spontaneous contractions of single fibers. This is the only place in the body where there is no overlying subcutaneous tissue. Hyperthyroidism and parahyperthyroidism may also cause a fibrillating tongue. If the tongue is truly atrophic its edges will be scalloped. The patient is then asked to protrude the tongue in the midline. Difficulty in protruding the tongue, deviation from the midline and involuntary movements are noted. Myotonia of the tongue can be demonstrated by having the patient place the extruded tongue on a tongue blade and then place a vertical tongue blade on the tongue which is then tapped. The indentation that occurs from the myotonic contraction may last for several seconds.

Alternating syndrome:

Jackson syndrome. It's paralysis of structures innervated by the tenth, eleventh, and twelfth cranial nerves, including the soft palate, larynx, half of the tongue, and the sternomastoid and trapezius muscles. It's caused by occlusion of the vertebral artery. Its signs are: Paralysis of the soft palate and vocal cords on one side which cause dysphagia and dysphonia. Loss of pain sensation and temperature sense on the other side. Horner's syndrome may be associated.

Avellise syndrome. It usually results from occlusion of the vertebral artery. Its signs are: Paralysis of the soft palate and vocal cords on one side which cause dysphagia and dysphonia. Loss of pain sensation and temperature sense on the other side, including the extremities, trunk, and neck. Horner's syndrome may be associated. In the original description, the vagus and glossopharyngeal nerves were involved; concomitant involvement of the neighbouring cranial nerves was observed later.

Common conditions affecting 9th, 10th and 12th nerve function

Motor neurone disease. When motor neurone disease is causing loss of motor neurones from the lower cranial motor nuclei in the medulla, the bulbar palsy can eventually lead to extreme difficulty in speech (anarthria) and swallowing. Inanition and aspiration pneumonia are commonly responsible for such patients' deaths. The tongue is small, weak or immobile, and fasciculating.

Infarction of the lateral medulla Infarction of the lateral medulla, following posterior inferior cerebellar artery occlusion, is one of the most dramatic cerebrovascular syndromes to involve speech and swallowing. Ipsilateral trigeminal, vestibular, glossopharyngeal and vagal nuclei may be involved, along with cerebellar and spinothalamic fibre tracts in the lateral medulla.

Guillain–Barré syndrome Patients with Guillain–Barré syndrome, acute, post-infectious polyneuropathy, may need ventilation via an endotracheal tube or cuffed tracheostomy tube. This may be necessary either because of neuropathic weak- ness of the chest wall and diaphragm, or because of bulbar palsy secondary to lower cranial nerve involvement in the neuropathy.

Recurrent laryngeal nerve palsy The recurrent laryngeal nerves are vulnerable to damage in the neck and mediastinum, e.g. aortic aneurysm, malignant chest tumours, malignant glands and

surgery in the neck (especially in the region of the thyroid gland). A unilateral vocal cord palsy due to a unilateral nerve lesion produces little disability other than slight hoarseness. Bilateral vocal cord paralysis is much more disabling, with marked hoarseness of the voice, a weak 'bovine' cough (because the cords cannot be strongly adducted) and respiratory stridor.

Myasthenia gravis. Bulbar muscle involvement in myasthenia gravis is quite common in this rare condition. The fatiguability of muscle function, which typifies myasthenia, is frequently very noticeable in the patient's speech and swallowing.

Control materials to the preparatory stage of the class:

Questions (right answer in bold):

For bulbar syndrome it is typical: **a) dysarthria, dysphagia, hoarseness, nasal voice, diminished gag reflex;** b) dysphagia, dysarthria, emotional incontinence, crying (or laughing)

What is bulbar palsy? a) it's a syndrome of upper motor neuron paralysis that affects the corticobulbar system above the brain stem bilaterally; **b) it's a syndrome of lower motor neuron paralysis, affecting muscles innervated by cranial nerves (mainly IX-XII)**

A 32-years-old male patient complains of tongue deviation to the left and weakness of the right extremities. During examination tongue muscles atrophy and fibrillation on the left; hemiparesis with high muscle tone and Babinski sign on the right side were detected. Ground the topical diagnosis. Point the syndrome.

Answer: **Jackson syndrome**

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

Electronic information resources
1. Medical Books On-line Library (Neurology) – free download
http://medbookshelf.info/category/neurology/

Practical class No 7.

Theme: Neuropathy of the trigeminal nerve and its individual branches. Iatrogenic trigeminal neuropathy.

Actuality of theme. The early diagnosis of the affection of the trigeminal nerve and its individual branches can prevent the dangerous complications.

Particular goals:
a student must know:
1) Zone of innervation and function of trigeminal nerve.
2) Symptoms of lesion of trigeminal nerve.
3) Iatrogenic trigeminal neuropathy

to be able: to explore patients with the lesion of trigeminal nerve, to make topical diagnosis, to give an appropriate treatment.

Tasks for class

Common terms: Trigeminal neuralgia, acute zoster and postherpetic neuralgia, trigeminal neuropathy, trigeminal neuropathy

Theoretical questions:

1. Clinical classification of diseases of trigeminal nerve.
2. Pathogenesis, clinical course of diseases of the trigeminal nerve.
3. Treatment of diseases of trigeminal nerve.
4. Indications for surgery.

Practical skills:

Exam patients with diseases of the trigeminal nerve.
Give an appropriate treatment

Table of contents of the class:

The fifth nerve is a mixed sensory and motor nerve. It conducts sensory impulses from the greater part of the face and head; from the mucous membranes of the nose, mouth, and paranasal sinuses; and from the cornea and conjunctiva. It also innervates the dura of the anterior and middle cranial fossae. The cell bodies of the sensory part of the nerve lie in the gasserian, or semilunar, ganglion. This, the largest sensory ganglion in humans, lies in the medial part of the middle cranial fossa at the base of the cranium. The central axons of the ganglion cells form the sensory root. These fibers, on entering the mid pons, divide into short ascending and long descending branches. The former are concerned mainly with tactile and light pressure sense and synapse with second-order neurons in the principal sensory nucleus. Proprioceptive afferents from facial muscles and the masseter terminate in the mesencephalic nucleus. The fibers that mediate pain and temperature

sensation do not end in these nuclei but form the unique anatomy of the long descending branches of the spinal trigeminal tract. The latter pathway, which contains both facilitatory and inhibitory fibers, together with its nucleus, extends from the junction of the pons and medulla to the uppermost segments (C2 or C3) of the spinal cord (as evidenced by the relief of facial pain after medullary trigeminal tractotomy). The spinal nucleus is a continuation of the spinal tract of Lissauer and substantia gelatinosa; the main sensory nucleus is a continuation of the nucleus of the medial lemniscus. From all parts of the principal trigeminal sensory and spinal nuclei, second order fibers cross to the opposite side and ascend to the thalamus. They come to lie in the most medial part of the spinothalamic tract and lateral part of the medial lemniscus. These systems of fibers are called the trigeminothalamic tract. In addition, the secondary trigeminal neurons project to the facial and hypoglossal nuclei bilaterally, the saliva- tory nuclei, the cuneate nuclei of the upper cervical segments, and other cranial nerve nuclei. The main sensory and spinal trigeminal nuclei receive fibers from the reticular formation, the thalamus, the nucleus solitarius, and the sensory cortex. The peripheral branches of the gasserian ganglion form the three sensory divisions of the nerve. The first (ophthalmic) division passes through the superior orbital fissure; the second (maxillary) division leaves the middle fossa through the foramen rotundum; and the third (mandibular), through the foramen ovale.

The motor portion of the fifth nerve, which supplies the masseter and pterygoid muscles, has its origin in the trigeminal motor nucleus in the midpons; the exiting fibers pass under neath the gasserian ganglion and become incorporated into the mandibular nerve. The masseter and pterygoid muscles are utilized in chewing and are implicated in a number of brainstem reflexes, the best known of which is the jaw jerk. Tapping the chin with the jaw muscles relaxed stimulates proprioceptive afferents that terminate in the mesencephalic nucleus of the midbrain, which sends collaterals to the motor nucleus of the fifth nerve and causes the masseters to contract. This reflex is enhanced in spastic bulbar (pseudobulbar) palsy. Another pontine reflex that utilizes afferent trigeminal sensory nerves is the blink reflex. Tapping of the brow or bridge of the nose evokes bilateral blink through activation of the orbicularis oculi muscles (facial nerve efferents). Touching the eyelids and cornea (corneal reflex) does the same. Because of their wide anatomic distribution, complete interruption of both the motor and sensory fibers of the trigeminal nerve is rarely observed. In contrast, partial affection of the trigeminal nerve, particularly of the sensory part, is not uncommon, the main symptoms being numbness and pain. The various cranial nerve and brainstem syndromes in which the fifth nerve is involved.

Methods of examination of the trigeminal nerve.

To determine precisely which portion of the trigeminal nerve complex is affected, the examiner needs to initially test touch, temperature, and pain sensation within the distribution of each of the three major divisions.

Examining the corneal reflex provides another useful clinical tool. Application of a wisp of cotton to the cornea normally leads to an eye blink, provided the facial nerve is intact, permitting a blink to occur when the sensory portion of this reflex arc is preserved. When there is a significantly asymmetrical corneal response or actual unilateral loss of this reflex, there is evidence of ophthalmic division CN-V neuropathy.

The muscles of mastication are the temporal, masseter and pterygoids. The pterygoids move the jaws from side to side while chewing and the masseter and temporal muscles clench the jaws. Placing the fingers lightly on the masseter muscle and then asking the patient to bite down tests the latency of contraction and its force. As the jaws are opened they are pushed forward. The opposing pterygoids are balanced. If one is weak, the jaw deviates to the side of the weak muscle. If the lesion is supranuclear (corticobulbar fibers are lesioned), the jaw will deviate to the opposite side. The symmetry of the temporal fossa and the angles of the jaw should be noted. The examiner places his or her hand against the side of the jaw and instructs the patient to push against it. Ipsilateral weakness or deviation occurs from a contralateral supranuclear lesion or an ipsilateral nuclear lesion. The ability to swallow without choking is accomplished by the tensor veli palatini muscle which closes

off the nasopharynx. Lower motor pathologies such as motor neuron disease or tumor are associated with hollowing at the temple and flattening of the angle of the jaw.

Masticatory weakness is often very difficult to test when the change is subtle. Normally both lateral pterygoid muscles pull the jaw anteriorly or forward. When there is a unilateral motor fifth nerve lesion, the healthy unopposed pterygoid pulls the jaw across the midlines, thus leading to a deviation of the jaw to the side of the paretic motor fifth. This occurs when the motor nucleus, root, or mandibular division of the fifth cranial nerve is damaged.

Jaw jerk: The jaw jerk is helpful in localization in the brainstem and, in conjunction with other reflexes, gives information in regard to the patient's reflex status. It is dependent on the mesencephalic tract of the fifth cranial nerve which mediates proprioceptive information from jaw muscles. The patient is instructed to open the jaw slightly and the examiner places a forefinger below the lower lip and gently taps it downward with the reflex hammer. There is a slightly palpable upward movement. It is frequently unobtainable in normal people.

Trigeminal neuralgia (TN) is the most common type of facial pain neuralgia. The pain typically occurs in the distribution of one of the branches of the trigeminal nerve (cranial nerve V), usually on one side. Rarely, it can affect both sides, although simultaneous bilateral trigeminal neuralgia is uncommon. It involves both the mandibular and maxillary divisions of the trigeminal nerve in 35% of affected patients. Isolated involvement of the ophthalmic division is much less common (2.8% of TN cases).

According to Penman in 1968, the prevalence of TN is approximately 107 men and 200 women per 1 million people. The incidence is 4-5 cases per 100,000. The disease begins after age 40 in 90% of patients and is slightly more common in women. Rushton and Olafson found that approximately 1% of patients with multiple sclerosis (MS) develop TN, whereas Jensen et al stated that 2% of patients with TN have MS.

A lack of clear definitions for facial pain has hampered the understanding of trigeminal neuralgia. The condition has no clear natural history, and no long-term follow-up study of the progression of the disorder has ever been published. In an attempt to rationalize the language of facial pain, recently, a new classification scheme that divides facial pain into several distinct categories was introduced:

Trigeminal neuralgia type 1 (TN1): This is the classic form of trigeminal neuralgia in which episodic lancinating pain predominates. (also known as ―Typical TN).

Trigeminal neuralgia type 2 (TN2): This is the atypical form of trigeminal neuralgia in which more constant pains (aching, throbbing, burning) predominate. (also known as ―Atypical TN).

Trigeminal neuropathic pain (TNP): This is pain that results from incidental or accidental injury to the trigeminal nerve or the brain pathways of the trigeminal system.

Trigeminal deafferentation pain (TDP): This is pain that results from intentional injury to the system in an attempt to treat trigeminal neuralgia. Numbness of the face is a constant part of this syndrome, which has also been referred to as anesthesia dolorosa or one of its variants.

Symptomatic trigeminal neuralgia (STN): This is trigeminal neuralgia associated with multiple sclerosis (MS).

Postherpetic neuralgia (PHN): This is chronic facial pain that results from an outbreak of herpes zoster (shingles), usually in the ophthalmic division (V1) of the trigeminal nerve on the face and usually in elderly patients.

Geniculate neuralgia (GeN): This is typified by episodic lancinating pain felt deep in the ear.

Glossopharyngeal neuralgia (GPN): This is typified by pain in the tonsillar area or throat, usually triggered by talking or swallowing.

Differential diagnosis:

TN presents with multiple episodes of severe and spontaneous pain that usually lasts seconds to minutes. The pain is often described as shooting, lancinating, shocklike, or stabbing. The episodes frequently are triggered by painless sensory stimulation to perioral trigger zones, eg, a patch of facial skin, mucosa, or teeth innervated by the ipsilateral trigeminal nerve. Triggers include touch, certain head movements, talking, chewing, swallowing, shaving, brushing teeth, or even a cold draft. The most commonly affected dermatomal zones are innervated by the second and third branches of the trigeminal nerve.

The episodes may be repetitive, recurring, and remitting randomly. Pain-free intervals, which might last for years early in the course of TN, typically grow shorter as the disease progresses. During episodes of pain, some many patients have difficulty talking, eating, and maintaining facial hygiene out of fear of triggering the pain.

Standard bedside neurological examination findings are normal in TN. Patients may refuse examinations of the face, fearing the triggering of pain. Male patients may present with an area of the face, the trigger zone, that is unshaven and unkempt. The finding of significant numbness in the trigeminal distribution of TN suggests secondary TN and more extensive damage to the trigeminal nerve.

Most of the following conditions are not easily confused with TN:

Trigeminal neuropathy: Sensory loss is usually prominent; constant burning pain is common.

Herpetic and postherpetic neuralgia (PHN): This condition usually affects the first branch of the trigeminal nerve. The diagnosis of PHN usually requires the outbreak of shingles (herpes zoster) in the forehead or eye.

Acute herpetic neuralgia is the norm in shingles, but pain that persists after the lesions have healed is PHN. The risk of PHN development is directly related to patient age.

Neoplasms: These may present as a compressing mass or neoplastic cell infiltration of the trigeminal nerve. Pain is usually more constant than in TN1, and facial numbness is more common.

Granulomatous inflammation (eg, tuberculosis, sarcoidosis, Behçet syndrome, collagen vascular diseases): These and other vasculitides may affect the trigeminal nerve and simulate TN.

Other conditions that may mimic TN include odontogenic pain, geniculate neuralgia, glossopharyngeal neuralgia, temporomandibular disorders, cluster headache, hemicrania, and SUNCT (short-lasting, unilateral neuralgia from headache attacks with conjunctival injection and tearing) syndrome.

The diagnosis of facial pain is almost entirely based on the patient's history. In most cases of facial pain, no specific laboratory tests are needed. A blood count and liver function tests are required if medications are contemplated, specifically with carbamazepine.

Oxycarbazine can cause hyponatremia, so the serum sodium should be tested after institution of therapy. Brain imaging, particularly MRI using heavily T2 weighted (FIESTA) sequences, is critical to exclude a neoplastic cause of TN.

Although rarely indicated, appropriate blood work for rheumatic diseases, such as scleroderma (trigeminal neuropathy is reported in up to 5% of patients with this collagen vascular disease) and systemic lupus erythematosus, should be undertaken in patients with atypical features of facial pain and a systemic presentation of collagen vascular disease. Appropriate blood work includes a sedimentation rate, antinuclear antibody titer, double-stranded DNA, anti-Sm antibody, lupus erythematosus cell preparation, and complete blood count to look for hematological abnormalities (eg, hemolytic anemia, leukopenia, thrombocytopenia). Particularly in the case of scleroderma, creatinine kinase and aldolase levels may be elevated with muscle involvement. Antibody titers to SCL-86 and SCL-70 may also be present.

Pathophysiology

The etiology of most cases of TN is chronic vascular compression and injury to the trigeminal nerve at its entrance into the brainstem (pons). In one study, 64% of the compressing vessels were identified as an artery, most commonly the superior cerebellar (81%). Venous compression was identified in 36% of cases.

Vascular compression of the trigeminal nerve appears to cause demyelination and remyelination of the nerve with persisting abnormalities of myelination (dysmyelination).

The most common theoretical explanation for TN proposes that high-frequency ectopic impulses are either generated from or augmented by areas of dysmyelination. These abnormal discharges may ignite a chain reaction of neuronal depolarization in the trigeminal ganglion. The subsequent cascade of neuronal activity is progated centrally into the trigeminal nucleus and is then perceived by the patient as an overwhelming burst of pain.

Although most cases of TN are caused by vascular compression, other structural disease is present in secondary TN, which can produce either typical or atypical pain. For example, a mass such as a meningioma may displace and damage the nerve, resulting in pain. Alternatively, inflammation secondary to multiple processes may be due to the underlying lesion.

In MS, lesions in the pons at the root entry zone (REZ) of the trigeminal fibers have been demonstrated. This is one form of "symptomatic" trigeminal neuralgia related to visible pathology.

Treatment options:

Treatment options for TN include medicines, surgery, and complementary approaches.

Since most patients incur TN when older than 60 years, medical management is the logical initial therapy. Medical therapy alone is adequate treatment for 75% of patients.

According to Dalessio et al, medications work by interrupting the temporal summation of afferent impulses that precipitate the attack. Once a patient experiences breakthrough pain on a single agent, a second and even a third additional medication may be required to restore relief. Medical therapy often is sufficient and effective, allowing surgical consideration only if pharmacologic treatment fails. Because this disorder may remit spontaneously after 6-12 months, patients may elect to discontinue their medication in the first year following the diagnosis. Most must restart medication in the future.

Anticonvulsant medicines—used to block nerve firing—are generally effective in treating TN. These drugs include carbamazepine, oxcarbazepine, topiramate, clonazepam, phenytoin, lamotrigin, and valproic acid. Gabapentin or baclofen can be used as a second drug to treat TN and may be given in combination with other anticonvulsants.

Tricyclic antidepressants such as amitriptyline or nortriptyline are used to treat pain described as constant, burning, or aching. Typical analgesics and opioids are not usually helpful in treating the sharp, recurring pain caused by TN. If medication fails to relieve pain or produces intolerable side effects such as excess fatigue, surgical treatment may be recommended.

The most effective medication for the treatment of trigeminal neuralgia (TN) is carbamazepine. It acts by inhibiting the neuronal sodium channel activity, thereby reducing the excitability of neurons. The effective dose ranges from 600-1200 mg/d, with serum concentrations between 40-100 mcg/mL. However, many adverse CNS effects (eg, vertigo, sedation, ataxia, diplopia) are associated with carbamazepine, which may make it difficult to use in elderly patients. The dose may be tapered once pain is controlled, since remission may occur. A complete blood count (CBC) must be obtained during the first few weeks of therapy and yearly thereafter. Agranulocytosis and aplastic anemia are extremely rare adverse effects, but suppression of the WBC count is not uncommon. This mild suppression of the WBC count does not warrant discontinuation of carbamazepine therapy. Hepatic function should also be monitored. Up to 70% of patients receive complete or acceptable partial relief, at least temporarily. Oxycarbazine is a newer agent that may have fewer side effects, but it can cause hyponatremia, which should be monitored with serial serum sodium measurements in the first few weeks of therapy.

Gabapentin, lamotrigine topiramate, and several other newer anticonvulsants are being used to treat TN. Further outcome studies on their use in the treatment of TN are needed.

Surgical treatment is indicated for patients whose TN is intractable despite medical therapy, in those who are intolerant to the adverse effects of the medications, and in those in whom previous procedures failed. In some studies, more than 50% of patients with TN eventually had some kind

of surgical procedure. Experience would indicate that medical management eventually fails in most patients with TN, and those patients undergo surgery.

Microvascular decompression (MVD) is the most invasive of all surgeries for TN, but it also offers the lowest probability that pain will return. A neurectomy, which involves cutting part of the nerve, may be performed during MVD if no vessel is found to be pressing on the trigeminal nerve. Neurectomies may also be performed by cutting branches of the trigeminal nerve in the face. When done during MVD, a neurectomy will cause permanent numbness in the area of the face that is supplied by the nerve or nerve branch that is cut. However, when the operation is performed in the face, the nerve may grow back and in time sensation may return. Peripheral neurectomy, although safe and effective, is rarely used but may be of value in patients who have TN and a limited life span. MVD is usually indicated for patients younger than 70 years who are at lower risk for complications during general anesthesia, although healthy older patients may tolerate the procedure well. MVD is commonly is performed in younger, healthier patients, especially those with pain isolated to the ophthalmic division or in all 3 divisions of the trigeminal nerve and in those with secondary TN. MVD is now the most common surgery performed for TN and is the classic and most effective surgical procedure. It involves a posterior fossa craniotomy and dissection of vascular elements that compress the trigeminal nerve in the subarachnoid space. Surgeons perform the operation under general anesthesia, incising the skin behind the ear and performing a 3-cm craniectomy.

Acute Zoster and Postherpetic Neuralgia.

Neuralgia associated with a vesicular eruption due to the herpes zoster virus may affect cranial as well as peripheral nerves. In the region of the cranial nerves, two syndromes are frequent: herpes zoster auricularis and herpes zoster ophthalmicus. Both may be exceedingly painful in the acute phase of the infection. In the former, herpes of the external auditory meatus and pinna and sometimes of the palate and occipital region—with or without deafness, tinnitus, and vertigo—is combined with facial paralysis. This syndrome, since its original description by Ramsay Hunt, has been known as geniculate herpes, supported only by the postmortem study of Denny-Brown and RD Adams. Pain and herpetic eruption due to herpes zoster infection of the gasserian ganglion and the peripheral and central pathways of the trigeminal nerve are practically always limited to the first division (herpes zoster ophthalmicus). Ordinarily, the eruption will appear within 4 to 5 days after the onset of the pain; if it does not, some cause other than herpes zoster will almost invariably declare itself. Nevertheless, a few cases have been reported in which the characteristic localization of pain to a dermatome, with serologic evidence of herpes zoster infection, was not accompanied by skin lesions. The acute discomfort associated with the herpetic eruption usually subsides after several days or weeks, or it may linger for several months. Treatment with acyclovir will shorten the period of eruption and pain, but the drug does not prevent its persistence as a chronic pain. It is mostly in the elderly that the pain becomes chronic and intractable. Usually it is described as a constant burning, with superimposed waves ofstabbing pain, and the skin in the territory of the preceding eruptionis exquisitely sensitive to the slightest tactile stimuli, even though the threshold of pain and thermal perception is elevated. Allodynia is frequent. This unremitting postherpetic neuralgia of long duration represents one of the most difficult pain problems with which the physician has to deal. Some relief may be provided by application of capsaicin cream, use of a mechanical or electrical stimulator, or administration of phenytoin, gabapentin, or carbamazepine.

Antidepressants such as amitriptyline and fluoxetine are helpful in some patients, and Bowsher has suggested, on the basis of a small placebo-controlled trial, that treatment with amitriptyline during the acute phase may prevent persistent pain. The addition of a phenothiazine to an antidepressant (e.g., amitriptyline 75 mg at bedtime and fluphenazine 1 mg tid) has proved to be a useful measure, but the long-term use of phenothiazines carries with it all the well-known risks, including that of inducing a movement disorder.

Probably equivalent results are obtained by a combination of valproic acid and an antidepressant, as reported by Raftery. King has reported that two (5-grain) aspirin tablets crushed

and mixed with cold cream or chloroform (15 mL) and spread over the painful zone on the face or trunk relieved the pain for several hours in most patients with postzoster neuralgia. Ketamine cream has been suggested as an alternative. The authors have limited experience with these topical treatments. Extensive trigeminal rhizotomy or other destructive procedures should be avoided, since these surgical measures are not universally successful and may superimpose a diffuse refractory dysesthetic component on the neuralgia (anesthesia dolorosa).

Trigeminal neuropathy. A varietyof diseases may affect the trigeminal nerve. Most present with sensoryloss on the face or with weakness of the jaw muscles. Deviation of the jaw on opening indicates weakness of the pterygoids on the side to which the jaw deviates. Some cases are due to Sjo¨gren's syndrome or a collagenvascular disease such as systemic lupus erythematosus, scleroderma, or mixed connective tissue disease. Among infectious causes, herpes zoster and leprosyshould be considered. Tumors of the middle cranial fossa (meningiomas), of the trigeminal nerve (schwannomas), or of the base of the skull (metastatic tumors) maycause a combination of motor and sensorysigns. Lesions in the cavernous sinus can affect the first and second divisions of the trigeminal nerve, and lesions of the superior orbital fissure can affect the first (ophthalmic) division; the accompanying corneal anesthesia increases the risk of ulceration (neurokeratitis).

Loss of sensation over the chin (mental neuropathy) can be the only manifestation of systemic malignancy. Rarely, an idiopathic form of trigeminal neuropathyis observed. It is characterized by numbness and paresthesia, sometimes bilaterally, with loss of sensation in the territoryof the trigeminal nerve but without weakness of the jaw. Gradual recoveryis the rule. Tonic spasm of the masticatory muscles, known as trismus, is symptomatic of tetanus or may occur in patients treated with phenothiazine drugs.

Damage the branches of the trigeminal nerve, facial and cranial injuries and fractures are probably the most common but they do not often come to the attention of neurologists. The most superficial branches of the nerve—the supratrochlear, supraorbital, and infraorbital— are the ones usually involved. The sensory loss is present from the time of the injury, and partial regeneration may be attended by constant pain, often demanding nerve block or sectioning. Of the various inflammatory and infectious diseases that affect the trigeminal nerves or ganglia, herpes zoster ranks first. Persistent pain after herpetic infection of the fifth nerve is a serious problem, not responding well to any type of treatment. Herpes simplex virus has been isolated from the ganglion in as many as 50 percent of routine autopsies, but in nearly all patients this virus is associated only with lesions of the skin and lips.

Middle ear infections and osteomyelitis of the apex of the petrous bone may spread to the ganglion and root, also implicating the sixth cranial nerve (Gradenigo syndrome).

Human immunodeficiency virus (HIV) infection has not been clearly implicated in infection of the fifth nerve (as it has in the seventh nerve), but reactivation of latent herpes zoster is seen with acquired immune deficiency syndrome (AIDS).

The trigeminal root may be compressed or invaded by intracranial meningiomas, acoustic neuromas, trigeminal neuromas, cholesteatomas, and chordomas and by tortuous branches of the basilar artery. Sinus tumors and metastatic disease may also implicate the nerve, causing pain and a gradually progressive sensory loss. The ophthalmic division of the fifth nerve may be involved in the wall of the cavernous sinus in combination with the third, fourth, and sixth nerves by a variety of processes, including thrombosis of the cavernous sinus. Tumors of the sphenoid bone (myeloma, metastatic carcinoma, squamous cell carcinoma, and lymphoepithelioma of the nasopharynx) may involve branches of the trigeminal nerve at their foramina of entry or exit.

The mandibular division may be compressed by the roots of an impacted third molar (wisdom) tooth. Several times the authors have observed numbness of the chin and lower lip (infiltration of the mental nerve) as the first indication of metastatic carcinoma of the breast and prostate and from multiple myeloma. Massey and colleagues have described 19 such cases ("numb-chin" syndrome).

Neurologists often encounter instances of slowly evolving unilateral or bilateral trigeminal neuropathy in which sensory impairment is confined to the territory of the trigeminal nerve, sometimes associated with pain and paresthesias and disturbances of taste. Loss of facial sensation can occur as part of a widespread sensory neuropathy that occurs as a remote effect of cancer or as part of Sjo¨gren disease. More common is an association between isolated trigeminal neuropathy and immunemediated connective tissue disease. Of 22 such cases described by Lecky and colleagues, 9 had either scleroderma or mixed connective tissue disease, and a similar number had either organ- or nonorgan-specific serum autoantibodies. The symptoms may involve the other side years later. Hughes has also reported cases of trigeminal neuropathy with scleroderma, lupus erythematosus,and Sjo¨gren disease. We have seen several patients with Sjo¨gren disease in whom the trigeminal neuropathy and the associated antibodies or inflammation of the minor salivary glands were evident well before the characteristic sicca syndrome or other systemic manifestations of the disease. The condition may remain troublesome for years. Pathologic data are limited but point to an inflammatory lesion of the trigeminal ganglion or sensory root. Stilbamidine and trichloroethylene are known to cause sensory loss, tingling, burning, and itching exclusively in the trigeminal sensory territory.

Spillane and Wells stressed an isolated trigeminal neuropathy (it has been called Spillane's trigeminal neuritis in some texts). Four of their 16 patients had an associated paranasal sinusitis, but subsequent reports have failed to substantiate a causal relationship between sinusitis and cranial neuritis. One wonders how many of these individuals had connective tissue disease. A less common form of idiopathic trigeminal sensory neuropathy with which we have limited experience has a more acute onset and a tendency to resolve completely or partially, in much the same manner as Bell's palsy, with which it is sometimes associated (Blau et al). A recurrent variety of uncertain origin has been reported in the dental literature.

We have had experience with two patients whose facial numbness was a component of an upper cervical disc syndrome that included numbness on the same side of the body. Cases such as these are reported sporadically in the literature.

A pure unilateral trigeminal motor neuropathy is a clinical rarity. Chia has described five patients in whom an aching pain in the cheek and unilateral weakness of mastication were the main features. Electromyography (EMG) showed denervation changes in the ipsilateral masseter and temporalis muscles. The outcome was favorable.

In most cases of trigeminal neuropathy except those due to tumor and herpes zoster, the results of gadolinium-enhanced magnetic resonance imaging (MRI) are normal, as is the cerebrospinal fluid (CSF). The function of the nerve may be studied by the electrical recording of blink reflexes. A few laboratories have developed an evoked potential test specifically of the trigeminal nerve.

Temporomandibular Joint Pain (Costen Syndrome).

This is a form of craniofacial pain consequent on dysfunction of one temporomandibular joint. Malocclusion due to ill-fitting dentures or loss of molar teeth on one side, with alteration of the normal bite, may lead to distortion of and ultimately degenerative changes in the joint and to pain in front of the ear, with radiation to the temple and over the face. The diagnosis is supported by the findings of tenderness over the joint, crepitus on opening the mouth, and limitation of jaw opening. The favored diagnostic maneuver involves palpating the joint from its posterior aspect by placing a finger in the external auditory meatus and pressing forward. The diagnosis can be made with some confidence only if this entirely reproduces the patient's pain. CT and plain films are rarely helpful, but effusions have been shown in the joints by MRI.

Management consists of careful adjustment of the bite by a dental specialist and should be undertaken only when the patient meets the strict diagnostic criteria for this condition.

The temporomandibular joint may also be the source of pain when involved by rheumatoid arthritis and other connective tissue diseases.

Facial Pain of Dental or Sinus Origin.

Maxillary and mandibular discomfort are common effects of nerve irritation from deep caries, dental pulp degeneration, or periodontal abscess. The pain of dental nerve origin is most severe at night, slightly pulsating, and often associated with local tenderness at the root of the tooth in response to heat, cold, or pressure. It is usually eradicated by infiltrating the base of the tooth with lidocaine.

Trigeminal neuritis following dental extractions or oral surgery is another vexing problem. There may be sensory loss in the tongue or lower lip and weakness of the masseter or pterygoid muscle. Sometimes the onset of "atypical facial pain" (see below) can be dated to a dental procedure such as tooth extraction, and, as usually happens, neither the dentist nor the neurologist is able to find a source for the pain or any malfunction of the trigeminal nerve. Roberts and coworkers as well as Ratner and associates have pointed out that residual microabscesses and subacute bone infection account for some of these cases. They isolated the affected region by local anesthetic blocks, curetted the bone, and administered antibiotics, following which the pain resolved. The removed bone fragments showed vascular and inflammatory changes and infection with oral bacterial flora.

Materials for self-control of quality of preparation:

A 43-year-old woman describes lancinating pains radiating into the right side of her jaw. This discomfort has been present for more than 3 years and has started occurring more than once a week. The pain is paroxysmal and routinely triggered by cold stimuli, such as ice cream and cold drinks. She has sought relief with multiple dental procedures and has already had two teeth extracted. Multiple neuroimaging studies reveal no structural lesions in her head. Assuming there are no contraindications to the treatment, a reasonable next step would be to prescribe which of the following?

a. Clonazepam, 1 mg orally three times daily
b. Diazepam, 5 mg orally two times daily
c. Divalproex sodium, 250 mg orally three times daily
d. Indomethacin, 10 mg orally three times daily
e. Carbamazepine, 100 mg orally three times daily

A 39-year-old left-handed woman is being treated with carbamazepine for lancinating pain in her left face. The pain is paroxysmal, usually occurring without apparent reason, but seems sometimes to be brought on by a cold breeze. Both trigeminal neuralgia and atypical facial pain involve pain that may be which of the following?

a. Lancinating
b. Paroxysmal
c. Associated with anesthetic patches
d. Abolished with resection of the gasserian ganglion
e. Unilateral

A 29-year-old woman comes to the emergency room with facial pain of new onset. She has stabbing pains on the left side of her face just below her eye. These last less than 1 s at a time, but are so severe that she winces involuntarily with each pain. The pain seems to be triggered by drinking cold fluids. The only other problems she has noticed are clumsiness in her right hand and blurred vision in her right eye. Both of these have been present for more than 2 years and have not interfered with her normal activities. Which CN affected in this patient?

Answer: **V CN.**

A 53-years-old female is detected: expression muscles palsy on the left: downturning mouth, nasolabial fold is smoothed, palpebral fissure is dilated. Patient cannot knit one's brow on

this side; Bell's symptome is positive. There is lacrimation from the left eye. Hearing and sense of taste are preserved. Ground the topical diagnosis.
Answer: **lesion of the left facial nerve at the level of the foramen stylomastoideum**

Recommended literature

Basic:

- Neurology: textbook / I.A. Hryhorova, L.I. Sokolova, R.D. Herasymchuk et al.; edited by I.A. Hryhorova, L.I. Sokolova. – Kyiv : AUS Medicine Publishing, 2020. – 624 p.

- Netter Atlas of Human Anatomy: Classic Regional Approach: Professional Edition with NetterReference Downloadable Image Bank (Netter Basic Science) 8th Edition By Frank H. Netter MD / Publisher : Elsevier; 8th edition (April 25, 2022). - 712 p. ISBN-10 : 0323793738 ISBN-13 : 978-0323793735

- Neuroanatomy through Clinical Cases 3rd Edition By Hal Blumenfeld / Publisher : Sinauer Associates is an imprint of Oxford University Press; 3rd edition (February 28, 2021).- 1056 p. ISBN-10 16053596299: ISBN-13 : 978-1605359625

- Pocket Neurology (Pocket Notebook Series) Third Edition By M. Brandon Westover MD PhD Publisher : LWW; Third edition (October 16, 2021). - 390 p. ISBN-10 : 1975169034 ISBN-13 : 978-1975169039

Additional:

- Topical Diagnosis in Neurology. Anatomy, Physiology, Signs, Symptoms / Mathias Baehr, Michael Frotscher (6 edition) – Thieme, 2019 - 332 p.

- Adams and Victor's Principles of Neurology / Allan Ropper, Martin Samuels, Joshua Klein, Sashank Prasad (11th edition). - McGraw-Hill, 2019. - 1664 p.

- Clinical Neuroanatomy Made Ridiculously Simple: Color Edition 6th Edition by Stephen Goldberg M.D. / Publisher: MedMaster; 6th edition (September 14, 2022).- 112 p. ISBN-10 : 1935660519 ISBN-13 : 978-1935660514

- Clinical Neurology and Neuroanatomy: A Localization-Based Approach, Second Edition 2nd Edition by Aaron Berkowitz / Publisher : McGraw Hill / Medical; 2nd edition (July 21, 2022).- 384 p. ISBN-10 : 1260453367 ISBN-13 : 978-1260453362

- Handbook of Neurosurgery 9th Edition by Mark S. Greenberg / Publisher : Thieme; 9th edition (October 23, 2019).- 1784 p. ISBN-10 : 1684201373 ISBN-13 : 978-1684201372

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