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Introduction

Nowadays one of the main conditions of higher medical establishment's activity is teaching of English – speaking foreign students. As there is a problem of this process' methodical equipment, such as English Neurology textbook according to the studying program, the staff of I.Y. Horbachevskyy Ternopil State Medical University Neurology Department has prepared English syllabus "Neurology in lectures" under the editorship of prof. Shkrobot S.I. It reckons in the existing differences between studying programs in English – speaking countries and Ukraine, some differences in the names of diseases and classification.

The syllabus includes certain lectures of propaedeutics and special neurology for the students of medical faculty.

All the data of anatomy, physiology of nervous system and propaedeutics of neurological pathology should be studied by the fourth year students of medical faculty for better understanding of topical diagnostics and neurological syndromes. All the diseases are described according to the Xth international classification (1997). There are described different forms of the diseases, the peculiarities of the course of disease, diagnosis, modern methods of treatment, social and working rehabilitation and prevention of the diseases.

In lectures of special neurology one can find the questions of etiology, pathogenesis and diagnosis of vascular, inflammatory and demyelinating diseases, brain tumors, peripheral diseases of nervous system, autonomic disorders, hereditary diseases, intoxications and so on. A great attention is paid to urgent treatment of dynamic cerebral circulation disorders, strokes, inflammatory and demyelinating diseases of nervous system.

The syllabus is printed according to the demands of Central methodical cabinet of higher medical education in Ukraine.

Functions of nervous system. Conditioned and unconditioned reflexes. Motor system (symptomatic and topical diagnostics of movement disturbances)

Neurology as science

Neuropathology (from Greek neuro – nerve, pathos – disease, logos – science) – is a part of clinical medicine, which is involved in nervous diseases and its role in pathology of other organs and systems of human body.

The main structural, functional, genetic and anatomic unit of nervous system is neuron – nervous cell. Its main function is to accept and carry out impulse.

The main function of nervous system is unification and regulation of different physiological processes. That means that nervous system unites, integrates and subordinates all the parts of human body and provides its connection with environment.

The base of nervous system activity is **reflex principle**. Reflex – is a reaction of our organism to various outside and inside effects. It is provided by nervous system.

The reflex consists of afferent part (which accepts information), central part (that keeps information), and efferent part (that creates response). As a result we have a circle – like structure – receptor (primary information center) – programme center – executive apparatus (fig. 1, 2).

All the reflexes can be divided into different

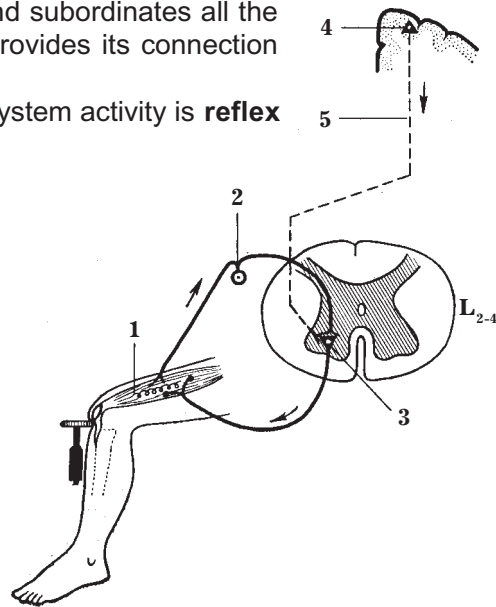


Fig. 1 Scheme of knee jerk reflex arch:

1. Receptors of nervous – muscular cords
2. Dorsal root ganglion of spinal cord
3. Afferent neuron
4. Efferent neuron
5. The central neuron of Pyramidal tract

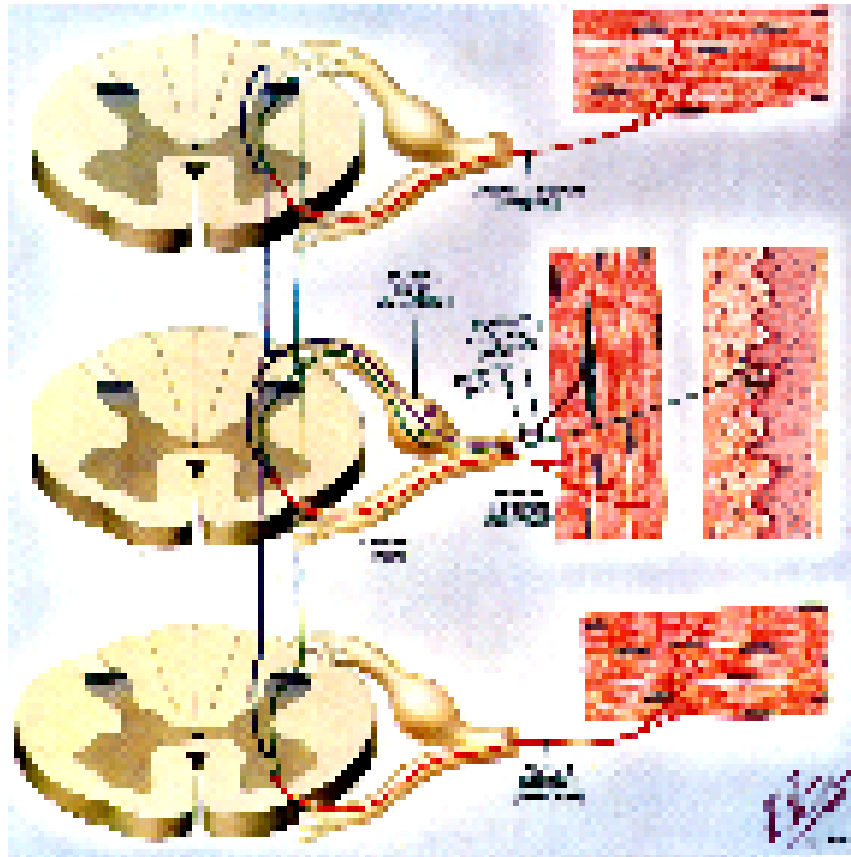


Fig. 2 Spinal effector mechanism (Frank H. Netter, M.D.)

groups – simple and complex, inborn and trained, conditioned and unconditioned.

Unconditioned reflexes:

1. They are inborn ones.
2. They are phylogenetically old, that means they were formed in course of phylogenesis.
3. They are based on certain anatomic structures (segments of spinal cord or brain stem).
4. They exist even without brain cortex control.
5. They are inherited.

6. They can be regulated by brain cortex.
7. They are basis for the conditioned reflexes.

Conditioned reflexes:

1. They are the result of the individual experience and are formed during ontogenesis.
2. They are unstable, that means they need constant support.
3. They aren't based on certain anatomic structures.
4. They are fixed in brain cortex.

There are such conditioned reflexes as speaking, writing, reading, calculation, practice.

Neurology studies unconditioned reflexes. That helps evaluate the state of nervous system. Conditioned reflexes are studied by psychiatrists.

Unconditioned reflexes are divided into such groups:

- Superficial and deep.
- Simple and complex.
- Proprioceptive (stretch, periosteal, joint).
- Exteroceptive (dermal, from mucose membrane).
- Interoceptive (from mucose membrane of internal organs – for example urination in case of internal sphincter irritation).

In clinical practice we evaluate the following reflexes (fig. 3-8):

Reflex	The group of reflex	Muscles	Nerves	Segments
1	2	3	4	5
Subeyebrow	Deep, periosteal	M. orbicularis oculi	Trigeminal N (V) – Facial N (VII)	Medulla oblongata and pons
Corneal (lid)	Superficial, from mucose membrane	M. orbicularis oculi	Trigeminal N (V) – Facial N (VII)	Medulla oblongata and pons
Jaw Jerk (mandibular, chin masseter) (Bechterev's)	Deep, periosteal	M. masseter	Trigeminal N (V) – Mandibular N (sensory and motor)	Medulla oblongata and pons
Pharyngeal	Superficial, from mucose membrane	Mm. constrictores pharyngis and others	Glossopharyngeal N (IX) and Vagus N (X) (sensory and motor)	Medulla oblongata

Продовження табл.

1	2	3	4	5
Palatal (palatine)	Superficial, from mucose membrane	Mm. levatores velli palatini	Glossopharyngeal N (IX) and Vagus N (X) (sensory and motor)	Medulla oblongata
Biceps	Deep, stretch	M. Biceps brachii	Musculocutaneous N	C5-C6
Triceps	Deep, stretch	M. Triceps brachii	Radial N	C7-C8
Radial (carporadial, brachioradial)	Deep, periosteal	Mm. pronatores flexores, digitorum, brachioradialis, biceps	Median N, Radial N, Musculocutaneous N	C5-C8
Scapulo-humeral (scapuloperiosteal)	Deep, periosteal	Mm. teres major, subscapularis	Subscapular N	C5-C6
Upper superficial abdominal	Superficial, dermal	Mm. transversus, obliquus, rectus abdominis	Intercostals Nn	Th7-Th8
Middle superficial abdominal	Superficial, dermal	Mm. transversus, obliquus, rectus abdominis	Intercostals Nn	Th9-Th10
Lower superficial abdominal	Superficial, dermal	Mm. transversus, obliquus, rectus abdominis	Intercostals Nn	Th11-Th12
Cremasteric	Superficial, dermal	M.cremaster	Genitofemoral N	L1-L2
Knee jerk, or patellar reflex (quadriceps)	Deep, stretch	M.quadriceps femoris	Femoral N	L3-L4
Achilles (ankle jerk)	Deep, stretch	M. triceps surae	Tibial N (Sciatic N)	S1-S2
Plantar (sole)	Superficial, dermal	Mm. flexores digitorum pedis and others	Sciatic N	L5-S1
Anal	Superficial, dermal	M. sphinter ani externus	Anococcygei Nn	S4-S5

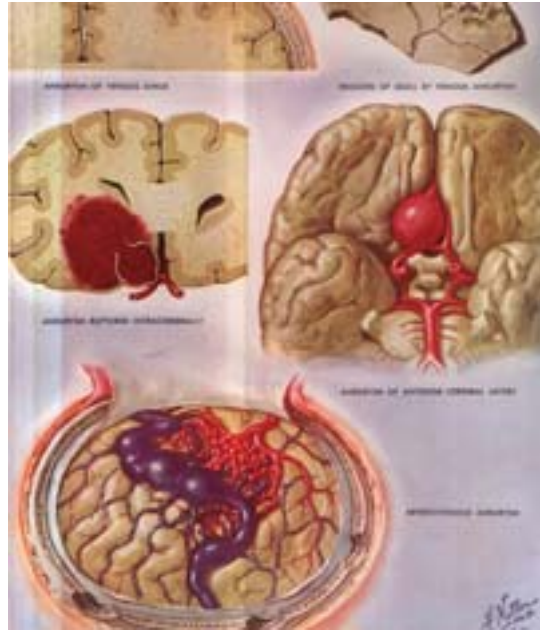


Fig. 3 Examination of abdominal reflexes.

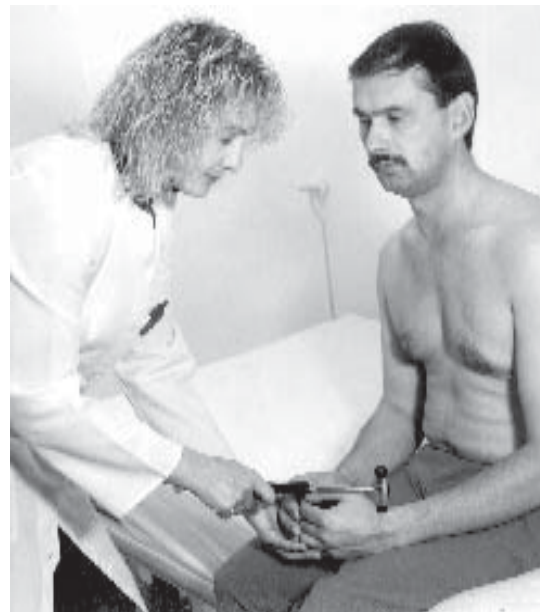


Fig. 4 Examination of carpo-radial reflex.



Fig. 5 Examination of biceps reflex.



Fig. 6 Examination of triceps reflex.

Normally the reflexes are lively and the same on both sides ($D=S$). Hyper-reflexion, hyporeflexion or areflexion are abnormal signs. If the reflexes change on one side we can write down: $D > S$; $S > D$; $D = S$; $D < S$; $S < D$.

Put the pathologic sign always on the first place.

Motor system

This system provides conduction of nervous impulse from brain cortex to muscles. The way of this impulse is known as **motor way** or **tractus corticomuscularis**. It consists of two neurons:

1. central.
2. peripheral.

Upper and lower extremities, neck, trunk and perineum muscles' innervation

The **first (central) neuron** is called **tractus corticospinalis** (fig 9).

The second (peripheral) neuron is called **tractus spinomuscularis**.

The fibers of tr. corticospinalis are of Betz cells origin. Most of its fibers

originate from anterior central gyrus, posterior parts of upper and middle frontal gyres and paracentral lobe (area 2, 4, 6). The Betz cells in central anterior gyrus are presented vice versa to the parts of the body:

- in upper part the muscles of lower extremities are presented

- in middle part – the muscles of upper extremities are presented

- in lower part – the face muscles are presented.

There is crossed innervation of muscles. The axons of Betz cells that create tr.



Fig. 7 Examination of knee reflex.



a



б

Fig. 8 Examination of ankle (Achilles) reflex (a,b).

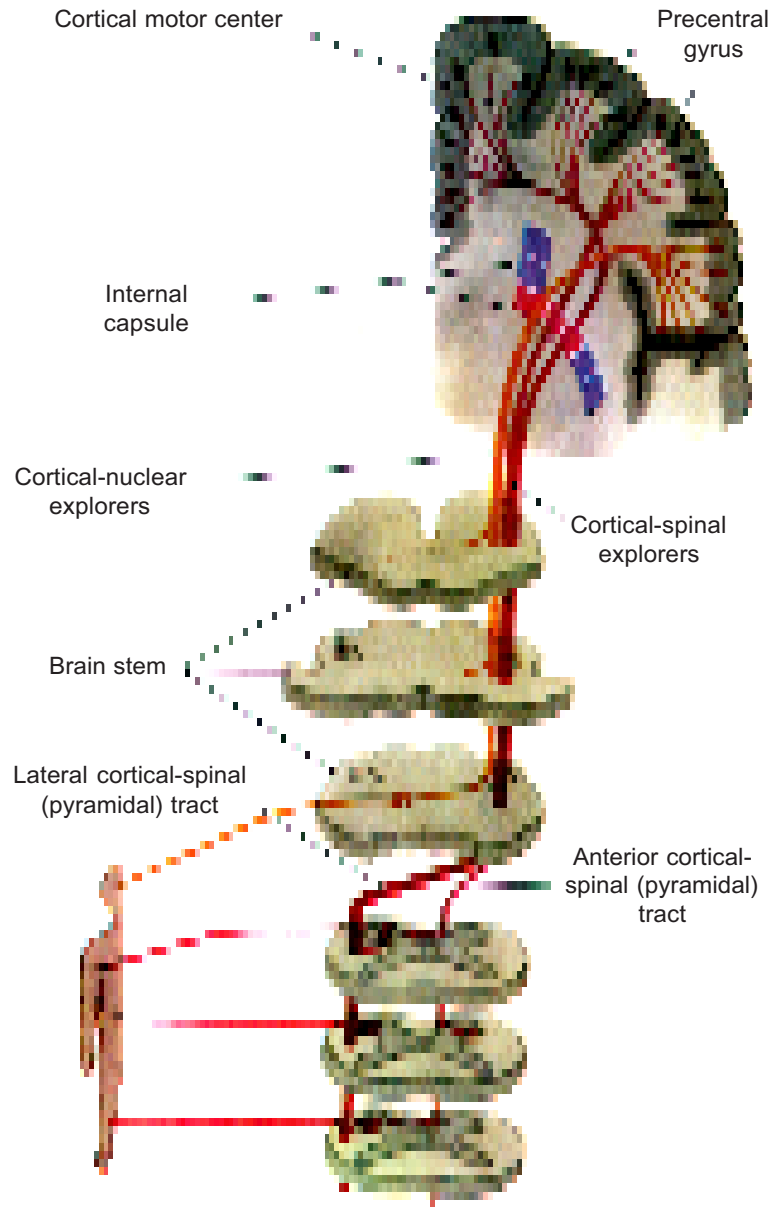


Fig. 9 Scheme of Pyramidal (cortical-spinal) and cortical-nuclear tracts

corticospinalis go through *radiate crown* to *internal capsule* (fig 10) via its anterior 2/3 of posterior leg. Then the axons of motor way go through the peduncles, pons to medulla oblongata to form *pyramids*. 80 % of all fibers make decussation on the border between

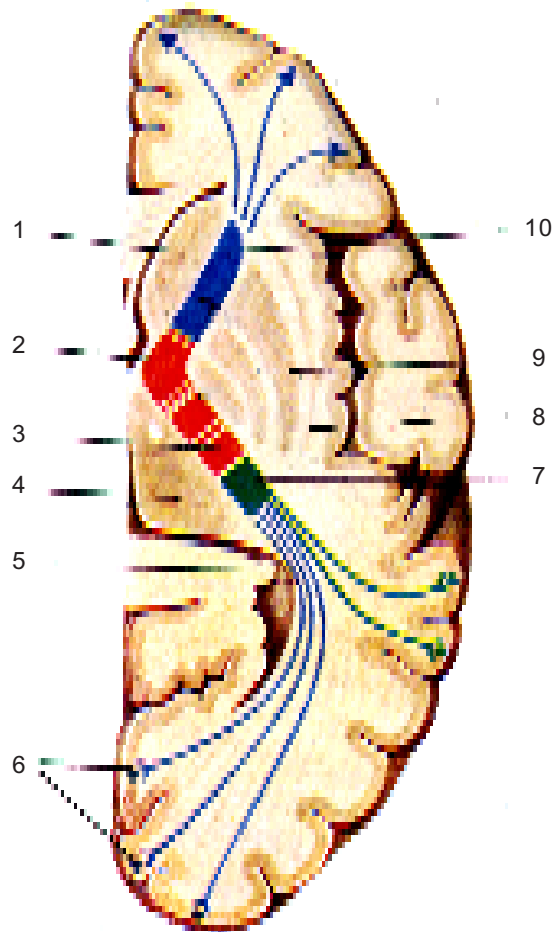


Fig. 10 Internal capsule structures (for Phlexig):

1. The head of nucleus caudatus;
2. The knee of Internal capsule;
3. The posterior leg of Internal capsule;
4. Thalamus;
5. Gratiole's tract;
6. Fissura calcarina of the Occipital lobe;
7. Posterior therty part of the Internal capsule;
8. Pallidum;
9. Putamen;
10. The anterior leg of Internal capsule

medulla oblongata and spinal cord. The crossed fibers go to the lateral funiculi of spinal cord on the opposite side and create **tr. corticospinalis lateralis**. The last provides lower and upper extremities muscles innervation. The rest – 20 % of all fibers aren't crossed. They go to funiculus anterior and create **tr. corticospinalis anterior** (fasciculus Turka). This one provides neck, trunk, and perineum muscles innervation. The fibers of tr. corticospinalis are finished in motor neurons of spinal cord anterior horns.

The **second neuron** – peripheral – **tractus spinomuscularis**

Neurons of C1-C4 anterior horns innervate neck muscles, C5-Th1-2 – muscles of upper extremities, Th2-Th12 – trunk muscles, L1-S2 – muscles of lower extremities, S3-S5 – muscles of pelvic organs. The second neuron originates from anterior horns alpha-motorneurons of spinal cord. Axons of these neurons go within anterior roots and then join with posterior ones to form the spinal nerve. Each spinal nerve gives 4 branches (fig 11):

1. ramus anterior (together they form plexus – cervical, brachial, lumbar and sacral);
2. ramus posterior (it is spinal nerve, which innervates posterior trunk muscles);
3. ramus meningeus;
4. ramus comunicante albi.

Thus, the motor impulse goes from anterior horns through anterior roots, spinal nerve, plexus and peripheral nerves to muscles.

That's the reason to make following conclusions:

1. The muscles of upper and lower extremities have unilateral cortical innervation from contralateral hemisphere.
2. The muscles of neck, trunk and pelvic organs have bilateral innervation from both hemispheres. In case of unilateral pathologic focus these structures do not suffer.

Face, tongue and pharynx muscles innervation

This way is called **tractus corticomuscularis**.

The first central neuron is called **tractus corticonuclearis**.

The second peripheral one is called **tractus nucleomusculares**.

The first neuron cells are situated in the lower part of anterior central gyrus. The axons go through **radiate crown**, the knee of **internal capsule** to brain stem (that means peduncles, pons and medulla oblongata). There are nuclei of CNs in brain stem.

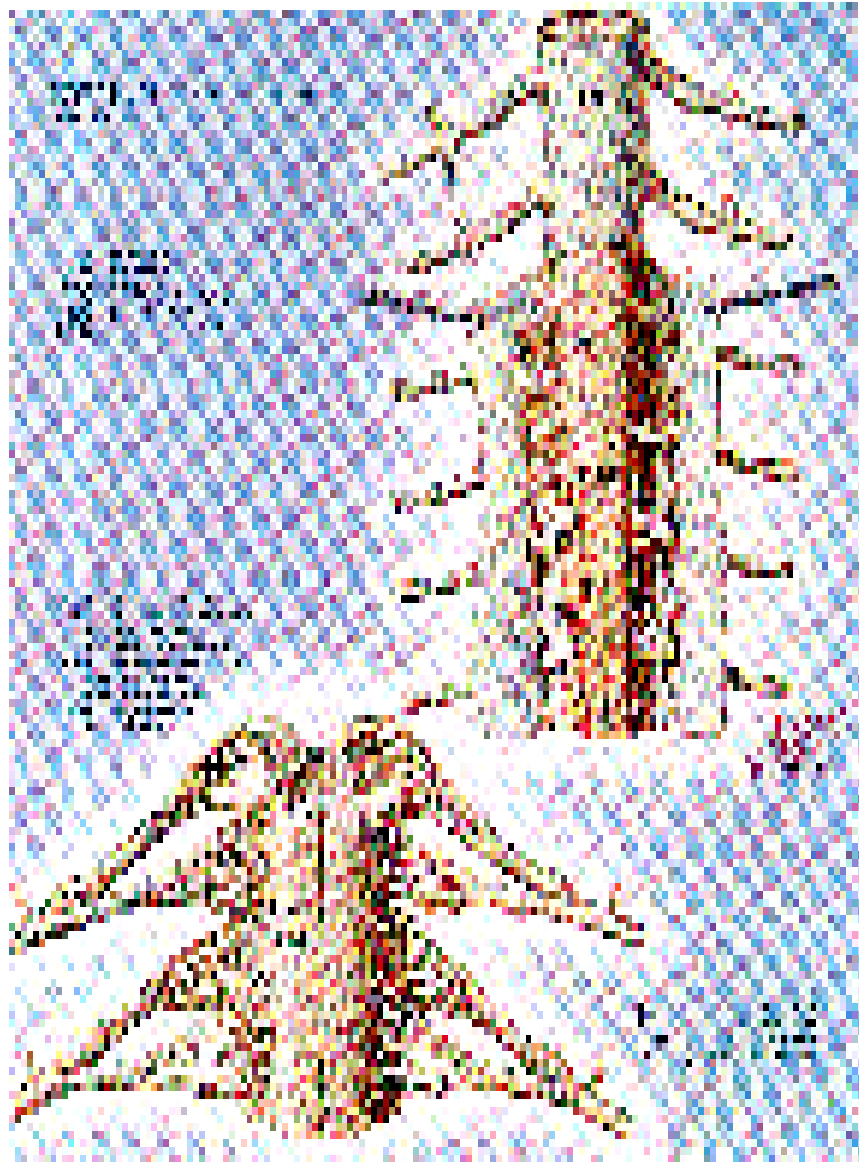


Fig.11 Spinal membranes and nerve roots (Frank H. Netter, M.D.)

And one more peculiarity – the fibers of tractus corticonuclearis make decussation above all the nuclei. This decussation is incomplete. The only exception is lower part of VII CN nucleus and nucleus of XII CN. In this case decussation is complete.

The second neuron is situated in motor nucleus of CNs. This way to face muscles is called **tractus nucleomuscularis**.

Thus we can make the following conclusions:

1. The face muscles have bilateral cortex innervation except the mimic muscles and tongue muscles that have unilateral innervation from the opposite hemisphere.
2. The muscles of upper and lower extremities, lower mimic muscles and tongue muscles have unilateral cortical innervation.
3. All the other muscles (the muscles of neck, trunk, perineum, extraocular muscles, muscles of mastication, pharyngeal and palatal muscles) have bilateral cortical innervation.

Movement disturbances

In case of complete lesion of motor way (tractus corticomuscularis) **paralysis (plegia)** occurs. That means the absence of active movements.

In case of incomplete lesion of motor way **paresis** occurs (fig 12, 13). That means active movements disorders – hemi-, tetra-, mono-, tri- and paraparesis.

Paralysis is divided into:

- Central (spastic);
- Peripheral (flaccid).



Fig. 12. Bare test (upper).



Fig. 13. Bare test (lower).

Central or spastic paralysis is caused by the lesion of central neuron and its fibers (tr. corticospinalis or tr. corticonuclearis).

Peripheral or flaccid paralysis is caused by the lesion of peripheral neuron (tractus spinomuscularis or tractus nucleomuscularis).

The main features of central or spastic paralysis are:

1. It is a diffuse paralysis.
2. There is spastic hypertonus of muscles.
That means:
 - Tonus is increased in the group of flexors in upper extremities and in the group of extensors in lower extremities;
 - “clasp – knife” symptom;
 - in course of evaluation tonus decreases.
3. Hyperreflexion of stretch and periosteal reflexes.
4. There are pathologic reflexes. They are considered to be reliable signs of central paralysis.

There are following subcortical oral pathological reflexes:

Bechterev’s sign. It is caused by impact of hammer on the upper lip. Reciprocal reaction consists in reduction m. orbicularis oris, a circular muscle of a mouth (reflex contraction of the lips forward).

Nasolabial reflex (Astvatsaturov’s sign) It is caused by impact of hammer on top of a nose. Reciprocal reaction is reflex contraction of the lips forward too.

Sucking (lip or snout) reflex. It is elicited by scraping the hard palate with a tongue blade or an applicator stick, or by applying other stimuli to the lips or shaped irritation of them; in the answer are observed sucking movements by lips.

The distance-oral reflex (Karchikjan’s sign) It is caused by hammer approximation a mouth of the patient: even before impact by it arises reflex contraction of the lips forward (reduction m. orbicularis oris).

Palm-mental reflex (Marinesku-Radovichi’s sign) It is caused by shaped irritations of skin of a palm above tenar, reduction contraction of m. mental on the given side, with light displacement of a skin of a chin up.

All the pathologic reflexes from the lower extremities are divided into: flexing and extending (fig 14-19). To extending reflexes belong:



Fig.14. Examination of Babinski sign



Fig. 15. Examination of Oppenheim's sign



Fig. 16. Examination of Gordon's sign

Babinski reflex. It is caused by pressed the end of applicator stick against the sole of the foot on the lateral side of the heel, from whence it is carried forward toward the little toe and, when the ball of the foot is reached, across it toward the base of the great toe. The response consist of extension of the great toe, usually associated with fanning (abduction and slight flexion) of the other toes.

The Babinski sign is referred to as being present or absent, not positive or negative.

Oppenheim's sign. It is caused by applying firm pressure on the shin with the fingers positioned, beginning just below the knee and carrying the stimulus rather slowly downward to the ankle. The response – extension of the great toe.

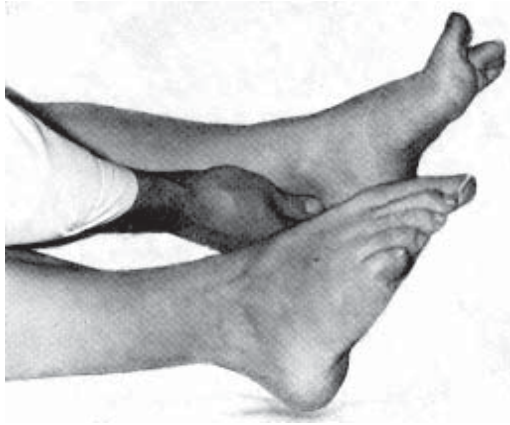


Fig. 17. Examination of Sheffer's sign



Fig. 18. Examination of Rossolimo sign



Fig. 19. Examination of Jukovski sign

Gordon's sign. It is caused by squeezing the calf, the toe movement.

Chaddock's sign. Stroking the lateral aspect of the foot below the external malleolus may elicit the abnormal toe movement.

To flexion ones belong:

Rossolimo reflex: it is a muscle-stretch reflex obtained by tapping the plantar surfaces of the toes with a reflex hammer. A modified method of elicitation is by producing a very quick, short stretch of the toe flexors by briskly brushing them in the direction of extension with a slapping motion of hands, the dorsum or plantar surface the fingers making contact with the toes.

Bechterev's sign. It is a muscle-stretch reflex obtained by tapping the external surfaces of feet of bending of fingers with a reflex hammer.

Jukovski sign. It is caused by hammer impact on a sole under fingers; response is reflex flexing of II-V fingers.

There are following pathologic reflexes on upper extremities:

Bechterev's sign. It is a muscle-stretch reflex of bending of fingers obtained by tapping the back of hand with a reflex hammer.

Jukovski sign. It is caused by hammer impact on a palm under fingers; response is reflex flexing of II-V fingers.

Rossolimo (Venderovych) reflex: It is a muscle-stretch reflex obtained by tapping the palmar surfaces of the fingers with a re-flex hammer; the response is reflex flexing of II-V fingers.

Tremner reflex. It is a muscle-stretch reflex obtained by tapping the palmar surfaces of the nail-phalax of II –V fingers. The response is fingers flexing.

Jakobson – Laske reflex. It is caused by hammer impact on processus styloideus; the response is reflex flexing of II-V fingers.

Klipel – Veil reflex. It is caused by passive bending of II – V fingers. The response is thumb flexing.

5. Protective reflexes (the reflexes of spinal automatism).

They are the signs of lesion of motor way. They are especially clearly expressed at cross lesion of a spinal cord (dissociation of underlying segments of the last from a brain). The result is squeezing of foot, sharp plantar flexion of toes (V.M.Bechterev). Serial putting irritations on one and the other leg, can result in imitation of automatisms of walking.

6. Pathologic synkinesis is involuntary movements in paralysed extremity. They are observed while moving by healthy extremity. Synkinesis are divided into:

- Global.
- Coordinatory.
- Imitating.

Peripheral (flaccid, atonic) paralysis

It occurs at lesion of tractus spinomuscularis or tractus nucleomuscularis (fig. 20-21).

The main features of peripheral paralysis are:

1. Areflexion or hyporeflexion.
2. Atonia or hypotonia.
3. Muscular atrophy.
4. Fasciculation of muscles.

5. It is limited paralysis.
6. There is reaction of degeneration.



Fig. 20. Peripheral paresis of right foot muscles.



Fig. 21. Peripheral paresis of right leg.

Table 1

Clinical signs of central (spastic) and peripheral (flaccid) paralysis

Sign	Central paralysis	Peripheral paralysis
Anatomical cause	Motor part of cortex damage (the first upper motor neuron)	Anterior horns of spinal cord and fibers of peripheral nerves damage (the second lower motor neuron)
The localization of paralysis	Diffuse	Local
Muscular tone	Spastic hypertonia	Hypotonia
Reflexes	Tendon deep reflexes are increased, superficial skin reflexes are decreased or absent	All unconditional reflexes are decreased or absent
Pathological reflexes	Babinski sign and other pathological reflexes are present	Absent
Defenses reflexes	Present	Absent
Pathological synkinesis	Present	Absent
Reaction of muscular degeneration	Absent	Present
Muscular atrophy	Absent	Present

The main symptoms of motor way lesion on different levels
(fig 22):

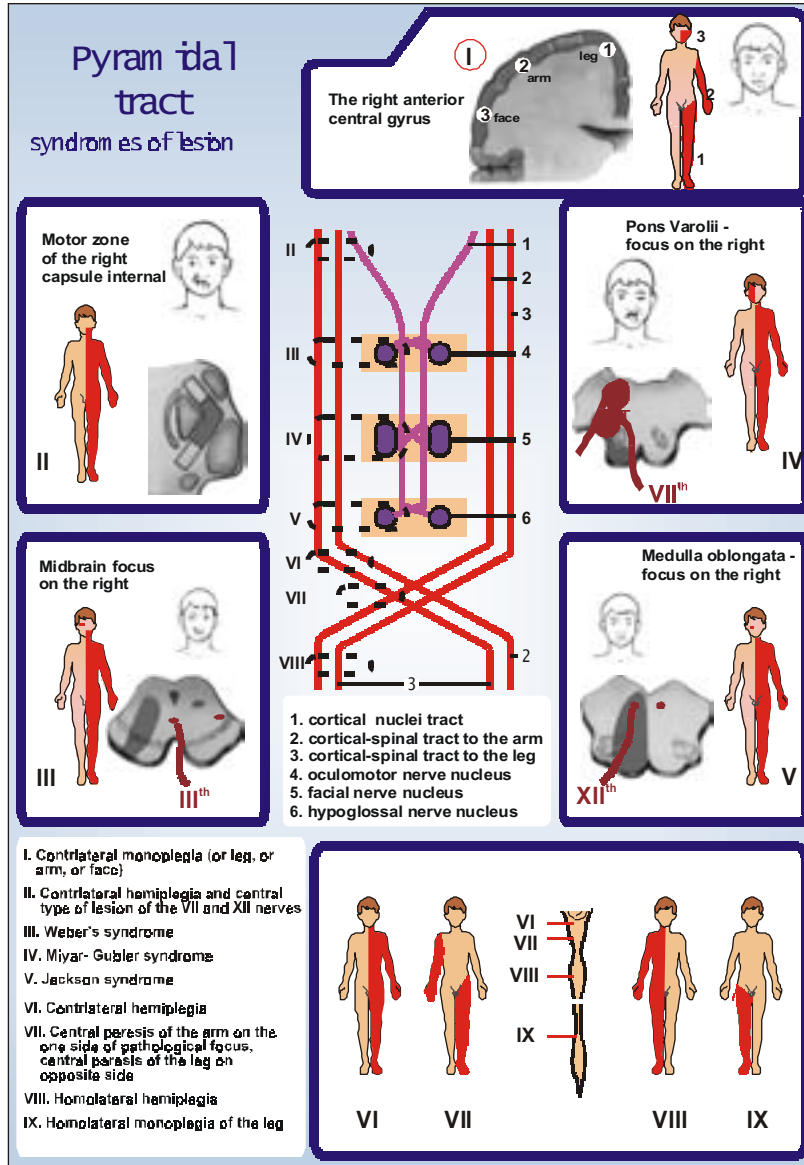


Fig.22

1. The lesion of anterior central gyrus

It usually causes monoplegia (or monoparesis) on the opposite side. If the focus is situated in upper part of anterior central gyrus, paralysis of lower extremity occurs. If it is in middle part of anterior central gyrus, we can observe paralysis of upper extremity. If it is in lower one, face suffers. In case of anterior central gyrus irritation Motor Jackson takes place. Motor Jackson is a set of local seizures that can cause generalized seizures.

2. The lesion of radiate crown

It usually causes central hemiplegia on the opposite side (that means that arm, leg, lower mimic muscles and tongue muscles are involved). By the way, if the process is much more expressed in upper part of radiate crown, paralysis in lower extremity dominates. If the process is much more expressed in middle part of radiate crown, paralysis in upper extremity prevails. If the process is much more expressed in lower part of radiate crown, paralysis in face muscles dominates. Besides hemianesthesia can join hemiplegia.

3. The lesion of internal capsule part of motor way

It can causes hemiplegia on the opposite side, central paresis of tongue muscles and lower mimic muscles. Hemihypesthesia often joins all the other symptoms and hemianopsia. Vernike – Mann posture is typical for this lesion.

4. The lesion of brain stem

It causes central paralysis on the opposite side and peripheral paralysis of face muscles on the side of lesion. It is known as alternating syndrome. The last are divided into peduncle, pontine and bulbar ones.

5. The lesion of pyramidal decussation part of motor way

It usually causes central paralysis of upper extremity on the side of lesion and paralysis of lower extremity on the opposite side. Sometimes tetraplegia or triplegia is observed.

6. The lesion of motor way in lateral funiculus of spinal cord

It causes central paralysis below the level C1-C4, C5-Th1, Th1–Th12, L1-S2.

7. The lesion of anterior horns or motor nucleus of CNs

It causes peripheral paralysis of certain muscles. At chronic process we can observe fasciculation of muscles. Also there are early atrophy and degenerative reaction.

8. Anterior roots lesion

it causes also peripheral paralysis. In most of cases it is observed only when several roots are damaged.

9. The lesion of nerve plexus

It causes peripheral paralysis, pain, sensory and autonomic disturbances.

10. The lesion of peripheral nerve

it causes peripheral paralysis of the muscle, innervated by this nerve. There is also pain, sensory and autonomic disturbances.

Sensation. Signs of sensation disturbances

Sensation and reception

Sensation is an ability of an organism to accept stimuli from external and internal environment.

Reception is a set of all afferent systems, which accept stimuli from external and internal environment and carry them out to the center.

Reception is wider concept, than sensation. One doesn't not feel everything he accepts.

Sensation is a part of reception, which one feels and can analyze by certain structures of his brain. It means that sensation is closely connected with activity of analyzers.

Analyzers and its structures

Analyzer is a sole functional system that consists of three parts:

1. Receiving apparatus (receptors) – receptor part
2. Sensory explorers – conductive part
3. Part of cortex, which receives information, analyzes and synthesizes it.

The main function of **Analyzer** is to accept and analyze stimuli. We distinguish the following analyzers:

- Visual
- Acoustical
- Sensual
- Testate

Reception apparatus

Receptors are sensitive structures that have ability to accept different changes of external and internal environment and transmit them as impulse.

Receptors are divided into:

1. Exteroreceptors (in skin and external mucose membrane)
2. Proprioceptors (in muscles, tendons, joints)
3. Interoreceptors (in inner organs, in vessels)

There are also telereceptors in ears and eyes.

1. **Exteroreceptors** accept superficial sensitiveness (light touch (tactile), pain and temperature sense). They are divided into **mechanoreceptors** (touch, pressure), **thermoreceptors** (cold, hot), **nociceptors** (accept pain).

The tactile sense is perceived by tactile **Merkel's bodies** on fingers tips.

Meysner's bodies on palms, soles, lips, on the end of the tongue are very sensitive to any touch.

Fater-Pachini's bodies in deep layers of skin perceive sense of pressure.

Cold receptors are situated in **flasks Krause's**.

Thermal receptors are located in **Puffin's bodies**.

More fibers react to cold stimuli than to thermal ones.

Pain is accepted by free nervous endings between epidermal cells.

2. Proprioceptors are situated in deep tissues (muscles, joints, tendons). The muscular receptors are variable. The most important of them are nervous – muscular cords. They react to tension of muscles. They are covered by a connective tissue case and are situated intra– and extrafusally between the fibers of striated muscles.

The **Goldie's** and **Matson's bodies** accept joint feeling. They are situated between muscles and tendons.

Besides, there are also osmoreceptors, chemoreceptors, baroreceptors and others.

The impulse is transmitted from the receptor apparatus to the cerebrum by means of nerve fibers. The last are axons of unipolar cells of dorsal root ganglia.

There are 3 types of fibers:

1. Type A – thin myelin fibers, which carry out deep and light touch sense; the speed of impulse transmission by these fibers is 40-60 m/s
2. Type B – myelin fibers, which carry out pain and temperature sense; the speed is 10-15 m/s
3. Type C – without myelin fibers, which carry out diffuse pain sense with speed 1-1,5 m/s.

Classification of sensation

Depending on the special interest of the investigator, sensation may be classified in many different ways. The neurologist, in his search for the location and cause of neurological disease, finds it convenient to classify sensation into: superficial and deep.

Each of the main groups includes different modalities which will be discussed separately. There are different classifications of sensation.

I. Classification, which is based on **the place of originating of stimuli**. According to this classification sensation is divided on:

1. Exteroceptive.
2. Interoceptive.
3. Proprioceptive.

II. Classification, which is based on **biological principle** of originating of sensation. According to this one sensation is divided into:

1. Prototypical (vital, nociceptive, thalamic). This ancient sensation is typical for the primitive nervous system of our ancestors.
2. Epicritical sensation is connected with cortex and it is based on the differentiation of stimuli according to their modality, intensity, localization etc.

III. In clinical practice usually we use classification, which is based on the **kind of stimuli**. According to clinical classification sensation is divided into:

1. Superficial.
2. Deep.
3. Complicated.

S u p e r f i c i a l sensation

This term includes the modalities of light touch, pain and temperature.

1. **Light touch (tactile) sensation** – is feeling of touch, which may be examined by touch of cotton, end of hammer, paintbrush;
2. **Superficial pain** – is a feeling of pain, which may be tested with a corsage pin or pinwheel (acutely or bluntly, pricks or does not prick) (fig.23).



Fig. 23. Examination of feeling of pain.



Fig. 24. Examination of temperature sensation.



Fig. 25. Examination of deep joint sense.

3. Temperature sensation – is feeling of cold or hot, which may be tested by application of glass tubes filled with iced (10°C) and hot (43°C) water to the skin (fig. 24).

4. Trihoesthesia – is a sensation of touch of hair;

5. Hydroesthesia – is a sensation of humidity;

6. Sensation of electrical current;

7. Feeling of tickling.

Deep sensation

This includes joint and vibratory sense and pain from the deep-lying somatic structures, namely muscle, ligaments, fascia, bone, and so on.

1. Joint sense – is a sense of position and passive movements.

2. Vibration sense.

3. Feeling of mass.

4. Feeling of pressure.

5. Kinesthesia.

1. The joint sense (bathyanesthesia) – is a deep sense, which is based on the ability to distinguish position and passive movements in joints.

Position sense or proprioception is tested by gently moving a terminal phalanx – in the lower extremities by vertical movements of the toes and in the upper extremities by similar movements of the thumbs and fingers (fig. 25). Examination of this feeling

is always started from movements in joints of fingers, then – in a carporadial joint and further – in ulna etc. The loss of joint sense, which is called bathyanesthesia, results in disturbance of muscular coordination and is known as **sensitive ataxia**.

Sensitive ataxia is divided on:

- a) static;
- b) dynamic.

Static ataxia in legs may be investigated by means of Romberg's test – patient is asked to stand directly with the extended forward arms and feet together. In case of ataxia difficulty of standing and instability occurs. It is magnified while eyes are closed.

Static ataxia in arms may be investigated by follows: we ask patient to extend forward arms and to place fingers separately. In case of ataxia consensually spontaneous (involuntary forced) movements (pseudoathetosis) in fingers of arms occurs.

Dynamic ataxia in arms may be examined by means of finger-nasal test, and in legs – heel-knee test.

2. The vibration sense (pallesthesia) – may be tested by placing the base of the tuning fork over a bony prominence (it can be back of the hand, feet) during vibration and again when the fork is stopped (silent control application) (fig.26). We must control how many seconds the patient feels vibration of a tuning fork (to the moment when he feels only pressure). Normally in arms it is – 15-20 s, in legs – 10-15 s.

4. The sensation of weight (baroesthesia) – is the ability to distinguish different weights, and it may be examined with the help of small weights, which are put in the patient's palm. Normally the patient distinguishes a difference of weight about 15-20 grams.



Fig. 26. Examination of vibration sense.

The loss of this ability is called barognosis.

5. **The sensation of pressure** – is determined by simple pressing of finger or instrument barethesiometer. The patient should feel pressure of different force and distinguish pressure from touch.
6. **Kinesthetic sense** is a sensation of movement of dermal fold (fig 27).



Fig. 27. Examination of kinesthetic sense.

Complicated sensation (Integrative function of parietal cortex)

The role of the cortex in sensory appreciation is discriminative. Destruction of the parietal cortex does not produce anesthesia for any modality of sensation except as a transitory phenomenon. The basic sensation of

pain, temperature, vibration, and touch are recognizable as such, but the ability to make fine sensory distinctions is impaired over the contra lateral side of the body – facial sensation being least affected for some reason.

1. **Stereognosis (Three-point distinction)** is the ability to identify familiar object placed in the palm of the patient by palpation when the eyes are closed. It is complicated kind of sensation, which is based on the reception of separate properties of object (weight, form, surface, and sizes), synthesis and analysis of all these pro-perties in the cerebral cortex and is particularly related to activity in the parietal lobes. For example, to identify by touch (with the closed eye) a pen, hammer etc.
2. **Graphism** – is the ability to determine figures and numbers traced on the skin with the closed eyes (fig. 28). **Graphesthesia** – impaired graphism is very sensitive indicator of parietal lobe damage.
3. **Localization sense** – is the ability to point an exact place of the stimuli.
4. **Discrimination sense (two-point discrimination)** – tests the ability of the patient to differentiate one stimulus from two. It may be



Fig. 28. Examination of graphism.

examined by Weber's circus. After the patient closes his eyes the doctor puts stimuli by circus branches on either one side or both sides of skin of his body. At first pulling branches together, and then enlarging distance between them. He marks thus on what distance the patient feels two simultaneously put stimuli as two, and on what as one. The test leads are most sensitive of fingers, tough. The results of examine estimate under the special table.

5. Baragnosis – is the impaired ability to distinguish different weights.

Anatomy of Superficial sensation pathways (fig 29).

The way which carries out pain, temperature and part of tactile sense has three neurons.

The first neuron is situated in unipolar cell bodies. The last are located in dorsal root ganglia of the spinal cord and homologous ganglia of the cranial nerves (ganglion intervertebral or ganglion spinalis). Their dendrites are routed on peripherals within plexuses, peripheral nerves. There they are finished in various sensory skin receptors. The axons of these unipolar cells enter the spinal cord through the dorsal roots in a basis of dorsal horns, where they are finished.

The second neuron – the cells of the second neuron are situated in dorsal horns of the spinal cord. The axons create **tractus spinothalamicus**. The axons of these neurons cross the midline through the ventral commissura and go to the opposite lateral funiculus and then run in the lateral **spinothalamic tracts**. These tracts run upwards to the brain stem, where they pass through the oblong brain, the Varoliy's pons, and peduncles of brain and are finished in **nuclei of thalamus**.

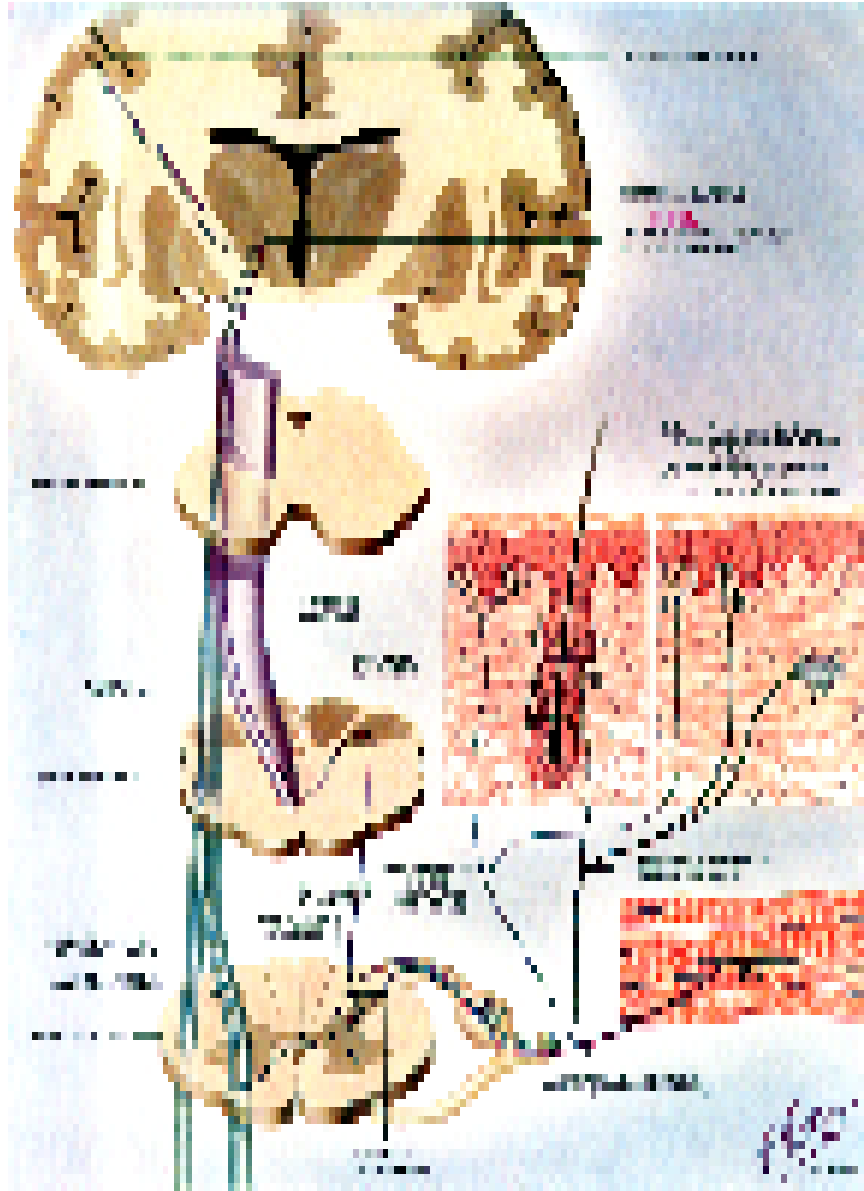


Fig. 29. Somesthetic system, body (Frank H. Netter, M.D.)

The features of spinothalamic tracts, which have diagnostic value:

1. The decussating in front of white matter occurs not in a horizontal plane at a level of segment, but obliquely from below upwards during 1-2 segments. Therefore if we have lesion of lateral funiculus, the sensitive disturbance occurs on the opposite side 1-2 segments below the level of a pathological focus.
2. The caudal contributions to the spinothalamic tract are pushed laterally by the incoming contributions from higher up results in a lamination of the tract, with the fibers from the lower segments of the spinal cord placed more dorsolaterally on each side.

Taking into account this fact it is possible to make the conclusion. In case of extramedullary pathological process (for example, the tumors squeezing a lateral fiber of lateral funiculus of spinal cord) disturbances of sensation will accrue from below upwards (at first on foot, then on leg, thigh, and the trunk, further in an arm (hand)), that is the **ascending** type of sensitive disturbance. In case of intramedullary pathologic process (when first lesion of medial fibers is in lateral funiculus of spinal cord) sensitive disturbance will be distributed from above downwards, that is **descending** type of sensitive disturbance.

The third neuron is located in the nucleus of thalamus. The axons form **thalamocortical tract** and pass through internal capsule, then within radiate crown, and are ended in post central gyrus and parietal lobes of brain hemisphere, and in upper parts of a gyrus – the sensation from lower extremities, on the middle – from upper extremities, in lower – from the face and tongue are ended.

Anatomy of Deep sensation explorers

This pathway has also 3 neurons.

The first neuron The unipolar cell bodies are located in the dorsal root ganglia of the spinal cord and homologous ganglia of the cranial nerves (ganglion intervertebral or ganglion spinalis). The dendrites are routed on peripherals within plexuses, peripheral nerves, where they are ended in various sensory receptors in muscles, tendons and joints. The axons of these unipolar cells enter the spinal cord through the dorsal root and run in dorsal funiculus on one side of the spinal cord, where it divides into two paths – medial thin Holl's

pathway and lateral Burdach's pathway. In Holl's pathways fibers pass from segment Th₄ and below, and in Burdach's pathway, from segment Th₄ and higher. That means the Holl's path carries out deep sense from lower extremities and bottom of a trunk, and Burdach's path – from upper extremities and top of a trunk.

This feature has topical and diagnostic value: at extramedullary processes (for example, in cervical part of spinal cord) the disturbance of deep sense accrue for the descending type, and, on the contrary, at intramedullary processes of spinal cord disturbance of deep sense occurs for the ascending type.

The second neuron is in Holl's and Burdach's nuclear of oblong brain. The axons of the second neuron create bulbothalamic tract. The fibers of this path are crossed on olives level of oblong brain, on the pons of brain stem they join fibers of spinothalamic tract lateral and create a medial closed loop. The medial closed loop (lemniscus medialis) consists of fibers of spinothalamic tract and bulbothalamic tract. The axons of the second neurons carry all sorts of sensation from opposite side of the body. The medial closed loop is ended in ventral nucleus of thalamus.

The third neuron – is in thalamus, from which cells thalamocortical tract starts. The axons of this path go through internal capsule, radiation crown and are ended in a postcentral gyrus, partially in the right central gyrus and in parietal lobes. It is necessary to tell, that a part of fibers from the second neurons of deep sense are routed not to a thalamus, but to a cerebellum through lower legs of a cerebellum.

The part of impulses from muscles, tendons, joints, deep tissues run to a cerebellum (to its worm) after spinocerebral paths. For example, in dorsal horns of spinal cord there are cells, which axons borrow (occupy) lateral funiculus and rise to brain stem as spinoreticular, spinoolivar, spino-vestibular, spinotectal pathways.

Sensation innervation of the face is carried out as follows (fig. 30):

The first neuron of face, nasal sinuses, oral cavities and nose sensory conductors are situated in trigeminal (Gasser's ganglion). Those for larynx mucose membrane, pharynx, radix of tongue – in ganglions of wandering and glossopharyngeal nerves (ganglion superior and inferior). Those for 2/3 front of tongue – in a geniculate ganglion (ganglion geniculi) of Facial nerve.

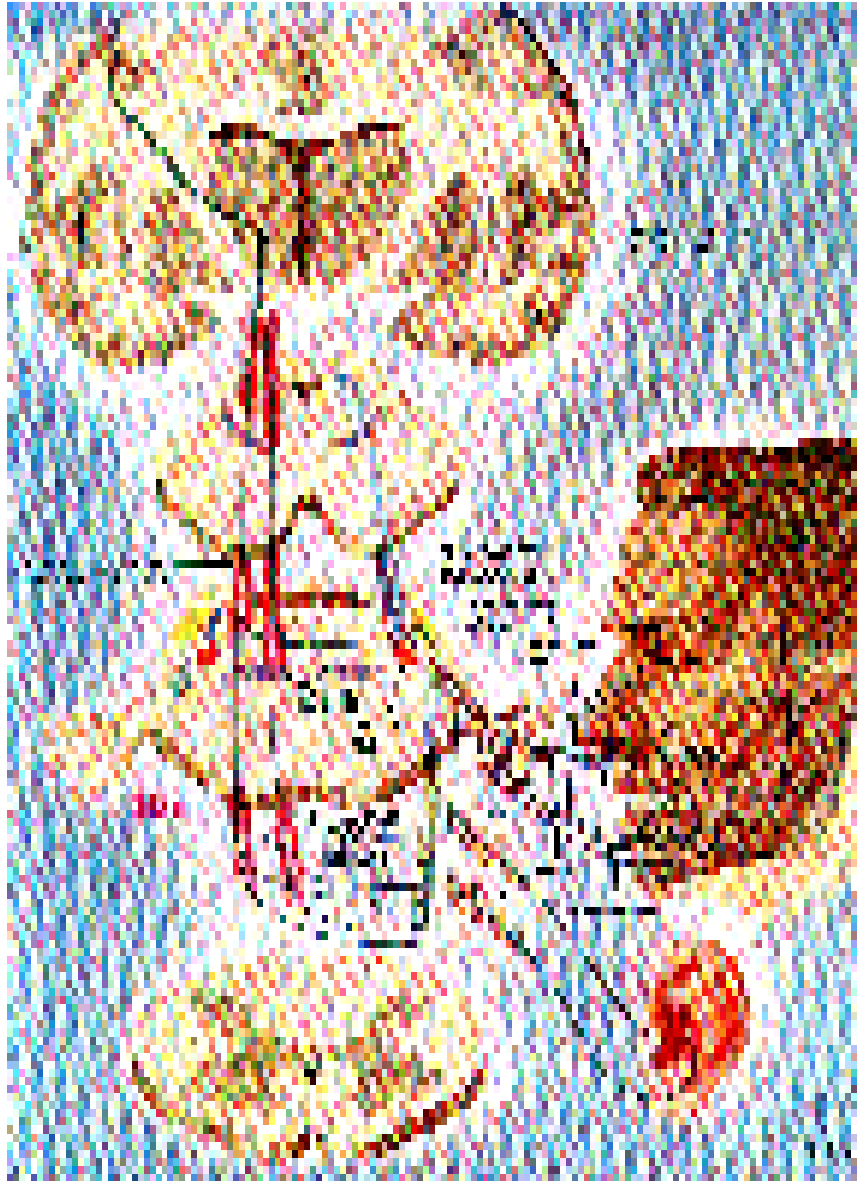


Fig. 30. Somesthetic system, face (Frank H. Netter, M.D.)

The axons of the first neurons as a part of sensitive roots of cranial nerves (V, VII, IX, X) are routed to a brain stem.

nucleus tractus spinalis n. trigemini
nucleus terminalis
nucleus alae cinerea

The second neurons are situated in the sensitive nuclei of the adequate cranial nerves. The axons of the second neuron make decussation and after that join medial closed loop, in which it passes to ventral-lateral nuclei of a thalamus.

The third neuron is in thalamus. The axons form thalamocortical tract, pass through internal capsule, a radiate crown and are ended in lower parts of postcentral gyrus.

Symptoms of sensory disturbances (sorts and types of sensory disturbances)

Depending on qualitative and quantitative changes of sensation in clinic we distinguish the following **objective sorts of sensory disorders**:

1. **Anesthesia** – complete loss of any sorts of sensation. For example:
 - Analgesia** – loss of pain sense.
 - Thermoanesthesia** – loss of a temperature sense.
 - Bathyanesthesia** – loss of deep joint sense.
 - Astereognosia** – loss of stereognostic sense.
 - Topanesthesia** – loss of localization sense.
 - Pallaesthesia** – loss of vibratory sense.
2. **Hypoesthesia** – lowering of sensation.
3. **Hyperesthesia** – sensitization as result of lowering a threshold of energization in cortex of brain.
4. **Dyesthesia** – distortion of sensitivity, when instead of one stimulus the patient feels absolutely other. For example, warm touch one feels as cold.
5. **Hyperpathia** – results from rise of a threshold of energization, when there are strong, unpleasant, badly localized sensations of stimuli. Thus the mild stimuli are not received absolutely. In basis of hyperpathia the disturbance of the analytical function of cortex lays.
6. **Synesthesia** – sensation of stimuli not only in a place of its plotting, but also in the other place.
7. **Polyesthesia** – means sensation of one stimulus as several ones.

8. **Alloheyria** – sensation of stimuli in symmetrical sites on an opposite body part.
9. **Alloesthesia** – sensation of stimuli in the other place.
10. **Dissociation of sense** – phenomenon of fallout of some kind of sensitivity while saving others in the area of segment innervation.

Subjective sorts of sensory disturbances:

1. **Paresthesia** is a creeping sensation, cold, burning sensation, fever, numbness, itch, the pricking etc. Frequently paresthesia is the first sign of nervous system lesion.
2. **Pain.** The pain sensations can arise at stimuli by the pathological process of sensitive analyzers at any level (from receptors up to cortex). Pain is one of the most common complaints to be brought to the physician attention. The initial goal of the neurologist is to ascertain whether the pain represents disease of the nervous system as contrasted with visceral, ischemic, musculoskeletal, or psychosomatic causative factors. The most common pains of neurological origin, except headache, are those that originate from lesions of the peripheral nerves and the spinal roots. Of less frequency but of no less importance are those kinds of pains that reflect dysfunction of the sensory tracts of the central nervous system or thalamus.

Determine the following sorts of pain:

1. **Local pain** – is pain, for example, at palpation of the nervous trunk. That is pain, which coincides with the place of lesion.
2. **Projectional pain** - is a pain in zone of innervation not only in place of stimuli, but also distally on a course of nerves or roots. To projection belongs the stump neuralgia - pain in absent segments of an extremity after its ablation. Or other pain example: during a trauma of a ulna nerve in the field of a ulna joint the pain gives back in V fingers of a paintbrush.
3. **Irradiating pains** – are pains, which are distributed from one nerve branch to another, not struck. For example, at neuralgia of the first branch of trigeminal nerve the pain is distributed to zone of innervation of the second or the third branches, in upper or lower jaw, in ears etc.
4. **Displayed pains** – are pains in zones Zacharyin-Hed's at dis-

Zacharyin-Hed's zones

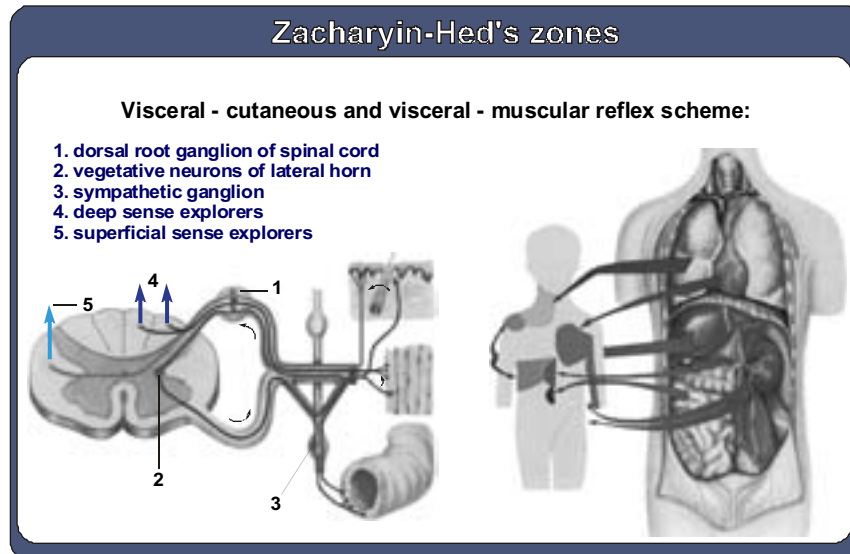


Fig. 31

eases of inner organs, when irradiation arises to certain zone on skin through cells of dorsal horns of spinal cord. For example, pain on ulnar territory of the left forearm and paintbrush at angina pectoris (fig 31).

5. **Causalgia** (Greek causes – burning sensation, algos – pain). It is intensive thermalgia originating, for example, at traumas. It is pain without stimulation.
6. **Reactive pains** – are pains that originate at expansion of nerves. The pains can arise at palpation of pain points and at band spread of nervous trunks.

There is a set of pain points, for example, point of an exit of branches of trigeminal nerve, supraclavicular and subclavian point Herb's for humeral plexus, scapular point Lasarev's, junta spinal points, pain points at palpation of acanthus vertebra, interposes intervals, point Hara's at pressing of transversal processes lumbar vertebrae, point Raymist's – point of lumbar-sacral concatenation, pain points Valle's – on course Sciatic nerve, pain point of Femoral nerve at pressing of middle of pupart sheaf. Signs of tension the following: Laseque's sign, Wassermann's sign, Nery's sign, Matskevich's sign and many others



Fig. 32. Laseque's sign.



Fig. 33. Matskevich's sign.

(fig 32, 33). The pain points and tension signs will be shown to you on practical lesson during learning a special neurology.

Types of sensory disturbances (fig 34).

Determine the following types of sensory disturbances:

1. Peripheral.
2. Segmental (sectional).
3. Conductive.

1. The peripheral type occurs at lesion of dendrites of the first neuron of all sorts of sensation. The peripheral type is divided on:

- a) **Mononeuritic (or neural) pattern** – is observed at lesion of one peripheral nerve and consists of disturbance of all sorts of sensation in innervative zone of this nerve. There is a pain in the field of nerve, sometimes hyperpathia, hyperalgesia, causalgia, tension signs of nerve, pain at palpation.
- b) **Polyneuritic pattern** – is observed at multiple, frequently symmetric lesion of all peripheral nerves. Appears by sensory disturbance in distal parts of extremities as “socks” on legs and “gloves” on arms. The “stocking-glove” pattern of sensory loss is typical for peri-pheral neuropathy. But sometimes cerebral or spinal lesion may cause distal sensory loss, usually of a single extremity in the

Types

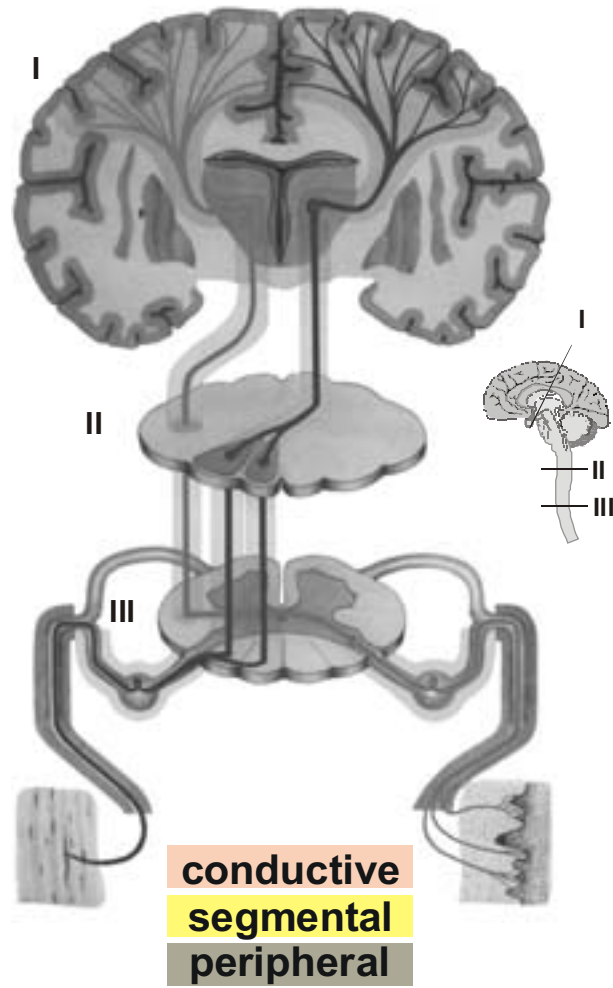


Fig. 34

case of cerebral disease, and often in association with hyperreflexia and the Babinski sign in cases of either cerebral or spinal lesions.

- c) **Plexal pattern** – occurs at lesion of dorsal root ganglia and appears by sensation disturbance in innervative zone of a plexus. In this case there are pains, tension signs of nerves going from a plexus, movement disturbance – peripheral paresis of muscles group, which innervated from this plexus.

It is necessary to point, that at lesion of a peripheral nerve, many peripheral nerves and plexuses, in which near the sensitive fibers pass also movements pathway, simultaneously with sensitive disturbance there are the signs of flaccid paralysis or paresis.

2. **The segmental (sectional) type** disturbance of sensation is observed at lesion of sensitive fibers at segment level of spinal cord, and, means, at lesion of dorsal root ganglion, dorsal roots, dorsal horns of spinal cord, front white soldering.

Subtypes of a sectional type:

- a) Segmental – radicular
b) Segmental – dissociated
- a) **Segmental – radicular pattern** occurs at a lesion of dorsal root or simultaneous lesion of root and dorsal root sensitive ganglion. At lesion of dorsal root there is a loss of all sorts of sensation in its zone innervation according to the segmental type. The sensitive disturbance is appeared as transversal strip on a trunk and longitudinal strip on extremities (in human being there are 36 sensitive segments (31 spinal segments are on trunk and extremities and 5 segments at the expense of trigeminal nerve on the face) (fig 35).

This type of disturbance of sensation arises at radiculopathies, at extramedular tumors. At lesion of dorsal root ganglion occurs herpes exanthema in a zone of innervation of the struck segment (at a ganglionitis or ganglioneuritis) as bubbles (so-called herpes zoster), sharp pains and anesthesia in a segment.

- b) **Segmental – dissociated pattern.** It is observed at lesion of dorsal horns of spinal cord and front grey soldering. Thus the disturbance of sensation appear as loss or lowering pain and thermoanesthesia and saving tactile and joint sense in given segment. Such disturbance are called dissociated and result from that in dorsal horns and front grey soldering pass

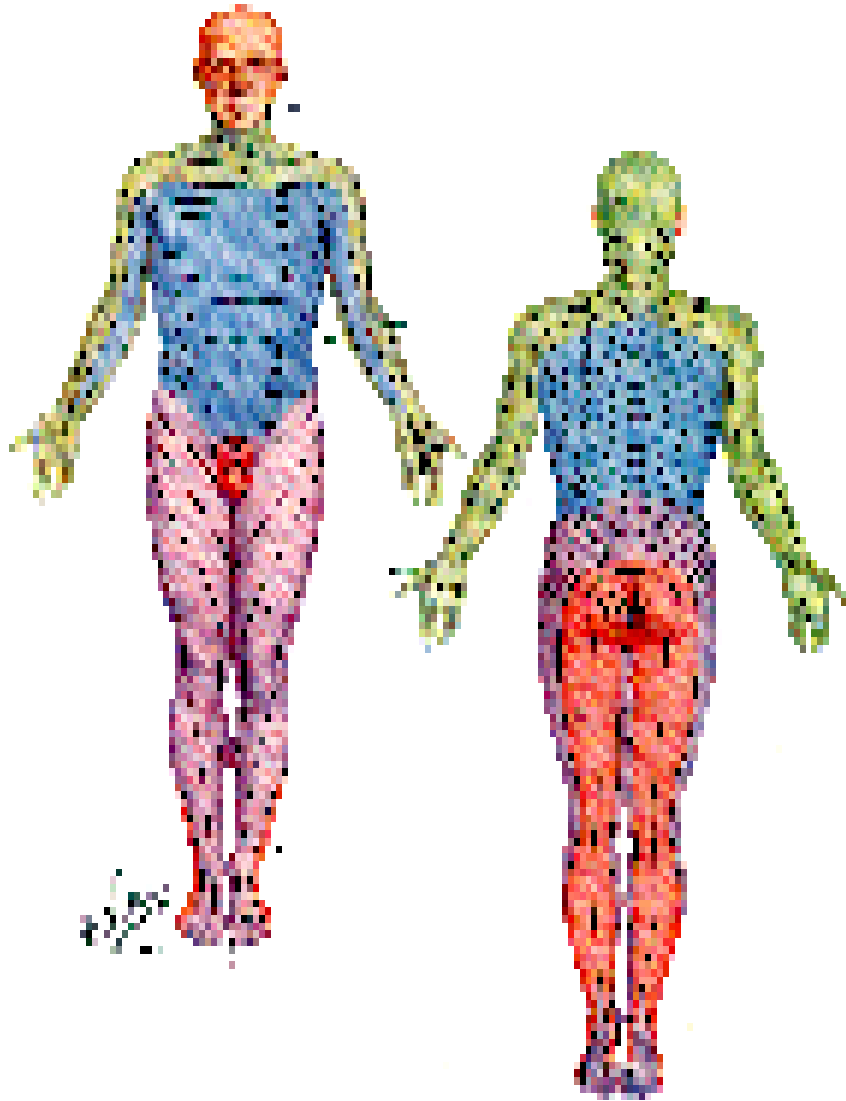


Fig. 35. Dermal segmentation (Frank H. Netter, M.D.)

explorers of superficial sensation, and from the explorers of deep feeling that do not go to a dorsal horn of spinal cord (recollect anatomy).

The dissociated type of disturbance of sensitivity more often arises at a myelosyringosis, when the sensitive disturbance are observed in certain dermatomes as “jacket” or “half jacket” at lesion of dorsal horns of spinal cord in thoracic segments, or “trousers” – at lesion of dorsal horns of spinal cord in lumbar segments.

3. Conductive type.

The lesion of sensory explorers in spinothalamic tract, Holl’s and Burdach’s pathways, bulbothalamic tract, medial closed loop and thalamocortical tract cause conductive type of sensory loss. This type is divided on:

1. Spinal

2. Cerebral

The sensory disturbance from the defined level of a lesion and downwards is typical for both subtypes.

1. Spinal pattern can be:

- a) **Complete transversal** (is observed at a lesion that involves a diameter of a spinal cord, at which all sorts of sensation below that level of a lesion drop out, pain and temperature sense drop out on 1-2 segments below than level of a lesion, and deep – from the same level. Usually, the deficits are in the lower trunk and legs, are bilateral and almost symmetric.
- b) **Half transversal** or **Brown-Sequard pattern** – arises at a lesion of a lateral half diameter of a spinal cord, thus the deep feeling drops out on the side of a lesion, and pain and temperature sense – on the opposite side 1-2 segments lower.
- c) **Descending**
- d) **Ascending**, depending on extra– or intramedular lesion
- e) **Monotype**
- f) **Hemitype**

2. Cerebral pattern is divided on:

- a) **Brain stem pattern** (alternating). At lesion of sensory fibbers in brain stem there is fallout of sensation on the face according to the segmental type on the side of lesion. Lesions in the pons and below in medulla and cord may result in dissociated sensory

loss on one or both sides of the body because of different levels of crossing of the sensory pathways. A lesion of the lateral medulla (lateral medullar syndrome, called Wallenberg's syndrome) will cause loss of pain sensation on the same side of the face and the opposite side of the body. In this situation, touch is preserved in areas where there is loss of pain perception.

- b) **The thalamic pattern** (at a lesion of thalamus) is observed:
1. Hemihypoesthesia of all sorts of sensation on opposite side from the pathological focus.
 2. Hyperpathia – the disturbance of deep feeling prevail
 3. There are thalamic pains (burning, intolerable)
 4. Hemiataxia – as result of lowering deep joint feeling.
- c) **Capsular pattern** – in case of a lesion of sensory fibers in back leg of internal capsule arises hemi anesthesia of all sorts of sensation on opposite sides and hemiataxia owing to fallout of deep feeling.
- d) **Cortical pattern** – arises at a lesion of a postcentral gyrus and upper parietal gyrus. Thus the sensation drops out on monotype in an arm either in a leg, or on the face depending on localization of a lesion in a postcentral gyrus.

The cerebral or thalamic disorders, when the lesions are above the pons, cause sensory disturbances that involve one entire side of the body. This pattern is also found in cases of hysteria, but then the line of demarcation usually is precisely in the midline, whereas in organic deficits demarcation is short of the midline. At **stimuli of a postcentral gyrus** cortical subtype paresthesia appears on the opposite side to the focus, on the face, in arm or in leg. At lesion of parietal share combined and deep sorts of sensation suffer (develops astereognosis or fallout of joint feeling). The syndrome of disturbance of joint feeling can be shown as **afferent paresis**. This syndrome for the first time was described by Fester in 1936. Appears – disturbance of movement functions owing to disturbance of joint sense. Such movement disturbances are characterized by disturbance of coordination of movements, awkwardness, and deceleration of movements. The syndrome of afferent paresis can be tag of a lesion of parietal share of a brain.

Syndromes of lesion of sensory explorers at different levels (fig 36, 37).

1. The lesion of peripheral nerve – appears by fallout of all sorts of

sensation in the field of a nerve, pains, paresthesia.

The pain and paresthesia produced by lesions of the peripheral dermal nerves are usually limited to the region supplied by the affected nerve or nerves.

In lesion of nerves composed of both somatic motor and sensory fibers, corroboration in diagnosis may be afforded by the detection of weakness, wasting, decreased muscle stretch reflex, and electromyography findings of the denervation in the muscles supplied by the affected nerve peripheral to the site of the lesion.

Signs of autonomic fiber involvement may include alteration in sweating, temperature, and distribution of hair.

In peripheral neuropathy, the subjective disturbances are the same as in dermal neuropathy but are confined to the distal portion of the extremities, usually most prominent in the lower limbs. In mononeuritis multiplex, the lesions are disseminated; the several nerves are involved at random.

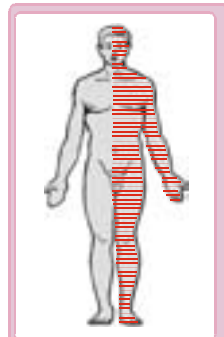
2. The lesion of **plexus** – appears by fallout of all sorts of sensation on one extremity, pains, paresthesia, and vegetative disturbances. Diseases of the brachial or lumbar-sacral plexus are usually associated with pain which may be maximal in the proximal limb with variable extension diffusely or to a portion of the involved extremity.
3. Lesion of **sensory (dorsal) nerve root** – loss of all sorts of sensation for the sectional type in zone of innervation of the certain segment, and also pain, lowering of reflexes.
4. Lesion of a **sensory (dorsal) root and ganglion** – same manifestations + herpes zoster.
5. The lesion of a **sensory (dorsal) horn of a spinal cord** cause the same manifestations, as well as at lesions of sensory (dorsal) root, dissociated disorders of sensation only are observed.
6. The lesion of **front grey soldering** cause sectional – dissociated disorders of sensation symmetric on both sides as “butterfly”.
7. The lesion of **dorsal funiculus of spinal cord** – deep feeling drops out on one side according to the conductive type
8. The lesions of **lateral funiculus of spinal cord** – pain and temperature sense drops out on the opposite side according to the conductive type.
9. The lesion of **half of diameter of a spinal cord**, Brown-Sequard sign – on the side of the focus deep sense drops out according to

Sensory disturbances

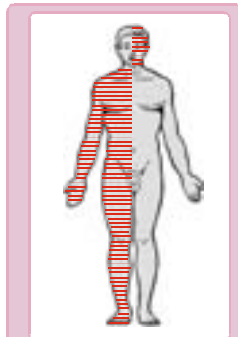
		Focus of lesion	Syndrome
	I		medial closed loop
	II		spinal-thalamic tract, nuclei of superficial and deep sensation of trigeminal nerve
	III		spinal-thalamic tract, nucleus of superficial sensation of trigeminal nerve
	IV		medial loop decussate
	V		lesion of the half of spinal cord
	VI		spinal-thalamic tract
	VII		posterior column of spinal cord
	VIII		anterior grey soldering
	IX		posterior horn of the spinal cord
	X		posterior root of the spinal cord

Fig. 36.

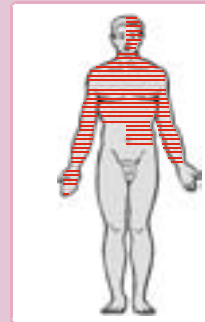
Sensory disturbances



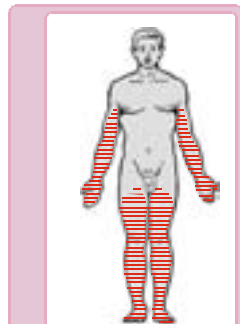
**Capsule
internal lesion**



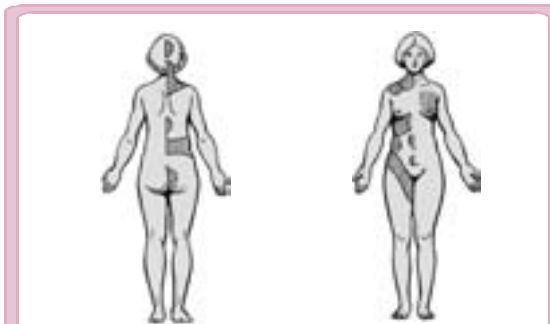
**Brain stem lesion
Alternate type**



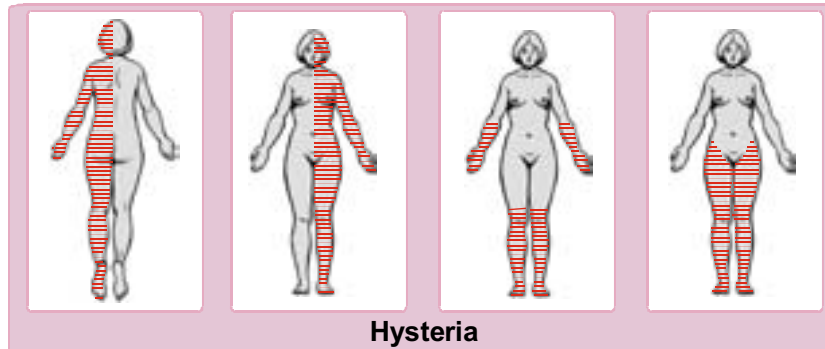
**Lesion of posterior
horns of the spinal cord
and anterior grey
matter
(*"jacket"* type)**



polyneuropathy



Zacharyin-Hed's zones



Hysteria

Fig. 37.

the conductive type from level of lesion. Paresis of an extremity, the zone of an anesthesia at level of lesion and radicular pain, is observed on opposite side – drops out pain and temperature sense 2 segments lower than level of focus.

10. Lesion of **diameter of a spinal cord** – there is anesthesia of all sorts of sensation according to the conductive type lower than a level of focus: deep sense from a level of focus, superficial sense – 2-3 segments lower. Central paralysis. Defective control of the urinary bladder and anal sphincter according to the central type. Trophic disorders.
11. Lesion of a **medial closed loop** – hemianesthesia, sensitive hemiataxia.
12. Lesion of **thalamus**:
 - hemianesthesia,
 - sensitive hemiataxia,
 - hemianopsia,
 - hemialgia.

Since the thalamus is concerned with sensory impulses from the opposite side of the body, the pain resulting from lesions within it is confined to the opposite side of the body. In large lesions the entire opposite half of the body, including the head, may exhibit hyperpathia discomfort. In less extensive lesions the pain may be limited to large contiguous portions of the body, such as the whole lower extremity and lower part of the trunk or the side of the head, upper extremity, and chest.

Characteristically, thalamic pains appear as the patient is recovering from a thalamic infarct. These pains are persistent and are greatly aggravated by emotional stress and fatigue. They are usually described as burning, drawing, and feeling of pulling, swelling, and tenseness, above all they have a peculiar, highly distressing quality.

13. Lesions of sensory pathways in **internal capsule** :
 - hemianesthesia,
 - hemianopsia,
 - hemiplegia.
14. Lesions of **postcentral gyrus** – monoanesthesia on the opposite side.
15. **Stimulations of postcentral gyrus** – sensory “Jackson” - the feeling of tingling or numbness, that appear suddenly with the duration near 30-60 seconds.

Examination of sensation.

On examination ask if the patient is aware of any abnormal sensation or loss of sensation. Paresthesia (tingling, numbness, burning, crawling, coldness, or dead feeling) are common symptoms of disorders of the sensory system at any level. Although the patient should not watch the examination, when you are attempting to determine a zone or level of sensory loss, he should always first indicate the region where sensation is abnormal and where it becomes normal. He may be able to do this with as much accuracy as can result from examination.

If sensation is reduced in some region, start the examination in that region and advance to the normal zone by successive, identically applied pinprick stimuli at intervals of about 2 cm. Ask him to tell you when the pinprick sensation changes or when it becomes normal.

Pain sense is examined with the help of a pin wheel or corsage pin. The examination starts from the segments of face, then cervical segments, the upper chest, and then goes to the hands and feet, comparing sensitivity on the two sides of the body, then distal to proximal areas, and finally upper to lower aspects of the trunk.

Pain and temperature sense are closely associated in the nervous system. Reduction in or loss of either modality has the same meaning, and a deficit in one will usually be accompanied by a deficit in the other.

Test for temperature discrimination Set up the test by putting crushed ice and water in one tube and hot tap water in the other. Apply the cold and hot tubes in irregular alternation, letting each dwell on the skin long enough to register cold and heat. Start with a normal area of reference and frequently check the ability to sense hot or cold there. Ask the patient to report the sensation. Normally, the patient's reports will show quick discrimination and few mistakes. Compare the two sides and various areas, as indicated, for the ability to discriminate cold from hot, much as one test for pain. Rough levels or zones of loss should be estimated.

Vibratory sense and position sense are closely allied functions and are diminished or lost when the posterior columns of the spinal cord are diseased, in cases of peripheral neuropathy and lesions of the brain stem and cerebrum.

Always test first for loss of **vibratory sense** in the hands and

feet and then more proximally when peripheral losses are found. Use a 256-HZ tuning fork. Apply the end of the fork firmly to the dorsum of the great toe or to a distal knuckle of a finger. Ask the patient what he feels. Be sure he feels vibration.

To **test for position sense**, hold the sides of the patient's great toe. Have the patient watch while you demonstrate up and down movement. Then ask him to call out "up" or "down" with eyes closed as you move the toe. Normally a few degrees of movement will be sensed and the direction identified.

Testing for touch is done in a manner comparable to testing for pain. A cotton ball is used, with a small piece pulled out to reduce the area of contact. Apply this to a reference area to acquaint the patient with the sensation. Ask him to close his eyes and say "yes" each time he feels the cotton. Compare the regularity of response on the right with that on the left side and the distal with that on the proximal aspects of the extremities. Then ask the patient if he believes there is any difference in the contrasted areas.

Testing the ability to identify small objects in the hands without visualization is an important part of the sensory examination. Use cotton ball, pencil, key, and so on. The normal person will hold the object with the fingertips, turn it around, follow its contours, rub and manipulate it in a knowing way, and usually give a correct answer. If the patient cannot identify some or all of the objects, the presence of posterior-column disease should be suspected, or, if the patient's difficulty is unilateral and accompanied by little or no other sensory loss, the existence of Parietal lobe disorder is probable.

Graphesthesia refers to the ability to recognize letters or numbers by feeling them being traced on the skin. Generally this is done on the palm. Make the shape of the numbers as well defined as possible. Before testing with the patient's eyes closed, draw several numbers while his eyes are open to make sure that he understands the test.

Two-point discrimination is commonly determined on fingertips and shins. For the fingertips, hold two pins. Touch simultaneously with the sides of the points. Apply one pin, then two held at a certain distance apart, in irregular alternation and ask the patient to report "one" or "two". Normally a distance greater than 5 mm is all that is necessary for the patient to detect that there are two points instead of one. If distances appreciably greater are needed, a sensory defect is suggested. The examiner should measure the distance at which two points can be discriminated from one and thus decide whether normal limits are exceeded.

Extrapyramidal system and Cerebellum. The main anatomic and physiological data Physiological functions and pathologic syndromes at lesion of cerebellum

Motor dysfunction can be caused by lesions of muscles, neuromuscular junction, peripheral nerve or central nervous system.

The pyramidal system, extrapyramidal system and cerebellum are the main components of central nervous system which are involved in motor function coordination.

Extrapyramidal system consists of (fig. 38):

- cortical areas 4, 6, 8.
- the basal ganglia: n. caudatus, n.lenticularis (putamen, globus palidus).
- the nuclei of brain stem (black substance, red nucleus, vestibular nuclei, reticular nuclei, nucleus of Darkshevych, Lues' body, lower olives).
- spinal cord: g-motor neurons and a-small motor neurons, which are located in anterior horns of the spinal cord.

There are **two parts of Extrapyramidal system**:

- pallidum (globus palidus, black substance, red nuclei, vestibular nuclei, nucleus of Darkshevych, lower olives, Lues body).
- striatum (cortical areas 4,6,8, n. caudatus, putamen).

Pallidum is phylogenetically older than striatum. That's why in newborn babies pallidum dominates. And only at the age of 4 – 5 months old striatum starts to influence on motor functions.

The main connections between different parts of Extrapyramidal system:

Pallidum and striatum are closely connected with each other by means of such pathways:

- **nigrostriatal** (dopaminergic) – black substance – nucleus caudatus – it inhibits the neurons of striatum;
- **strionigral** (GABA-ergic)– nucleus caudatus (GABA) – black substance – it controls production of dopamine.

That means inhibition of inhibition. Normally there is a balance between GABA and dopamine.

Extrapyramidal system receives impulse from thalamus by means of such **afferent pathways**:

- T – EPNS – T.
- T – cortex – EPNS – T.
- T – cortex – pons – cerebellum – thalamus.

To the **effluent pathways** of extrapyramidal system belong:

- Tr. olivospinalis;
- Tr. rubrospinalis;
- Tr. vestibulospinalis;
- Tr. tectospinalis;
- Tr. reticulospinalis.

The main functions of Extrapyramidal system are:

1. It prepares muscles to smooth economical movements.
2. It determines the posture.
3. It makes automatical involuntary regulation of conscious movements.
4. It provides automatical stereotyped movements and reflex protective movements.
5. It provides motor manifestation of emotions.

Normal function in the basal ganglia appears to depend on a homeostatic relationship between various neurotransmitter agents – DA and GABA. Disturbances of their homeostasis result in one or another of the symptoms which are generally attributed to this area of the brain. In general, dopamine deficiency allows for cholinergic hyperactivity and can be correlated with the akinetic-rigid disorders such as Parkinsonism. Dopamine hyperactivity and/or cholinergic hypoactivity result in the hyperkinetic phenomena encountered in Huntington's chorea.

Considerable extent disorders are directed to a better understanding of role and reestablishing balance of these neurotransmitter agents.

There are two **main syndromes** of the Extrapyramidal system's lesion: **Parkinson's syndrome** and **syndrome of involuntary movements**.

In 1817 James Parkinson was the first who described the major manifestation of this syndrome. In 1874 this disease was called after James Parkinson – Parkinson's disease. In 1920 Tretiakov was the first who noticed that the greater cell loss in the substantia nigra, the lower

EXTRAPYRAMIDAL SYSTEM

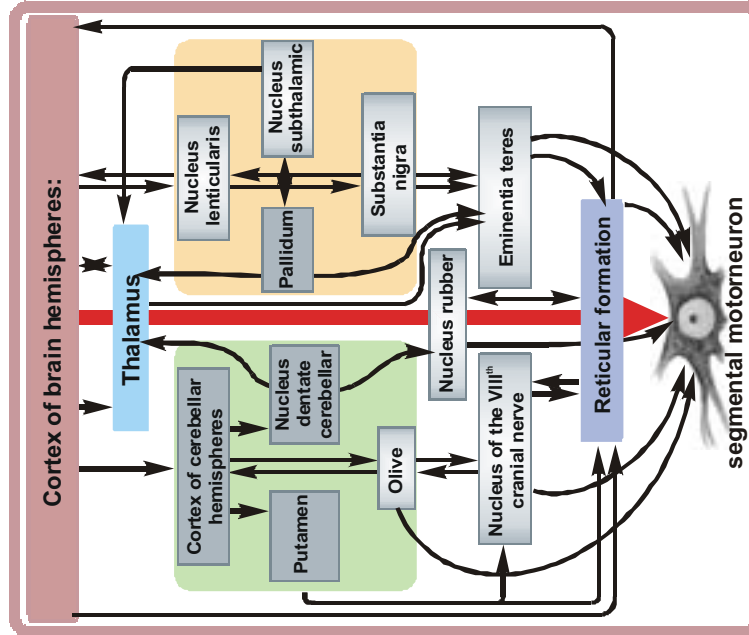
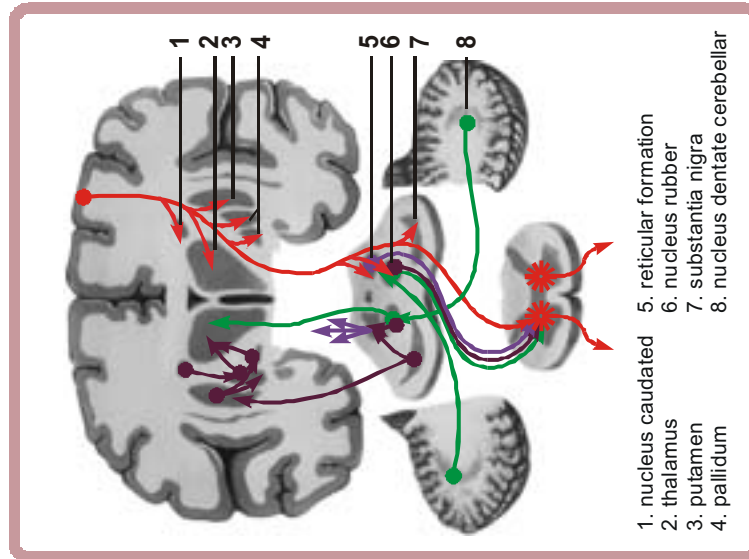


Fig.38.

concentration of dopamine is in striatum and more severe the degree of clinical Parkinsonism. The exact mechanism, however, by which selective damage to substantia nigra occurs, is unknown nowadays exactly. But according to the modern investigation it is considered that in base of this disease is inborn deficiency of tyrosinetransferase enzyme which provides transformation of tyrosine in dopamine.

In the light of these findings Parkinsonism may be defined in biochemical terms as an inborn dopamine deficiency state. Normally, there is a balance between the effects of acetylcholine and dopamine. About 60 – 187 persons per 100 000 population suffer from this disease all over the world. In Ukraine about 133 people per 100 000 population have Parkinson's disease. An average age of manifestation is 45 – 52 years old.

There are three basic symptoms of Parkinson's syndrome:

1. Hypokinesia (Akinesia);
2. Rigidity;
3. Tremor.

The main pathogenetic mechanisms are:

1. Great cell loss in the substantia nigra, low concentration of dopamine in striatum, the influence of striatum on pallidum. As a result akinesia occurs.
2. The main cause of rigidity is increasing of tonic reflex on muscles tension.
3. The main source of tremor is thalamus (its nucleus ventrolateralis).

Hypokinesia (Akinesia).

- The gait and posture are constantly affected as to produce a stereotyped picture: a lack of mobility of facial expression with infrequent blinking of the eyelids, the head and the shoulders are stooped forward, the arms are slightly abducted, the forearms are partly flexed, and the phalangeal joints are extended at the interphalangeal joints.
- Poverty of movement which result from the bradykinetic and akinetic state.
- The gait is shuffling and the steps are slight.
- Parallel footprints.
- The loss of associated swinging of the arm or arms when walking – (acheiropkinesis).
- A lack of mobility of facial expression (Bechterev's symptom).

- Infrequent blinking of the eyelids (Mary's symptom).
- Fixed look.
- Inertia of rest (that means it is very difficult for patient to start moving).
- Inertia of movement (As the patient starts to walk, the movements of lower extremities may be quite slow. As though in an effort to «get going» he may lean forward, causing him to hurry his steps. This results in a shuffling of the feet which may increase in rapidity until the patient is almost running, the so-called propulsive gait. Similarly, a deviation of the center of gravity to one side or backward produce lateropulsion or retropulsion).
- Micrography – handwriting is too small.
- Speech is quite and inexpressive (bradyphasia).
- Paradoxical kynesia is possible after strong impression or great emotions.

Rigidity (plastic type of increased muscle tone):

- Cogged-wheel symptom;
- Tonus increases in course of evaluation of nervous system state;
- Tonus is expressed in the same manner in the group of flexors and extensors.

Tremor:

- Are much more expressed in distal parts of extremities, sometimes tremor of lips or lower jaw can occur;
- It looks like coins counting;
- It is much more expressed while resting. It disappears or decreases while moving;
- Its frequency is 3 – 6 times per second.

Besides these basic features of Parkinson's disease sometimes can occur:

- Bradyphrenia (thoughts are too slow);
- Bradymnesia (recollection is slow too);
- While speaking such patients are boring (akairia – Astwatsaturov symptom);
- Usually they are in a bad mood. Depression is very typical for the patients with Parkinson disease;
- Sometimes they have autonomic disorders. Parasympathetic nervous system dominates in such patients – they have running sa-

liva (aerial symptom), hyperhidrosis, fatty skin and type of hair, bradycardia and arterial hypotension.

At **early stages** of Parkinson disease following tests can be used:

1. The symptom of air pillow or **Vartenberg symptom** – The patient is lying down. One props up his head a little bit and then quickly takes his hand out. Normally the head is falling down. But in patients with Parkinson disease the head stays in the same position for a while.
2. The symptom of **Noica–Haneva** – While evaluations of muscle tone one asks the patient to raise his opposite extremity. In patients with Parkinson disease the tonus suddenly increases.
3. **Westfahl's phenomena** of paradoxical muscle constriction – While foot extension it stays in the same position for a while.
4. Hand extension test.
5. Test of knee flexion – The patient is lying on his abdomen; his lower extremities are bended in knees. In patients with Parkinson disease the legs are fixed in this position for several minutes.

According to the prevalence of this or that symptom there are different **clinical forms of Parkinson disease**:

1. Rigid (hypokinesia dominates);
2. Trembling;
3. Mixed: rigid – trembling or trembling – rigid.

The degree of severity of clinical Parkinsonism (according to Petelin):

The first degree – expressed one or two main symptoms. The patient preserves professional and home activity.

The second degree – The patient is disabled professionally.

The third degree – The patient cannot take care of himself.

Besides Parkinson disease (or **idiopathic Parkinsonism**) there is **symptomatic** one. It is caused by:

1. Craniocerebral trauma (for example, in Muhammad Ally).
2. CO, Mn intoxication.
3. Brain tumor in subcortical ganglia.
4. Encephalitis (Economo).

5. Strokes.
6. Cerebral atherosclerosis.
7. Medicinal parkinsonism (reserpinum, neuroleptics).

Another syndrome of Extrapyrarnidal system, which usually develops at striatum lesion, is **hyperkynetic–hypotonic**. The main **clinical signs** of this syndrome are:

1. Muscular hypotonia;
2. Involuntary movements – hyperkynesis.

All **involuntary movements** are characterized by:

- the amplitude of the movement;
- involved muscle;
- the rate;
- the duration of contraction and the relaxation.

The same kind of the involuntary movement might result from organic disease in one patient and from an emotional disturbance in another (fig 39).

1. Chorea

- spontaneous, irregular, purposeless and asymmetric movements;
- they are present at rest and subside during sleep;
- there are some symptoms, which are very typical for chorea: eyes and tongue symptom (or Hersonsky's symptom) – the patients are unable to maintain tongue protrusion for more than a few seconds; Hordon's II symptom – while checking knee – reflex crus stays in the position of extension for a while and then slowly goes down; Cherni symptom – pathologic sudden abdomen at breath;
- The most common diseases with chorea syndrome are Huntington's chorea (inherited disease), rheumatic subcortical encephalitis or chorea Sydenhams (juvenile disease), atherosclerotic chorea, chorea gravidarum, electric chorea.

2. Athetosis

- movements are slower and more sustained than choreiform movements;
- they affect primarily the distal portion of extremities;
- snakelike movement of any combination of flexion, extension, adduction and abduction in varying degrees;
- they are regularly associated with increased muscular tone;
- it is supposed that athetosis is the result of nucleus caudated lesion.

3. Choreoathetosis

- is a term selected to describe movements that are intermediate between chorea and athetosis.

4. Ballism and hemiballism

- its the more or less continues gross abrupt contractions of axial and proximal muscles of the extremities;
- in the most cases this movement disorder is confined to one side of the body (hemiballism);
- it may be associated with hypotonia.

5. Myoclonus

- is a jerking movement of one or more muscle groups (for example palatine, tongue, pharynx, larynx, diaphragm and skeletal muscles);
- usually only one muscle group is involved;
- they are synchronous in most of cases and sometimes they are asynchronous;
- their frequency is about 15 – 18 per minute;
- they may be induced by visual, tactile, or auditory stimuli (stimulus-sensitive myoclonus) or by the initiation of the voluntary movement (intention myoclonus).
- myoclonus – epilepsy is typical for chronic form of forest spring form encephalitis.

6. Torsion spasm

- twisting or turning movements;
- the muscles of

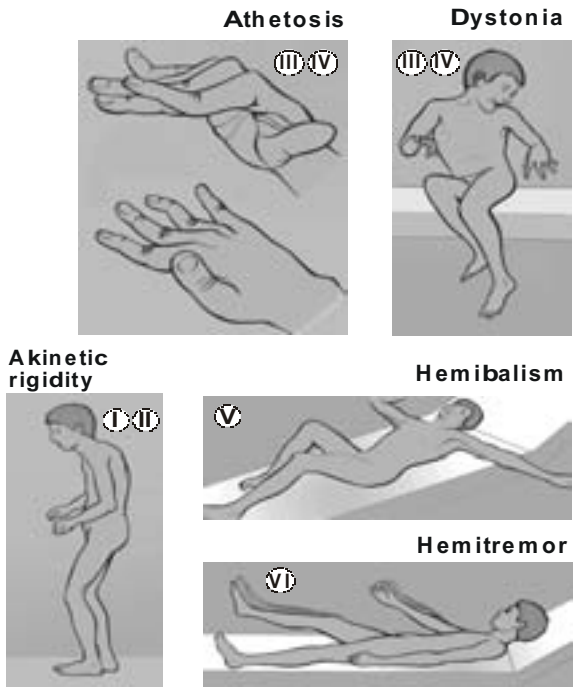


Fig. 39. Kinds of Hyperkinesis

- trunk and neck are involved;
- sometimes torticollis can occur;
 - usually it is the result of putamen lesion.

7. Tics

- are an involuntary compulsive stereotyped movements;
- they may be simple or complex;
- tics may involve any portion of the body (they are most common about the face where they are manifest as blinking, grinning, smirking, lip licking, nose wrinkling).

8. Facial cramp

- is tonic seizure in facial muscles.

9. Tremor

- is rhythmical jerking of arms, legs or head;
- its frequency is about 4 – 6 per second.

Cerebellum

Cerebellum is located in the posterior cranial fossa. It contains two large lateral hemispheres, flocculo-nodular lobe and three pairs of peduncles. Cerebellum is a reflexional organ of coordination of movements, equilibrium and muscular tone (fig 40, 41).

The gray matter of cerebellum is presented by the cortex of hemispheres and nuclei of cerebellum (fig 42).

- Nucleus fastigii – phylogenetically the oldest one, which is closely connected with vestibular nuclei.
- Nucleus globosus, nucleus emboliformis.
- Nucleus dentatus – this one is considered to be phylogenetically young structure.

There are next **layers in the cortex** of the cerebellum:

1. Granular one – it consists of small cells with short dendrites and long axons.
2. Ganglionar one – it consists of Purkinie cells.
3. Molecular one – it consists of basket cells.

The white substance of cerebellum is presented by:

1. Associative fibers, which connect gyres with each other.
2. Commissural fibers, which provide connection between the hemispheres of cerebellum.
3. Projection fibers that provide connection of cerebellum with other structures.

Cerebellum and its communications

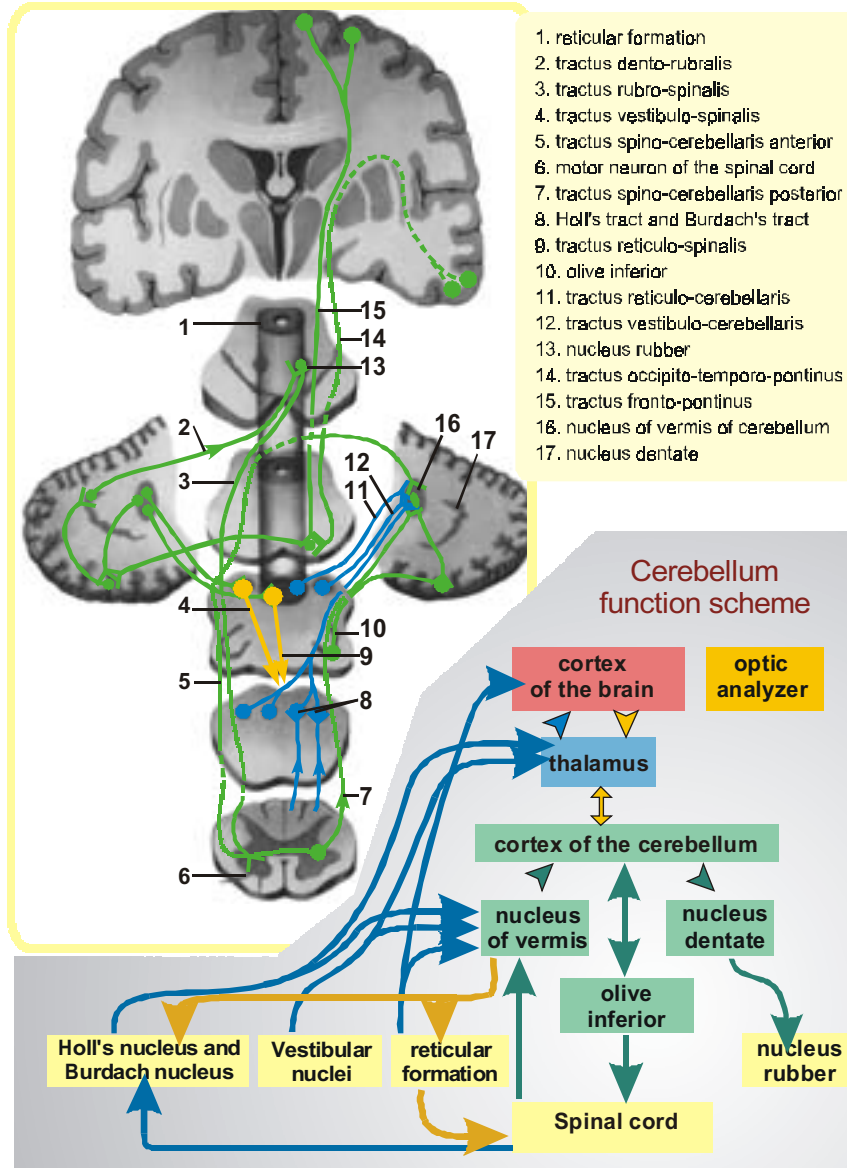


Fig. 40.

Cerebellum is connected with all the other parts of CNS by means of its peduncles (fig 42):

The lower peduncles (corpora restiformia) provide connection with oblong brain and spinal cord:

- Tr. spinocerebellaris dorsalis (Flexig's);
- Tr. vestibulocohlearis (from nuclei vestibularis to nucleus fastigii);
- Tr. olivocerebelaris (from lower olives to nucleus dentatus);
- Fibre arcuate externe (from nuclei Holl and Burdach to hemispheres and vermis).

The middle peduncles (pedunculum cerebellaris medii) provide connection with pons. They are presented by fibers of tr. pontocerebellaris. They connect nuclei of pons with the opposite hemisphere of cerebellum.

The upper peduncles of cerebellum (pedunculi cerebellaris superior) connect cerebellum with middle brain. They include two systems:

- Afferent one – from spinal cord to cerebellum – tr. spinocerebellaris ventralis (Hover's);
- Efferent one – from cerebellum to the structures of extrapyramidal nervous system – tr. cerebellotegmentalis et tr. dentorubralis.

A special attention we should pay to **the way of cerebellum correction**. It consists of six neurons. The impulse runs from motor area of brain cortex to the pons, then some of impulses go to the spinal cord and the others go to the cerebellum in order to confirm information about coordination of movements. Then it runs to the anterior horn.

The first neuron – tr. fronto-temporo – occipito– pontinus;

The second neuron – tr. pontocerebellaris (pontino– cerebellaris decussation);

The third neuron – tr. cerebello – dentatus;

The forth neuron – tr. dentorubralis (Vernekink's decussation);

The fifth neuron – tr. rubrospinalis (Forel's decussation);

The sixth neuron – tr. spinomuscularis.

Thus brain cortex and nuclei rubri are connected with opposite hemispheres of cerebellum. And the segments of spinal cord have homolateral connection.

The main functions of cerebellum are:

- body equilibrium;
- regulation of muscle tone;
- coordination of movements ;
- synergy.

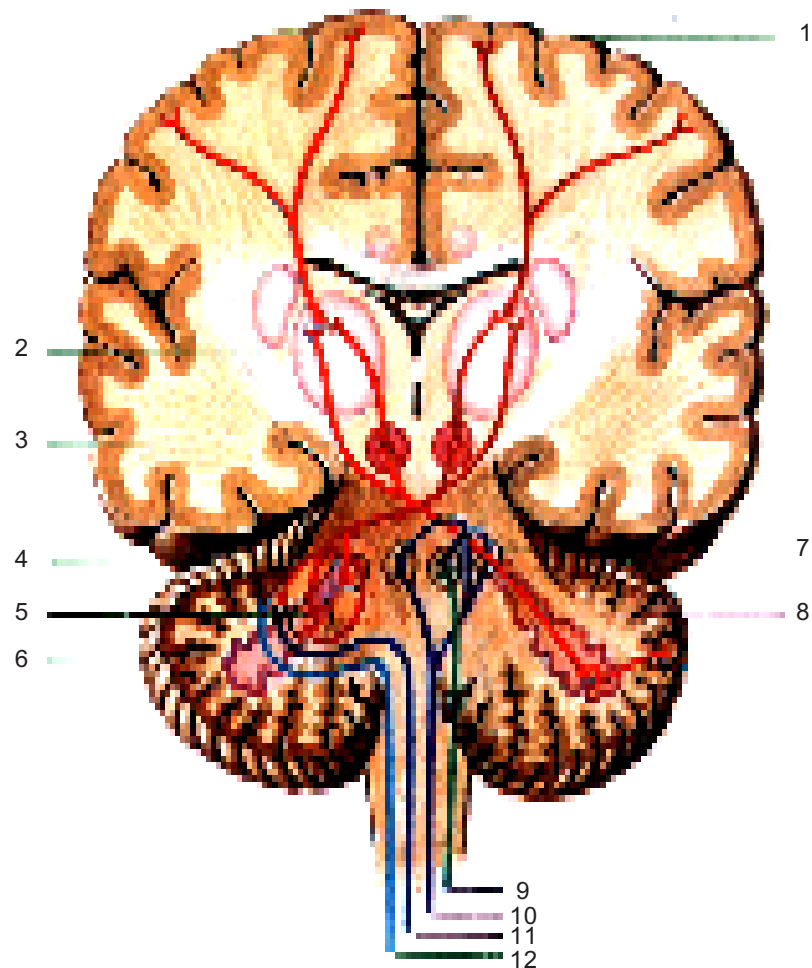


Fig. 41. Cerebellum (Nucleus and Relations):

1. cortex of the brain; 2. ventral nucleus of Thalamus; 3. red nucleus;
4. n. fastigii; 5. n. globosus; 6. n. emboliformis; 7. n. dentatus;
8. tr. dentorubralis and tr. dentothalamicus; 9. tr. vestibulocerebellaris;
10. tr. cerebello-gracilis and cerebello-cuneatus; 11. tr. spinocerebellaris ventralis (Hover's); 12. tr. spinocerebellaris dorsalis (Flexig's)

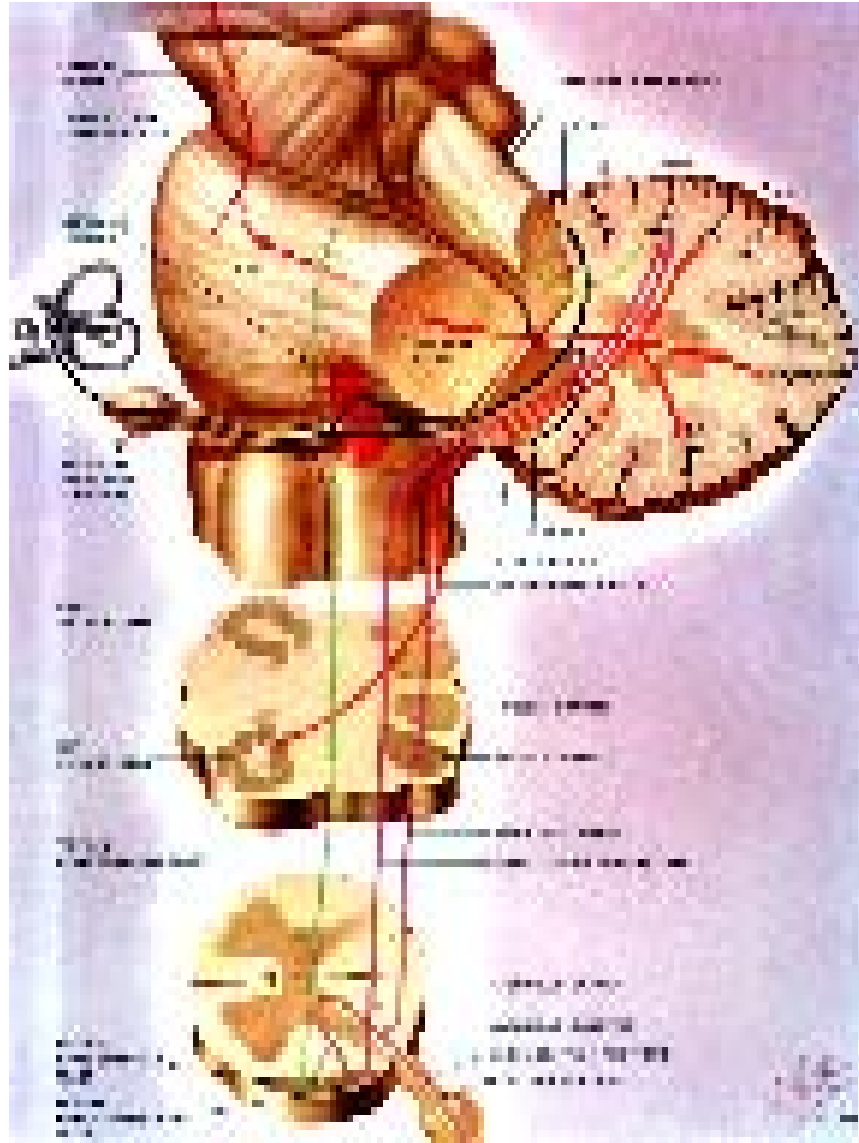


Fig. 42. Cerebellum, afferent pathways (Frank H. Netter, M.D.)

Equilibrium and regulation of muscle tone are the functions of flocculo-nodular lobe (vermix). Coordination of the movement and synergy are the functions of the cerebellum hemispheres.



Fig. 43. Romberg test.

Cerebellum lesion produces **cerebellar ataxia**. There are two kind of ataxia:

1. static one (it develops at lesion of vermix);
2. dynamic (it develops at lesion of hemispheres).

Static ataxia means standing and walking disorders. Usually it is checked by means Romberg test (fig. 43). The **Romberg test** is called positive when unsteadiness is increased by closure of the eyes. As a rule, it is present at such diseases as tabes dorsalis, combined degeneration, or polyneuritis with a

loss of proprioceptive sensation in the muscle of lower extremities.

Dynamic ataxia can be observed while moving. Its main features are (fig 44):

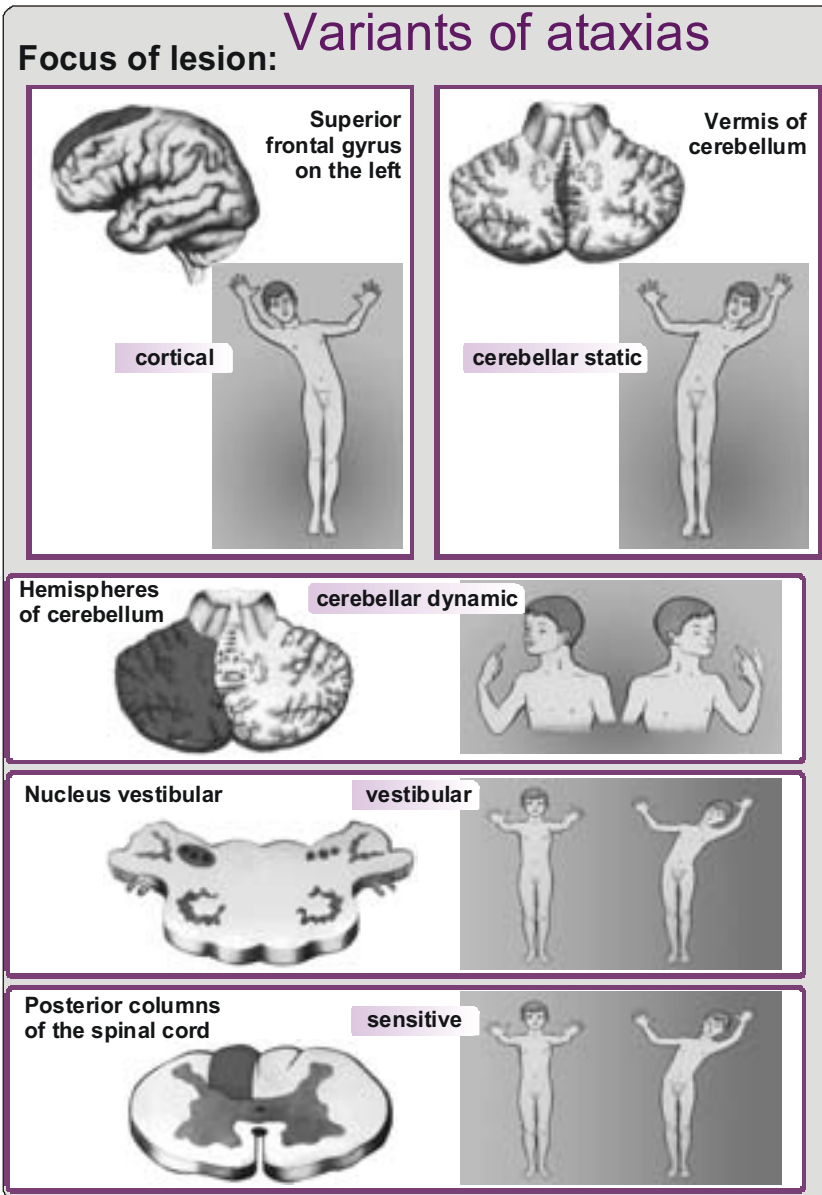


Fig. 44.

1. **Nystagmus** originating in the central nervous system may be horizontal, vertical, vertical rotary, or dissociated (different in each eye). It is rhythmic, having a quick and a slow phase, and the direction of the quick and slow movements depend on the position of eyes. It has long duration, lasting for months or years.
2. Scanning speech.
3. Intention tremor (Finger-Nose Test, Heel-to-Knee Test) (fig. 45, 46).
4. Macrographia.
5. Missing while coordinatory tests checking.
6. Dysmetria (disturbed ability to gauge distances).
7. Muscular hypotonia
8. Adiadochokinesia (disturbed ability to perform rapid alternating movements).
9. Asynergia.

Cerebellar lower peduncles lesion can cause:

- Cerebellar ataxia;
- Bulbar syndrome;
- Sometimes pathology of Holl's and Burdach's nuclei is associated with this lesion. Then ataxia is complex and is called cerebellar – sensitive ataxia.

Cerebellar upper peduncles lesion can cause:

- Cerebellar ataxia at the side of lesion;
- Trochlear Nerve lesion;
- Midbrain lesion symptoms.

Nucleus ruber lesion can cause:

- Cerebellar ataxia in the opposite extremities;
- Resting tremor;
- Webber's syndrome or paresis of convergence may occur.

Pontino – cerebellar angle lesion manifests as:

- Cerebellar disorders and pathology of VII, VIII, V, VI pairs of CNs at the side of lesion;
- Pyramidal and sensory hemisyndrome on the opposite side.



Fig. 45. Finger-Nose test.



Fig. 46. Heel-to-Knee test.

Brain cortex. Localization of functions. Syndromes of brain lesion

Scientists consider that our brain is the most complicated mechanism which has ever been constructed. The weight of the human brain is from one to two kg. It has a volume of about 3.21 litres and consists of about 12 billion cells. Each cell is connected to the other directly or indirectly by nerve fibers. The brain is the centre of a wide system of communication. The nervous cells of the cortex are the most delicate and younger of all the cells of the human body. To estimate the function of different areas of the brain many experiments have been carried out by the investigators.

Anatomically cortex is a plate of grey substance that covers external surface of hemispheres. There are external surface, internal one and base of hemispheres. There is sulcus cerebri centralis on the external surface that separates Frontal and Parietal lobes. There is also sulcus cerebri lateralis that separates Temporal lobe from Parietal and Frontal one. Occipital lobe is separated from Parietal and Temporal one by means of sulcus that continues sulcus parietooccipitalis. Thus there are four lobes on the external surface of brain. They are Frontal, Parietal, Temporal and Occipital one (fig. 47).

External surface

There are three gyri on the external surface of the Frontal lobe. They are upper, middle and lower one. And they are separated by sulcus frontalis superior et inferior.

Parietal lobe is divided by sulcus interparietalis into upper and lower parietal lobes. The lower one contains gyrus supramarginalis and gyrus angularis.

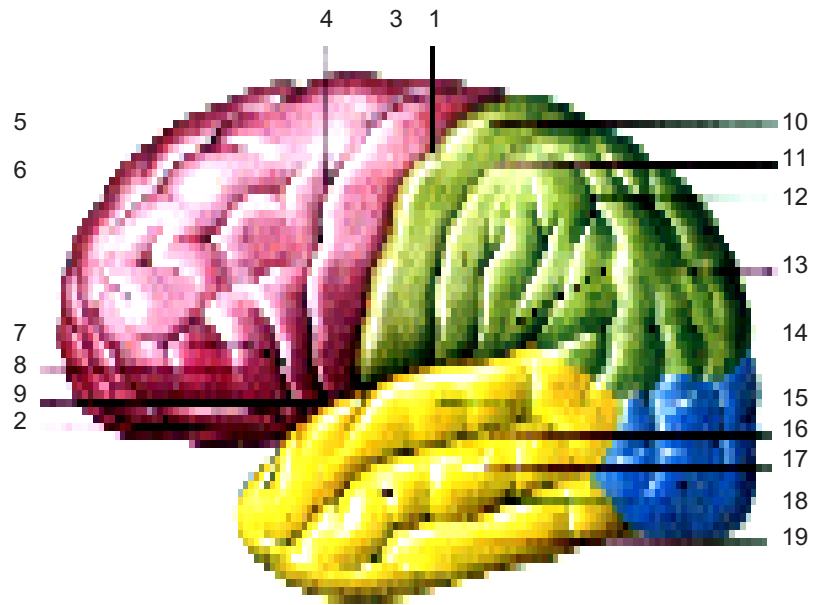
Temporal lobe is divided by sulcus temporalis into upper, middle and lower temporal gyri.

Internal surface

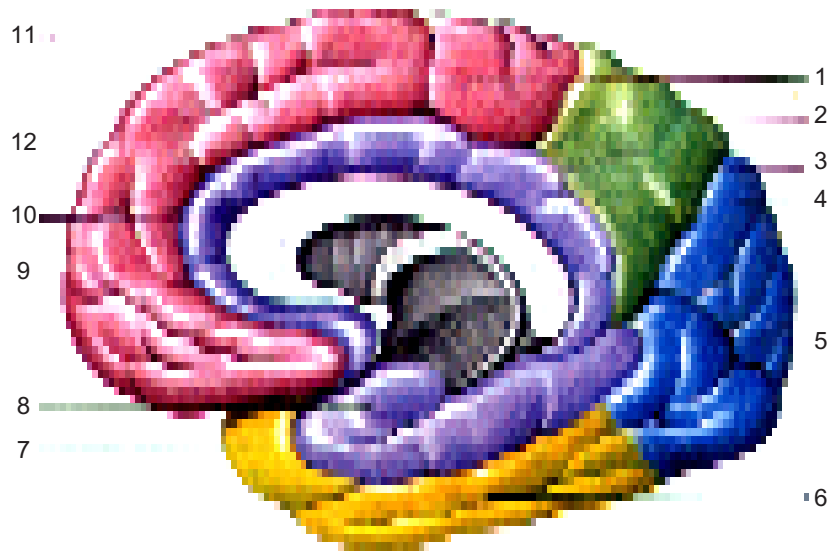
On the internal surface Occipital lobe is separated from the Temporal one by means of sulcus parietooccipitalis. There is Cuneus which is located a little bit upper then fissura calcarina and gyrus Lingualis which is located a little bit lower then fissura calcarina. There is also gyrus Hyppocampi in the anterior part of the temporal lobe.

The basis of hemispheres

There are frontal lobes, occipital and temporal one, bulbus and tractus



A



B

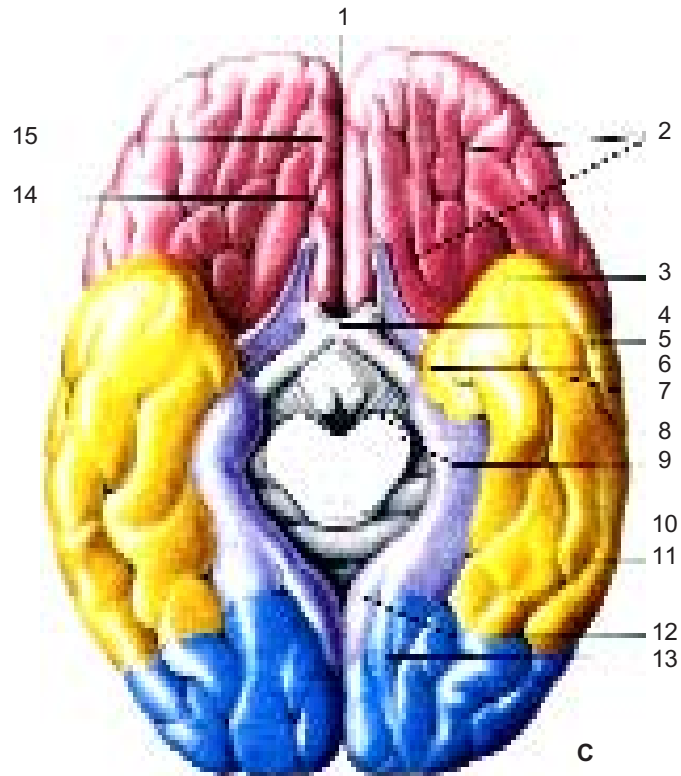


Fig. 47. Brain hemispheres: A. External surface of the left hemispheres: 1. sulcus cerebri centralis; 2. orbital surface of the inferior frontal gyri; I. Frontal lobe: 3. precentral gyrus; 4. sulcus precentralis; 5. frontal gyrus superior; 6. frontal gyrus medial; 7. frontal gyrus inferior; 8. opercular zone of frontal gyri inferior; 9. sulcus cerebri lateralis; II. Parietal lobe: 10. postcentral gyrus; 11. sulcus postcentralis; 12. sulcus interparietalis; 13. gyrus supramarginalis; 14. gyrus angularis; III. Occipital lobe; IV. Temporal lobe: 15. temporal gyrus superior; 16. sulcus temporalis superior; 17. temporal gyrus medial; 18. sulcus temporalis medial; 19. temporal gyrus inferior; B. Internal surface of the hemispheres: 1. paracentral lobe; 2. precuneus; 3. sulcus parietooccipitalis; 4. cuneus; 5. lingual gyrus; 6. parietal-occipital lobe lateral; 7. parahippocampi gyrus; 8. uncus; 9. —; 10. corpus callosum; 11. frontal gyrus superior; C. The basis of hemispheres: 1. central fissure of the brain; 2. sulci orbitalis; 3. Olfactorial nerve; 4. hiasma opticum; 5. sulcus temporalis medial; 6. uncus; 7. temporal gyrus inferior; 8. mammelake bodies; 9. peduncles; 10. sulcus temporo-occipitalis lateralis; 11. parahippocampi gyrus; 12. sulcus collateralis; 13. —; 14. lingval gyrus; 15. sulcus olfactorius.

olfactorii, hiasma opticum, peduncles, pons, oblongate brain and cerebellum.



Fig. 48. Cerebral cortex structure (Frank H. Netter, M.D.)

The microscopic structure of cortex includes six layers (fig 48).

They are:

1. molecular level under the meninges which has a little cortical cells;
2. external level which has a lot of small cells;
3. the level of small and middle pyramidal cells;
4. internal level;
5. ganglion level consists of big pyramidal cortical cells;
6. polymorphic cells level near the white matter .

In cortex of the brain there are 10 or 13 billions of nervous cells. Among them the 8 bill cells have big and middle size (3-5-6 levels) and near the 5 bill have small size (from different levels). Beside the cortex cells, there are many other cells of glia (microgliocytes, oligodendrocytes, astrocytes).

There are **three types of cortex fibers**:

1. **conductive** (for example, Pyramidal tract) – to connect cortex of the brain with other underlying structures of nervous system.
2. **associative** (for example, fibrea arcuatae, fasciculus uncinatus) – to connect different part of the brain cortex together.
3. **commissural** (for example, corpus callosum, commissural anterior) – to connect right and left hemispheres of the brain together.

The main functions of brain cortex:

1. It is the top level of CNS.
2. It provides active movements. It is the source of motor way.
3. Cortex is the structure of conscious reception of sensory stimuli.
4. It is anatomical basis for conditioned reflexes.
5. It is the structure of analysis and synthesis of all stimuli from internal organs and environment.
6. It provides our individual experience.
7. It is the main structure responsible for mental activity and language.
8. It regulates and controls the other structures of nervous system.

Frontal lobe Localization of functions

Frontal lobe provides motor activity, motor mechanisms of speech, behaviour, mental activity and memory of human being.

1. Precentral gyrus provides motor activity of the opposite side of the body.

The projection of certain muscle groups is represented vice

verse to its location on the body:

- The upper part of precentral gyrus – is responsible for the lower extremity;
 - The middle part of precentral gyrus – is responsible for the upper extremity;
 - The lower part of precentral gyrus – is responsible for the muscles of face, tongue, pharynx, and larynx.
2. Posterior part of upper frontal gyrus is the center of straight walking and straight standing. It provides movements of the body and is connected with opposite hemisphere of cerebellum.
 3. Posterior part of middle frontal gyrus is the center of eyes movements in the opposite direction. It is connected with the fasciculus longitudinalis posterior in the brain stem.
 4. The center of writing is located not far from the center of head and eyes movements in the opposite direction.
 5. The center of Broca or the center of motor expressive speech is situated in the posterior part of left lower frontal gyrus.

The main symptoms of lesion and irritation

1. The lesion of precentral gyrus causes central paralysis and paresis. Usually it manifests as monoparesis or monoplegia on the opposite side.
2. The irritation of precentral gyrus causes Jackson's epilepsy in the certain muscular groups (according to the involved part of the brain cortex). It occurs on the opposite side and isn't associated with the loss of consciousness.
3. The irritation of posterior part of upper lobe gyrus causes sudden seizures of the opposite muscles with the loss of consciousness.
4. The irritation of posterior parts of middle frontal gyrus causes movements of head and eyes in the opposite direction.
5. The irritation of opercular part of lower frontal gyrus causes rhythmical chewing, licking and smacking of one's lips.
6. The lesion of anterior parts of upper and middle frontal gyri causes frontal ataxia. The last one manifests as astasia (inability to standing) and abasia (inability to walking without paresis). There is fox-like gait.
7. The lesion of posterior parts of middle frontal gyrus causes gaze into the side of lesion. It is called cortical gaze paralysis.
8. The lesion of posterior part of lower frontal lobe (the center of

Broca) causes Broca's aphasia.

Broca's aphasia occurs in case of motor encoding area involved and means the impairment of spontaneous speech, repetition, reading aloud, naming (but recognizes object) and writing, retained the comprehension of both spoken and written language function. Testing:

1. Speech is slow, nonfluent, produced with great effort, and poorly articulated. There is marked reduction of total speech, which may be "telegraphic" with the omission of small words or endings.
2. Comprehension of written and verbal speech is good.
3. Repetition of single words may be good, though it is done with great effort; phrase repetition is poor, especially phrases containing small function word (eg, "no if's, and's, or but's").
4. The patient always writes aphasically.
5. Object naming is usually poor although it may be better than spontaneous speech.
6. Hemiparesis (usually greater in the arm than in the leg) is present, as the motor cortex is close to Broca's area.
7. The patient is aware of his deficit; he is frustrated and frequently depressed.
8. Interestingly, the patient may be able to hum a melody normally. Curses or other ejaculatory speech may be well articulated.

According to Lurie there are three types of motor aphasia. They are:

1. afferent;
 2. efferent;
 3. dynamic.
1. **Afferent motor aphasia** is associated with the lesion of lower parts of postcentral gyrus which provide innervation of oral muscles. In this case articulation of sounds suffers. That means the loss of automatically speech, repetition, naming. This kind of aphasia is connected with oral apraxia.
 2. **Efferent motor aphasia** is Broca's aphasia. It means it occurs at lesion of the center of Broca. In this case articulation is preserved. But the patient cannot spell some sounds, words and phrases. At completely aphasia the patient doesn't speak at all. At partial aphasia he uses only nouns.
 3. **Dynamic motor aphasia** is usually caused by lesion of cortical zone in front of Broca's center. For this type of aphasia asponantic speech is typical. The patient refuses to speak in active manner. But he is

able to repeat certain sentences, words, answer the questions.

9. The lesion of posterior part of middle frontal gyrus causes : **Agraphia** – the loss of ability to write. The patient with agraphia will be unable to take dictation, to copy letters he sees, or to imitate letters or words that are written for him. Milder degrees of agraphia will show up as poorly formed letters, reversals of letters, and substitutions.
10. Subcortical reflexes can be observed at frontal lobe lesion.
11. Grasp phenomena. The patient grasps everything that touches his palm.
12. Counteraction phenomena. The patient counteracts the doctor in his attempt to change the patient's extremities position.
13. Mental disorders are developed at diffuse lesion of frontal lobe or the lesion of pole of frontal lobe. It is known as **frontal mental disorders**. It manifests as:
 - apathy;
 - torpid mental reactions;
 - slackening of memory and attention;
 - the absence of criticism;
 - inadequate appreciation of disease severity;
 - euphoria;
 - gross humor;
 - the patients are childish;
 - untidiness.
14. Sometimes general hypokinesia can be observed, because of extrapyramidal system lesion.
15. **Frontal apraxia**. It is a disturbance of purposeful movement.

Parietal lobe

The main function of Parietal lobe is associated with space, time orientation and analysis of information from sensory stimuli.

Localization of functions

1. There is nucleus of **general sensitivity analyser** in the postcentral gyrus. Sensitivity from lower extremities is represented in upper parts of postcentral gyrus, sensitivity from upper extremities is represented in middle parts of postcentral gyrus, and sensitivity from face is represented in lower parts of postcentral gyrus.
2. **Complicated kinds of sensitivity** are located in upper parietal lobe. They are stereognosis, sense of localization, discrimination, sense

- of weight, pressure, position of different parts of the body in space.
3. The **center of praxis** is situated in gyrus supramarginalis.
 4. The **center of reading and calculation** are located in the gyrus angularis.

The main symptoms of parietal lobe lesion

1. The lesion of postcentral gyrus causes conductive sensory disorders on the opposite side of the body. They are called monoanesthesia.
2. The irritation of postcentral gyrus causes the attacks of sensory Jackson's epilepsy. That means parasthesia in certain extremities of the opposite side.
3. The lesion of upper parietal lobe on left side causes bathyesthesia, the loss of light touch sense, the loss of sense of localization and discrimination, astereognosis (the patient is unable to identify object by touch. Stereognosis is the ability to identify an object by handling it).
4. The lesion of upper parietal lobe on right side can cause:
 - **Autotopagnosia** (somatotopagnosia) includes disturbances in recognition of the patient's own body or body parts. Autotopagnosia may be limited to one part of the body or may be more general, involving relationships of the body as a whole. the limited form includes lack of recognition of one half of the body or one extremity and inability to recognize and name individual fingers (finger agnosia) or otherbody parts. in mild form the patient may have a sense of distortion of the involvedpartsof the body and disturbances of laterality.this may be tested by asking the patient to name various parts of his own body and to identify right and left.
 - **Pseudomelia** – the patient has a sensation of presence (pseudopolymelia) or the absence (pseudoamelia) of additional extremities.
 - **Anosognosia** is lack of awareness or denial of the existence of disease. An obvious example is a denial by the hemiplegic patient that he is paralyzed.

These symptoms can be observed at right parietal lobe lesion.
5. **Apraxia** occurs at lesion of gyrus supramarginalis lesion of the dominant hemisphere.

Apraxia – is a disturbance of purposeful movements which is not due to elementary motor or sensory impairments. Apraxia is divided on:

1. motor or kinetic;
2. ideational;
3. constructional and dressing;

Kinetic Apraxia The patient has lost the ability to make fine skilled movements such as finger wiggling, opposition, writing, or piano playing.

Ideational Apraxia In ideational apraxia there is inability or failure to comprehend, develop, or retain the concept of what is desired. Such a patient will have difficulty in understanding what is desired and will fail to complete the desired act. This becomes apparent when he is asked to carry out a series of simple acts such as “Put the pencil in the cup and hand me the matches.” After requiring that the request should be repeated, he may pick up the pencil and fail to do anything more. The patient with ideational apraxia has lost the idea.

Ideokinetic Apraxia. Another common form of apraxia is known as “ideokinetic” or “ideomotor” apraxia. This occurs when there is a break in transmitting or converting the idea into the appropriate motor act. Despite knowing what is desired, the patient is unable to carry out a desired complex performance. He may, for example, hesitantly touch his forehead when asked to touch his nose. He may recognize a comb but be unable to use one to comb his hair when asked. Then, surprisingly, he may carry out the same acts automatically that he is unable to perform volitionally on request. Perseveration of a previous motor act is common in such patients. Ideokinetic apraxia generally subsides spontaneously with the passage of time.

Constructional and Dressing Apraxia Constructional apraxia may be demonstrated by having the patient try to copy geometric forms or draw the face of a clock. Further testing includes having the patient try to arrange sticks in a specific pattern or to arrange Kohs blocks in a desired design. Dressing apraxia is tested simply by asking the patient to put on a shirt or bathrobe. To increase the difficulty of the test, one arm of the garment can be turned inside out prior to the test.

6. **Alexia and acalculia** occurs at lesion of gyrus angularis of the dominant hemisphere.

Alexia means that the patient is not able to read, write (optical agraphy). Severe defects of reading with inability to recognize letters or words have been known as **visual verbal agnosia or word blindness**.

Acalculia means one is not able to count. For example, aphasic patients or patients with intellectual deficit from diffuse brain damage frequently have difficulty with arithmetic.

Temporal lobe Localization of functions

This lobe receives information from auditory, smell, taste stimuli and speech sounds.

1. Auditory zone is located in upper temporal gyrus.
2. Smell zone is located in gyrus parahippocampalis.
3. Taste zone is located not far from the smell one.

These zones of each hemisphere have connections with both – right and left receptors.

4. Wernicke center (the center of sensory speech) is located in the left upper temporal lobe.
5. The center of mnemonic speech (the center of naming) is situated in posterior parts of temporal and lower parts of parietal lobes.

The main symptoms of lesion

1. **Wernicke's aphasia** occurs in case of auditory association area involved and means the impairment of comprehension of spoken language, repetition of spoken language, naming comprehension of written language, spontaneous speech, the spoken language is fluent but impaired (jargon). Testing:

1. Speech is fluent with normal rhythm and articulation, but it conveys information poorly because of circumlocutions, use of empty words and incorrect words (paraphasic errors).
2. The patient uses wrong words and sounds –ie, makes paraphasic errors (“treen” for train; “here is my clover” for here is my hand).
3. The patient is unable to comprehend written or verbal speech.
4. The content of writing is abnormal, as is speech, though the penmanship may be good.
5. Repetition is poor.
6. Object naming is poor.
7. Hemiparesis is mild or absent, since the lesion is far from the motor cortex. A hemianopsia or quadrantopsia may be

present.

8. Patients may not realize the nature of their deficit and often are not depressed in the acute stage.
2. **Anomic (nominal) aphasia** is impairment of naming objects and spontaneous speech. Spoken language fluent but rambling and vague. Retained the repetition, comprehension of both spoken and written language.
 1. This type of aphasia may be seen at small lesions in the angular gyrus, toxic or metabolic encephalopathy, or with focal space-occupying lesions far from the speech area, but which exert pressure effects.
 2. Speech is fluent but conveys information poorly because of paraphasic errors and circumlocutions (written language is impaired in the same way). Even though this aphasia is termed anomic aphasia (difficulty in naming objects), anomia is not unique to this type of aphasia.
 3. The patient can understand both written and spoken speech.
 4. There is no hemiplegia.
 5. Comprehension and repetition are normal.
3. **Semantic aphasia** occurs at lesion of temporal-parietal-occipital border of left hemisphere. The patients cannot realize the difference between "The brother of the father" and "the father of the brother".
4. Lesion of the temporal lobe tends to produce **quadrantic defects** which extend to the point of fixation without macular sparing. Lesion of the temporal lobe around the inferior horn of the lateral ventricle give rise to defects in upper quadrants of the homonymous half-fields on the side opposite the lesion.
5. Lesion of the temporal lobe may produce **temporal ataxia**: the attacks of vestibular-cortical dizziness, associated with auditory hallucinations.
7. **Auditory verbal agnosia**. It is rare cases of isolated loss of word recognition. Such patients can read, speak, and write appropriately but are unable to understand or respond to what is said to them.
8. **Smell and taste agnosia**. Patient unable to recognize well-known substances by smelling and tasting them.
9. Irritative lesions of the temporal lobes produce the **auditory, smell, and taste hallucinations**. Usually it is the first symptom of epi-

leptic attack.

10. **Petit mal** is a typical symptom of temporal lobe lesion. Usually it manifests as short lasting loss of consciousness.
11. Development of **sleep like state** is very typical for temporal lobe lesion. In this case it seems to the patient that everything around him is unreal, unnatural, but at the same time it seems to him that he has seen it before.

Occipital lobe Localization of functions

Visual zone is located in the internal surface of occipital lobes in the depth of fissura calcarine. The region above the sulcus calcarinus is called cuneus and it represents the lower quadrants of visual fields. The region below the sulcus calcarinus is called gyrus lingualis and it represents upper quadrants of visual fields.

The symptoms of lesion

1. Quadrant hemianopsia. At cuneus lesion the lower quadrant visual fields are lost. At gyrus lingualis lesion the upper visual fields are lost.
2. Homonymous hemianopsia of opposite visual fields occurs at lesion in fissura calcarina on the internal surface of occipital lobe.
3. Scotoma is a defect within the visual fields.
4. Hemiambliopia – decreased vision.
5. An early symptom of visual analyzer lesion is the loss of color sense in the opposite visual fields.
6. At lesion of sulcus calcarinus seldom cause total blindness. Usually central or macular vision of both eyes is preserved.

The lesion of convex surface of occipital lobe causes next symptoms:

7. **Visual agnosia**: Visual object recognition is simply the ability to recognize objects that are seen. Formal testing may not be necessary if it is apparent that the patient can name and use various objects. If there is any doubt, his ability can be tested by presenting him with one object and having him pick out the matching object from a group; or, for convenience, pictures may be used instead of objects.
8. **Metamorphopsia** means disability to recognize objects' contours. It seems to the patient that they are destroyed, damaged.
9. Irritation of the occipital lobe can cause **visual hallucinations**. Thus **simple hallucination (fotoma)** occurs at irritation of sulci calcarini. They manifest as light or coloured phenomena, for ex-

ample flash, fire, shadow.

10. **Complex hallucinations** occur at external surface of occipital lobe irritation. They manifest as figures, moving subjects. As a rule they are aura of epileptic attack.

Thus for the **right hemisphere lesion** three groups of symptoms are typical:

- body scheme agnosia (autotopagnosia, anosognosia, pseudomelia);
- mental disorders (euphoria, memory impairment, confabulations);
- parakynesis (involuntary movements of healthy extremities – automatic gesticulation).

For the **left hemisphere lesion** the next symptoms are typical:

- aphasia;
- agraphia;
- alexia;
- acalculia;
- apraxia (kinetic, ideational, constructional).

Mental function and Level of consciousness

1. **Amnesia** is memory impairment. It manifests as disability to preserve and restore previous knowledge.
2. **Anterograde amnesia** is a kind of memory impairment that concerns those events which happened just after consciousness disorder or psychic disorders.
3. **Retrograde amnesia** is a kind of memory impairment that concerns those events which have happened before consciousness disorder or psychic disorders.
4. **Somnolence (Grade 1)** The somnolent patient may be roused by various stimuli and will then make appropriate motor and verbal responses. When aroused, such a patient may be clear mentally but often is somewhat confused. Illusions, delusions, hallucinations, or delirium is common in such patients. In somnolence the patient generally drifts back to sleep when the stimulus ceases. Spontaneous movements and spontaneous speech or muttering are usual. The patient may be restless or may show paucity of movement.
5. **Stupor (Grade 2)** In stupor the patient will often have considerable spontaneous movement. He will respond to pain, tactile stimuli,

loud auditory stimuli, or bright lights. The usual response is one of withdrawal, but occasionally a combative response may be elicited. Repeated and persistent stimuli will often rouse the patient to the point where he will respond briefly to questions or follow very simple commands. Spontaneous movements are common, and there may be twitching or picking motions. Control of bowel and bladder is variable.

6. **Semicoma (Grade 3)** At this level organized withdrawal or other. Simple adaptive movements occur in response to painful stimuli. Persistent tactile stimulation or shaking may produce a similar response. Verbal responses are limited to groaning or muttering. As soon as the stimulus ceases, the patient resumes his previous status. Reflex responses are present, but the patient is usually incontinent. Spontaneous movements are uncommon unless the patient is roused.
7. **Deep Coma (Grade 4)** In deep coma the patient makes no response to any stimulus or at most will move slightly to a very painful stimulus. Effective stimuli which may produce a response are limited generally to deep pain. This may be elicited by pressure over the styloid process in the neck, the supra-orbital nerve, the root of the fingernails, or periosteal surfaces of bone. Squeezing muscle bellies or tendons have a similar effect. Care must be observed not to harm the patient while producing the painful stimulus and, if relatives are present, it is wise to explain the need for the procedure. The pectoralis muscle may be squeezed unobtrusively without giving the appearance of harming the patient. The muscle-stretch reflexes, Babinski signs, corneal reflex, and even pupillary responses tend to disappear. There are no spontaneous movements, and the musculature is flaccid. The patient is incontinent of urine and feces. The pulse is usually rapid, the respirations are periodic, and the blood pressure may tend toward shock levels.
8. When confusion becomes combined with illusions, hallucinations, and severe anxiety or panic, the result is **delirium**.
9. **Dementia** is severe psychic processes impairment without involving cheerfulness.
10. **Pseudowakeful States**. Of particular interest clinically are the pseudowakeful states. When this condition is fully developed, the

patient lies or sits with eyes open but fails to follow objects or lights. Similarly he fails to turn his eyes toward a noise. He remains mute. In less marked instances, the patient may follow objects or people slowly with his eyes; he may turn slowly toward a sound and look as if he were about to speak but does not. This appearance has been described as a "*reptilian stare*." The patient responds to external stimuli in a manner similar to that of the patient in stupor or semicoma. Some patients in a pseudowakeful state are restless and hyperkinetic, and pluck at the bedclothes.

11. Some patients in a pseudowakeful state lack spontaneous movements and speech (**akinetic mutism**). The pseudowakeful states may be seen in coma of "toxic" origin or in focal lesions implicating the diencephalon, either primarily or secondarily by pressure.

The autonomic nervous system. Anatomy. Functions. The symptoms of lesion

The autonomic nervous system is a part of nervous system that regulates the activity of internal organs, glands, blood and lymphatic vessels, smooth and striated muscles and organs of sensation (fig. 49).

The modern physiology defines **Autonomic nervous system as a part of nervous system due to which the activity of internal organs and metabolism is regulated.**

There is a close connection and similarity between the autonomic and somatic nervous system.

- The morphological and functional unit of both of them is a neuron
- The main functional unit is a reflex arch.
- The autonomic nervous fibers go within cranial nerves and spinal nerves.

Anatomy

The autonomic nervous system is divided into such parts:

- Central (all the structures that are within the brain and spinal cord)
- Peripheral (all the others structures)

The **central autonomic nervous system** is divided into:

- above – segmental level (the limbic system, reticular formation, hypothalamus);
- segmental level.

The last one according to the structure and functional peculiarities is divided into: **sympathetic** and **parasympathetic** nervous system.

The **peripheral part of autonomic nervous system** is presented by:

- The nodus of Ashof – Towar;
- The plexus of Meisner and Auerbach;
- Sympathetic noduses;
- Solar and hypogastrical plexuses;
- White and gray fibers.

There are two main **functions of autonomic nervous system**:

- Ergotropic (homeokinesis) – adjustment to the changes of environment and providing the needs of the organism.
- Trophotropic (homeostasis) – it supports the constant internal reactions and provides anabolic processes.

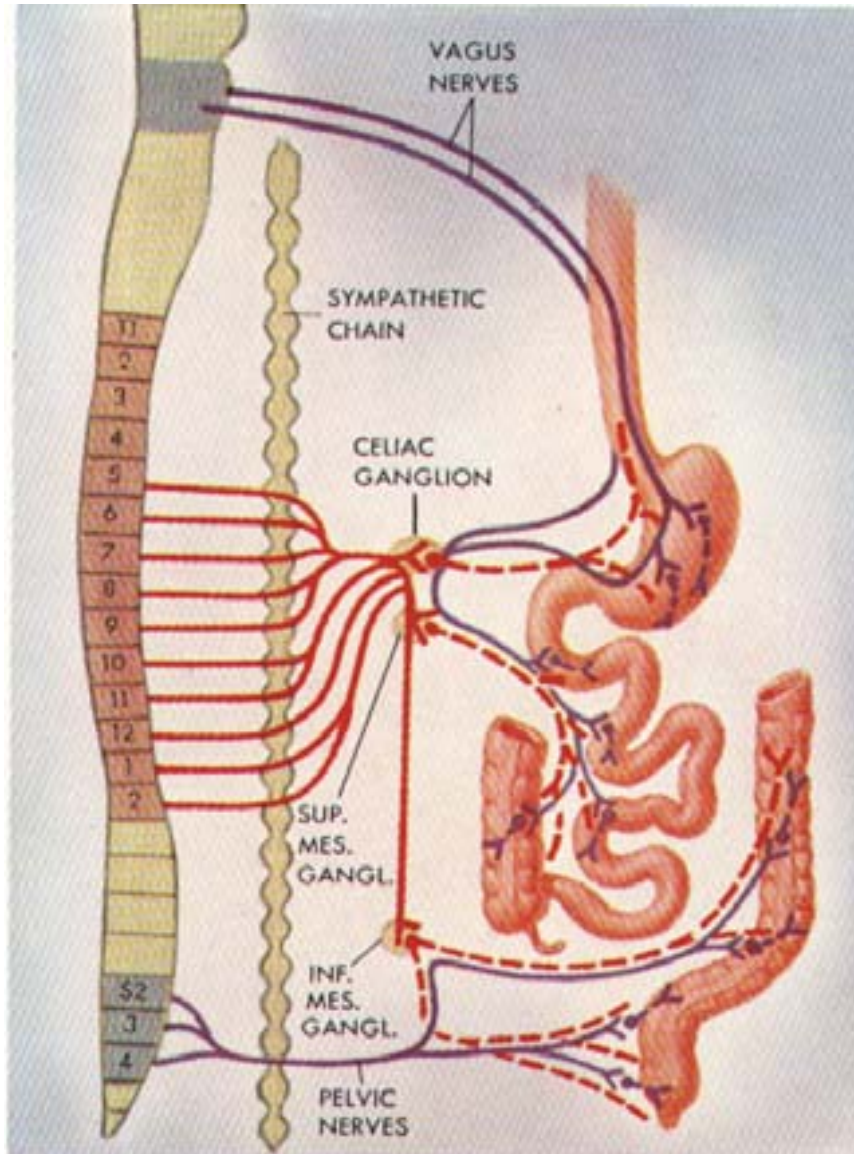


Fig. 49. The autonomic nervous system (Frank H. Netter, M.D.)

The limbic system was described by French neuro – anatomist Broca in 1878. Mc Lein continued its studying in 1952. It consists of:

- Bulbus olphactorius, tractus olphactorius, trigonum olphactorii, substantia perforata anterior;
- Septum pellucidum;
- Gyrus cinguli;
- Gyrus hippocampalis;
- Orbital part of frontal lobe;
- Corpus amygdaloideum;
- The pole of temporal lobes.

Function

1. Emotional reactions.
2. The reception of afferent impulses from internal organs.
3. It is a memory substratum; it preserves information about previous genetically inherited experience.
4. It provides motivation to thirstiness, hunger, sexual desire.
5. It regulates the state of sleepiness and liveliness.
6. It indirectly regulates the function of internal organs.

The symptoms of lesion

1. Emotional disturbances (euphoria or depression).
2. Anorexia or bulimia.
3. Sleeping disorders.
4. Sexual disturbances.
5. Memory disorders.

The lesion of temporal lobe leads to the changes of eating behaviour. For example mice catch and put into the mouth different objects and even snakes in spite of the fact that normally they are afraid of snakes.

The lesion of corpus amygdaloideum leads to increasing of appetite and obesity (the experiment was held on cats).

Hypersexuality was noticed in experiments in spite of the animals' family and sex.

The lesion of gyrus cinguli is associated with memory disorders especially on current events. All these events are forgotten in 2 – 3 min. But those events that are associated with emotions, visual, smell, auditory influences are much more stable.

The irritation of some structures leads to aggression.

Reticular formation

It is a tonus motor of the brain, which works constantly in the brain stem. It was described by Deiters, Bechterev, and Ramon & Kachal. It consists of great number of cells, the axons of which are going in different directions and create a reticule.

In spite of the other cells of nervous system the cells of reticular formation accept pain, light, temperature and humoral impulses and send them to the brain cortex.

Thus **the main functions of reticular formation** are:

- To support the brain cortex tonus, its state of liveliness which is necessary for the normal activity. The reticular formation energizes the brain.
- It supports the certain level of activity of autonomic centers (the activity is very similar to that of sympathetic nervous system. There is also the same mediator – noradrenalinum).

Function

1. The control of sleepiness and liveliness.
2. To accept the information from the environment.
3. To keep in tonus all the forms of behaviour, those have long – lasting character.

The symptoms of lesion

- The low activity of RF – the patient is unconscious.
- Decreased activity of RF – sleepiness.

The lower parts of RF have general and long – lasting influence on consciousness and behavior, the upper ones have short – lasting and specific influence.

Hypothalamus

Hypothalamic region has 32 pairs of nuclei of cranial nerves. They can be divided into three groups.

- Anterior – it is associated with parasympathetic function;
- Middle – endocrine – trophic;
- Posterior – has mainly sympathetic influence.

The peculiarities of activity:

1. Motor cortex receives 440 capillaries per 1 mm³, visual cortex – 900/mm³, hypothalamic region – from 1650 to 2600/mm³.
2. Almost all the arterial brain systems give their branches to hypothalamic region. That makes impossible disturbances of its activity.

3. There is no space of glia between the vessels and gangliocytes. That provides quick reaction to the changes of internal surroundings.

Hypothalamus is closely connected with cortex, thalamus, extrapyramidal nervous system, nuclei of brain stem and spinal cord, reticular formation and hypophysis.

The function of hypothalamus

1. Regulation of heart – vascular activity.
2. Regulation of lipid, water, mineral metabolism.
3. Thermoregulation.
4. Regulation of vessels' and tissue membranes penetrance.
5. Regulation of endocrine glands' function.
6. Constant internal surroundings support.
7. Adaptation.
8. Biorhythm.
9. Emotional behaviour.

Hypothalamus rules the internal world by means of **three ways**:

- Through the nervous impulses.
- Humoral.
- Hormonal.

The very important role belongs to the releasing factors. In hypophysis there are **7 tropic hormones** that activate the production of certain hypophysis hormone:

- Corticoliberinum.
- Tirioliberinum.
- Luliberinum.
- Foliliberinum.
- Somatoliberinum.
- Prolactoliberinum.
- Melanocytoliberinum.

There are **3 inhibiting hormones**:

- Prolactostatinum.
- Melanocytostatinum.
- Somatostatinum.

Hypothalamic syndromes are those that are associated with the lesion or deficiency of hypothalamus.

1. **Autonomic – vascular – visceral** – is associated with crisis of paroxysmal character.

- Sympathetic – adrenal..
 - Vago – insular.
 - Mixed.
2. **Neuro – endocrine – metabolic** – is associated with increasing or decreasing of hypophysis function (Itsenko – Kushing, acromegaly, early climax, impotence, non sugar diabetes, tyreotoxicosis).
 3. **Neuro – trophic** is associated with trophic disturbances (dryness, neurodermitis, ulcers, bed sores, acute perforates of stomach and esophagus) (fig. 50).



Fig. 50. Neuro-trophic disturbances.

4. **Neuro – muscular** – hypothalamus provides chemical and biochemical activity of extrapyramidal nervous system and cerebellum.
 - Myasthenia.
 - Myotonia.
 - Paroxysmal myoplegia.

5. **Thermoregulation disturbances** – the temperature is 37,1 – 37,5, there is asymmetry under the arms, in the mouth and in rectum. Sometimes this symptom has paroxysmal character and is associated with trembling.

6. **Sleeping disorders**

- Insomnia.
- Lethargy (a special form – narcolepsy) – sudden attack of sleepiness that can happen in any place and position of the patient. Sometimes they are associated with catalepsy (the loss of muscle tonus).
- Sleeping inversion.

Parasympathetic nervous system is presented by:

- Mesencephalic level (nuclei of Perle and Yakubovich), the fibers are going within the III CN and provide innervating of m. Sphincter pupillae, m. Ciliaris.
- Bulbar level (n. salivatorius superior et inferior, n. dorsalis nervi Vagi) within VII, IX, X CN's innervate parotid, sublingual, submandibular glands and internal organs (except the pelvic organs).
- Sacral level – the cells of lateral horn S3 – S5 – innervating of pelvic organs.

There is the arch of pupil reflex on mesencephalic level. The symptoms of lesion are:

- Spasm or paralysis of accommodation.
- Mydriasis.
- Direct or indirect symptom of Argil – Robertson.

The symptoms of lesion of bulbar level:

1. Salivation or xerostomy..
2. Tears or xerophthalmia.
3. Dyspnoea, Biot, Chein – Stocks types of breathing.
4. Tachycardia, arterial hypertension, arrhythmia, asystoly.

Sympathetic nervous system consists of the cells of lateral horn of spinal cord from C8 to L2. The axons within the anterior roots leave the spinal cord. Some of them are finished in sympathetic trunk (it consists of 20 – 23 nodules) – 3 cervical, 10 – 12 thoracic, 3 – 4 lumbar, 4 pelvic. The rest fibers are going to the prevertebral nodules or plexuses.

Common features:

1. There are 2 neurons. The second neuron is located in ganglion.
2. Preganglionic fibers are myelin – associated.
3. Postganglionic fibers are without myelin.

Differential features:

1. Mediator

Sympathetic nervous system – adrenalin, noradrenalin

Parasympathetic nervous system – acetylcholine

2. The length of fibers

Sympathetic nervous system – short pre– and long postganglionic fibers

Parasympathetic nervous system – long pre – and short postganglionic fibers

Symptoms of lesion of segmental part of autonomic nervous system:

1. The lesion of **Lateral horns of spinal cord:**

- Autonomic– vascular disorders (pale, cyanosis);
- Trophic disorders (edema, arthropathy) (fig 51);
- Secreting disorders (dryness of the skin, hyperhidrosis).



Fig. 51. Neuro-trophic disturbances.

The zone of innervation of lateral horns of spinal cord:

C8 – Th3 – head and neck;

Th4 – Th7 – shoulders and arms;

Th8 – Th9 – body;

Th10 – L3 – pelvis and legs.

2. The lesion of **Segments of the spinal cord – S3 – S5:**
 - Retention of urine;
 - Truly urine incontinence;
 - Ishuria paradoxa.
3. The lesion of **Upper cervical sympathetic nodes:**
 - Causalgia;
 - Paresthesia, hypesthesia;
 - Vasomotor, secreting and trophic disorders in head and neck region;
 - Horner syndrome.
4. The lesion of **Nodus stellatum:**
 - Causalgia;
 - Paresthesia, hypesthesia;
 - Vasomotor, secreting and trophic disorders in head and neck region.
5. The lesion of **Upper thoracic noduses:**
 - Cardialgia;
 - Tachycardia;
 - Breathing disorders plus all the above named symptoms.
6. The lesion of **Lower thoracic and lumbar noduses:**
 - Visceral and autonomic disorders of the organs of abdominal cavity.
7. The lesion of **Solar plexus:**
 - Dull pain in the abdomen;
 - Increased aorta pulsation;
 - Instable arterial pressure;
 - Instable stool;
 - Poli – oligouria;
 - Glucosuria.
8. The lesion of **Posterior neck sympathetic nodus:**
 - Neck pain like “casque putting off”;
 - Photopsia;
 - Vestibular syndrome.

The disturbance of vegetative functions:

1. The paroxysmal signs:

Sympathy-adrenal attacks:

- a) skin is pail;
- b) xerostomia;
- c) dryness of hair and skin;
- d) tachycardia;
- e) high blood pressure;
- f) midriasis and widening of eye-slit;
- g) exophthalmia;
- h) tremor;
- i) gooseflesh;
- k) diarrhea.

Vagoinsular attacks:

- a) hyperemia;
- b) hyperhidrosis;
- c) oily skin and hair;
- d) bradycardia;
- e) low blood pressure;
- f) miosis;
- g) angina pectoris;
- h) salivation;
- i) breathlessness;
- j) abdominal spastic pain;
- l) frequent and abundant urination

2. The signs of lesion:

- | | |
|--|-------------------------------------|
| a) peri-arthritis; | a) incontinence of urine and feces; |
| b) epicondylitis; | b) ischuria /retention of urine; |
| c) miositis; | c) eye accommodation paralysis; |
| d) hyperkeratosis; | d) midriasis; |
| e) fissures of skin; | e) breath; |
| f) arthropatias; | f) dyspnea; |
| g) trophic ulcer; | g) apnea; |
| h) alopecia; | h) cardiac arrhythmia; |
| i) hyperpigmentation; | i) asystolia; |
| j) Horner's syndrome:
(ptosis, miosis, enophthalmia). | j) collapse. |

Methods of investigation of autonomic nervous system

There are numerous clinical, laboratory and instrumental methods of investigation of ANS. The choice of the method depends on our aim.

First of all we start with investigation of present state of ANS. For this purpose different tables are used.

Normally there is a balance between the sympathetic and parasympathetic nervous system. But it can be changed in different periods of our life.

Cerebrovascular diseases. Dynamic cerebral blood circulation disturbances

1. Epidemiology of cerebrovascular diseases

Among all the neurologic diseases of adult life the cerebrovascular ones clearly rank first in frequency and importance. At least 50 % of all the neurological disorders are of this type. The prevalence of strokes all over the world is about 1.5 – 7.4 per 1000 of the general population. In Ukraine it is 3.27 per 1000 of the general population. Cerebral stroke is the commonest vascular disease of the brain. Every year there are about 6 mln cases of stroke: among them – 1 mln in Western Europe, about 500 000 in the United States, 300 000 – in Russia, 200 000 – in Japan and 130 000 – in Ukraine.

Stroke after heart diseases and cancer is the third commonest cause of death according to the World Health Organization data.

During the last 15 years there has been a marked enlargement in the prevalence of stroke in Ukraine in about 3.4 times and in incidence in 3 times approximately.

2. The main reasons and risk – factors of cerebral blood – circulation disturbances.

There are a lot of reasons and risk – factors of cerebral blood – circulation disturbances. But the commonest are 3 of them:

1. Atherosclerosis of cerebral vessels and general atherosclerosis. In 75 % of all cases it is the main reason of all acute cerebral blood – circulation disturbances.
2. Hypertension – the frequency of hypertension in case of stroke is about 72 %.
3. Combination of atherosclerosis and hypertension.

Besides these main reasons there are some others which can cause cerebral blood – circulation disturbances.

4. Symptomatic arterial hypertension (for example caused by kidney diseases).
5. Heart diseases such as inborn and acquired cardiac abnormalities, arrhythmias, IHD, atherosclerosis, cardiosclerosis, angina pectoris, myocardial infarction.
6. Infectious and infectious – allergic vasculitis (at rheumatism, connective tissue diseases, lues).

7. Arterial hypotension.
8. Vasomotor dystonia.
9. Blood diseases (polycythemia, leucosis, haemophilia).
10. Kidney diseases.
11. Endocrine diseases (diseases of thyroid gland, pancreas, supra-renal glands).
12. Diabetes mellitus.
13. Toxic lesion of vessels at endogenous and exogenous intoxication (at acute and chronic kidney or liver failure, alcoholic intoxication).
14. Traumatic lesion of vessels (at hemorrhage – subdural, epidural, ventricular, parenchymatous).
15. Artery and vein compression (especially in cervical part of spinal cord – for example at osteochondrosis).
16. Inborn and acquired Willis circle abnormalities – occlusion, stenosis of magistral arteries of head (MAH) and neck, aneurism, constriction of vessels.
17. Brain tumors.

For these reasons' realization they should be combined with risk – factors, such as:

- Age (the elder person is, the highest risk of cerebrovascular disease is);
- Sex (at the age of up to 55 – 60 years fatality from cerebrovascular disease is higher in men, after 55 – 60 years – it is higher in women);
- Heredity (in particular to heart and cerebrovascular diseases);
- Alcohol abuse;
- Cigarette smoking;
- Hyperlipidemia and hyperglycemia;
- Arterial hypertension;
- Hypodynamia;
- Meteorological dependence (especially people with labile autonomic nervous system);
- Personal type (picnotic type), stress, high carbohydrate diet.

Combination of three and more factors increases risk of development of acute neurological deficit (fig. 52-54).

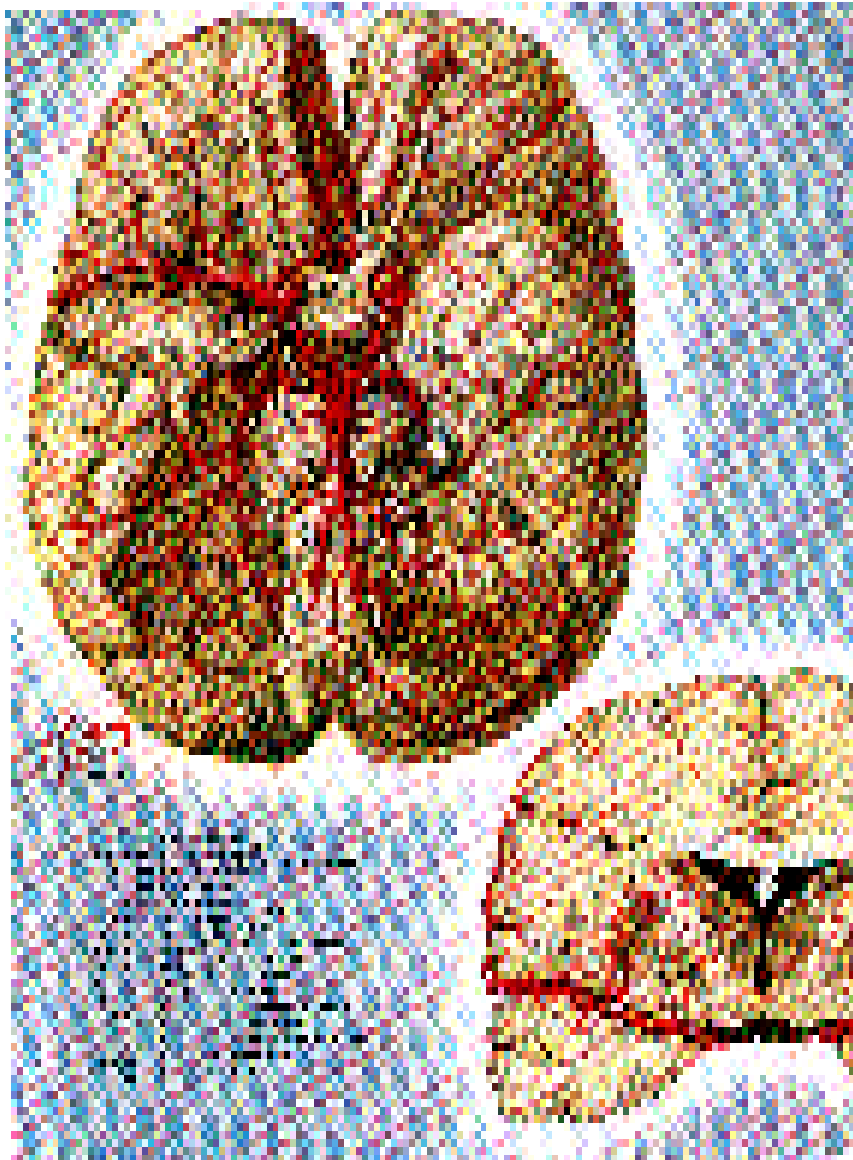


Fig. 52. Blood supply of the brain (Frank H. Netter, M.D.)

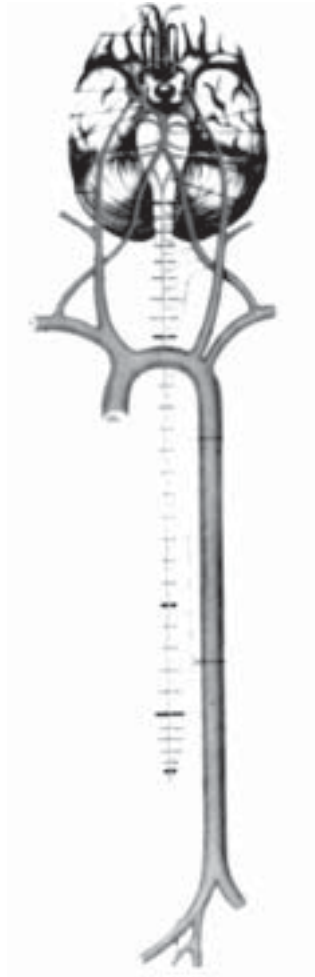


Fig. 53. Blood supplying of the brain and spinal cord (sheme).

Classification of cerebrovascular diseases

According to the World Health Organization **classification all cerebrovascular diseases** are divided into 3 groups:

A. **Premonitory and initial** symptoms of brain blood supply insufficiency.

B. **Acute** cerebral blood circulation disturbances:

1. Dynamic cerebral blood circulation disturbances;
2. Acute hypertonic encephalopathy.
3. Hemorrhage – subdural, epidural
4. Intracerebral hemorrhage
5. Brain infarction (nonembolic)
6. Brain infarction (embolic)

C. Dyscirculative encephalopathy or chronic cerebral blood circulation insufficiency or slowly progressive insufficiency of cerebral blood circulation.

Dynamic cerebral blood circulation disturbances (DCBCD)

DCBCD – are acute brain dyscirculation events which usually are developed against the main diseases. They are associated with temporary general and focal brain symptoms. They have tendency to involution during 24 hours.

To DCBCD belong:

1. Transient ischemic attacks (TIA) – they take about 1/3 of all DCBCD.
2. Hypertonic crisis – it takes about 2/3 of all DCBCD.

This disease is very common and takes about 1/3 of all cases of cerebrovascular diseases.

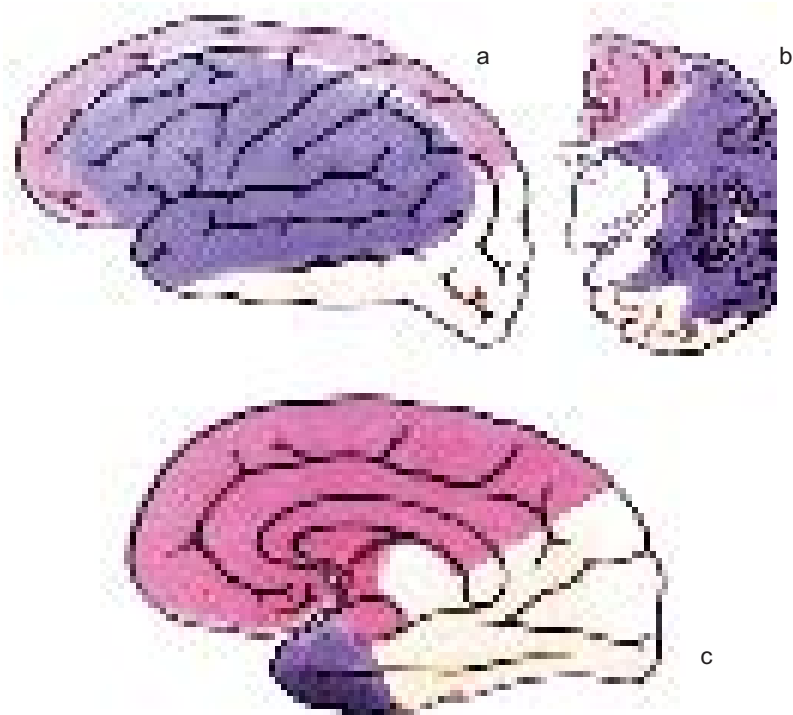


Fig. 54. Blood supplying of the hemispheres:
 Red – territory of the anterior cerebral artery;
 Blue – territory of the middle cerebral artery;
 Yellow – territory of the posterior cerebral artery

Etiology

TIA are usually associated with:

- Atherosclerosis.
- Stenosis of MAH.
- Heart diseases (abnormalities, myocardial infarction).
- Sometimes vasculitis, systemic vascular diseases.

In 30 – 40 % of patients with TIA stroke is developed in 5 years. In 20 % of them stroke is developed in 1 month, in 50 % – it is developed in a year. The possibility of stroke development is higher at repeated TIA and depends on age of the patient (it is getting higher in 1.5 times with every 10 years).

Pathogenesis

The main pathogenic mechanisms are:

A. Thromboembolic

B. Hemodynamic

They are very closely associated with each other.

A. The commonest of them are **thromboembolic**. That means microembolism of cerebral vessels.

Microembols are divided into **arteriogenic and cardiogenic**.

Arteriogenic microembols are small units that originate from clots and are the result of destroyed atherosclerotic plaques.

Thrombocytic embols are crumbling. That is the main reason of rapid involution of neurological deficit.

Cardiogenic microembols can cause TIA in patients with arrhythmias, heart abnormalities, after myocardial infarction, in patients with rheumatic endocarditis.

B. **Haemodynamic mechanisms**

1. Atherosclerotic stenosis of cerebral vessels or MAH, especially in association with hypotension, arrhythmias and myocardial infarction.
2. Thrombosis of neck magistral artery.
3. In the **subclavian steal syndrome** the patient has a narrowed subclavian artery and the arm "steals" blood from the basilar artery via the vertebral artery. There may be a cervical bruit and a difference in blood pressure between the arms. At times of arm exercise the patient may experience vertebrobasilar insufficiency.
4. Spasm of cerebral vessels and as a result perivascular edema and hypoxia of brain tissue.
5. Inborn stenosis, abnormalities of MAH.
6. Compression of vertebral artery by osteophytes at cervical osteochondrosis.
7. Vessels insufficiency (contradiction between real and demandable blood supply). This can occur at heart failure, hypotension, internal bleeding.

Clinical picture of TIA

TIA is usually characterized by focal neurological symptoms. The last usually dominate over general brain symptoms. Thus TIA is regional DCBCD. They are usually acute and develop suddenly.

There are two main groups of TIA's symptoms:

- a. focal;
- b. general brain symptoms.

General symptoms, if they present, usually manifest as headache, dizziness, short loss of consciousness.

Focal symptoms depend on the vessel territory.

TIA's in carotid distribution (They take 30 % of all TIAs)

Carotid distribution – is a territory of internal carotid arteries and their branches – ophthalmic artery, anterior cerebral artery, and middle cerebral artery. Via anterior cerebral artery carotid distribution supplies anterior part of frontal lobe, internal surface of hemisphere to sulcus parietooccipitalis, via middle cerebral artery – the cortex of frontal, parietal, temporal lobe, internal capsula and nucleus (fig 53).

1. The most common symptoms are subjective sensory disorders, such as numbness, tingling in face and extremities; and objective sensory disorders such as hyperesthesia of superficial sensation in face and extremities, sometimes deep sensation in fingers and toes.
2. Very often motor disorders together with sensory ones are observed. They are central paresis of extremity or fingers with hyperreflexia, pathologic signs of Babinski, Rossolimo. Hemiparesis or hemiplegia is observed only in severe cases.
3. Sometimes distorting language (transient aphasia) is observed.
4. When TIA occurs in the internal carotid artery territory ophthalmic – pyramidal syndrome is developed. It usually manifests as blindness or reduction of vision on the same side and hemiparesis on the opposite side. It is known as **Lasko – Radowich syndrome**.
5. Focal Jackson motor or sensory epileptic attacks are observed in patients with MAH pathology.

TIA's in vertebrobasilar distribution (They take about 70 % of all TIAs).

This territory provides blood supply of brainstem, cerebellum, occipital lobe cortex, mediobasal structures of temporal lobes (fig 37).

1. **Vestibular syndrome**. It manifests as systemic dizziness, occipital headache, nystagmus, equilibrium disorders.
2. **Brainstem – cerebellum syndrome**. It manifests as equilibrium, coordination and synergy of action disorders.

3. **Paresis of oculomotor muscles** with convergence disorders, diplopia, cross – eye.
4. **Bulbar syndrome** with swallowing, voice and speech disorders.
5. **Alternation syndromes** are quite rare.
6. **Vision disorders** of cortex character such as photopsias, hemianopsia, quadric hemianopsia and optic phenomena.
7. **Atonic – adynamic syndrome.** It is observed at acute ischemia of reticular formation and lower olives in medulla oblongata in case of “drop – attacks “ – the attacks of sudden loss of muscle tone that cause patient’s falling down without loss of consciousness. These attacks manifest at cervical spinal cord disorders in case of sudden turning out or head retroversion. Sometimes symptom of Sistine chapel can occur when loss of muscle tone is associated with loss of consciousness.
8. **At ischemia of mediobasal structures** of temporal lobe one can Korsakov syndrome observe – the attacks of temporary memory disorders on current events associated with confabulator component and disorientation.
9. **Paroxysmal hypersomnic and kataleptic syndromes** with autonomic – vascular crisis are observed at ischemia of hypothalamic structures.
10. **Syndrome of temporal epilepsy.**
11. The **subclavian steal syndrome.**

Except carotid and vertebrobasilar cerebral ischemic crisis there are associated dynamic cerebral blood circulation disturbances such as:

- Coronary – cerebral crisis;
- Aorto – cerebral crisis;
- Liver– cerebral crisis;
- Cholecysto – cerebral crisis.

TIA with neurological symptoms that clinically disappear within 24 hours but leave changes at CT such as low density zones are regarded as “ **minor ischemic strokes** “.

It is important to mention that the duration of TIAs is from several minutes up to 24 hours. But usually it is 10 – 15 minutes.

One of TIA’s characteristic features is **relapse**, when attacks are observed 3 – 5 times per year. There is one more peculiarity, which is necessary to remember – the attacks in vertebrobasilar territory are more common and are frequently repeated in spite of TIA in carotid territory.

But TIA in carotid territory prognosis is much more serious. Usually in one or two years after first attack stroke is developed.

Another peculiarity is that if TIA is observed several times per 24 hours it means there is pathology of MAH. Transient in word TIA concerns only neurological clinical picture, but has nothing to do with hemodynamic cerebral disturbances, as they are observed during the next 3 weeks.

Hypertensive cerebral crisis

It takes about 13 – 15 % of all acute disturbances of cerebral blood circulation.

Pathogenesis In course of a set of experiments it was found out that the spasm of veins during increasing of blood pressure displays a reaction of cerebral blood circulation regulation. The spasm of veins protects our brain from excessive perfusion. In case of rapid and severe increasing of blood pressure the system of self-regulation cannot compensate it. That is the main reason of **disturbance of self-regulation system of blood circulation and brain hyperemia.** It also promotes brain edema, blood circulation deficiency and ischemia of brain tissues.

Clinical features of hypertensive crisis:

- General.
- Regional (focal).
- Mixed.

Hypertensive crisis is a manifestation of hypertension or symptomatic arterial hypertension exacerbation. It manifests as general cerebral and focal symptoms.

Except high blood pressure severe headache, heavy head, psychomotor agitation, obscured consciousness, heart beating, shortness of breath, heart pain, nausea, vomiting, sometimes epileptic attacks are observed in case of hypertensive cerebral crisis. It is also usually accompanied by autonomic disturbances, such as sweating, feeling of cold in extremities, change of face colour, heart and breathe rate.

According to the system of hemodynamics there are three types of crises:

- Hyperkinetic.
- Hypokinetic.
- Eukinetic.

1. **Hyperkinetic type** usually is accompanied by increasing of heart outflow with increased heart index more than 4.5 l per min in m and normal general peripheral resistance.
2. **Hypokinetic type** is accompanied by decreasing of heart outflow with decreased heart index to 2.8 l per min in m and high general peripheral resistance.
3. **Eukinetic type** is accompanied by normal heart outflow and slightly increased general peripheral resistance.

The main clinical features of hyperkinetic crisis are:

1. There is a rapid development of crisis without portents.
2. Mainly systolic blood pressure is increased (More than 180-200 mm).
3. General cerebral symptoms are well – expressed. That means psychomotor agitation, severe headache with nausea and vomiting.
4. There are well – expressed autonomic disorders, such as shortness of breath, red or pale skin, polyuria.
5. The duration of crisis is up to several hours.

The main clinical features of hypokinetic crisis are:

1. There is gradual development against long lasting hypertension background.
2. Mainly diastolic blood pressure is increased.
3. There are well – expressed EKG – changes: slow intra – ventricular conductivity, decreasing of ST – segment.
In this case the risk of ischemic stroke is too high.

The main clinical features of eukinetic crisis are:

1. A rapid development.
2. Both systolic and diastolic blood pressures are increased.
3. The symptoms of acute left – ventricular insufficiency and lung edema can be observed.

According to the duration hypertensive crises are divided into:

1. Mild, which last up to 1 hour;
2. Mediate (they last several hours);
3. Severe (they last from 5 –6 hours up to 24 hours).

The frequency:

• **Mild hypertensive crisis** are:

Frequent – means they are observed more than 4 times per month;

Moderate frequent – means they are observed 3 – 4 times per month;

Rare – means they are observed 1 – 2 times per month.

• **Mediate and severe hypertensive crisis** are:

Frequent – means they are observed more than 5 times per year;

Moderate frequent – means they are observed 3 – 5 times per year;

Rare – means they are observed 1 – 2 times per year.

Diagnostics of dynamic blood circulation disturbances

When the symptoms of blood circulation disturbances last from several minutes up to hour there are no problems with diagnosis of dynamic blood circulation disturbances, such as transient ischemic attack or hypertensive crisis. In case of rapid development of general cerebral and focal symptomatic and duration up to several hours usually the diagnosis of dynamic blood circulation disturbances is put afterwards, as nobody knows the results (sometimes stroke is developed in such a way).

Very often transient ischemic attack or hypertensive crisis is the first sign of heart – vascular system disorders. That's why **it is very important to reveal the main disease**. That's why we need additional methods of patient's examination.

Additional methods of patients' with DCBCD medical examination:

1. Ultrasonic doplerography – finds out the absence or presence of stenosis and occlusions of magistral arteries of head and neck.
2. Rheoencephalography – finds out asymmetry of blood circulation, the state of vessel tonus and elasticity of vessels.
3. EEG – finds out diffuse and local changes of brain bioelectric potentials.
4. X –ray examination of cervical part of spinal cord – finds out osteochondrosis, spinal abnormalities.

5. EKG – finds out the state of coronal vessels, rhythm disorders, and coronary insufficiency.
6. Otoneurological examination is recommended in case of expressed vestibular syndrome in order to exclude labyrinth pathology.
7. Ophthalmic examination – finds out sclerotic or hypertensive changes on eye fundus.

Blood analysis:

8. Coagulation test – finds out increased aggregation of thrombocytes, erythrocytes, hematocrit.
9. Biochemical blood analysis – finds out proteinemia, increased cholesterolin, b– lipoprotein, pre-b– lipoprotein.

Differential diagnosis

Dynamic blood circulation disturbances are usually differentiated with:

1. Migraine crisis;
2. Partial epileptic attacks;
3. Vestibular crisis;
4. Multiple sclerosis;
5. Brain tumor;
6. Hypoglycemia;
7. Faint.

Treatment of dynamic blood circulation disturbances

Hospitalization is necessary for the patients with:

- a. Severe cerebral – vascular crisis that can result in stroke;
- b. Repeated attacks with well – expressed focal symptoms;
- c. Severe hypertensive crisis with high blood pressure;
- d. Coronary – cerebral attack, suspected myocardial infarction or angina pectoris.

The main principals of DBCD treatment:

1. To normalize blood pressure.
2. To improve brain hemodynamic.
3. To improve microcirculation and rheologic properties of blood, to prevent aggregation of blood cells.
4. To decrease vessels' penetrance.
5. To prevent brain edema, to decrease intracranial hypertension.
6. To improve heart activity.

7. To improve brain metabolism.
8. To overcome autonomic syndrome.

Treatment of hypertensive crisis

1. **At all kinds of crisis** hypotensive medications are used:
 - Clofelin 1ml 0.01 % i/v in 20 ml of physiologic solution;;
 - Dibazol 0.5% 2 – 4 ml i/m or i/v;
 - Euphyllini 2.4% 10.0 i/v in physiologic solution;
 - Antagonists of Calcium (finoptini 0.5% 2 ml i/v in 10 ml of physiologic solution);
 - Cardioselective b-adrenoblockers (athenolol 25 – 50 mg per day);
 - Nifedipine 10– 20 mg a day;
 - Inhibitors of ACE – Enalapril 5 – 10 mg a day;
 - In case of too high blood pressure with left ventricle insufficiency and lung edema ganglioblockers are used – benzohexonium 2.5 % 0.5 – 1 ml i/m or s/c in glucose solution by drops.
2. **In case of hypertensive hypokinetic crisis** we usually begin with:
 - Dibazol 0.5% 4 – 6 ml i/m and i/v;
 - b-adrenoblockers – Anaprilini 0.1% 5 ml i/v.

But the last are contraindicated at bradycardia, disorders of atrio-ventricular conduction.
3. **In case of hypokinetic crisis** we start with peripheral vasodilators:
 - Apresin – 2% 1 ml i/v by drops;
 - Diazoxid 1.5% 20 ml 3 – 4 times per day.
4. **At eukinetic crisis** we usually use clofelini, antagonists of calcium, spasmolytics.
5. **At all kinds of hypertensive crisis with psychoemotional and autonomic reaction** we prescribe i/v by drops:
 - Aminazini 2.5% 1 ml in 200 ml of physiologic solution;
 - Sibazoni 0.5% 2 ml i/m;
 - Pipolfeni 2.5% 2 ml i/m.
 1. In order to prevent brain edema we use euphyllini 2.4 % solution 10 ml with 2 ml lazix i/v;
 2. To decrease brain hypoxia we use Na oxybutiras 20% 10 ml i/v in glucose solution.

Treatment of TIA:

1. Blood pressure normalization

- a. At heart failure and systolic blood pressure (BP) less than 120 mm glycosides are used:
 - Corglyconi 0.06 % 1.0 + strophantini 0.05 % 1.0 in 20 ml of 40 % glucose i/v;
 - Cordiamini 1 ml s/c;
 - Sulfocamfocaini 10 % 2ml i/m;
 - Cofeini 10 % 1ml s/c;
 - Mesatoni 1– 0.05 % 1 ml s/c or i/m at low BP;
 - Prednisoloni 60 – 120 mg i/v by drops with 200 ml 5 % glucose.
- b. At hypertension hypotensive therapy and spasmolytics are used in usual doses.

4. **To improve brain hemodynamic** vasoactive medications are used:

- Euphilini 2.4 % 10 ml i/v with 10 ml 0,9 % NaCl;
- Cavintoni 10 – 20 mg i/v with 20 ml 0.9 % NaCl;
- Sermioni 4 –8 mg i/v by drops in 150 ml 0,9 % NaCl;
- Ksantinoli nicotinas 15 % 2 ml i/m.

3. **To improve microcirculation and rheologic properties of blood, to prevent aggregation of blood cells.** (It is especially important in case of well expressed focal neurological symptoms in order to prevent stroke).

• Direct anticoagulants:

– Heparini 5 –10 000 U. s/c 3 –4 times per day during 3 –5 days, than 2 500 U. 4 times per day during 3 – 4 days;

– Fraxiparini 0.3 x 2 s/c;

• Indirect anticoagulants:

– Pelentani 0.1 – 0.3;

– Fenilini 0.015 – 0.03;

– Syncumar 0.004 2-3 times per day during 2-3 weeks.

4. Antiagregants:

• Aspirini 70 –80 mg once a day;

• Ticlidi 0.25 2 times per day during 2– 3 months;

• Trentali 2% 5 ml i/v by drops (it is contraindicated at myocardial infarction, do not use it with heparin or on an empty stomach);

• Agapurini 1 pill 3 times per day;

• Reopoliglucini 200 –400 ml i/v by drops;

• Solcoserili 10 –20 ml i/v by drops during 5 –7 days;

- Ksantinoli nicotinas 15 % 1 –2 ml i/m;
- Kurantili 0.025 3 times per day (it is contraindicated at low BP, heart failure);
- Plavix 75 mg per day;
- Parmidini 0.5 3 times per day.

5. To prevent brain edema, to decrease intracranial hypertension:

- Furasemidi 40 –80 mg per day;
- Lasix 1% 2ml i/v or i/m;
- Manitoli 10 –20 % 200 ml i/v;
- Dexoni 4-6 mg i/v or i/m;
- Albumini 5 % 100 ml i/v;
- Vit E 5% 2 ml i/m, Aevit 1 ml i/m, Unitioli 5 ml i/m.

6. To improve brain metabolism:

- Nootropil 20 % 5– 10 mli/v;
- Instenoni 2 ml i/m or i/v by drops;
- Solcoserili 10 –20 ml i/v by drops;
- Actovegini 2 ml i/m or i/v (it is contraindicated at cardiovascular failure, oliguria, lung edema);
- Cerebrolisini 15 % 1.0 ml i/m or i/v.

7. Antioxydants:

- Vit E 1 ml i/m;
- Tiatrisolini 2 ml i/m;
- Emoxipini 1.0 i/m.

8. Symptomatic treatment:

- At vomiting and hiccup – cerukal, aminasini, galoperidol, validol;
- At headache – tramadol, analgini;
- At agitation – sedatives and anxyolytics.

Surgical treatment – is used at stenosis of general or internal carotid artery (when stenosis is more than 70 % according to ultrasonographic data). Thrombinectomy is used.

Prevention means in term diagnosis and treatment of main disease (first of all atherosclerosis, hypertension, and heart diseases).

Working efficiency The persons with DCBCD are regarded temporary disabled. In case of frequent and repeated DCBCD it is necessary to employ patient in order to exclude mental and physical strain or direct him to MSEC to fix up the group of disability.

Strokes

Definition

In broadest sense, the World Health Organization has defined stroke as “rapidly developing clinical signs of focal (at times global) disturbance of cerebral function, lasting more than 24 hours or leading to death with no apparent cause other than that of vascular origin”.

Epidemiology

According to World Health Organization data about 150 to 740 people per 100 000 population suffer from stroke all over the world. In the United States of America there are about 400 000 patients with stroke every year. In Europe there are about 1 mln people with stroke every year. In Ukraine according to 2001 data 130 890 patients had stroke and 54 000 of them died. During the last 30 years the frequency of stroke has increased significantly all over the world.

Classification of stroke Stroke is classified by the pathology of the underlying focal brain injury into either infarction or hemorrhage. Infarction is much more common than hemorrhage. The proportion between them is 4:1 (80 % to 20%).

Hemorrhagic stroke means **hemorrhage**. **According to the localization of hemorrhage it is divided into such groups:**

1. Intracerebral (when the hemorrhage is into the substance or parenchyma of the brain).
2. Membrane
 - a) subarachnoid (when the bleeding originates in the subarachnoid spaces surrounding the brain);
 - b) epidural and subdural (traumatic).
3. Combined
 - a) subarachnoid – parenchymatous;
 - b) parenchymatous– subarachnoid;
 - c) parenchymatous–ventricular;
 - d) ventricular.

Etiology

The most common causes of hemorrhage are:

1. Hypertension.

2. Symptomatic arterial hypertension (at kidney diseases, systemic vessel processes).
3. Inborn arterial and arterial – venous malformations.
4. Blood diseases (leucosis, polycythemia).
5. Cerebral atherosclerosis.
6. Intoxications, such as uremia, sepsis.

Pathogenesis

Hemorrhage results from rupture of the vessel anywhere within the cranial cavity or diapedesis in case of increased penetrance of vessel's wall. Rupture of the vessel is much more common and takes about 80 % of all stroke cases while diapedesis occurs only in 20 % of cases.

Hypertension has been implicated as the main cause of hemorrhage. Hypertension is the cause of a weakening in the walls of arterioles and the formation of micro aneurysms. Among elderly nonhypertensive patients with recurrent hemorrhages, amyloid angiopathy has been implicated as an important cause. Others causes include arterial-venous malformations, aneurysms, bleeding disorders or anticoagulation, trauma, tumors, cavernous angiomas and drug abuse.

Pathomorphology

In case of hemorrhagic stroke we distinguish hematoma – like hemorrhage and transudation – like hemorrhage.

According to the localization there are:

1. Lateral hemorrhage (they are located laterally compared with the internal capsule. They take about 40 % of all hemorrhages).
2. Medial hemorrhage (they are located medially compared with the internal scapule and take about 10 % of all hemorrhages).
3. Combined hemorrhages (they take the whole region of basal nuclei: subcortical nuclei, thalamus, and internal capsule. They take about 16 % of all hemorrhages).
4. Cerebellar hemorrhages (6 – 10 %).
5. Brain stem hemorrhages (5 %).

Primary ventricular hemorrhages are very rare. In case of large hemorrhage brain edema is developed. The last is associated with dislocation of brain stem which is the main cause of patients' death.

The main cause of death is rupture into the ventricular system and brain stem hemorrhage with lesion of main vital centers.

Clinical picture There are three main periods of stroke:

1. Acute (up to 3 – 4 weeks);
2. Renewal (up to 1 year);
3. Residual.

Acute period is divided into such stages:

1. Precursors;
2. Apoplectic stroke;
3. Focal signs.

Usually hemorrhage has rapid development in day time during physical or emotional stress. The patients are usually young. And they have risk – factors in anamnesis. Precursors are very rare.

Two main groups of symptoms are typical for hemorrhage – general cerebral and focal. Typically general cerebral symptoms prevail over focal ones in case of hemorrhage.

General cerebral symptoms mean severe headache, vomiting, consciousness disorders.

While examining patients with consciousness disorders we should pay attention to the possibility of contact with patient, his ability to follow commands, to say what has happened with him, to orient in the space and time and so on.

Sometimes **sopor** occurs at the beginning of hemorrhage, which can develop in coma in a few hours.

Coma is characterized by deep consciousness disorder, disturbance of breathing and heart activity. The patient doesn't respond to stimuli.

At **atonic coma** all reflexes are lost, blood pressure is decreased and there is breathing disturbance.

In case of coma development response to stimuli is absent, eyes are closed, mouth is opened, face is red, lips are cyanotic, neck vessels are pulsing, there is breathing disturbance, skin is cold, pulse is strained and slow, blood pressure is increased, temperature increases in 24 hours. Patient is lying on his back. All muscles are relaxed. Pupils are changed (there can be anisocoria, cross – eyes, sometimes gaze paresis can be observed). Mouth angle is a little bit lower. On the opposite side hemiplegia is often observed: the arm is

falling down like bane, there is hypotonia of muscles, reflexes are low, and Babinski sign is often observed too.

Sometimes meningeal signs, vomiting and dysphagia are observed too. Retention of urine or involuntary urination can also occur. In case of cortex irritation epileptic attacks can be developed. Large hemisphere hemorrhage is often complicated by **secondary brain stem syndrome**. It manifests as:

1. progressive breathing disorders;
2. disturbance of heart activity;
3. disturbance of consciousness;
4. eye movements disorders;
5. changes of muscle tonus (hormetonia);
6. autonomic disorders (sweating, tachycardia, hyperthermia).

Brain stem hemorrhage is associated with tetraparesis, alternating syndromes, eye movement disorders, nystagmus, goggle disorders, cerebellar syndromes.

Pons hemorrhage manifests as ptosis, gaze paresis, increased muscular tone (hormetonia).

Cerebellar hemorrhage usually starts with dizziness, severe headache in occipital lobe, vomiting. Eye movement disorders, ptosis, Gertvig – Mazhandi syndrome, Parino syndrome are observed. It is also associated with cerebellar symptoms, such as nystagmus, dysarthria, hypotonia, and ataxia. Paresis of extremities is not common.

The most common complication of intracerebral hemorrhage is **rupture into the ventricle system**. This is usually associated with worsening of patient's state, hyperthermia, breathing disorders, and hormetonia. Hormetonia manifests as changes of muscle tone in extremities, when hypotonia is changed into hypertonia in a few seconds or minutes.

At right hemisphere hemorrhage involuntary movements in nonparalysed extremities are observed and they are called parakinesis.

On the **side of lesion** there is anisokoria, Bechterev phenomena, painful trigeminal and occipital points, automatic movements, gaze paralysis.

On the **opposite side** – positive Babinski's sign, hypotonia, hyporeflexia, pathological signs, foot is turned outside, mouth angle is located lower than normally.

The course of the disease

The patients with brain hemorrhage are in very severe state. 70% to 95 % of them died. About 40 % – 45 % of them die during the first day, the rest of them – live 3 or 5 days.

The main **cause of death** is compression of brain stem as a result of brain edema and rupture into ventricular system associated with disorders of vital centers.

In case of good prognosis the patients usually recover from coma, then reflexes appear, general cerebral symptoms regress, the patients start to gorge, then they start to move, sensation and speech renew also.

Diagnostics

In case of rapid development of all symptoms with loss of consciousness, high blood pressure and presence of focal symptoms there is no problem with putting diagnosis. But when the hemorrhage starts gradually without loss of consciousness, then it is much more difficult to put a diagnosis. In this case instrumental and laboratory examination has a great meaning.

In **blood** usually leucocytosis, related lymphopenia, hyperglycemia (up to 8 – 10 mmole/l) is observed.

In **liquor (CSF)** which is flowing out under high pressure during lumbar puncture a great number of erythrocytes are found.

On **eye fundus** – retinal hemorrhages, hypertonic angioretinopathy and Salus symptoms are observed.

At **echoencephalography** there is dislocation of middle structures on 6 –7 sm to the healthy side.

By means of **angiography** we can find out aneurysm, dislocation of blood vessels, to find out zone “without vessels”.

CT and **MRI** find out hyperdense focuses (fig. 55-58).

Differential diagnosis

1. Infarction of brain (thrombembolic).
2. Epistatus.
3. Uremic coma.
4. Diabetic coma.
5. Traumatic hemorrhage.
6. Brain tumor with inside hemorrhage.

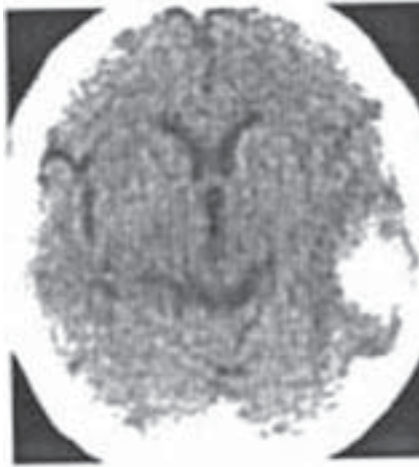


Fig. 55. Lateral hemorrhage (they are located laterally compared with the internal capsule)

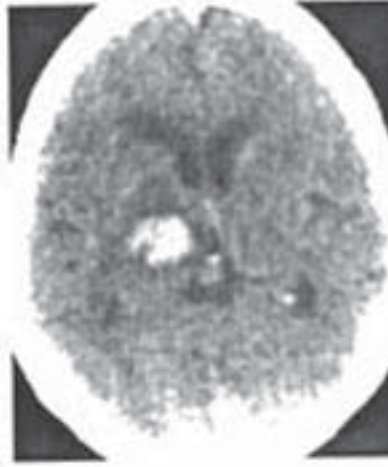


Fig. 56. Medial hemorrhage (they are located medially compared with the internal capsule)



Fig. 57. Ventricular hemorrhage.



Fig. 58. Brain stem hemorrhage

Subarachnoid hemorrhage (SH)

Etiology The most common cause of SH is aneurysm rupture. The other causes include hypertension, atherosclerosis, blood diseases, rheumatism, brain tumors, and uremia and so on. **The rupture risk – factors** are physical or emotional stress, changes of blood pressure, alcohol usage.

According to **Samoiylov (1990)** there are 8 kind of **etiologic factors of subarachnoid hemorrhage**:

1. Aneurysmatic (50 – 62 %) – aneurysm rupture (fig 59).
2. Hypertensive (at hypertension).
3. Atherosclerotic (15 %).
4. Traumatic (5 – 6 %).
5. Infectious – toxic (8.5 %).
6. Blastomatose (at tumors).
7. Pathohemic (at blood diseases).
8. Cryptogenic (4 – 4.8 %).

Clinical features

The **development of the disease is rapid**, without precursors. Although some of patients have symptoms of aneurysm: headache in frontal lobe, paresis of CN (Oculomotor N is often damaged).

The **first signs of SH** are severe headache or feeling of hot liquid flowing in the brain. At the beginning this pain is local (in the region of occipital lobe) than it becomes more diffuse. Later pains in neck, back appear, sometimes they irradiate in legs. Simultaneously with headache vomiting and nausea occur. Besides these symptoms there are often **general cerebral symptoms**, such as short loss of consciousness, psychomotor excitement, seizures.

In a few hours or the next day **meningeal syndrome** occur. It manifests as rigidity of occipital muscles, symptoms of Kernig, Brudzinski, and general hyperesthesia. Significant focal neurologic symptoms are not common. Only in case of basal hemorrhage CNs suffers (that is the reason of ptosis, cross – eye, diplopia, paresis of mimic muscles).

That's why lesion of CNs is typical for basal aneurysm rupture.

In acute stage of hemorrhage a lot of patients have **focal symptoms**: paresis of extremities, sensory and speech disorders. Usually it can be explained by associated brain hemorrhage, arterial spasm and as a result local ischemia. Spasm is usually developed on the 3rd – 4 th day and lasts till 3rd – 4 th week.

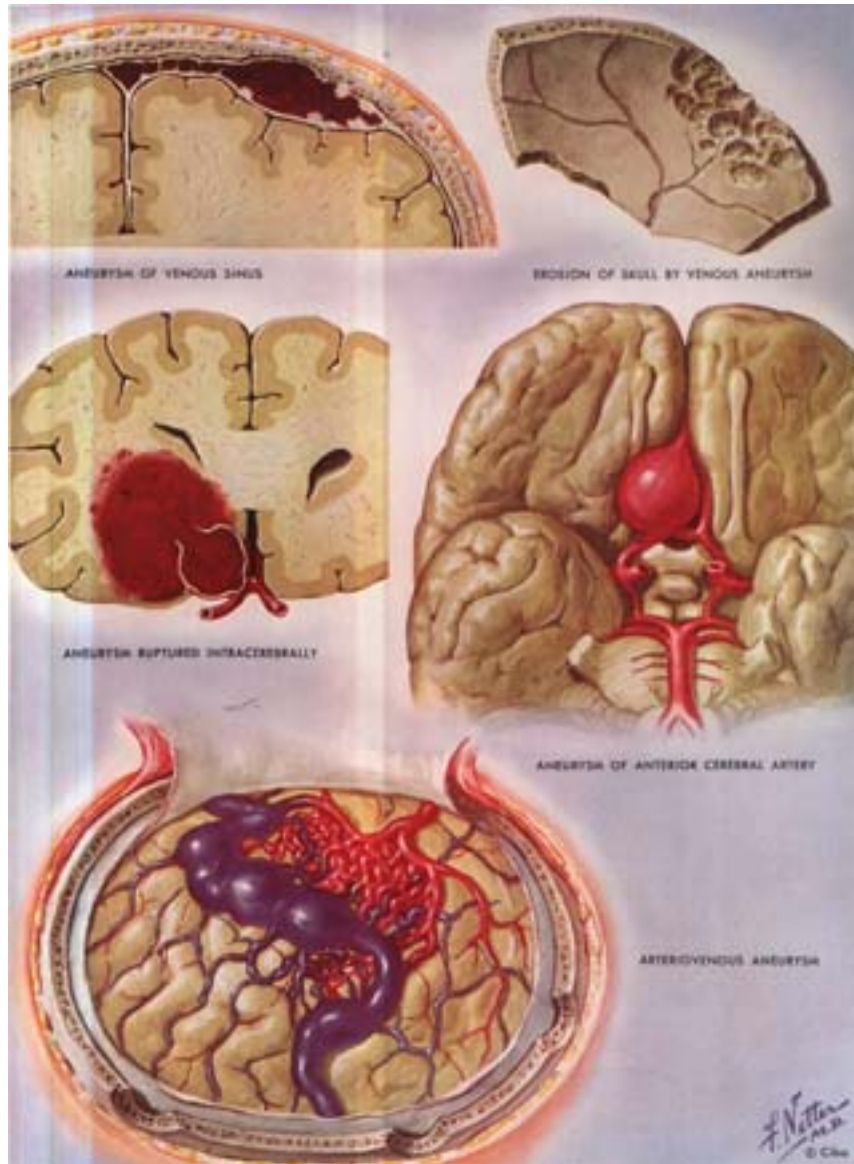


Fig. 59. Intracranial aneurysms (Frank H. Netter, M.D.)

On the second or third day almost all patients with SH have increased temperature to 37.5 – 38°. Leucocytosis is also observed. In liquor fresh blood is found out.

In severe cases significant disorders of heart – vascular system and breathing are observed.

The main **complications of SH** are:

1. Brain edema;
2. Recurrent SH;
3. Occlusive hydrocephalus;
4. Brain infarction.

Prognosis usually is not good. As during the 2nd – 4th weeks recurrent hemorrhage can occur which can cause patient's death.

Thus we usually put the **diagnosis** of SH in case of:

1. Stroke – like development with general cerebral and meningeal symptoms and absence of significant focal neurologic deficit.
2. The presence of blood in liquor (bleeding liquor during first day and yellow liquor on 3rd – 5th day).
3. Retinal hemorrhages are on eye fundus.

Differential diagnosis:

1. Meningitis.
2. Acute food toxic infection.
3. Infectious diseases.

Lumbar puncture, CT (fig. 60), MRI, US helps to put diagnosis; angio-graphy helps to find localization and sizes of aneurysm.

Brain infarction

Classification

Brain infarction is divided into thrombotic, nonthrombotic and embolic. Thrombotic and embolic occurs in case of plugging of extra cranial or intracranial vessel by thrombus, embolus or atherosclerotic plaque.

Nonthrombotic one occurs at angiospasm in case of atherosclerosis, vascular insufficiency.

Etiology

The most common causes of brain infarction are:

- Atherosclerotic lesion of MAH;
- Combination of atherosclerosis with hypertension;

- Chronic ischemic heart disease with rhythm disorders;
- Combination of atherosclerosis with diabetes;
- Rheumatism, heart abnormalities (inborn and acquired);
- Vasculitis.

Pathogenesis



Fig. 60. Subarachnoid hemorrhage

1. Thrombotic brain infarction occurs at thrombosis in case of atherosclerotic lesions, disturbance of rheologic blood properties, central hemodynamic disorders.

2. Embolic brain infarction occurs at embolus plugged cerebral vessel.

The sources of embolus are:

- a) parts of aortal or arterial thrombus;
- b) parts of thrombus in case of heart abnormalities, such as mitral stenosis, aortal abnormality, rheumatic or bacterial endocarditis, myocardial infarctions, cardiosclerosis, cardiomyopathy;
- c) embolism can occur at thrombophlebitis, lung abscess, malignant tumors, and sepsis;
- d) fat embolism occurs at bone fractures, after surgery associated with trauma of subcutaneous tissue;
- e) gas embolism occurs at surgery on lungs, at pneumothorax.

Usually it is difficult to distinguish thrombosis and embolism. That's why we use the term "thromboembolism".

Pathophysiology

As a result of brain infarction in case of blood circulation less than 10 ml per 100 g of brain tissue in 1 min **zone of focal necrosis** is formed (fig. 61).



Fig. 61. Strokes: 1. Ischemic; 2. Hemorrhage.

During the first 6 hours this zone is surrounded by region with borderline blood circulation (18 – 20 ml per 100 g in 1 min). This region is called **penumbra** (fig. 62). The neurons within this region are preserved structurally but their function suffers. During first 3 – 6 hours we can renew their function, that's why this period is known as **“therapeutic window”**.

After 6 hours zone of brain infarction is formed completely. Pathobiochemical and pathophysiological changes are observed in penumbra region. One of the main reasons of neurons death is accumulation of glutamat, which causes brain edema and disturbance of synaptic transference. The number of intracellular enzymes is increased. That causes activation of thrombocytes, microcirculation disorders, and ischemia. As a result ruination of neurons occurs.

Nonthrombotic brain infarction

Usually occurs at disturbance of self – regulation of brain blood circulation, in case of MAH abnormalities and long lasting angiospasm.

Clinical features:

Acute period of brain infarction is divided into three stages:

1. Precursors
2. Apoplectic stroke
3. Focal signs

To **precursors** belong transient ischemic attacks (TIA) in the same region where brain infarction is developed. Brain infarction usually is developed in elderly people at any time. **Gradually** during se-

veral hours focal neurologic symptoms are developed. Sometimes the beginning of brain infarction (BI) is rapid, especially in case of embolic stroke. But one of the most important differential features of BI is **the prevalence of focal symptoms over the general cerebral ones.**

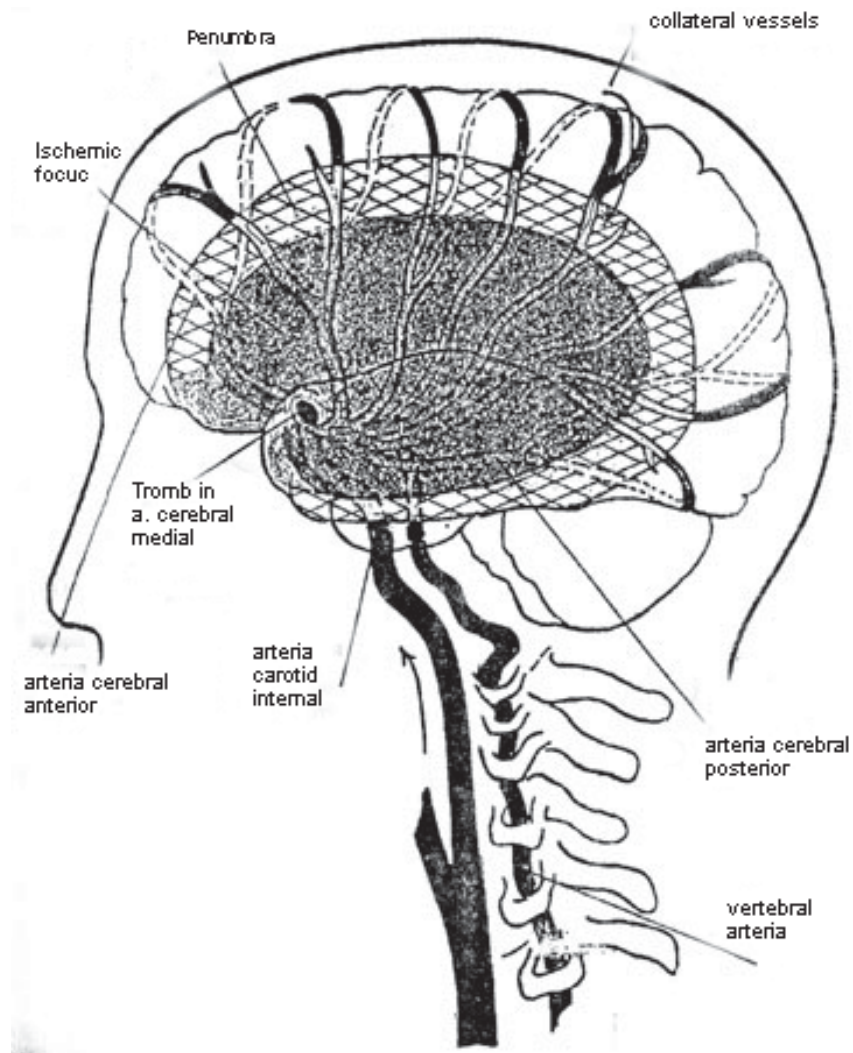


Fig. 62. Penumbra

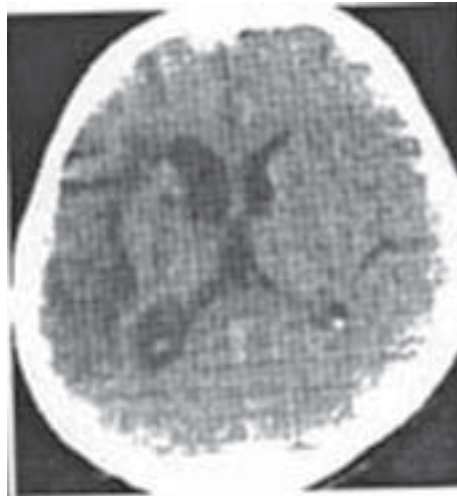


Fig. 63. Ischemic stroke in distribution of medial cerebral artery



Fig. 64. Distribution of medial cerebral artery

General cerebral symptoms manifest as headache, vomiting, consciousness disorders. These symptoms are usually observed at thrombembolic stroke and are increased according to the brain edema.

Focal symptoms depend on localization of the infarction, damaged vessel and state of collateral blood circulation. The most common brain infarctions are in the region of middle cerebral artery.

Middle cerebral artery blood – supplies basal nuclei, internal capsule, a part of temporal lobe, middle and lower parts or pre – and post central gyri, opercular region, a part of parietal lobe, gyrus angularis, posterior parts of upper and middle frontal gyri (fig. 63, 64).

If the artery's main trunk is occluded hemiplegia, hemianesthesia, gaze paresis, speech disorders (such as motor, sensory, total aphasia) occur when the lesion is in left hemisphere and apraxia –

agnostic syndrome occur (apraxia, astereognosia, autotopagnosia, pseudomelia) when the lesion is right hemisphere.

If the artery's cortical branches are occluded motor and sensory disorders in upper extremity, hemianopsia, sensorimotor apha-

sia, apraxia, alexia, acalculia occur when the lesion is in left hemisphere and anosognosia, and autotopognosia occur when the lesion is right hemisphere.

If the artery's posterior branches are occluded hemianesthesia, bathianesthesia, astereognosis, afferent paresis of extremities, hemianopsia, sensory aphasia, agraphia, alexia, acalculia and apraxia occur.

Anterior cerebral artery blood – supplies the cortex of frontal lobe (superior frontal gyrus), superior part of anterior central gyrus, superior part of posterior central gyrus, corpus callosum, a part of superior parietal lobulus, orbital part of frontal lobe, lobulus paracentralis.

Infarction in the area of the anterior cerebral artery causes spastic hemiparesis with the prevalence in proximal part of upper extremity and distal part of lower extremity. This kind of BI is associated with the symptoms of oral automatism, psychiatric disorders, dysphagia, dysphonia, atasia, abasia, motor aphasia, retention of urine.

If the internal carotid artery is occluded before ophthalmic artery alternating optic – hemiplegic syndrome takes place. That means blindness or visual disorders on the side of lesion and hemiparesis on the opposite side. There is absence of internal carotid artery pulsation on neck.

If the internal carotid artery is occluded intracranial hemiplegia, hemianesthesia, well expressed general cerebral symptoms as a result of brain edema, compression or dislocation of brain stem are observed.

When the infarction is in the region of **vertebral-basilar vessels** occipital lobes and brain stem suffers.

Infarction in **cortical branches** region manifest as visual disturbances: homonymic hemianopsia with preserved macular vision or upper quadrantic hemianopsia, sometimes visual agnosia and metamorphopsia can occur. If the lesion is in the left hemisphere alexia, sensory and semantic aphasia can be observed.

If the process is distributed on mediobasal part of temporal lobe, memory disorders, such as Korsak syndrome usually take place.

Posterior cerebral artery blood supplies occipital lobe, posterior part of lower and middle temporal gyri, basal and mediobasal part of temporal lobe, deep thalamocollicular branches blood supply thalamus, hypothalamus, posterior – lower parts of cortex of parietal lobe (fig. 65).

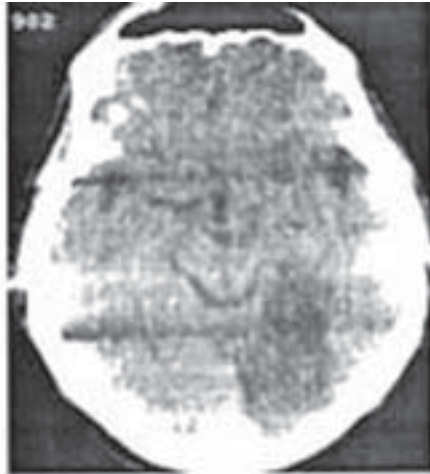


Fig. 65. Ischemic stroke in distribution of posterior cerebral artery

Infarction in the region of the posterior cerebral artery causes hemianopsia, visual agnosia, hemianesthesia, hyperpathia, desorientation in space and time, disorders of space – optical gnosis.

Infarction in the region of deep branches of posterior cerebral artery that blood supplies thalamus, posterior part of occipital lobe, corpus callosum, radiate crown, causes thalamic syndrome of Dejerin – Russi. The last manifests as hemianesthesia, hyperpathia,

dysesthesia, thalamic pains, hemianopsia, pseudoathetosis, hemiataxia, and dynamic hemiparesis on the opposite side.

Infarctions in the region of vertebral artery Vertebral artery blood supplies brain stem; oblongate brain, cerebellum, cortex of occipital lobe, part of cervical part of the spinal cord. It can be damaged extra – or intra cranially.

In case of extracranial lesion systemic dizziness, hearing, visual, eye movements, vestibular, equilibrium disorders and paresis with sensory disturbances in extremities are observed. Some patients have “drop– attacks“.

In case of intracranial lesion alternating syndromes of oblongate brain occur.

Infarctions in the region of **basilar artery** cause the lesion of pons, cerebellum, hypothalamus, cortex of occipital lobe. In case of acute occlusion loss of consciousness, eye movements disorders, pseudobulbar syndrome, tetraplegia, muscle tone disturbance, sometimes even cerebellar symptoms and cortical blindness occur. Most of these patients die because of the vital functions disorders.

When the lesion is in **pons** syndromes of Miyar – Hyubler, Fowill, Brisso – Siquar and alternating hemianesthesia occur.

Infarction in the region of **mesencephalon** causes Parino syndrome, which manifests as gaze paresis upwards.

Infarction in the region of **brain peduncles** causes Weber or Benedict alternating syndrome. Sometimes Claude and Foa – Nikolesku syndrome can be observed.

Diagnostics of Brain Infarction The main peculiarities are:

- Before stroke period in the previous history (TIA in anamnesis);
- The beginning of the stroke is gradual;
- Data of somatic and neurological status;
- Additional methods of diagnostics.

Usually the patients with BI have rheologic disturbances. **Liquor** is pellucid, without significant changes. There is focus of pathologic activity on **EEG**. **USG** finds out occlusion, stenosis of carotic and vertebral arteries. **CT** reveals hypodensive focus on the second day. **MRI** helps to find out small focuses and those, located in the brain stem.

Differential diagnosis:

1. Traumatic hemorrhage, trauma of brain.
2. Myocardial infarction.
3. Epilepsy.
4. Uremic coma.
5. Hyperglycemic coma.
6. Hypoglycemic coma.
7. Brain tumor with inside hemorrhage.

While differentiation we should pay attention on anamnesis, run of the disease, symptoms, blood pressure, MAH state, eye fundus state, EKG, laboratory data, LP data.

Alcoholic coma is characterized by alcohol odour, psychomotor agitation, red eyes and face, seizures, normal blood pressure, low reflexes, enlargement of liver.

Uremic coma is characterized by ammonium odour, oliguria, edema, ascites.

Diabetic coma is characterized by acetone odour, Kussmaul breathing, dry skin, hypotonia, hyporeflexia, hyperglycemia, glucosuria, acetone and sugar in urine.

Hypoglycemic coma is characterized by wet skin, normal breathing, seizures, and low sugar level in blood.

Epileptic coma is characterized by seizures, there is episyndrom in anamnesis. EEG helps to put a diagnosis.

Course of the disease There are three main kinds of stroke course:

1. **Favorable**, when all damaged functions are renewed.
2. **Remittent** when the patient state is worsening because of associated pneumonia or other complications.
3. **Progressive**, which usually causes death of the patient.

The most severe are hemorrhages. Mortality in this case is from 60 to 90 %. Especially increases mortality in case of hemorrhage in ventricles, brain stem and cerebellum.

Favorable prognosis is in case of limited hemorrhages, small in size, without brain edema and secondary brain stem syndrome development.

Mortality at brain infarction is in 20 – 27 % of all cases.

Strokes treatment

There are nondifferential and differential kinds of stroke treatment.

Nondifferential treatment includes:

1. Prevention and treatment of pulmonary insufficiency;
2. Liquidation of heart – vascular disorders;
3. Brain edema treatment;
4. Normalization of water – electrolytes balance, acid – alkali balance;
5. Osmose correction;
6. Improving of brain metabolism;
7. Liquidation of hyperthermia and other autonomic disorders.

1. Prevention and treatment of pulmonary insufficiency:

- a) the patient is lying on the bed with his head elevated;
- b) cleaning of patient's oral cavity;
- c) tracheostomia (at inspiratory muscles paralysis);
- d) at lung edema patient is given oxygen; narcosis, Bobrov's apparatus, 2 ml 1 % lazix, 2 ml 1% dimedroli, 2 ml 0.1 % atropini i/m are used;
- e) antibiotics are used in order to prevent pneumonia.

2. Liquidation of heart – vascular disorders:

- a) At increased blood pressure we use:
 - Clofelini 1 – 3 ml 0.01 % solution i/m, i/v;
 - Dibasoli 3 – 4 ml 1 % solution i/v;
 - Droperidoli 1 ml 0.25 % solution i/v;
 - Rausedili 1 – 2 ml 0.1 % i/v, i/m;

- In case of hyperkynetic type of blood circulation we use b– adrenoblockers (anaprilini, obzidani, inderal);

- In case of hypokynetic type of blood circulation we use peripheral vasodilators (Natrii nitroprussidi, appresini) in combination with euphyllini;

b) At low blood pressure we prescribe:

- Dexamethazoni 4 – 8 mg i/v by drops in physiological solution;

- Prednizoloni 60 – 120 mg i/v by drops in physiological solution

- In order to improve heart activity we use strofantini, corgliconi, cordiamini.

3. Brain edema treatment:

Diuretics, corticosteroids, albumini, ganglioblockers, 20 % mannit, manitoli, glycerini, lazix, diakarbi are used.

4. Normalization of water – electrolytes balance and acid – alkali balance:

We should estimate patient's necessity in water according to his secretion, the level of Na in blood, hematocritis. An average water necessity is 35 ml per kg, in patients with loss of consciousness it is 50 ml per kg. We should correct patient's hyper– or hyponatriemia, hyper– or hypokaliemia. 4 % solution of natrium bicarbonates i/v, trisaminum is used at metabolic acidosis. KCl i/v is used at metabolic alkalosis.

5. Osmose correction:

Normally blood osmose is within 280 – 295, urine osmose is 600 – 900 mosm per liter.

At stroke usually we have hyperosmose, which manifests as increased hematocritis, hyperagrigation.

6. Improving of brain metabolism:

Vit E, piracetami, aminaloni, cerebrolysin natrii oxybutiras are used.

7. Liquidation of hyperthermia and autonomic disorders:

At hyperthermia we use:

- Reopirini 5 ml i/m

- Analgini 50 % 2 ml i/m, i/v;

- Aspizoli 0.5 g i/v.

At autonomic disorders we introduce:

- Sibazoni 0.5 % 2 ml i/m, i/v;

- Haloperidoli 0.5 % 1 ml i/m;

- Dimedroli 1 % 2 ml i/m;
- Natrii oxybutiras 20 % 10 ml i/v.

Differential treatment of haemorrhage:

The main directions of treatment are:

1. To lower increased blood pressure.
2. To liquidate brain edema and lower intracranial pressure.
3. To increase coagulative properties of blood and decrease penetrance of vessels' wall.
4. To prevent and treat cerebral vessels spasm.
5. To normalize vital and autonomic functions and prevent complications.
6. To treat hypoxia and brain metabolism disorders.

1. To lower increased blood pressure:

Clofelini, b-adrenoblockers (anaprilini, obzidani, inderali), Calcium antagonists (nifidipini, adalat) IACE (capoten, enalapril, renarapril) are used.

At **too high blood pressure** ganglioblockers are used:

- Pentamini 5% 1 ml i/v by drops;
- Benzohexonium 2.5 % 1 ml i/v by drops;
- Arfonad 5 ml 5 % i/v in physiologic solution by drops.

2. To liquidate brain edema and lower intracranial pressure:

The same measures as in nondifferential treatment are used.

3. To increase coagulative properties of blood and decrease penetrance of vessels' wall:

a) CaCl₂ 10 – 20 ml 10 % i/v;

Vicasoli 1 – 2 ml 1 % i/v;

Ascorbinic acid 2 – 5 ml 5 – 10 % solution I/m.

b) **Antifibrinolytics:**

EAKA 100 ml 5 % solution 1–2 times per day I/v by drops during 5 – 7 days. Then we use it orally – 3 g every 3–4 hours up to 3 weeks.

c) **At decompensated fibrinolysis** we use:

Trasiloli 20 – 30 000 U i/v by drops in 250 ml of physiologic solution;
Hordox 5 000 U i/v by drops every other day.

d) **to normalize microcirculation** we use:

Dicinoni 2 ml 12.5 % solution 2 – 3 times per day during 10 days, then 2 tablets (0.5 g) every day;

Ascorutini, Rutamini (1 ml i/m 1 – 2 times per day).

4. To prevent and treat cerebral vessels spasm:

Antagonists of Calcium are used:

Nimotop is introduced i/v 15 mg per kg per day during 5 – 6 hours. On the 5th – 7th day it is used orally 60 mg every 4 hours during 7 – 10 days.

5. To normalize vital and autonomic functions and prevent complications.

6. To treat hypoxia and brain metabolism disorders:

The same measures as in nondifferential treatment are used.

7. Symptomatic treatment:

At **severe headache**:

- Baralhini 5 ml i/v, i/m;
- Combination of analgini (4 ml 50 % solution) with 1 ml 1 % dimedroli and novocaini 5 ml 0.5 % solution i/m;
- Promedoli are used.

At **vomiting**:

- Haloperidoli 1 – 2 ml 0.5 % solution i/m;
- Droperidoli 1 ml 0.25 % solution i/m are used.

At **seizures**:

- Sibazoni 2 – 4 ml 0.5 % solution i/v, i/m;
- Natrii oxybutiras 10 ml 20 % solution i/v are used.

Surgical treatment is used at lateral or lobar hemorrhages less than 100 ml.

At subarachnoid hemorrhages surgical treatment is recommended during first 48 hours or on the second week.

In renewal period we prescribe:

- Cerebrolizini 5 ml i/m, i/v 2 – 3 times per day;
- Piracetami 10 ml 20 % solution i/v, i/m;
- Instenoni 2 ml i/m;
- Actovegini 2 ml i/m twice a day.

Differential treatment of brain infarction

The main directions are:

1. To renew blood circulation in zone of ischemia.
2. To correct rheologic and coagulative properties of blood, to improve microcirculation.
3. To prevent disorders of cerebral metabolism.
4. To decrease brain edema.

5. To treat brain hypoxia.

1. To renew blood circulation in zone of ischemia:

- a). Actilaza 100 mg i/v by drops every 2 – 3 hours;
- b) Inhibitors of glutamat excretion (difenin, nimotop, MgSO₄) are used. Nimotop is used 15 mg in 1500 ml of physiologic solution i/v by drops, or in tablets 30 – 60 mg 4 times per day.

In order **to improve perfusion** we use cavinton 20 mg i/v by drops.

At hyperperfusion we use:

- Euphyllini 10 ml 2.4 % solution i/v;
- Pentoxiphyllini 5-10 ml i/v;
- Diuretics (manitol 15 % 100 – 200 ml i/v by drops);
- Albumini 100 ml i/v by drops.

2. To correct rheologic and coagulative properties of blood, to improve microcirculation:

- a) **anticoagulative therapy** is used only at progressive stroke:
 - Heparini 5 000 U s/c 4 times per day during 5 – 7 days, the 2 500 U during next 3 –4 days;
 - Fraxiparini is considered to be even more effective s/c.
- b) **Antiagregants** are used also:
 - Pentoxiphyllini 5 – 10 ml 2 % solution i/v by drops during 10 days, then 200 mg 3 –4 times per day up to 1 month;
 - Sermioni 4 mg i/v by drops during 10 days, then 1 tablet 3 times per day up to 1 month.
 - Ticlid 250 mg twice a day;
 - Aspirini 250 mg once a day;
 - Dipyridamoli 1 – 2 ml i/v by drops during 10 days, then 25 mg 2 –3 times per day.

c) Hemodilution is reached by introduction of reopoliglucini 400 ml i/v by drops during 5 – 7 days.

3. To decrease brain edema:

- Glycerini 1 g per 1 kg is used i/v;
- Dexamethazoni 16 – 32 mg i/v, i/m per day.

4. To treat brain hypoxia:

- Vit E 1 ml i/m;
- Piracetam 10 – 20 ml i/v by drops.

In case of stenosis, occlusion of MAH surgical treatment can be used.

Prognosis During the first two days the state of patients with

brain infarction is very severe. In a few days it is going better. Recurrent stroke can occur during the first year, sometimes during the first 2 or 3 years.

Prevention means in time treatment of heart – vascular diseases, hypertension and rhythm disorders.

sMeningitis

Meningitis is an acute infectious disease with involvement of the arachnoid and pia of the brain and spinal cord by pathogenic microorganisms. The most common **organisms that cause meningitis** are bacteria, viruses, sometimes fungus, micoplasma and rickettsia.

Classification

1. According to the inflammatory process in the meninges and cerebrospinal fluid (CSF) changes all meningitis are divided into **serous and suppurative, purulent**. At serous meningitis there is increased number of lymphocytes in CSF, while at suppurative and purulent meningitis neutrophiles dominate.
2. According to the pathogenesis there are **primary and secondary meningitis**. Primary meningitis are developed without previous general infection or infectious disease of any organ. Secondary ones are developed in case of general or focal infectious disease, that means that they are complications of different infectious diseases. For example tuberculous, syphilitic meningitis are considered to be secondary meningitis.
3. According to the localization of pathologic process there are: **general, basal, convexital meningitis**.
4. According to the rate of development there are **fulminant, acute, subacute, chronic meningitis**.
5. According to the severity there are **mild, moderate and severe meningitis**.
6. According to the etiology there are **bacterial, viral, fungal meningitis**.

Pathogenesis

There are three ways of meninges infection:

1. In case of open head or spinal trauma, fractures of skull basis associated with liquorrhea.
2. Direct extension (sinusitis, mastoiditis, middle ear infection).
3. Hematogenic way of infection (the source of infection is localized in intestine, stomach, nose).

Organisms multiply rapidly in cerebrospinal fluid with inflammation of meninges. Neutrophiles react with release chemicals. Bac-

terial toxins and inflammatory response cause brain edema. Increased metabolic demand and reduced blood flow result as inadequate tissue perfusion. That causes raised intracranial pressure and systemic effects of infection.

Pathomorphology

The main changes at suppurative and purulent meningitis are:

- a) Infection and inflammatory response are generally confined to the subarachnoid space and the arachnoid and pia.
- b) Brain and meninges edema.
- c) Blood repletion of cortical veins
- d) Internal hydrocephalus
- e) Perivascular infiltration of cortical cells

At serous meningitis meninges and brain edema, dilatation of liquor spaces is usually observed.

At tuberculous meningitis basal exudation occurs. As a result of commissural process hydrocephalus and subarachnoid space obstruction can occur.

Clinical features

In spite of the etiology there are three common syndromes for all meningitis:

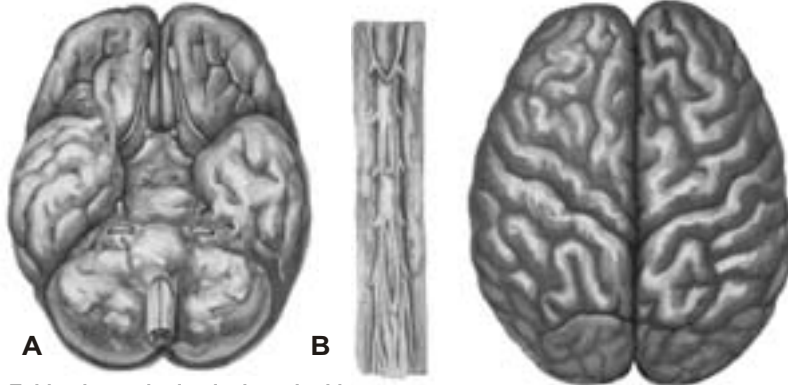
1. General – infectious
2. Meningeal
3. CSF changes

1. General – infectious syndrome means headache, shiver, inflammatory changes of blood, rash, tachycardia, tachypnoe with rhythm disorders.

2. Meningeal syndrome (fig. 66) manifests as neck stiffness, Kernig sign, Brudzinski signs (upper, middle and lower ones), Lessage symptom (in children), Bechterev's cheek bone phenomena. Increased intracranial pressure causes bulging of an unclosed anterior fontanelle in children.

Upon meningeal symptoms **general cerebral signs** are developed. They are headache (as a result of increased intracranial pressure), vomiting, nausea, general hyperesthesia, changes of consciousness, psychomotor agitation, and seizures.

Meningeal syndrome



Epidemic cerebral-spinal meningitis
 a) external view of the brain
 b) external view of the spinal cord

Hemorrhagic leptomeningitis

Meningeal symptoms (complex):

1. Headache
2. Vomiting
3. Consciousness disorders
4. Total hyperesthesia
5. Meningeal symptoms: (Kernig's sign, nuchal rigidity, and meningeal pose)
6. Seizures



Meningeal pose



Meningococemia
Hemorrhagic exanthema
(Testing of nuchal rigidity)



Testing of Kernig's sign

Fig. 66.

3. CSF changes manifest as cellular – protein dissociation, colour changes, cerebrospinal fluid is cloudy (purulent), the sugar content is decreased; clot formation on standing is observed at tuberculous meningitis.

The results of CSF examination have an important meaning for differential diagnosis and prescription of etiologic treatment. In order to put a diagnosis we should make bacteriologic and serologic examinations. Usually we use immunologic express – methods of investigation. They are:

- Countercurrent immunoelectrophoresis (CIE)
- Latex agglutination

These methods are used in order to reveal specific antigens (protein components of organisms). It is necessary to find out sensation of organisms to antibiotics in order to prescribe a proper therapy.

Epidemic cerebrospinal meningitis

Etiology

It is usually caused by *Meningococcus Weichselbaum*. The source of infection is ill person or carrier. Meningococcal infection can manifest as suppurative meningitis, asymptomatic bacterio – carriage, nasopharyngitis, arthritis, meningococemia.

The entrance is mucose membrane of pharynx and nose. Meningococcal infection can be transmitted by air drops or contact. In most instances the spread of meningococcus is haematogenous. Usually this disease is observed in winter and spring. Meningococcus is instable at high temperature, humidity and day light.

Pathogenesis

Meningococcus in upper respiratory ways usually causes primary nasopharyngitis. The last in typical cases is rapidly cured. In those persons who are less resistant to infections meningococcus is getting into the blood and then it is spread in the human body. In severe cases meningococemia develops, which is usually associated with typical hemorrhagic rash. Endotoxin can even cause endotoxic shock.

Clinical features and diagnosis

Incubation period of meningococcal infection lasts from 2 to 10 days (usually about 3 – 7 days). The onset of meningococcal men-

ingitis is acute (about several hours). The temperature is elevated at 39 – 40°.

The main peculiarities of clinical picture are:

1. General cerebral syndrome is well – expressed and increases rapidly. Meningeal syndrome increases during 2 – 3 days (fig. 67).
2. Focal neurologic symptoms manifest as lesion of III-rd and IV– th CN's – diplopia, ptosis, cross – eye, anizokoria.
3. Herpes labialis is often observed on the 2 – nd or 5 –th day. Sometimes hemorrhagic rash occurs, that is the sign of meningococemia.
4. Sometimes brain tissue is involved. Then there is clinical picture of meningoencephalitis. That means occurrence of hyperkinesis, ataxia, nystagmus, paralysis, seizures, sometimes epistatus.
5. At severe meningoencephalitis inflammation of ventricles occurs. The last manifests as hornetonia, edema of optic nerves' disk, disturbances of breathing and cardiac activity.
6. Cerebrospinal fluid is cloudy (purulent), the cell count in the fluid is usually between hundreds and thousands/mm³. The protein content is increased to 10 – 15 g/l. The sugar content is decreased. Meningococcus can be found in CSF.

According to the clinical picture there are three main forms of meningitis. They are mild, moderate and severe one. Fulminant hypertoxic form is very rare.

Complications of meningococcal meningitis Severe forms can be associated with pneumonia, myocarditis, pericarditis. The



Fig. 67. Kernig's sign.

most dangerous complications are acute brain edema and bacterial endo-toxic shock.

The course of meningococcal meningitis There are fulminant, acute, abortive and recidivous course of the disease. The most common are the first and the second forms.

Diagnosis It is usually based on clinical features (acute onset, general infectious, general cerebral symptoms and meningeal syndrome, hemorrhagic rash), CSF examination and revealing of meningococcus.

Differential diagnosis We should differentiate this disease with:

- other forms of meningitis;
- meningism at general infections (that means the absence of CSF changes in case of meningeal syndrome);
- subarachnoid hemorrhage (usually general – infectious syndrome is absent, there is no blood in CSF).

Prognosis The disease lasts from 2 to 6 weeks. In case of adequate and in time treatment it is usually cured.

Only functional changes of CNS such as asthenic syndrome, mild disorders of mental development may be observed after meningococcal meningitis. Focal symptoms such as paresis of extremities, CN's, hydrocephalus, seizures, decreased vision, hearing loss, and arachnoiditis are very rare.

Secondary purulent meningitis

Etiology, pathogenesis

Secondary purulent meningitis are observed in case of pyogenic source in the body. The disease is developed after direct extension from pyogenic source (for example pyogenic middle ear infection) or by means of metastasis from faraway pyogenic focuses (abscess, ulcerous endocarditis).

Usually secondary purulent meningitis is caused by different coccus (pneumococcus, staphylococcus), Hemophilus influenzae.

Clinical features

The onset of the disease is accompanied by chills and fever to 40°. Then meningeal syndrome, seizures, consciousness disorders, focal neurologic symptoms, autonomic disorders occur. Sometimes the clinical picture is very similar to sepsis.

In CSF the cell count is very high. The protein content is significantly increased. Bacteria (staphylo–, streptococcus) are usually found.

Treatment

The measures of treatment of primary suppurative and secondary purulent meningitis are the same. But the secondary purulent meningitis needs liquidation of the source of infection. If necessary we can use surgical methods of treatment.

Treatment of suppurative, purulent meningitis

The patient should be hospitalized in horizontal position and isolated before the revealing of the type of meningitis.

We use 3 kinds of treatment:

1. Etiotropic.
2. Pathogenic.
3. Symptomatic.

1. Etiotropic treatment

A. Antibiotics

Just after the putting diagnosis we should prescribe antibiotics. It's a pity but sometimes it is difficult to reveal the organism that causes the disease. And we lose time. That's why very often we start with antibiotic that has wide spectrum of action – **Penicillinum**.

We use it in dose 300 000 U per kg (that means about 24 – 32 mlns of U per day 6 – 8 times i/m). At severe cases the dose is increased to 48 mlns. When the treatment is started later or the patient is in coma the dose is 60 mlns of U per day. And the first 4 – 12 mlns are used i/v. Simultaneously we prescribe 3 mlns of U of Nystatinum. The treatment lasts from 7 to 10 days. The main sign which indicates us it's time to put off the medication is CSF sanitation (that means cell counting less than 100/mcl, lymphocytes not less than 75 %).

- For purulent meningitis treatment we can prescribe half– synthetic Penicillinum (ampicillinum) in dose 300 – 400 mg per kg per day i/v or i/m.
- Cephalosporines are also used for the treatment. For example such as Cephazolinum, Cephalomycinum, Ceftriaxon, Cefotaxim. They are used 1 g 4 times per day i/m or ii/v.

- Aminoglycozides, such as Gentamycini, Kanamycini, Amikacini are also prescribed. Amikacinum is used 0.5 g 3 times per day. Gentamycinum is used 80 mg 2 – 3 times per day.
- The most effective are combinations of different antibiotics. For example:
 1. Penicillinum and Aminoglycozides (Gentamycinum, Kanamycinum);
 2. Ampicillinum and Aminoglycozides;
 3. Penicillinum and Cephalosporines of the second or third generation;
 4. Aminoglycozides and Cephalosporines;
 5. Tienamycinum.

B. Sulphanilamide

Sulfamonomethoxine, Sulfapyridazinum, Sulfadimethoxinum are used 2 g twice a day during the first day, then 2 g once a day.

C. The treatment of secondary purulent meningitis includes **treatment of the source of infection** (inflammation process in lungs, middle ear, and nose). Sometimes we use surgical methods of treatment in order to liquidate the source of infection.

2. Pathogenetic treatment includes:

- Treatment of intoxication
- Correction of heart – vascular and breathing disturbances
- Liquidation of brain edema
- Correction of water – electrolytes balance.

Treatment against intoxication

1. To **liquidate hypovolemia** we prescribe Rheopolyglucinum 400 ml per day, 100 ml 5 % Albuminum, Plasma 100 ml, Haemodesum 200 ml per day.

An average quantity of liquids is about 2 – 2.5 l per day. After introduction of 1 – 1.5 l we prescribe 2 – 4 ml of Lasix.

We also use polarized compound (500 ml 5 % Glucose, 150 ml 1 % KCl, 10 U of Insulinum, 10,0 Pananginum, ATP, Cocarboxylase) and diuretics. In order to overcome metabolic acidosis we use Na hydrocarbonas 100 ml 4 %.

2. **Hormones** are necessary in case of infectious – toxic shock

treatment. We use Dexamethazonum 8 mg twice a day or Hydrocortisone 15 mg per kg. Glycosides are also used in this case.

Treatment against brain edema

1. **Diuretics** such as Manitol, Lasix, Glucocorticoids (Dexametasonum), Albuminum.

Another medicines are prescribed as components of pathogenetic treatment

1. **Heparinum** is prescribed in dose 5 000 U 4 times per day.

2. **Antihistamine medications** (CaCl₂, Ca gluconas, Suprastinum, Dimedrolum, Tavegil) are used too.

3. **Nootrops** such as Piracetam, Nootropil, Actovegin, Instenon, and Cerebrolisyn are used.

3. Symptomatic treatment

At **acute kidney insufficiency** we use hemodialysis.

At **acute suprarenal glands insufficiency** we introduce physiological solution, Rheopolyglucinum plus 125 – 500 mg Hydrocortisone, 30 – 60 mg Prednisolone, 8 – 16 mg Dexamethasone, 500 – 1000 mg Ascorbin acid, Cordiamini, Strophanthinum.

At **brain hypoxia** we use nootropils and neuroleptics (Na oxybutiras, Relanium 0.5 % 2 ml, Sibazonum 2 – 4 ml).

At **severe pain** we use analgesics (Analginum, Baralginum, Tramadolium).

Prognosis About 20 – 25 % of all patients with suppurative, purulent meningitis die. In case of different complications and late diagnosis 60 – 80 % of all patients die. Death is much more common at secondary purulent meningitis and at pneumococcal infection.

Serous meningitis

According to the etiology there are:

- Bacterial serous meningitis (tuberculous, syphilitic meningitis)
- Viral meningitis (acute lymphocytic choriomeningitis, parotid meningitis, influenza– meningitis, enteroviral, herpes – adenoviral – meningitis)
- Fungal meningitis

Tuberculous meningitis

Tuberculous meningitis is always secondary to tuberculosis elsewhere in the body. The primary focus of infection is usually in the lungs

but may be in the lymph glands, bones, nasal sinuses or any organ of the body. This disease is observed in both – children and adults.

Pathomorphology

The process is usually most intense at the base of the brain, ependyma, III –rd and IV – th ventricles, choroids plexus. The inflammatory process may extend for a short distance into the cerebral substance. Proliferative changes are frequently seen in the inflamed vessels of the meninges, producing a panarteritis. All these changes lead to the CSF dynamics disturbances and hydrocephalus. The most typical feature of tuberculous meningitis are minute tubercles on the base of the brain (fig 68).

Clinical features and diagnosis

1. Tuberculous meningitis develops very slowly.
2. There are three periods of the disease:
 - a) **prodromal** stage (with loss of appetite, general weakness, headache, irritability, increased temperature and sometimes vomiting) lasts for 2 – 3 weeks;
 - b) **meningeal** stage;
 - c) **paralytic** stage.
3. Meningeal syndrome is not well – expressed.
4. Temperature is increased up to 38° sometimes 39°.
5. Focal neurologic signs – paresis and paralysis of Oculomotor n. (III), Abducens n. (IV), Facial n. (VII), Optic n. (II). Sometimes Vestibulocochlear n. (VIII) is involved.
6. Encephalitic syndrome manifests as paresis, paralysis, aphasia, hyperkinesis, cerebellar disturbances.
7. Subacute course of the disease. Sometimes acute course is observed in children. Chronic course may be in those persons who had used antituberculosis treatment previously.

Diagnosis

The CSF findings are quite characteristic. These include:

- Increased pressure;
- Slightly cloudy;
- Increased protein content to 1 – 5 g per l;
- Moderate pleocytosis of 100 – 300 cells/mmi, 70 – 80 % of them are lymphocytes;

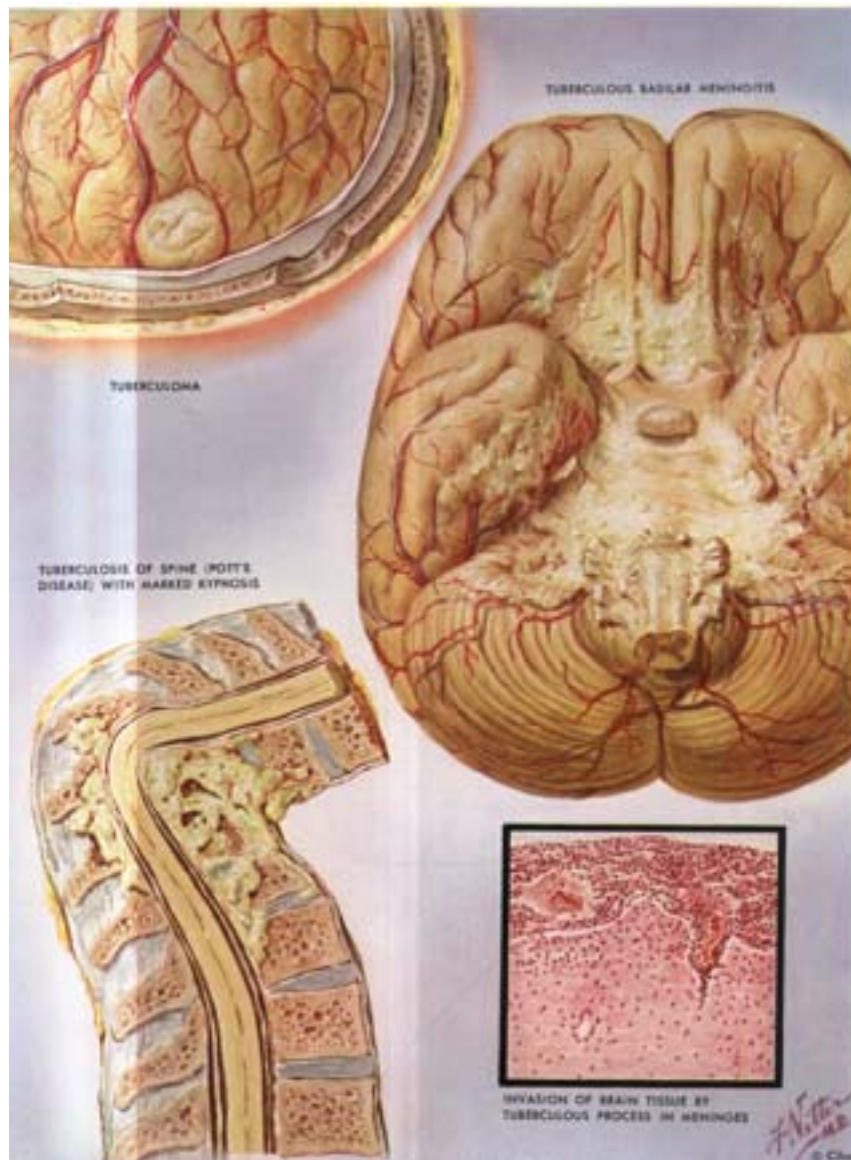


Fig. 68. Tuberculosis of the Brain and Spine (Frank H. Netter, M.D.)

- Decreased sugar content with values in the range of 0.15 to 0.3 g/l;
- Formation of a clot on standing during 12 – 24 hours, where bacilli of tuberculosis are usually found out.

In blood sometimes leucocytosis and increased SR (sedimentation rate) is observed.

Differential diagnosis Spinal form of tuberculous meningitis should be differentiated with spinal tumor or myelopathy.

Treatment

Etiotropic treatment includes:

- Isoniazid 15 mg/kg 3 times per day;
- Rifampicinum 600 mg once a day;
- Pyrazinamide 30 mg/kg (1.5 – 2.5 g per day) twice a day.

They are used during three months. Then we use only Isoniazid and Rifampicinum during 7 months.

The most recent medications are combinative ones such as Rifogal, Rifater (Rifampicinum 120 mg, Isoniazid 50 mg, Pyrazinamide 300 mg).

Streptomycinum is used in dose 1 g once a day.

Ethambutol – 25 mg/kg per day

Usually we use all these medications about three months, as all of them have side effects such as ototoxicity, lesion of optic nerve, diarrhea, thrombocytopenia, hepatitis.

Pathogenetic and symptomatic treatment is the same as at suppurative and purulent meningitis.

Prognosis Mortality is about 10 %. Residual symptomatics includes epileptic attacks, eye movement disorders, hemiparesis, hearing loss, neuroendocrine disturbances.

During the first year of the disease the patients are considered to be disabled, during the next 2 or 3 years they have limited abilities.

Serous viral meningitis

Acute lymphocytic choriomeningitis

The **cause of the disease** is Armstrong virus. **Viral reservoir** is domestic mice. Human being is infected by means of food infected by mice. The main way of disease transmittance is hematogenic.

Clinical features

1. Incubation period lasts for 1 – 2 weeks.
2. The onset is with headache, running nose, sore throat.
3. Moderate meningeal syndrome against high temperature – 39° – 40°.
4. On 7–th or 10–th day general–infectious and meningeal signs disappear.
5. CSF is pellucid, pleocytosis is 300 – 500 cells, most of them are lymphocytes. The content of protein and sugar is the same.
6. There are some atypical forms of meningitis, such as:
 - Flu – like;
 - Encephalomyelitic;
 - Poliradiculoneuritic;
 - Visceral.
7. Alopecia and orhitis can occur at the 7 th week of the disease.

Diagnosis According to the virusologic and serologic examination we put the diagnosis. For these we use blood and CSF during the beginning of the disease.

Treatment

Etiotropic treatment

1. Interferon 500 000 U. It is prescribed in dose 1 amp. once a day during 7 days or Reaferonum 1 mln U i/m
2. Virolex – 250 mg 3 – 4 times per day or Aciclovir 200 mg 3 times per day, or Virasolom 2 caps. 3 – 4 times per day during 7 days.
3. Stimulators of interferonum production – Prodigiosan – 1 ml 0.005% i/m once every three days, Cicloferonum 2 ml i/m according to the schema.
4. Acidi ascorbinici, vit. B1, B6.
5. Antibiotics in order to prevent pneumonia.

Pathogenetic treatment for all types of meningitis:

1. Desintoxication (Rheopolyglucinum 400 ml i/v per day, Plasma 100 ml i/v per day).
2. Dehydratation (Mannit, Lasix, Albuminum).
3. Desensibilization (Suprastin, Dimedrol, Klaritin).
4. Hormones (Dexamethazonum, Prednizolonum).
5. Nootrops. (Is the same as at all the others meningitis).

Prevention To liquidate mice in the houses.

Enteroviral meningitis

The causes of the disease are Coxsacki and ECHO (enteric cytopathogenetic Human organs) virus.

Clinical features

1. The disease is much more common in summer and autumn;
2. Epidemic focuses are very typical;
3. It is highly contagious disease;
4. Incubation period lasts for 10 days approximately;
5. The onset is acute with high temperature, well – expressed general cerebral syndrome (loss of consciousness, psychomotor agitation, seizures);
6. Abdominal pain, diarrhea;
7. Enlargement of liver and spleen;
8. Myalgia;
9. Herpes –angina;
10. Rash;
11. Lymphatic nodules enlargement;
12. CSF – pleocytosis 100 – 500 cell/mm³, most of the cells are lymphocytes;
13. In 2 – 3 days the temperature is decreased, meningeal syndrome disappear.

Diagnosis

1. Virusologic examination of smears;
2. RN;
3. RCC.

Treatment

Pathogenetic and symptomatic treatment is the same as at all the others meningitis.

- gamma-globulin is used in dose 6 ml i/m once a day up to 5 days;
- RNA is prescribed in dose 30 mg 6 times per day up to the normalization of temperature;
- Antibiotics and sulphanilamides are prescribed in case of complications (otitis, pneumonia, tonsillitis);
- At myalgia we use Indometacinum, Metindolum, Ortofenum.

Parotid meningitis

Usually this disease is observed in children, especially boys. The signs of meningitis appear on the 3-rd or 5 th day after parotitis. During that period virus can get into the other organs, such as pancreas and testis and as a result they can cause pancreatitis and orchitis. The virus is transmitted by air drops. Incubation period lasts for 3 weeks.

Clinical features and diagnosis

The onset of the disease is acute. There are well expressed general infectious, meningeal and general cerebral syndromes. In children general weakness, sleepiness, sometimes agitation, hallucinations, consciousness disorders, general seizures are expressed.

There are different **forms of parotid meningitis**:

- Serous meningitis;
- Meningism without meningitis;
- Clinically asymptomatic meningitis.

The **CSF findings** are quite characteristics. These include:

- Increased pressure;
- Slightly cloudy;
- Increased protein content to 0.6 – 1 g per l;
- Moderate pleocytosis;
- Sugar content without changes.

Clinical picture mainly depends on increased intracranial pressure and not cytosis. All symptoms of the disease tends to disappear in 7 – 10 days. CSF returns to normal content a little bit longer.

Complications Orchitis, pancreatitis.

Treatment the same measures are used as at others forms of meningitis.

Brain abscess

Encapsulated or free pus in a substance of the brain tissue following an acute purulent infection is known as brain abscess. Abscesses may vary in size from a microscopic collection of inflammatory cells to an area of purulent necrosis involving the major part of one hemisphere. Abscess of the brain has been known for over two hundred years but the surgical treatment started with Macewen in 1880.

Etiology.

Brain abscesses arise either as direct extension from infections:

- within the cranial cavity (mastoid, nasal sinuses, osteomyelitis of the skull),
- from infections secondary to fracture of the skull,
- as metastases from infection elsewhere in the body.

Brain abscess occurs in about 2% of patients with congenital heart disease. It is generally believed that metastatic brain abscesses are usually multiple.

The infecting organism may be any of the common pyogenic bacteria, but *Staphylococcus aureus*, *Streptococcus viridans*, hemolytic streptococcus, Enterobacteriaceae and anaerobes such as *Bacteroides* are most commonly found. Pneumococci, meningococci and *Haemophilus influenzae* are rarely recovered from brain abscesses. Not infrequently the abscess will be sterile by the time operation is performed. Brain abscess is an infrequent complication of infection with the *Entamoeba histolytica*.

Pathology The pathological changes in brain abscess are similar regardless of whether the infection extends to the brain directly from an epidural or subdural infection, by a retrograde thrombosis of veins or by metastasis through the arterial system (fig 69-70).

In the first stage there is a suppurative necrosis of brain tissue (purulent encephalitis). When immunological forces control the spread of the infection the macroglia and fibroblasts proliferate in an attempt to surround the infected and necrotic tissue. The success of the body in limiting the spread of infection and the density of the capsule are the main factors in the final outcome. Edema of the adjacent portion of the cerebrum or of the entire hemisphere is a common finding.

Incidence. Brain abscesses are relatively rare and constitute less than 2% of the patients who are referred to a neurosurgeon for intracranial surgery. Brain abscess may occur

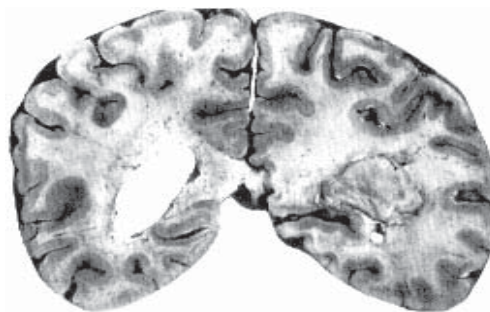


Fig. 69. Abscess of the temporal-occipital zone.

it any age but it is more common in the first to third decades of life. The high incidence of mastoid and nasal sinus disease at this period. Males are slightly more frequently affected than are females.

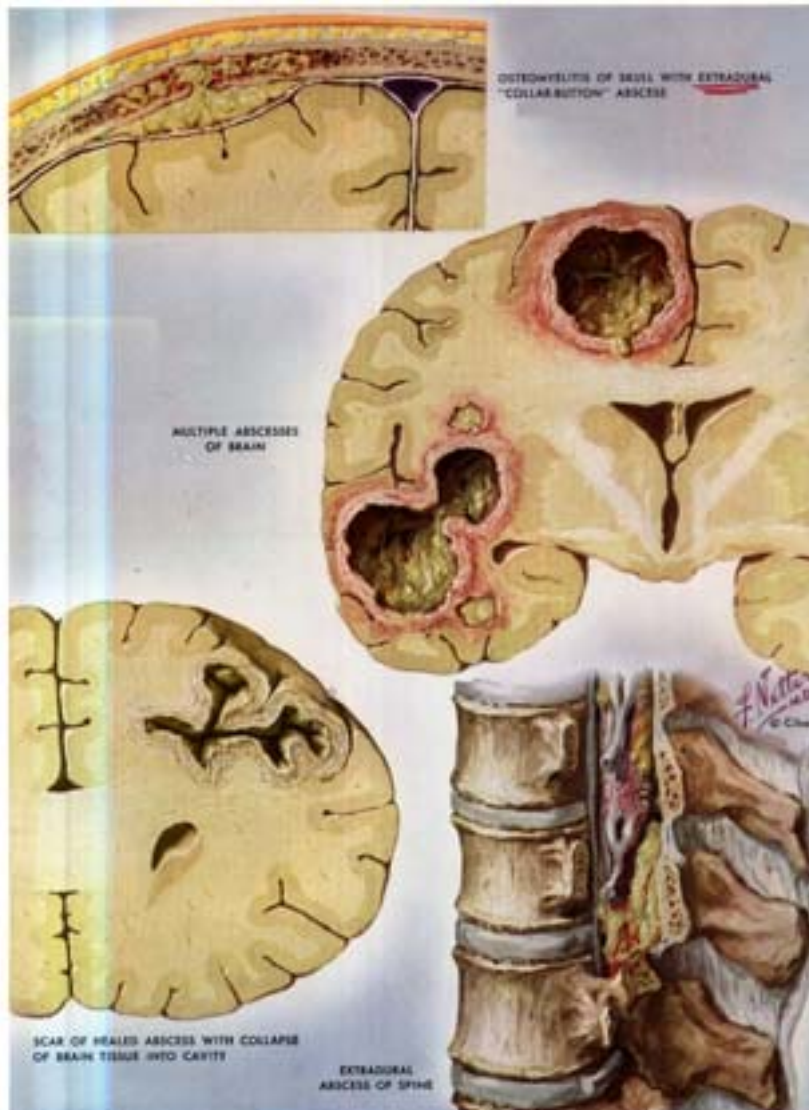


Fig. 70. Abscess of the Brain and Spine (Frank H. Netter, M.D.)

Clinical features. The symptoms of brain abscess are essentially the same as those of any expanding lesion in the brain.

General symptoms.

1. Symptoms of infection are lacking unless the focus which gave rise to the abscess is still active.

2. Chills and fever at the onset of the invasion of the nervous system are said to be present in some cases.

3. Since edema of neighboring brain tissue is common, increased intracranial pressure develops rapidly. Thus headache, nausea and vomiting are early symptoms. Convulsions, focal or generalized, are common.

4. The body temperature is normal or subnormal except when it is elevated as the result of other complications or activity of the septic focus which was the source of the abscess.

5. The pulse and respiratory rates are normal unless the intracranial pressure is greatly increased.

6. Signs of injury to the third or sixth cranial nerve as a result of increased intracranial pressure are occasionally seen.

Focal signs include:

1. Hemiparesis or hemiplegia when the abscess is in the cerebral hemispheres;

2. Torpidity and mental confusion are especially prominent with abscesses in the frontal lobe;

3. Hemianopsia and aphasic disturbances, when the temporal or parieto-occipital lobes are involved;

4. Ataxia, intention tremor, nystagmus and other symptoms of cerebellar and vestibular dysfunction when the abscess is in the cerebellum.

5. Abscesses in the brain stem are distinctly rare, and the diagnosis is usually established at necropsy. Fever, headache, cranial nerve palsies, hemiparesis, dysphagia and vomiting were the common symptoms.

Diagnostics.

The diagnosis of brain abscess can be made without difficulty when convulsions, focal neurological signs or increased intracranial pressure develop in a patient with an acute or chronic infection in the middle ear, mastoid, nasal sinuses, heart or lungs or with congenital heart disease. The diagnosis may be very difficult in case of the absence any focus of infection.

Examination of the blood and urine is normal unless there is activity in the septic focus or unless complications are present.

1. In the cerebrospinal fluid are related to the size, location and stage of development of the abscess and to the presence or absence of acute meningitis. When the latter is present, the cerebrospinal fluid findings are those commonly seen in acute purulent meningitis. The pressure is elevated in almost all cases. The fluids are usually clear and colorless but they may be cloudy or turbid in the early stages and a fine clot may form in fluids with a high cell count. The cell count varies from normal to a thousand or more cells per mm³. The protein content is moderately increased in about 75% of the cases. The sugar content of the fluid is normal.

2. The optic disc are choked in over 50 % of the cases, depending to some extent upon the site of the abscess. inequality in the degree of the choking in the two eyes has no lateralizing value.

3. The electroencephalographic changes are similar to those which are found in cases with other space-occupying lesions.

4. Roentgenograms of the skull may show separation of sutures in infants or children and an increase in the convolution markings. Widening of the sella turcica, thinning of the clinoid processes and displacement of the pineal gland may occur in adults.

5. Computerized tomography (CT) scan and MRI with contrast enhancement permits accurate determination of capsule formation and precise localization of the abscess.

Differential diagnosis.

The common differential diagnoses which must be considered are:

1. Brain tumor;
2. Extradural or subdural abscess;
3. Sinus thrombosis;
4. Encephalitis and meningitis.

Most of these conditions can usually be differentiated by the history of the development of symptoms and the results of roentgenography of the skull, electroencephalogram and CT and MRI brain scan.

Prognosis. The outcome of untreated brain abscess is, with rare exceptions, fatal. The mortality in surgically treated cases varies depending upon various factors:

- Location and degree and encapsulation of the abscess,
- site of the original infection,
- presence of complications
- whether there are single or multiple abscesses.

Treatment. The treatment of brain abscess is surgical evacuation of the pus. The operation is usually delayed until the abscess is firmly encapsulated.

The surgical treatment of brain abscess varies from complete extirpation to repeated drainage through a trephine opening. The ideal treatment is complete removal of the abscess within its capsule.

Any method of surgical treatment should be combined with the administration of antibiotics in full therapeutic doses both pre- and postoperatively.

Encephalitis

Encephalitis – means inflammation of the brain tissue. In pathogenesis of encephalitis besides infectious one there are three more factors. They are:

- Infectious – allergic;
- Toxic (in this case it is known as encephalopathy);
- Allergic.

Classification

Encephalitis are divided into:

- Primary;
- Secondary.

Primary encephalitis

I. Viral:

1. Seasonal (acaridan, gnat);
2. Without seasonal characteristics:
 - a. Enteroviral;
 - b. Herpetic;
 - c. Flu – associated;;
 - d. HIV – associated.
3. Caused by unknown virus – epidemic.

II. Microbial (at neuro – lues, rheumatism).

Secondary encephalitis

1. Parainfectious (at measles, small pox, rubella, scarlet fever, poliomyelitis).
2. Postvaccinal (after antirabies, antipoliomyelitic, antidiphtheric vaccine).

According to the spreading of the pathological process encephalitis are divided into:

1. Leukoencephalitis – when white substance of the brain is mainly involved;
2. Polioencephalitis – when gray substance of the brain is mainly involved;
3. Panencephalitis – when there is diffuse lesion of the neurons of main pathways of brain and spinal cord.

The most common are parainfectious and postinfectious encephalitis. They take about 50 % of all encephalitis.

Among the primary encephalitis the most common are viral encephalitis (such as enteroviral, flu – associated, herpetic ones)

The common syndromes for all encephalitis are:

1. General – infectious;
2. General – cerebral;
3. Focal.

Epidemic encephalitis

Etiology. Etiological agent is unknown. It is supposed that this disease is caused by virus.

Pathogenesis. Virus penetrates through the mucous membrane of the nose, pharynx into the subarachnoid space of the brain and causes inflammation. This disease is much more common in winter and spring.

The source of infection is ill persons or healthy carriers

The incubation period lasts from 1 to 14 days. Infection is transmitted by air drop way.

It is much more often observed in 20 – 30 years old persons.

Pathology

Vascular – inflammatory changes are mainly localized in central ganglions of hemispheres and brainstem. Usually the lesions are observed in midbrain, red nuclei, black substance, thalamus, hypothalamus and reticular formation (fig 71).

In acute stage exudation and proliferation takes place.

In chronic stage degenerative changes of the neurons are observed.

Clinical features

There are two stages of the disease:

- Acute.
- Chronic.

There are three main symptoms in **acute stage**:

1. Fever;
2. Eye movements disorders;
3. Sleep disorders.

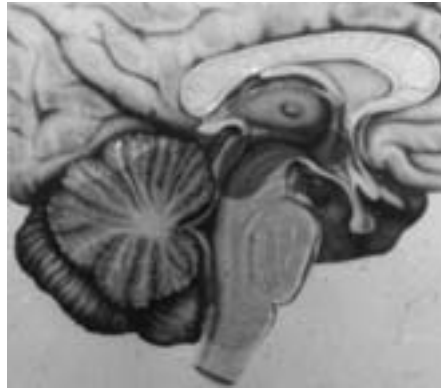
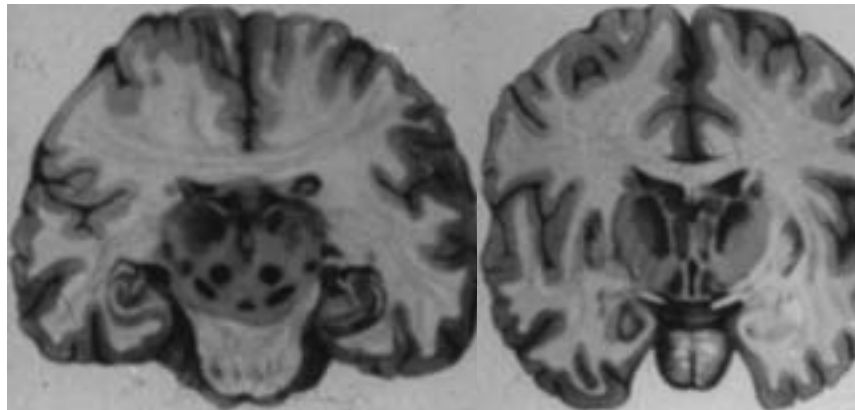


Fig. 71. Localization of lesion:
a) acute stage

All of these symptoms are present at **typical form** of epidemic encephalitis.

1. Fever. It manifests as increased temperature to 38 – 39° C against general cerebral symptoms, such as headache, nausea, vomiting, muscle pain. Sometimes there are complaints on upper respiratory ways disorders. This period lasts for about 2 weeks.

2. Sleep disorders. This syndrome usually manifests as



b) chronic stage

pathologic sleepiness. Sometimes pathologic insomnia can occur. In some cases the patient sleeps in day – time and cannot fall asleep at night.

3. Eye movement disorders. These symptoms usually occur in case of nuclei of Oculomotor nerve lesion. It manifests as diplopia, anizokoria, gaze paresis, converse symptom of Argil – Robertson, ptosis, cross eye, midriasis, nystagmus, ophthalmoplegia (fig. 72).



Fig. 72. Ophthalmolpegia externa

Besides these three main syndromes of acute period sometimes we can observe the others. They are:

1. Vestibular disorders;
2. Lesion of VII, XII, V CN's;
3. Hyperkinesia;
4. Hypothalamic disturbances: disturbances of sweating, salivation, fatty skin, loss of flesh.

Against general cerebral symptoms consciousness disorders appear such as – sopor, psychomotor agitation, fear, hallucinations.

In blood analysis there are leucocytosis (lymphocytic), increased SR.

In CSF there is slight lymphocytic pleocytosis to 20

– 40 cells in 1 mm³, slightly increased sugar and protein content.

Besides typical form of acute stage sometimes **atypical forms** are observed. They are:

1. Abortive – when all the symptoms of acute stage are not well expressed.
2. Oculocephalgic – when eye movements disorders and severe headache dominate over the other symptoms.
3. Vestibular – when vestibular syndrome dominates (there are dizziness, vomiting, ataxia).
4. Hyperkinetic – against the symptoms of acute stage hyperkinesia occur. Sometimes in this case sleep disorders, amyostatic symptoms and thalamic pains are observed.
5. Flu – associated – It looks like flu – infection. General infectious symptoms prevail.
6. Rare forms:
 - Epidemic hiccup;

- Peripheral form;
- Pseudoneurastenic.

Course of the disease Acute stage lasts from 4 days to 1–4 months. It is characterized by remission and exacerbations. Usually it is cured. In 35 – 50 % of all patients the results of acute stage is a chronic one.

Chronic stage (fig. 73).

It manifests as Parkinson syndrome in 90 % of patients or Hyper-kinetic syndrome in 10 %. There are three main groups of symptoms at Parkinson syndrome:

1. Akinesia or hypokinesia;
2. Plastic hypertonia;
3. Static tremor.

Chronic stage lasts for several years. Usually it is not cured. Differential diagnosis should be made with Parkinson disease, vascular, posttraumatic, neuroleptic, toxic Parkinson disease.

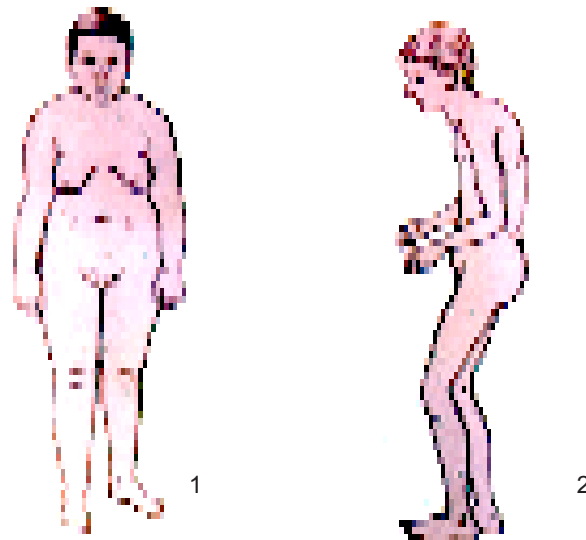


Fig. 73. Chronic stage syndromes of epidemic encephalitis:
1. Neuroendocrine disorders; 2. Parkinsonian syndrome

Parkinson disease – is hereditary degenerative disease which usually occurs at the age of 45 – 60 years. Tremor prevails over hypokinesia. It is a family disease. The symptoms of acute stage are absent.

Vascular Parkinsonism – occurs at the age of 60 and later. There are well expressed symptoms of cerebral atherosclerosis – memory disorders, the symptoms of discirculative encephalopathy (vestibulo – cerebellar, cephalic, pyramidal syndromes). There are vascular sclerotic changes on eye ground.

Neuroleptic Parkinsonism – occurs at long lasting usage of Galloperiolium, Tizercinum. At first autonomic disorders appear, then Parkinson syndrome occurs. It is also associated with hyperkinesia of oral muscles. There is dissociation of typical symptoms, for example akinesia with tremor without plastic hypertonus.

Traumatic Parkinsonism – is the result of cranial trauma. Clinical picture is characterized by memory disorders, symptoms of hydrocephalus, seizures, pseudobulbar syndrome, symptoms of CN's lesion and another symptoms.

Toxic Parkinsonism – is usually developed at Mn, CO intoxication. It is usually associated with polyneuropathy, psychiatric disorders, seizures, choreoatetosis.

Treatment

Acute stage

Etiotropic treatment

1. Interferonum 500 000 U. It is prescribed in dose 1 amp. once a day during 7 days or Reaferonum 1 mln U i/m.
2. Virolex – 250 mg 3 – 4 times per day or Aciclovir 200 mg 3 times per day, or Virasolom 2 caps. 3 – 4 times per day during 7 days.
3. Stimulators of interferonum production – Prodigiosan – 1 ml 0.005% i/m once every three days, cycloferoni, amizoni.
4. Acidi ascorbinici, vit. B1, B6.
5. Antibiotics in order to prevent pneumonia.

Pathogenetic treatment for all types of encephalitis (see treatment of meningitis):

1. Desintoxication.
2. Dehydratation.
3. Desensibilization.
4. Hormones.
5. Nootrops.

Symptomatic treatment

Chronic stage

1. Synthetic holinolytics: Cyclodolum (0.01, 0.005), Romparkin, Parkopan (0.001, 0.002);
Stimulators of dophamine secretion.
2. Amantadine medications increase sensation of dophamine receptors to dophamine, excite dophamine receptors. Midantan (0.1 3 times per day), Amantadinum.
3. Inhibitors of MAO (Jumex) – 5 mg 1 – 2 times per day.
4. Stimulators of dophamine receptors – Bromcriptine, Akineton, Norakin (0.001 – 0.002).
5. Medications that decrease converse catch of dophamine. Amitriptilinum, Amipraminum, Melipraminum.
6. Substitutional therapy: Sinemet 3 – 6 tablets per day. Nacom – 3 – 6 tablets per day.
7. In order to decrease tremor we use b – adrenoblockers: Anaprilinum 10 mg 3 times per day, Amitriptilinum 25 mg 3 times per day.
8. In order to decrease muscle tonus: Midocalm, Baclofen are used.
9. Nootrops.
10. Physiotherapeutic methods.

Spring – summer encephalitis

Etiology and pathogenesis

Etiological agent is a neurotropic virus of spring – summer encephalitis. It is resistant to the low temperature and it is unstable at temperature higher than 70⁰ C. Reservoir of virus is Acaridan. Simultaneously they are transmitters of the infection. The disease is much more common in spring and summer because of the biological peculiarities of Acaridan. This disease is widely spread in Siberian, in Ukraine – in Carpathian and Volyn region, also in Western European countries.

The person is usually infected by means of two ways. They are:

- bite of Acaridan;
- while using of row milk, cheese (made of wild goats).

Then with blood the virus penetrates into the central nervous system. In the third day after the bite the virus can be found in blood of the person. The incubation period lasts from 8 to 20 days.

Pathology

There are hyperemia, brain edema, inflammatory changes, foci of necrosis, vasculitis mainly in the oblongate brain and spinal cord. In chronic stage there are fibrous changes in meninges, arachnoid cysts in the anterior horns of the spinal cord and in the oblongate brain.

Clinical features (fig. 74).

At first headache and general weakness appears. The temperature is increased to 39–40° C. There are such syndromes:

- a) general – infectious (there is leucocytosis, increased SR in blood).
The temperature curve has 2 hills. The space between them is equal to several days. The second hill means that the virus is already in the central nervous system.
- b) General – cerebral (headache, vomiting, seizures, disorders of consciousness, hallucinations).
- c) Meningeal – neck stiffness, Kernig sign, Brudzinski signs.
- d) CSF disorders – (cytosis up to 100 cells, protein content up to 2 g/l).
- e) Sometimes there are diarrhea and abdominal pain.

Clinical forms of acute stage:

1. Polioencephalomyelitic (typical form);
2. Poliomyelitic;
3. Meningeal;
4. Encephalitic;
5. Meningoencephalitic;



Fig. 74. Acute stage syndromes of spring-summer encephalitis:

1. Meningeal posture; 2. "hanging head"; 3. Peripheral paresis of mimic muscles.

6. Not well expressed;
7. Polyradiculoneuritic.

1. Polioencephalomyelitic (typical form)

On the third – fourth day of the disease there are:

- a) Flaccid paresis or paralysis in the shoulders, neck muscles (as a result of lesion in the anterior horns of the cervical segments). There is symptom of the “hanging head”.
- b) Bulbar syndrome with paresis of the muscles of pharynx, larynx, breathing disorders.

2. Poliomyelitic

This form is associated with flaccid paralysis and paresis of extremities.

3. Meningeal

This form manifest as acute serous meningitis. There is pleocytosis in CSF to 300 cells (lymphocytes).

4. Encephalitic

There are bulbar, pontine, midbrain, subcortical, capsular and hemispheres syndromes.

5. Meningoencephalitic

There are symptoms of two previous described forms.

6. Not well expressed

General infectious syndrome dominates over the others.

7. Polyradiculoneuritic

This form is characterized by the symptoms of polyradiculoneuritis (flaccid paralysis, polineuritic type of sensory disorders, pain)

Chronic stage

This stage is characterized by such symptoms as:

1. Kozhevnikov epilepsy – it is associated with myoclonus in certain group of muscles and periodic epileptic attacks.
2. Poliomyelitic form – means progress of the acute stage.
3. ALS syndrome.
4. Syringomyelitic syndrome.
5. Syndrome of multiply encephalomyelitis.

The course of the disease In the acute stage the symptoms are usually developed during the first 10 days. Then it slightly de-

creases. Renewal of functions lasts from 2 to 3 years. There is high mortality at meningoencephalitic and poliomyelitic forms associated with bulbar syndrome.

Diagnosis

1. Epidemic anamnesis (endemic zone, the bite of Acaridan).
2. The presence of virus in blood and CSF with its identification.
3. Serologic reactions.

After the disease the patients usually have strong immunity.

Differential diagnosis

1. Serous meningitis.
2. Acute poliomyelitis.
3. Typhus.

The treatment of the acute stage

There are specific and nonspecific methods of treatment.

Specific treatment:

1. 10 % Gamma globulin (made of serum of those people who live in endemic regions or were vaccinated against the etiologic agent of this disease).
2. 10 % Gamma globulin (made of serum of hyperimmunized hoarses). It is used in dose 3 – 6 ml during 2 –3 days.
3. The serum of those people who had this disease in dose 8 – 10 ml endolumbal or 50 ml i/m once a day during 2 – 3 days.
4. Blood transfusion from those people who had this disease.

Non – specific treatment:

1. RNA – dose 25 mg 6 times per day during 5 – 6 days.
2. Antibiotics in order to prevent pneumonia.
3. Hormones (Dexon 16 – 24 mg per day).
4. Diuretics (Diacarb, Sorbit, Manitol).
5. Vitamins B1, B6, C.
6. Desintoxication (Hemodes, Rheopoligluclin, Rheogluman, 5 % Glucosa).
7. Symptomatic treatment (glycosides, artificial ventilation, tracheostomy).

Renewal period:

1. Vitamins B1, B6, B12;
2. Proserinum, Halantaminum;
3. Biostimulators (Aloe);
4. ATP, Cocarboxilasa;
5. Anabolics;
6. Nootrops;
7. Physiotherapy;
8. Sanatorium treatment.

Prevention:

1. Active immunization with cultural vaccine three times with revaccination after 4–12 months.
2. Passive immunization with hoarse hyperimmune serum in dose 10 – 20 ml i/m.

Primary poliseasonal encephalitis

To this group belong those encephalitis which are caused by Coxacci viruses (A9 B3 B6) and ECHO (2, 11, 24), that means enteroviral, herpetic, flu – associated.

Enteroviral encephalitis

Pathology

There is well – expressed perivascular edema (especially in cortex, basal nuclei), focal destructive changes of white substance and inflammatory changes of vascular walls.

Clinical features

Encephalitic signs are developed on the second – fifth days against expressed general – infectious and general – cerebral symptoms. There are such syndromes as:

- Hemispheres
- Brainstem
- Cerebellar

1. **Hemispheres syndrome** manifests as seizures against increased temperature, paresis, paralysis. Sometimes it is associated with aphatic disorders and hyperkinesis.
2. **Brainstem syndrome.** It is characterized by CN's disorders, such as diplopia, ptosis, cross eye (III CN), dizziness, nausea, vomiting, decreasing of hearing (VIII CN), dysarthria, dysphonia, dys-

phagia (caudal group of CN's).

- 3. Cerebellar syndrome** manifests as static and dynamic ataxia against general infectious and general cerebral syndromes.

Diagnosis The diagnosis of this type of encephalitis is put according to the presence of focal symptoms on the second – fifth day after upper respiratory ways disorders or sub febrile temperature and general weakness. There is increased content of lymphocytes and increased pressure in CSF.

The course of the disease Usually the course of the disease is favourable with full regress of neurologic symptoms. Sometimes slight disorders of CN's, paresis or aphasic disorders remain after the disease.

Treatment:

1. Antibiotics;
2. DNA – daza – 25 mg every 4 hours during 10 – 12 days;
3. Hormones (Dexamethazonum 8 – 16 mg a day);
4. Desensibilizative (Claritinum, Suprastinum, Tavegil, Loratidinum);
5. Hemodes, Rheopoliglucinum, Rheoglumanum in order to desintoxicate;
6. Symptomatic treatment.

Herpetic encephalitis

Herpes simplex virus can damage different organs and systems of organs (skin, mucous membrane, liver, nervous system).

It is widely spread. Almost 70 – 90 % of all grownups have antibodies to it. It can persist in patient's body for a long time. At unfavorable conditions in those persons who have low immune response it can be activated.

It penetrates into the central nervous system with blood flow.

Pathology There is brain edema, focuses of necrosis and hemor-rhages with neurons death. The lesion of mediobasal parts of temporal lobes and lower orbital parts of frontal lobes is very typical for the herpetic encephalitis. This can be explained by the fact that the virus penetrates through the bulbus ophthalmicus or trigeminal

ganglion.

Clinical features The clinical picture is the same as at any other encephalitis (increased temperature, general cerebral symptoms).

Peculiarities:

1. General epileptic attacks;
2. Focal symptoms of temporal and frontal lobe:
 - Smell and taste hallucinations;
 - Anosmia;
 - Memory disorders;
 - Psychiatric disorders;
 - Hemiparesis;
 - Hyperkinesia.

The course of the disease The disease has severe course and rapid progress. Mortality is too high (50 – 70 % in case of absence of treatment). As a result of acute brain edema coma is developed with cutting – in syndrome which usually leads to patient's death.

Diagnosis

1. **CSF.** There is increased pressure, lymphocytic pleocytosis (from 100 to 500 cells in mmi), slightly increased protein content, sometimes erythrocytes in CSF.
2. **EEG.** There are periodic high amplitude quick waves in temporal lobes.
3. **CT and MRI scan.** There are hypodensive zones as a result of inflammatory changes as well as edema, small hemorrhages in frontal and temporal lobes.
4. Positive **serologic reactions.**
5. **Immunofluorescentive method.**

Treatment

Herpetic encephalitis has effective specific treatment:

1. Acyclovir (Zovirax, Virolex) is used in dose 200 mg 5 times per day orally or i/v in dose 10 – 30 mg per kg 3 times per day. We usually use it during 10 – 14 days. Herpevir 200 mg 5 times per day during 7-10 days. This medication has small toxicity and is excreted by kidneys. Its side effects are nausea, headache, dermatitis, phlebitis. It is usually prescribed when we do not know exactly etiologic

agent.

2. DNA – daza 25 mg i/m every 4 hours.
3. Interferonum 500 000 U i/m once a day.
4. Mannit, Manitol, Lasix i/v by drops.
5. Rheopoliglucinum, Rheoglumanum, Hemodes i/v by drops.
6. Immunomodulators (Immunoglobulin, T – activin, Timalinum).
7. Symptomatic treatment.

Prognosis It depends on patients age, state of consciousness disorders. Mortality now is about 25 %. Half of those who survived have residual symptoms such as Korsakov syndrome, dementia, epileptic attacks, and speech disorders.

Flu – associated encephalitis

It is caused by A1, A2, A3, B viruses and is usually the result of flu. The virus has toxic influence on the brain vessels receptors.

Pathogenesis The virus has neurotoxicity and causes circulation disturbances in the brain. It also stimulates immunologic reactions.

Pathology There are thrombovasculitis, small hemorrhages and perivascular changes in the brain tissue.

Clinical features

Common flu is characterized by general infectious and general cerebral symptoms – headache, painful eye movements, muscle pain, sleepiness. Usually in case of favourable course all these symptoms tend to disappear. Sometimes it is complicated with flu associated encephalitis, especially in those persons who have chronic diseases, brain diseases, chronic intoxication or brain trauma.

The disease starts rapidly with increased temperature, general cerebral, meningeal and focal symptoms after the flu.

The main syndromes are:

1. Cortical syndrome:

1. First of all general weakness, depression, irritation, emotional instability appear;
2. Psychotic disorders manifest as hallucinations, paranoids, delirium;
3. Seizures are usually tonic – clonic with involuntary urination

and tongue biting.

2. **Brainstem syndrome** manifests as vestibular syndrome, lesion of III, IV, VI CN's with eye movements disorders, ptosis, diplopia, cross – eye. Sometimes VII and XII CN's and pyramidal pathways are involved (in this case we can observe paresis of mimic muscles, tongue deviation, asymmetric stretch reflexes, pathologic reflexes of Babinski and others. In most severe cases spastic paralysis and paresis are developed).
3. **Hypothalamic syndrome** means changes of pulse, heart rate, skin colour
4. **Cerebellar syndrome** means ataxia, coordination disorders.
5. **Subcortical syndrome** usually manifests as hyperkinesia against low muscle tone.

In case of toxic form of flu associated encephalitis **hemorrhagic flu associated meningoencephalitis** can be developed. It is characterized by brain vessels tonus disturbances, small hemorrhages. Usually this disease is much more common among the young persons at the age of 16–40. The disease starts rapidly with general cerebral symptoms such as headache, sleepiness, loss of consciousness, vomiting, psychomotor agitation and well expressed meningeal syndrome.

This disease should be **differentiated** with subarachnoid hemorrhage. Besides general cerebral syndrome there are focal one or two side cerebral symptoms (speech disorders, lesions of CN's, bulbar syndrome, frontal release signs). There are changes on eye ground, lymphocytic pleocytosis, increased protein content and blood traces in CSF.

Hemorrhagic encephalitis has severe **course** and usually leads to patients' death. If the person survives some of the neurologic symptoms remain. When headache, dizziness and nausea remain for a long time and are increased at slight physical exercises then it is considered to be the sign of commissural arachnoiditis.

At flu associated encephalitis there is leucocytosis or leucopenia in blood and lymphocytic pleocytosis in CSF.

The course of the disease in most cases is favourable. The disease lasts for a month and is usually cured.

Treatment:

1. Desintoxication;

2. Diuretics;
3. Gamma globulinum 6 mg per day i/m, Rimantadinum 0.1 3 times per day;
4. Hormones (Dexamethazonum, Prednisolonum);
5. Antibiotics in order to prevent pneumonia.

Encephalitis at general infections

Etiology and pathogenesis

This type of encephalitis is very common at early childhood. Allergic process takes a special place in its **pathogenesis**. This encephalitis is usually associated with various infections in children, such as measles, scarlet fever, rubella, small pox.

Measles encephalitis and encephalomyelitis

Measles encephalitis is the most typical form of parainfectious encephalomyelitis. The frequency of this disease is 1:1000.

Pathology There is fibrous edema of vessels' walls and perivascular focuses of demyelination.

Clinical features

The disease develops rapidly on the third – fifth day after the rash when the temperature is usually normal. Then suddenly it is going worse, the temperature increases to 39 – 40° C, the child becomes sleepy, has consciousness disorders, psychomotor agitation, seizures, sometimes meningeal syndrome is associated. In some cases focal signs can be observed such as paresis, paralysis, hyperkinesia, coordination disorders, lesion of II, III, VII CN's.

When the spinal cord is involved in the process there is also paralysis of lower extremities, sensory disorders according to the conductive type, disturbance of function of pelvic organs.

There is mild lymphocytic pleocytosis to 200 cells per 1 mm³, increased content of protein in the CSF.

The course of the disease is severe. Mortality is 10 – 25 %. There are residuals such as seizures, hyperkinesia, and mental retardation.

Small pox encephalitis

The prevalence of this disease is 1 per 10 000. It has infectious – allergic character.

Pathology There is inflammation of brain tissue and focuses of demyelination. In severe cases there are purulent – hemorrhagic focuses.

Clinical features

The disease is usually developed on the third – seventh day after the rash. In 50 % of children there is benign cerebellar ataxia. In the rest ones there are pyramidal and extrapyramidal symptoms against general infectious, general cerebral and meningeal syndromes. There is lymphocytic pleocytosis to 100 – 200 cells, increased protein content in the CSF.

The course of the disease is favourable. Mortality is 10 %.

Residual symptoms – paresis, hyperkinesis, seizures.

Treatment:

1. Desensibilizative medications;
2. Hormones;
3. Against edema medications;
4. Dezintoxication;
5. Nootrops;
6. Symptomatic treatment;
7. Vitamins B1, B6, C.

Postvaccinal encephalitis

This group of encephalitis are developed after the vaccination.

Pathogenesis This group of encephalitis belong to the allergic leucoencephalitis

Pathology There are small hemorrhages, brain edema, focuses of demyelination in the brain and spinal cord.

Clinical features

Usually this disease develops in vaccinated children, especially at late vaccination and revaccination. The disease develops on the 7th – 12th day after the vaccination and starts with increased temperature to 39 – 40^o C, severe headache, vomiting, consciousness disorders, seizures. Sometimes meningeal signs

appear. In some cases paresis, paralysis and coordination disorders are observed. There is mild lymphocytic pleocytosis in the CSF.

The course of the disease Usually it is favourable. Although in some cases paresis and paralysis remain for a long time. The main peculiarity of the course of the disease after antirabies vaccination is the clinical picture of acute encephalomyelopoliradiculoneuritis with well expressed bulbar syndrome, disturbance of breathing and heart – vascular activity, which can cause death of the patient.

Treatment:

1. Desensibilizative medications;
2. Hormones;
3. Diuretics;
4. Antiseizures medications;
5. Antipyretics.

Acute toxic encephalopathy

This encephalopathy develops after the viral infection and is usually associated with lipid degeneration of some internal organs, especially liver. It is much more common in children than in adults.

Etiology It is unknown till nowadays. It is considered that the disease is the result of viral effects and intoxication. The risk of the disease increases in case of Aspirin usage.

Pathology Brain edema without inflammation, liver enlargement.

Clinical features

After the viral infection when there is normal temperature, there are no respiratory or gastro – intestinal disorders suddenly it is going worse, the temperature increases again, sleepiness and consciousness disorders appear. This is known as **first stage of Rayer syndrome**.

The **second stage** – delirium and epileptic – attacks.

The **third stage** – superficial coma develops without any focal signs.

The **4th and 5th stage** – deep coma develops, sometimes cut-

ting in syndrome can occur and it can cause the death of the patient.

The disease can be stopped at any stage or can progress. The typical features are liver enlargement with its' function disorders and pancreas disturbances.

There are no changes in CSF. There is leucocytosis in blood.

Treatment:

1. Symptomatic;
2. Dehydration (Manitol);
3. ALV (artificial lung ventilation);
4. Correction of liver functions (Glucosa, Vicasolum, hepatoprotectors).

Neurosyphilis

Definition

Syphilis is a chronic system infection, which is developed after sexual relations in case of local infections.

Etiology

The etiologic cause of this disease is *Treponema pallidum*. In 1913 it was identified in the brain by Noguchi and Moore.

Epidemiology

According to the German scientists – Rittr and Prange (1987) the incidence of neurosyphilis is 15 persons per 100 000. Neurosyphilis takes 0.1% of all organic neurological diseases. Significant clinical signs of nervous system involvement are reported in 1– 2% of all patients with neurosyphilis. This disease is much more often observed in men (70%) than in women (30%).

Following the introduction of penicillin the incidence of neurosyphilis declined. One of the very important peculiarities is that among those infected with human immunodeficiency virus (HIV), about 15% have serologic evidence of syphilis and about 1% have neurosyphilis.

Pathogenesis

In early neurosyphilis, lymphocytes and other mononuclear cells infiltrate the meninges. The inflammatory reaction also involves the cranial nerves and provokes axonal degeneration. When the inflammation affects small meningeal vessels, occlusion due to endothelial proliferation may lead to ischemic necrosis of brain and spinal cord. This process may cause demyelination, myelomalacia at the periphery of the cord, or transverse myelitis.

The pathology of dementia paralytica develops slowly. After an inflammatory meningeal reaction, lymphocytes and plasma cells infiltrate small cortical vessels and sometimes extend into the cortex itself. The cortical inflammatory response provokes loss of cortical neurons and glial proliferation. Spirochetes can be demonstrated in the cortex in dementia paralytica, but only rarely in other forms of neurosyphilis.

In tabes dorsalis, the mononuclear inflammation of meninges

and blood vessels is followed by insidious degeneration of the posterior roots and posterior fiber columns of the spinal cord, as well as the cranial nerves occasionally.

Classification

Syphilis is divided into:

Primary – begins with chancre and lasts until it is cured or until the rash appears.

Secondary – it develops after the appearance of specific pimple rash on skin, condylomas on mucous membrane.

Latent – early and late. The border between them is 2 years from the moment of infection. The patients do not have clinical signs of the disease but all the serologic reactions are positive. These patients are divided into such groups:

- 1/3 of them have no clinical signs of syphilis. Non – treponema blood – tests are negative, but treponema blood – tests are positive.
- 1/3 of them have no clinical signs of syphilis. Non – treponema and treponema blood – tests are positive.
- 1/3 have tertiary syphilis:
 1. 50 % – gummy syphilis;
 2. 25 % – cardiovascular syphilis;
 3. 25 % – neurosyphilis.

Tertiary (late) syphilis.

Syphilis is a contagious disease, especially in case of specific changes on skin and mucous membrane. Latent syphilis in case of absence of skin changes and tertiary syphilis aren't contagious. After the first injections of penicillin the patients become not contagious.

Neurosyphilis is divided into 2 forms:

- Early (mesodermal);
- Late (parechymatous).

Clinical forms:

Early mesodermal syphilis:

1. Asymptomatic neurosyphilis;
2. Acute syphilitic meningitis;
3. Chronic basal syphilitic meningitis;

4. Early meningovascular syphilis;
5. Syphilitic meningomyelitis;
6. Syphilitic mono – and polyneuropathy.

Late mesodermal syphilis:

1. Late syphilitic meningitis;
2. Late vascular syphilis;
3. Late meningovascular syphilis;
4. Late pupil monosyndrome;
5. Gummy brain and spinal cord;
6. Cerebrospinal syphilis.

Late ectodermal (parenchymatous) neurosyphilis:

1. Tabes dorsalis;
2. Amyotrophic spinal syphilis;
3. Spastic spinal paralysis;
4. Progressive paralysis.

Clinical features. Diagnosis

Early mesodermal forms of neurosyphilis

Nowadays the most common among all the forms of neurosyphilis is **asymptomatic syphilitic meningitis**. Usually it is associated with:

1. General cerebral signs:
 - Headache;
 - Head noise;
 - Dizziness;
 - Vomiting;
 - Painful eyes movements;
 - General hyperesthesia.
 2. Intoxication symptoms (sometimes):
 - Weakness;
 - Nervousness;
 - Insomnia;
 - Depression.
 3. Focal signs:
 - Radicular pain;
 - Transient insufficiency of some cranial nerves (II, III, VI, VII, VIII).
- During the 12 – 18 months from the moment of infection there are inflammatory changes in CSF:
- Lymphocyte pleocytosis;

- Insignificant increasing of protein content;
- Positive Wasserman, Kann reactions.

The frequency of asymptomatic neurosyphilis is 30% from all the forms of neurosyphilis.

Acute general syphilitic meningitis

It is very common among the young people in case of inadequate treatment. There are 3 clinical forms:

1. Acute syphilitic hydrocephalus;
2. Acute basal syphilitic meningitis;
3. Acute syphilitic meningitis with focal neurological signs.

On the background of fever there are:

- Severe headache;
- Dizziness;
- Nausea, vomiting;
- Well –expressed meningeal symptoms;
- Pathological reflexes;
- Anizoreflexia, paresis;
- Seizures;
- CN's lesion (II, III-VI);
- Consciousness disorders;
- Sometimes pelvic disorders and trophic disturbances (such as bed – sores).

CSF:

- Increased pressure up to 300 – 500 mm;
- Lymphocyte cytolysis;
- Protein content – 0.6 – 1.2 g/l;
- RW ++++.

The course of the disease is favourable. In case of adequate treatment the meningeal syndrome is liquidated in 2 – 3 days, CSF is normalized in 7 – 10 days. Very often this form can be transformed into a chronic one.

Chronic basal syphilitic meningitis

It develops subacutely, sometimes during several weeks. The **typical features** are:

1. Early CN's lesion:

- Oculomotor nerve (cross – eye, ptosis, anisokoria);

- Facial nerve (Bell's palsy);
- Vestibular-Cochlear nerve;
- Optic nerve (choked discs on eye ground, atrophy, neuritis);
- Trigeminal nerve (pain, sensory disorders);
- Isolated paralysis of certain external or internal eye muscles.

2. General cerebral and meningeal signs

Rare forms of neurosyphilis:

- Early meningovascular syphilis;
- Syphilitic meningomyelitis;
- Syphilitic neuropathy and polyneuropathy.

Early meningovascular syphilis:

1. General cerebral signs;
2. Meningeal signs;
3. Focal symptoms (hemiparesis, aphasia, general seizures).

Syphilitic endarteritis The result of it can be brain and spinal cord infarction. It is characterized by gradual development of focal signs. It is much more common in men than in women. The symptoms of the disease manifest 5 – 30 years after the infection. The progress of the disease is very progressive.

Syphilitic meningomyelitis It is characterized by:

1. Sudden onset;
2. Acute course of the disease;
3. Rapid development of lower paralysis;
4. Well expressed trophic disturbances;
5. Conductive hyposthesia;
6. Pelvic disorders.

Spinal meningovascular syphilis It is an acute infarction in the region of a. spinalis anterior.

Clinical features:

1. Lower flaccid paralysis;
2. Conductive hyposthesia;
3. Pelvis disorders;
4. There are also specific changes in CSF.

Additional methods of neurosyphilis diagnosis:

1. Positive Wasserman test in blood and CSF;
2. Positive serologic tests;
3. Positive Lange reaction in CSF;
4. Lymphocyte pleocytosis and increased protein content in CSF at meningeal forms.

The typical changes in CSF:

1. Increased protein content from 0.5 to 1.5 g/l;
2. Lymphocyte cytolysis (50 – 100 cell in 1 mcl);
3. Lange reaction of paralytic or meningeal type;
4. Positive reaction of Wasserman.

Late mesoderm syphilis

Clinical features appear 7 – 8 years after infection. There are such forms as:

- Late syphilitic meningitis;;
- Late vascular syphilis;
- Late meningovascular syphilis;
- Late pupil monosyndrome.

To them belong also:

- Gummy brain and spinal cord;
- Cerebrospinal syphilis.

Late syphilitic meningitis The typical features are:

- Without fever onset;
- Gradual development;
- Recrudescence chronic course of the disease.

Neurological symptoms:

1. General cerebral symptoms:
 - Attack – like severe headache;
 - Vomiting.
2. Meningeal symptoms:
 - Neck stiffness;
 - Kernig – sign;
3. CN's lesion:
 - Oculomotor nerve;
 - Argil – Robertson syndrome – myosis, anizokoria, pupils defor-

- mation, the absence of pupils reaction at chemical tests;
- Optic nerve – decreased vision, hemianopsy, chocked discs on eye ground, atrophy;
 - V, VI, VIII CN's.

Vascular syphilis

Inflammatory changes are observed in the wall of the vessels. The meninges remain intact. The disease is very similar to brain infarction. As a result of numerous vessels lesion there are repeated brain infarctions associated with new focal signs. Hemorrhages are very rare.

Late meningovascular syphilis Typical focal neurological signs:

- Hemiparesis, hemiplegia;
- Sensory disturbances;
- Aphasia;
- Memory disorders;
- Alternating syndromes.

The peculiarity of this form is combination of general cerebral and meningeal syndromes.

Late pupil monosyndrome:

- Anizokoria;
- Bilateral pupils' deformation;
- Direct Argil – Robertson syndrome.

Combination of pupils' monosyndrome and absence of Achille and knee reflexes, sensory disorders is known as **pretabes or pupile–radicular syndrome**.

Gummy brain It is a syphilitic without-vessels granuloma. It creates separated focus of lesion. It is a localized form of meningitis. Clinical picture is very similar to brain tumor. The most typical place of its localization is fossa interpeduncularis.

Clinical features:

- Increased brain pressure (headache, vomiting, chocked discs on eye ground, epileptic attacks);
- Focal signs:
 - Paralysis;
 - Sensory disorders;

- Aphasia;
- Epileptic attacks;
- Agnosia;
- Choked discs on eye ground;
- Protein – cell dissociation in CSF.

Gummy brain is characterized by:

- Pupils deformation;
- Slight pupils reactions;
- Low Achille and knee reflexes.

Cerebrospinal syphilis It is very common disease. Usually cerebral symptoms dominate over the spinal ones.

CSF examination reveals:

- Moderate increasing of protein content (0.5 – 1.0g/l);
- Insignificant mononuclear cytolysis (20 – 70 cells in mcl);

There are also positive serologic reactions, such as reaction of immobilization of *Treponema pallidum*, reaction of immunofluorescentation in blood and CSF.

Late ectodermal forms of neurosyphilis

Tabes dorsalis

Nowadays it is a rare form of neurosyphilis. Usually it is observed 15 – 25 years after infection. It is the result of posterior columns and posterior roots of spinal cord degeneration. Some of cranial nerves, paravertebral and spinal ganglions can be injured.

There are 3 **clinical stages**:

- Neuralgic;
- Ataxic;
- Paralytic.

Neuralgic stage:

- Parasthesia (numbness, tingling);
- Pain (it has lancinating character. It is very severe and it appears very sudden. Its duration is from 1 – 2 seconds, sometimes longer. Usually it is the first sign of the disease);
- One of the typical features is visceral crisis – the attack of neurological pain in the internal organs with the loss of function of this organ. The most common are stomach crisis, also intestinal cri-

sis, laryngeal crisis, heart crisis, liver, and kidney and urine bladder crisis;

- One of the typical and early sign of tabes dorsalis is Argil – Robertson syndrome –
 1. Decreased pupils reaction on light at preserved convergence and accomodation;
 2. Anizokoria;
 3. Deformation of pupils;
 4. Myosis.

Knee and Achille reflexes are decreasing. There are lesions of Optic N (primary gray atrophy), VIII CN (dizziness and decreasing of hearing), Oculomotor N (ptosis, outside cross – eye), Abducens N (inside cross eye), Trigeminal N.

Ataxic stage

The dominating symptoms are disorders of coordination in extremities and unstable standing and walking. These symptoms can be explained by disturbances of deep sensation. Usually the symptoms of lesion dominate in lower extremities. The patients feel the flour badly. It is difficult for him to walk in darkness or with his eyes closed. The muscle tonus is decreased.

Paralytic stage

It is characterized by severe disorders of motor function. There are no true paresis or paralysis. But because of the static and dynamic ataxia the patients are not able to stand and walk. The most common symptoms are:

- Trophic and pelvic disorders;
- Retention of urine and feces, sometimes incontinence of urine and feces;
- Trophic skin disorders.

Psychological disturbances:

- Changes of personality;
- Memory disturbances;
- Consciousness disorders.

Sometimes tabes dorsalis is associated with progressive paralysis. The last manifests as memory disturbances, insomnia, and

euphoria. There are also speech, writing and reading disorders. This disease is known as **taboparalysis**.

Diagnosis:

- Pupils deformation;
- Lancinating pain;
- Retention of urine;
- Parasthesia;
- Back hyperesthesia;
- Tabes crisis and ataxic disorders.

CSF:

- Insignificant increasing of protein content;
- Lymphocyte pleocytosis;
- Positive Wasserman test.

Progressive paralysis

It is developed 10 – 20 years after syphilis infection. The main clinical features are – memory and personality disturbances, loss of acquired skills, positive Argil – Robertson test, paresis, sensory disturbances, epileptic attacks. There are also increased protein content (0.45 – 0.6 g/l), positive Wasserman test and paralytic Lange curve.

Treatment

The treatment of neurosyphilis begins with the treatment of early syphilis. Usually penicillin is used as standard treatment. It can be introduced in 2 ways:

1. I/v 12 000 000 – 24 000 000 U per day 2 – 4 mlns of U every 4 hours. It is used during 10 – 14 days. Then 2 400 000 U of Retarpen is used once a week during 3 weeks.
2. I/m 2 400 000 U per day in association with Probenecid (500 mg 4 times per day) during 10 – 14 days. Then 2 400 000 U of Benzatin – Benzylpenicillin is used once a week in a course of next 3 weeks.

The effectiveness of treatment is determined according to the blood tests and CSF examination. That's why Lumbar puncture is made just after the penicillin treatment and then every 3 months. The result is normal CSF. That means that such patients are observed

during several years after the disease.

The patient is considered to be cured only in case of completely normal CSF 2 years after the infection.

Prevention of syphilis:

- Prevention of syphilis infection;
- Adequate treatment of early syphilis;
- Adequate observation of the patient with control blood tests and CSF – examination.

Neurological signs of lumbar and cervical osteochondrosis.

Among all the spinal diseases associated with neurological signs the most common are degenerative – dystrophic processes such as osteochondrosis and spondyloarthrosis.

Osteochondrosis (OC) is one of the most common diseases. About 80% of all people have attacks of low back or cervical pain during their life – time. It takes 68% of all temporary working disability in neurology.

Spinal osteochondrosis is a degenerative lesion of cartilage – between vertebral disc associated with reactive changes in neighbouring vertebrae, between vertebral joints and connecting apparatus.

Etiology of osteochondrosis

According to the main modern theory – osteochondrosis is a polyfactorial disease that develops in case of two conditions:

1. Decompensation of trophic systems. It is associated with other diseases (those of gastro – intestinal tract, liver, pancreas, and enzyme – associated diseases). All of these diseases lead to the disorders of homeostasis
2. Local excessive load on spinal segments. They can be explained by inborn peculiarities of spinal cord (sacralization, lumbalization, narrow spinal canal).

Pathogenesis of osteochondrosis

Usually osteochondrosis is developed at the age of 30 – 50. The main pathological process begins in between vertebral discs. Disc consists of pulp nuclei, fibrous ring and hyaline plate. Disc is a natural amortisseur.

The first stage. Because of certain factors (such as dysmetabolism, arthritis and so on) degenerative changes of pulp nuclei develop. That means destruction of chondrocytes with releasing of hyaluronidase, chondrochapsines, and papain. That causes destruction and depolymerization of mucopolysaccharide complex. As a result pulp nuclei loses its hydrophilous properties, becomes small and mobile.

The second stage. There are degenerative changes in fibrous ring. During the movements the main load is on the internal structures of fibrous ring. They broke and cause disc protrusion. The last can be developed later in disc hernia.

The third stage. There are changes of hyaline plates that are called subchondral sclerosis. In this stage reactive process is developed. Disc hernia is vasculated, fibroused and later osteophytes are growing along the bodies of vertebrae. In case of spinal segments increased mobility the reactive changes in neighbouring bodies of vertebra and joints are developed. It is known as associated spondyloarthrosis.

X –ray signs of osteochondrosis (fig 75).

1. Low height of between vertebral discs.
2. Sclerosis of final plates.
3. Osteophytes on the edges of vertebrae's bodies. They are located perpendicularly to the spinal axis.
4. Local scoliosis.
5. Spondylolisthesis – dislocation of neighbouring vertebrae's bodies.
6. Local kyphosis (instead of lordosis)

At distorted spondylolysis (as a result of degenerative – dystrophic changes in long spinal ligaments) there are osteophytes, which are located parallelly to the spinal axis along the spinal cord and there is no changes of vertebral discs height.

Between vertebral disc's hernia can be located in different places according to the spinal canal. These positions are:

- Medial (clinical features of horse tail lesion);
- Paramedial (a little bit out side the medial position – several radices are compressed);
- Posterior – medial (much more out side the medial position – 1 or 2 radices are compressed);
- Foraminal (near the foramina vertebral – 1 radix is compressed).



Fig. 75. Osteochondrosis of intervertebral disc L4-L5. Low height between vertebra, sclerosis of final plates, osteophytes.

Pathogenesis of neurological signs (fig 76).

1. Irritation of vessels and nerves with vascular spasm.
2. Compression of vascular – nervous complex.
3. Edema (perivascular edema and radicular edema).
4. Reactive process of neighbouring structures (straining of muscles).
5. Autoimmune reactions (a part of disc becomes antigen).

According to the nervous structures that are involved in case of disc's hernia there are compressive and reflex syndromes.

Compressive syndrome occurs at compression and deformation of radix, vessels and spinal cord.

Reflex syndrome occurs at irritation of different receptors (such as Lushka nerve). That means reflex muscular – tonic disturbances, extension of muscles, and pain at palpation of muscles. That is usually the reason of local pain and pain in distance. Such zones of pain are called trigger zones. Pathogenetical process in these zones is known as neuroosteofibrosis, the painful nodes in muscles are called nodes of Kornelius, muscular hypertonus – hypertonus of Muller.

Neurological signs of osteochondrosis at lumbar – sacral level

Classification:

I. Reflex syndromes:

- Lumbago (backache);
- Lumbalgia;
- Lumbar ischialgia (muscular – tonic, neuro – dystrophic, autonomic vascular).

II. Compressive radicular syndromes.

III. Compressive vascular radicular – spinal syndromes (radicular ischemia):

1. Acute:
 - A. Transient;
 - B. Strokes.
2. Chronic ischemic myelopathy.

All the neurological signs of osteochondrosis have common symptoms – the symptoms of vertebrogenous syndrome.

Vertebrogenous syndrome points that the pathological process is associated with spinal cord.

- Limitation of movements in lumbar – sacral part of spinal cord

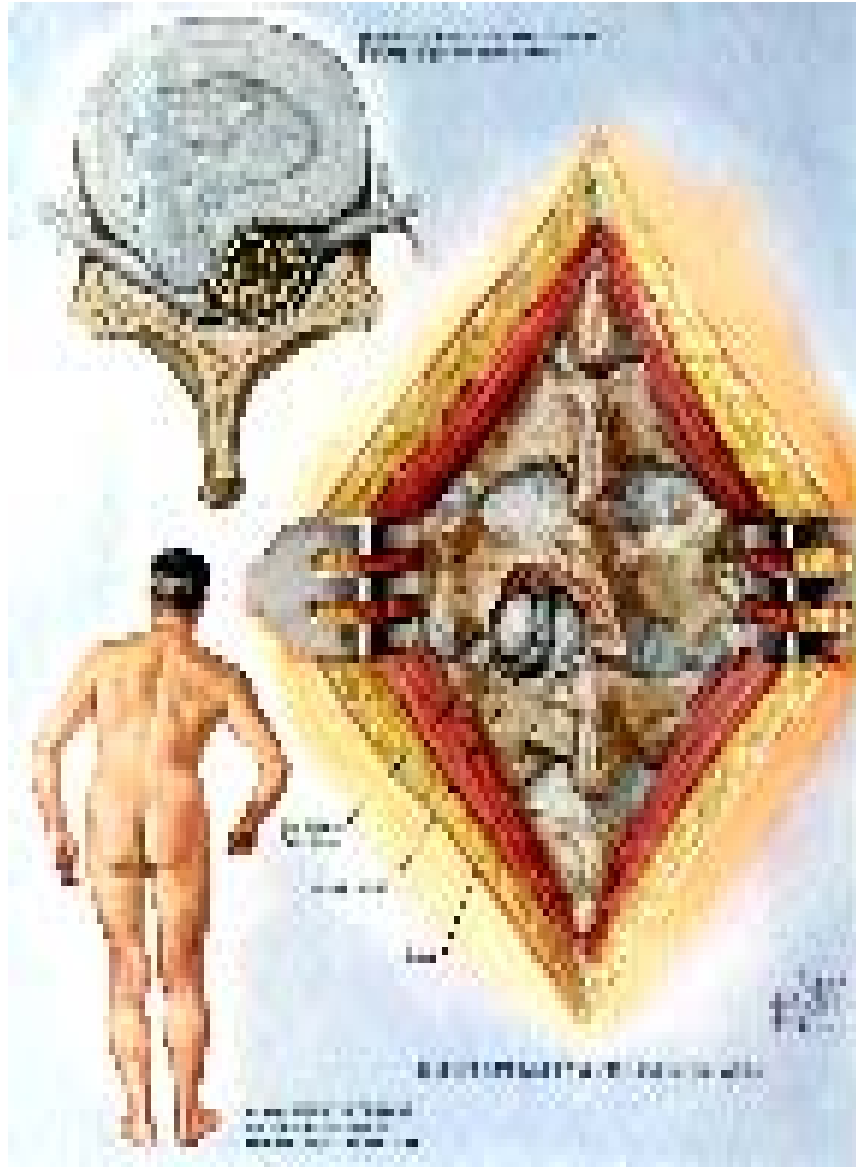


Fig. 76. Herniation of the Lumbar intervertebral discs (Frank H. Netter, M.D.)

(bending forward, backward) and increasing of pain while movements, coughing and laughing.

- Protective straining of long back muscles.
- Extension of lumbar lordosis, cyphosis in lumbar – sacral division.
- Scoliosis, sometimes with rotation.
- Painful paravertebral points.
- Painful vertebral processes.
- Discharge – postures and symptoms:
 - Knee – elbow position;
 - While standing the patient keeps his leg aside in order to make the load less on his leg;
 - While lying in the bed he bends his leg in all joints.
- The symptoms of spinal cord instability (it is difficult for the patient to stand, to wash himself, but it is much more easier to walk).

I. Reflex syndromes on lumbar – sacral level are divided into:

- muscular – tonic;
- vascular;
- neuro– dystrophic.

Clinically all the reflex syndromes at lumbar – sacral level can be divided into:

1. Lumbago;
2. Lumbalgia;
3. Lumbar ischialgia.

1. Lumbago It is acute, sudden, attack – like low back pain. It usually begins at lifting something heavy, catching cold or sometimes spontaneously. Pain is very severe and the patient stays in the same position for a long time. At first the pain cannot be localized. Later the patient can localize it and refers the pain to the low back region. It lasts for 1 – 3 up to 6 days.

2. Lumbalgia The pain is not so severe. It has subacute or chronic character. It lasts for weeks or even months. The pain can be increased or decreased according to different factors.

3. Lumbar ischialgia The source of pain impulse is receptor. The pain irradiate into hips, leg.

The points of pain:

- Pain along the crista iliaca;
- The point of iliosacral joint;
- The point of m. gluteus minimus (just under the crista iliaca);

- The point of m. gluteus medius (1 sm lower);
- The point under the backside fold;
- Trochanter os iliaca;
- Along the ischiadic nerve (the posterior surface of hip and fossa subpoplitea).

The symptoms of strain: (fig. 32-33)

- Lasegue's symptom – in case of straining and lifting the leg the low back pain appears;
- Neri symptom – there is pain in leg at bending head forward;
- Matskevych symptom – there is pain in the anterior surface of the leg at knee bending while lying on abdomen;
- Wasserman symptom – the same clinical picture at lifting the leg;
- Sequar symptom – there is pain on posterior surface of leg at foot flexing;
- Turin symptom – the same clinical picture at toe's flexing;
- Bechterev's symptom – there is pain at knee – flexed leg extension;
- Dejerine's symptom – there is pain in posterior surface of the leg at coughing, sneezing.

Types of lumbar ischalgia:

1. Muscle – tonic;
2. Neurodystrophic;
3. Autonomic – vascular.

1. Muscle – tonic Clinical features are connected with secondary lesion of nerves according to the compressive – ischemic type, the type of tunnel syndrome as a result of muscles spasm and straining.

A. Piriformis syndrome M. Piriformis is located under m. gluteus maximus. It is attached to the internal edge of Trochanter Major and anterior surface of sacroiliac joint.

Under the muscle sacroiliac ligament is situated. Between the muscle and the ligament nervus ischiadicus and a. ischiadica are located. These structures can be irritated or compressed at long – lasted straining of m. Piriformis.

Clinical features:

- Painful palpation of Trochanter major;
- Painful m.Piriformis;

- Symptom of Soobrase (painful cross-legged position);
- Symptom of Bone – Bobrovnikova (painful abduction of leg);
- Popelyansky intermittent claudication (while walking the patient is forced to sit down because of the pain. That is the result of spasm of the vessels.

Except the Popelyansky intermittent claudication there are also:

- Myelogenic intermittent claudication ;
- Caudal intermittent claudication;
- Intermittent claudication at obliterated endarteriitis:
 - Insignificant sphincter disorders (pause before the urination) as a result of n. pudendus irritation;
 - Insignificant signs of n. ischiadicus lesion (muscles hypotrophy, low Achille reflex, hyposthesia, pain).

2. Neurodystrophic form of lumbar ischialgia is a sign of neuromyofibrosis. There are:

A. Sacroiliac periartrosis – pain and limitation of movements in hip joint. The patients cannot run and so on.

B. Knee joint periartrosis– sudden pain in knee joint.

C. Popliteal syndrome It is the result of neuroosteofibrosis in popliteal fossa in the place of m. ischiocrural attachment.

Clinical features:

- Pain in fossa poplitea while patient's standing, palpation;
- Cramps – sudden painful tonic straining of m. Triceps surae; Pains in m. soleus.

D. Coccygodynia Pain is in the coccyx's region. The last is connected with sacrum via discs. Degenerative changes in disc; straining and painful pelvic muscles cause coccygodynia. Usually it is observed at pregnancy, after childbirth, at long sitting.

Clinical features:

1. Long lasting aching pains;
2. Paresthesia in coccyx region, which is increased at sitting, defecation and decreased at standing;
3. That often leads to patients' depression.

E. Neurodystrophic changes of Achille tendon (it is very rare)

The **peculiarity** of neurodystrophic changes is:

- One side lesion on the side of lumbar ischialgia;
- The lesion of large joints;

- Connection of exacerbation with low back pain.

3. Autonomic – vascular form of lumbar ischialgia

- Vasospastic;
- Vasoparetic;
- Mixed.

Clinical features: It appears on background of low back pain, freezing and cyanosis, hyperhidrosis, insignificant autonomic – trophic changes of lower extremities (hyperkeratosis, dryness of skin, edema).

II. Compressive radicular syndromes (radiculopathy)

Hernia of intervertebral discs in lumbar region causes compression of L5 – S1 radices, sometimes L3 – L4. In compressed radices there are edema, venous stasis, aseptic inflammation. Clinical features of radiculopathy consist of clinical features of lumbar ischialgia and symptoms of radix's loss of functions.

Clinical features of radiculopathy.

1. Radix L5 (Disc L4 – L5):

- Pain in the external edge of hip, on the anterior –external surface of crus until the internal surface of foot and great toe;;
- Sensory disorders (hypalgesia, analgesia) in the same zones;
- Paresis of great toe extensors and foot extensors;
- Hypotonia and hypotrophy on the anterior surface of crus;
- The patient cannot stand on heels.

2. Radix S1 (Disc L5 – S1):

- Pain in external – posterior surface of hip, crus, foot, the IV – th and Vth toes;
- Sensory disorders (hypalgesia, analgesia) in the same zones;
- Paresis of toes flexors;
- Absent or low Achille reflex.

3. Radix L4 (Disc L3 – L4):

- Pain in anterior – internal surface of hip;
- Sensory disorders (hypalgesia, analgesia) in the same zones prevail over motor ones;
- Weakness of m. Quadriceps femoris;
- Hypotrophy of m. Quadriceps femoris;
- Knee reflex is low or sometimes increased.

4. Radix L2 – L3:

Compression of these radices is very rare. Clinical features include pain and sensory disorders on anterior – medial surface of hip:

- Symptoms of Matskevych, Wasserman;
- Low knee reflex;
- Weakness of m. Quadriceps femoris;
- Cruralgia;
- Symptoms of lesion of horse tail;
- Irradiation of pain into lower part of abdomen, genital organs.

Syndrome of compression of horse tail It is created by radix L2 – S5. It is observed at hernia of discs L4 – L5.

Clinical features:

- Significant pain in legs;
- Sensory and motor disorders in certain zones of innervation (flaccid paralysis, segmental-radicular type of sensory disturbances);
- Pelvic disorders (incontinence of urine and feces).

III. Compressive vascular radicular – spinal syndromes on lumbar level

There are:

1. Acute vascular – radicular syndromes (transient, strokes);
2. Chronic (chronic myelopathy).

Acute These ones are observed at disc's hernia, narrow spinal canal, as a result of spondilolisthesis.

Transient

1. **Myelogenic Popelyansky intermittent claudication** (transient ischemia of conus and epiconus). Clinical features:

- Weakness in legs without pain during long lasting walking;
- Paresthesia;
- Micturition.

2. **Caudal intermittent claudication of Verbista** (transient ischemia of horse tail radices). Clinical features:

- Pain in feet, cruses, anal region;
 - Weakness in feet;
 - Retention of urine;
- Symptoms last for about 5 – 7 minutes.

Strokes

They are developed rapidly after long lasting lumbalgia or lumbar ischialgia. There are such forms as:

A. Paralysis ischias (at radix spinal artery L5 – S1 compression).

Clinical features:

- Foot weakness without sensory disorders;
- Absence of Achille and sole reflex.

B. Syndromes of cone ischemia (S3 – S5):

- Anesthesia of ano-genital zone;
- Pelvic disorders (retention of urine);

C. Syndromes of epicone ischemia (L4 – S2):

- Flaccid feet paralysis with paresthesia;
- Absence of Achille reflexes;
- Sensory disorders in zones L4 – S2.

D. Syndromes of cone and epicone ischemia

- Paresis and paralysis of lower extremities, much more expressed in distal parts;
- Sensory disorders in zones L4 – S5;
- Pelvic disorders.

E. Syndrome of a. Adamkevych

- Central or peripheral paralysis (paresis) of lower extremities;
- Conductive sensory disorders from umbilicus and downwards;
- Pelvic disorders according to the central type;
- Bed – sores.

Chronic compressive vascular syndromes – dyscirculative myelopathy.

It occurs at graduate compression of spinal vessels, hypertrophy of ligamentum flavum. Lumbar compressive spinal syndromes are very rare. Compressive myelopathy occurs at discs hernia. There are such syndromes:

A. Epicone syndrome (It occurs at protrusion of Th10 – L1 discs with lesion of L4 – L5 – S1 – S2 segments).

Clinical features:

- Low back pain with irradiation in posterior surface of leg;
- Feet paresis;
- Hypotonic and hypotrophic crus' muscles;
- Absence of Achille and sole reflexes.

B. Cone syndrome It occurs at protrusion of L1 – L2 discs with lesion of S3 – S5 segments. Clinical features:

- Perineum anesthesia;
- True urine, feces incontinence and sometimes its retention;
- Bed – sores. These signs are dominant.

Neurological signs of osteochondrosis at cervical level

Anatomical peculiarities of cervical part of the spinal cord

A. C1 and C2 vertebrae are joined without between vertebral discs. Rotation is dominating movement in these vertebrae.

B. The body of C3 and the rest cervical vertebrae aren't separated by the disc completely. Disc is only in anterior and posterior part. There are also hook – like processes. Before them artery is located, after them nerve is situated.

C. Transverse processes of cervical vertebrae have transverse holes. These holes are used by vertebral artery to come through.

D. If there are some changes in between vertebral discs, the main pressure is on hook –like processes, the artery and nerve.

Classification of neurological signs of osteochondrosis on cervical level

1. Reflex symptoms:

- Stiff neck;
- Cervicalgia;
- Cervical cranialgia;
- Cervical brachialgia:
 - Muscle – tonic syndrome;
 - Autonomic – vascular syndrome;
 - Neurodystrophic.

2. Compressive radicular syndrome

3. Compressive – spinal syndrome

4. Compressive vascular radicular – spinal syndrome:

- Acute:
 - Transient;
 - Strokes.
- Chronic ischemic myelopathy

5. A. vertebralis syndrome (radiculopathy of C8 radix, disc C7– C8)

1. Reflex syndromes:

Stiff neck It is sudden acute pain in neck that lasts from several days up to 1 – 2 weeks.

Cervicalgia It is severe dull pain in cervical part of the spinal cord. Usually it appears in the morning, while coughing. There are signs of vertebrogenous syndrome in cervical level – limitation of movements in cervical part of the spinal cord, painful paravertebral points and vertebral processes. There are positive symptoms of muscles straining.

Cervical cranialgia. It is the result of:

1. Irritation of a. vertebralis sympathetic plexus – posterior cervical sympathetic syndrome.

2. Irritation of cervical muscles, fibrous tissues receptors.

Clinical features of posterior cervical sympathetic syndrome:

- Cranialgia – occipital pain with irradiation in temporal, parietal parts;
- Vestibulo – cochlear disturbances – dizziness, vomiting;
- Eyes symptoms – eyes pain, tears;
- Autonomic upper quadrant syndrome – asymmetry of blood pressure, temperature, pulse, sensation, cardiac pain and so on.

Cervical brachialgia:

1. Muscle – tonic form;

2. Neurodystrophic form;

3. Autonomic – vascular one.

1. Muscle – tonic form It can manifest as:

Scalenus – syndrome It is connected with straining of m. scalenus. The muscle starts from transversal processes C3 – C4 and it is attached to the first rib. There are subclavian artery, vein and lower truncus of brachial plexus between the muscle and the rib.

Clinical features:

- There are pains above and under clavicle at the muscle straining;
- There are pains at head movements with irradiation in the arm;
- Edema in above clavicle region;
- Positive Adson test – during the arm adduction there is pain over a. subclavia and slow pulsation in a. radialis;

- Weakness of hand;
- Tenar hypotrophy;
- Hypalgesia of the hand ulnar surface;
- Hand edema;
- Paleness of the hand.

M. pectoralis minor syndrome At this muscle straining the distal part of vascular – nervous trunk is pressed. Clinical features:

- Pain in anterior thoracic part and in ulnar surface of hand;
- Hand weakness;
- IV – th – V – th fingers parasthesia.

2. Neurodystrophic form of cervical brachialgia

- Shoulder – scapula periartrosis;
- Shoulder – hand syndrome;
- Epicondilosis.

Shoulder – scapula periartrosis is the result of muscle – tonic and neurodystrophic tissue disorders. Clinical features:

- Pain and limitation of movements in shoulder;
- Painful palpation of caput os humeri;
- Limitation of arm movements (the patient cannot comb his hair);
- The symptom of frozen shoulder;
- M. deltoideus, m. supraspinatus and infraspinatus atrophy.

There are two stages of this syndrome – algic and dystrophic. In the dystrophic stage not only active movements are limited but the passive ones also.

Shoulder – hand syndrome This syndrome includes clinical features of shoulder – scapula periartrosis and autonomic – trophic changes of hand. Clinical features of algic stage:

- Severe hand pain;
- Edema, hyperemia and cyanosis;
- Hyperesthesia;
- Decreased muscle strength and limitation of movements.

Clinical features of dystrophic stage:

- Muscle atrophy;
- Osteoporosis on X – rays examinations.

Shoulder epicondylitis It is very common in tennis players.

Clinical features:

- Elbow pain;
- Insignificant hypalgesia on external surface of hand.

3. Autonomic – vascular form

Clinical features are the same as in case of lumbar ischialgia.

II. Compressive radicular syndromes on cervical level

1. Radiculopathy C6 radix (C5 – C6 discs):

- Pain, parasthesia and hypalgesia on anterior external surface of arm;
- Weakness, hypotrophy of m. biceps brachii;
- Absent or low flex elbow reflex.

2. Radiculopathy C7 radix (C6 – C7 discs):

- Pain, parasthesia and hypalgesia;
- Weakness, hypotrophy of m. triceps brachii;
- Low extensor elbow reflex.

3. Radiculopathy C8 radix (C7 – C8 discs):

- Pain, parasthesia and hypalgesia;
- Low extensor elbow and carpo – radial reflex.

III. Compressive spinal syndromes

Clinical features are developed during several months or years, sometimes rapidly, acutely. Clinical features:

1. The syndrome of bilateral ventral compression of spinal cord:

- Flaccid upper paralysis;
- Central lower paralysis;
- Conductive sensory disorders;
- Pelvic disturbances.

2. Syndrome of lateral column compression:

- Flaccid upper paralysis;
- Central lower paralysis;
- Conductive sensory disorders on the opposite side.

IV. Compressive vascular radicular – spinal syndromes.

1. Acute:

A. Transient.

- B. Stroke.
- 2. Chronic.

1. **A. Transient** – Sensory and motor disorders are liquidated in course of 1 – 2 weeks.
 - B. Stroke** (It is acute ischemia of radix or spinal cord):
 - At radix artery compression radix stroke is developed;
 - At anterior spinal artery compression only anterior 2/3 of spinal diameter are injured.
2. **Chronic – cervical myelopathy:**
 - Anterior horns lesion syndrome;
 - Lateral columns syndrome;
 - LAS – syndrome;
 - Syringomyelia syndrome.

V. Vertebral artery syndrome:

1. Drop – attacks (reticular formation ischemia);
2. Syncope (pyramidal tract ischemia).

Additional methods of diagnostics:

1. Spinal X – ray – examination;
2. CT of spinal cord.

Treatment

1. Orthopedic.
2. Medicines:
 - At edema: Euphyllinum 2.4% 10.0; Lasix 2.0; Dexamethasonum 4 – 8 mg; NaCl 0.9% 200.0 i/v by drops;
 - Spasmolitics: Platiphillinum, No – spa, Baralginum i/m
 - Analgesics: Reopirini, Voltareni 2.5% 3.0; Tramadol, Aminasini;
 - Non steroids medicines: Ketanov, Dicloberl, Movalis, Ranselex, Celecoxib;
 - Myorelaxants: Midocalm, Baclofen;
 - Chondro– protectors: Rumalon, Alflutop, Arthron-complex, Structum;
 - Vitamins and biostimulators;
 - Physical methods.

3. Surgical methods. The main indications:
 - Horse tail compression;
 - Long lasting (3 – 6 months) pains;
 - Huge hernia (over 15 mm);
 - Acute compression of radicular – spinal artery.
4. Sanatorium.

Prevention

1. Hypokinesia prevention;
2. Moderate physical activity;
3. Treatment of chronic diseases.

Peripheral neuropathies. Neuritis. Neuralgia. Polyneuritis.

Definition

It is multiple symmetrical lesions of peripheral nerves, associated with motor, sensory and autonomic – trophic disorders mainly in distal parts of extremities. If there is any infection among etiological factors this pathology of peripheral nerves is called polyneuritis. If there is an intoxication or hereditary factor in etiology it is known as polyneuropathy.

Classification

I. Etiological

A. Polyneuritis

1. Infectious

- a. Bacterial (neuropathy of leprosy, lues, leptospirosis and so on);
- b. Infectious – allergic (at scarlet – fever, infectious mononucleosis, diphtheria).

B. Polyneuropathy

1. Allergic (vaccinal, serum);
2. Toxic (bacterial, exogenous, iatrogenic);;
3. Dismetabolic (diabetic, uremic, alcoholic, hepatic);
4. Paraneoplastic;
5. Inherited;
6. Ischemic;
7. Radial.

II. Morphological

1. Axonal;
2. Periaxonal;
3. Interstitial.

III. Clinical

1. Motor;
2. Sensory;
3. Sensitive;
4. Autonomic;
5. Mixed.

Common clinical picture of all polyneuritis and polyneuropathies.

One of the common features of all polyneuropathies and polyneuritis is the presence of polyneuritic syndrome that includes three types of disorders:

1. Motor disorders (these ones prevail at motor form) such as flaccid paralysis or paresis with hypotonia, hypotrophy and hyporeflexia (fig. 20).
2. Sensory disturbances in distal parts of extremities such as subjective (pain, parasthesia) and objective (anesthesia or hyposthesia of superficial and deep types of sensation) disorders:
 - a. Sensory form is developed when superficial sensation is lost;
 - b. Sensitive form is developed when deep sensation suffers.
3. Autonomic – trophic disorders are dominant at autonomic form (fig. 50).

In case of any polyneuritis or polyneuropathy the presence of polyneuritic syndrome is obligate, although every polyneuritis or polyneuropathy has its own clinical peculiarities.

The peculiarities of clinical picture of polyneuritis

1. Luetic polyneuritis
 - Sensitive forms (deep muscle – joint and vibration sensation suffers) prevail;
 - Sensitive ataxia dominates in clinical picture;
 - Diagnosis: serum reactions of Wasserman, RIF;;;
 - In treatment Penicillinum long – active antibiotics are used.
2. Leptospirosis polyneuritis
 - Polyneuritic disorders are developed on the background of specific somatic clinical picture;
 - Diagnosis: serum reaction;
 - In treatment antibacterial medications are used.
3. Leprosy polyneuritis.
 - Sensation is lost spot – likely;
 - There are rodentic – inflammatory infiltrates and spots on skin;
 - At first pain and temperature sensation suffers, then tactile one is involved;
 - There are severe autonomic disorders (ulcers, bone deformations).
4. Brucellosis polyneuritis
 - Superficial sensation on the extremities suffers spot – likely;

- Muscular pain;
- Asthenic syndrome;
- The other organs are involved;
- Typical profession;
- (+) Reaction of Rait – Hedelson.

In treatment antibacterial medications are used.

5. Viral polyneuritis (Flu – associated one and so on)

- It is developed on the background of upper respiratory ways disease and fever;
- The onset is acute (in course of 2 – 3 days);
- Severe intoxication;
- CNS is often involved (meningoencephalomyelopolyradiculoneuritis);
- Sensory forms without paresis are the most typical ones;
- Flaccid paralysis are very rare;
- Renovation of nerves' function takes up to 1 – 2 months.

The peculiarities of polyneuropathies' clinical picture

I. Allergic:

- Guillain – Barre
- Those associated with collagenous diseases;
- vaccinal.

1. Guillain – Barre polyneuropathy it is an acute autoimmune auto-allergic disease. In most cases it is developed after infections, trauma and surgery.

The peculiarities of clinical picture:

1. There is flaccid tetraparesis, much more expressed in proximal parts of extremities.
2. IX, X, VII CN's are often involved (bulbar syndrome with respiratory disturbances).
3. There are respiratory disorders in case of between ribs nerves involvement.
4. Sensory disorders are slightly expressed or absent.
5. It is often associated with meningoradicular syndrome, (+) Kernig, Lasseg, Neri signs.
6. Protein – cell dissociation in CSF.
7. The course of the disease has its own peculiarities: the clinical

picture develops rapidly during 7 – 10 days. By the way, the more rapidly it is developed, the better it will regress. In case of typical course of the disease almost all patients recover completely. In case of severe course of the disease (with bulbar and respiratory disorders) convalescence lasts up to 1 – 2 years and 17 % of all patients still have severe motor disorders even after 5 years.

Treatment:

- desintoxication;
- dehydration;
- desensibilization;
- plasmaferesis (8 – 10 seances);
- glucocorticoids.

2. Polyneuropathy at collagenous diseases

One of the most common polyneuropathy in this group is the one associated with periarteriitis nodosis.

The peculiarities of clinical picture:

- The disease starts as multiple mononeuropathy (there are signs of different nerves involvement, for example Ulnar N, Median N and so on);
- Later clinical picture of polyneuritic syndrome is developed;
- The typical changes of internal organs (especially those well blood supplied, such as kidneys, GIT, heart);
- Long lasting hyperemia;
- Skin rash;
- Inflammatory changes in blood;
- Polyarthralgia;
- Cachexy.

Diagnosis is put after morphological investigation of muscles.

To treat such patient Corticoids, Cytostatics are used.

3. Vaccinal polyneuropathy

Usually this group of the disease is developed after the usage of anti rabies vaccine (as an allergic reaction on vaccinal myelin.

The peculiarities of clinical picture:

- The clinical picture of serum disease with fever and arthralgia develops with the 4- th – 6 – th injection;
- The symptoms of brain and spinal cord lesion appear (such

as delirium, cross – eye, CN's lesion (bulbar syndrome, facial neuropathy), pelvic disorders);

- Polyneuritic syndrome appears in 2 – 6 days.

The treatment of such patients includes Glucocorticoids.

II. Toxic polyneuropathy.

1. Bacterial – toxic (diphtheric polyneuropathy, botulinum polyneuropathy).
2. Iatrogenic.
3. Exogenous (Pb, As, Hg, at POC poisoning).

1. Bacterial - toxic polyneuropathy

A. Diphtheric polyneuropathy

Diphtheric toxin is fixed mainly in peripheral nerves and its nuclei, anterior roots, root nerves.

The clinical picture is developed on the background of upper respiratory ways diphtheria in 5 – 6 days. Sometimes diphtheria is diagnosed afterwards because of the peculiarities of polyneuritic syndrome development.

The peculiarities of clinical picture:

- At the beginning of the disease there are accommodation paralysis and other eye movements disorders with development of diplopia and strabismus;
- Bulbar syndrome (IX – X CN's are involved) with respiratory disorders and pneumonia;
- Heart – vascular disorders (bradycardia, myocarditis with arrhythmia, tachycardia);
- In a 1 month polyneuropathy of lower extremities is developed. Mainly it is sensory – motor form;
- Polyneuropathy of upper extremities could be developed in a month afterwards.

In treatment anti diphtheric serum and Glucocorticoids are used.

B. Botulinum polyneuropathy

The main point of action of botulinum poison is the place of nervous – muscular synapse (as the poison doesn't come through the haemato-encephalic barrier). In this case the mechanism of acetylcholine metabolism is destroyed. Thus in the base of polyneuritic syndrome at botulinum polyneuropathy is nervous – muscular deficiency.

The peculiarities of clinical picture:

- The symptoms of the disease appear in a few hours after the usage of spoiled food stuffs (canned goods, sashis);
- Vomiting, diarrhea, stomachache, skin dryness;
- Then midriasis, accommodation paralysis, diplopia, swallow and speech disorders, neck muscles weakness appear;

Diagnosis is put after the results of biological test on mice. Usually blood, vomiting stuffs are used in order to find out the type of the toxin.

In treatment specific serum is used. In case of non-adequate treatment the patients die in 50% of all cases.

2. Iatrogenic polyneuropathy.

This group of the diseases is developed after usage of medications of different groups (Cytostatics, Glucocorticoids, Isoniazidum)

In case of Isoniazidum polyneuropathy there is the deficiency of vitamin B6. That's why phthisiatrician should prescribe vitamin B6 with Isoniazidum.

3. Exogenous polyneuropathy.

A. Lead polyneuropathy

Peculiarities:

- Contact with lead (in those persons who are working in lead enterprises, printing ones and so on). When the concentration of lead in blood is more then 80 mg per 100 ml of blood;
- Lead asthenia;
- Intestinal colic;
- Dark blue bordering on the gum;
- The symptoms of toxic hepatitis;
- Usually the peripheral nerves of upper extremities are involved (such as radial one);
- In lower extremities usually peroneal nerve suffers;
- Sensory disorders are not well expressed and in most cases they are subjective ones (muscle, bone and joints' pain).

In treatment complexions are used: Calcii tetacini 10% i/v, Unithiol i/m; general treatment.

B. Arsenic polyneuropathy.

It is developed at acute or chronic poisoning. Acute one is characterized by enteritis associated with polyneuritis.

Peculiarities:

- Sensory disorders prevail (pain, parasthesia, cramps, feet and hands hyperesthesia, poliesthesia);
- Atrophy of interostal spaces on hands and feet;
- Sensitive ataxia is often developed because of the deep sensation disturbances;
- CN's lesion (VIII, II, sometimes III, VII);
- Trophic disorders: feet and hands edema, white strips on nails. In treatment unitholium and desintoxication are used.

C. Hydrargyrum polyneuropathy.

- Enteral disorders;
- Fever;
- General cerebral symptoms;
- Polyneuritic syndrome;
- Cerebellar atxia;
- Extrapyrimal disturbances.

In treatment complexions and general therapy are used.

D. Polyneuropathy at POS.

At this type of polyneuropathy there is the blockage of cholinesterase. As a result the activity of acetylcholine increases.

An acute poisoning is associated with general somatic clinical picture. And in 2 – 3 weeks in 20 % of all patients polyneuropathy is developed.

Clinical picture:

- Diffuse fasciculations;
- Stomachache;
- Bradycardia with hypotonia;
- Autonomic horror;
- Respiratory disorders;
- Lung edema;
- Loss of consciousness;
- Hyperkinesis;
- Seizures;
- Coma, death.

The severity of polyneuropathy depends on the stage of intoxication in acute period, on hepatitis and nephritis clinical picture.

In polyneuropathic syndrome motor disorders are dominant. Sometimes neurological symptoms are associated with sphincter and cerebellar symptoms. Recovery is very slow. There is high percentage of disability.

Emergency: reactivators of cholinesterase: Dipiroxime, Izonitrozide, Atropinum i/v.

III. Dismetabolic polyneuropathies.

A. Alcoholic polyneuropathy.

It is developed in 32 – 97 % of all patients with chronic alcoholism. Among all types of polyneuropathy alcoholic one takes 36 – 40 %. In pathogenesis of this disease an important role belongs to the vitamin B deficiency because of the liver and gastro – intestinal tract disorders.

Besides alcohol directly influences on neuronal metabolism. Thus alcoholic polyneuropathy is demonstrational alimentary- toxic polyneuropathy.

According to the course of the disease there are acute and chronic form of the disease. At acute one paresis is developed rapidly, while at chronic one clinical picture manifests gradually.

The peculiarities of clinical picture:

- At first lower, then upper extremities suffer;
- There is well expressed lesion of peroneal nerve: feet extensors weakness, specific gait disorders;
- At the beginning stretch reflexes increase, then decrease and sometimes even disappear;
- Ankle reflexes disappear at the early beginning of the disease;
- Sensory disturbances at first are irritating: pain, parasthesia, hyperesthesia and hyperpathy dominate;
- There is sensitive ataxia because of the deep sensation disturbances;
- Polyneuropathy is associated with amnestic Korsakov syndrome;
- There are internal organs' changes – chronic hepatitis, cirrhosis and so on.

In treatment high doses of vitamin B, desintoxication, liver protectors, Berlition, Dialipon are used.

B. Diabetic polyneuropathy.

This type of polyneuropathy is developed in 65 – 70 % of all patients with diabetes.

The peculiarities of clinical picture:

1. Sensory disturbances on lower and upper extremities such as parasthesia, hyperesthesia, hypesthesia and pain are dominant.
2. There are autonomic disorders: dryness of skin, an- or

- hyperhydrosis, trophic ulcers and so on.
3. Knee and ankle reflexes are lost at the beginning of the disease.
 4. Severe paresis is very rare in this case.
 5. There is visceral form of diabetic polyneuropathy with progressive autonomic insufficiency.
 6. It is one of the most light and slowly progressive polyneuropathy.

IV. Paraneoplastic polyneuropathies.

The disease is developed on the background of malignant tumors or 2 – 3 years before the final diagnosis.

Sometimes polyneuropathy appears before or just after surgery.

V. Hereditary polyneuropathies.

1. Charkot – Marie – Tooth neural amyotrophy.
2. Interstitial polyneuritis in children.
3. Hereditary sensory radiculopathy.
4. Lipidosis polyneuropathies.

Charkot – Marie – Tooth neural amyotrophy

It is one of the most common hereditary polyneuropathy (fig. 77).

The peculiarities of clinical picture:

1. It is inherited according to the dominant or recessive type.
2. It manifests in middle age and slowly progresses.
3. Dystal amyotrophy and lower extremities weakness prevail.
4. The lower extremities look like turned over its head bottle.
5. The foot looks like Friedreich one.

The symptoms of lesion of peripheral nerves

1. Radial nerve

- Forearm extensors weakness;
- Hand extensors weakness;
- Fingers basal phalanx extensors weakness;
- Impossible supination;
- Impossible adduction of thumb;
- Lost sensation on dorsal thumb surface and in space between the I st and II nd small bones.

2. Ulnar nerve

- Hand flexors weakness;

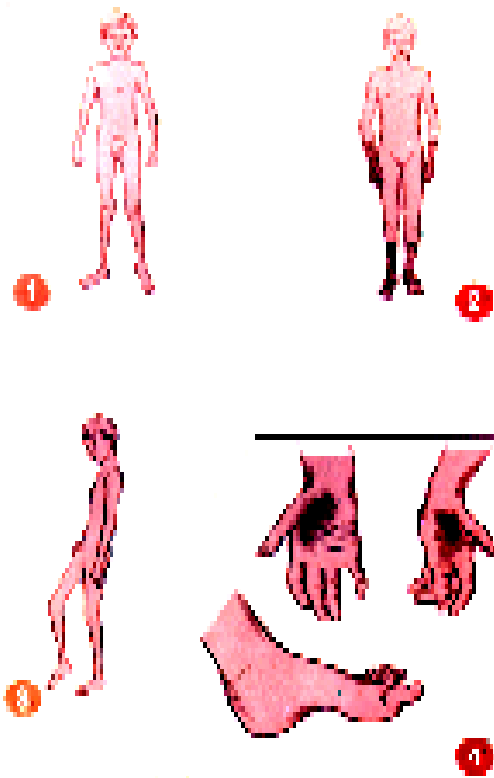


Fig. 77. Charcot-Marie-Tooth syndrome:
 1. distal atrophy of the extremities;
 2. polineuritic sensory disorders; 3. Cock's
 gate; 4. Fridraych's foot

- The IV th and V th fingers distal phalanx extensors weakness;
- Thumb abductors weakness;
- Hypothenar hypotrophy;
- Hand looks like bird paw;
- The patient cannot scrawl with the nail of the V th finger, play on the piano and keep the sheet of paper with the I st and II nd fingers;
- Anesthesia in the V th finger and hypothenar.

3. Median nerve

- Impossible pronation;;
- Hand flexors weakness (especially of the I, II and III fingers);
- Thenar hypotrophy;
- Hand looks like monkey paw;
- Disability to oppose the thumb to the rest fingers;
- Disability to make a fist as a result of thumb flexors weakness;

- + mill sign;
- Anesthesia on hands palm surface except the half of the IV th and V th finger;
- Severe autonomic disorders.

4. Peroneal nerve

- Feet and toes extensors weakness;
- Feet adductors weakness;
- Typical gait – cock like;
- Anesthesia of external edge of shin and dorsal surface of feet.

5. Tibial nerve

- Feet and toes extensors weakness;
- Feet abductors weakness;
- Disability to walk on tip toes;
- Anesthesia on sole and external surface of the feet.

6. Sciatic nerve

- Complete paralysis of foot and toes;
- Impossible flexion in knee joint and shin – foot joint;
- Shin and hip atrophy.

Facial nerve neuropathy (Bell's palsy)

Etiopathogenesis

The nerve lesion occurs due to the ischemic anoxia as a result of cold catching, inherited narrowing of facial nerve canal, local infectious – allergic processes. The main pathogenetic factors are edema and ischemia.

Clinical picture

There is facial mimic muscles paralysis, facial asymmetry, lagophthalm, Bell's sign, taste disorders and tears or dryness of the eye.

Treatment in acute period:

1. Dehydration (Euphillinum, Lasix, Dexamethasonum i/v);
2. Anti-inflammatory therapy (Glucocorticoids);
3. Non steroid anti-inflammatory drugs (Diclofenac, Movalis, Naclofen, Messulid);
4. Fonoforesis with Hydrocortisonum;
5. Microcirculation improving (Actovegin, Solkoseril, Pentoxiphillin).

At recovery:

1. Massage;
2. Electrophoresis with Proserinum, Nicotin acid;
3. Warming procedures;
4. Biostimulants (Plasmol, Aloe);
5. Vitamins B, PP;
6. Acupuncture.

Trigeminal neuralgia

The disease is polifactorial. An important role belongs to chronic processes, trauma, tumors and so on.

The peculiarities of clinical picture:

- Pain paroxysms that are caused by the movements of mimic muscles, talking, eating, face washing. They can continue up to 1 – 2 min;
- Pain tics – facial hyperkinesis;
- Trigger points – touching of these points can cause severe pain.

Treatment:

- anti seizures medications (Finlepsin);
- Non steroid drugs;
- Acupuncture.

Demyelinating diseases

Multiple sclerosis

Multiple sclerosis (MS) – is a chronic disease that begins most commonly in young adults and is characterized pathologically by multiple areas of central nervous system (CNS) white matter inflammation, demyelination, and glial scarring (sclerosis). The lesions are therefore multiple in spaces.

The clinical course varies from a benign, largely symptom – free disease to one that is rapidly progressive and disabling. Most patients begin with relapsing and remitting symptoms. At first recovery from relapses is almost complete, but then neurological disabilities accrue gradually. The lesions are therefore multiple in time and space.

The cause is elusive, although autoimmune mechanisms, possibly triggered by environmental factors in genetically susceptible individuals, are thought to be important.

Epidemiology

Age of onset is between 20 – 40 years. Usually it is 21 – 25 years, in women – 2 – 3 years earlier. In women the incidence of MS is 1.5 – 2 times higher than in men. Nowadays there are about 2 mln people with MS all over the world. The geographic distribution is uneven. Most of northern USA, southern Canada, northern Europe, southern Australia and New Zealand are areas of high prevalence.

In Ukraine the incidence of MS is 15 per 100 000 people. But it is much more higher in western regions (25 per 100 000 people) than in eastern and southern ones (6 – 8 per 100 000 people).

The main cause of the increased growth of the disease:

1. Better diagnosis;
2. Unitary diagnostic scales;
3. Increasing possibilities of treatment that leads to the growth of percentage of the patients with long lasting course of the disease;
4. True growth of MS incidence.

Etiology

The cause of MS is unknown. There are 2 groups of possible reasons of the disease:

- I. Genetic susceptibility.

- II. Environmental factors:
1. Infections (the virus can influence on nervous system directly or through the autoimmune mechanisms).
 2. Geographical (ground, water properties, the number of light days in a year).
 3. Toxic.
 4. Social conditions.
 5. Diet (domination of meat in the diet).
 6. Other factors (trauma).
- I. Compelling data indicate that susceptibility to MS is inherited. The major histocompatibility complex (MHC) on chromosome 6 has been identified as one genetic determinant for MS. The MHC encodes the genes for the histocompatibility antigens (the human leucocyte antigen [HLA] system) involved in antigen presentation to T cells. The class II haplotype DR15, DQ6, Dw2 is associated with increased risk of MS.
- II. 1. In MS demyelination could be precipitated by a viral infection. Measles, rubella, mumps, coronavirus, parainfluenza, herpes simplex, Epstein – Barr and human T – cell lymphotropic virus type I viruses all have been reported to be present in patients with MS. Perhaps no single virus is the trigger for demyelination in all patients with MS. Instead, several different viruses may be involved.
2. The geographic distribution of MS is uneven. In general, the disease increases in frequency with latitude in both the northern and southern hemispheres, although the rates tend to decrease above 65 degrees north or south. Most of northern Europe, northern United States, southern Canada, southern Australia and New Zealand are areas of high prevalence. Southern Europe, southern United States, Asia Minor, the Middle east, India, parts of northern Africa and South Africa have medium prevalence rates. Low prevalence areas are Japan, China, Latin and South America.
3. Toxins can cause changes of biochemical and antigen properties of myelin. Also they suppress remyelination and activate immune modulation. Such substances as Cu, Zn, Mn, Co and vitamin D and B12 deficiency can also cause demyelination.
4. Emotional stress and other social conditions play important role in pathogenesis of this disease.
5. Meat diet is considered to be one of the factors that can provoke MS.

Pathogenesis

Different etiologic agents provoke autoimmune mechanisms. The result of this process is myelin destruction. At the beginning of the process auto-allergic processes prevail over the other ones. Then immunodeficiency is developed. The typical features of MS pathogenesis are:

- Clinical and immune signs are closely connected with each other in MS patients. Usually immune signs are the first ones;
- There is disturbance of activating and suppressing cytokines balance;
- The immunity is changed in the course of the disease;
- There are signs of immune suppression and immune modulation according to the stage of the disease – exacerbation or remission.

Pathology

There are multiple areas of Central Nervous System white matter inflammation, demyelination and glial scarring (sclerosis). The lesions are multiple in space. They are located mainly in spinal cord, cerebellum, n.Opticus, brain white substance (fig. 78).



Fig. 78. Multiple sclerosis (Frank H. Netter, M.D.)

Clinical features

The beginning of the disease usually is slow. The first symptom is one of the next ones:

1. **Paresthesia**. It is the feeling of numbness or tingling in one of the extremity. It can be spread during the next 3 – 4 days and lasts for about 1 – 2 weeks, then gradually disappear.
2. **Motor disorders** – weakness in lower extremities. This symptom is much more common at the age of 25 – 40 years.
3. **Retrobulbar neuritis** is a progressive loss of vision, colour vision disturbances. It lasts for about several weeks.
4. **N. Oculomotorius disorders** (diplopia and cross eye).
5. **Pelvis disorders** (retention of urine, micturition).
6. Acute vestibular syndrome.
7. **Cerebellar disorders** – ataxia, disorders of coordination.

The **typical clinical features** of MS:

1. Motor disorders – 89 – 97%;
2. Ataxia – cerebellar, sensitive and vestibular – 62 – 74%;
3. Sensory disorders – pains and sensitive ataxia – 72 – 74%;
4. Brain stem symptoms – vestibular syndrome, dysarthria, CN's lesion – 47 – 58%;
5. Visual and eye movements disorders – 42 – 52%;
6. Autonomic disturbances – pelvic and sexual disorders – 46 – 60%;
7. Nonspecific symptoms – cognitive, memory disturbances, loss of attention – 62%;
8. Paroxysmal symptoms.

1. Motor disorders

Hemiparesis, lower paraparesis and monoparesis are common symptoms of MS. Upper extremities are injured very seldom. The typical signs of these symptoms are low muscle strength, the presence of pathological reflexes and low abdominal reflexes. There are also changes of muscle tonus – spastic hypertonus, hypotonus or dystonus. Hypotonus can be the sign of cerebellum and spinal cord posterior columns lesion.

2. Ataxia

There are dynamic and static ataxia, dysmetry, hypermetry, intention, asynergy.

3. Sensory disorders

Subjective sensory disturbances are early signs of MS. Then conductive sensory disorders are joined to them. Muscle – joint sense usually suffers at the fifth year of the disease and later. The loss of vibration sense points on posterior columns lesion.

4. Brain stem disturbances

There is vestibular symptom with dizziness, nystagmus and vestibular ataxia. Sometimes trigeminal pains are observed.

5. Visual and eye movements disorders

The typical features of MS are retrobulbar neuritis, subatrophy of optic nerve disc, decoloration of disc's temporal part. Eye movement disorders mean that there are syndromes of ophthalmoplegia.

6. Autonomic (pelvic) disorders

A. Syndrome of m. Detrussor hyperreflexion. That means urine bladder inability to accumulate urine. The main symptoms are micturition, increased frequency of urination, incontinence of urine, retention of urine.

B. Incomplete urine bladder emptiness. Dyssynergy of m. Detrussor and Sphincter.

7. Nonspecific symptoms

There are general weakness, cognitive disorders, memory, attention disturbances, behavioral disorders, depression, euphoria and fatigue syndrome.

8. Paroxysmal symptoms

They unite short lasting sensory and motor disorders. They can be observed 200 – 300 times per day. They are:

- a. Tonic muscles spasm (painful and short lasting);
- b. Dysarthria and ataxia attacks;
- c. Lermitt symptom – it is a short lasting feeling of electrical current along the spinal cord;
- d. Paroxysmal trigeminal pains;
- e. Atypical pains in extremities;
- f. Paroxysmal itching;
- g. Paroxysmal choreoatetosis;
- h. Paroxysmal nystagmus;
- i. Paroxysmal facial hemispasm;
- j. Epileptic attacks (focal and general).

Pains are very often observed at MS. They can be paroxysmal or chronic ones.

In some patients Uthoff's symptoms can be observed – it is the worsening of patients state after the hot bathroom or hot meal.

Clinical forms

1. Cerebral forms:
 - a) cortical (epileptic attacks, psychiatric disorders);
 - b) visual;
 - c) brain stem;
 - d) cerebellar.
2. Spinal forms:
 - a) Cervical;
 - b) Thoracic;
 - c) lumbar – sacral;
 - d) pseudotabes.
3. Cerebrospinal.

The course of the disease:

1. Acute
2. Subacute
3. Chronic – remittent, remittent – progressive, progressive – remittent, progressive

The most common is remittent course of the disease, when exacerbation follows remission.

The periods of the disease:

1. Exacerbation
2. Remission (complete, incomplete)
3. Stable period

MS degree: I, II, III, IV, V

- I – patient has difficulty to walk only after physical training;
- II – patient has difficulty to walk and weakness on 2-3 km;
- III – patient has spastic-paretic gait, difficulty to walk and weakness on 200-300 m;
- IV – patient can't to walk without help;
- V – patient can't to walk or has blindness.

Diagnosis

In order to put a right diagnosis we use clinical and paraclinical data. There are such types of diagnosis:

- Veridical MS;
- Credible MS;
- Possible MS.

To put veridical MS we have to reveal in patient at least 2 focuses of lesion and 2 exacerbations, or 2 exacerbations of 1 clinical focus and 1 paraclinical supposed focus.

For **MS diagnosis** we use:

I. Immune examinations of blood and CSF:

1. Usually there are increased Ig G, M, A contents;
2. Insignificant increasing of protein content and moderate pleocytosis in CSF;
3. Lymphocytosis, eosynophilia – in exacerbation stage; leukopenia, lymphopenia – in the period of remission;
4. Increased thrombocytes aggregation and fibrinogen content;
5. Increased Ig content in serum and decreased T – lymphocytes quantity.

II. The most accurate method is MRI. According to the accepted criteria there should be at least 3 focuses (2 of them should be located paraventricularly, 1 – subtentorially (that means in brain stem or cerebellum). The diameter of focuses should be at least 6 mm, or there should be 4 focuses, 1 of them periventricularly.

III. The method of evoked potentials. This is a method that reveals bioelectrical brain activity in response to the stimulation. This method is not a specific one for MS diagnosis.

Treatment

1. Pathogenetical treatment.
2. Symptomatic treatment.

1. Pathogenetical treatment:

- a. Corticosteroids and ACTH;
- b. Cytostatics and immune modulators, non specific immune suppressors;
- c. Cytokines, interferones;
- d. Antigen – specific immune therapy.

a. Corticosteroids and ACTH

These medicines have immune suppressive action and can shorten the period of exacerbation. The main representatives are – Prednisone, Methylprednisolone, Dexamethasone.

Prednisone is used orally 1 – 1.5 mg/kg/day twice a day during 10 – 14 days. Then during the next 2 months we decrease the dose gradually.

One of the most popular schema for **Methylprednisolone** usage is 500 – 1000 mg per day i/v in 500 ml of physiological solution during 3 – 5 days. Then Prednisone is used in dose 0.5 – 1 mg/kg during 3 – 7 days with gradually decreasing of dose during the next 2 – 3 weeks. This way of usage has much more expressed and quick effectiveness and insignificant outside effects.

Dexamethasone is used i/v or i/m according to the schema – 8 mg per day during 7 days, 4 mg – 4 days, 2 mg – 3 days. It is used at retrobulbar neuritis.

The peculiarities of Corticosteroids usage:

- Long lasting and frequent usage is undesirable;
- Usually H-2 blockers are used together with Corticosteroids;
- ACTH has immune suppressive activity, inhibits cellular and humoral immunity. It is used in dose 40 – 100 U i/m during 10 – 14 days;
- Plasmapheresis is used in case of exacerbation.

b. Cytostatics and immune modulators, non specific immune suppressors

These are the medicines of choice and are used at chronic progressive forms when there are no effects from the Corticosteroids. To them belong Asatioprine, Cyclophosfamidum, Cyclosporinum A. But all of these medicines have a lot of side effects.

The representatives of immune modulators are – T – activinum, Timalinum, Myelopid, Levamisolum. They are prescribed at progressive forms of MS.

T – activinum is used in dose 100 mcg s/c every evening during 5 days, then 1 – 3 injections every 10 days.

Timalinum is used in dose 10 mg i/m twice a day during 5 days, then every 10 days 2 injections are used.

c. Interferones

They are used mainly for exacerbations' prevention and selec-

tive immune correction. There are 3 types of Interferonum – γ , α , β .

- **α – Interferonum** has neither toxic nor treating activity;
- **γ – Interferonum** activates immune system and that's why it provokes exacerbations;
- **β – Interferonum** inhibits production of γ – interferonum, increases activity of T – suppressors, has antiproliferative, antiviral and immune modulating properties.
- **Rebif** – is a modern human β – interferonum produced by “Serono” production and is used for MS treatment. It is used in dose 6 – 12 mln s/c three times per week. It is one of the most effective modern medicines in MS patients, but unfortunately it is very expensive.

d. Antigen – specific immune therapy

One of the representatives of these medications is **Copaxone**, made in Israel. Cost of treatment is about 7 000 \$. It is used in dose 20 mg per day s/c during 6 – 24 months. It has selective immune modulating action.

Besides all of these mentioned medicines in MS patients we use also:

- Vitamins B group;;
- Desensibilizative medicines;
- Amino acids;
- Nootrops;
- ATP, Cocarboxylasa;
- Biostimulants;
- Entero and hemosorption;
- Antiplatelet;
- Antioxydants;
- Angioprotectors;
- Inhibitors of proteolytic enzymes;
- Regeneration stimulants.

Symptomatic treatment of MS

Pelvis disorders:

- Proserinum, Halantaminum decrease m. Detrussor hyperreflexion.
- α – Adrenoblockers decrease dysynergy of Sphincter and Detrussor.

Spasticity:

- Baclofen 5 mg 3 times per day.

- Sirdalude 4 mg 3 times per day.

Tremor:

- β – Adrenoblockers are used at postural tremor.
- Clonasepam, Carbamasepam are used at intention.

Hyperkinetic form:

- Adrenoblockers.
- Antidepressants.

Asthenia:

- Psychostimulants.
- Dopaminergic medicines.

Paroxysmal signs:

- Carbamasepinum.
- Filepsin.

Prevention of MS To avoid catching cold, acute respiratory infections, stress, pregnancy and childbirth, isolation and heating procedures.

Working ability The patients with MS usually have one of invalidity group:

- III – if they need much more comfortable working conditions;
- II – patients with well – expressed permanent neurological signs;
- I – patients that need somebody to care about them.

Acute multiple encephalomyelitis (AMEM)

It is an infectious – allergic disease that is characterized by acute multiple lesion of the brain and spinal cord.

The etiology of this disease is still unknown. Perhaps it is a virus.

The pathology consists of multiple perivascular demyelinating focuses and vascular inflammatory reaction. The process is localized in brain white substance, brain stem and spinal cord.

Clinical features

The disease develops acutely in most cases. In the beginning it is very similar to acute respiratory infection. There are headache, vomiting, general weakness, fever, psychomotor excitement. Sometimes general cerebral and meningeal syndromes are well – expressed. 3 – 7 days later focal signs appear.

Focal signs are variable. Different cerebral and spinal structures can be involved in the process.

In some patients there are symptoms of lesion of radices and peripheral nerves associated with pain, sensory disorders, amyotrophy and low reflexes.

Paraplegia in such patients often is associated with conductive sensory disorders, pelvis disorders and bed – sores.

The most common cerebral disturbances in such patients are nystagmus and eye movement disorders, CN's bulbar group lesion.

Clinical forms of the disease:

- **Encephalomyelopoliradiculoneuritis** – it is the most common form of the disease, which is characterized by the lesion of all parts of nervous system.
- **Polioencephalomyelitis** – it is characterized by the lesion of CN's nuclei and spinal cord gray substance.
- **Opticoencephalomyelitis and opticomyelitis** – are characterized by optic nerve neuritis and symptoms of lesion of brain and spinal cord.
- **Disseminated myelitis** – the spinal cord is damaged on different levels.

Treatment of the disease

Corticoids are used in order to treat such patients. Among them are Prednisolone and **Methylprednisolone**. The last one is used in dose 10 – 15 mg per kg i/v by drops per day. Later we can use it in pills – 1.5 – 2 mg/kg every other day.

Together with this medicine we prescribe **anabolics, K, Ca, vitamin C**.

In acute stage we prescribe **desensibilizing** and **dehydrating** medicines. In case of severe bulbar disorders we include resuscitation measures.

Plasmapheresis and vitamin B are also used.

In residual period we prescribe massage, dibasol, KJ, biostimulants, Lidasa, Seduxen, sanatorium treatment.

Amyotrophic Lateral Sclerosis. Syringomyelia.

Amyotrophic lateral sclerosis is a chronic progressive disease of nervous system and is defined pathologically as one in which there is degeneration of both upper and lower motor neurons of brain and spinal cord. It is characterized by unfavourable prognosis. The disease was first described by Charcot and Joffroy in 1869.

Epidemiology

The incidence rate of Amyotrophic Lateral Sclerosis of 2 – 5 cases per 100 000 population has been found in many countries. As many as 60 000 – 70 000 patients have this disease at any one time all over the world.

The disease is one of middle and late life. Only 10% of cases begin before age 40 years; 5% begin before age 30. In most series, men are effected one to two times more often than women. There is no known ethnic predilection.

The disease is found worldwide in roughly the same prevalence. However, about 50 times that number were found on the island of Guam.

Etiology and pathogenesis

The cause of ALS is not known. Most of scientists consider this disease to be a polietiologic one.

The main theories:

1. Connection of ALS with Prion Diseases. This can be proved by high content of Arginaza in the brain of animals in experiment and in the CSF of patients with ALS. Besides this we can find morphologic changes in the brain of such patients. They are spongiosis and synapse degeneration. This can be explained by the deficiency of Argininum.
2. Autoimmune theory. There are certain changes in cell – mediated and humoral immunity.
3. Genetic theory. The patients with familial forms of ALS have changes of gene, located in the 21st chromosome. This gene is responsible for the production of endogenous antioxidants. The deficiency of such antioxidants can cause activation of free radical oxidation that leads to the death of neurons of anterior horn.
4. Amino acids, neuromediators, neuropeptides metabolism disor-

ders. And those chemical substances are responsible for the apoptosis. That's why the patients with ALS have low content of Mg, Mn, AL, Se in biologic fluids.

The main reasons of ALS development are:

1. Insufficiency of spinal blood circulation.
2. Vertebrogenous pathology.
3. Trauma.
4. Infections.
5. Demyelination.
6. Metabolic disorders..
7. Tumours (rarely).
8. Radiation.

Pathomorphology (fig. 79)

The pathologic process is localized in:

- Anterior horns of the spinal cord;
- Pyramidal tract;
- Motor neurons of IX, X, XI, XII CN's, sometimes – V, VII.

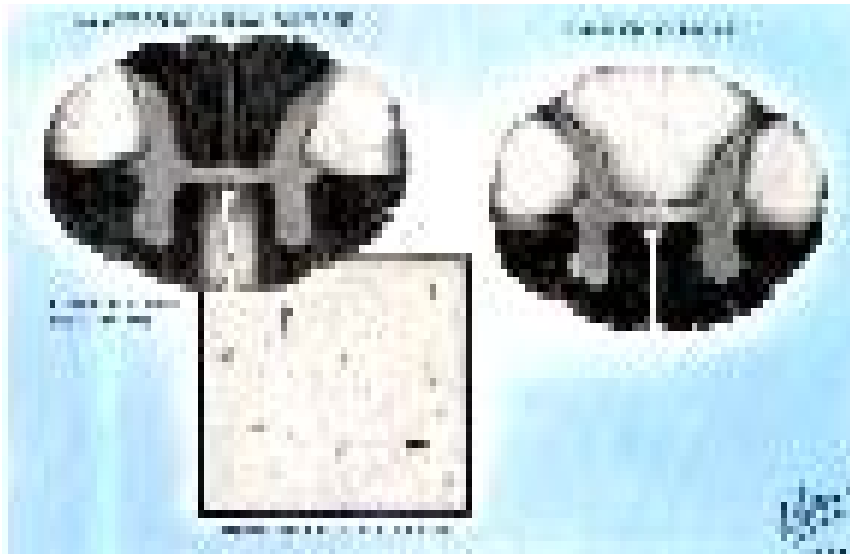


Fig. 79. Amyotrophic Lateral Sclerosis (Frank H. Netter, M.D.)

There is degeneration of third and fifth cortex layers of the anterior central gyrus (small and large pyramidal cells).

Besides there is demyelination of pyramidal tracts (in the internal capsule, lateral and anterior horns of the spinal cord) that cause the death of axial cylinder. That means that the fibres irreversibly die.

Classification

There are **three forms** of the disease:

1. Classical or sporadic ALS.
2. Familial ALS which is usually associated with:
 - Dementia;
 - Extrapyrarnidal disorders;
 - Extrapyrarnidal and cerebellar disorders;
 - Extrapyrarnidal and sensory disorders;
 - Peripheral neuropathy.
3. Complex – ALS – Parkinson disease – Dementia.

Familial ALS usually begins at the age of 45 – 48. This form can be inherited as autosomal dominant and autosomal recessive one.

According to the localization of the process at the beginning of the disease we can differ such clinical forms:

1. Cerebral;
2. Bulbar;
3. Cervical – thoracic ;
4. Lumbar – sacral.

Clinical features of the main forms of ALS and its diagnostics

According to the International Neurologic Community in 1990 the main criteria of ALS were established:

- The symptoms of lower motor neuron lesion;
- The symptoms of upper motor neuron lesion;
- Progressive course of the disease (during 6 months and more);
- No effects from medicines and physiotherapeutic treatment.

ALS is ruled out at:

- Sensory disorders;
- Pelvic disorders;
- Eye movements disorders;
- Autonomic disturbances;

- Also if it is not a familial form – in case of Parkinson disease and Dementia.

1. Anamnesis of the disease.

The first symptoms of the disease are weakness, especially in extremities (hands in particular), awkwardness during the performance of acute movements, reduced muscles of thenar and hypothenar, fasciculations. Forerunner symptoms of ALS are crampies (painful tension of calf muscles). These symptoms were observed in 30% of patients 3 – 6 months before the other signs of the disease.

2. Typical clinical picture:

- Flaccid and spastic paralysis;
- Muscles atrophy;
- Bulbar and pseudobulbar signs.

The reflexes at the beginning are very high with wide reflexogenic zones. There are all the groups of pathologic reflexes. But:

- Abdominal reflexes are preserved for a long time;
- There are no pelvic disorders.

Clinical picture of high (cerebral) form of ALS

It is observed in 1-2% of all patients with ALS. It is characterized by spastic tetraparesis and pseudobulbar syndrome. Sometimes at this form Extraparamidal nervous system (the signs of Parkinson disease) and cortex of frontal lobe (the signs of Dementia) is involved.

Clinical picture of bulbar form of ALS

It is observed in 25 % of all patients with ALS. At the beginning the process is localized in the medulla oblongata and causes the lesion of IX, X, XI, XII CN's that leads to the classic bulbar syndrome. Then it is joined by anterior horns amyotrophy and pyramidal disorders.

Clinical picture of cervical – thoracic form of ALS

It is observed in 50 % of all patients with ALS. The disease begins with the weakness and reduced muscles of proximal and later distal parts of the upper extremities. There is well – expressed hypotrophy, increased muscle tone according to the spastic type, hyperreflexia, pathologic signs. That

means that there is a mixed paresis. In the course of the disease the reflexes fade away, the peripheral palsy dominates, bulbar symptoms are joined.

Clinical picture of lumbar – sacral form of ALS

It is observed in 25 % of all patients with ALS. The first signs of the disease are connected with the peroneal muscle group lesion. This is associated with high stretch reflexes, pathologic signs. This form is characterized by slow progress.

Depending on the combination or domination of some clinical signs there are:

1. Classical type – when anterior horns and lateral columns are involved in pathologic process in equal way.
2. Poliomyelitic type – in 10 – 12 % of all cases the lesion of anterior horns (that means amyotrophy) dominates.
3. Spastic type – the conductive disorders dominate.

The stages of the disease:

- I. There are only subjective signs (complaints). This stage lasts for about 2 – 3 years.
- II. There are the clinical signs of the disease – focal and bulbar disorders. This stage lasts for about 6 months – 2 years.
- III. The process involves neighbour regions. This stage lasts for about 4 months – 2 years.
- IV. Terminal stage. The patients need somebody's care. This stage lasts for about 3 months – 1 year.

The additional methods of diagnostics

ENMG:

- Regular rhythmical fasciculations;
 - Decreased speed of impulse conductance along motor fibres but preserved speed of impulse conductance along sensory fibres.
- This method is very important in the diagnostics. Due to it we can:
- Localize the process;
 - To find out its expansion;
 - The degree of lower and upper motor neurons lesion;
 - To register fasciculations as usually it is one of the first symptoms of the disease.

MRI:

- We can diagnose atrophy and bilateral degeneration of cortico – spinal tracts. In case of high lesion we can find the focus in the posterior crus of the internal capsule.

PET:

- One can register the low content of glucosa in the region of anterior central gyrus. This sign by itself doesn't have very important meaning.

CSF – examination:

- Increased protein level;
- Increased activity of Arginaza.

Differential diagnosis should be made with following diseases:

1. Chronic stage of Acaridan encephalitis.
2. Cervical ischemic myelopathy, which is characterized by slow progress and often with the termination of the process:
 - Limited fasciculations, which are non – rhythmical;
 - A great attention should be paid to the X– ray – examination, MRI, ENMG;
3. Subacute poliomyelitis which is characterized by the absence of pyramidal tract lesion.
4. Spinal amyotrophy, which is characterized by slow benign course. Pyramidal signs are very rare at this disease.
5. Syringomyelia (anterior horns form). There are sensory and segmental trophic disorders.
6. The tumours of the cervical part of the spinal cord – intramedullar. Only MRI and CT examination can differ those pathologies at early stages.
7. Radiculoischemia on the lumbar level is characterized by peculiarities of development, vertebrogenous syndrome, radicular syndrome and partial regress in case of treatment.

The course of the disease and prognosis

There is progressive course of the disease. The result of ALS in most cases is death of the patient. Sometimes the periods of stable symptoms can be observed during 1 – 3 years.

The duration of life in such patients depending on the clinical form of the disease:

1. Cerebral form – 4 – 6 years.
2. Bulbar form – 1 – 3 years.
3. Cervical – thoracic form – 6 – 8 years.
4. Lumbar – sacral form – 10 – 12, 9 – 16 years.

The main principles of treatment:

1. For the examination and diagnosis putting the patients are sent to the neurologic hospital.
2. There is no effective treatment for ALS.
3. For the stabilization of the process there are used:
 - Vit B (2 – 4 ml per day during 30 days);
 - Vit E 1500 mg per day;
 - Biostimulators;
 - Anabolics (Nerobol 5 – 10 mg twice a day during 1.5 – 2 months, Retabolil – 1 ml (50 mg) once a week);
 - As pathogenetic treatment we prescribe Delargil 2 mg twice a day i/v during 10 days, then i/m up to 1 month;
 - Tireotropinum i/v by drops 500 mg;
 - Riluzan 100 mg per kg during 12 months;
 - At increased tonus myorelaxants are used;
 - Dipheninum is prescribed at crampy;
 - Antidepressive medicines;
 - Electro – therapy is excluded;
 - The patients in the third and fourth stage need somebody's care. Sometimes fixative equipments are used. At bulbar disorders the patients get food in a special way.

Syringomyelia

The term Syringomyelia connotes a chronic, progressive disorder that most often involves the spinal cord.

Syringomyelia or cavitations of the spinal cord (fig. 80) was first described by Esteinne in 1546 in «La Dissection Du Corps Humain». The precise term syringomyelia referring to cavitations of the spinal cord is attributed to Charles P. Ollivier D'Angers in 1827. He attributed the abnormal dilatation of the central canal to a developmental abnormality.

Syringomyelia is quite rare. It occurs more frequently in men than in women. The first sign of the disease appears in the second or third decade of life but sometimes it may begin in childhood or late adulthood.

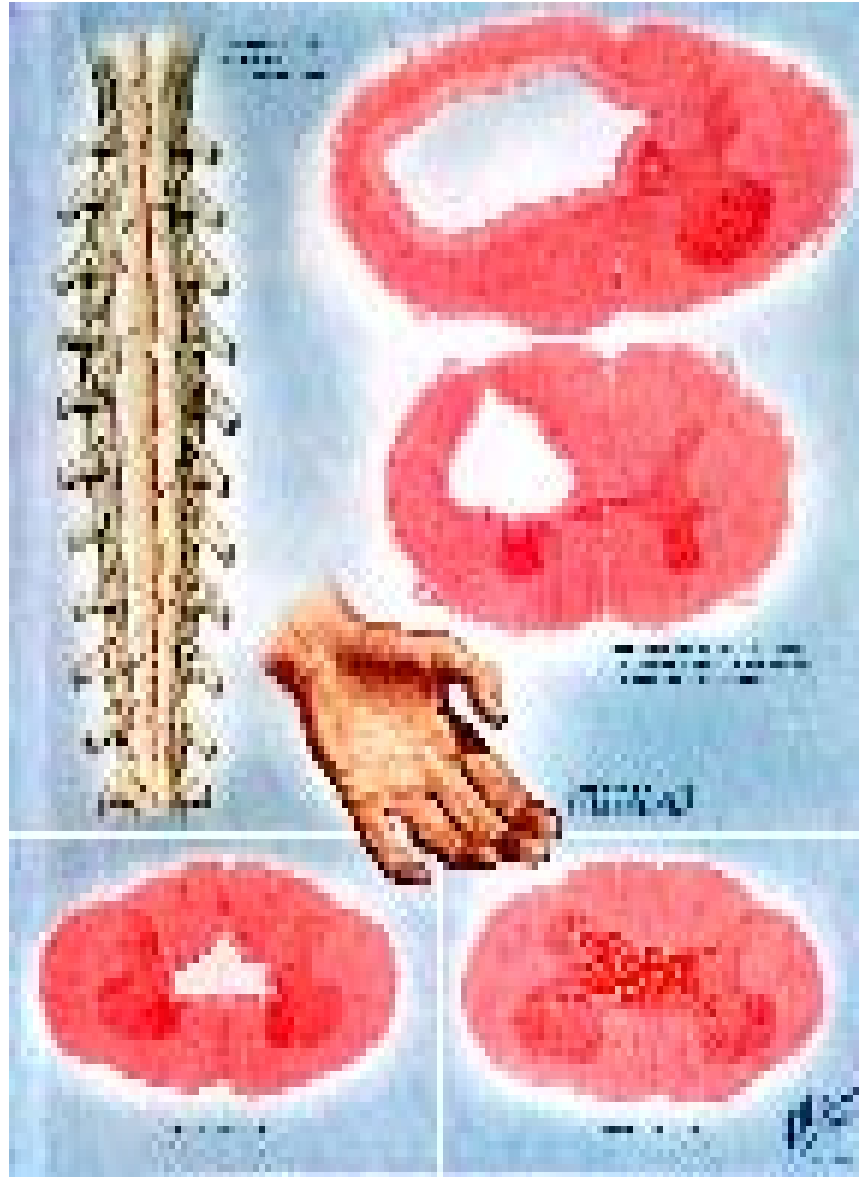


Fig. 80. Syringomyelia. (Frank H. Netter, M.D.)

Etiology

Normally on the 6th – 8th week of embryo development as a result of high production of CSF and increased pressure in the neural tube the connection between the ventricular system and subarachnoid space is established and the central canal is obliterated.

The occlusion of drain holes can cause the widening of central canal and creation of cavities.

But this theory has some weak places. That's why we should pay attention to the others possible reasons of creation of medullar cavity. They are:

- Necrotic myelitis;
- Hemorrhage;;
- Trauma of spinal cord;
- After meningitis.

In 1971 Grinand supposed that Syringomyelia is one of the slowly progressive infections. According to the pathology there are three types of Syringomyelitic cavities:

- Symmetric central – medullar – the simple wideness of canal;
- Central with paracentral expansion. They are usually located in the brain parenchyma and are the result of previous diseases.

Classification

There are several classifications according to the clinical features, expansion, and course of the disease, etiology, pathogenesis and the stage of the disease.

According to the clinical form (Borisova N.A. 1989) there are such forms:

- Posterior horns – 40 – 50 %;
- Anterior horns – 10 – 15 %;
- Autonomic – trophic – 5 – 10 %;
- Mixed – 30 – 40 %;
- Bulbar.

According to the expansion of pathologic process there are:

- Spinal (cervical – 2 – 4 %, cervical – thoracic – 70 – 80 %, thoracic – 10 %, lumbar – sacral – 1 – 2 %, total – 8 – 10 %);
- Brain stem;
- Brain stem – spinal.

According to the course of the disease the are:

- Stable – 25 %;
- Slowly progressive – 50 %;
- Rapid progressive – 25%.

According to the etiology and pathogenesis (1998):

- I. Idiopathic.
- II. Secondary:
 1. As a result of cranio – vertebral abnormality;
 2. After trauma;
 3. Associated with hydrocephalus;
 4. As a result of cervical part of spinal cord stenosis;
 5. As a result of lumbar myelitis;
 6. At Pegett disease;
 7. At cysts of posterior cranial fossa;
 8. At tumours of occipital large foramen;
 9. At extramedullar tumours and cysts;
 10. At brain tumours;
 11. As a result of arachnoiditis.

The risk factors are:

1. Syringomyelia in family members.
2. The signs of dysraphic state (facial asymmetry, high palate, eye lids hypertrophy, short neck, shoulders asymmetry, acromehalia, enuresis, scoliosis, shortness).
3. Permanent physical exercises, trauma.

Clinical features and diagnostic criteria:

1. The first signs of the disease are:

- The loss of pain and temperature sensation on upper extremities (as a result of trauma or burns);
- Pains in the region of shoulders;
- Trophic disturbances (ulcers);
- Horner syndrome.

30 – 40 % of patients with syringomyelia have subclinical course of the disease and the diagnosis is usually put during the preventive examinations.

2. At neurologic examination there are:

- Sensory disorders – dissociative loss of pain and temperature sensation “coat like“ and half coat like”. In case of progress of the disease there are signs of loss of deep sensation.

- Motor disorders – segmental (distal one side or bilateral peripheral paresis of upper extremities, seldom – lower extremities), central (spastic mono and paraparesis of lower extremities). In case of pathologic focus in the medulla oblongata we can observe the signs of bulbar palsy.
- Autonomic – trophic disorders – acrocyanosis, hyper – anhidrosis, oedema of extremities, neuro – dystrophic lesions of extremities (in 70 % of all patients). There are defects of joints, neuro – osteopathy with the lesion of extremity function. (Fig. 81).



Fig. 81. Syringomyelia. Cervical- thoracic form

In case of clinical features of CSF disturbances at syringomyelia sometimes the signs of limbico – reticular complex are joined.

- AVD.
- Neuro – endocrinologic metabolic syndrome.

A d d i t i o n a l m e t h o d s o f e x a m i n a t i o n

1. X – ray of cranial cavity, cranio – vertebral part, spinal cord, joints. This method helps to differentiate the disease with inborn defects and secondary pathologic changes.

2. MRI – this method affords us to find out the type of syringomyelia and indications to the surgical treatment.

3. CT – this method doesn't give us too much information.

4. ENMG – affords us to register pyramidal signs.

5. Esthesiometry helps us to localize the expansion of sensory disorders.

Differential diagnosis

1. Intramedullar tumour (first of all those that are localized in the cervical part of the spinal cord). MRI data helps to put the right diagnosis.
2. Hematomyelia. Differential features are – acute beginning after the trauma, gradual regress of the main symptoms.
3. ALS at the beginning of the disease in case of anterior horn form. We should pay attention to the rapid progress and additional methods of examination data.
4. Cervical ischemic myelopathy at anterior horn form, Reino disease and angiopathy at autonomic form.
5. Compressive–ischemic mononeuropathy (at the beginning of the disease).

Prognosis

In case of slow progress and slight neurologic symptoms the patients preserve working ability up to 6 – 25 years. Rapid progress leads to the loss of working ability during 2 – 5 years. Rapid progress is much more common in persons involved in high physical activity in industry and agriculture.

Treatment

1. Conservative treatment.

- A. X – ray therapy – at gliosis forms. The method is based on the fact that X – ray decrease glia proliferation and the progress of the disease. It is used 5 times per week. The dose is – 90 Rad. For the course we use 900 – 1000 Rad. (9 – 11 Gr). The course is repeated in 1 – 1.5 year.
- B. Radioactive P and J. The mechanism of action is the same as in previous case. **J** is introduced twice a week in dose 50 – 100 MKuri (150 – 170 MKuri for the whole course. Usually 4 – 5 courses are used every 1 – 1.5 year. **P** is prescribed in dose 150 – 170 MKuri every 3 – 4 days up to the dose 500 – 450 MKuri. This course is repeated in 3 months.
- C. Symptomatic treatment – Proserinum, Nicotinic acid, vit B, non-steroid antiinflammatory medicines. At severe pain – analgethics, antidepressants and neuroleptics are used.
- D. Physiotherapy – massage, treating bathes. Hitting procedures are forbidden.

Brain tumors and spinal tumors

The incidence of brain tumors is 4 – 6 persons per 100 000 people. Brain tumors account for 6 % of malignances and in 1 – 1.5 % of all cases are the reason of patho – anatomical sections. Brain tumors also account 4 – 5 % of all organic diseases of the brain.

There are several classifications of brain tumors. The last one was proclaimed by World Health Organization in 1993. According to this **classification all the brain tumors** are divided into 10 groups.

- I. Epithelium tissue tumors:
 1. Astrocytomas;
 2. Oligodendrogliomas;
 3. Ependymomas, subependymomas;
 4. Mixed gliomas (oligoastrocytomas);
 5. Choroid plexus tumors;
 6. Neuroepithelium tumors of unknown origin (spongioblastoma, glioblastoma);
 7. Neuronal – gangliomas, gangliocytomas;
 8. Pineal region tumors (pinealoma, pineoblastoma);
 9. Embryonal tumors (meduloblastoma).
- II. Tumors of spinal and cranial nerves (neurinomas, neurofibromas).
- III. Tumors of the meninges.
- IV. Lymphomas and tumor of hematogenic tissue.
- V. Tumors from embryo cells (chorion – carcinoma, teratoma, embryo carcinoma).
- VI. Cysts and tumors – like processes (dermoid and epidermoid cyst).
- VII. Tumors of Turkish saddle region (adenoma of Hypophysis region, carcinoma of Hypophysis region).
- VIII. Tumors that are expanding from neighbour's territory.
- IX. Metastasis tumors.
- X. Non classified tumors.

Epithelium tissue tumors are the most common ones (50 – 60 % of all cases). Tumors of the meninges account about 20 % of all brain tumors.

Clinical classification of brain tumors

According to the cerebellar tentorium the tumors are divided into:

- Above tentorial (they are located in the base of anterior and middle cranial fossa).
- Under tentorial (these ones are located in the cerebellum region, IV–th ventricle and medulla oblongata).

According to the brain tissue the tumors are divided into

- Out of brain tumors (meningiomas and neurinomas can be treated in radical way).
- Intra brain tumors (gliomas).

The typical features of brain tumors:

1. **Astrocytoma** (15%) is a benign slowly progressive tumor located in frontal, temporal, parietal lobes or in brain stem.
2. **Oligodendroglioma** (8%) is a benign intra brain slowly progressive tumor. It is hardly differentiated from brain tissue. It has petrifactions that are seen at craniogram. It is localized in large hemispheres (fig. 82).
3. **Ependymoma** (3%) is usually found in the region of ventricles (the central canal of spinal cord and the IV – th ventricle).
4. **Glioblastoma** (15%) or **spongioblastoma** is a very malignant intra brain tumor located in temporal lobe or even sometimes in both hemispheres. It has a rapid growth with expressed intoxication and metastasis along CSF pathways.
5. **Meduloblastoma** (4%) is a very malignant tumor located in cerebellum, the IV–th ventricle or in brain stem in children at the age of 1 – 10 years. It gives rapid metastasis along CSF pathways.
6. **Angioreticuloma** (2%) – gemangioblastoma – is a benign slowly progressive tumor. It consists of multiple cysts of different sizes and is well separated from the brain tissue. It is often located in cerebellum (fig 83).
7. **Meningeoma** (15%) – is a benign out of brain tumor. There are basal and convexial meningiomas. Convexial ones are often located parasagittally and basal ones are mostly located in the region of Turkish saddle (fig. 84).
8. **Neurinoma** (8%) – is a benign tumor from the covers of cranial (VIII, V) and spinal nerves (fig. 85).

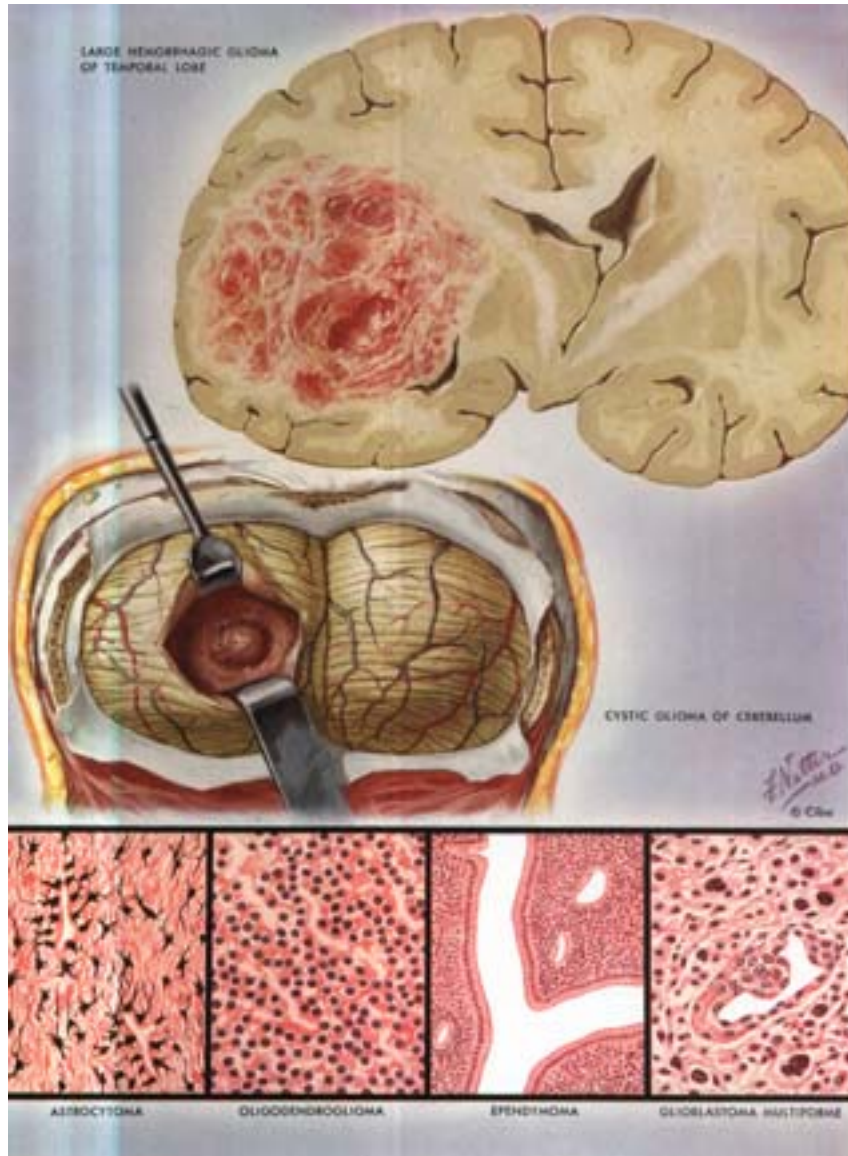


Fig. 82. Gliomata (Frank H. Netter, M.D.)



Fig. 83. Spinal cord tumors (Frank H. Netter, M.D.)

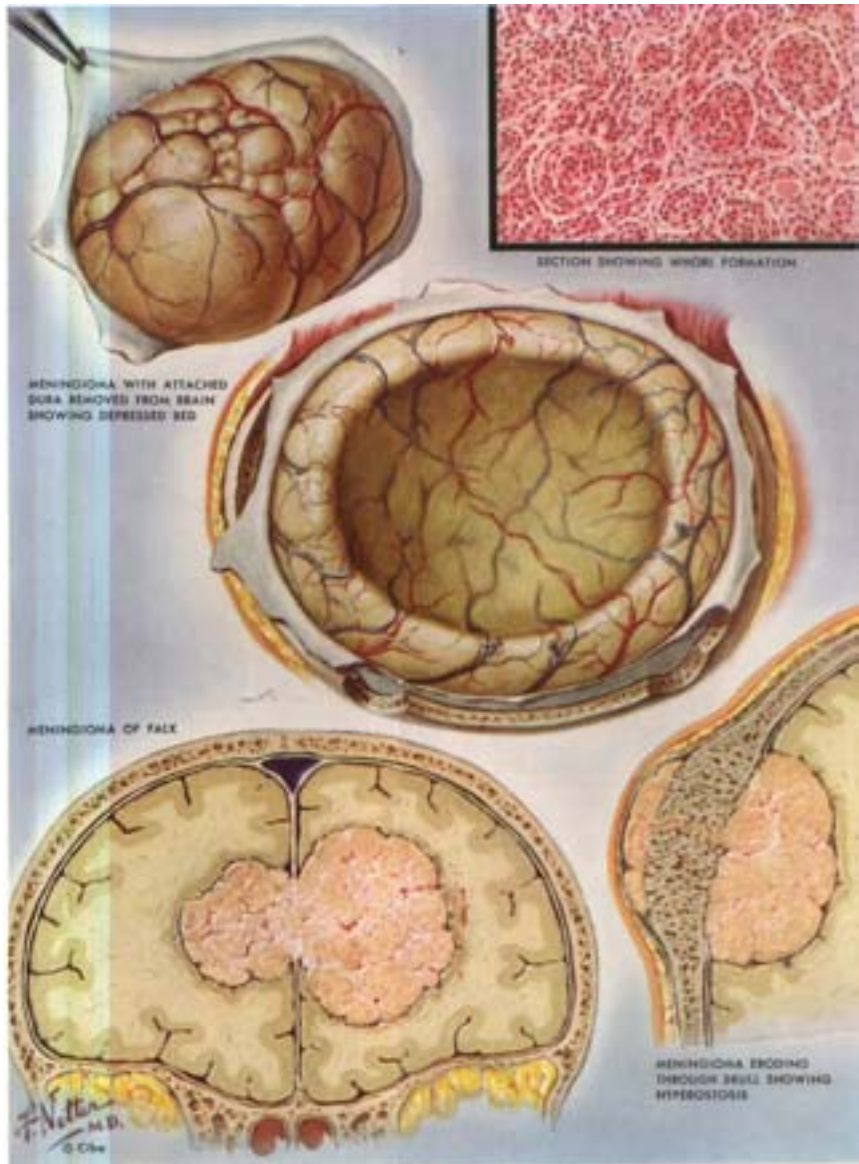


Fig. 84. Meningeomata (Frank H. Netter, M.D.)

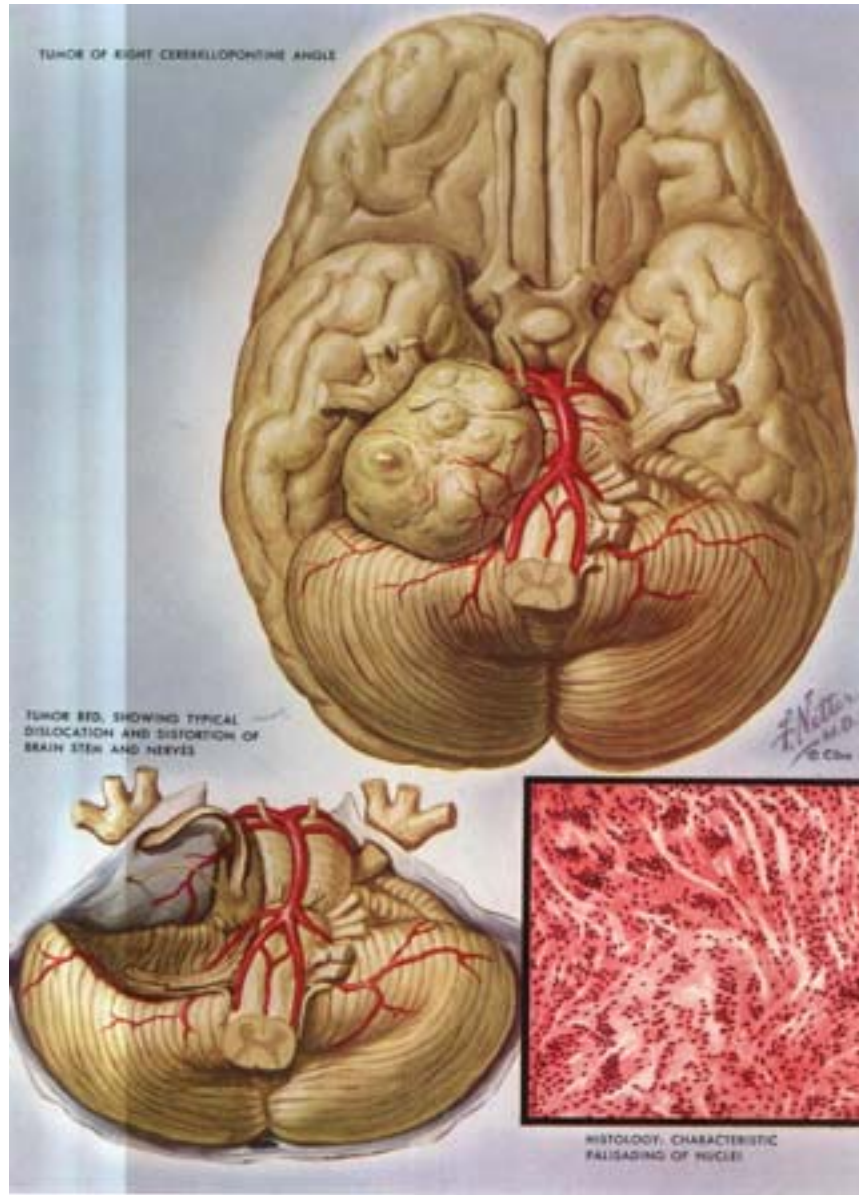


Fig. 85. Acoustic tumors (Frank H. Netter, M.D.)

9. **Pinealoma** is a benign tumor located in brain stem or posterior parts of ventricles'. It is characterized by secondary malignization.
10. **Adenoma of hypophysis** (10%) can be hormonally active or non active. There are adenomas with increased production of Prolactinum, Somatotropinum, ACTH, Tyriotropic hormone with typical clinical picture. Hormonally active tumors can expand into the neighbouring regions. Microadenomas in the Turkish saddle do not show clinical picture.
11. **Craniopharyngioma** is a tumor that grows from Ratke pocket. It is inborn benign one with great number of cysts and petrifications.
12. **Metastasis** (8%). Usually nervous system metastasis originates from lung, breast, kidneys, and stomach cancer. The main way of metastasis is haematogenous (fig. 86).

Etiology and pathogenesis Polietiological and dysontogenic theory.

Clinical picture

The clinical picture of brain tumors is associated with the following factors:

1. The direct influence of tumor on the brain tissue.
2. Increased intra cranial pressure.
3. Dislocation of different parts of the brain .

There are 4 **groups of clinical symptoms** in case of brain tumors:

- I. The symptoms associated with increased intra cranial pressure – **hypertension or general – cerebral symptoms.**
- II. **Focal symptoms** as a result of tumor's direct influence on brain tissue.
- III. **Neighbour symptoms** as a result of brain edema, blood circulation disturbances.
- IV. **Distance symptoms** as a result of increased intra cranial pressure and dislocation of some parts of the brain.

The clinical picture of tumor can consist of focal or general cerebral symptoms according to its location. If the tumor is far from the SCF pathways then focal symptoms dominate in clinical picture. If the tumor is close to the CSF pathways then the symptoms of hypertension dominate in clinical picture.

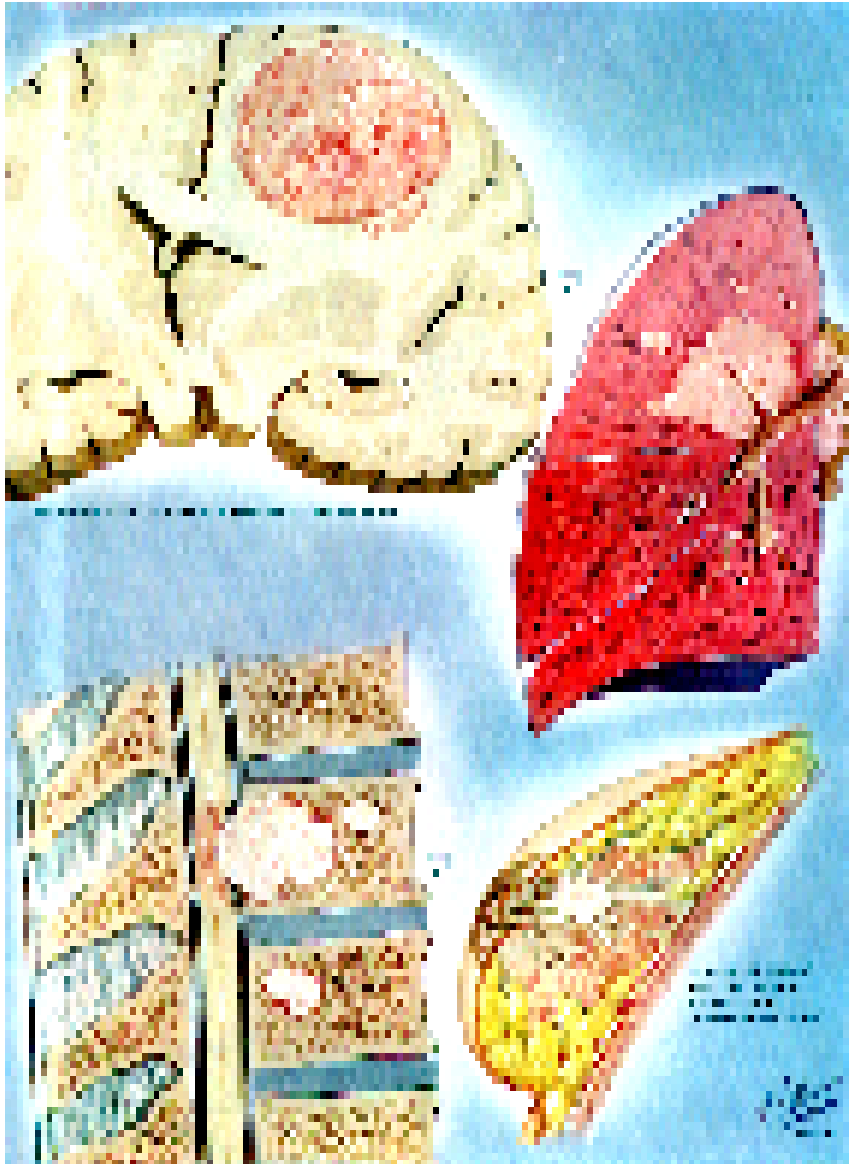


Fig. 86. Tumors metastatic to Brain and Spine (Frank H. Netter, M.D.)

I. General – cerebral (hypertensive) syndromes are the result of increased intra cranial pressure (fig. 87).

The main reasons of increased intra cranial pressure are:

- The tumor mass;
- Brain edema;
- Blood and CSF drain disturbances;
- CSF absorption disturbances.

Brain edema is developed as a result of:

- Mechanical influence of tumor;
- The influence of the substances that are created in course of tumor metabolism that leads to the additional production of CSF;
- Ischemia of brain tissue as a result of vessels pressure;
- Venous drain disturbances and blood stagnation.

Blood and CSF drain disturbances are observed when the tumor is located near large veins and venous collectors. The blood content in cranial cavity is increased. There is blood stagnation in cranial cavity. That causes increased secretion of CSF by choroids plexuses and leads to the increased intra cranial pressure. At the same time there are disturbances of CSF absorption that also leads to the increased intra cranial pressure.

The main clinical signs of intra cranial hypertension:

1. Headache;
2. Vomiting;
3. Choked disks of II CN;
4. Pulse, AP and breathing disturbances;
5. Epileptic attacks;
6. Psychiatric disorders;
7. CSF changes;
8. Craniogram changes.

The first three symptoms are most frequent but they are observed only in 50% of all patients.

1. Headache – is usually in the morning. It is puffed one. It is increased in case of coughing or physical exercises and it is decreased after vomiting. The main reason of headache is irritation of receptors as a result of increased intra cranial pressure. Head-

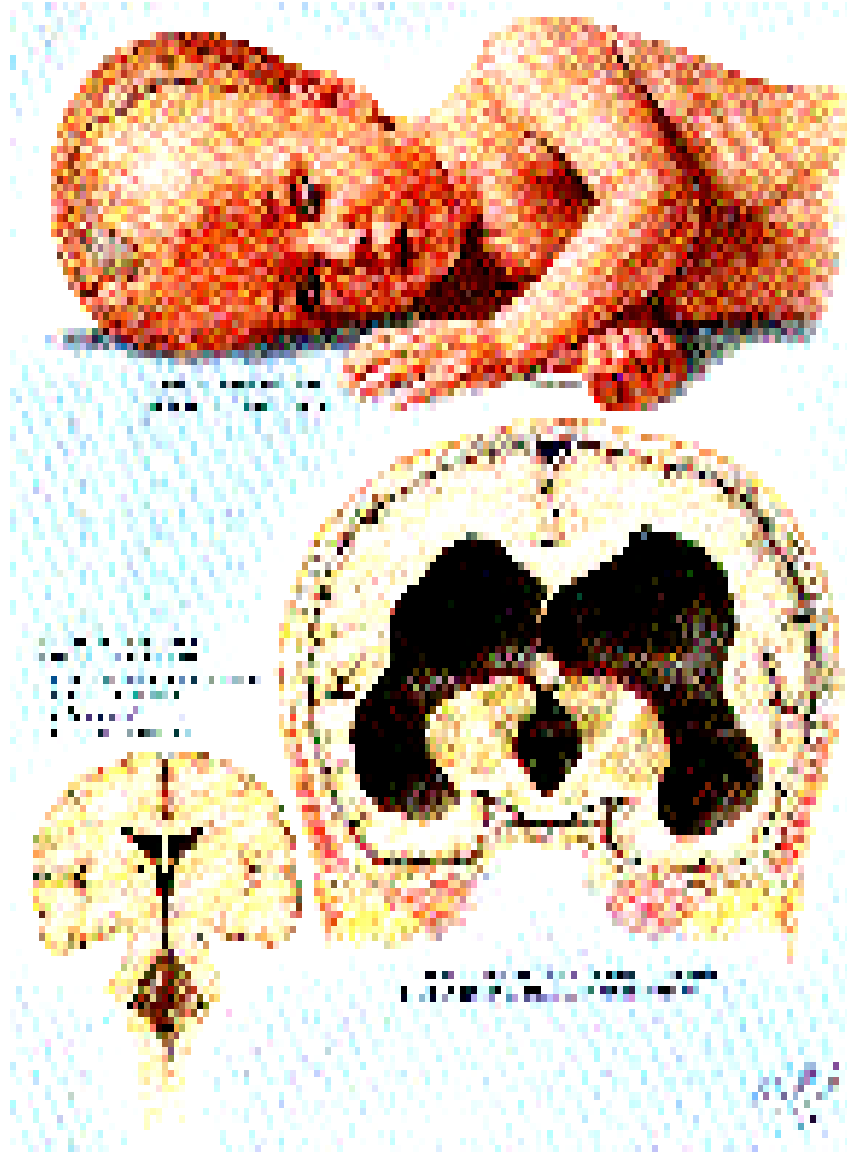


Fig. 87. Hydrocephalus (Frank H. Netter, M.D.)

ache is associated with painful trigeminal points, Bechterev's cheekbone phenomena.

2. **Vomiting** – is usually in case of severe headache and it is not associated with eating. The main reason of vomiting is irritation of Vagus nerve endings as a result of increased intra cranial pressure. It is a late symptom of tumor. Only in case of IV–th ventricle tumor it is an early symptom.
3. **Choked disks of optic nerve and visual disorders** – are associated with venous drain disturbances. Especially often this symptom is observed at tumors in posterior cranial fossa, cerebellar tumors, tumors of temporal lobe, IV–th ventricle. Choked disks of optic nerves lead to the low visual acuity and atrophy of optic nerve disks. There is the symptom of Foster – Kennedy atrophy of optic nerve disk on the side of tumor and choked disks on the opposite side.
4. **Pulse, AP, breathing disturbances** – are late symptoms of CSF hypertension. Pulse changes are due to the irritation of Vagus Nerve endings stimulation. Tachycardia is the result of brain stem pressure. Ahythmia and AP changes are the results of increased intra cranial pressure.
5. **Epileptic attacks** – are also the results of increased intra cranial pressure. Sometimes they are the first sign of cortical tumor.
6. **Psychiatric disorders** – are memory disturbances, loss of orientation, inhibition, sometimes even sopor and coma.
7. **CSF changes** – are increased CSF pressure, CSF is transient, with increased protein content at normal cytolysis (protein – cellular dissociation). The most expressed CSF changes are at basal tumors (basal meningiomas), neurinomas. CSF changes at convexital tumors are less significant.
8. **Craniogram changes** – are observed in a month after increased intra cranial pressure. They are osteoporosis of Turkish saddle, well expressed digital depressions, well expressed vessels picture, unclosed cranial sutures, osteoporosis of pyramids of temporal bone and edge of occipital foramen, increased skull sizes and thin skull bones.

II. Focal symptoms

Frontal lobe tumors:

1. **Frontal psychiatric disorders** – decreased attention, critics, be-

havioral changes, speech and motor depression, the loss of social and professional skills, untidiness, excitement, aggression, low mental activity, rude humor.

2. **In case of anterior central gyrus lesion** – there are central hemiparesis with domination in one extremity and facial paresis on the opposite side.
3. **In case of irritation of anterior central gyrus** there are epileptic attacks like “Motor Jackson “.
4. **In case of irritation of anterior adersive region** there are adersive attacks. They start with gaze and head turning into the opposite direction.
5. **Frontal ataxia** – which can later cause astasia – abasia. It is characterized by standing and walking disturbances with tendency to falling down into the opposite direction.
6. **Catching phenomena of Yanishevskyy.**
7. **Hypoosmia and anosmia** (at olfactory meningiomas).
8. **Motor aphasia** – when the process is localized in posterior part of lower frontal gyrus. Agraphy and gaze paresis is observed at tumor in posterior part of middle frontal gyrus.
9. **Foster – Kennedy symptom** – is the result of direct influence of tumor on optic nerve or increased intra cranial pressure.
10. **If the tumor is localized in paracentral lobe** there is lower paraparesis, urination central disorders.

Parietal lobe tumors

1. **In case of irritation of posterior central gyrus** there are sensory Jackson attacks with parasthesia sensation in one of the extremity.
2. **In case of posterior central gyrus lesion** there is loss of sensation according to the cortical monotype.
3. **The loss of deep sensation** in the extremities or afferent paresis of the opposite side extremity.
4. **Autotopognosia, anosognosia, pseudomelia, astereognosia.** (At right – side localization).
5. **Alexia, acalculia, agraphia.** (At left – side localization).
6. **Apraxia** (at tumors of gyrus supramarginalis).

Temporal lobe tumors

1. **Epileptic attacks** with auditory, olfactory, gustatory, visual and visceral hallucinations. The patients complain often on unpleasant internal sensations, depressions with time and space loss of orientation (especially when the tumor is localized between the temporal and parietal lobe).
2. **Olfactory, auditory, gustatory agnosia.**
3. **Homonimic hemianopsia** (when the tumor is deep in the posterior part).
4. **Sensory aphasia.**
5. **Amnesic aphasia.**
6. **Pseudocerebellar ataxia.**
7. **Vestibulo – cochlear dizziness.**
8. **Memory disorders on current events** at mediobasal parts lesion
9. **Psycho – motor automatisms.**

Those tumors very quickly lead to:

1. Dislocation symptoms and cutting – in;
2. Increased intra cranial pressure (headache, vomiting, choked disks of optic nerves);
3. The symptoms of III and V CN's lesion.

Occipital lobe tumors (They are very rare):

1. Simple visual hallucinations or photopsia (light, flash);
2. Homonimic hemianopsia (this symptoms appears at destruction of cortical neurons);
3. Colour vision disturbances (dyschromatopsia);
4. Visual agnosia;
5. Optic metamorphopsia;
6. Central gaze paralysis;
7. Quadrant hemianopsia;

General cerebral symptoms at occipital lobe tumors are very early.

Subcortical structures tumors:

1. They are located very close to the CSF pathways.
2. Hypertension and dislocation develop very early.

Clinical picture

1. Hyperkinesia.
2. Amyostatic syndrome with plastic hypertonus.
3. Hemiplegia, hemianesthesia, hemianopsia – capsule syndrome.

Thalamus tumors

There are hemianesthesia on the opposite side with burning – like unpleasant pains.

Corpus Callosum tumors

1. Psychiatric disorders with memory disturbances on current and past events.
2. Dementia.
3. Apraxia.
4. One or two – side paralysis.

Third ventricle tumors

1. Hormonal disorders (Non sugar diabetes, obesity).
2. The attacks of general weakness with muscle tone disorders.
3. Headaches with autonomic reactions.
4. Occlusive – hypertensive syndrome with memory disorders and psychiatric disturbances.

Chiasma and saddle tumors

Usually there are basal meningiomas, hypophysis adenomas, chiasmatic gliomas, optic nerve gliomas.

1. Endocrine disorders.
2. Primary atrophy of optic nerve disk.
3. Bitemporal hemianopsia.
4. III, IV, VI, V CN's lesion.
5. Foster – Kennedy syndrome.
6. Local hyperostosis or osteolysis in case of tumor's expansion into the base of the skull.

Posterior fossa tumors (tumors of cerebellum and IV – th ventricle and brain stem tumors).

In this case hypertension increases very quickly. There are radicular and nuclei lesions of CN's and alternating brain stem syndromes, bulbar syndrome and brain stem nystagmus.

At cerebellar tumors there are muscular hypotonia, coordination disorders, ataxia, enforced head position (the patient is lying on the tumor), occipital headache, asynergy, adiadochokinesis. There are early symptoms of increased intracranial pressure.

Fourth ventricle tumors (ependimomas)

These tumors are mainly developed in young persons. In elderly ones there is metastasis of bronchial cancer into the fourth ventricle. There are 3 main symptoms:

1. Headaches with irradiation into the neck and shoulder.
2. Morning vomiting because of IV – th ventricle irritation.
3. Dizziness and nystagmus.

All these symptoms are united into the **Bruns attacks** (severe headache, vomiting, skin autonomic reaction, breathing disorders, arhythmic pulse, enforced head position). The main reason is – acute occlusion of IV – th ventricle exit.

Sometimes it is associated with:

1. Neck's muscles tension.
2. Coordination disorders.
3. Tonic attacks, tachycardia, brain stem breathing disorders, hiccup.

Sudden death is very common in this case.

Brain stem tumors.

They are gliomas (benign tumors) and sarcomas (malignant tumors) or metastasis in brain stem.

1. At **midbrain tumors** there are peduncle alternating syndromes of Veber, Benedict.
2. At **pons tumors** there are alternating syndromes of Fovil, Miyar – Hubler.
3. At **medulla oblongate** there are syndromes of Jackson, Avelis, Shmidt, Valenberg – Zaharchenko.

Heart – vascular and breathing disorders are often joined.

The tumors of pontine – cerebellar angle (neurinoma of VII CN)

These tumors mainly appear at the age of 40 – 50 years. The typical signs are:

1. Slow growth.
2. Early focal signs.
3. Late increased cranial pressure.

The first signs of neurinoma are:

1. Progressive hearing loss (otiatric stage).
2. Dizziness and nystagmus.
3. The symptoms of V, VI, VII, VIII CN's lesion and cerebellar disorders (otoneurologic stage).
4. IX, X, XI, XII CN's lesion.
5. Enlargement of internal auditory pathway and osteoporosis of the top of temporal bone pyramid.

III. Neighbour regions syndromes.

These are developed because of the lesion of neighbouring regions, such as edema and blood circulation disturbances. These symptoms can regress while usage of dehydration therapy. Sometimes when the tumor is localized in the so-called "mute parts of the brain" these symptoms can manifest as focal ones that complicates the diagnosis of tumor's localization.

IV. Distance syndromes (dislocation and cutting – in)

The most common are temporal tentorial and axial occipital cutting – in.

The reasons and mechanisms of cutting – in:

Cranial cavity is divided by cerebellar tentorium and falx cerebri into several parts (right and left hemisphere, above and under tentorial parts). In case of tumor in one of these parts the pressure in cranial cavity is increased and brain mass is dislocated in the direction where the pressure is lower. This leads to the dislocation of other parts of the brain and cutting – in of brain stem, corpus callosum.

At temporo – tentorial cutting – in gyrus parahippocampalis is cutted – in Bisha hole. That clinically manifests as brain stem syndromes with downwards dynamics. At first the symptoms of brain peduncles appear, then pons and oblongate brain are involved, later decerebrative rigidity and disorders of consciousness, heart – vascular and breathing activity are joined.

At axial occipital cutting – in there is cutting – in of cerebellum into the foramen occipital magnum. That clinically manifests as occipital headache, neck – stiffness, enforced head position, later bulbar disturbances and cyanosis, breathing and heart – vascular disorders are joined. That often leads to death.

Tumors diagnostics

1. Neurological examination.
2. Ophthalmological examination with eye ground, visual fields and visual acuity examination.
3. Otoneurological examination (hearing, taste, smell and vestibular sensation).
4. Craniogram in two projections. If necessary it can be associated with additional pictures in special positions. On X-ray examination can be found the signs of intracranial hypertension, local hyperostosis, local bone defects, enlargement of physiological holes.
5. Echo-EG finds dislocation of middle structures or wideness of ventricles at hemisphere localization of the tumor.
6. CT and MRI give us information about the tumor's localization and its sizes.
7. Lumbar puncture is used rarely as it can cause cutting – in syndrome.
8. EEG finds the focus of pathologic activity.
9. PET finds the changes of blood circulation and metabolism in the region of brain where the tumor is localized.

Treatment

1. Surgical.
2. Surgical with next radiation therapy.
3. Medicines.

In case of most tumors the treatment is **surgical**. Before the surgery it is necessary to take into consideration all the indications and contraindications. During the surgery we can do complete radical extraction of tumor or partial extraction of tumor. Sometimes we use palliative surgery in order to improve CSF drainage. In case of cutting of cerebellum in order to save patient's life surgeon makes a hole with side ventricle's horn puncture. This decreases intra cranial pressure and compensates patient's state.

Recently bone – plastic skull trepanation is used. Almost all extra brain tumors are extracted without brain incision. Hypophysis tumors can be extracted through the nose, small neurinomas can be extracted through the labyrinth of temporal bone. Partially internal brain tumors are extracted.

Palliative operations are used when the direct operation is impossible. To palliative operations belong decompressive skull trepanation, perforation of IV– th ventricle ground, drainage.

Radiation therapy When it is established that the tumor is sensitive to radiation therapy, surgical treatment is associated with radiation therapy. It is made in two ways:

1. Radio-therapeutic treatment is made by means of distant radiation therapy – gamma – therapy, X – ray therapy and proton radiation (at the tumors in the base of the skull, brain stem, gliomas, medulloblastomas, and metastasis).
2. Radio – surgical treatment is made by means of radio – pharmaceutical medication introduction. This method is used at endocrine tumors – hypophysis adenomas, craniopharyngiomas, and tumors in the base of the skull.

Medicines are used mostly symptomatically in order to:

- Decrease intra cranial pressure and brain edema (diuretics, steroids);
- Anti-seizures medications;
- Nootrops (in postoperative period);
- Analgesics;
- Chemotherapy.

Spinal tumors

They are less common than brain tumors. They take about 12% of all tumors of central nervous system. They are usually diagnosed at the age of 30 to 50 years.

Classification

According to the growth of the tumors they are classified into 2 groups:

I – extramedullar (out of spinal cord. To them belong neurinomas, meningiomas).

II – intramedullar (within the spinal cord. To them belong ependimomas, astrocytomas, oligodendrogliomas).

The correlation between extra – and intra – medullar tumors is 1:4. Extra– medullar tumors amount about 80% and intra– medullar tumors about 20% of all spinal tumors. The most common are tumors of thoracic part of spinal cord (66%), there are also cervical (18%) and lumbar (15%) tumors.

There are primary and secondary (metastasis) spinal tumors. Metastasis usually originates from breast, esophagus, thyroid gland and prostate cancer.

Clinical features

Extra – medullar tumors (fig. 88)

There are 3 stages:

1. Radix – associated – not well expressed.
2. Stage of Brown – Sequard syndrome.
3. Stage of complete spinal cord diameter lesion.

Intra – medullar tumors (fig. 89).

There are 3 stages:

1. Segmental (segmental – dissociative disorders).
2. Stage of complete spinal cord diameter lesion.
3. Radix – associated.

At extra – medullar tumors superficial sensory disorders are slowly moving upwards, while at intra – medullar tumors these disorders are slowly moving downwards.

In order to localize the tumor we should pay attention at:

- The symptom of spinal process (pain in case of hammer percussion).
- Radix– associated pain.

Diagnosis

1. Lumbar puncture with CSF – dynamic tests and CSF – examination.
2. X– ray of spinal cord. There are such symptoms:
 - Widened between vertebra spaces.
 - Osteoporosis of the base of vertebra’s arches radices and widened central canal (the symptom of Elsberg – Dayke).
 - Destruction of vertebra’s bodies.
3. CT and MRI.
4. Myelography with contrast usage (omnopak, dimmer, conrey) gives us possibility to refine the presence of tumor and its level.

Treatment.

It is only surgical. Extra – medullar tumors and some of intra – medullar tumors (ependimomas, astrocytomas) are extracted radically.

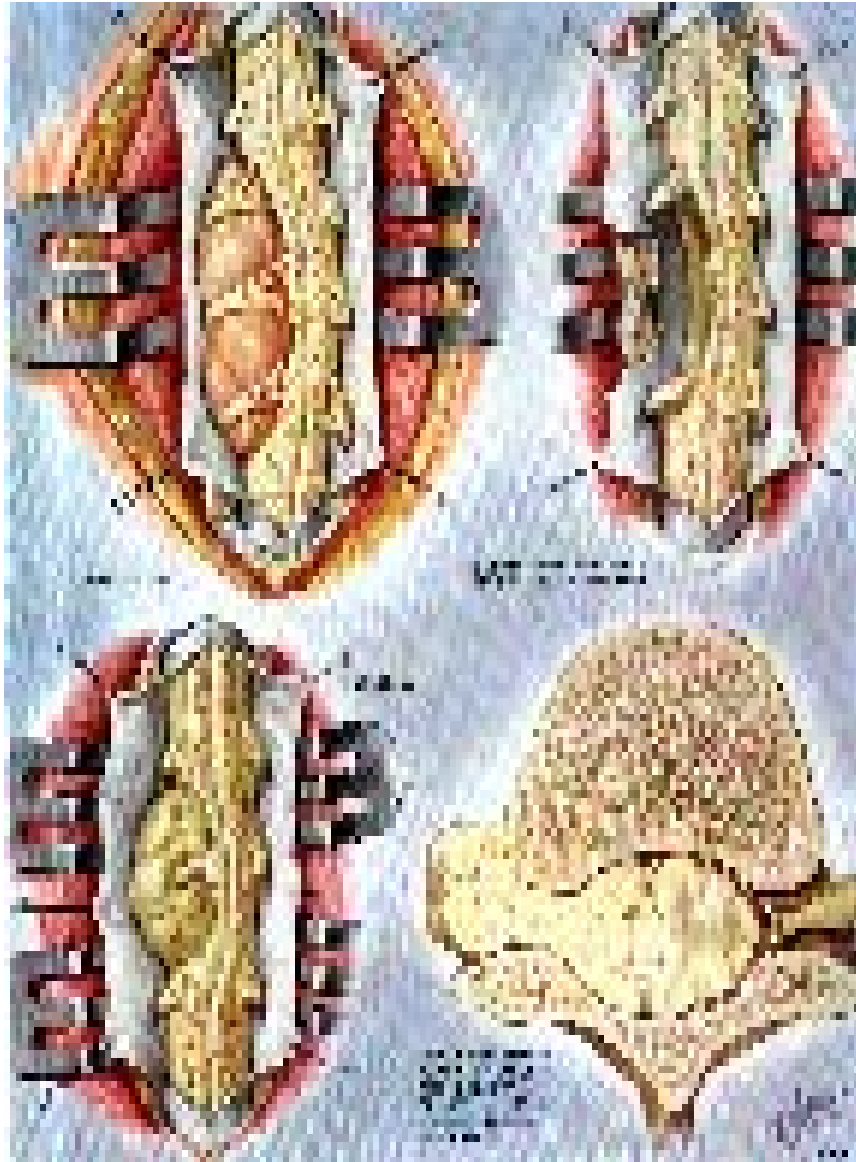


Fig. 88. Spinal cord tumors, extramedullary (Frank H. Netter, M.D.)

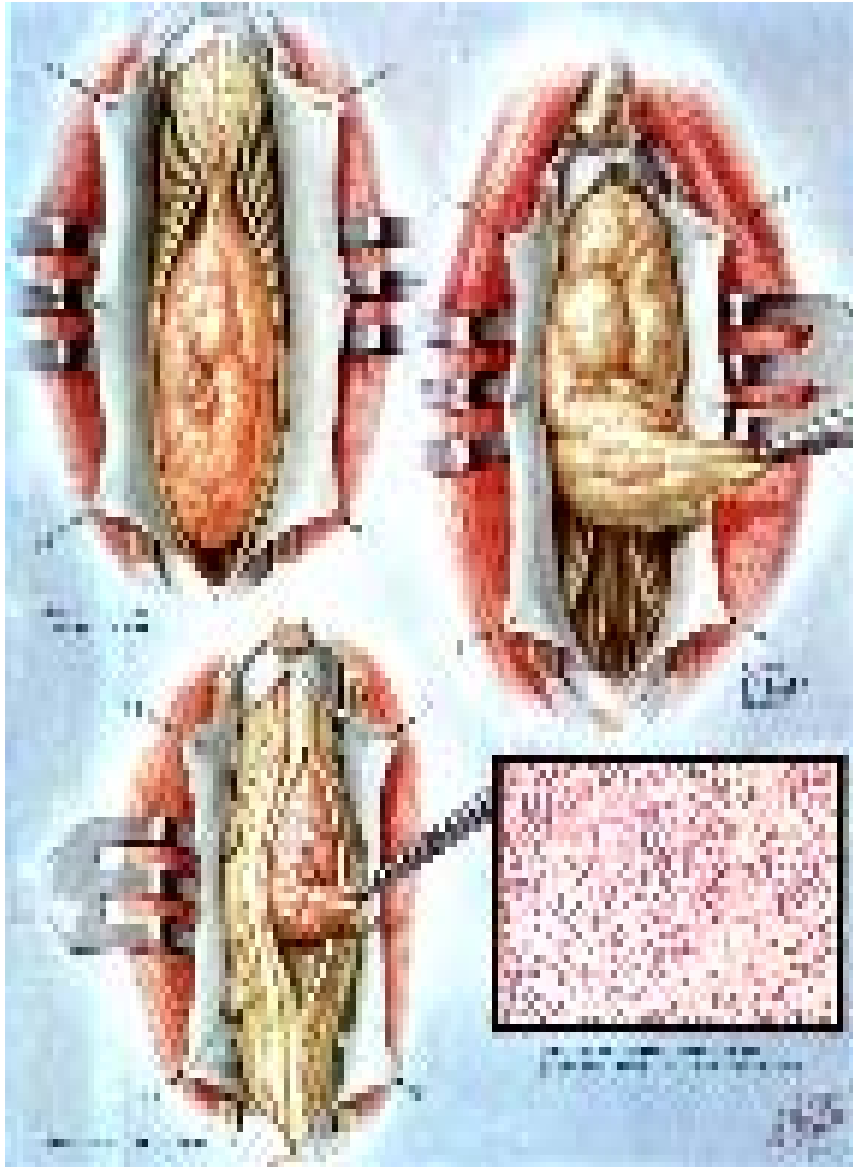


Fig. 89. Spinal cord tumors, intramedullary (Frank H. Netter, M.D.)

Epilepsy and convulsion syndromes

Epilepsy is a chronic disorder, which is characterized by the presence of:

- Epileptic focus.
- Recurrent attacks with various clinical signs.
- Personality disorders between attacks.
- Some specific paraclinical signs.

Epidemiology

The prevalence of epilepsy is 0.8 – 1.2%. About 40 million people are affected worldwide. Among them 6 million people live in Europe. In men the incidence of epilepsy is 1.5 times higher than in women. The disease begins at the age under 10 – in 31 %; 20 – in 29 %; 30 – in 30 %; 40 – in 14 %; elder – in 6 %. About 200 billions of Euro are spent for the treatment of epilepsy in Europe.

Epilepsy as a disease should be differentiated from:

- Epileptic reaction;
- Epileptic syndrome.

Epileptic reaction – is the response of the brain to the strong external and internal damaging factors (such as electro – shock, insulin shock, brain hypoxia, severe alcohol intoxication and so on). The main clinical features of epileptic reaction are abortive seizures or general tonic – clonic seizures.

Epileptic syndrome – is characterized by recurrent epileptic attacks on the background of pathologic focus in brain. The attacks are variable and depend on the localization of focus. Focal symptoms are obligatory in this case.

Risk factors:

1. Inheritance.
2. Organic brain diseases:
 - Prenatal (infections – cytomegalovirus, rubella, toxoplasmosis, toxicosis of pregnancy, diet disturbances);
 - Perinatal (physical trauma, child birth anoxia, metabolic disorders, neonatal infection);
 - Postnatal (infections, trauma, dehydration, toxins);

3. Disorders of brain function. Sleep disorders.
4. Paroxysmal states in childhood:
 - Newborns seizures;
 - Febrile seizures;
 - Affective – respiratory seizures.

Pathology

There are such changes:

- The results of organic diseases.
- The results of epileptic process.

Each epileptic attack causes hypoxic changes in brain and leads to the development of encephalopathy.

Pathologic physiology

1. There is a group of neurons with pathologic activity, which is called **epileptic focus**.
2. There is the ability to enforce and spread the activity.
3. Weakness of anti – epileptic protection. (It is provided by caudal parts of the brain).

Neurochemistry of epilepsy

1. Disorders of balance between glutamate (exciting neurotransmitter), GABA (inhibitory neurotransmitter) that leads to desynchronization.
2. Disorders of K / Na pump.

Immunology of epilepsy

There is increasing content of anti–brain antibodies. The primary attack causes disturbances of HEB. Immune system butts into strange to it nervous system. The result is production of antibodies and CIC that are fixed in brain tissue and favour its damage in new places.

Classification of epileptic attacks

I. Partial epileptic:

1. Simple:
 - a. Simple motor:
 - Focal motor without march;
 - Focal motor with march;

- Adversive;
 - Postural;
 - Phonatory simple.
- b. Simple sensory:
 - Somatosensory (with and without march);
 - Visual, acoustical, gustatory, smell);
 - c. Simple autonomic – visceral.
 - d. Simple with psychiatric disorders:
 - Aphatic;
 - Dymnestic;
 - With thinking disturbances (ideatory);
 - Emotional – affective;
 - Hallucinatory.
2. Complex:
 - a. Temporal pseudoabsence;
 - b. Automatisms.
 3. Simple with generalization (secondary general).

II. General attacks:

1. Absence:
 - Typical;
 - Atypical.
2. Myoclonic.
3. Tonic – clonic.
4. Tonic.
5. Clonic.
6. General atonic.

III. Non classified.

IV. Epileptic status.

Clinical features

I. A. General seizures.

Epileptic general tonic – clonic attack (grandmal) usually begins with short initial stage that lasts several seconds. The last can manifest as:

- Bilateral general muscle jerks;
- Loss of consciousness;
- Autonomic changes;
- Enlargement of pupils;

Karlov called this stage initial one.

The tonic stage lasts about 10 – 20 seconds. During this stage seizures involve all the muscles. Usually seizures dominate in the extensors but at the beginning flexors are even more involved. The eyes are opened and are looking upwards. Mouth is opened too. Seizures start from axial muscles and then involve extremities. Shoulders are lifted and adducted. Muscles of lower extremities are seldom involved. Opisthotonus and “obstetrician arm” are often observed. Extension of great toe is a common symptom in this case. Because of the diaphragm muscles contraction epileptic shouting is often associated with the attack.

Then tonic stage is converted in clonic one. That means that on the background of tonic muscles straining there is trembling of the muscles. Then between the muscles straining relax pause appear. This stage is often associated with tongue biting, clonic vocalization. This phase lasts 30–40 seconds. During the tonic–clonic attack there are severe autonomic disorders – apnoe, cyanosis, small skin hemorrhages, pulsation of carotic arteries. Pulsation is frequent, AP is increased. There is also midriasis with pupils areflexia, hyper-salivation that usually manifest as bloody foam. After attack the next stage can be divided into early and late ones.

Early stage lasts 1 – 5 min and is characterized by: after the last clonic attack a new phase of tonic contraction appears. The last is very similar to the one at the beginning. But the seizure dominates in the face, especially in chewing muscles and causes trismus. Usually the extremities aren't involved. The eyes are looking upwards. There is midriasis. This stage is finished by muscles atonia that leads to involuntary urination. Corneal reflexes are absent. Deep reflexes are increased. There is loss of consciousness. And that means that the patient is in coma.

Late after attack (recovery) stage is characterized by decreasing of midriasis, normal superficial reflexes, decreased deep reflexes and Babinski sign. The behavior of the patient is often automatic. When the patients are conscious they can complain on headache, muscles pain and complete amnesia. This period lasts 5–15 min.

There can be different types of general tonic – clonic attack because of the age of the patient. In children tonic stage can last longer

than clonic one. In newborn babies there is often difference between right and left hemisphere seizures. Sometimes in children general tonic – clonic attacks are associated with vomiting and feces incontinence.

Tonic attacks are seldom in grown – ups. In children tonic attacks are often associated with atypical absentia epileptica.

There are three types of epileptic tonic attacks:

1. Axial – body and facial muscles are involved in attack. There is spasm of respiratory muscles and breathing stop at expiration.
2. The same signs plus less involvement of extremities muscles.
3. Global means involvement of body and extremities muscles in the same way.

These attacks are associated with loss of consciousness. There is midriasis, tachycardia, increased AB and so on.

General tonic – clonic attacks are very dangerous for the patient. They can cause trauma, aggression and sudden death as a result of autonomic disorders and respiratory disturbances, acute suprarenal insufficiency.

Clonic epileptic attacks

General typical clonic attacks are often observed in newborn babies. There is loss of consciousness, autonomic disorders, rhythmic clonic seizures. Between the attacks of clonic muscles jerks there is muscle hypotonia.

If the attacks last 1–2 min the consciousness recovers quickly. But these attacks can last 4–5 min and even more. Then after the attack coma can be developed.

B. Without seizure attacks

Absentia epileptica are characterized by sudden and short – lasting (2–30 sec) loss of consciousness and EEG – peculiarities. They are associated with absent gaze, interruption of patient's activity, autonomic disorders (paleness or hyperemia of face, midriasis). The attack is finished suddenly. The patient doesn't remember anything about it. This picture is typical for simple attack, when motor activity, seizures or loss of muscles tonus are absent.

If absentia is associated with any motor component it is called complex absentia.

Complex absentia can be divided into myoclonic, atonic, tonic

and with automatisms.

Myoclonic absentia is characterized by loss of consciousness, rhythmic bilateral myoclonus in face and upper extremities. There are jerks of eyelids, periorbital muscles, mouth edges, eye bulbs. The patient can lose some objects he is holding in his arms.

Atonic absentia is characterized by decreasing of postural tonus, hanging head and sudden drops.

Tonic absentia is associated with looking of eyes upwards. There is domination of either extensor or flexor component, symmetric or asymmetric.

Absentia with automatism can be the sign of focal attack and absentia. The main condition for automatism is incomplete loss of consciousness. Differential diagnosis in this case is very complicated. That's why EEG should be made for such patients.

Typical absentia are associated with bilateral symmetric complexes "top – waves" with frequency 3 per sec in frontal – central lobe.

Typical absentia are much more common in children and can be caused by hyperventilation or light. They are very refractory to the antiepileptic drugs.

Atypical absentia are associated with such EEG changes:

1. Bilateral symmetric complex of "top – wave" which are rhythmically repeated with frequency 2 per sec.
2. Epileptic rhythm of gathering with frequency 10 per sec.
3. Epileptic rhythm of gathering with frequency 20 per sec.
4. Complex of multiple spikes – waves with frequency 4 – 6 per sec.

Atypical absentia are resistant to hyperventilation and paroxysmal light.

II. Focal attacks

Focal attacks are those that clinically and on EEG manifest as the beginning of activation of neuron system of certain part of brain hemisphere.

There are three groups of focal attacks:

1. Simple focal.
2. Complex focal.
3. Focal attacks with secondary generalization.

The main differential feature of complex focal attacks from simple

ones is loss of consciousness.

Focal attacks are characterized by different symptoms – motor, sensory, autonomic or psychiatric that depends on focus localization and the peculiarities of morpho–functional organization of epileptic system. Motor attacks are caused by discharges in certain part of motor cortex. Somato–motor or motor Jackson attacks are seizures in certain muscle group according to the focus localization. They can be local or involve other group of muscles according to the topical localization in the brain cortex.

In case of epileptic discharges in motor speech center speech disorders or involuntary vocalization – involuntary repetition of words are observed.

Sensory attacks manifest as simple or complex sensory disorders, such as somatosensory, visual, acoustical, olfactory, taste attacks and epileptic attacks of dizziness.

Somatosensory Jackson attacks are associated with numbness, tingling in some part of the body. They can be localized or involve other parts of the body. They are caused by epileptic discharges in Post – Rolandic region. Very often the attack begins as somatosensory and then converts into somatomotor.

As for visual, acoustical, olfactory, taste attacks and epileptic attacks of dizziness they can manifest as simple disorders or complex illusion or hallucinations.

Treatment

The main principals of epilepsy treatment should be – emergency, accordance to stages, following.

On the way to hospital:

1. To release breathing air ways.
2. Digitalis drugs.
3. Sibazonum 0.01g.

In ambulance:

- a) Tracheobronchial tree drain:
 1. Sibazonum 30 ml in 150 ml of physiological solution, in 10 min we add the medication up to 100 – 120 mg;
 2. Magnesiii sulfas 25% 10.0 in glucose 40 %;
 3. Anesthesia with nitrous oxide;
 4. Dosed anesthesia;

5. Aminazinum 25% 1-2 ml;
6. Atropinum 0.1% 1.0 s/c;
7. Cardiac, antihistamine, diuretics;
8. Natrii tiopentali 1g in 10 ml of physiological solution.

Epileptic status

- To provide permeability of respiratory airways.
- To evaluate the function of heart – vascular and respiratory systems.
- To provide free way to veins.
- Lorazepam 4mg i/v or Diazepam 10 mg.

In the hospital

- To take blood for analysis.
- % of urea, electrolytes.
- liver function.
- % of glucose.
- % of blood gas.
- etiology of attack: hypoglycemia – 50% solution of glucose 50 mg
– at alcohol abuse – Tiaminum

Next half an hour:

- to introduce the medication through the naso – gastral probe.
- Fenitoin 18 mg per kg or Phenobarbitalum 15 mg per kg i/v by drop 100 mg per kg.

In 30 minutes:

- General anesthesia.
- EEG.

Surgical methods of treatment:

1. Resections

- Anterior temporal lobectomy;
- Selective amygdalohypocampoectomy;
- Calosotomia;
- Hemispherectomy.

2. Stereotaxic

- Destruction of deep temporal structures. This procedure on lateral part normally decreases seizures, on medial ones – aggression.

3. **Radio – surgical with gamma – knife. Gamma** – waves from 201 sources are focused on certain aims. The effectiveness of this procedure is 70 – 80 %.

4. **Electrostimulative** – stimulation of certain structures:

- Nucleus dentatus, the caput of nucleus caudatus (the price is 3.5 – 3 000 \$)
- Stimulation of n. vagus – this method is one of the newest one. It is indicated at partial seizures with secondary generalization.

Hereditary diseases of nervous system

All the hereditary diseases of nervous system are divided into:

1. Diseases with **involvement of nervous – muscle synapse**:
 - A. Myasthenia;
 - B. Myasthenic syndromes.
2. Hereditary diseases with **involvement of pyramidal system**:
 - A. Spastic paraplegia of Shtrumpel;
 - B. Family spastic paralysis with amyotrophy, oligophrenia, retina degeneration (described by Kellin);
 - C. Family spastic paralysis with ichthyosis and oligophrenia.
3. Diseases with **involvement of extrapyramidal system**:
 - A. Hepato – cerebral degeneration;
 - B. Dystonia;
 - C. Double athetosis;
 - D. Huntington disease;
 - E. Parkinson disease;
 - F. Myoclonus – epilepsy;
 - G. Tourette syndrome;
 - H. Hereditary trembling;
 - I. Rulph seizure;
 - J. Palpebral – mandibular synkinesis.
4. **Hereditary ataxia**:
 - A. Spinal ataxia of Fridreich;
 - B. Hereditary cerebellar ataxia of Pier – Mary;
 - C. Olivo – ponto – cerebellar degeneration;
 - D. Refsum disease;
 - E. Rusi – Levina disease;
 - F. Marinesku – Shagrena disease;
 - G. Lichtenshtein – Knorr disease.
5. Diseases with **involvement of neuro – muscular junction**:
 - I. Progressive muscular dystrophies:
 - A. Dushenne muscular dystrophy;
 - B. Late Becker muscular dystrophy;
 - C. Family visceral myopathy;
 - D. Facioscapulohumeral muscular dystrophy Landuzi – Degerina;
 - F. Scapulo peroneal form of Davidenkova;

- G. Erba dystrophy;
- H. Ophthalmoplegic myopathy;
- I. Distal myopathy of Wallender;
- J. Inborn nonprogressive forms of myopathy.
- II. Amyotrophy as a result of peripheral neuron lesion:
 - A. Spinal amyotrophy of Werding – Hoffman;
 - B. Proximal amyotrophy of Kukelberg – Welander;
 - C. Kennedy amyotrophy;
 - D. Sharkot – Marie – Tooth disease;
 - E. Hypertrophic neuritis.
- 6. Family – hereditary **myotonia**:
 - A. Myotonia Thomsena (Myotonia Congenita);
 - B. Atonic myotonia;
 - C. Paramyotonia of Alenburg;
 - D. Chondro – dystrophic myotonia.
- 7. Hereditary **diseases with paroxysmal states**:
 - A. Paroxysmal family myoplegia;
 - B. Episodic hereditary adynamia;
 - C. Mac – Ardel disease;
 - D. Episodic myotonic adynamia of Bekker.

Myasthenia

Myasthenia gravis (MG) is caused by a defect of neuromuscular transmission due to an antibody-mediated attack on nicotinic acetylcholine receptors (AChR) at neuromuscular junctions. It is characterized by fluctuating weakness that is improved by inhibitors of cholinesterase.

Incidence MG is a common disease. An apparent increase in the incidence of the disease in recent years is probably due to improved diagnosis. According to Phillips and Torner (1996), the prevalence rate is 14 per 100,000 (or about 17,000 cases) in the United States. Before age 40 years, the disease is three times more common in women, but at older ages both sexes are equally affected.

Etiology

How the autoimmune disorder starts is not known. In human disease, in contrast to experimental MG in animals, the thymus gland is almost always abnormal; there are often multiple lymphoid follicles with germinal centers (“hyperplasia of the thymus”), and in about

15% of patients, there is an encapsulated benign tumor, a thymoma. These abnormalities are impressive because the normal thymus is responsible for the maturation of T-cells that mediate immune protection without promoting autoimmune responses.

There are few familial cases of the disease, but disproportionate frequency of some HLA haplotypes (B8, DR3, DQB1) in MG patients suggests that genetic predisposition may be important. Other autoimmune diseases also seem to occur with disproportionate frequency in patients with MG, especially hyperthyroidism and other thyroid disorders, systemic lupus erythematosus, rheumatoid arthritis, pernicious anemia, and pemphigus.

Pathogenesis

The polyclonal IgG antibodies to AChR are produced by plasma cells in peripheral lymphoid organs, bone marrow, and thymus. These cells are derived from B cells that have been activated by antigen-specific T-helper (CD4+) cells. The T-cells have also been activated. In this case by binding to AChR antigenic peptide sequences (epitopes) that rest within the histocompatibility antigens on the surface of antigen-presenting cells.

The AChR antibodies react with multiple determinants, and enough antibody circulates to saturate up to 80% of all AChR sites on muscle. A small percentage of the anti-AChR molecules interfere directly with the binding of ACh, but the major damage to endplates seems to result from actual loss of receptors due to complement-mediated lysis of the membrane and to acceleration of normal degradative processes (internalization, endocytosis, lysosomal hydrolysis) with inadequate replacement by new synthesis. As a consequence of the loss of AChR and the erosion and simplification of the endplates, the amplitude of miniature endplate potentials is about 20% of normal, and patients are abnormally sensitive to the competitive antagonist curare. The characteristic decremental response to repetitive stimulation of the motor nerve reflects failure of endplate potentials to reach threshold so that progressively fewer fibers respond to arrival of a nerve impulse.

Most AChR antibodies are directed against antigenic determinants other than the ACh binding site. Nevertheless, the summed effects of the polyclonal anti-AChR antibodies with differing modes of action result in destruction of the receptors. Physiologic studies indicate impaired postsynaptic responsiveness to ACh, which

accounts for the physiologic abnormalities, clinical symptoms, and beneficial effects of drugs that inhibit acetylcholinesterase.

Clinical features

The typical sign of myasthenia is muscles weakness. One of the most specific features of this weakness is its increasing with movements. The disease develops at the age of 20 – 30 years old. Before age 40 years, the disease is three times more common in women, but at older ages both sexes are equally affected. The disease is developed subacutely or chronically in most cases.

According to the course of the disease there are such forms:

1. Progressive;
2. Stationary;
3. Myasthenic episodes.

Clinical forms:

1. Ophthalmic.
2. Bulbar.
3. Skeletal.
4. General.

Ophthalmic

The most common signs are ptosis, diplopia and eyes movement disturbances (fig. 90).

Typical features:

1. Asymmetric lesion.
2. Dynamic symptoms (the signs increase in the evening).

Ophthalmoplegia is a very common symptom.

The other ones are:

- Weakness of mimic muscles – especially oral muscles.
- Weakness of chewing



Fig. 90. Myasthenia. Ophthalmic form

muscles.

- Weakness of pharyngeal, laryngeal muscles and muscles of tongue.
- Tongue muscles function disorders.
 - Breathing disturbances.
 - Extremities function disturbances (especially proximal parts).
 - Neck muscles weakness – hanging of the head.
- Body muscles weakness that leads to duck – like gait.
 - Sensory and pelvic disorders usually are not observed.

Diagnosis

1. Complains on general weakness that increases in the evening.
2. Early symmetric lesion of external eyes muscles.

Tests for this disease revealing

The patient is asked to look upwards or inside during 30 seconds in order to cause ptosis. He is asked to read text aloud in order to cause dysarthria. The patient is asked to make 100 chewing movements in order to reveal the weakness of these muscles.

Proserine test. Proserine is introduced in dose 1.5 – 3 ml s/c, sometimes Atropinum is used in order to prevent side effects. In 20 – 40 min all the signs of myasthenia disappear. In 2 – 3 hours all the symptoms appear again.

EMG – myasthenic reaction. Test is positive in 85% of patients with skeletal form.

Muscle biopsy – muscle atrophy and signs of degeneration.

CT reveals timoma signs. In 90% of all patients antibodies to ACHR are found.

Differential diagnosis:

- Botulism.
- Neurasthenia.
- ALS.
- Polineuropathy.
- Muscles dystrophy.
- Inflammatory myopathy.
- MS.
- Stroke in v/b region.
- Brain stem tumor.

Treatment

1. Compensation of neuro – muscular transference.
2. Thymus influence.
3. Correction of immune disorders.
1. **Anticholinesterase medicines** –
Caliminum – 30 mg 3 times per day.
Proserinum – 0.5 – 1.5 mg s/c.
2. **K drugs** 3 – 4 g per day.
3. **Corticoids** – we start from 15–20 mg a day, than increase gradually on 5 mg every 3 day.
4. **Anabolics** – Retabolil 50 mg once every 3 days, 5 – 6 injections.
5. **Immune suppressors** – Asatioprinum in dose 50 – 150 – 200 mg per day.
6. **Plasmapheresis** – at acute and progressive form.
7. **Radiation therapy of thymus.**
8. **Methabolic drugs.**

At myasthenia crisis:

- Plasmapheresis;
- Ig i/v (2 g per kg 2 – 5 days);
- Corticoids (100 mg prednisonum);
- Proserinum 1 – 2 ml i/v;
- SLV, oxygen;
- Halloperidolum at excitation.

Cholinergic crisis

There fasciculations, seizures, bradycardia, salivation, hyperhydrosis and abdominal pain.

Treatment – Atropinum 1 ml 0.1 % s/c or i/v.

Parkinsonism

Parkinsonism – is a chronic progressive neurodegenerative syndrome that is characterized by motor disorders as a result of extrapyramidal system involvement.

Parkinson disease (PD) – is a chronic progressive degenerative disease of CNS that manifest as voluntary movements disorders. PD was described by James Parkinson for the first time in 1817 as shaking paralysis.

Epidemiology

The prevalence of PD is 133 per 100 000 people. This disease is considered to be one of the most common ones among old people after dementia, epilepsy, cerebral vascular diseases. The beginning of the disease is at the age of 55. The most common factors that lead to the disease are – old age, inheritance, toxic agents.

Old age Every 10 years the person loses 8% of all neurons. But the symptoms of PD manifest only when 80% of all neurons will be lost.

Inheritance The inheritance of the disease can be proved by:

- Association of PD with dementia;
- Rapid progress of the disease.

Toxins There are a lot of toxic substances that can provoke PD.

The other reasons of PD development are:

- Viral infections;
- Cerebral vessels sclerosis;
- Severe cranial trauma;
- Long lasting usage of neuroleptics, reserpinum medicines.

Pathogenesis

The core biochemical pathology in parkinsonism is decreased dopaminergic neurotransmission in the basal ganglia. Degeneration of the nigrostriatal dopamine system results in marked loss of striatal dopamine content. Drug-induced parkinsonism is the result of blockade of dopamine receptors or depletion of dopamine storage. It is not known how hydrocephalus or abnormal calcium metabolism produces parkinsonism. Physiologically, the decreased dopaminergic activity in the striatum leads to disinhibition of the subthalamic nucleus and the medial globus pallidus, which is the predominant efferent nucleus in the basal ganglia. Understanding the biochemical pathology led to dopamine replacement therapy; understanding the physiologic change led to surgical interventions, such as pallidotomy, thalamotomy, and subthalamic nucleus stimulation.

Clinical features

The main signs of PD are:

1. Hypokinesia;
2. Rigidity;

3. Resting trembling;s
4. Loss of postural reflexes.

The clinical features of tremor, rigidity, and flexed posture are referred to as *positive phenomena* as reviewed first; bradykinesia, loss of postural reflexes, and freezing are negative phenomena. In general, the negative phenomena are the more disabling. *Rest tremor* at a frequency of 4 to 5 Hz is present in the extremities, almost always distally; the classic "pill-rolling" tremor involves the thumb and forearms. Rest tremor disappears with action but reemerges as the limbs maintain a posture. Rest tremor is also common in the lips, chin, and tongue. Rest tremor of the hands increases with walking and may be an early sign when others are not yet present. Stress worsens the tremor.

Rigidity is an increase of muscle tone that is elicited when the examiner moves the patient's limbs, neck or trunk. This increased resistance to passive movement is equal in all directions and usually is met by a ratchety "give" during the movement. This so-called cogwheeling is caused by the underlying tremor even in the absence of visible tremor. Cogwheeling also occurs in patients with essential tremor. Rigidity of the passive limb increases while another limb is engaged in voluntary active movement.

The *flexed posture* commonly begins in the arms and spreads to involve the entire body. The head is bowed, the trunk is bent forward, the back is kyphotic, the arms are held in front of the body, and the elbows, hips, and knees are flexed. Deformities of the hands include ulnar deviation of the hands, flexion of the metacarpal-phalangeal joints, and extension of the interphalangeal joints (striatal hand). Inversion of the feet is apparent, and the big toes may be dorsiflexed (striatal toe). Lateral tilting of the trunk is common.

Akinesia is a term used interchangeably with bradykinesia and hypokinesia. *Bradykinesia* (slowness of movement, difficulty initiating movement, and loss of automatic movement) and *hypokinesia* (reduction in amplitude of movement, particularly with repetitive movements, so-called decrementing) are the common features of parkinsonism, although they may appear after the tremor. Bradykinesia has many facets, depending on the affected body parts. The face loses spontaneous expression (masked facie: *hypomimia*) with decreased frequency of blinking. Poverty of spontaneous movement is characterized by loss of gesturing and by the patient's tendency to sit

motionless. Speech becomes soft (*hypophonia*, and the voice has a monotonous tone with a lack of inflection (*aprosody*). Some patients do not enunciate clearly (*dysarthria*) and do not separate syllables clearly, thus running the words together (*tachyphemia*). Bradykinesia of the dominant hand results in small and slow handwriting (*micrographia*) and in difficulty shaving, brushing teeth, combing hair, buttoning, or applying makeup. Playing instruments is impaired. Walking is slow, with a shortened stride length and a tendency to shuffle; swing decreases and eventually is lost. Difficulty rising from a deep chair, getting out of automobiles and turning in bed are symptoms of truncal bradykinesia. Drooling saliva results from failure to swallow spontaneously, a feature of bradykinesia, and is not caused by excessive production of saliva. The patients can swallow properly when asked to do so, but only constant reminders allow them to keep swallowing. Similarly, arm swing can be normal if the patient voluntarily and, with effort, wishes to have the arms swing on walking. Pronounced bradykinesia prevents a patient with parkinsonism from driving an automobile; foot movement from the accelerator to the brake pedal is too slow.

Bradykinesia is commonly misinterpreted by patients as weakness. Fatigue, a common complaint in parkinsonism, particularly in the mild stage of the disease before pronounced slowness appears, may be related to mild bradykinesia or rigidity. Subtle signs of bradykinesia can be detected even in the early stage of parkinsonism if one examines for slowness in shrugging the shoulders, lack of gesturing, decreased arm swing, and decrementing amplitude of rapid successive movements. With advancing bradykinesia, slowness and difficulty in the execution of activities of daily living increase. A meal normally consumed in 20 minutes may be only half eaten in an hour or more. Swallowing may become impaired with advancing disease, and choking and aspiration are concerns.

Loss of postural reflexes leads to falling and eventually to inability to stand unassisted. Postural reflexes are tested by *the pull-test*, which is performed by the examiner, who stands behind the patient, gives a sudden firm pull on the shoulders, and checks for retropulsion. With advance warning, a normal person can recover within one step. The examiner should always be prepared to catch the patient when this test is conducted; otherwise, a person who has lost postural

reflexes could fall. As postural reflexes are impaired, the patient collapses into the chair on attempting to sit down (*sitting en bloc*). Walking is marked by festination, whereby the patient walks faster and faster, trying to move the feet forward to be under the flexed body's center of gravity and thus prevent falling.

The freezing phenomenon (motor block) is transient inability to perform active movements. It most often affects the legs when walking but also can involve eyelid opening (known as *apraxia of lid opening* or levator inhibition), speaking (palilalia), and writing. Freezing occurs suddenly and is transient, lasting usually no more than several seconds with each occurrence. The feet seem as if "glued to the ground" and then suddenly become "unstuck," allowing the patient to walk again. Freezing typically occurs when the patient begins to walk ("start-hesitation"), attempts to turn while walking, approaches a destination, such as a chair in which to sit (destination-hesitation), and is fearful about inability to deal with perceived barriers or time-restricted activities, such as entering revolving doors, elevator doors that may close, and crossing heavily trafficked streets (sudden transient freezing). Freezing is often overcome by visual clues, such as having the patient step over objects, and is much less frequent when the patient is going up steps than when walking on a level ground. The combination of freezing and loss of postural reflexes is particularly devastating. When the feet suddenly stop moving forward, the patient falls because the upper part of the body continues in motion as a result of the inability to recover an upright posture. Falling is responsible for the high incidence of hip fractures in parkinsonian patients. Likely related to the freezing phenomenon is the difficulty for parkinsonian patients to perform two motor acts simultaneously.

The main clinical forms:

1. Trembling.
2. Rigid.
3. Mixed.

Severity stages:

I – loss of activity, but that doesn't influence on professional activity and working ability.

II – moderate loss of professional activity.

III – the patients need someone to look after him.

Treatment

Treatment of parkinsonism in general is based on the treatment of PD. At present, treatment is aimed at controlling symptoms because no drug or surgical approach unequivocally prevents progression of the disease. Treatment is individualized because each patient a unique set of symptoms, signs, response to medications, and a host of social, occupational, and emotional needs that must be considered. The goal is to keep the patient functioning independently long as possible. Practical guides are the symptoms and degree of functional impairment and the expected benefits and risks of therapeutic agents.

Drug Therapy

Although pharmacotherapy is the basis of treatment, physiotherapy is also important. It involves patients in their own care, promotes exercise, keeps muscles active, and preserves mobility. This approach is especially beneficial as parkinsonism advances because many patients tend to remain sitting and inactive. Psychiatric assistance may be required to deal with depression and the social and familial problems that may develop with this chronic disabling illness. Electroconvulsive therapy may have a role in patients with severe intractable depression.

Selection of the most suitable drugs for the individual patient and deciding when to use them in the course of the disease are challenges for the treating clinician. In many Parkinson-plus disorders, the response to treatment is not satisfactory, but the principles for treating PD are used in treating these disorders as well. Because PD is chronic and progressive, treatment is lifelong. Medications and their doses change with time as adverse effects and new symptoms are encountered. Tactical strategy is based on the severity of symptoms.

Carbidopa is listed as the peripheral dopa decarboxylase inhibitor, but in many countries *benserazide* is also available. These agents potentiate the effects of levodopa, thus allowing about a fourfold reduction in dosage to obtain the same benefit. Moreover, by preventing the formation of peripheral dopamine, which can act at the area postrema (vomiting center), they block the development of nausea and vomiting. *Domperidone* is a dopamine receptor antagonist that does not enter the central nervous system; it is used

to prevent nausea, not only from levodopa but also from dopamine agonists. Domperidone is not available in the United States. Of the listed dopamine agonists, only bromocriptine, pergolide, pramipexole and ropinirole are available in the United States; they are reviewed in a following section. Because it is water soluble, apomorphine is used as an injectable, in countries where available, as a rapidly acting dopaminergic to overcome "off states. Cabergoline has the longest half-life. Catecholamine-O-methyl transferase inhibitors extend the elimination half-life of levodopa.

Amantadine, selegiline, and the anticholinergics are reviewed in following sections. Because the anticholinergics can cause forgetfulness and even psychosis, they should be used cautiously in patients most susceptible (those older than 70 years). The antihistaminics, tricyclics, and cyclobenzaprine have milder anticholinergic properties that make them useful in PD, particularly in the older patient who should not take the stronger anticholinergics.

Antidepressants are needed for treating depression. Because of its anticholinergic and soporific effects, amitriptyline can be useful for these properties as well as for its antidepressant effect. The serotonin uptake inhibitors are also effective in treating depression of PD but may aggravate parkinsonism if antiparkinsonian drugs are not given concurrently. Diazepam is usually well tolerated without worsening parkinsonism and can help to lessen tremor by reducing the reaction to stress that worsens tremor.

Cisapride can help to overcome constipation, a common complaint in patients with PD or the Shy-Drager syndrome. Clozapine, a selective dopamine D4 receptor antagonist, can ameliorate levodopa-induced psychosis without worsening parkinsonism, but weekly monitoring of white blood cells is necessary to prevent irreversible agranulocytosis. The drug is discontinued if the white blood cell count declines. Quetiapine, a related drug, does not need hematologic monitoring and therefore can be tried first to overcome psychosis.

Surgery

The surgical approaches are not considered in the early stages of PD but are reserved for patients who have failed to respond satisfactorily to drugs. *Thalamotomy and thalamic stimulation* (target

for both is the ventral intermediate nucleus) are best for contralateral intractable tremor. Tremor can be relieved in at least 70% of cases. Although a unilateral lesion carries a small risk, bilateral operations result in dysarthria in 15% to 20% of patients. Thalamic stimulation seems to be safer and can be equally effective against tremor, but it runs the risks associated with foreign bodies and thin electronic wires that can break. *Pallidotomy* (target is the posterolateral part of the globus pallidus interna) is most effective for treating contralateral dopa-induced dystonia and chorea but also has some benefit for bradykinesia and tremor. The target in the globus pallidus interna is believed to be the site of afferent excitatory glutamatergic fibers coming from the subthalamic nucleus, which is overactive in PD. *Lesions of the subthalamic nucleus*, although effective in relieving parkinsonism in animal models, are hazardous in humans because hemichorea or hemiballism may result. Instead, *stimulation of the subthalamic nucleus* is used and appears to be the most promising in reducing contralateral bradykinesia and tremor. Subthalamic nucleus stimulation in a patient appears to reduce symptoms of PD that respond to levodopa in that patient. It is not effective against symptoms that do not respond to levodopa. This type of surgery often allows a marked reduction of levodopa dosage, thereby reducing dopa-induced dyskinesias and treating parkinsonian symptoms. *Fetal dopaminergic tissue implants* are being investigated. This surgical procedure may reduce bradykinesia and rigidity in younger patients but is less effective in those over age 60; it is not effective against tremor. Its long-term effect is not established, but in some patients it has replaced bradykinesia with persistent dyskinesia in the absence of levodopa. Until this problem can be solved, transplantation surgery is not a useful option.

Levodopa is uniformly accepted as the most effective drug available for symptomatic relief of PD. If it were uniformly and persistently successful and also free of complications, new strategies for other treatment would not be needed. Unfortunately, 75% of patients have serious complications after 5 years of levodopa therapy.

Hereditary diseases with involvement of Pyramidal system Spastic paraplegia of Shtrumpel

This disease is the result of pyramidal tracts and cerebellar

connections degeneration.

The disease is genetically recessive in most cases but in some families it show dominant inheritance.

Clinical features

The first signs of the disease are observed at the age of 10–15. The typical signs of the disease are lower spastic paraplegia with increased muscle tonus, high stretch reflexes, pathological reflexes. Usually the lesion of lower extremities is symmetrical. Sometimes motor disorders can be developed in upper extremities. In some cases pseudobulbar symptoms are joined.

The typical signs of the disease:

- The dominance of spastic tonus over motor disorders;
- Well preserved abdominal reflexes;
- The absence of pelvic disorders.

The typical clinical picture of spastic paraplegia is often associated with cerebellar symptoms and symptoms of posterior spinal columns. The progress of the disease is slow.

Recessive form of spastic paraplegia differs by early beginning and much more severe course of the disease. In some families this disease is observed in men only.

Differential diagnosis:

- LAS;
- Multiple sclerosis;
- Vascular myelopathy;
- Cerebral palsy.

Hereditary diseases with involvement of Extrapyramidal system

Hepatocerebral dystrophy (HCD)(Wilson – Konovalov disease)

This disease is connected with disorders of ceruloplasminum metabolism. Ceruloplasminum is a blood protein responsible for Cu transport. It is produced in liver. Pathologically there is accommodation of Cu in subcortical ganglions (especially n. Lenticularis), brain cortex, cerebellum, liver, spleen, iris.

The disease is genetically autosomal – recessive. And it is

observed in male and female with the same frequency.

Clinical signs of the disease

The first signs of the disease are observed in early childhood. There are neck stiffness, different hyperkinesis and psychiatric changes. Sometimes seizures can be observed. There is also liver enlargement. One of the most specific changes is Kaizer – Fleishner ring in the iris.

According to the Konovalov classification there are 4 main neurological types of the disease:

1. Rigid – arhythmokinetic;
2. Trembling – rigid;
3. Trembling;
4. Extrapyramidal – cortical.

Sometimes the disease manifests only as liver insufficiency and neurological signs are joined later.

Diagnosis

- Family history.
- The typical signs of the disease – Kaizer – Fleishner ring, lesion of liver, low quantity of ceruloplasminum in the blood, increased quantity of Cu in urine.

Differential diagnosis:

- Huntington disease;
- MS;
- Chronic stage of epidemic encephalitis.

Torsion dystonia

The pathology of the disease includes degenerative changes of subcortical ganglions, subthalamic nuclei and n. Dentatus of cerebellum as a result of neuromediators production and metabolism disturbances.

Hyperkinetic form of the disease has autosomal – dominant type of inheritance. Rigid form of the disease is characterized by autosomal – recessive type of inheritance.

Clinical features of the disease

The disease begins in early childhood and it is characterized by permanent progression. The typical signs of the disease are hyperkinesis that increases with every movement. The hyperkinesis

may have a look of tonic body and extremities muscle straining. Spastic torticollis is usually one of the earliest symptoms of the disease. There are no mental disorders in typical cases. There are generalized form of the disease and local ones, such as spastic torticollis and chirospasm.

Diagnosis Family history and the evaluation of pathological process dynamics are necessary for the diagnosis putting.

Differential diagnosis

- Atypical form of Economo encephalitis.

Huntington disease

It is a progressive hereditary disorder that usually appears in adult life. It is the result of systemic degeneration of extrapyramidal structures and brain cortex.

It has autosomal – dominant type of inheritance.

Clinical features of the disease

The disease usually appears in adult life and it is very rare in children. Male and female can suffer from this disease.

The main clinical symptoms of classic form are:

- Choreic movements;
- Extrapyramidal rigidity;
- Slowly progressive dementia.

Rare forms are:

- Akinetic – rigid syndrome;
- Extrapyramidal immobility in children;
- Epileptic attacks;
- Myoclonia.

Diagnosis:

1. Clinical and genetic analysis.
2. CT and MRI of brain (atrophic changes of brain hemispheres)
3. EEG.
4. DNA – analysis.

Differential diagnosis:

1. Chorea.
2. Hepato – cerebral degeneration.

Double atetosis

The typical sign of the disease are involuntary movements in face, body and extremities muscles.

The disease has autosomal – dominant type of inheritance. Both – male and female suffer from this disease.

Clinical picture of the disease

The typical sign of the disease are slow worm – like movements in fingers and toes. Usually the lesion is bilateral. But sometimes hemiatetosis can be observed. Spasmus molibilis is also one of the typical sign of the disease. The signs of the disease are developed usually just after birth and are preserved during the whole life of the patient.

Diagnosis:

- Peculiarities of clinical picture.
- Family anamnesis.

Differential diagnosis:

- Hepato-cerebral dystrophy.
- Huntington disease.

Myoclonus – epilepsy

It is one of the most rare hereditary diseases. The pathology of the disease includes specific inclusions in patient's liver and brain.

The disease is inherited according to autosomal – recessive type.

Clinical features of the disease

The disease begins in childhood and progresses constantly. Most of the patients die from the associated diseases and increasing dystrophy.

There 4 main symptoms of the disease:

1. General epileptic attacks;
2. Myoclonic hyperkinesis;
3. Extrapyramidal rigidity;
4. Psychiatric degradation.

Differential diagnosis:

- Chronic stage of epidemic encephalitis.

Generalized Tourette tic

The disease is characterized by local face, larynx muscles. That causes gait disorders and complex movements. Sometimes mental disorders can be observed.

The genetic base of the disease is still being studied.

Clinical features

The disease develops in childhood and progresses gradually.

Diagnosis:

- Anamnesis;
- The results of complex neurological examination;
- Emotional state evaluation;
- Dynamic changes of clinical changes.

Differential diagnosis:

- Torsion dystonia.
- Huntington disease.

Essential tremor of Minor (hereditary trembling)

The most earliest symptom is hands' trembling, which is preserved while resting and increases at emotional stress. Sometimes head trembling is associated. There are no other neurological signs of the disease. Sometimes there are some of extrapyramidal disorders – rigidity, gait disorders.

The disease is inherited according to the autosomal – dominant type.

Clinical features

The disease begins at the age of 50. Men suffer much more often than women from this disease.

Differential diagnosis:

- Parkinson disease;
- Huntington disease;
- Tirotoxic tremor.

Hereditary ataxias

Spinal Friedreich ataxia

The disease is characterized by spinal cord degeneration and degenerative – dystrophic changes in posterior and lateral columns.

The disease is characterized by autosomal – recessive type of inheritance.

Clinical features of the disease

The disease begins at the age of 10 – 12 and then slowly progresses. The main clinical signs are sensitive – cerebellar ataxia, nystagmus, muscle hypotonia and areflexia, gait disorders. At the beginning of the disease there is deep sensation disorders according to the conductive type on lower extremities. In the course of the disease coordination disorders, scan speech, body and upper extremities ataxia appear. The disease is characterized by some bone abnormalities, cardiomyopathy, mental disorders and the symptoms of lesion of pyramidal tracts.

Differential diagnosis:

- MS.
- Neurosyphilis.

Hereditary cerebellar ataxia of Pier – Mary

The main signs of the disease are:

- The beginning at the age of 30 – 50;
- Cerebellar ataxia;
- Dysarthria;
- Hyperreflexia;
- Spastic muscle hypertonia.

The inheritance of the disease is autosomal – dominant.

Clinical feature of the disease

The disease begins gradually with gait disorders, disorders of coordination, nystagmus, dysarthria. There are high reflexes, increased muscle tonus according to spastic type (mainly in lower extremities), pathologic reflexes. Eye movements disorders are often observed in such patients. There are mental, memory and emotional disorders.

The course of the disease is progressive.

Differential diagnosis:

- Olivo-ponto-cerebellar degeneration;
- MS.

Olivo-ponto-cerebellar degeneration

It is the group of the diseases that are connected by system lesion of cerebellar cortex, pons and lower olives. Sometimes the neurons of anterior horns of the spinal cord and basal ganglia are involved. The inheritance of the disease is autosomal – dominant.

Clinical features of the disease

The disease begins at the age of 15 – 20, sometimes 30 years. Cerebellar symptoms dominate in clinical features. There are also extrapyramidal and pyramidal symptoms, peripheral polyneuropathy. Sometimes retina is involved in pathological process. Mental disorders are often observed.

There are several types of olivopontocerebellar degeneration:

- Olivopontocerebellar degeneration of Mentsel. The disease begins at the age of 20 – 25. There are cerebellar ataxia, bulbar disorders, extremities paresis, high reflexes and pathological reflexes.
- Olivopontocerebellar degeneration of Degerina – Tomas. It develops at the age of 7 – 12. There are cerebellar disorders, distal hyperesthesia, areflexia, ptosis, convergence disturbances.
- Olivopontocerebellar degeneration of Holms. It develops at the age of 20 – 25. It is characterized by cerebellar ataxia, dementia, ophthalmoplegia.
- Olivopontocerebellar degeneration with macular degeneration. It develops at the age of 20 – 25 with progressive decreasing of visual acuity, central scotoma. In several years ataxia, intention, choreic hyperkinesia and spastic lower paraparesis are developed.

Differential diagnosis

- Ataxia of Friedreich.

Diseases with involvement of neuro – muscular junction:

a. Progressive muscular dystrophy (Myopathies)

Myopathies are conditions in which the symptoms are due to dysfunction of muscle with progressive weakness, impaired relaxation

(myotonia), cramps or contracture (in McArdle disease), or myoglobinuria. Dystrophies are myopathies with four special characteristics:

1. They are inherited.
2. All symptoms are due to weakness.
3. The weakness is progressive.
4. There are no abnormalities in muscle other than degeneration and regeneration, or the reaction to these changes in muscle fibers (infiltration by fat and connective tissue), and there is no storage of abnormal metabolic products.

Etiology

These inherited diseases are separated from each other on the basis of clinical and genetic criteria, but the inherited biochemical abnormality has not been identified in any of them. Therefore, the pathogenesis of these disorders is not known.

Pathology

In the majority of the cases, the significant pathological findings are confined to the muscles. There may be a few degenerative changes in the ventral horn cells or a slight reduction in their number, but as a rule the peripheral and central nervous systems are normal. In the early stages of the disease, the muscle fibers are rounded and enlarged to more than twice their normal size.

With progress of the disease, there is a longitudinal splitting of some of these large fibers with resulting admixture of fibers of various sizes. This splitting of the fibers is accompanied by hyaline degeneration of the myoplasma, evidence of regeneration, an increase in the number of sarcolemmal nuclei and replacement of the muscle substance by fat and connective tissue.

Classification

They divide the cases into such groups:

- a. Duchenne pseudo – hypertrophic muscular dystrophy;
- b. Late Becker muscular dystrophy;
- c. Facioscapulohumeral muscular dystrophy Landuzi – Degerina;
- d. Erba dystrophy.

Clinical features of all myopathies. Symptoms

The symptoms are essentially those due to the muscular weakness.

In the majority of cases the proximal musculature is more severely affected than that of the distal parts of the extremities. The child is clumsy in walking and has difficulty in climbing up and down stairs. Toe walking is a common early symptom. The weakness of the shoulder proximal muscles makes it difficult for the child to raise the arms over the head or lift heavy objects. The weakness of the pelvic proximal muscles gives rise to the characteristic waddling gait and attempts to turn result in much commotion but little progression because the knees cannot be raised properly. The boy may fall frequently and then has trouble rising without assistance.

Signs. Cardiac failure as the result of involvement of the heart muscles has been reported in a few cases. In the majority of cases, the dystrophy is limited to the muscles of the trunk and extremities. The gait and posture are characteristic. There is an advanced degree of lumbar lordosis as a result of weakness of the trunk muscles. There is a stoppage, waddling gait. Movements of the arm may be accompanied by winging of the scapula. Weakness of the shoulder proximal muscles causes the child to slip through the hands when

attempts are made to lift him by placing the hands in the axillar (fig. 91).

Another characteristic and diagnostic feature of the disease is the manner in which the patient rises from the supine to the erect position (Gower's sign). The patient first turns over onto the abdomen and raises the trunk to the crawling position. He then places the feet firmly on the floor with the aid of his arms and gradually elevates the upper part of the body by "climbing up his own trunk" with the arms (fig. 92). With progression of the weakness, the patient is able to rise from the floor only by

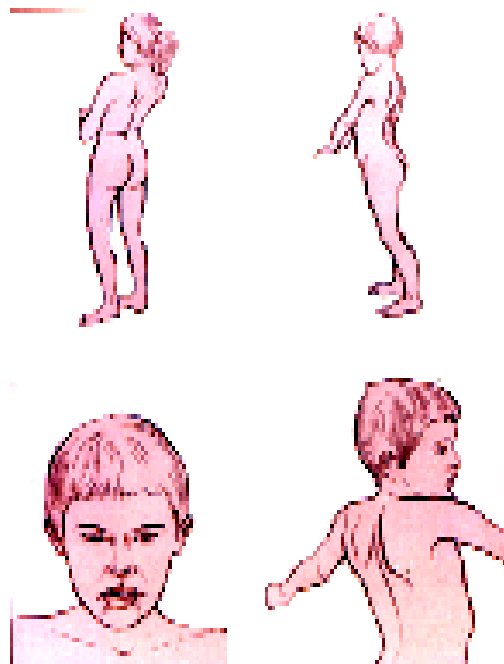


Fig. 91. Erba-Rotta myodystrophy.



Fig. 92. Dushenne myodystrophy. “Climbing up his own trunk” with the arms.

pulling himself up with his hands on a chair or some other fixed object.

On palpation the hypertrophic muscles feel firm and rubbery. The wasted muscles are often difficult to feel on account of the overlying fat. Pseudohypertrophy may precede the onset of wasting, or it may affect muscles which have never hypertrophied. The involvement of the musculature is usually symmetrical. There are some variations in the degree of weakness on the two sides but involvement limited entirely to one side does not occur. Abnormal movements and fibrillary twitchings of the muscles are not present.

The sensory examination is normal and there are no sphincter disturbances. The deep reflexes may be lost early in the course of the disease or they may persist in wasted muscle. The knee jerks usually disappear before the ankle jerks. Cutaneous reflexes are preserved and the plantar responses are usually flexor.

Dushenne pseudo – hypertrophic muscular dystrophy occurs entirely in males and the onset is usually in the first four years of life.

The transmission is as sex-linked recessive trait and the

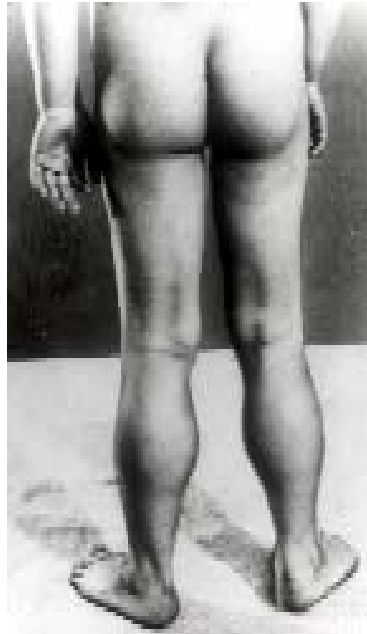


Fig. 93. Dushenne pseudo-hypertrophic muscular dystrophy.

mutation rate is high. The rate of progression is relatively rapid, with death in the second or third decade. A milder form of X-linked recessive dystrophy was described by Becker. The clinical features are similar to those of Duchenne dystrophy but the onset is later in childhood or adolescence and the rate of progression is much slower, so that survival into adult years is common.

In Duchenne dystrophy the child is clumsy in walking and has difficulty in climbing up and down stairs. Toe walking is a common early symptom. There are no cerebral symptoms, but mental retardation seems to be unduly common in the Duchenne type. In Duchenne dystrophy, pseudohypertrophy (fig. 93) is present in some muscles of the extremities and in others it is entirely absent. More

often, there is wasting of some muscle groups and pseudohypertrophy in others. The gastrocnemius, deltoid and triceps are most frequently affected by the pseudohypertrophy.

Facioscapulohumeral muscular dystrophy Landuzi – Degerina type occurs in both sexes. The onset of symptoms may be at any time from early childhood to late adult life. There are many mildly affected abortive cases.

Transmission is by an autosomal dominant gene (fig. 94).

When the facial muscles are affected in the Landuzi-Dejerine type, the expression is mask-like, the lips are prominent, the eyes are imperfectly closed in sleeping and facial movements are absent in laughing or crying as well as on voluntary efforts in whistling and the like. Involvement of the masticator, palatal and pharyngeal muscles may occur, but is rare.



Fig. 94. Landuzi-Degerin myodystrophy.

Erba-Rotta dystrophy (limb-girdle type) occurs in either sex and the onset of symptoms is usually in the first three decades of life. There are various modes of inheritance, but it is commonly transmitted as an autosomal recessive trait (fig. 95).

Examinations

Routine examinations of the blood, urine and cerebrospinal fluid are normal. The excretion of creatinine is decreased in proportion to the amount of loss of muscle substance, in a similar manner to that occurring in other diseases accompanied by muscular wasting. There is an increase in the amount of creatine excreted in the urine, and there is impairment of the ability of the body to store ingested creatine. The significance of the creatinuria is not known. There is no consistent or specific pattern of amino acid excretion.

Increased serum levels of aldolase, lactic dehydrogenase, phosphohexoisomerase, transaminase and creatine phosphokinase have been reported. Serum enzyme levels are most consistently



Fig. 95. Erba-Rotta dystrophy

elevated in the early stages of the Duchenne variety of muscular dystrophy. Detection of the disease in the preclinical stage of this form of the dystrophy can be made by determination of the serum enzymes, and detection of the carrier state in unaffected females is manifested by an increase in the serum of creatine phosphokinase. When the serum CPK is definitely increased in a potential carrier of the Duchenne gene, it is likely that the woman is a carrier, but borderline or normal values do not exclude this possibility because even in known carriers (for instance, a woman with both an affected brother and an affected son),

CPK is abnormal in only about 80% of the cases. A mild degree of degenerative change in the muscles has also been found in some asymptomatic carriers.

Among the biochemical abnormalities that suggest an abnormality of surface membranes in Duchenne dystrophy are impaired responses of adenyl cyclase to epinephrine and fluoride in muscle, and several abnormalities of erythrocyte membranes, including sodium-potassium ATPase, membrane phosphorylation, osmotic fragility, and some morphological characteristics.

Ultrastructural study of muscle has revealed gaps in the plasma membrane of muscle. In contrast to normal surfaces, these gaps seem to be permeable to large molecules such as the protein, horseradish peroxidase, or a dye, procion yellow. Additionally, freeze-fracture studies showed decreased numbers of membrane particles. These abnormalities bear upon theories of pathogenesis, but have not yet had an impact on diagnosis of individual cases.

The electromyogram is of considerable value in diagnosis. The pattern of voluntary effort recorded by means of concentric needle electrodes is characterized by disintegration of motor unit potentials, many of which become polyphasic and of short duration.

Course

There is considerable variation in the course of these diseases. The prognosis is most favorable when the onset of symptoms occurs after the second decade of life. As a rule, there is a gradual increase in the weakness of the muscles which are first involved and a slow progress of the wasting to unaffected muscles. The small muscles of the hands and feet are usually last to be affected.

Contractures may appear and atrophic changes in the bones have been reported in a few cases. *Formes frustes* with preservation of general good health to old age are not rare in facioscapulohumeral dystrophy, and it is not unusual to find patients who have suffered with the disease for four and five decades but are still able to walk. In the Duchenne form it is common for the disease to progress within a period of five to fifteen years to the stage where the patient is confined to a bed or a wheel-chair. Death may occur from intercurrent infection or from involvement of the respiratory musculature.

Diagnosis

The diagnosis of Duchenne dystrophy can usually be made without difficulty by the onset of muscular weakness in childhood, the presence of pseudohypertrophy of the muscles, the characteristic distribution of the weakness, the family history and the increased serum enzyme activity. When the onset is relatively late in life, as in limb-girdle or facioscapulohumeral forms, the diagnosis is made by the distribution of the weakness, the loss of deep reflexes and the absence of evidence of involvement of the spinal cord or peripheral nerves. Determination of the serum enzymes, electromyography and biopsy of the muscles are of value in establishing the diagnosis.

Differential diagnosis

Progressive muscular dystrophy must be distinguished from the diseases of infancy and childhood which are accompanied by muscular wasting. Limb-girdle and facioscapulohumeral dystrophy must be distinguished from diseases of adult life which are

accompanied by muscular wasting:

1. myotonic muscular dystrophy;
2. proximal spinal muscular atrophy;
3. peroneal muscular atroph;
4. amyotrophic lateral sclerosis;
5. atypical forms of polyneuritis;
6. syringomyelia and myositis.

The differential diagnosis between named diseases and progressive muscular dystrophy can be made by the lack of family history, much more rapid course, inflammatory response in muscle. Genetic analysis has shown that there is an X-linked dystrophy, the Becker type, less severe than the Duchenne type; recognition of Becker cases is facilitated by the recognition of other affected individuals in the same family who are still walking after age twenty, but in sporadic cases, the separation of Becker cases from either Duchenne or limb-girdle cases may be difficult.

In polyneuritis, particularly the Guillain-Barre form, the muscular weakness may occasionally be greatest in the girdle muscles. The acute onset of the symptoms, the increased protein content in the cerebrospinal fluid and the subsequent regression of symptoms should serve to establish the diagnosis.

Pseudohypertrophy of the muscles may occasionally be seen in syringomyelia but the other features of the disease should leave no doubt in regard in the diagnosis.

Treatment. There is no treatment which has proven to be effective in arresting the course of the disease. Stretching of contractures, bracing and tendon-lengthening operations are advocated with varying degrees of enthusiasm in different centers. When known carriers of the Duchenne gene become pregnant, fetal sex determination permits interruption of pregnancy for boys, but there is no way of identifying.

Amyotrophy as a result of peripheral neuron lesion

- A. Spinal amyotrophy of Werding – Hoffman.
- B. Proximal amyotrophy of Kugelberg – Welander.
- C. Sharko – Mary – Tooth disease.

Infantile muscular atrophy (**Werding – Hoffman**) is recognized by its onset in infancy, the presence of fibrillations and the rapidly

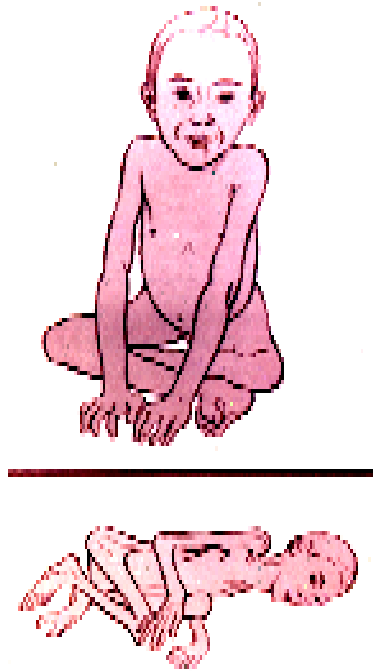


Fig. 96. Werdnig-Hoffmann amyotrophy.



Fig. 97.

fatal course (fig. 95-96).

Juvenile muscular atrophy (**Kugelberg-Welander**) may be suspected if fasciculations are seen in a patient who otherwise seems to have limb-girdle or facioscapulohumeral dystrophy because of proximal limb weakness. In some patients, however, fasciculations are not clinically visible and motor neuron disease is identified only by electromyography and muscle biopsy. This distinction is of some importance because, in general but not always, the prognosis of juvenile neurogenic disease is less serious.

Hereditary neuropathies Peroneal muscular atrophy. Peroneal muscular atrophy (Charcot-Marie-Tooth syndrome) (fig. 98) includes several genetic disorders of the peripheral nervous system that most



Fig. 98. Charcot-Marie-Tooth syndrome

severely affect the peroneal and other distal muscles of the legs.

Inheritance is usually autosomal dominant and, less frequently, autosomal recessive.

Foot deformities are frequent and may be the only apparent feature of the disease in mildly affected family members. Impairment of sensation in a stocking-and-glove distribution is usually present, though sensation is preserved in some families. Achilles reflexes are absent and other tendon reflexes may be diminished.

Clinical features

The pathologic changes are of three types. There are:

1. Demyelination of the peripheral nerve with axonal loss and some hypertrophy of the Schwann cells, producing hypertrophic neuropathic changes. In Type I Charcot-Marie-Tooth syndrome, symptoms begin in the first or second decade of life with foot-drop and a stoppage gait. Distal muscle atrophy produces a "stork leg" deformity (fig. 98); intrinsic hand muscle atrophy develops later. Distal tendon reflexes are diminished or absent and a stocking-and-glove sensory defect is present. Scoliosis and high pedal arches or club feet are common. Peripheral nerves are often

palpably enlarged. Tremor is prominent in some patients; the clinical constellation of Charcot-Marie-Tooth syndrome with tremor is termed the Roussy-Levy syndrome.

2. Neuronal loss of anterior horn cells and posterior nerve root ganglion neurons in the lumbar and sacral segments (hereditary motor and sensory neuropathy, Type II). The first symptoms of peroneal muscular atrophy often appear in adult life, but foot deformities may be evident much earlier. The progression is slow with muscle weakness and wasting confined to the feet and sometimes involving the leg muscles. The involvement may be asymmetric. Atrophy and weakness of distal muscles, stocking-and-glove sensory impairment is minimal or absent, and depression of tendon reflexes in the lower limbs but normal in the arms. Deficits are usually less severe and nerves are not palpably enlarged.
3. Anterior horn cell involvement with secondary axonal loss and demyelination of motor fibers.

Examinations

1. Lumbar puncture. CSF protein content is frequently elevated in Type I Charcot-Marie-Tooth syndrome, but is normal in Type II. The CSF is otherwise normal, as are blood and urine.
2. Nerve conduction velocities. Motor and sensory nerve velocities are very slow in the peripheral nerve demyelinating type but are normal or only slightly delayed in the other two types.
3. Nerve biopsy. Nerve biopsy is normal and should differentiate the three types of the disease.

Differential diagnosis

Peroneal muscular atrophy is considered when there is atrophy of the peroneal muscles, foot deformity, and a positive family history. Even if family history is reported to be negative, relatives should be examined because some affected individuals are asymptomatic, or deny mild symptoms.

1. **Friedreich ataxia** should be considered if distal neuropathy and a positive family history are accompanied by nystagmus, dysarthria, ataxia, or Babinski signs.
2. **Familial amyloidosis** may resemble peroneal muscular atrophy, and can be recognized by sural nerve biopsy.

3. **Dejerine-Sottas** syndrome also resembles peroneal muscular atrophy, but onset is earlier, nerve hypertrophy is more prominent, the elevation in CSF protein content is greater, and nerve conduction rates are slower than in Type I Charcot-Marie-Tooth syndrome.
4. The distal motor and sensory deficits in **Refsum disease** are associated with deafness, retinitis pigmentosa, scaly skin, and elevated serum phytanic acid.
5. **Lipomas** and other masses in the lumbosacral canal may cause neurogenic foot deformities and distal weakness, but sensory loss is in a radicular pattern, the arms are spared, and the family history is usually negative.
6. **Myotonic muscular dystrophy** may present with a similar pattern of distal atrophy and is a dominantly inherited disorder, but is distinguished from Charcot-Marie-Tooth syndrome by the presence of myotonia, cataracts, frontal balding, the absence of sensory abnormalities, and by the results of EMG studies.

Course and Treatment. The progression of disability is slow in Type I Charcot-Marie-Tooth syndrome and very slow in Type II. Death does not occur as a result of this syndrome and complete incapacitation is rare. There is no specific treatment. Braces for correction of the foot-drop and hand deformities can be helped ambulatory by splinting and orthopedic procedures.

Hereditary sensory neuropathy. These genetic disorders affect sensory fibers in peripheral nerve and sometimes autonomic fibers as well. Dominantly inherited, or Type I, hereditary sensory neuropathy causes progressive sensory loss beginning in the first or second decade. There is progressive loss of pain, thermal sensibility, light touch, and proprioception. Tendon reflexes are lost. Ulcerations may develop on the feet and fingers owing to unperceived injuries. The disorder is the result of a selective degeneration of sensory neurons in the dorsal root ganglia.

Other types of hereditary sensory neuropathy include an autosomal recessive form that resembles the dominantly inherited disorder, congenital sensory neuropathy with anhydrosis, hereditary sensory neuropathy with spastic paraparesis, and familial dysautonomia (**Riley-Day syndrome**). Familial dysautonomia is an

autosomal recessive condition most common in Jews, and may be a result of an inherited defect in synthesis of nerve growth factor. It presents in infancy with lack of pain and temperature sensibilities, gastrointestinal dysfunction, poor regulation of blood pressure, and absence of fungiform papillae on the tongue.

Hereditary muscle diseases associated with Myotonia

Myotonia is a sustained contraction of muscles that may be induced by voluntary contraction, percussion, or electrical stimulation. The primary failure of myotonia is a delayed relaxation due to increased excitability of muscle demonstrated on electromyography, which records discharges of repetitive action potentials following muscle contraction. Myotonia occurs in myotonia congenita (of Thomsen), myotonic dystrophy (atonic myotonia), paramyotonia congenita (of Alenburg).

Myotonia Congenita (Thomsen myotonia)

Myotonia congenita is an hereditary condition of delayed relaxation following voluntary muscle contraction. Myotonia congenita is caused by a genetically determined abnormality in the voltage gated chloride channel. This results in a blocking of membrane chloride conductance and action potentials can be triggered by a smaller than normal current, resulting in a train of impulses that are self-maintaining, following termination of the stimulating pulse.

Thomsen Myotonia (a mild form) is autosomal-dominant trait of inheritance or Becker myotonia (a more severe form) inherited as an autosomal-recessive trait.

Clinical features of disease

The first symptoms of myotonia appear in infancy or childhood as inability to relax a muscle following contraction. The symptoms tend to increase during childhood and adolescence but may lessen in severity in adult life.

The main clinical symptoms of classic form are:

- Myotonia occurs only in skeletal muscle, and the patient has difficulty initiating movement and making certain movements.
- Once repetitive movements are initiated, they can be continued without difficulty.

- Sudden movements may initiate a sustained contraction sufficient to throw the patient off balance.
- Patients with myotonia congenita have a well-developed musculature and have been described as herculean in appearance.
- Percussion of an affected muscle produces percussion myotonia, a dimpling of the skin, and sustained contraction. Percussion may also be followed by local swelling of muscle, termed myedema.

Diagnosis

1. Clinical and genetic analysis
2. Electromyography (traction or percussion of the muscle produces a series of prolonged potentials that persist when the patient is instructed to relax the contracted muscle. Contraction and relaxation of the muscle produces a typical sound on electromyographic examination, which has been termed the “dive bomber” effect)

Treatment

1. Mexiletine 150 mg q12h, increasing to 300 mg q8h, is the drug of choice to reduce myotonia.
2. Phenytoin (Dilantin) 100 mg q12h and increasing the dose until the serum levels are within therapeutic range is also an effective treatment.
3. Procainamide HC1 (Pronestyl) 50 mg/kg/day given in divided dosage four times a day and quinine 10 mg/kg/day in divided dosage.
4. Intractable cases may respond to acetazolamide (Diamox) 8 to 30 mg/kg/day in divided dosage.

Myotonic Dystrophy (atonic myotonia)

Myotonic dystrophy is a multisystem disorder inherited as an autosomal dominant trait through a locus on chromosome 19. The disease is characterized by progressive muscular weakness, myotonia, cataracts, cardiac abnormalities, hypogonadism, and frontal balding. Myotonic dystrophy is the product of an expanded CTG repeat in the 3' untranslated region of a gene that encodes a protein kinase DM-PK on chromosome 19. There is an expansion of

the repeat sequence in myotonic dystrophy and a positive correlation between repeat size and clinical severity. Protein kinases phosphorylate neurotransmitters to achieve a physiological response on specific target cells. Failure of protein kinase activity may cause ion channel dysfunction in myotonia. Expression is maximal in cardiac muscle in myotonic dystrophy but is also present in skeletal muscle and brain.

Clinical Features of the disease

The signs and symptoms of the disease appear at any time from birth to age 40 years. Hydrops fetalis has been reported in congenital myotonic dystrophy in a newborn infant who presented with hypertonia, edema, pleural effusion, and respiratory distress. Myotonia is usually the first symptom and affects the hand with later involvement of other muscles, particularly those of the lower limbs. Muscle wasting also affects the hands first then spreads to the facial muscles, muscles of mastication, the sternocleidomastoids, the flexors and extensors of the forearm, the quadriceps, and the dorsiflexors of the feet. The facial appearance is characteristic, with bilateral ptosis and wasting, which has been termed "hatchet face."

Patients with myotonic dystrophy usually have involvement of other organ systems. These include:

1. Cardiac abnormalities (prolapsed mitral valve, atrial flutter, cardiac arrhythmias, syncopal attacks). Subclinical cardiac involvement is not uncommon and may be responsible for sudden death in some cases.
2. Brain involvement (mental retardation, progressive dementia). Magnetic resonance imaging (MRI) shows cerebral atrophy and areas of increased signal intensity in the white matter in most adults with myotonic dystrophy.
3. Ophthalmic abnormalities (subcapsular lens opacities, which enlarge and eventually impair vision).
4. Endocrine abnormalities (primary gonadal failure and gonadal atrophy, impotence, loss of libido, and testicular atrophy, mild diabetes mellitus).
5. Skin and skeletal abnormalities (frontal balding, a high-arched palate).
6. Smooth muscle abnormalities (impairment of mobility in the gastrointestinal tract, with dilatation of the colon).

7. Respiratory abnormalities (respiratory insufficiency, hypoventilation is a feature in the late stages of the disease when there is an increased risk of aspiration pneumonia).
8. Hearing loss (sensorineural hearing loss).
9. Peripheral neuropathy of axonal type is responsible for the areflexia, distal sensory impairment, and fasciculations.

Children born to mothers with myotonic dystrophy may present with congenital myotonic dystrophy in the neonatal period. This condition is characterized by hypotonia, facial diplegia, dysphagia, mental retardation, a high-arched palate, and tented lips. Myotonia develops later in the course of this condition.

Diagnosis

1. DNA analysis using a DNA probe allows direct identification of the myotonic dystrophy mutation in DNA extracted from peripheral blood lymphocytes.
2. The effect of temperature change. Submersion of the hands in cold water for several minutes may facilitate the appearance of myotonia.
3. The electromyogram is characteristic with an increase in insertional activity and typical myotonic discharges following voluntary contraction of muscle.
4. Cardiac involvement affects predominantly the conduction system in the heart. Atrial fibrillation, atrial flutter, a prolonged PR interval, and various conduction defects may be present.
5. The MRI shows the presence of increased signal intensity in the white matter.
6. Computed tomography (CT) scans. Microcephaly, calcification of the basal ganglia, and cerebral atrophy may be demonstrated by CT scan.
7. Chest and skull x-rays. Thickening of the calvarium and enlargement of the frontal sinuses are often present on skull x-rays.
8. Pure tone screening and impedance audiometry will detect sensorineural hearing loss.
9. The diagnosis can be established by a muscle biopsy, which shows characteristic findings of a dystrophic process.

Treatment

1. The calcium channel blocking agent, nifedipine, which has no effect on cardiac conduction, decreases myotonia in doses of 10 to 20 mg q8h.
2. The respiratory infection should be treated with appropriate antibiotics, postural drainage, and chest percussion.
3. Patients with muscle weakness need supportive care, including splinting and use of an electric cart or wheelchair.
4. Cardiac involvement need appropriate treatment for arrhythmias.

Prognosis

Myotonic dystrophy is a chronic condition with progressive deterioration in most cases. In noncongenital forms of the disease, death occurs between ages 40 and 60 years due to cardiac or respiratory dysfunction. In congenital muscular dystrophy, there is a 25 percent chance of death before 18 months of age, and a 50 percent chance of survival until the mid-thirties.

Chondrodystrophic Myotonia

Disease is inherited as an autosomal recessive trait and is characterized by myotonia, short stature, blepharospasm, muscle hypertrophy, and skeletal deformities. The affected infant presents with hip contractures or dislocation. This is followed by other joint contractures, progressive growth failure, and myotonia. There is a puckering of the mouth, blepharospasm, and small palpebral fissures. Intelligence is normal. Electromyographic findings are typically those of myotonia. Muscular atrophy can occur in older children.

Hereditary diseases with paroxysmal states

Familial periodic paralysis

The familial periodic paralysis is inherited as an autosomal dominant trait.

1. Hypokalemic Periodic Paralysis is an inherited condition of episodic muscle paralysis associated with hypokalemia.

Etiology of Familial hypokalemic periodic paralysis is mutation in chromosome 1q31-32. Mutation in this gene modifies the function of the **DHP** receptor by altering calcium channel current in hypokalemic periodic paralysis.

Muscle biopsy shows the presence of vacuoles, containing glycogen, in muscle fibers, which are present during an attack, and which may decrease in number immediately after an attack.

Clinical Features Hypokalemic periodic paralysis is more common in men and occurs predominantly during the teens and twenties. Attacks begin at night and the patient awakens with weakness of all skeletal muscles except those involved in respiration and speech. However, there are reports of respiratory involvement and some deaths from respiratory failure, but this is a rare complication. Involved muscles are firm and tender to palpation. The neurological examination is normal, with sparing of muscles supplied by cranial nerves and those involved in respiration and speech. The attacks last from several hours to days. Several factors are: large meals with a high carbohydrate content; exertion; trauma; heavy alcohol ingestion; upper respiratory tract infection; cold weather; and administration of insulin, thyroid hormone, steroids, epinephrine, thiazides, or licorice.

Diagnosis

1. The movement of potassium, sodium chloride, phosphate, ions, and water into the muscle fibers during an attack is reflected in decreased serum potassium level below 3.5 mEq/L.
2. Urinary excretion of potassium and sodium is decreased.
3. Electromyography shows decreased amplitude, number, and duration of motor unit potentials or electrical silence during periods of paralysis. Muscle fiber velocity is reduced between attacks.
4. Provocative tests to induce hypokalemia, using glucose and insulin, require close monitoring by electrocardiography because hypokalemia may induce cardiac arrhythmia. A 10-min bicycle exercise test is a safer alternative. Patients with hypokalemic periodic paralysis experience a minimal increase in serum potassium levels 10 and 30 min after exercise, compared to a control population where the increase is significant.
5. Corticotropin 80 to 100 IU intravenously can be used in suspected periodic paralysis of either hypo- or hyperkalemic type. Corticotropin will induce an attack of paralysis with appropriate changes in serum potassium concentration in each condition.

Treatment

1. Oral or parenteral administration of potassium 130 mEq/day.
2. Acetazolamide 125 mg q.o.d., increasing by increments to achieve an optimum dosage, to a maximum of 500 mg q12h orally, is the drug of choice to prevent attacks.
3. Spironolactone 100 mg daily or bid is effective in reducing the number of attacks.
4. Other drugs that may be of benefit include the carbonic anhydrase inhibitor dichlorphenamide, the calcium channel blocking agent, verapamil, or lithium.
5. Predisposing factor should be avoided.

2. Hyperkalemic Periodic Paralysis is a condition inherited as an autosomal dominant trait, where periodic paralysis is associated with elevation of serum potassium levels. The disease has been linked to allelic defects in a gene that encodes for the alpha subunit of the tetrodotoxin-sensitive skeletal muscle sodium channel, localized to chromosome 17q23 1-25.3. There is non-inactivation of this channel during an episode of paralysis, with influx of sodium resulting in sustained membrane depolarization.

Muscle biopsy may be normal or may show some nonspecific features, such as large variation in fiber size and central nuclei, increased subsarcolemmal glycogen and the presence of vacuoles in some cases.

Clinical Features

Attacks begin in childhood and occur during the day, usually while resting after exercise. Each attack may be preceded by a sensation of heaviness and stiffness in the muscles and paresthesias in the face and extremities. Episodes usually last 1 h. In addition to exercise, paralysis can be precipitated by exposure to cold, hunger, administration of potassium, emotional stress, and pregnancy. Attacks may be prevented by eating a carbohydrate after exercise. Muscle weakness affects the lower extremities predominantly, but upper extremity and neck muscles can be involved. Mild myotonia can be experienced during muscle weakness. Permanent muscle weakness is an occasional complication.

Diagnosis

1. Serum potassium levels are increased during an attack, but normal potassium levels have been recorded in some cases, giving rise to so-called normokalemic periodic paralysis.
2. Serum sodium levels decrease as sodium enters muscle fibers.
3. Urinary potassium increases during an attack, resulting in a drop in serum potassium levels and recovery.
4. Electromyography may reveal electrical silence during paralysis or fibrillations, positive sharp waves, and myotonic discharges during paresis. Motor unit potentials are decreased in number and duration.
5. Provocative test with administration of 2 to 10 g potassium chloride will induce an attack within 1 to 2 h. Electrocardiographic monitoring should be performed to detect any abnormalities related to hyperkalemia.

Treatment Attacks can be prevented by thiazide diuretics—hydrochlorothiazide 25 to 75 mg/day. Acetazolamide, albuterol, and metaproterenol are also effective.

Myotonic muscular dystrophy is distinguished from the cases of muscular dystrophy of relatively late onset by the distal weakness, myotonia, and the other characteristic features of the disease, i.e., cataracts, testicular atrophy and early baldness.

Additional Cranial nerves

Anatomy of Olfactory nerve (fig 99).

The nerve endings that arise from bipolar cells located in the mucous membrane of the upper portion of the nasal cavity.

The axons of the bipolar cells pass in bundles through the cribriform plate of the ethmoid bone to enter the olfactory bulb on the floor of the anterior cranial fossa.

The axons of the mitral cells pass through the olfactory bulb and divide into medial and lateral olfactory striae.

Fibers in the medial olfactory striae terminate in the paraolfactory area, subcallosal gyrus, and the inferior portion of the cingulate gyrus.

Fibers from the lateral olfactory striae enter the gyriform area, which contains the uncus and the anterior portion of the hippocampal gyrus.

Clinical examination of Olfactory nerve. Odoriferous substances, such as wintergreen and camphor, are used when testing olfactory sensation. The ability to perceive such substances is tested separately in each nostril by having the patient sniff the test substance while he or the examiner closes the other nostril. The patient is asked first to state whether he perceives an odor, and if he makes a positive answer he is then asked to identify the smell.

Anatomy of Visual system

Retina The receptors for visual stimuli are of two types—rods and cones—and are the outer segments of the cells in the outer nuclear cell layer. The rods and cones are specialized photosensitive receptors and are located in the outer nuclear layer of the retina. The afferent processes synapse with dendrites of bipolar cells lying in the bipolar cell layer. The bipolar cells in turn synapse with dendrites of ganglion cells lying in the ganglion cell layer of the retina. The unmyelinated axons of the ganglion cells converge in centripetal fashion in the superficial, nerve fiber layer of the retina and form the optic nerve.

Optic nerve The optic nerve is situated slightly to the nasal side of the retina. The fibers that compose the optic nerve become myelinated as they pierce the lamina cribrosa of the sclera. The optic nerve traverses the optic foramen of the orbit accompanied by the ophthalmic artery and is

Cranial nerves

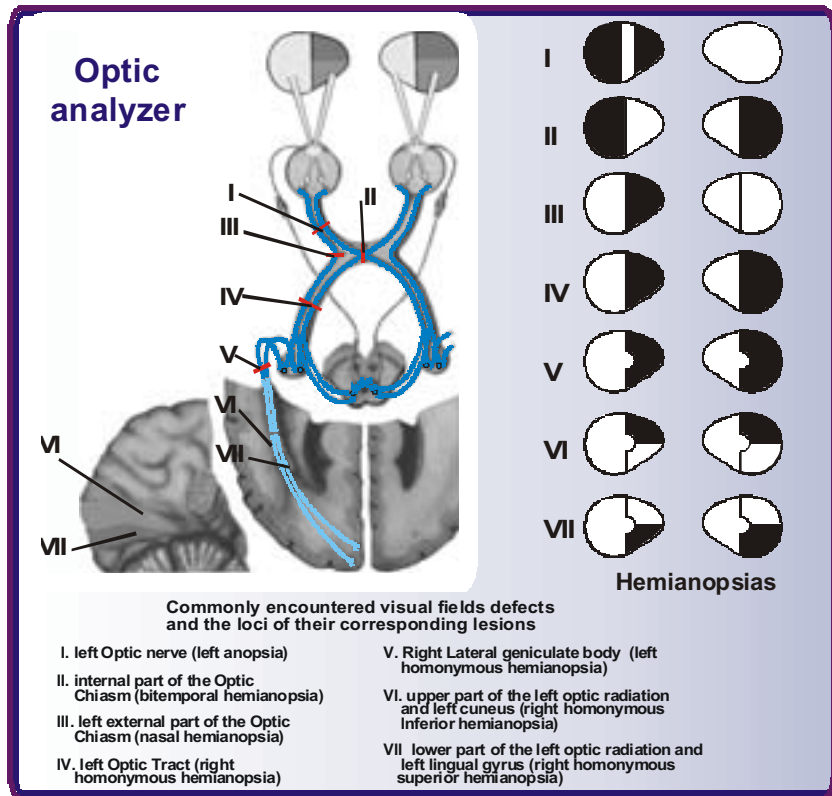
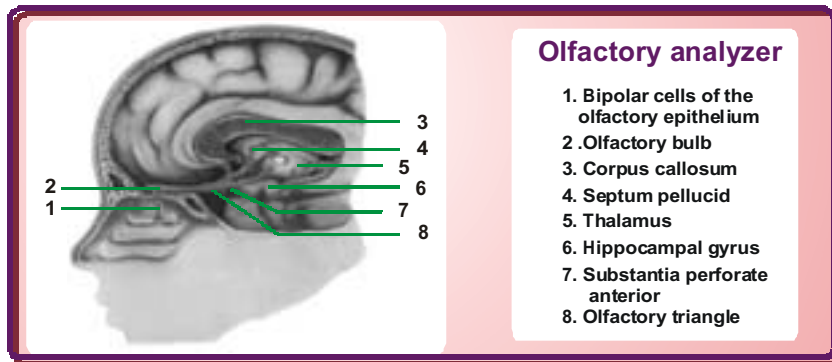


Fig. 99.

joined by the central artery of the retina. The course within the cranial cavity is short and the optic nerve is closely related to the olfactory tract, internal carotid, and anterior cerebral arteries as it terminates in the optic chiasm.

Optic chiasm The optic chiasm is located superior and slightly anterior to the pituitary fossa, pituitary gland, and infundibulum. Nerve fibers from the nasal half of each retina decussate in the optic chiasm and enter the opposite optic tract. Fibers from the inferior nasal quadrant of the retina loop forward into the opposite optic nerve just before they turn into the optic tract.

Optic tract The optic tract receives fibers from the temporal half of the ipsilateral retina and the nasal half of the contralateral retina. The optic tract passes posterolaterally around the cerebral peduncle and terminates in the lateral geniculate body of the thalamus.

Optic radiation The optic radiation arises from the lateral geniculate body and passes laterally in the retrolenticular portion of the internal capsule anterior to the descending portion of the lateral ventricle. Fibers from the lower visual fields, pass dorsally along the lateral wall of the posterior horn of the lateral ventricle and terminate in the superior lip of the calcarine fissure. Fibers from the upper visual fields, pass forward in the roof of the temporal horn of the lateral ventricle. They then loop backward (Meyer's loop) and pass inferiorly into the parietal lobe in the lateral wall of the posterior horn of the lateral ventricle and terminate in the inferior lip of the calcarine fissure.

Clinical examination of Optic nerve. *Visual acuity* of each eye is tested separately for distant and near vision. Distant vision is determined by use of the Snellen test chart, which is placed 5 m from the patient. The *field of vision* may be defined as that portion of space in which objects are visible during fixation of gaze in one direction. One of the most reliable of the confrontation test may be referred to as outline perimetry (The examiner covers one eye of the patient by holding a small card over it. The patient is instructed to fix his gaze on one eye of the examiner and to inform him when first he sees a pencil which the examiner slowly moves to the patient's field of vision. The pencil is held a few inches from the patient's face outside of his field of vision and is moved forward to it. For accurate testing of cooperative patients the peripheral portions of the field are explored on a perimeter with the aid of an arc which can be turned in any desired meridian and which is marked in degrees to 90°. Another method (ophthalmoscopy and ophthalmodynamometry) are repeated by the ophthalmologist (fig. 100-101).



Fig. 100. Visual fields by confrontation test.

Fig. 101. Testing visual fields by

Clinical examination of Oculomotor, Trochlear and Abducens nerves include: general observation, determination of ptosis and examination of the pupil (size, testing the reaction of the pupil to light, the convergence reflex). Ocular rotations are tested by asking the patient to turn the eyes in the six cardinal directions of gaze and by having the eyes converge on a near point. Paralysis or weakness of a single or several ocular muscles or of conjugate movements (movements of both eyes in the same direction) and the presence or absence of nystagmus on conjugate deviation is observed. Normally, there is a great variation in the limits of upward and downward rotation, but differences between the two eyes are readily recognized and are important in the diagnosis of paralysis of single ocular muscles (fig. 102).

Light reflex The afferent pathway for the pupillary light reflex is activated by light, which stimulates the rods and cones in the retina. The afferent pathway passes via the optic nerve with partial decussation in the optic chiasm and then continues bilaterally through both optic tracts and the brachium of the superior colliculi to the pretectal region of the midbrain. From the pretectal region the fibers pass forward close to the aqueduct to enter the Edinger-Westphal nucleus on the same side or pass through the posterior commissure to enter the nucleus on the opposite side.

The efferent side of the reflex is completed by fibers that pass from the neurons in the Edinger-Westphal nucleus through the oculomotor nerve to the ciliary ganglion, and thereafter by short ciliary nerves to the constrictor muscles of the iris (fig. 103-107).

Cranial nerves

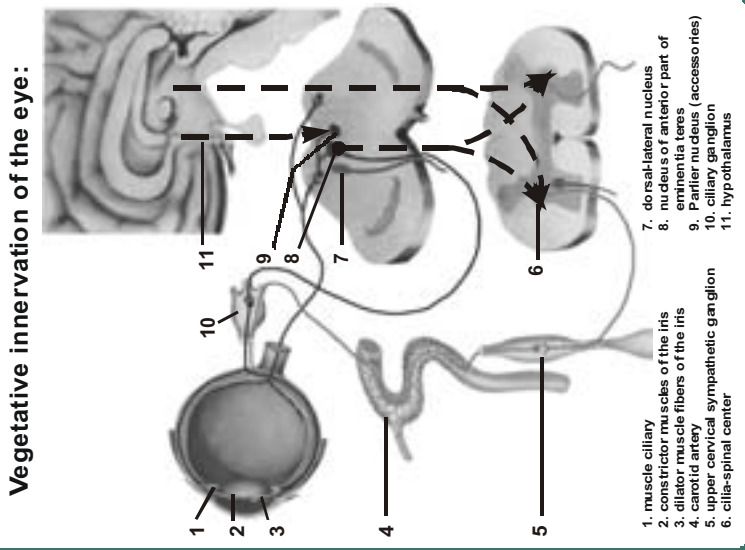
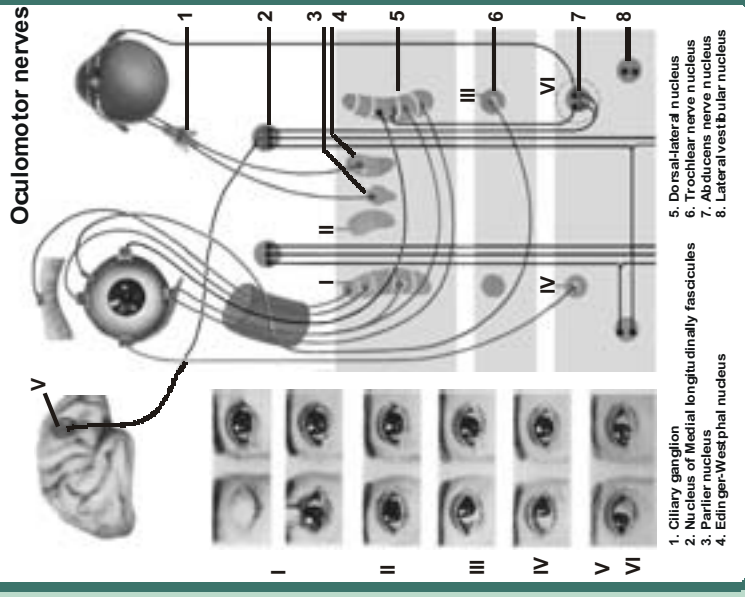


Fig. 102.

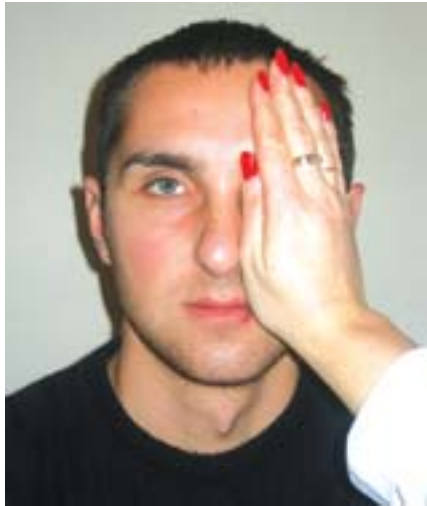


Fig. 103. Testing of light reflex.



Fig. 104. Testing of oculomotor function.

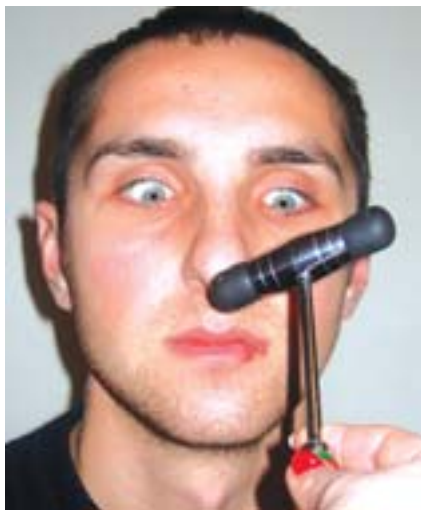


Fig. 105. Testing of convergence.



Fig. 106. Lesion of Oculomotor nerve. Outside cross-eye.



Fig. 107. Lesion of Abducens nerve, inside cross-eye.

Alternate midbrain syndromes:

Weber's syndrome – a lesion in the base of the cerebral peduncle affects the root of the third cranial nerve and the corticospinal pathway, producing Oculomotor nerve palsy and a contralateral hemiplegia.

Benedict's syndrome – Oculomotor nerve palsy and a contralateral choreoathetosis and intention tremor.

Clod's syndrome – Oculomotor nerve palsy and a contralateral cerebellar ataxia.

Nerve	Symptoms
Olfactory nerve	patient can't identify the smell, he may be unable to name the test substance; hyposmia, anosmia; olfactory hallucination
Optic nerve	amblyopic, scotoma; bitemporal hemianopsia, binasal hemianopsia, homonymous hemianopsia; quadrantic hemianopsia, retinal lesions («choked disk»; «retrobulbar neuritis», primary optic atrophy, secondary optic atrophy, retinal emboli), optic hallucination
Oculomotor nerve	Ptosis; diplopia; outside cross eye, rotation of the eye outward and slightly downward; paralysis of convergence; inability to move the eye upward, inward, or downward; exophthalmus; midriasis (dilatation of the pupil) with iridoplegia and cycloplegia; paralysis of accommodation
Trochlear nerve	the eye can't be turned downward when it's rotated inward, diplopia downward
Abducens nerve	the ipsilateral eye is turned in toward the nose and abduction of the eye is impaired

Clinical examination of Trigeminal nerve includes testing pain, thermal, and other sensations in the area supplied by the trigeminal nerve. The corneal reflex is tested by having the patient look to one side while the cornea is lightly touched with cotton wool has been twisted into a compact cylinder or drawn out into a point. After testing corneal sensation, the patient is instructed to close the eyes, and the cotton is carefully and gently inserted into the right (or left) nostril. Normally the patient will draw away slightly and wrinkle the nose. Then the other nostril is tested. The temporal and masseter muscles are examined

by applying palpates the muscles and attempts to separate the jaws by applying pressure downward on the chin (fig. 108-111).

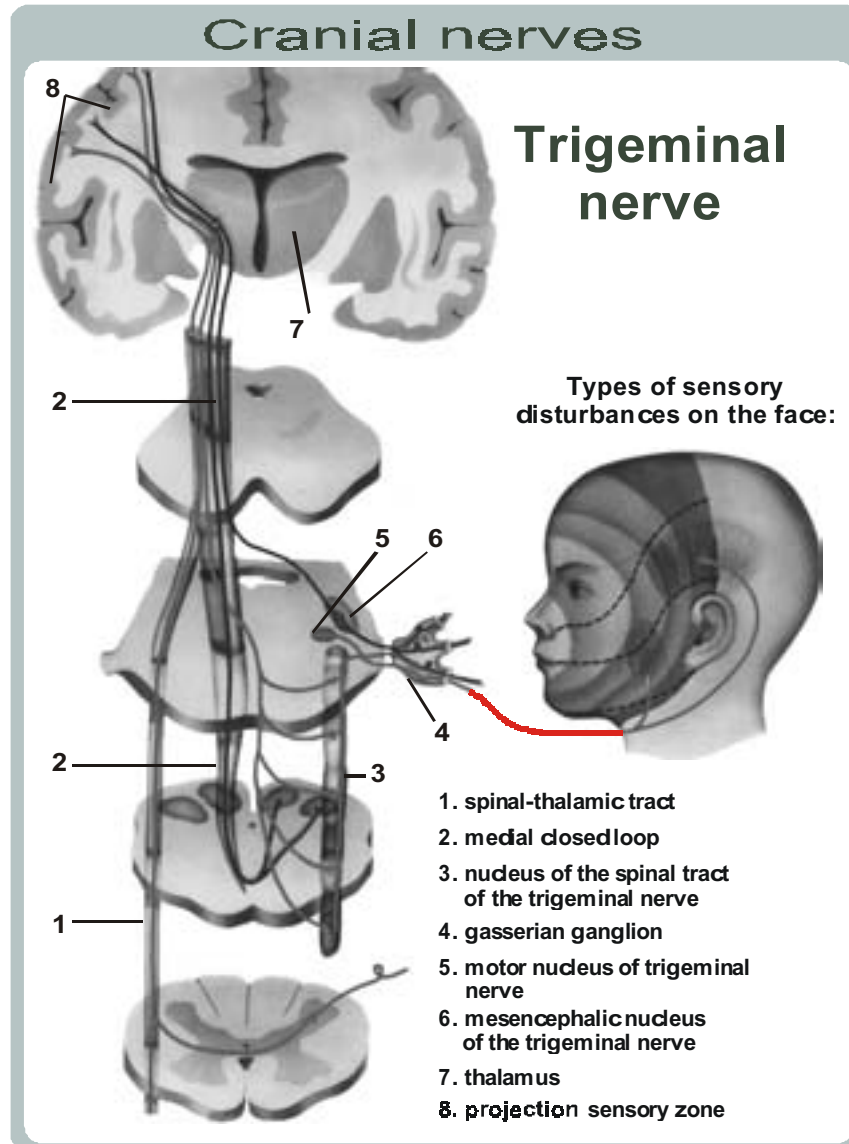


Fig. 108.



Fig. 109. Examination of chin reflex.



Fig. 110. Examination of corneal reflex.



Fig. 111. Examination of masseter muscles.

Clinical examination of Facial nerve function begins with the initial observation of the patient as he talks and smile. The motor portion of the nerve innervates muscles which wrinkle the forehead, close the eyes, purse the lips, and retract the buccal angles in a smile or grimace. The patient is asked to show his teeth by retracting the buccal angles, to whistle, and to press the lips against the pressure of the examiner's fingers. Slight unilateral weakness may be detected by asking the patient to blow the cheeks out fully. The lower facial musculature can be tested in stupor or uncooperative patients by observing the wincing reaction occasioned by pressing firmly over the styloid processes just posterior to the angle of the jaw. In infants the facial movements are observed during crying. The platysma can be observed to contract when the patient makes a maximal effort to draw the lower lip and angle of the mouth downward and outward, at the same time tensing the skin over the anterior surface of the neck. Taste is examined with the use of sugar, tartaric acid, sodium chloride, quinine, or similar substances. The patient is instructed to protrude the tongue; a small quantity of the test substance is gently rubbed on one side of the tongue and the patient signals identification of the test substance before drawing the tongue into the mouth to prevent diffusion of the taste to the opposite side or to the posterior third of the tongue, thus obscuring the tes (fig. 112-113).

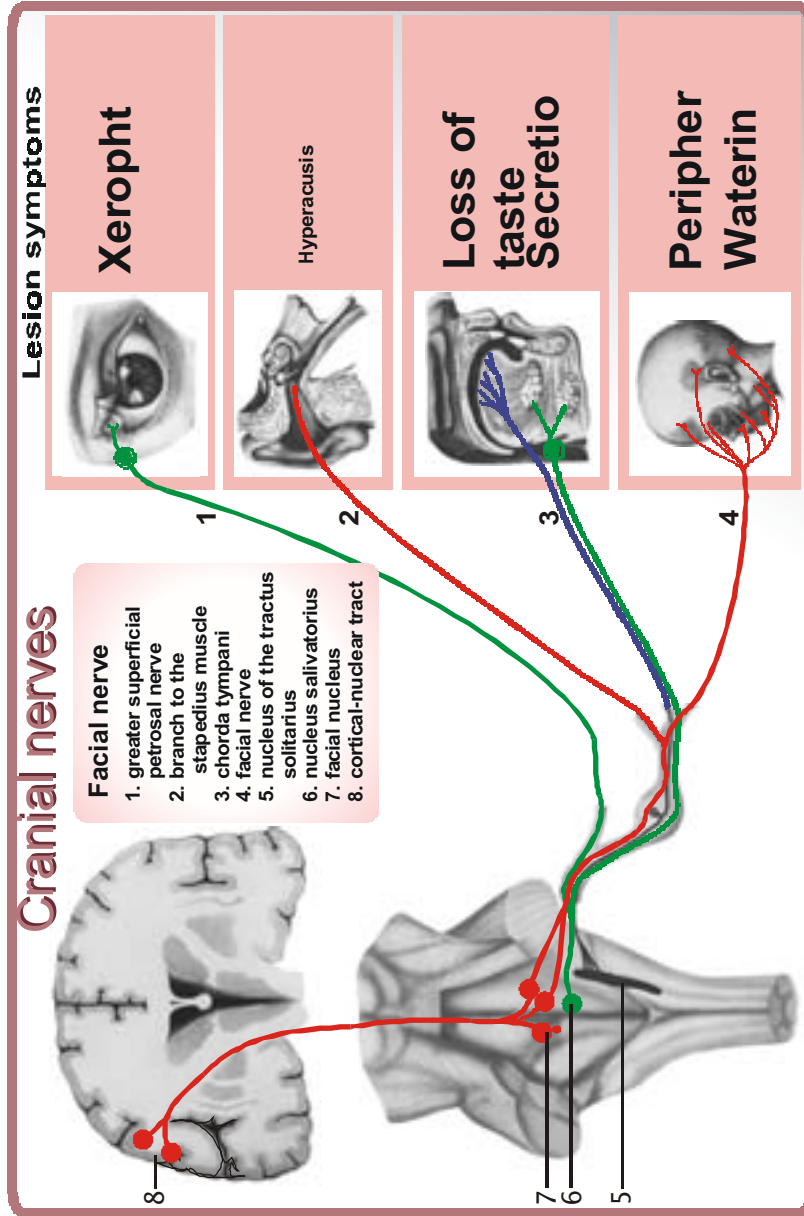


Fig. 112.



Fig. 113. Lesion of Facial nerve (Bell's palsy)

Clinical examination of Acoustic nerve include test of hearing (The so-called «dollar watch» is a commonly used test object. In a quiet room the «dollar watch» may be heard approximately 6 m from the normal ear). Another object for testing air conduction is a vibrating tuning fork of medium pitch (C=256 vibration per second). To measure hearing sense accurately, the electric audiometer is used and an audiogram is made (fig. 114-115).

Nerves	Symptoms
Trigeminal nerve	facial pain, sensory disturbance in the face; corneal reflex is decreased or absent; temporal and masseter muscles are atrophic or hypotrophic, atonic or hypotonic; jaw is seen to deviated toward the side of the weakened muscle;
Facial nerve	Facial asymmetry; patient can't wrinkle the forehead, close eyes, purse the lips, retract the buccal angles in a smile or grimace; impairment of taste on the anterior two third of the tongue; Bell's symptom; corneal reflex is decreased or absent;
Acoustics nerve	hypakusis; giddiness; tingle; dizziness or nausea; vertigo; vibrating turning fork of medium pitch (C=256) Rinne, Weber tests; audiometry; Barany test; electronystagmography.

Alternate pons' syndromes:

Raymond syndrome – a fallout of sensation on the face according to the segmental type on the side of the lesion both fallout pain and thermoesthesia on a trunk and extremities on opposite side.

Miyar- Gubler syndrome – peripheral palsy of the facial muscles on the site of the lesion and a contralateral hemiplegia.

Fovill syndrome – peripheral palsy of the facial muscles and the external rectus eyes' muscle on the site of the lesion and a contralateral hemiplegia.



Fig. 114. Rinne test.



Fig. 115. Weber test.

The ninth nerve (**Glossopharyngeal nerve**) is tested by touching the posterior wall of the pharynx with a wooden tongue depressor or applicator stick. The normal response is prompt contraction of the pharyngeal muscles, with or without gagging. However, the finding of a normal gag reflex after intracranial section of the ninth nerve suggests that the posterior pharyngeal wall is also supplied by the tenth cranial nerve. The testing of the taste sensation on the posterior one third of the tongue is technically too difficult to be of much clinical value.

Clinical testing of **Vagus nerve** is difficult in spite of the great size and many functions of nerve. Unilateral paralysis of the motor portion of the vagus produces ipsilateral paralysis of the palatal, pharyngeal muscles. The voice is hoarse or brassy as a result of weakness of the vocal cord, and speech has a nasal twang in lesions producing weakness of the soft palate, particularly if bilateral. Lesion of the recurrent laryngeal

branch of the vagus nerve produce weakness or paralysis of the ipsilateral vocal cord and the voice is coarse and husky. The soft palate is observed

as the patient says «ah». Normally the median raphe rises in the midline. However, if one side is weak, there will be deviation to the intact side. In unilateral involvement of the vagus, swallowing is ordinarily not impaired, but in bilateral lesions there will be dysphagia and regurgitation of the fluids through the nose. The sensory findings associated with vagus nerve lesion are difficult to test clinically.

The **Accessory nerve** is examined by having the patient turn his head forcibly against the examiner's hand away from the muscle being tested while the sternocleidomastoid muscle is observed and palpated. The patient next forcibly elevates his shoulders while the examiner palpates the action of both upper trapezium and attempts to depress the shoulders. Similarly, the lower portion of the trapezium is tested by having the patient brace the shoulders backward and down. Unilateral paralysis of the trapezium is evidenced by inability to elevate and retract the shoulders and by difficulty in elevating the arm above the horizontal. The trapezium ridge is depressed, exposing the levator scapulae, the scapula appears rotated, the upper end laterally and down, the lower end up and in.

Clinical examination of **Hypoglossal nerve**. The patient is asked to protrude the tongue in the midline and to move it rapidly in and out of the mouth or to wiggle it from side to side (fig. 116). An upper motor neuron lesion may cause some opposite loss of function of the hypoglossal nerve, although each nucleus receives upper motor neuron impulses from both sides of the cortex. A bilateral upper motor neuron lesion will cause the alternate motion rate of the tongue to be slow. When the hypoglossal nucleus or nerve is involved, there will be deviation of the protruded tongue toward the side of the lesion, and atrophy may be manifest by wrinkling of the tongue and loss of the substance on the affected side. The patient is asked to curl the tongue upward, attempting to touch the nose, and downward, to lick off the lower lip. He is instructed to push out the cheek on each side while the examiner tests the strength of the tongue by pushing against it through the bulging cheek. At times direct palpation of the tongue will confirm the suspicion that half tongue is atrophic.



Fig. 116. Examination of Hypoglossal nerve.

Nerves	Symptoms
n. Glossopharyngeal n. Vagus	the voice is hoarse or brassy; dysphagia and regurgitation of fluids through the nose; the testing of taste sensation on the posterior one third of the tongue; ipsilateral paralysis of the palatal, pharyngeal, and laryngeal muscles; bilateral lesion (dyspnea, apnea, periodic respiration – Cheyne-Stokes breathing)
n. Accessory	the patient can't turn his head; the trapezium ridge is depressed; the scapula appears rotated; evidenced by difficulty in elevating the arm above the horizontal.
n. Hypoglossal	deviation of the protruded tongue toward the side of the lesion; dysarthria;

Bulbar syndrome produces the lesion of the n. Glossopharyngeal, n. Vagus and n. Hypoglossal (nucleus or radix).

- dysarthria
- dysphagia
- dysphonia

Pseudobulbar syndrome produces the bilateral lesion of the tr. Cortical-nuclear.

Bulbar syndrome	Pseudobulbar syndrome
gag reflex is absent or decreased	gag reflex is present
the tongue is atrophic	the tongue is not atrophic
the pathologic oral reflexes is absent	the pathologic oral reflexes are present
paralysis is unilateral or bilateral	paralysis is only bilateral
may be (dyspnea, apnea, periodic respiration – Cheyne-Stokes breathing)	–

Alternate intramedullary syndromes of the medulla oblongata:

Jackson's syndrome – Hypoglossal nerve palsy on the site of the lesion and opposite hemiplegia.

Aweli's syndrome – peripheral palsy of the palatal and laryngeal muscles on the site of the lesion and a contralateral hemiplegia.

Schmidt's syndrome – peripheral palsy of the muscles (palatal, laryngeal, tongue and m. sternocleidomastoideus, m. trapezius) on the site of the lesion and a contralateral hemiplegia.

Valenberg – Zakcharchenko' syndrome – on the site of the lesion (nystagmus, Horner's syndrome, peripheral palsy of the palatal and laryngeal muscles, a fallout of sensation on the face for the segmental type, cerebellar ataxia) and a contralateral hemiplegia, hemianesthesia.

**CSF.
Meningeal and Cerebrospinal Fluid's Hypertension
Syndromes.**

Characteristics of Normal Cerebrospinal Fluid

Appearance	clear and colorless
Pressure	Less than 180 mm of CSF.
Cells	0 to 5, usually lymphocytes (more than 8 cells is abnormal)
Total protein	0,15 to 0,33 mmol/ liter
Glucose	2,2 to 3,3 mmol/ liter
Chlorides	120-130 mEq. Per liter

Cerebrospinal fluid's hypertension syndrome:

1. Diffuse headache (in the morning);
2. Dizziness;
3. Nausea, vomiting;
4. Convulsive seizures;
5. Lesion of cranial nerves;
6. Prostration;
7. Protein- cell dissociation;
8. Papilledema;
9. Increased pressure of the cerebrospinal fluid.

Meningeal syndrome

Meningeal syndrome manifests as neck stiffness, Kernig sign, Brudzinski signs (upper, middle and lower ones), Lessage symptom (in children), Bechterev's cheek bone phenomena. Increased intracranial pressure causes bulging of an unclosed anterior fontanelle in children.

The nuchal rigidity - the patient can't to flex the head and place the chin on the chest.

The Kernig's sign supplies the same information as the straight leg raising test is one of the classic sign in neurology for aid in diagnosis of meningitis. With the patient in the supine position the examiner flexes the hip and then extends the knee as far as possible without producing significant pain. Normally the knee can be extended so that the angle between the posterior surface of the thigh and leg is approximately 135 degrees. Results of the test are considered positive if extension of the knee is limited decidedly by

involuntary spasm of the hamstring muscles and, as a rule, if pain evoked.

Brudzinski's signs: 1) when the head is flexed on the chest - the knees are flexion too; 2) when the examiner to do the Kernig's test - the contralateral leg is flex.

Pain phenomena: pain points of Trigeminal nerve, occipital points, pain in eyeballs.

Upon meningeal symptoms general cerebral signs are developed. They are:

- headache (as a result of increased intracranial pressure),
- vomiting, nausea,
- general hyperesthesia,
- hallucination, prostration,
- photophobia,
- phonophobia,
- changes of consciousness,
- psychomotor agitation,
- seizures.

Technics of lumbar puncture.

1) Lumbar puncture should be carried out in an operating room equipped for the purpose and with nursing personnel experienced in the procedure when possible.

2) The patient is placed on his side on a firm but padded table of adequate size and given a small pillow on which to rest his head. The patient is disrobed sufficiently to expose a generous portion of his back from about the midgluteal to the midthoracic region. The area thus exposed is cleansed thoroughly with surgical soap and swabbed in turn with alcohol, ether, and an appropriate antiseptic solution.

3) The widest suitable interspace for puncture is selected by observation and palpation. Usually the fifth lumbar interspace between the sacrum and the fifth lumbar vertebra can be identified. When the doctor injected a local anesthetic agents (may be 1 per cent Lidocaine solution).

4) The lumbar puncture needle should not be inserted sooner than 1 minute after injection of the local anesthetic. During this interval the physician can discuss the procedure with the patient.

5) For obtaining pressures we use an open-end glass manometer of 1 mm. The adapter with manometer attached is connected to the spinal puncture needle, allowing as little cerebrospinal fluid to escape as possible during the procedure. The initial pressure is obtained. If the initial pressure is high (more than 180 mm of cerebrospinal fluid), every effort must be made to be certain that the elevated pressure is not due to extraneous factors. In some cases we must carefully executed Queckenstedt test (The patient is encouraged to remain relaxed and to continue breathing at the normal rate while an assistant compresses both jugular veins firmly with the palmer surfaces of the extended fingers. This is more easily accomplished while the assistant is standing behind the patient. Compression is maintained by the assistant for 10 seconds and suddenly released, after which 10-second intervals are called out to the examiner and the pressure readings made by the latter at 10-second intervals are recorded. An average normal result might be recorded as «11-30-11», indicating that the initial pressure was 110 mm. of cerebrospinal fluid, that it rose to 300 mm within 10 second after compression of the jugular veins, and that it returned to the initial level of 110 mm within 10 seconds after release of compression. A partial dynamic block might be recorded as «11-20-16-14-11', while complete block would be represented by readings of «10-10-10».

6) We take 7 to 10 ml of cerebrospinal fluid for the laboratory tests.

7) After that, the needle is simply withdrawn and a sterile dressing is applied to cover the punctured wound.

CSF changes manifest as:

1. cellular – protein dissociation,
2. protein – cellular dissociation,
3. colour changes,
4. cerebrospinal fluid is cloudy (purulent),
5. the sugar content is decreased;
6. clot formation on standing is observed at tuberculous meningitis.

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Посібник

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