FUNDAMENTALS OF NEUROSURGERY

A Handbook for Medical Students

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Preface

Neurosurgery is one of the medical specialties that has made a substantial progress during the last three decades. It is largely due to the rapid development of contemporary cutting-edge imaging modalities which can diagnose CNS lesions that would otherwise be invisible for the naked eye. The introduction of microsurgical and stereotactic radiosurgical techniques into routine neurosurgical practice has made possible the reach of cerebral and spinal lesions that for a long time have been deemed to be unapproachable. In addition, all this can be achieved at the cost of minimal or no trauma to the adjacent neural tissue.

Despite current advances, we cannot but admit that the outcome of the disease depends on the stage at which it was diagnosed. Delayed diagnosis of CNS lesions can lead to irreversible damage which could not be restored even with the use of appropriate treatment. Therefore, ambulatory practicians should be familiar with the fundamental neurosurgical pathologies in order to be able to properly guide patients to specialized units that can offer up-to-date diagnostic and treatment options.

In this book, we have made an effort to provide basic knowledge of the epidemiology, etiology, clinical presentation, diagnosis and treatment of the versatile neurosurgical diseases. Thus, future clinicians should be able to make correct diagnosis on time.

The authors

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CHAPTER 1 ***B. Kitov, I. Kehayov, E. Hristov***

NEURORADIOLOGY

The CNS can be investigated by the following groups of methods:

- non-invasive – which are based on equipment characteristics;
- invasive – they usually require surgical manipulation prior to instrumental investigation;
- interventional – basically, these can be used as both diagnostic and treatment methods which require surgical manipulation by means of endovascular catheters, balloons, etc.

1. Non-invasive methods

1.1. Conventional methods

These include different plain skull X-ray projections as well as plain X-rays of the spine.

Traditionally, the radiographic investigation of suspected calvarial and intracranial lesions begins with a series of plain skull radiographs. A variety of projections and techniques for plain radiographs relate radiographic features to specific intracranial lesions as well as some systemic disease. The major handicap of radiography of the skull arises from its inability to image intracranial lesions directly. Direct imaging of intracranial lesions is best achieved by the use of computer-assisted imaging techniques, computerized tomography (CT), and magnetic resonance imaging (MRI). These provide the capability of obtaining high-resolution images and specific localization of intracranial lesions. As a result, the interest in plain skull radiographs has recently declined. Its principal application today is as a screening or adjunctive imaging technique in the evaluation of suspected calvarial lesions, such as bony metastatic disease, reactive or inflammatory bone disease, congenital skull deformities, and skull trauma (fractures). For the astute clinician, plain radiographs of the skull also reveal evidence of elevated intracranial pressure or space-occupying lesions, such as thinning of the cortical floor of the sella turcica, displacement of the pineal gland from the midline, widening

of the bony sutures in children, and enlargement of the vascular grooves. The most useful views used to accomplish these objectives include:

- **lateral projection:** Most useful for observation of patterns of intracranial and craniovertebral junction pathology;
- **20° antero-posterior (AP):** Demonstrates the position of the calcified pineal gland;

In addition to the lateral and AP views, the standard skull series typically include:

- **25° postero-anterior (Cauldwell) view**: Useful in demonstration of fractures and lesions of the midfacial sinuses;
- **30° antero-posterior (Townes) view:** Useful in the demonstration of lesions involving the mastoid and petrous bones;
- **submental vertex (Hirtz) view**: May demonstrate abnormalities of the skull base foramina.

Additional applications of the skull x-ray to the neurosurgeon include the evaluation of ventricular or cisternal shunts for continuity and placement and the intraoperative localization of foreign bodies or outlining adjacent structures as is required in pituitary surgery.

Plain X-rays of the spine are useful preliminary investigations for patients presenting with spinal pain. Particular note should be taken of:

- vertebral alignment;
- presence of posttraumatic deformities such as fractures of the vertebral body and/or lamina;
- presence of degenerative disease with narrowing of the neural foramina and spinal canal;
- evidence of metastatic tumour with erosion or sclerosis of the vertebral body, pedicles or lamina;
- enlargement of a neural foramen indicating a spinal schwannoma;
- congenital abnormalities such as spina bifida.

1.2. Computed tomography (CT)

The introduction of CT in 1972 initiated a major evolution in the field of diagnostic neuroimaging with its ability to analyze the absorption of X-rays by all tissues to produce an image of high resolution in the transaxial (horizontal) plane. At the present time, CT is readily available in most major hospitals, making it the most widely used neurodiagnostic modality. CT is accurate, fast, and well tolerated by patients - even those who have difficulty remaining motionless or who require life support systems. The physical principle underlying the CT image is the same as in all radiography - the selective absorption of x-rays (photons) by tissues of different densities. Tissues with greater density – for example, bone - will exhibit more absorption of the X-ray beam and will be displayed on the CT image as areas of high density (white). Tissues with less absorption (attenuation) of X-ray beams will be displayed as areas of lower density (varying shades of gray to black).

Routine examinations of the brain with CT are performed with axial 10-mm slices through the entire brain and the base of the skull. Some specific anatomic regions - such as the posterior fossa, sella, orbits, temporal bone, or paranasal sinuses can be evaluated with thinner sections in axial, axial oblique, and/or coronal imaging planes. It is vital to recognize the advantages and limitations of CT relative to other imaging techniques such as MRI in the evaluation of intracranial pathology. Selection of the study of choice depends on multiple factors, such as history of acute or chronic trauma, the presence of cardiac pacemakers or metallic implants, the monitoring needs of the patients, or the presence of bone pathology versus soft tissue pathology. Generally, CT will be the initial study in patients presenting suspected intracranial hemorrhage that might be the result of head trauma, hypertension, or rupture of an aneurysm or infarction, as well as suspicion of intracranial tumors, either primary or metastatic. CT of the head is the study of choice as well for evaluation of craniofacial trauma to include facial and skull base fractures. Thus, a non-enhanced CT of the head is recommended for evaluation of acute head trauma, subarachnoid haemorrhage, acute stroke, follow-up of hydrocephalus or shunt malfunction, and for general evaluation of patients with dementia. A CT of the head should be obtained with

intravenous contrast enhancement as a screening study for possible metastases, headaches of unknown etiology without neurological deficit, suspected aneurysms or vascular malformations, intracranial infections or abscess, or primary and metastatic brain tumors. CT supplemented by intrathecal contrast is excellent in evaluating CSF fistulas. CT of the head with 3-dimensional reconstruction is the study of choice for preoperative and postoperative evaluation of facial anomalies and craniosynostosis.

Nevertheless, there are certain limitations of CT. Routine CT is suboptimal for evaluation of the prepontine and cerebellopontine angle cisterns unless special angulation is used and thin sections are obtained. This is due to the absorption of the X-ray beam by the thick petrous bones ("Hounsfield artifact"). The evaluation of the sellar region by CT should be performed in the coronal plane with intravenous contrast; however, the patient's position for the study may be awkward and uncomfortable, resulting in inconsistent image quality. Therefore, MRI is the study of choice for suspected lesions in the region of the sella. Indirect and direct signs of intracranial pathology can be seen on CT:

A/ Direct CT signs:

- hyperdense (white) lesions – with higher density than the normal brain tissue: acute intracranial hematomas, metallic foreign bodies, calcifications, some meningiomas, etc.
- hypodense (dark or black) lesions – with lower density than the normal brain tissue: brain oedema, ischaemic lesions, intracranial cysts, etc.
- isodense (gray) lesions – their density is similar to that of the normal brain, for example, vestibular schwannomas.

B/ Indirect CT signs:

- compression, deformation, dislocation or amputation of the ventricles;
- presence of focal or diffuse brain oedema;
- presence of dilatation of the ventricular system typical for hydrocephalus.

In cases of suspected brain tumor, the CT scanning should be enhanced by the application of contrast agent. It gives a clear

delineation of the pathological process from the surrounding brain oedema and makes possible the visualization of isodense lesions which can be omitted during plain CT scanning (Fig. 1.1).

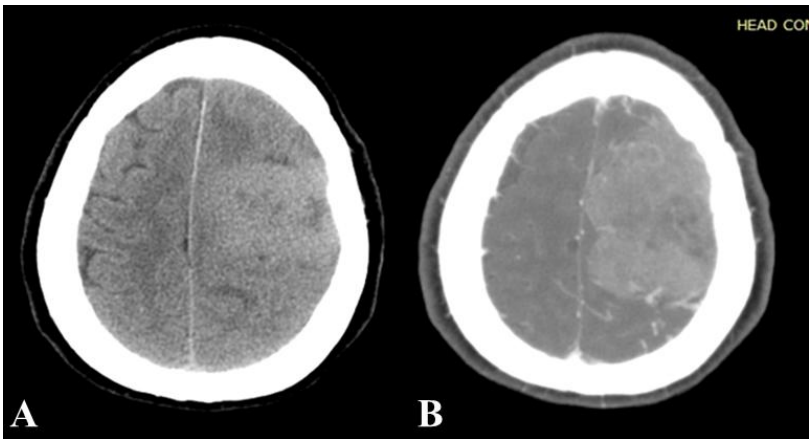

Fig. 1.1. CT image of convexity meningeoma:
A/ without contrast enhancement; **B/** with contrast enhancement

1.3. Magnetic Resonance Imaging (MRI)

The next major evolution in neuroimaging following CT in 1972 occurred with the clinical application of magnetic resonance imaging (MRI) in 1982. MRI is a noninvasive computer-assisted imaging technique that differs from CT in the type of energy used. The MRI is based on measuring the phenomenon of nuclear magnetic resonance of the hydrogen atom in various tissues. Magnetic resonance imaging uses a strong magnetic field and the application of radiofrequency pulses to generate 2-dimensional or 3-dimensional images. Nuclei of certain atoms, particularly hydrogen, respond to a strong magnetic field by aligning with or against the longitudinal axis of that magnetic field. This process is called magnetization. The number of hydrogen atoms aligned in the direction of the magnetic field is slightly greater than the number aligned against it, producing a net magnetization in the direction of the magnetic field. The magnitude of this net magnetization is related to the number of hydrogen nuclei in each volume of tissue (proton density). The direction of the net magnetization may be altered by the addition of energy in the form of a radiofrequency (RF) pulse of appropriate frequency. This extra energy flips the magnetization vector away from its alignment with the primary magnetic field. When the RF pulse is terminated, the

hydrogen protons will begin to realign in the direction of the external magnetic field, releasing the excess energy initially used to deflect them from alignment. The rate at which this realignment occurs depends on the rate that the added energy is released to the surrounding environment. This process is called longitudinal relaxation. The time required for 63 percent of the magnetization vector to return to alignment with the external magnetic field is designated ‘T1’, or longitudinal relaxation time. A second component of relaxation occurs in the transverse plane around the axis of the magnetic field. The magnetization vector in the transverse plane is the sum of many nuclei rotating at slightly different frequencies, which are forced together by the application of the external RF pulse (coherence). The net magnitude of the transverse vector and its signal strength diminishes as the nuclei fan out within the transverse plane (loose coherence). The time required for 63 percent loss of the transverse coherence is designated ‘T2’, or transverse relaxation time.

Different tissues will exhibit different T1 and T2 relaxation patterns, making their magnetic behavior different enough to be recognized as separate entities in the MR image. The excess energy released by the protons as they realign with the magnetic field is radiated to the environment as a radio frequency "signal." The amount of signal (signal intensity) emitted per unit of volume will be correlated to a gray scale, where high signal intensity will be white and absence of signal will be black. The signal intensity of a tissue is related to its T1 and T2 relaxation times and its proton density. MR images can be obtained in such a way as to favor the demonstration of structures with predominant T1 or with T2 characteristics.

T1-weighted images produce a strong MRI signal that results in anatomic images (Fig. 1.2A). In a T1-weighted image, cerebrospinal fluid (CSF), cortical bone, air, and rapidly flowing blood will have negligible signals, appearing dark. The gray matter and white matter will show different shades of gray, with the gray matter appearing slightly darker. Fat is intensely bright on T1-weighted images, which is an advantage when outlining orbital fat or the epidural space or marrow in the spinal column. Subacute and chronic hematomas are seen as high signal intensity. The abundant signal of the Tl-weighted image is ideal for demonstrating detailed intracranial or spinal

anatomy, for example, in evaluating the cerebellopontine angle cistern or the pituitary fossa. This is due to its high contrast between normal structures and the adjacent darker CSF. T1-weighted images, however, are relatively insensitive to small changes in the water content of tissue. Therefore, small brain lesions that do not cause anatomic distortion are often invisible.

T2-weighted images are much more sensitive in the detection of small changes in the water content of the tissue examined (Fig. 1.2B). Regions of increased water content (edema) are imaged as regions of high signal intensity superimposed on a darker background. Because of the lack of a strong background tissue signal, T2-weighted images do not display the anatomy as sharply as the Tl-weighted image does. Nevertheless, the high sensitivity of T2-weighted images for the detection of changes in tissue water content is the primary reason for the increased sensitivity of MRI over CT in the detection of brain and spinal cord pathology. In T2-weighted images, the dominant tissue producing the highest signal intensity will be bulk water, making CSF the dominant bright signal factor in comparison to gray and white matter. Cortical bone, air, and rapidly flowing blood still present negligible signals (black). Fat in T2-weighted images presents lower signal intensity than in T1-weighted images. Areas of demyelination, edema, infarction, or tumor infiltration will have higher water content (more hydrogen atoms) and will produce higher signal intensities than the surrounding tissues. A definite advantage of MRI over all other imaging modalities is its ability to image multiple direct planes (axial, sagittal, coronal, or any degree of oblique projections) without a change in the patient's supine position. The absence of signal from surrounding bone allows excellent anatomic detail of structures adjacent to bone, both intracranially and throughout the spinal canal. Most MRI examinations of the brain include sagittal, coronal, and axial projections. The axial images are most commonly obtained to evaluate the brain. The coronal images are most useful in evaluating sellar and parasellar regions. With both, the axial or the coronal images, side-to-side comparison is possible. Direct sagittal images are extremely useful when evaluating midline pathology of the brain, such as sellar tumors, pineal masses, brainstem tumors, vermian atrophy, obstructive hydrocephalus, or congenital malformations. They are also

helpful in visualizing the location of any other internal pathology in the lateral view.

Gadolinium (Gd), a heavy rare-earth element, is being used in special solutions as the intravenous contrast agent for MRI of the central nervous system (CNS). Although free Gd is toxic, its compound chelated to diethylenetriamine pentaacetic acid (DTPA) or other chelating agents exhibits "paramagnetic behaviour". When deposited within tissue, this compound greatly decreases the T1 relaxation time of the surrounding tissue, resulting in high signal intensity on T1-weighted images. Because of the nature of the bloodbrain barrier, Gd will only move outside of the intravascular space in areas of absence or breakdown of the blood-brain barrier. Damage to the blood-brain barrier is a nonspecific alteration that occurs with pathology within the brain, as seen with tumors, infection, and infarction. Gadolinium-DTPA, the current agent employed for contrast MRI, has been shown to be safe, with very few reported allergic reactions in over 5 million administered doses. The use of paramagnetic contrast increases the sensitivity of MRI for detection of specific disease processes, particularly small tumors. The pattern of contrast enhancement in MRI allows for better tissue characterization of the visualized lesions. Pituitary gland, infundibulum, choroid plexus, pineal gland, and dural reflections lack a blood-brain barrier and will normally enhance with Gd. Slowly flowing blood within the cavernous sinus or in the cortical veins will also exhibit normal enhancement.

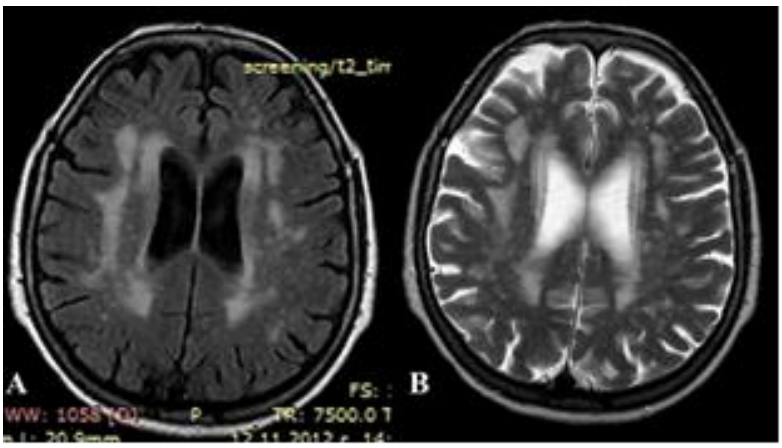

Fig. 1.2. MRI of the brain:
A/ T_1-wieghted image; **B/** T_2-weighted image

Contraindications to MRI include:

- the presence of ferromagnetic materials in soft tissues of the body, such as old intracranial aneurysm clips or intraocular metallic foreign bodies that could move in the fluctuating magnetic field and damage structures on which they are located;
- cardiac pacemakers that could malfunction in the magnetic field or large metallic implants that may become heated by magnetic induction;
- since the effect of high magnetic fields and radiofrequency energy on fetuses has not been determined, pregnant patients are generally excluded at the present time;
- patients with claustrophobia may not be able to remain in the MR unit for the required time. Quality MRIs require patients to remain motionless for 10 to 15 min;
- the magnetic environment restricts the use of conventional electronic monitoring devices. Therefore, critical or unstable patients are difficult to monitor and require special attention or equipment while undergoing MRI.

2. Invasive methods

2.1. Cerebral angiography (A^o). Contemporary cerebral angiography employs electronic digital imaging with computerized subtraction (DSA) and/or biplanar arteriography and fluoroscopy, an automatic contrast injector, and automatic film changers. The Seldinger's technique or a modification thereof, in most cases provides the safest means of selective arterial injection; however, on rare occasions a retrograde brachial or direct carotid injection may be necessary to obtain a complete study (Fig. 1.3.). Physicians requesting or performing cerebral angiography should be familiar with the normal cerebrovascular anatomy, commonly occurring anatomical variations, and typical angiographic patterns, as well as appearance of various lesions. Cerebral angiography can be used for:

- high vascularized tumors (meningiomas, hemangioblastomas, etc.);

- tumors bordering major intracranial arterial and/or venous vessels (parasagittal or parasellar meningiomas, etc.);
- vascular brain diseases (cerebral aneurysms, AVMs, thrombosis or stenosis of the Circle of Willis vessels), as well as for post-operative follow-up (Fig. 1.4);
- some patients with epilepsy;
- diagnosis of brain death when organ explantation is needed.

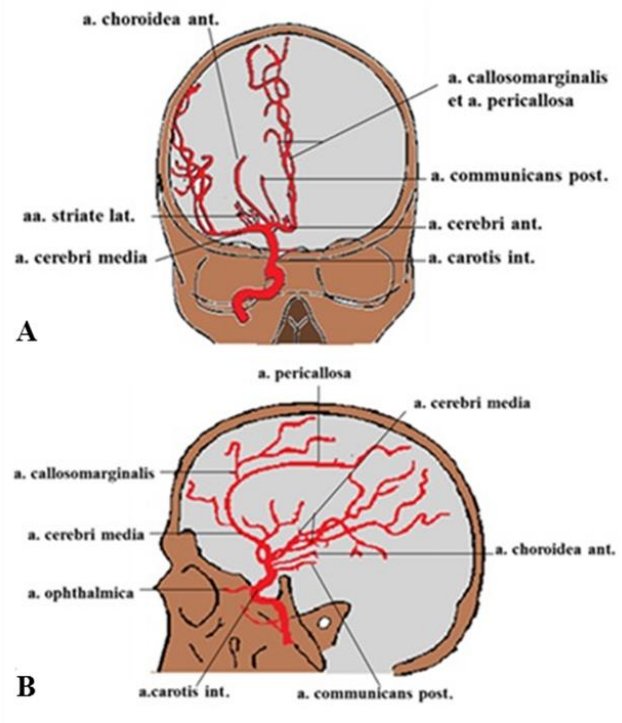

Fig. 1.3. Schematic drawing of the normal brain vesels on the digital substraction cerebral angiography: **A/** AP projection; **B**/ lateral projection

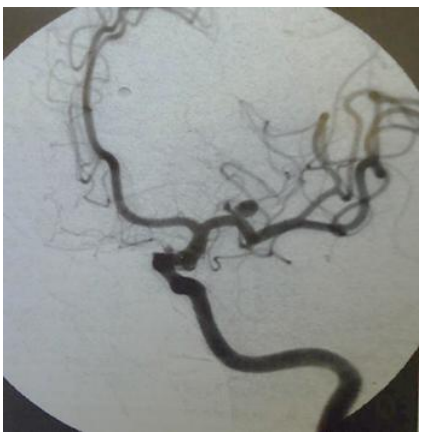

Fig. 1.4. DSA – middle cerebral artery aneurysm

2.2. Myelography. This method includes injection of contrast agent inside the spinal subarachnoid space. It is used for investigation of the spinal canal and its content (Fig. 1.5). The contrast agent is injected by lumbar puncture and/or lateral cervical puncture at C1-C2 level under radiographic guidance.

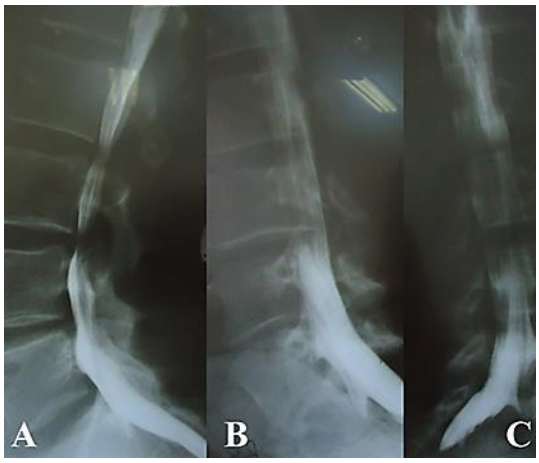

Fig. 1.5. Lumbo-sacral myelography (radiculosaccography):
A/ lateral projection; **B/** and **C/** left and right oblique projections

Indications to myelography are:

- spinal tumors;
- spinal injuries;
- disc herniations;
- spinal stenosis;
- spinal parasitic disease;
- spinal infection;
- spinal anomalies (meningocele, myelomeningocele, diastematomyelia, etc.).

3. Interventional methods

3.1. Endovascular embolization is used for the treatment of AVMs and some highly vascularized brain tumors. Pathologic vessels are embolized by means of special embolic agents depending on the lesion localization and vascularity.

3.2. Endovascular angioplasty and recanalization is performed by means of balloon catheters and stents. It can be used for

arterial stenoses. Fibrinolisis with Ateplase, Urocinase or Streptocinase can also be performed in cases of arterial thrombosis.

3.3. Endovascular coiling includes the insertion of platinum coils into the aneurysmal sac which are detached by induction of low electrical current (0.5mA). The coil induces thrombosis of the aneurismal sac. Endovascular coiling is a contemporary alternative to open microsurgical clipping of aneurysms.

3.4. Endovascular balloonization is usually used for the obstruction carotid-cavernous fistulas. It includes the insertion of a special catheter and the balloon is placed at the site of the pathologic vascular shunt between the cavernous portion of the carotid artery and the cavernous sinus. The elimination of the shunt is verified by intraoperative angiography.

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CHAPTER 2 ***B. Kitov, S. Raykov, A. Petkova***

HEAD INJURIES

Introduction

The severity, mortality and disability rate in head injured patients is high which poses substantial problems for the medical and state authorities. Head injuries commonly affect young and employed people in their active age. The outcome from sustained head injury depends on the following predictive factors:

- severity and localization of the head injury;
- presence of associated injuries (trauma affecting other organs and systems besides head injury - thoracic and abdominal trauma, limb fractures, etc.);
- presence of combined injuries as a result of damage caused by several destructive factors (mechanical trauma in combination with radiation, thermal, chemical and other influence).
- quickness and quality of first aid;
- availability of appropriate imaging studies;
- availability of high-quality treatment and reanimation.

Epidemiology: In Europe, more than 100 000 people die and 1 000 000 get injured in road accidents each year. In 70% of cases the mortality is caused by head injuries.

In USA, head injury is a major cause for death in people below the age of 45 and third as a reason for death in the age group 45-60. 75% of the mortality in road accidents is caused by head injuries.

In Bulgaria, 1 000 people die in road accidents annually and in most cases there is presence of head trauma.

Etiology: Head injuries occur in:

- road accidents – 55-65%;
- falls from height – 20% (common in children and elderly);
- employment accidents;
- sports trauma;
- assaults.

Classification: Head injuries can be divided into two major groups depending on the damage of the skin and soft tissues covering the skull:

- **opened** – there is skin wound;
- **closed** – there is no skin and soft tissue wound.

Injuries to the skull coverings (with or without brain damage):

1. **Closed injuries of the skull coverings:**
   - **cephalhematoma** – accumulation of blood between the eriostemon and the skull (Fig. 2.1);
   - **subfascial hematoma** – common in infants and early childhood. These hematomas usually subside spontaneously;
   - **caput succedaneum** – due to trauma during labour. It is accumulation of blood and interstitial liquid in the subcutaneous tissues affecting larger area.

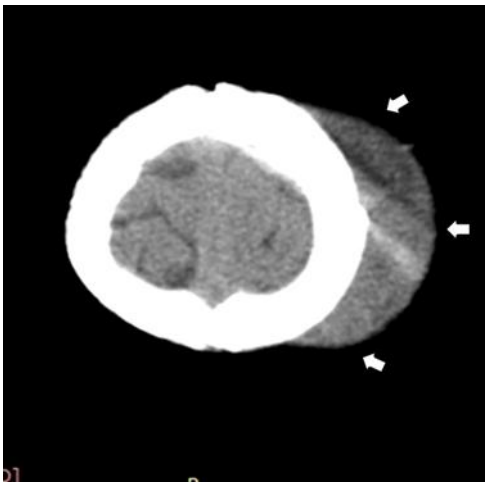


Fig. 2.1. CT of cephalhematoma

2. **Opened injuries of the skull coverings:**
   - **vulnus scissum** – due to cuts with sharp objects;
   - **vulnus punctum** – due to vertical penetration from sharp objects;
   - **vulnus lacerocontusum** – due to hit with flat and dull objects;
   - **vulnus morsum** – as a result of bites;
   - **vulnus sclopetarium** – gunshot wounds.

Types of opened injuries:

- **penetrating** – there is tear of dura mater;
- **non-penetrating** – dura mater is spared.

Wound contamination is common and poses a high risk of infection which necessitates wound revision and antiseptic lavage.

Classification of closed head injuries:

- **cerebral concussion (commotio cerebri) –** mild head injury;
- **cerebral contusion (contusio cerebri)**:
  - mild – 66%;
  - moderate – 24%;
  - severe – 10% (with primary or secondary brain stem contusion).
- **cerebral compression (compressio cerebri):**
  - epidural (or extradural) hematoma
  - subdural hematoma
  - intracerebral hematoma
  - subdural hydroma
- **skull fractures:**
  - **fractures of the cranial vault:**
    - ✓ linear – single and multiplex
    - ✓ comminuted without fragment dislocation
    - ✓ comminuted with fragment dislocation (depression fractures)
  - **skull base fractures:**
    - ✓ anterior cranial fossa
    - ✓ middle cranial fossa
    - ✓ posterior cranial fossa
    - ✓ combined - affecting more than one fossa
  - **mixed fractures** (of the skull vault and base)

Cerebral concussion (Commotio cerebri)

It is the mildest grade of brain injury without morphological changes to the brain seen on CT and MRI. The damage is only functional. Bed rest for 7 to 20 days is usually sufficient treatment.

Clinical presentation: There are signs of increased intracranial pressure without focal neurological deficit. Loss of consciousness is short within seconds or several minutes due to temporary interruption

of impulse transmission from the brain stem reticular formation to the cerebral cortex. Usual complaints include somnolence, fatigue, emotional and vegetative instability which are transitory. Mild to moderate headache is usually present and deteriorates with physical and mental loading. Nausea, vomiting and dizziness are also commonly present. Retrograde and anterograde amnesia is short-lasting and covers a period of 1 or 2 hours.

Diagnosis: The diagnosis is made upon the typical clinical complaints. All imaging studies are negative. Follow-up for a period of 24 hours is necessary.

The clinical symptoms of cerebral concussion should be monitored for a period of 24 hours.

Treatment: Patients should keep bed rest regimen for 7 to 14 days and avoid the intake of excitatory substances (alcohol, coffee, coke, smoking, etc.) Symptomatic agents are also prescribed (pain-killers, anti-emetic drugs, etc.).

Cerebral contusion (Contusio cerebri)

It is characterized by morphological changes to the brain seen on CT and MRI. Their localization and severity depends on the biomechanics and complex pathological mechanisms of the head injury.

Biomechanics of cerebral contusion

It depends on the mechanism of impact of the traumatic agent which determines the localization and severity of the contusion. According to the mechanism of impact traumatic brain injuries can be divided into:

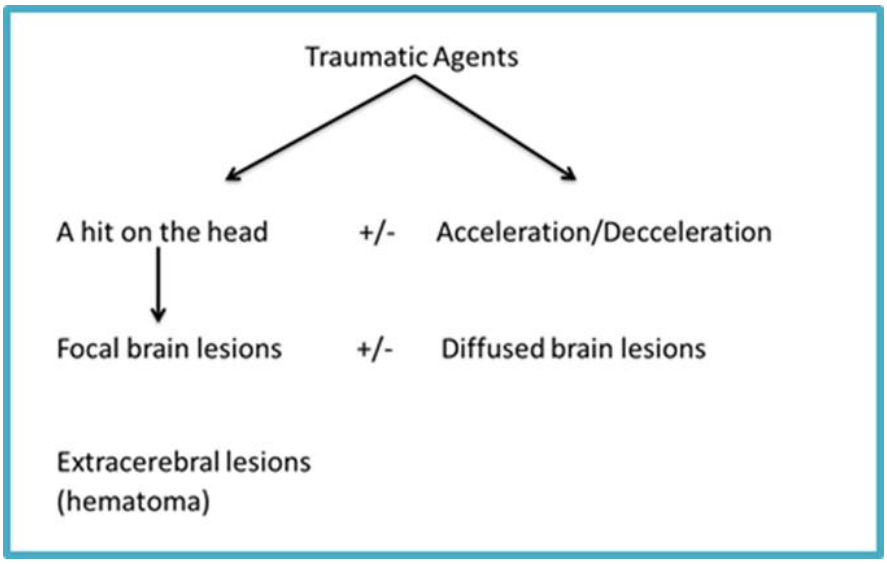
- **static** – the head is stationary
- **dynamic** – the head is in motion. Dynamic injuries occur either as a result of acceleration (the head is set into motion by the traumatic agent) or deceleration (sudden halt of the moving head).

Mechanical trauma of the head can cause local and general deformation of the skull depending on the characteristics (size and

weight), speed and direction of action of the traumatic agent. If the speed exceeds the bone elasticity, the skull fractures. The kinetic energy of the impact is transmitted to the underlying brain thus creating percussion wave and "cavitation" phenomenon resulting in the setting in motion of the brain parenchyma. The occurring linear and rotational acceleration/deceleration (Table 2.1.) can lead to:

- **focal contusion at the site of impact or to the opposite site of impact (so called "coup" and "contre coup" injuries)** (Fig. 2.2). They are usually bilateral affecting the basal surfaces of the frontal and temporal lobes.

Table 2.1. Biomechanics of head injury



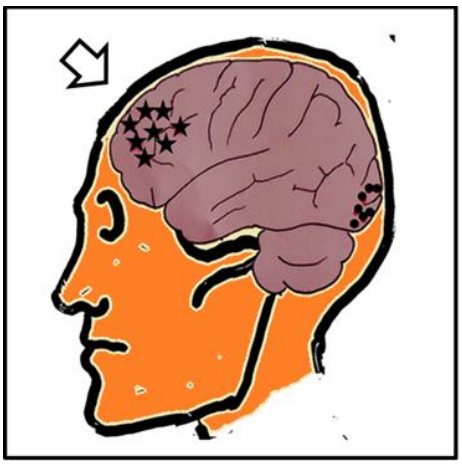


Fig. 2.2. Focal lesions: 1. At the site of impact (Coup)(★); 2. At the opposite site (contre coup) (●)

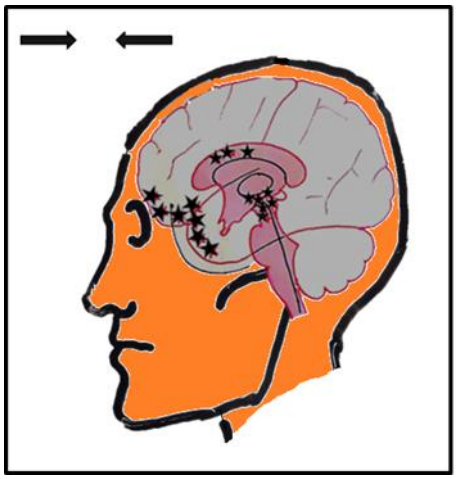


Fig. 2.3. Linear lesions: acceleration/ deceleration: 1. Basal focal lesions (★) 2. Diffuse axonal injury (★)

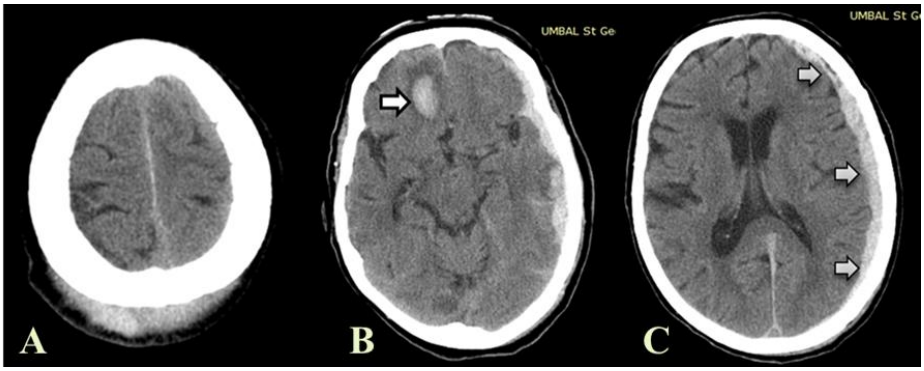


Fig. 2.4. CT images illustrating the „contre coup“ mechanism: **A/** subcutaneous swelling in the occipital region – at the site of the hit; **B/** intracerebral hematoma in the right frontal lobe; **C/** left-sided acute subdural hematoma;

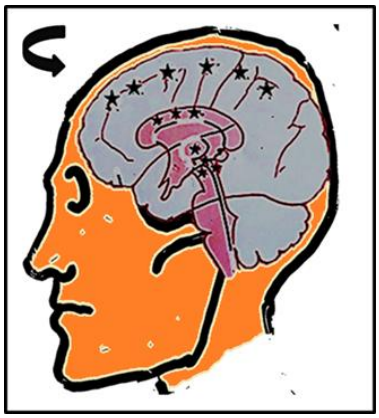


Fig. 2.5. Rotational acceleration/ deceleration - superficial lesions and diffuse axonal lesions(•)

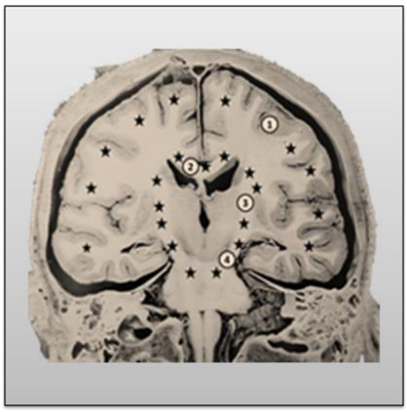


Fig. 2.6. Localization of diffuse axonal injuries: 1. Subcortical region; 2. Corpus callosum; 3. Capsula interna; 4. Pons cerebri

- **tearing, shearing and twisting of axons at the border between gray and white mater.** The most common localization is the subcortical zone of corpus callosum, basal ganglia and upper midbrain. This type of injury is termed diffuse axonal injury (DAI) or shear injury (Fig. 2.3, Fig. 2.5 and Fig. 2.6).

Pathology of cerebral contusion

Aforementioned, the mechanical factor causes brain injury (focal and diffuse) defined as primary and irreversible. Secondary brain injury results from the primary brain injury and can be reversible. Secondary injuries develop immediately after the initial trauma and in most cases determine the clinical course and disease outcome. The

progression of secondary injuries depends on the adequate blood circulation and pulmonary ventilation.

Classification of primary brain injury

1. **Focal injury**:
   - **cerebral contusion**
     - ✓ cortical contusion;
     - ✓ basal ganglia contusion;
     - ✓ primary brainstem contusion;
     - ✓ hemorrhages (subarachnoid, parenchymal, intraventricular).
   - **extracerebral hematomas**
     - ✓ epidural;
     - ✓ subdural.
   - **primary injury of cranial nerves**;
   - **diffuse axonal injury (DAI)**.
2. **Secondary brain injuries** develop on two major levels:
   - **local** – in the region of the focal brain contusion and its periphery;
   - **generalized –** changes in the intracranial pressure and cerebral circulation.

Processes accelerating secondary injuries on a local level:

Zones of neuronal destruction, breakdown of blood-brain barrier, intra-parenchymal hemorrhages and micro-thromboses are observed in the region of focal brain contusion. It leads to a release of large amounts of 'free radicals' (O^{2-}, OH^{-}). They cause damage to cell membranes and increase in intra-cellular calcium level which activates cellular phospholypases causing membrane destruction. This process predominantly affects glial cells and together with brain ischemia leads to hyperhydratation of the intra-cellular space, termed as cytotoxic edema**.**

Breakdown of microcirculation: Initially, it is damaged by the compression caused by the intracranial hemorrhages and the edema of endothelial and peri-capillary glial cells. Subsequently, brain ischemia leads to increase in lactate levels. The acidosis in the extra-cellular space causes vasodilation and breakdown in cerebral blood flow auto-regulation. The destruction of cell membranes causes a release of prostaglandins and other vaso-constrictive substances which additionally disturbs microcirculation.

Processes accelerating secondary injuries on a general level:

Increased intracranial pressure **(ICP):** When at rest, the normal ICP ranges between 8-12 mmHg. The three major components of the intracranial space are the brain parenchyma (1300 – 1500 cc), the cerebrospinal fluid or CSF (100 – 150 cc) and the circulating blood (100 – 150 cc). The last two components represent the 'cushions' of the system which by means of changing their volume support normal ICP – a phase of compensation.

When ICP exceeds 20 mmHg, the decompensation stage develops.

Etiology of increased ICP in brain contusion:

- **brain edema**: mixed vasogenic and cytotoxic edema develop at different stages of the disease;
- **impairment of cerebral vascular auto-regulation - vasoplegia**: Vasodilation leads to increased intracranial blood volume, thus increasing the ICP. This process can be focal, surrounding the contusion focus, but later becomes generalized. It is aggravated by acidosis or acute impairment in cerebral vascular auto-regulation due to damage of the cardio-vascular center in the brain stem;
- **intracranial hematomas** (epidural, subdural, intracerebral).

Increased ICP and brain dislocation phenomena: Brain structures can get herniated in the normal intracranial anatomical apertures:

- **subfalcine herniation:** medial aspect of the frontal lobes herniated below falx cerebri (Fig. 2.7). Clinically, it presents with depressed level of consciousness;
- **tentorial herniation:** The medio-basal surface of the temporal lobe (gyrus parahippocampalis) herniates below incisura tentorii known as Bichat's cleft. This type of herniation causes direct midbrain compression (Fig. 2.7 and Fig. 2.8). The symptoms include:
  - ocular nerves palsies and ophtalmoplegia;
  - motor weakness due to compression of the cerebral peduncles;

- nuchal rigidity;
- impairments in termoregulation and vascular tone regulation;
- bradycardia, coma and death.

- **tonsilar herniation:** Insertion of the cerebellar tonsils in foramen occipitale magnum which causes direct compression of medulla oblongata (Fig. 2.7). Clinical presentation includes:
  - cerebellar symptoms;
  - bulbar paralysis and/or pyramidal signs;
  - breathing and cardio-vascular impairment;
  - coma and death.

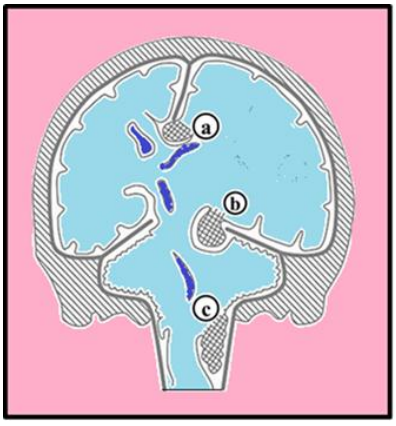


Fig. 2.7. Types of brain herniations: **a/** subfalxine; **b/** tentorial (temporal); **c/** tonsilar

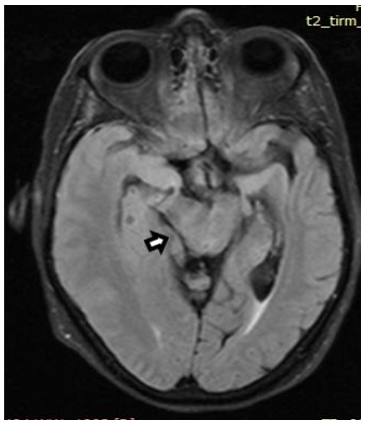


Fig. 2.8. MRI image of temporal herniation: the medial part of the temporal lobe compresses the brainstem

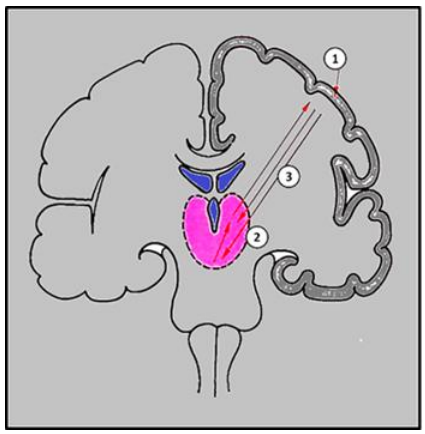


Fig. 2.9. Centers and pathways of consciousness
1. Cerebral cortex;
2. Diencephalic and mesencephalic centers;
3. Interhemispherial connections

Clinical presentation of cerebral contusion

Depressed level of consciousness due to increased ICP and focal neurological deficit are the most common symptoms. Altered consciousness presents with inadequate reaction to internal and external stimuli. It can be caused by:

- **diffuse axonal injury** which leads to interruption of neural transmission between the brainstem and cerebral cortex. This type of impairment is observed immediately after the sustained trauma (Fig. 2.9);
- **increased ICP** due to intracranial hematoma and brain edema.

Grades of altered consciousness: Currently, we use the Glasgow Coma Scale to assess the level of depressed consciousness. The score is formed by summing the sub-scores from the three major types of response (Table 2.2).

Table 2.2. Glasgow Coma Scale (GCS)

Type of response	Reaction	Points
Eye opening	Spontaneous	4
	To speech	3
	To pain	2
	No response	1
Best motor response	Obeys command	6
	Localize to pain	5
	Flexion to pain (withdrawal)	4
	Abnormal flexion	3
	Extension to pain	2
	No response	1
Best verbal response	Oriented, adequate	5
	Confused	4
	Inappropriate	3
	Incomprehensible sounds	2
	No response	1
Total		**3 to15**

The total score on the GCS is used to assess the severity and prognosis of the head injury.

Table 2.3. Severity of cerebral contusion according the the GCS score

Severity - grade	Points
Mild	12 - 15
Moderate	9 - 12
Severe	3 - 8

Table 2.4. Outcome depending on the GCS score

GCS score	3 p.	4 p.	5 p.	6 p.	7 p.
Mortality	65%	45%	35%	24%	10-15%

Alterations in the state of consciousness should be assessed after securing patent airways, adequate breathing and stable circulation and before the administration of sedatives.

Focal neurological deficit: It can be extremely diverse depending on the localization of the lesion and includes different neurological signs (impairment of cranial nerves, visual and speech disturbances) or hemi-type syndromes (motor and sensory), syndrome of meningeal irritation, etc. Epileptic seizures are also common. The presence of neurological deficit after head trauma presumes cerebral contusion. The absence of neurological deficit does not rule out cerebral contusion because the lesion can be located in functionally ineloquent or 'silent' brain zones.

The presence of focal neurological deficit following head injury presumes the diagnosis of cerebral contusion. The absence of focal neurological deficit does not rule out cerebral contusion because the lesion may be located within functionally non-eloquent brain areas.

Cases of severe cerebral contusion which present initially with signs of brainstem injury are grouped separately. In these cases, brainstem signs and symptoms result from direct brainstem impairment (Fig. 2.10).

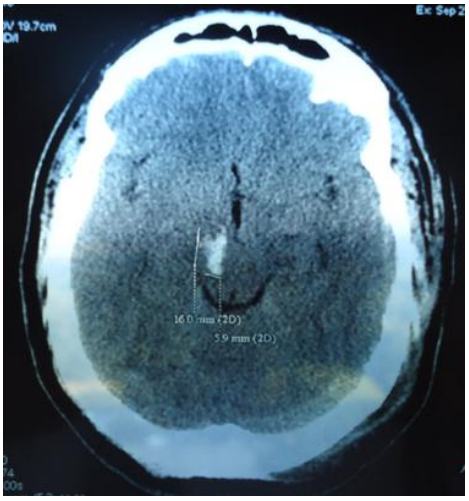


Fig. 2.9. CT of brainstem (midbrain) contusion

Diagnosis

- **skull X-rays** can demonstrate skull fractures and eventually pneumocephalus as a result of skull base fracture;
- **computed tomography** is a diagnostic procedure of choice. It can demonstrate single or multiple lesions - their size and localization (cortical, sub-cortical or brainstem) as well as the presence of dislocation phenomena and brain herniation.

Treatment of cerebral contusion

The treatment aims at limiting the secondary brain injuries such as cerebral edema, brain ischemia and increased ICP.

Common measures

- cardio-pulmonary resuscitation secures patent airways, adequate breathing and stable circulation. Blood gas analysis is used to assess acidosis and blood oxygenation rate;
- stabilization of hemodynamics - mean arterial blood pressure should be within 90-110 mm Hg to secure adequate cerebral perfusion;
- elevation of head and thorax at 30^{o} alleviates cerebral venous drainage and decreases ICP.

Control and correction of other vital parameters:

- lowering body temperature, eventually support mild hypothermia;
- central venous pressure within 4-6 mm Hg;

- CBC: hematocrite 30-40%, serum osmolality 300-320 mOsm/kg.

Brain edema treatment:

- moderate hyperventilation - causes cerebral vasoconstriction;
- diuretics:
  - osmotic (mannitol 0,5-1g/kg, sorbitol 40% - 1-2 ml/kg i.v.;
  - non osmotic **(**furosemide – suppresses CSF production).
- steroids are not as efficient in influencing edema as in brain tumors but can be used - Dexamethasone 80-120 mg i.v. initially x 8 mg every 2h gradual lowering of dose afterwards;
- antioxidants - vit. "C", vit. „E";
- cerebral protection – barbiturates (depress cerebral metabolism and oxygen demands making neurons more resistant to ischemia).

Treatment of epileptic seizures and status epilepticus

- barbiturates (Phenobarbital, Thiopental).

Support of other systems:

- **urinary system** – per-urethral catheter is mandatory to control adequate diuresis. Prophylaxis of urinary infection is also compulsory;
- **gastrointestinal system** – insertion of naso-gastric tube in comatose patients is necessary to secure adequate feeding. GIS complications are common in severe head injury - 17-20% (erosions, stress-ulcers, etc.);
- **pulmonary system** – prophylaxis of aspiration, hypostatic lung infection and atelectasis;
- **skin** – routine care, decubitus treatment.

Cerebral compression (Compressio cerebri)

Classification:

- **traumatic intracranial hematomas:**
  - epidural hematoma;
  - subdural hematoma;
  - intracerebral hematoma.
- **traumatic subdural hydroma;**
- **traumatic pneumocele.**

Epidural hematoma

Epidural hematoma is a blood collection between dura mater and lamina interna of the skull bone. Anatomically, the epidural space does not exist due to the tight adherence of dura mater to the skull bone. Accumulation of blood in this space is possible when there is detachment of the dura mater due to skull fracture and subsequent bleeding. Torn branch of the middle meningeal artery caused by skull fracture is the source of bleeding in more than 90% of cases which determines the most common localizations (Fig. 2.11, Fig. 2.12 and Fig. 2.16). In some cases the bleeding results either from laceration of dural sinuses and diploic veins or from torn pachionnian granulations. Usually, epidural hematomas have acute clinical onset and only rarely they are chronic.

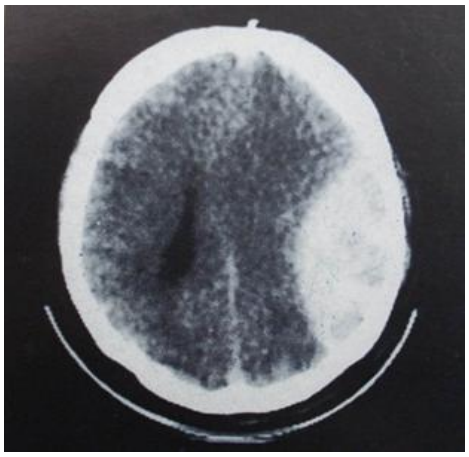


Fig. 2.11. CT image of epidural hematoma

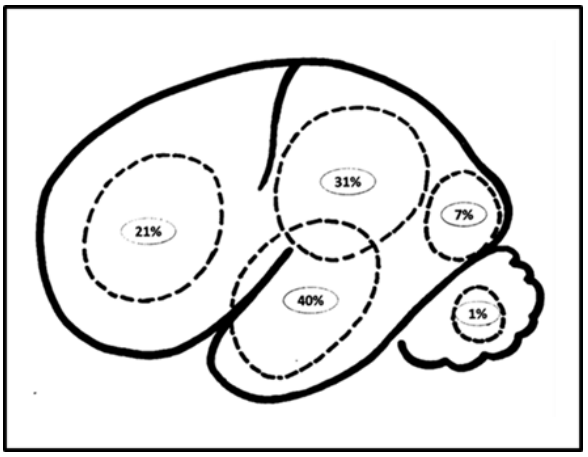


Fig. 2.12. Localization of epidural hematomas (Sichez et al., 1984)

Clinical presentation

A/ Typical clinical presentation:

- sustained head injury with short loss of consciousness;
- lucid interval when the patient is fully alert and shows no neurological deficit;
- in short term, usually hours, a depression of consciousness and neurological deficit develops;
- skull X-rays demonstrate the presence of skull fracture crossing a branch of the middle meningeal artery.

B/ Atypical clinical presentation:

- there is no loss of consciousness;

- lucid interval is very short or absent;
- symptoms of increased ICP can last longer in fronto-polar and vertex localizations;
- skull X-rays does not demonstrate skull fracture.

Subdural hematoma

Subdural hematoma is accumulation of blood between dura and arachnoid matter (Fig. 2.13). Subdural space is anatomically present which determines the larger area and volume of this type of hematoma (300-400cm^3). Usually, the source of bleeding is either a torn bridging vein (passing from the brain surface to a dural sinus) or a torn cortical artery in a zone of cerebral contusion. According to the period of formation and time of diagnosis subdural hematomas can be:

- acute – within first three days;
- subacute – from 4th to 20th day;
- chronic – after 21st day- there is capsule formation. (Fig. 2.13).

A/ Clinical presentation of acute subdural hematoma

- commonly combines with cerebral contusion;
- there is progression of headache, deterioration of consciousness and focal neurological deficit depending on the localization.

B/ Clinical presentation of chronic subdural hematoma

- patient has sustained head injury which he/she does not remember;
- mild to moderate headache;
- mild neurological deficit.

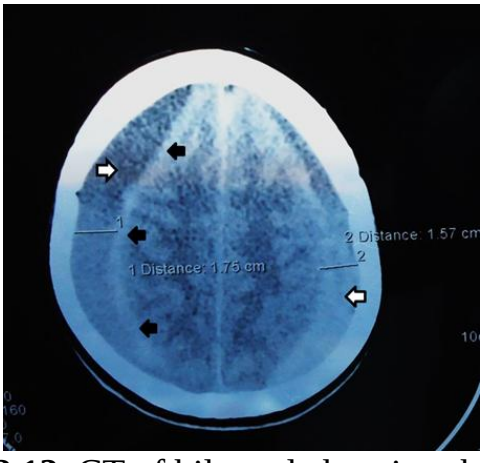


Fig. 2.13. CT of bilateral chronic subdural hematomas(⇨)hemolised blood;(➧) capsule

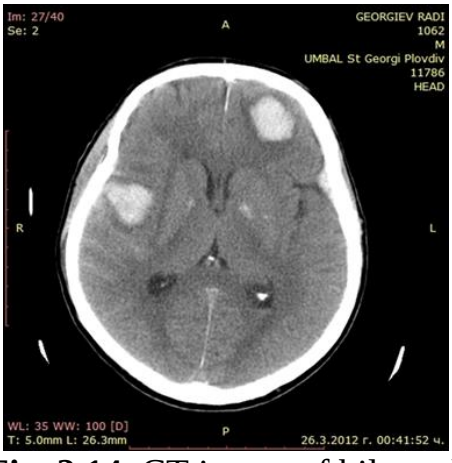


Fig. 2.14. CT image of bilateral intracerebral hematomas

Intracerebral hematoma

Intracerebral hematoma is an accumulation of blood in the cerebral parenchyma of the brain hemispheres and rarely in the cerebellum and brainstem (Fig. 2.14, and Fig. 2.15). The source of bleeding is the small parenchymal arterial and venous vessels. Commonly, they combine with hemorrhagic contusion lesions.

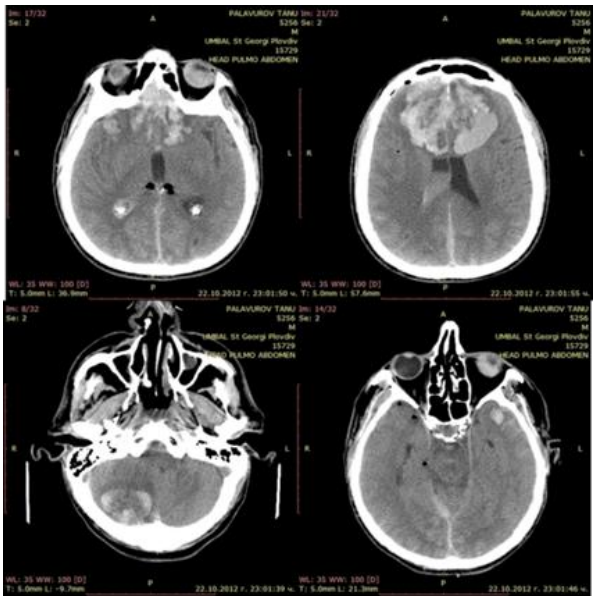


Fig. 2.15. CT images of multiple supratentorial and infratentorial intracerebral hematomas and pneumocephalus

Subdural hydroma

It was described for the first time by H. Naffziger in 1924. Sybdural hydroma is an accumulation of CSF in the subdural space. It is caused by a tear of arachnoid mater, through which the CSF enters the subdural space. Due to ball-valve mechanism the CSF cannot return in the arachnoid space (Fig. 2.17 and Fig. 2.18), which causes gradual enlargement of the collection resulting in brain compression. The content of the hydroma (CSF) is usually clear and pellucid but can also be xanthochromic due to subarachnoid hemorrhage. Long-lasting subdural hydroma can form a capsule.

Traumatic pneumocele

It presents an accumulation of air in the intracranial space. It can be caused by skull base and peri-nasal sinus fractures along with tears of dura mater. The escape of CSF is substituted by entering of air.

Pneumocele is a sign of skull base fracture but only rarely does it cause compression (Fig. 2.16 and Fig. 2.19)

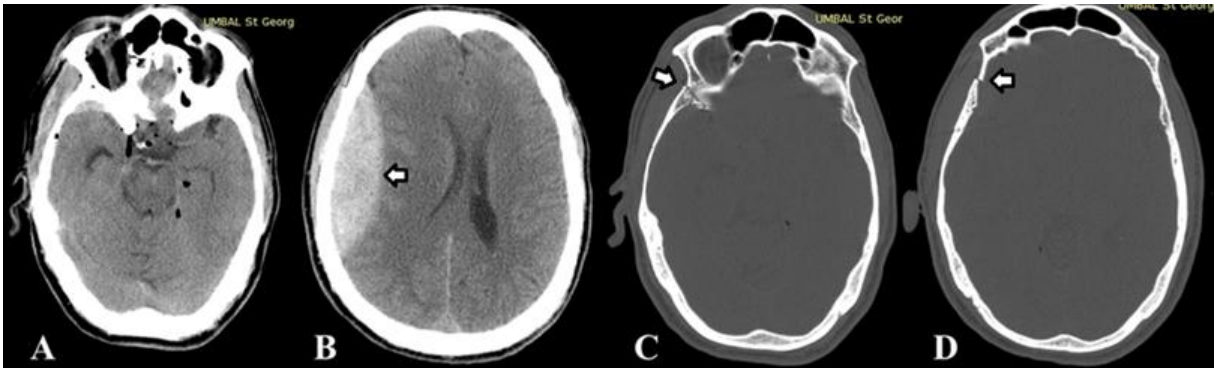


Fig. 2.16. CT images of: **A/** pneumocephalus; **B/** epidural hematoma; **C/** skull base fracture and **D/** calvarian fracture

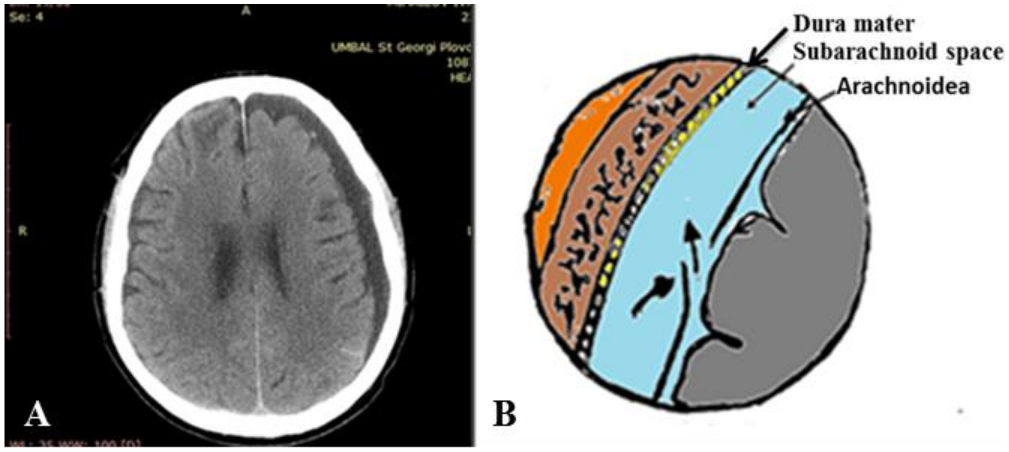


Fig. 2.17. A/ CT image of subdural hydroma; **B/** ball-valve mechanism

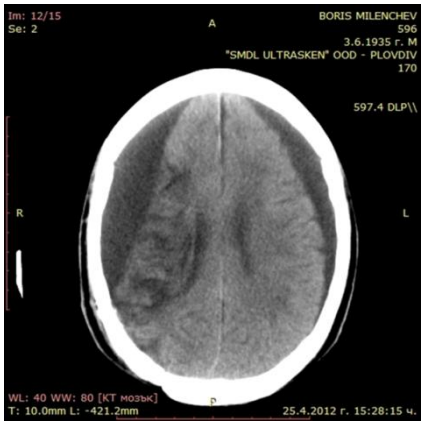


Fig. 2.18. CT image of bilateral subdural hydroma

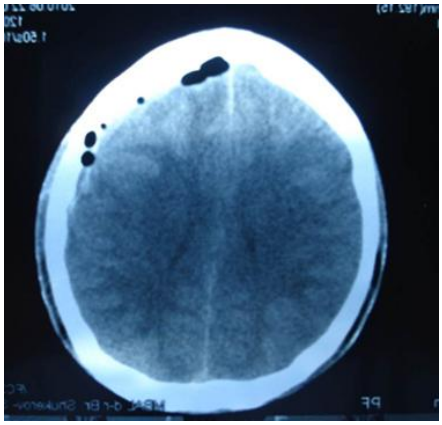


Fig. 2.19. CT image of pneumocephalus

Diagnosis of intracranial hematomas

- **skull X-rays** can demonstrate the presence of skull fractures. Linear fractures crossing a groove of the middle meningeal artery can cause epidural hematoma.
- **computed tomogrdaphy** gives detailed information about intracranial blood collections, its precise volume and localization as well as the presence of contusion lesions and brain edema. CT image of epidural hematoma resembles biconcave lens (Fig. 2.11 and Fig. 2.16) while subdural collections are usually sickle-like lesions (Fig. 2.13). CT density can predict the age of the hematoma. Acute hematomas are hyperdense or white (fresh blood clots); subacute hematomas can be isodense or mimic the gray colour of the brain (Fig. 2.20); chronic hematomas are hypodense or dark (as a result of hemoglobin degradation), but with hyperdense capsule (Fig. 2.13). CT appearance of hematomas may have prognostic value, for example, severe midline shift in acute subdural hematomas is associated with increased mortality rate.

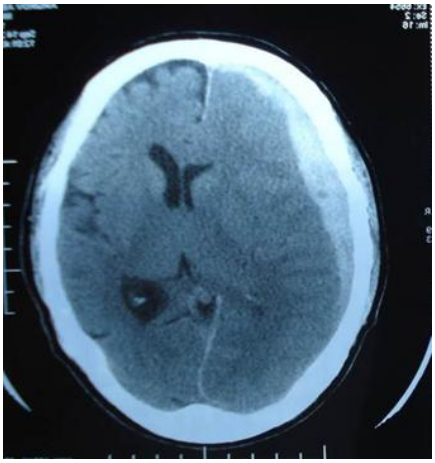


Fig. 2.20. CT image of acute subdural hematoma

Treatment

The presence of epidural and subdural hematomas is an indication for urgent surgical treatment. Intracerebral hematomas can subside spontaneously. Surgery is indicated for those which cause

mass-effect with compression and dislocation of brain structures and deterioration in the neurological status.

Skull fractures (Fracturae cranii)

They can be divided into:

- **Fractures of the cranial vault (fracturae calvariae)**:
  - linear fractures (single and multiple)
  - comminuted fractures:
    - ✓ non-dislocated (fragments do not compress the underlying brain).
    - ✓ dislocated or depressed (fragments compress the underlying brain) (Fig. 2.21 and Fig. 2.22).

Depressed cranial vault fractures are indicated for surgery because they cause brain compression.

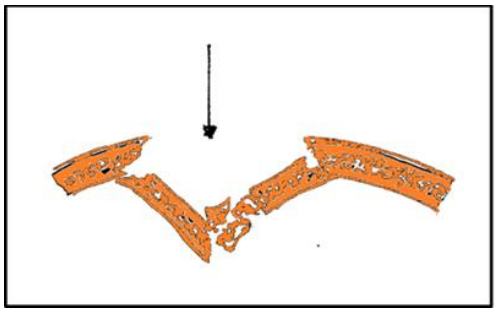


Fig. 2.21. Depressed skull fracture caused by object with small surface

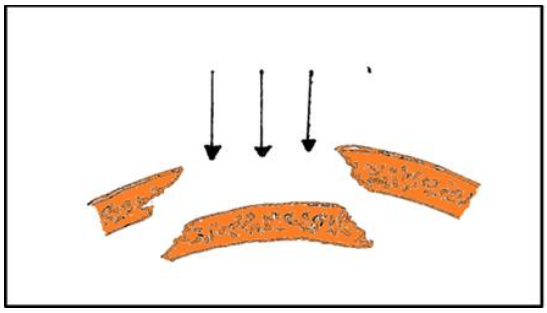


Fig. 2.22. Depressed skull fracture caused by object with large surface

Depressed skull fractures often tear the dura mater and the cortex of the underlying brain. Fragments compressing the brain impair the local blood flow and cause ischemia. Gradually, it causes the formation of glial scar which can become a source of epileptic seizures.

Clinical presentation: It depends on the size of the fragments and rate of compression and the area of brain being compressed. Associated cerebral contusion also adds to the clinical symptoms.

Diagnosis

- **skull X-rays** (AP and lateral) show the size and localization of the fracture (Fig. 2.23, Fig. 2.24);

- **CT** (with bone window) readily demonstrates the skull fracure (Fig. 2.25, Fig. 2.26) CT can also show the presence of cerebral contusion and/or hematoma.

Treatment: Treatment of linear and comminuted non-dislocated fractures is conservative. Depressed cranial vault fractures are treated surgically. The aim of surgery is to remove depressed fragments and necrotic brain, restore the integrity of dura mater, thus avoiding brain scarring. Skull defects are also restored.

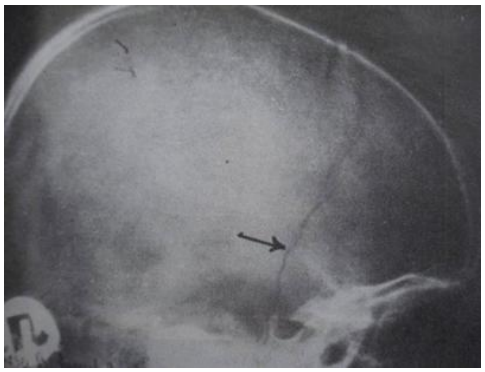


Fig. 2.23. Lateral skull X-ray linear fracture

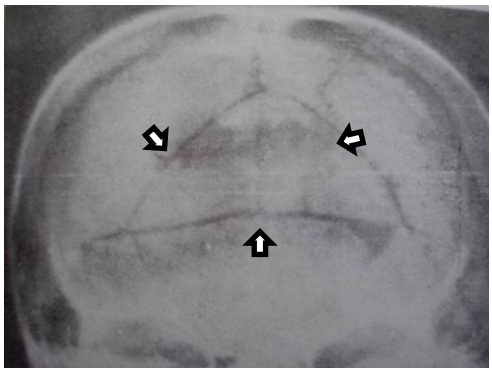


Fig. 2.24. AP skull X-ray depressed skull fracture

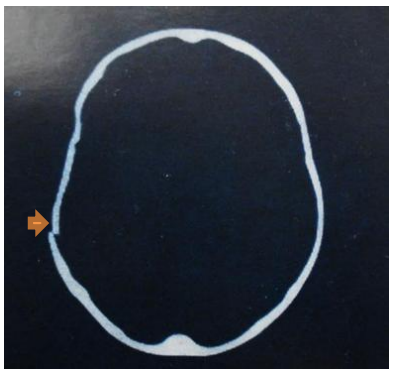


Fig. 2.25. CT with bone window – linear fracture

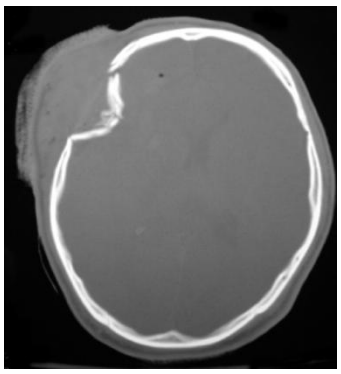


Fig. 2.26. CT with bone window - depressed skull fracture

Skull base fractures (Fracturae basis cranii)

They can be found in 20-25% of head injured patients and combined with cranial vault fracures.

Classification:

- fractures of the anterior cranial fossa;
- fractures of the middle cranial fossa;
- fractures of the posterior cranial fossa.

Skull base has numerous foramina through which important vessels and cranial nerves pass. Its external surface borders the paranasal sinuses and its internal surface faces the basal cisterns. These characteristics determine the typical clinical symptoms and possible complications of the skull base fractures.

Clinical presentation

A/ Anterior cranial fossa fractures may present with:

- nose bleeding (epistaxis);
- periorbital ecchymosis, i.e. 'raccoon' or 'black' eyes with subconjunctival hemorrhage;
- CSF rhinorrhea, brain detritus flowing out from the nose (Fig. 2.27);
- optic nerve injury - visual impairments, sometimes amaurosis.

B/ Middle cranial fossa fractures may present with:

- hematotympanoma, otorrhagia, CSF otorrhea, CSF leakage into the nasopharynx through the Eustachian tube;
- facial nerve injury. Peripheral facial paralysis with positive Bell's sign may become evident immediately after the trauma due to direct injury by the fractured pyramid of the temporal bone or later due to nerve hemorrhage and edema;
- statoacoustic nerve injury with hypacusis or anacusis, loss of balance and dizziness (vertigo);
- mastoid ecchymosis known as 'Battle's sign'.

C/ Posterior cranial fossa fractures may present with:

- caudal cranial nerves injury

Pneumocephalus seen on CT examination may present in all types of skull base fractures!

CSF leakage is observed in 10-15% of skull base fractures (Fig. 2.27). It presents a serious complication due to the direct communication between the environment and the intracranial space posing a great risk of infection (meningitis).

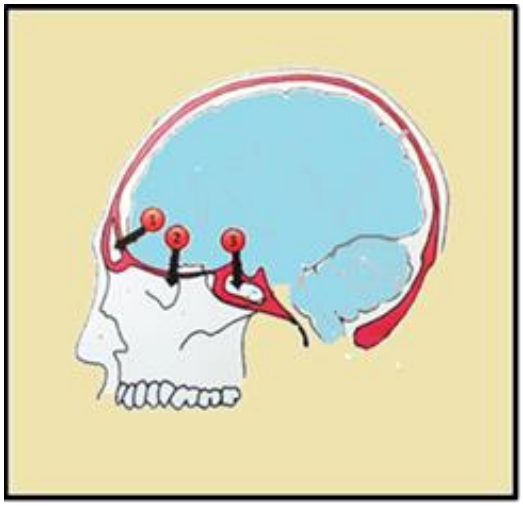


Fig. 2.27. Pathways of CSF rhinorrhea: 1. Through the frontal sinus; 2. Through lamina cribrosa; 3. Through the sphenoid sinus

Ear tamponade is not efficient and may increase the risk of infection. Patients should lie opposite to the side of liquorrhea, drugs lowering the intracranial pressure should be used diuretics (Acetazoleamid, Furosemid, Mannitol). Serial lumbar punctures are sometimes indicated. Antibiotic prophylaxis is mandatory.

Ear tamponade is not efficient and may increase the risk of infection.

Diagnosis

- **skull X-rays:** Basilar fractures require special X-ray projections such as Stenvers, Schuller, Hirtz, etc., and may be difficult to diagnose.
- **computed tomography** using bone window can demonstrate basilar fractures in some cases (Fig. 2.28). Pneumocephalus is an indirect sign indicating these fractures.
- CT cisternography with water-soluble non-ionic contrast media (Omnipaque, Amnipaque) can locate the site of CSF leakage.

Negative scanning and X-ray examinations do not rule out the presence of basilar fracture if typical clinical signs are present!

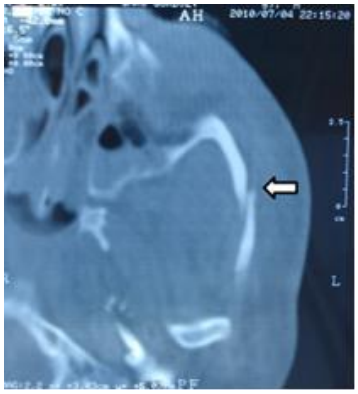


Fig. 2.28. CT image of middle cranial fossa fracture

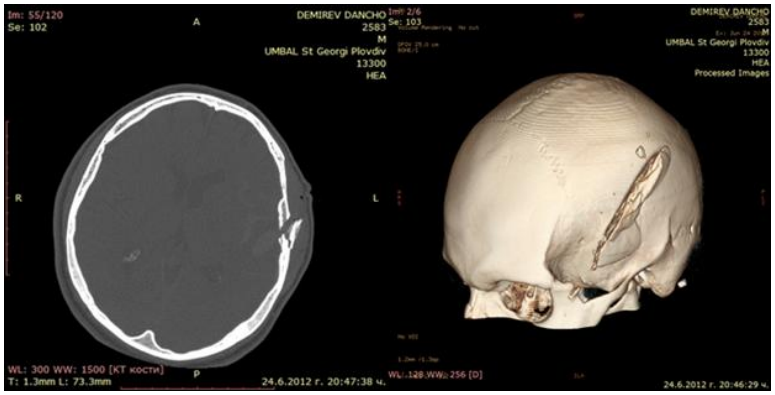


Fig. 2.29. CT image of opened penetrating head injury

Opened and penetrating injuries

Stab and incision wounds fall into this group. (Fig. 2.29) They share common mechanism, patophysiology and clinical presentation of injury:

- traumatic mechanism – mechanical energy of the traumatizing object causes damage to the tissues it passes;
- there is no acceleration and deceleration of the head which makes „contre-coup“ injuries extremely rare;
- the traumatizing object damages limited area;
- the agent is a source of infection to the intracranial space.

Diagnosis:

- examination of the head and skull tells the nature of the traumatizing agent;
- skull X-rays – show the site of entry of the agent and presence of alien bodies;
- CT can demonstrate the presence of intracranial hematomas and brain edema.

Treatment: Surgery aims at removing intracranial hematomas and alien bodies. Otherwise, medical treatment is indicated to prevent from infection and alleviate brain edema.

Gun-shot head injuries

They can be caused by projectile or shrapnel. They can be:

- penetrating (dura mater is torn);
- non-penetrating (dura mater is intact).

According to the mechanism they divide into (Fig. 2.30):

- **transit** – the projectile crosses the intracranial space. The entry wound is usually smaller than the exit wound. The hit wave causes severe damage to the brain tissue;
- **blind** – the projectile enters but does not exit the intracranial space;
- **tangential** – the projectile crosses soft tissues and skull of head and can tear the meninges and injure the brain;
- **ricochet –** the projectile does not have enough energy and speed to penetrate the skull and ricochets.

Clinical presentation: It depends on the mechanism of injury, the kinetic energy of the projectile and the subsequent intracranial infection.

Diagnosis: Skull X-rays and CT scanning are often used.

Treatment: Surgical treatment is indicated for all gun-shot head injuries. The aim of surgery is to remove the fractured skull fragments, remove intracranial hematoma, brain debris and alien bodies. Restoration of dura mater and scalp integrity is also necessary.

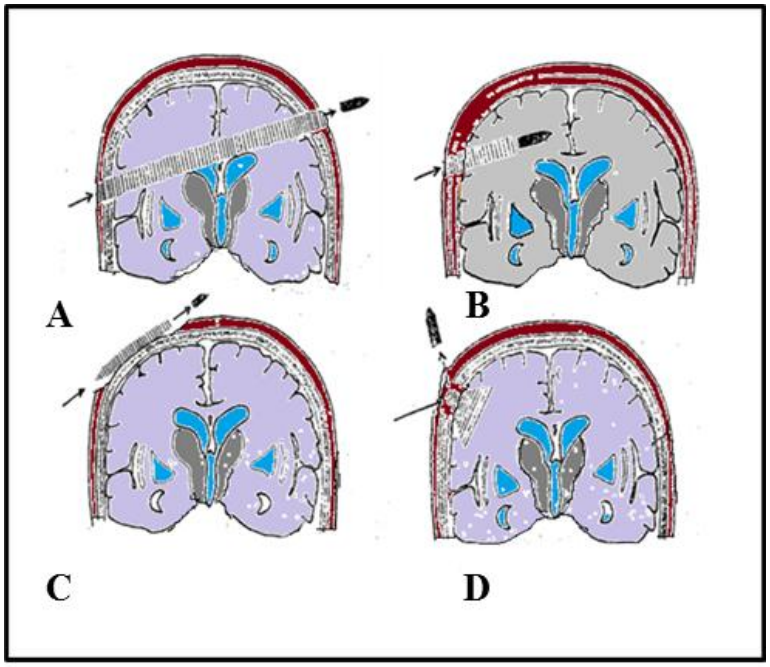


Fig. 2.30. Types of gun-shot wounds:
A/ transit; B/ blind; C/ tangential ; D/ ricochet

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CHAPTER 3 *H. Zheliazkov, I. Kehayov, A. Davarski*

SPINAL INJURIES

Introduction

Spinal injuries (SI) are less frequent than head injuries. SI are substantial social and personal problem for the majority of cases because they lead to high rate of disability and affect young individuals. The incidence of SI varies between 1-4%. Spine fractures contribute for 10-20% of all skeleton fractures.

The incidence of the affected spine segments is different: cervical fractures present 20-25% of SI, thoracic fractures 20-35%, and lumbar fractures 35-40%.

The spinal cord is affected in 25-40% of cases with SI. Focal neurological deficit is established in 70-90% of the lower cervical and high thoracic fractures and dislocations. Mortality varies from 6% to 34%. Mortality in cervical injuries reaches 33%, in thoracic injuries 8-9% and in lumbar injuries 6-7%. Associated injuries lead to an increase in mortality rate.

Etiology

- traffic accidents 40-50%;
- falls 18-23%;
- water jumps 8-10%;
- sports accidents 4-7%;
- penetrating SI (gun-shot and stab injuries) 7-14% of cases.

Classification. There is a great diversity of classifications of SI.

A/ According to the general surgical principles SI can be:

- **closed** – there is no skin laceration. Morphologically, they can be divided into:
  - SI without damage to neural structures;
  - SI with damage to neural structures;
  - neural damage without spine fracture and dislocation.
- **opened -** there is skin laceration. They can be divided into:
  - **non-penetrating** – without damage to the spinal canal;
  - **penetrating-** there is damage to the spinal canal, commonly, in stab and gunshot wounds. Gunshot wounds can be divided into:

- ✓ **transit** – the projectile passes through the spinal canal;
- ✓ **blind** – the projectile enters but does not exit the spinal canal;
- ✓ **tangential** – the projectile ricochets in the bony structures surrounding the canal but does not enter it.

B/ According to the damage of the vertebral column SI can be:

- fractures (vertebral body fractures (transverse, compression, transverse, burst-fracture, etc.); fractures of the tansverse and spinous processes) (Fig. 3.1A); dislocations (due to ligamentous and vertebral facet damage) (Fig. 3.1B-C);
- fracture – dislocations;
- distortions (due to ligamentous injuries) (Fig. 3.2).
- traumatic disc herniations.

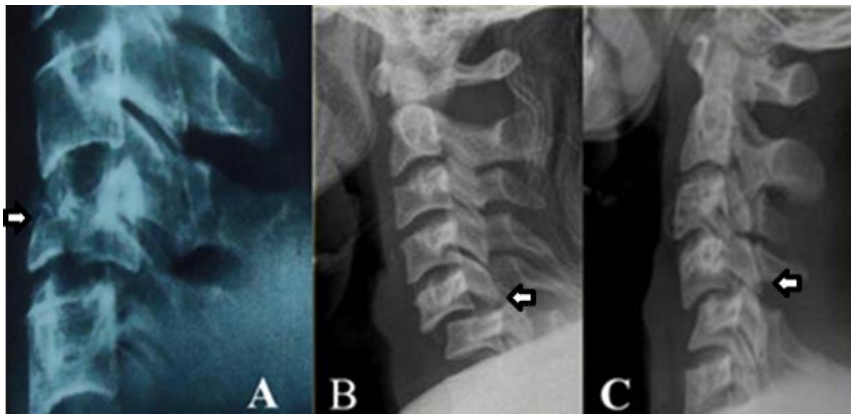

Fig. 3.1. Lateral X-ray: **A/** fracture of C5 vertebral body; **B/** dislocation at C5-C6 level; **C/** dislocation at C4-C5 level

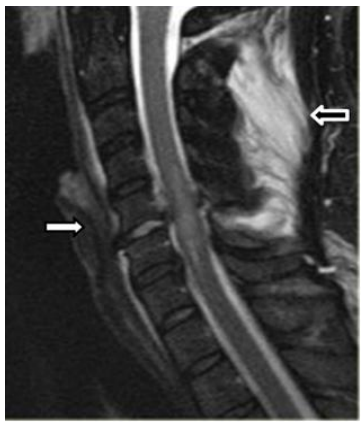

Fig. 3.2. MRI of cervical distortion: (➡) damage to the posterior ligaments; (⇨) damage to the anterior

longitudinal ligament and traumatic disc herniation at C5-6 level

C/ According to the mechanism of spinal injury

- **hyperflexion** – the most common type. It affects the transition areas from mobile to rigid spine (cervico-thoracic, thoraco-lumbar region). According to the spine load, this mechanism results in two major types of spine damage:
  - **axial load** It can lead to compression fracture or burst fracture. In the cervical region it may cause ligamentous and facet damage leading to dislocation (Fig. 3.3);

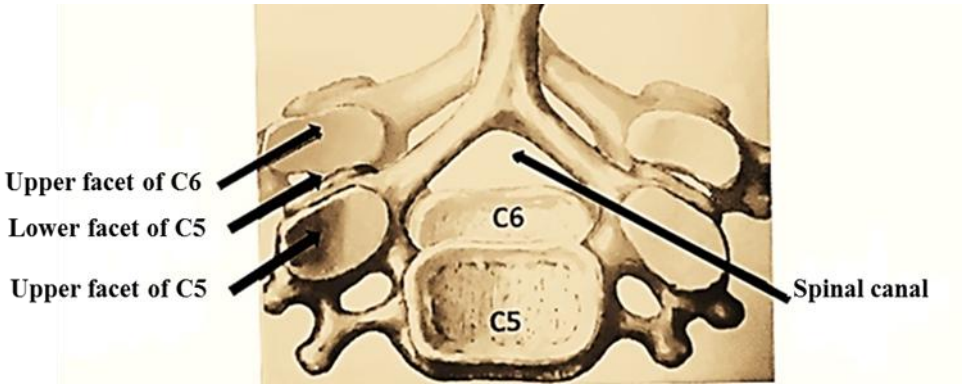

Fig. 3.3. Anterior dislocation at C_{5-6} level due to hyperflexion injury

  - **lateral load –** causes fractures of vertebral laminae and processes.
- **hyperextension** It leads to tearing of the anterior longitudinal ligament, anterior half of the intervertebral disc in combination with fractures of the posterior elements - laminae and processes (Fig. 3.4). Hyperextension injuries in adult patients with degenerative cervical spine can lead to neurological deficit without damage to the vertebral column seen on imaging studies. It results from compression of the anterior spinal artery and spinal cord by osteophytes and thickening of the yellow ligament. The condition is known as **central cord syndrome.**
- **axial compression** – seen in falls from height on the legs or pelvis – commonly causes burst fractures (Fig. 3.5).
- **rotation mechanism -** typical for the mobile cervical and lumbar spine. It can cause fractures of all vertebral elements - anterior, posterior and lateral.

- **lateral hyperflexion.** In these cases the load is transmitted to the lateral parts of the veretebrae and the articular processes.
- **combined mechanism –** hyperflexion with rotation, hyperextension with rotation, axial load with rotation, etc.

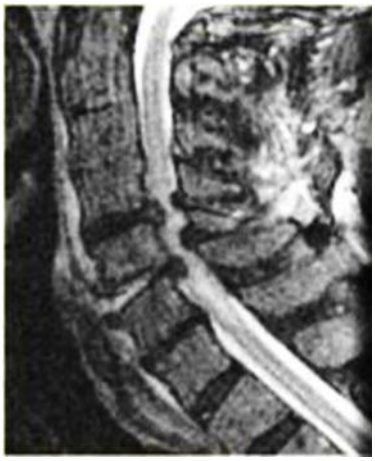

Fig. 3.4. MRI of hyperextension injury at C5-6 level and tearing of C5-6 disc

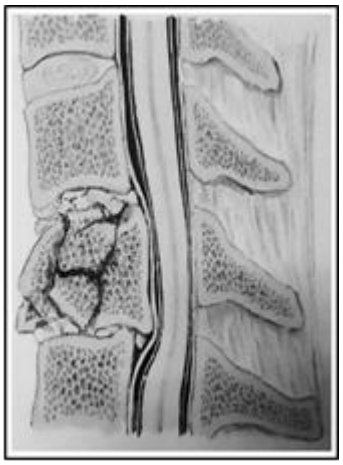

Fig. 3.5. Compression fracture.

D/ According to spinal stability

Stability or instability of the spine determines the severity, prognosis and treatment of SI. According to T. Whithesides (1977), the spine is stable when it can bear the normal loads and shear forces of body weight and motion without causing deformation and additional neurological damage. It is of extreme importance for the physician to determine if a certain vertebral fracture is stable or unstable. In 1983 Denis & McAfee, independently from each other, introduce the 'three-column' theory for the spinal stability. According to this theory, instability is present if two or more columns are damaged (Fig. 3.6).

E/ According to the localization of the SI

- occipito-cervical injuries: occipital condyles, C_1 and C_2 veretebrae form the occipito-atlanto-axial complex;
- sub-axial injuries ($C_3 – C_7$);
- thoracic injuries ($Th_1 – Th_{10}$): the thorax contributes to the increased stability of this segment;

- thoraco-lumbar injuries ($Th_{11} – L_2$): this region is commonly affected by SI;
- lumbar injuries ($L_3 – L_5$);
- sacral injuries.

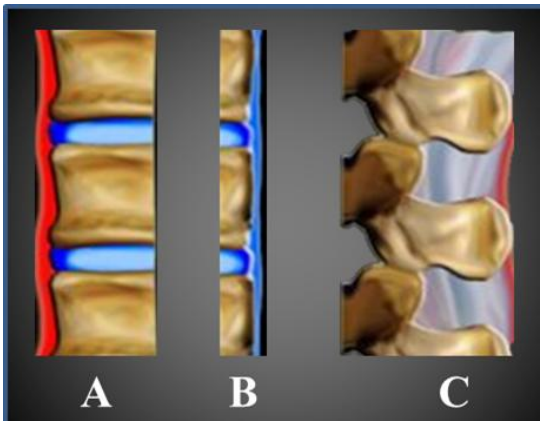

Fig. 3.6. The three columns of the spine according to F. Denis:
A/ Anterior column (anterior longitudinal ligament, anterior half of the vertebral body and disc);
B/ Middle column (posterior half of the vertebral body and disc, posterior longitudinal ligament);
C/ Posterior column (pedicles, articular facets and joints, laminae, spinous processes, yellow ligament, interspinous and supraspinous ligaments)

F/ According to morphological damage to the spinal cord

- spinal cord concussion (Commotio medullae spinalis) There are no morphological changes in the cord. There are only functional changes (including neurological deficit) which are transitory and regress within 24 hours;
- spinal cord contusion (Contusio medullae spinalis) There are always morphological changes detected with MRI. They can result from primary or secondary injury. Contusion can lead to:
  - complete anatomical interruption of the spinal cord (Fig. 3.7);
  - incomplete anatomical interruption of the spinal cord (Fig. 3.8);
  - complete axonal interruption (impaired function without anatomical interruption);
  - incomplete axonal interruption (partial damage of spinal cord conduction without anatomical interruption);
  - haematomyelia – intraparenchymal cord hemorrhage affecting one or more segments. Following resorption, a post-traumatic syrinx is usually formed.

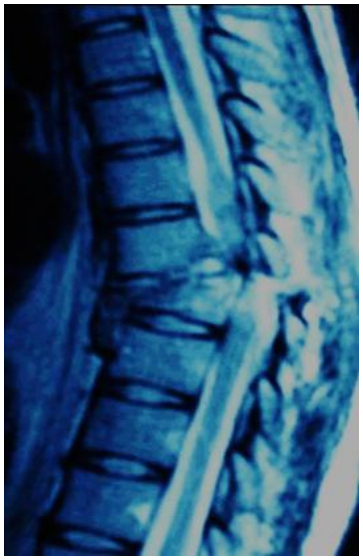

Fig. 3.7. MRI image of complete anatomical interruption

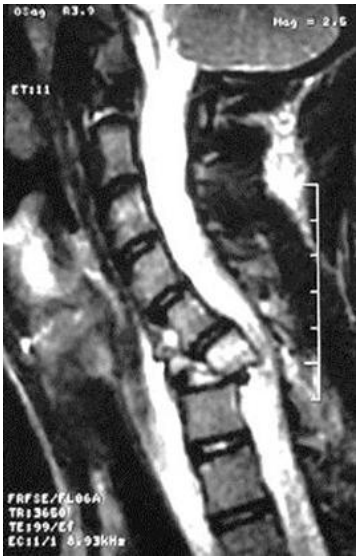

Fig. 3.8. MRI image of incomplete anatomical interruption

- spinal cord compression (Compressio medullae spinalis): Compression may be caused by:
  - bone fragments;
  - blood (Fig. 3.9);
  - dislocations;
  - traumatic disc herniation;
  - alien body.

Fig. 3.9. MRI of spinal epidural hematoma

Pathology of spinal cord injuries

Pathology of spinal cord injury is similar to that of traumatic brain injuries (TBI). Spinal cord injuries can be primary (focal and diffuse) and secondary. Treatment efforts should aim at preventing complications as a result of secondary injuries in order to minimise spinal cord damage. There are two major pathogenic mechanisms underlying spinal cord damage:

- **vascular mechanism -** the impairment in regional blood flow, local vasoconstriction and loss of autoregulation as well as the endothelial damage alter vascular permeability can result in thrombosis which will determine the severity of secondary damage and prognosis;
- **neuronal mechanism -** it includes direct damage to cell membranes with consequent cytotoxic and vasogenic oedema which eventually leads to formation of posttraumatic cysts and functio laesa.

Neurological syndromes resulting from SI

- **spinal shock:** Initial neurological symptoms following acute SI often result from the spinal shock. It is an acute block of spinal cord conductive functions below the level of injury. Decreased muscle tone and absence of somatic and autonomous reflexes are common (flaccid paralysis, hypotonia, bladder and bowel incontinence). It results from a rapid onset of hyperstimulation which leads to overload and blockage of neurotransmission. The spinal shock usually lasts from several days to several weeks followed by the development of the typical neurological signs of central damage (spastic paralysis, hyperreflexia, pathological reflexes, etc.);
- **complete anatomical interruption:**
  - **1st stage** – presents immediately after the acute trauma and includes: anesthesia, total loss of motor function (plegia) distal to the level of injury, hypotonia, loss of reflexes, bowel and bladder disturbances;
  - **2nd stage** – it takes several weeks to develop the typical clinical presentation anesthesia, signs of central paralysis

including hyperreflexia with abnormal reflexes, spasticity, urinary retention.

- **anterior cord damage** – motor paralysis below the level of injury with sparing of proprioception and vibratory sensation (Fig. 3. 10);
- **posterior cord damage** – loss of proprioception and vibratory sensation below the level of lesion with sparing of motor functions (Fig. 3.11);
- **lateral cord syndrome (Brown-Sequard)** - motor paralysis, loss of proprioception and vibratory sense on the same side and loss of pain and temperature sensation on the side opposite to the lesion, typically beginning 1 to 2 dermatomal levels below the injury (Fig. 3.12);
- **conus medullaris syndrome (S_3 – S_5 spinal cord segment)** – presents with urinary and bowel disturbance;
- **cauda equina syndrome –** presence of radicular pain and asymmetric dermatomal type of sensory and motor deficit in the lower extremities;
- **central cord syndrome –** typically occurs in patients with cervical stenosis who sustain hyperextesion injury. It is characterized by disproportionately bigger motor impairment in the upper extremities than in the lower extremities, bladder dysfunction (usually urinary retention), and variable degree of sensory loss below the level of the lesion.

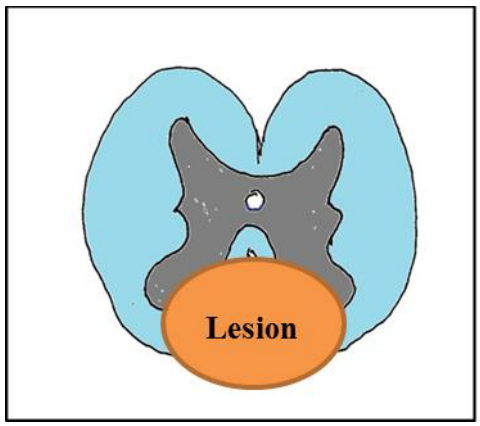

Fig. 3.10. Anterior cord damage

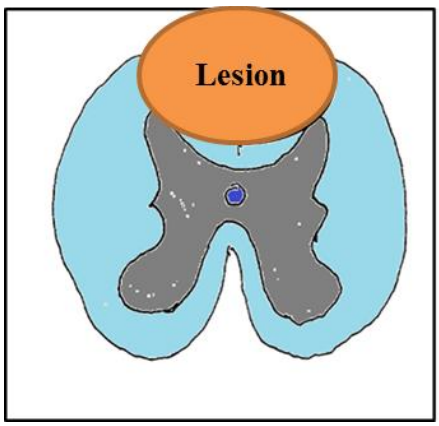

Fig. 3.11. Posterior cord damage

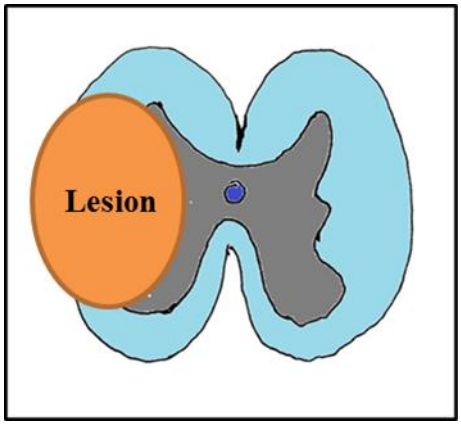

Fig. 3.12. Lateral cord syndrome

Diagnostic stages of SI

- **1st stage: assessment and support of vital functions.** It includes patient assessment based on history, observation and physical examination. It is of great importance to secure patent airways for adequate breathing. If there is loss of spontaneous breathing the patient should be intubated (without head deflexion or rotation). It is necessary to support stable hemodynamics as well as prophylaxis of other systems and organs.

All traumatic patients who are unconscious should be dealth with as patients potentially suffering from spinal injuries – they should be transferred immobilized and all initial diagnostic and therapeutic manipulations should avoid neurological damage due to spinal instability.

- **2nd stage: thorough neurological examination in order to diagnose the level of injury of the spinal cord:**
  - **severe injury above C_4** – paralysis of the respiratory muscles and the diaphragm;
  - **spared C_5 segment** – the patient is able to move his/her shoulders;
  - **spared C_6 segment** – the patient is able to move his/her shoulders, to do flexion in the elbows, and to do supination of the palms;

- **spared C_7 segment** – all of the above mentioned plus extention in the elbow and wrists;
- **spared C_8 segment** – all of the above mentioned plus flexion in the wrists;
- **spared Th_1 segment** – all of the above mentioned plus fine movements of fingers;
- **thoracic spinal cord injury –** in order to define the level of injury the line which delineates the decreased sensation can be used: mammillary line (men) – Th_4; processus xyphoideus – Th_5 - Th_6; umbilicus – Th_{10}; pubis - Th_{12}. Loss of the abdominal reflexes may also be helpful in determining the level of injury:
  - ✓loss of upper abdominal reflex – Th_5;
  - ✓loss of middle abdominal reflex – Th_{10};
  - ✓loss of lower abdominal reflex - Th_{12}-L_1.
- **conus medullaris injury (Th_{11} – L_1)** – presents with urinary and bowel disturbance;
- **cauda equina injury (below L_2)** – presents with urinary incontinence plus radicular pain and asymmetric dermatomal type of sensory and motor deficit in the lower extremities.

Diagnosis of spinal injuries

Apart from the physical and neurological examination, precise diagnosis should be confirmed by performing additional imaging and laboratory studies necessary for correct preoperative planning and appropriate treatment strategy.

- **radiograph (X-ray) examinations:** each patient who has sustained SI should be examined by X-ray in antero-posterior and lateral view. The physician should be able to assess:
  - the integrity of the bony elements comprising the spine and their relationships (presence of fractures, dislocations, spinal alignment, etc.);
  - abnormalities in the adjacent paraspinal structures;
  - the presence of non-traumatic degenerative and pathological conditions such as spinal and foraminal stenosis, conjenital

malformations (block-vertebrae, dysplasia), ankylosing spondylitis (Bechterew's disease), neoplastic lesions;

- **lumbar puncture:** gives information about the type, biochemical, cytological and bacterial CSF abnormalities. The Queckenstedt and Stookey probes can detect blockage of subarachnoid channels;
- **myelography:** can demonstrate compression of the spinal cord and/or cauda equina and epidural CSF leakage due to tears of the meninges;
- **computed tomography (CT):** it gives details about bony structures and surrounding elements (spinal cord, intervertebral discs, etc.). CT reconstruction in three planes (coronal, sagittal and axial) affords assessment of traumatic injuries to the veretebrae, articulo-ligamentous complex, and compression of the spinal canal by bone fragments, alien bodies and traumatic disc herniation;
- **magnetic reissonance imaging (MRI):** in contrast to CT, MRI can examine larger parts of the spine and spinal cord. It gives superior details about soft tissue changes, especially, those in the spinal cord which cannot be detected by CT.

Treatment of spinal cord trauma

The treatment of patients with spinal cord injuries begins at the place of the accident. There should be no flexion or extension at the place of the injury and immobilization is mandatory during transport. Depending on the location of the injury and its type, there are two therapeutic approaches:

- **non-surgical treatment.**
  - **manual reposition.** It can be used for unilateral or bilateral facet dislocation by a well-trained professional, followed by segment immobilization.
  - **cervical traction.** This method is preferable because reposition is achieved by progressively increasing traction through the brackets of Crutchfield, Gardner-wells, Winke

or Halo-traction. This method is applied in cases of cervical dislocations or fracture-dislocations.

- **postural reposition.** It is applied for thoracic and lumbar spine injuries with mild to moderate wedge deformation of the vertebral body without neurological symptoms. This is achieved through a gradual change of the body's position and subsequent immobilization.

- **surgical treatment.** Surgical treatment is a method of choice for severe SI (reduced vertebral body height more than 50%, compression of the spinal cord and instability of the segment). Different operative approaches can be used depending on the area that is damaged, the type of injury and the individual experience of the surgeon. Generally, anterior, lateral, posterior or combined approaches are used as each of them requires certain stabilizing system.

The main goal of the surgical treatment is to achieve reposition of the affected segment, decompression of neural structures (spinal cord and nerve roots) and stabilization of the affected segment.

Specific spinal cord injuries according to the type and location of the damage

1. **Spinal cord injuries of the occipito-atlanto-axial complex.**
   - **occipital condyle fractures.** These are rarely observed as isolated fractures. They are more often combined with C_1 fractures. Cases in which medulla oblongata is affected, usually end up fatally. In most cases, the clinical manifestation includes local pain in the occipitocervical region and, occasionally, hypoglossal nerve injury;
   - **atlas fracture (Jefferson fracture).** This fracture is relatively rare - 2% of all spinal injuries and 3 to 10% of cervical injuries. It is most commonly seen after a fall on the head. Due to the extreme load of the vertical spine axis, the sides of the atlas are pressed against the occipital condyle and the body of C_1, thus breaking the anterior and posterior arches (Fig. 3.13). Heavy neurological deficit is rare because of the "spontaneous decompression" caused by the spreading of the bone ring of

C_1. The most common symptoms include pain and stiffness of the muscles in this area. Condyle fractures are three types:

- **type I** – bilateral fracture of the posterior arch;
- **type II** – unilateral fracture that passes through the anterior and posterior arch and the articular facets;
- **type III** – typical Jefferson fracture with fracture of anterior and posterior arches (Fig. 3.13).

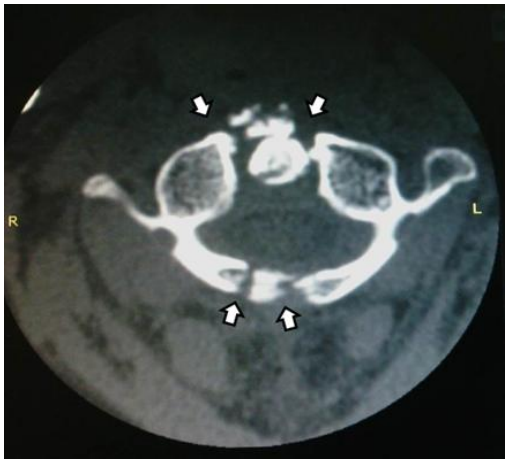

Fig. 3.13. CT image of Jefferson fracture

- **fractures of the dens axis.** C_2 fractures represent 5-15% of all cervical spinal injuries. They are most commonly due to axial load. The different types of C_2 vertebral fractures can result from additional rotation and lateral flexion. The dislocation of the dens is usually a result of hyperextension or hyperflexion injury. The clinical symptoms include pain and paravertebral muscle spasm. Neurological deficit is observed in only 7-10% of the cases. Sometimes fractures affect both C_1 and C_2 (Fig. 3.19). Dens axis fractures are three types (Fig. 3.14):
  - **type I (apical)** - breaking of the tip of the dens axis. It occurs rarely - in approximately 8% of the cases (Fig. 3.14 and Fig. 3.15);
  - **type II (59%) –** the fracture passes through the base of the dens. It is unstable fracture (Fig. 3.14, Fig. 3.16, Fig. 3.17, Fig. 3.18, Fig. 3.20);
  - **type III (33%) –** fracture line runs through the body of C2, and the articular facets (Fig. 3.14, Fig. 3.19).

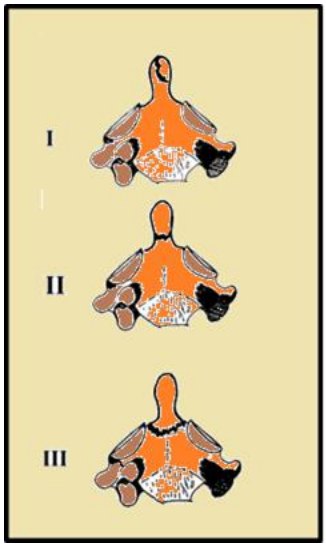

Fig. 3.14. Types of fractures of dens axis (A. Flander, 2008)

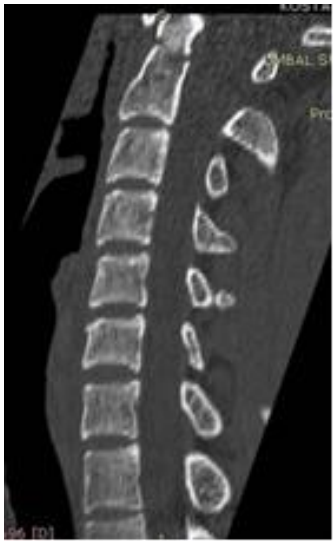

Fig. 3.15. CT image of the dens axis fracture (type I)

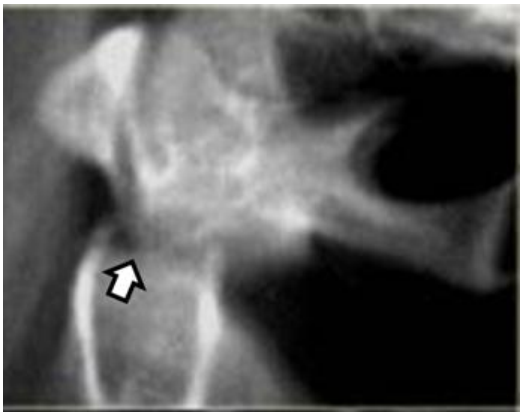

Fig. 3.16. Spondilography of fracture of dens axis (type II)

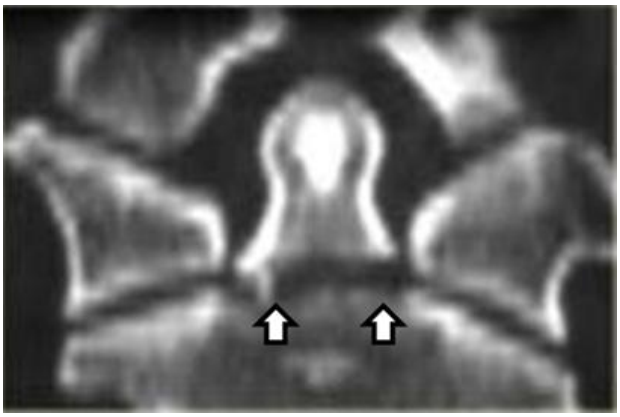

Fig. 3.17. CT image of fracture of dens axis (type II)

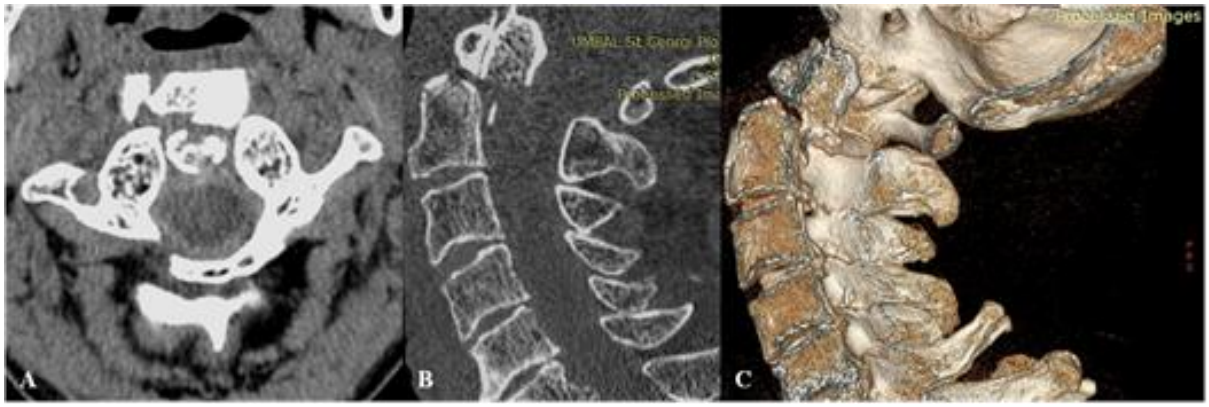

Fig. 3.18. CT image of the fracture of dens axis (type II):
A/ axial projection; **B/** sagittal reconstruction; 3D reconstruction

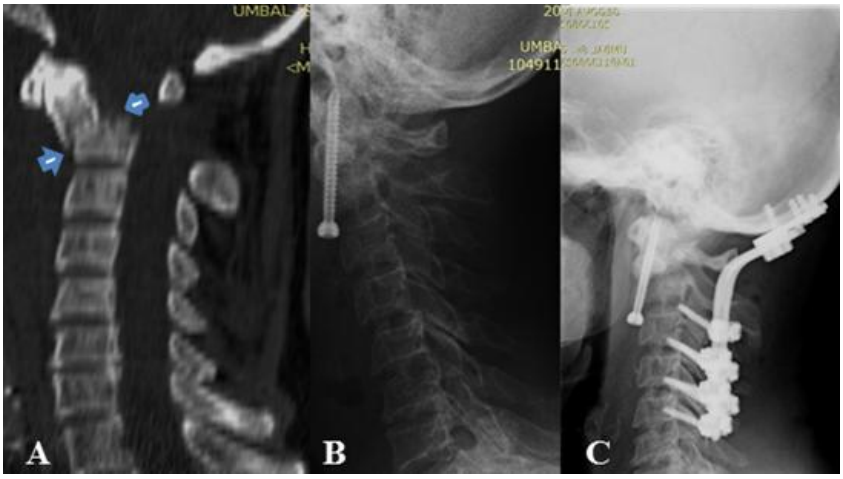

Fig. 3.19. Fracture of dens axis (type III):
A/ Sagittal CT reconstruction; **B/** postoperative radiograph - anterior stabilization; **C/** postoperative radiograph - combined anterior and posterior stabilization

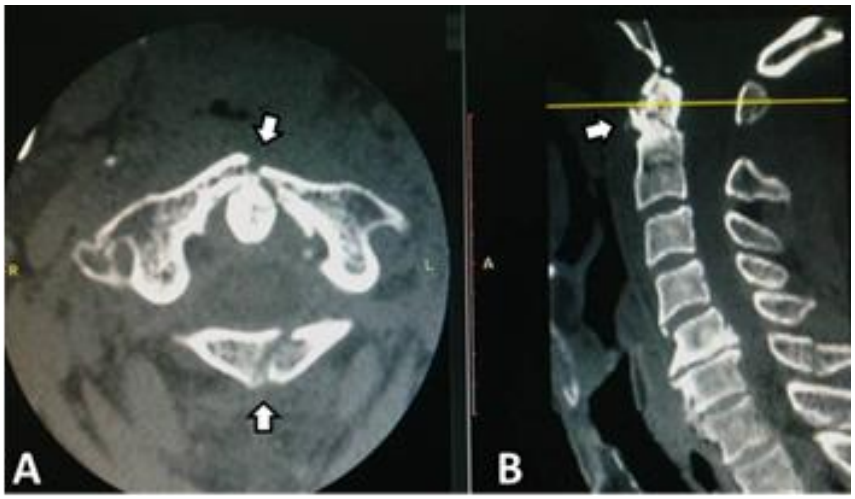

Fig. 3.20. CT image of simultaneous fracture of C1 and C2 (type II): A/ Jefferson fracture; B/ C2 fracture (type II)

- **traumatic spondylolisthesis of C_2 (Hangman's fracture)**. The combination of axial load and hyperextension results in fracture of the intraarticular part of C_2 and tearing of the anterior and posterior longitudinal ligaments as well as C_2-C_3 intervertebral discs. In these cases the head, C_1 and the body of C_2 move forward, while the arch of C_2 and the rest of the spine remain in place. Severe damage to the spinal cord is rarely observed due to the "spontaneous decompression". Depending on the divergence of the C_2 fractured parts (body and arches),

and the position of the anterior compared to the posterior segment, there are 3 types of fractures:

- **type I –** the distance between the anterior and posterior segment is less than 3 mm without change in the normal configuration (Fig. 3.21a, Fig. 3.22 and Fig. 3.23);
- **type II –** the distance between the two segments is more than 3 mm, as the upper segment is tilted forward and downward (Fig. 3.21b);
- **type III –** the distance between segments is similar to type I, but the whole C_2 vertebra is positioned forward compared to C_3 and the inferior articular facet of C_2 passes in front of the upper articular facet of C_3.

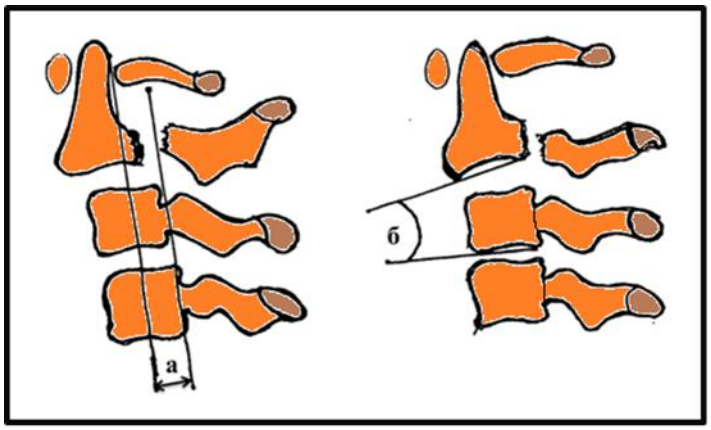

Fig. 3.21. Fractures of the dens axis:
a/ distance between segments; **b/** tilt measurement

- **atlanto-axial dislocation.** It is usually observed in childhood due to the anatomic and physiological characteristics of the paediatric spine;
- **basilar invagination.** Arthritic (rheumatoid, psoriatic) diseases can lead to pathological changes in the ligamentous apparatus which causes non-traumatic upward migration of C_2 into the foramen magnum (Fig. 3.24 and Fig. 3.25).

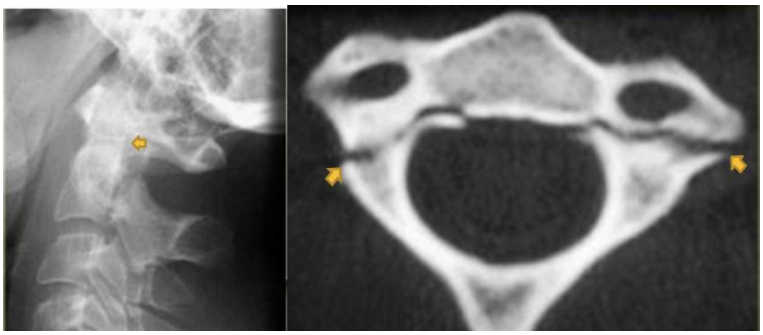

Fig. 3.22. Spondilography and CT of the Hangman's fracture - type I

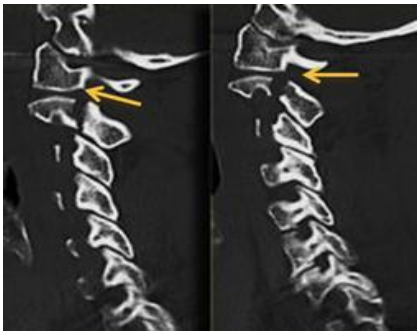

Fig. 3.23. CT of the Hangman's fracture - type I

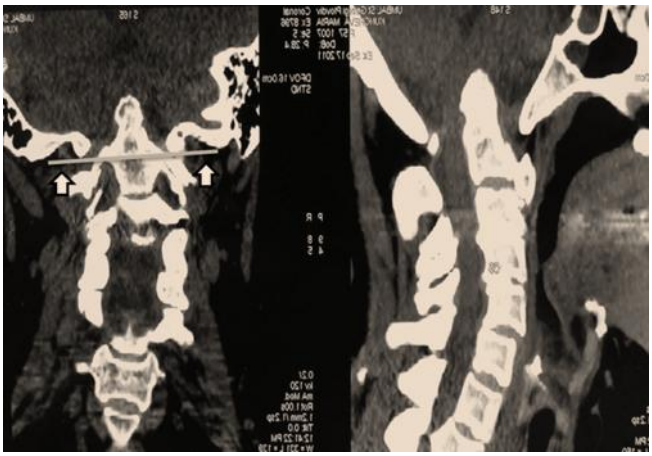

Fig. 3.24. CT image of basilar invagination – dens axis migrates upward and enters the posterior cranial fossa. The top of dens axis exceeds the Chamberlain's line on the sagittal image (not shown). The top of C2 is normally at or below this line

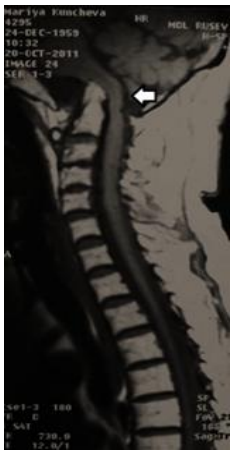

Fig. 3.25. MRI image of basilar invagination

2. Subaxial spinal cord injuries ($C_3 – C_7$)

A/ Compression fractures

- **wedge fractures.** They are the most common fractures in the cervical area. In these fractures axial load plays a major role, as it violates only the anterior column;
- **burst fractures.** The injury mechanism is identical to the type of fracture described above but the vertical load is greater thus violating the anterior and middle spinal column causing severe polyfragmented fracture of the vertebral body often leading to posterior displacement of bone fragments causing spinal cord compression. This type of fractures are unstable and require stabilization;

- **teardrop fractures.** The mechanism includes axial load in combination with hyperflexion which results in breaking of a piece of the antero-inferior part of the vertebral body (Fig. 3.26, Fig. 3.27);
- **unilateral articular dislocation (subluxation)**. Caused by a flexion-rotation mechanism in which the inferior articular facet of the upper vertebra is situated in front of the superior articular facet of the lower vertebra (Fig. 3.1B).

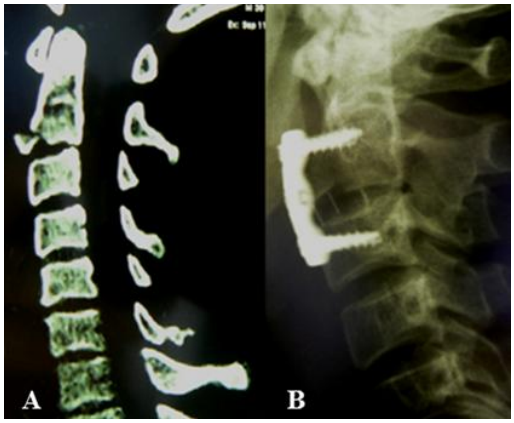

Fig. 3.26. Teardrop fracture of C_2: A/ CT reconstruction; B/ postoperative (X-rey) anterior stabilization

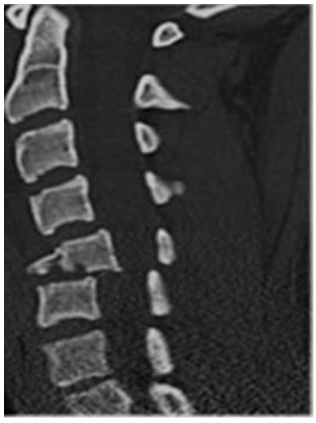

Fig. 3.27. CT of teardrop fracture of C_5

- **bilateral articular dislocation.** In these cases, both inferior articular facets of the upper vertebra pass in front of the superior articular facets of the lower vertebra. There are tears of the articular capsules and ligaments, which allow the upper vertebral body to slip forward and cause neurological deficits (Fig. 3.1C).

B/ Hyperextension fractures. This traumatic mechanism leads to ruptures of the anterior longitudinal ligament, the intervertebral disc as well as a fracture of the vertebral arches and processes.

3. **Spinal cord injuries of the thoracolumbar region.** This part of the spine includes three biomechanical areas (Fig. 3.28):
   - **upper thoracic region (Th1 - Th8) -** rigid zone with a normal kyphosis. This area is usually subject to flexion compression fractures caused by axial load (Fig. 3.28).
   - **thoracolumbar region (Th9 - L2) -** zone of transition between kyphotic and rigid thoracic spine and lordotic and

mobile lumbar spine. This is the most vulnerable area for spine injuries. A variety of mechanisms can play a role for SI here but the most common ones are axial compression, hyperflexion and rotation. Compression (wedge) and burst fractures are often observed (Fig. 3.28, Fig. 3.29, Fig. 3.30, Fig. 3.31, Fig. 3.32, Fig. 3.33, Fig. 3.34).

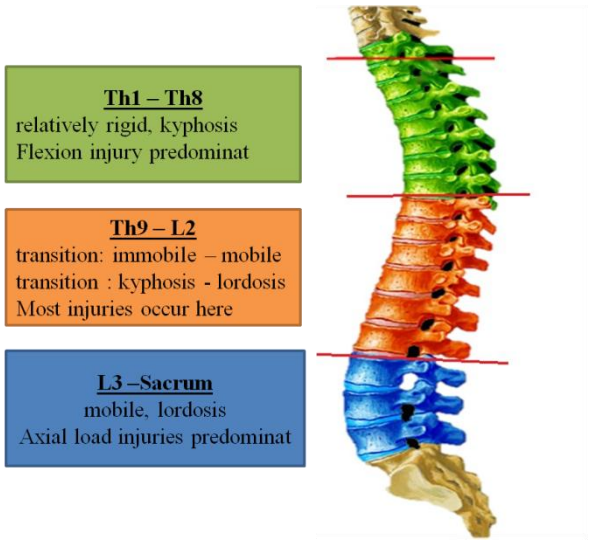

Fig. 3.28. Three biomechanical segments of thoraco-lumbar region

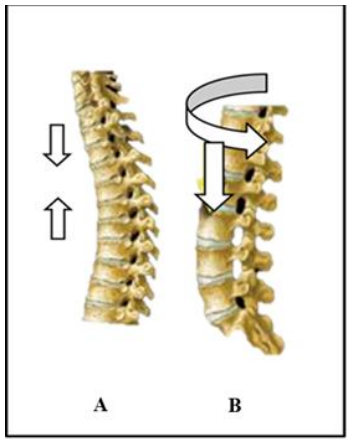

Fig. 3.29. Mechanisms of occurrence of spinal injuries in the upper thoracic, thoracolumbar and lumbar region: **A/** compression; **B/** hyperflexion with rotation

- **lower lumbar region (L_3-L_5 vertebrae)** - mobile part with normal lordosis. Axial loads usually lead to compression (wedge) and burst fractures (Fig. 3.30, Fig. 3.31, Fig. 3.32 and Fig. 3.34). Chance (or "seat belt") fracture and fracture dislocations can rarely be observed in this region.

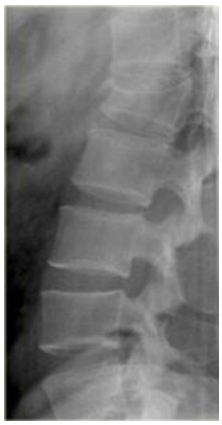

Fig. 3.30. X-ray image of a compression (wedge) fracture of L_1

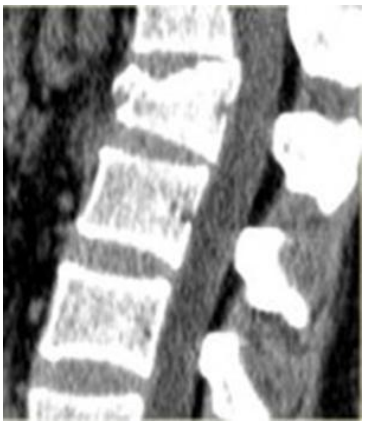

Fig. 3.31. CT reconstruction of a compression (wedge) fracture of L_1

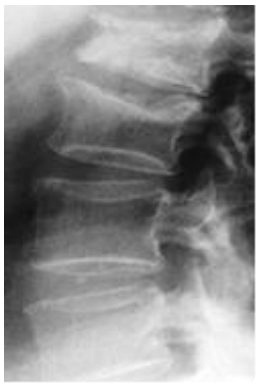

Fig. 3.32. X-ray image of compression farcture at two levels

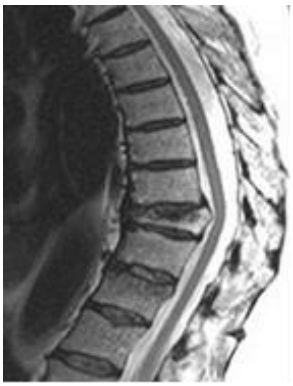

Fig. 3.33. MRI image of compression (wedge) fracture

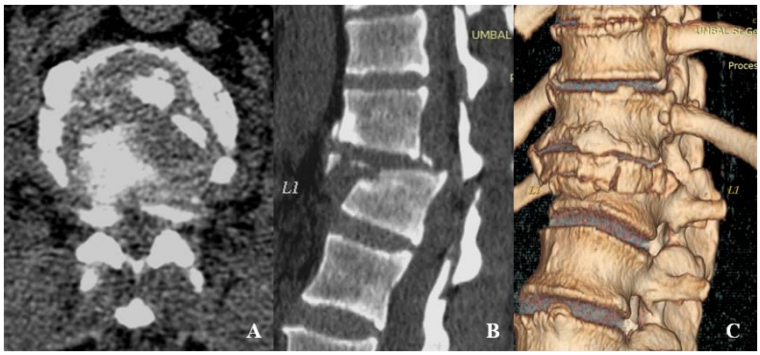

Fig. 3.34. CT image of fracture-dislocation of L_2 causing spinal cord compression: **A/** axial projection; **B/** sagittal reconstruction; **C/** 3D reconstruction

- **Seat belt fracture (Chance fracture).** This fracture results from hyperflexion and rotation of the spine around the seat belt in combination with vertical distraction force. The

horizontal fracture line passes through the spinous process, vertebral arch, the pedicles and the vertebral body (Fig. 3.34);

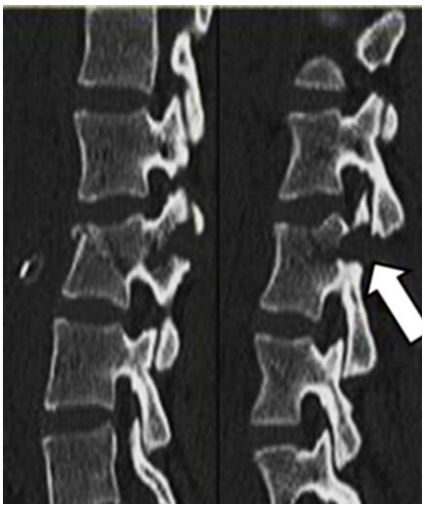

Fig. 3.35. CT image of chance (seat belt) fracture

- **fracture-dislocations.** They result from combined mechanisms (flexion-rotation, flexion-distraction). All three spinal columns are affected which makes this fractures unstable. Neurological deficit is common.

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CHAPTER 4 *B. Kalnev, I. Koev, G. Bogilov*

INTRACRANIAL TUMORS

Introduction

The incidence of brain tumors (BT) varies between 10-50/100000 per year. They represent 5-10% of all cancer cases and are the cause of death in about 0.5-2.5% of cases. Approximately 15-20% of brain tumors are paediatric. Two-thirds of intracranial tumors are malignant and 1/3 are benign. Male/female ratio is 55/45.

Etiology

The etiology of brain tumors is unknown. Influence of hereditary factors has been detected only in some cases (neurofibromatosis type 1 and 2, hemangioma of the cerebellum, some dermoid cysts and teratoma). There are no reliable data regarding the impact of the environment on the occurrence of brain neoplasms, but some reports show that they can be induced by local or intravenous infusion of some chemical agents (venyl chloride, etc.).

Histology

In general, primary BT arises from normal brain cells or abnormal cellular structures such as remnants of the embryonic development (Tables 4.1 and 4.2). Secondary BT results from hematogenous dissemination of systemic cancer (Table 4.3).

Table 4.1. Brain tumors arising from "normal" cells present in the intracranial histological structures

Cell structure	(%)	Type of tumor
Glial Cell • Astrocytes • Oligodendrocytes • Ependymal Cells	45	 Astrocytomas, Glioblastoma Oligodendroglioma, Oligodendroblastoma Ependymoma, Ependymoblastoma
Leptomeningeal cells	15	Meningioma, Atypical and malignant meningioma
Schwann Cells	8	Neuroma (schwannoma)
Connective tissue cells		Fibroma, sarcoma
Immature embryonal cells		Medulloblastoma, medulloepithelioma
Epiphyseal cells		Pinealocytoma, pinealoblastoma
Adenohypophyseal cells		Pituitary adenoma, pituitary carcinoma

Table 4.2. Brain tumors arising from remnants of the embryonic development

Histological structure	Type of tumor
Notochordal cells	Chordoma
Fat cells	Lipoma
Germ cells	Germinoma, teratoma
Cellular remnants of the Rathke's pouch	Craniopharyngioma

Table 4.3. Brain tumors derived from histological elements from other parts of the body.

Histological elements from other parts of the body.	Type of tumor
Lungs, breast, prostate, kidneys, genital organs, digestive system	Metastases

Features

Brain tumors differ from other neoplasms because:

a) They develop within limited intracranial space and exert pressure on the brain as a space-occupying process. Thus, benign brain tumors can also be life-threatening lesions because they can cause brain herniation. Intracranial lesions cannot be examined by physical methods.

b) Most of the brain tumors do not disseminate outside the nervous system

Topography of brain tumors

1. **Basic topography -** the extension of the dura mater that separates the cerebellum from the basal surface of the occipital lobes is called tentorium cerebelli (Fig. 4.1):
   - **supratentorial space** harbors both cerebral hemispheres, diencephalon and lateral ventricles. The left and right hemispheres are separated by the falx cerebri (Fig. 4.1);
   - **subtentorial space (**also known as posterior cranial fossa) harbors the cerebellum, the brain stem and the 4th ventricle (Fig. 4.1).

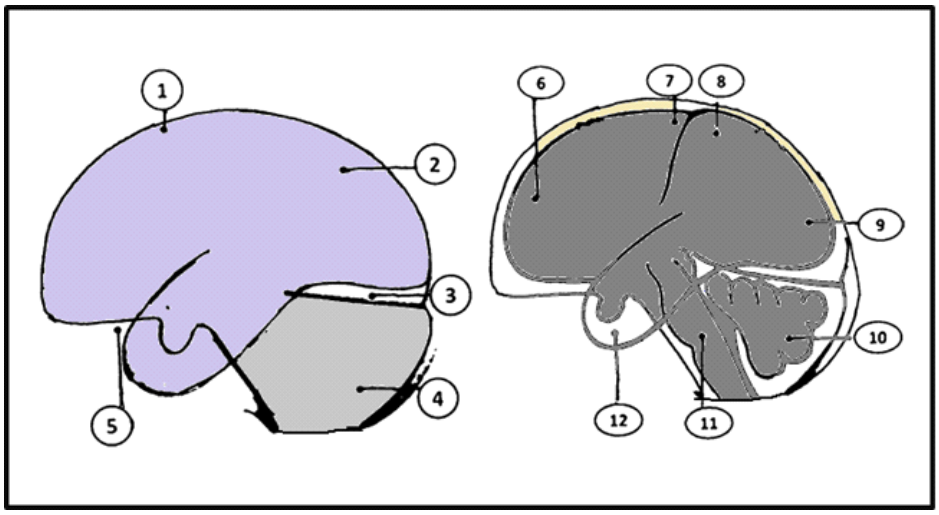

Fig. 4.1. Anatomical distribution of intracranial space:
1. Convexity; 2. Supratentorial space; 3.Tentorium cerebelli; 4. Infratentorial space; 5. Base of the brain; 6. Frontal lobe; 7. Motor area (Frontal part of the Rolandic zone); 8. Sensory area (parietal part of the Rolandic zone); 9. Occipital lobe; 10. Cerebellum; 11. Brain stem; 12. Temporal lobe

2. **According to their relationship to brain parenchyma tumors can be divided into:**
   - **intraparenchymal tumors**. The most common are:
     - not well-demarcated (glioblastoma, astrocytomas);
     - relatively well-demarcated (metastases, hemangio-blastoma).
   - **extraparenchymal tumors** (meningioma, neurinoma, cholesteatoma, etc.);
   - **intraventricular tumors** (ependymoma, meningioma, plexus chorioideus papilloma, colloid cysts, etc.).

3. **According to the functional importance of the affected brain area**:
   - **tumors developing within eloquent (functionally hyperactive) brain areas –** Rolandic zone, dominant temporal and frontal lobes, basal ganglia, brainstem, diencephalic - pituitary area);
   - **tumors developing within non-eloquent (functionally hypoactive) brain areas –** non-dominant frontal and temporal lobes.

The location of the brain neoplasm determines the clinical presentation of the disease, especially its debut.

Tumors growing within eloquent brain areas result in rapid development of focal neurological deficit while those which grow within non-eloquent brain areas present with symptoms of raised ICP.

4. **According to the relationship of the tumor to the CSF pathways**
   - Brain tumors located in close vicinity to or within the ventricular system can block CSF circulation and cause dilation of the ventricular system (hydrocephalus), which in turn leads to intracranial hypertension. This condition may be caused by tumors of the lateral ventricles (ependymoma, choroid plexus papilloma), tumors filling or compressing the 3rd ventrical (colloid cysts craniopharyngioma) as well as the tumors blocking the Sylvian aqueduct or the 4th ventricle (ependymoma, medulloblastoma, large acoustic neuroma, etc.) (Fig. 4.2).

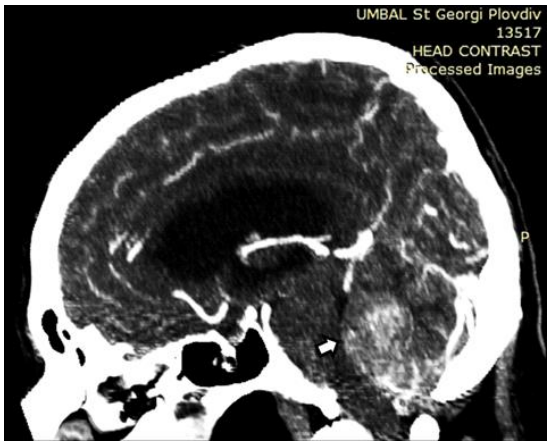

Fig. 4.2. CT with contrast enhancement of ependymoma of the IV ventricle causing obstructive hydrocephalus

5. **According to the relationship of the tumor to the cerebral vasculature (arterial and venous)**
   - the close relationship of brain tumors to important cerebral vessels can sometimes influence the extent of tumor removal and possible complications (for example meningiomas invading dural sinuses or basal tumors which are adherent to important arterial vessels such as the carotid and basilar arteries and their branches) (Fig. 4.3).

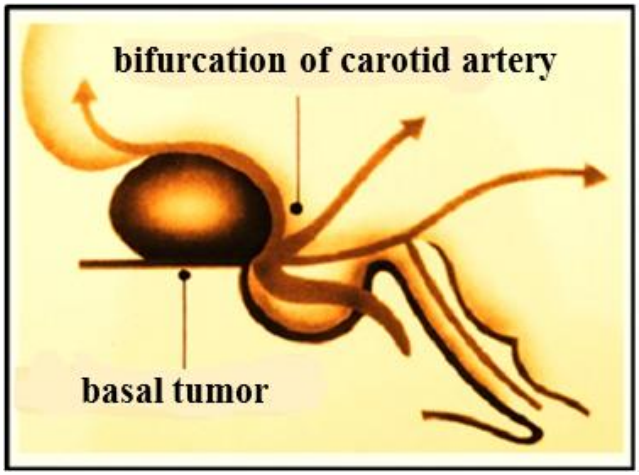

Fig. 4.3. Relationship between skull base tumor and the internal carotid artery branches

Symptomatology of brain tumors. Clinical symptoms of intracranial tumors can be related to two major syndromes.

1. **Focal (local) syndrome**. The focal syndrome is caused by the direct involvement of neural structures (brain parenchyma and cranial nerves) by the brain tumor and alteration in the regional blood supply. It is manifested by focal neurological deficit but can also trigger partial or generalized epileptic seizures.

The focal syndrome depends on:
a) tumor localization; b) tumor size and c) histological type

2. **Syndrome of increased intracranial pressure** (Fig. 4.4)

The syndrome of increased intracranial pressure depends on: a) tumor size; b) tumor histology and c) relationship of the tumor to CSF pathways

Symptoms of intracranial hypertension

- **headache** is caused by irritation of meningeal branches of the trigeminal nerve which innervate dura mater. The typical relapsing morning headaches with severe intensity can irradiate to the occipital or frontal-orbital region. Usually, this headache is accompanied by nausea and vomiting which do not relieve its intensity. The headache may sometimes depend on the position of the head (in case of intraventricular tumors). Frontal tumors may be associated with less severe headaches which can be neglected by the patient;

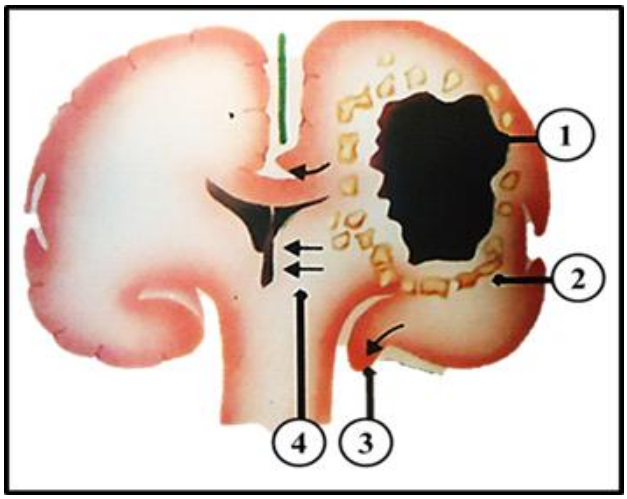

Fig. 4.4. Intraparenchymal tumor causing brain herniation:
1. Tumor; 2. Perilesional edema; 3. Temporal herniation; 4. Compression of the lateral ventricle

- **vomiting** is a result of irritation of the vagal nerve and streching of the meninges. It is not associated with gastrointestinal disorders and does not depend on food intake. It usually occurs later in the clinical course. Early onset of refractory vomiting which can be exacerbated by movements of the head is due to direct irritation of the vagal nuclei from 4^{th} ventricule tumors (so-called Bruns syndrome). In this case, vomiting presents as focal syndrome. Vomiting can be absent and the patient can suffer from nausea only;
- **papilledema** is registered by ophthalmic fundoscopy. It can eventually lead to secondary atrophy of the optic nerves. If left untreated, it results in blindness;

The absence of papilledema does not rule out the diagnosis of intracranial tumor

- **behavioral changes** can be a sole long-lasting symptom of brain tumors characterized by drowsiness, apathy, indifference, intellectual and cognitive decline. In some cases, the clinical presentation is dominated by these symptoms (typical of frontal and 3^{rd} ventricle tumors);
- **ocular symptoms**. Diplopia or "double vision" is due to the raised ICP which causes pressure on the abducens nerves. Their vulnerability is determined by their long course along

the cranial base;

- **symptoms of advanced intracranial hypertension syndrome.** The neurological examination reveals nuchal rigidity along with compensatory bradycardia and arterial hypertension. If left untreated, raised ICP leads to brain herniation (tentorial, subfalcine, tonsilar, etc.) which causes brain stem compression manifested by mydriasis, decerebration, autonomic disturbances and death.

Evolution

Brain tumors are characterized by gradual and progressive development of the clinical symptoms. The type of clinical evolution depends on tumor localization and histology. For example, fronto-basal meningiomas grow slowly within years and reach large size before presenting clinically, whereas medulloblastoma can cause acute hydrocephalus within weeks. However, it should be noted that the clinical course can follow acute or subacute evolution due to disturbed regional blood supply (ischaemia) or developement of severe cerebral edema leading to acute onset of intracranial hypertension.

Diagnosis of brain tumors

Diagnosis need to prove the existence of a brain tumor, to specify its location and its possible nature.

Brain tumors should be suspected if: a)epileptic (partial or generalized) seizures are registered; b)there is presence of focal neurological deficit; c)there are symptoms of raised ICP; d)the symptoms develop progresively

Diagnosis aims at establishing the presence of brain tumor, its localization and probable histology.

- **skull X-rays.** In children skull X-rays may show widened cranial sutures and the presence of “impressiones digitatae” (imprint of the brain gyri on the skull). In adults skull X-rays can identify changes in the cranial bones (in case of osteomas and meningiomas), presence of abnormal calcifications (typical of meningioma, oligodendroglioma or craniopharyngioma), enlargement or destruction of sella turcica (typical of pituitary adenomas), or expansion of the internal acoustic meatus (in case of vestibular neuroma);
- **computed tomography (CT)** examination should be

performed with multiple thin slices before and after contrast enhancement. CT scan confirms the presence of bone or brain lesions (single or multiple), the location of the tumor, its relationship to CSF pathways and cerebral vessels, the presence of peritumoral edema and the degree of "mass effect" (compression of the ventricles, midline shift and incipient herniation) (Fig. 4.5, Fig. 4.6, Fig. 4.7 and Fig. 4.8);

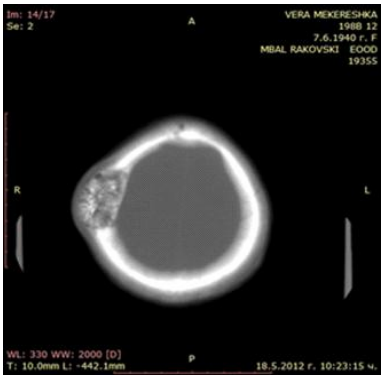

Fig. 4.5. CT image of osteoma affecting the skull

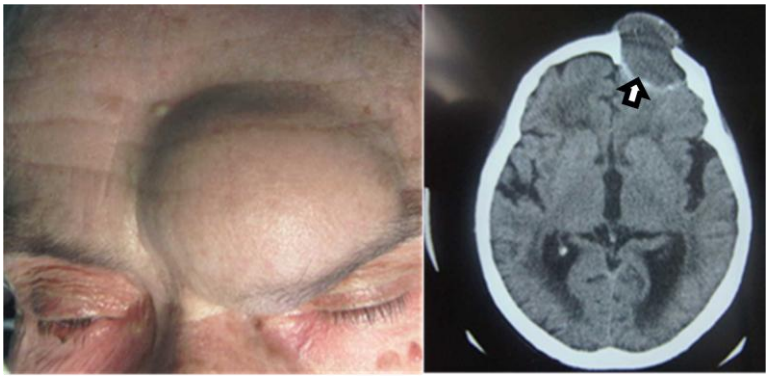

Fig. 4.6. CT image of osteoma in the skull with bone destruction

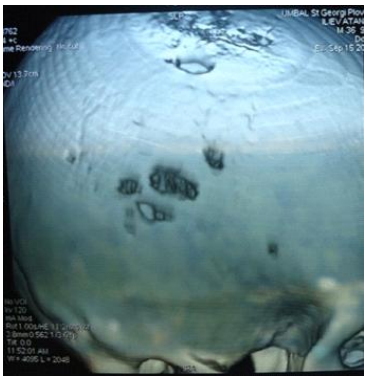

Fig. 4.7. CT - bone lesion caused by meningioma

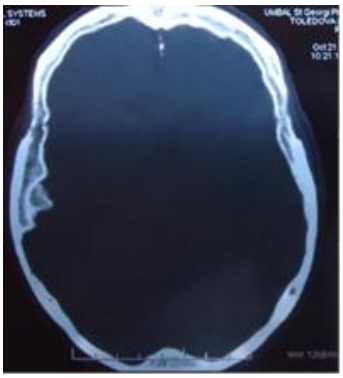

Fig. 4.8. CT - bone lesion caused by meningioma

- **magnetic resonance imaging (MRI) – a method of choice.** MRI provides superior details compared to the CT scan. It demonstrates more precisely tumor localization in three planes (axial, coronal and sagittal) and its relationship to the surrounding structures (neural, vascular and CSF) (Fig. 4.9);
- **cerebral angiography (CA)** can demonstrate the relationship of the tumor to major vessels and its blood supply (parasagittal meningioma, basal tumors, etc.).

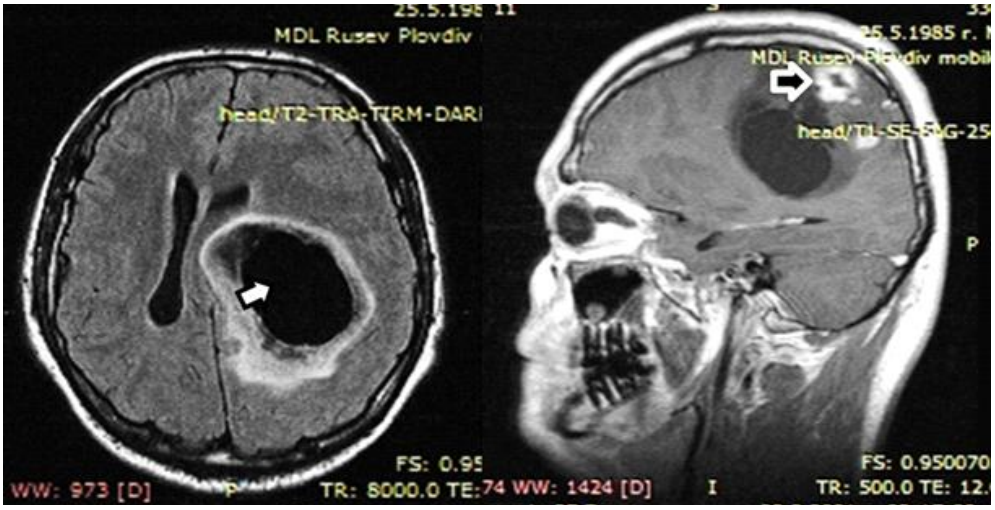

Fig. 4.9. MRI (axial and sagittal projection) of parietal oligodendroglioma: (➪) - cystic part; (➪) - calcification

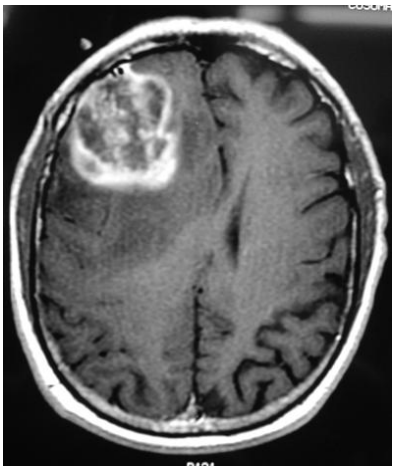

Fig. 4.10. MRI image of glioblastoma

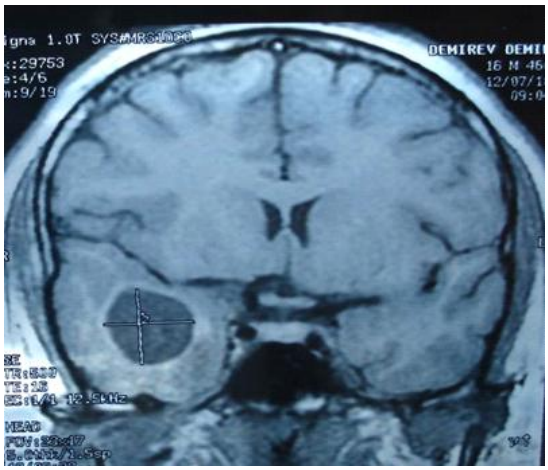

Fig. 4.11. MRI image of low-grade astrocytoma

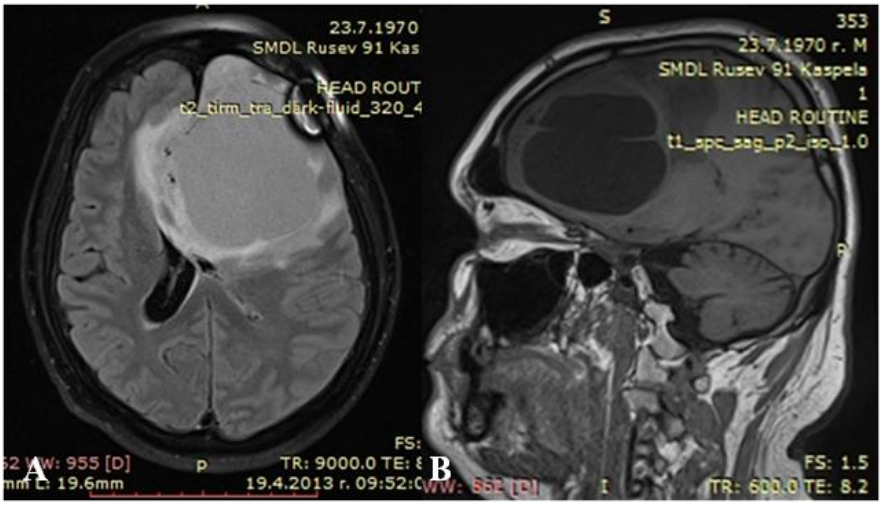

Fig. 4.12. MRI of astrocytoma grade II:
A/ axial image; B/ sagittal image

Brain tumors - basic differences, clinical presentation and treatment options

I. Supratentorial tumors

A. Lobar tumors. Histologic features:

1. Gliomas – the most common primary brain tumors in adults (60%) (astrocytomas, anaplastic astrocytoma, oligodendroglioma, oligodendroblastoma, mixed oligodendroastrocytoma). Generally, these are infiltrating tumors without clear demarcation from the surrounding brain tissue which often impedes complete tumor removal. As a rule, these tumors always recur. Glioblastoma is the most malignant tumor of this group, with rapid evolution and poor prognosis (average life expectancy varies between 6-8-12 months) (Fig. 4.10). Astrocytoma (fibrillary type) and oligodendrocytoma are slow growing tumors showing lower malignancy. They total extirpation is possible in certain cases. (Fig. 4.11 and Fig. 4.12).

2. Metastases – due to modern imaging modalities in clinical practice the incidence of secondary brain neoplasms reaches 30%-40% of all brain tumors. The peak incidence is between 45-60 years. Brain metastases could disseminate to the brain from different primary sources: lung cancer - 30-35%, breast cancer - 18-23%, gastrointestinal cancer – 6-7%, renal cancer – 5-6%. In 15-20% of the cases the primary source remains unknown. Brain is frequently affected by metastases from melanoma and choriocarcinoma (Fig. 4.13); 80% of metastases are located supratentorially. They can be:

- parenchymal metastases – 60% to 70%;
  - dural metastases – 1-3%;
  - menigeal carcinomatosis – 5-10%;
  - metastasis in the pituitary gland – rare.

In 50%-60% of the cases the metastases are single (Fig.4.14). Two metastases are found in 20% of the cases; three or more – in 13% of the cases. Frequently, the clinical presentation is due to only one of the metastases (Fig. 4.14 and Fig. 4.15).

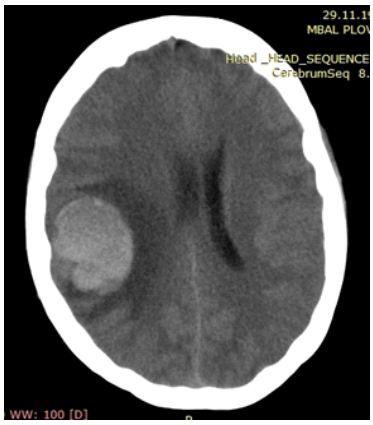

Fig. 4.13. CT image of metastasis of melanoma

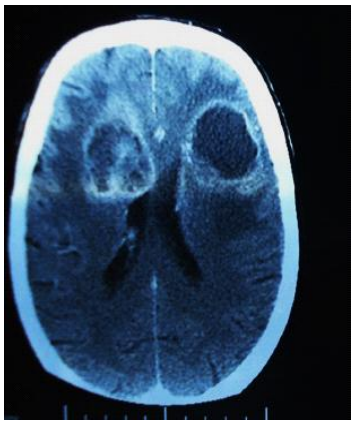

Fig. 4.14. CT image of bilateral metastases

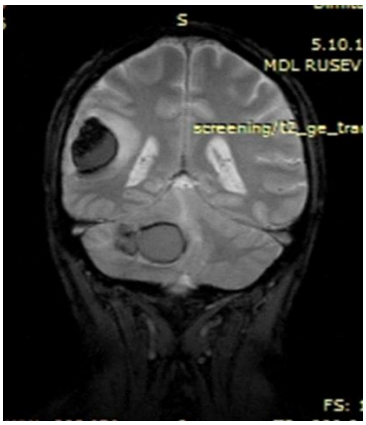

Fig. 4.15. MRI of supratentorial and subtentorial metastases.

3. **Meningiomas (15-23%)** are benign primary brain tumors with slow growth which originate from arachnoid cap cells. The peak incidence is between 30 and 60 years but in 75% of cases they are diagnosed over the age of 50. The female/male ratio is 2:1. Only 4-5% of meningiomas are found in children up to 15 years. Only 10% of meningiomas are located subtentorially. According to their attachment zone, menigiomas can be divided into:
   - **convexity meningiomas** represent about 25% of all cases. They are usually fed by dural arteries and branches of the middle cerebral arteries. From a surgical point of view they are easily approachable and can be totally removed (Fig. 4.16 and Fig. 4.17);

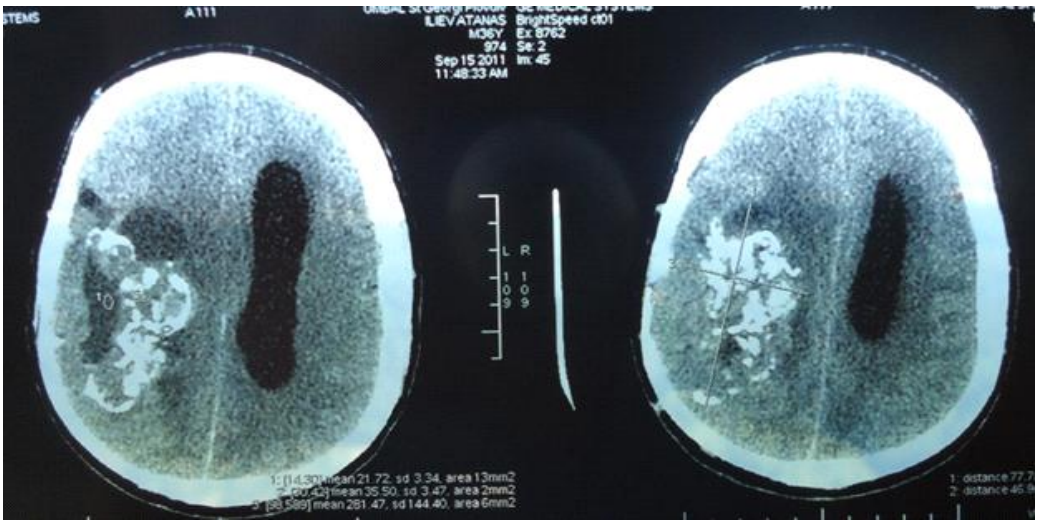

Fig. 4.16. CT image convexity meningioma with calcification

- **parasagittal meningiomas** are the most common type (28 - 35%). These tumors are located in close vicinity to the superior sagittal sinus without free space between the wall of the sinus and the place of tumor attachment to the dura. They are divided into three groups depending on their location in relation to the sinus:
  - anterior third of the sinus - located between the beginning of the sinus and the coronal suture;
  - middle third of the sinus - from the coronary suture to the lambdoid suture (Fig. 4.18, Fig. 4.19);
  - posterior third of the sinus – from the lambdoid suture to the confluence of the sinuses.

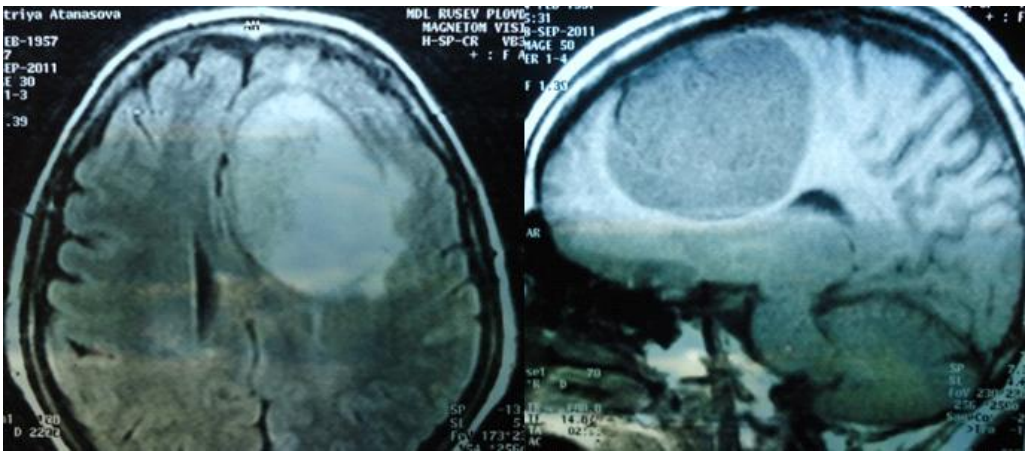

Fig. 4.17. MRI images (axial and sagittal projection) of convexity meningioma

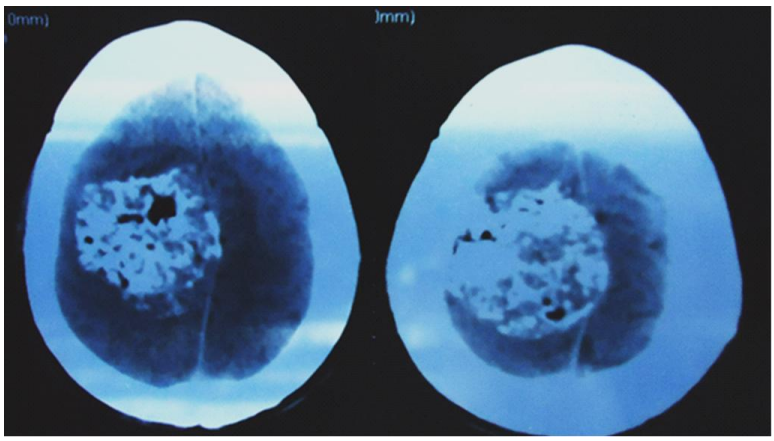

Fig. 4.18. CT image of a parasagittal meningioma in the middle third with calcifications

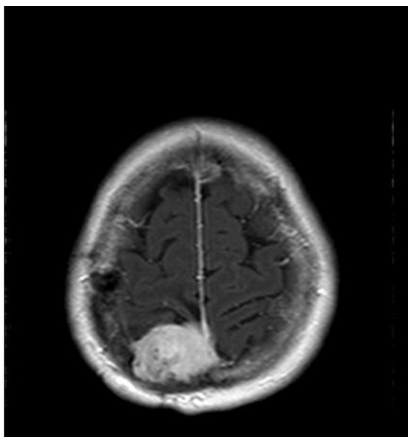

Fig. 4.19. MRI parasagittal meningioma in the middle third

- **falx meningiomas**. This type of tumors are attached to falx cerebri. They are covered by the brain parenchyma, often with bilateral and asymmetrical localization (Fig. 4.20).

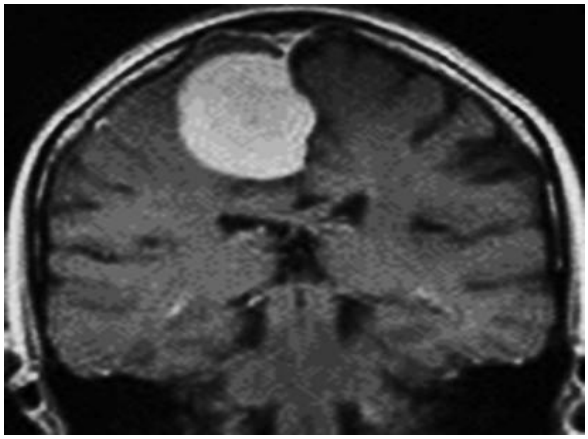

Fig. 4.20. MRI of falx meningeoma

Clinical features of lobar tumors - depend on location and histology of the lesion

1. Frontal tumors

They can be asymptomatic for a long period of time. Frontal tumors can present with symptoms of raised ICP. Generalized epileptic seizures are relatively common (40%). Psychiatric disorders are also typical – personality and mood disturbances, intellectual and cognitive decline, irrelevant behavior, etc. Motor deficit occurs later in the course of the disease. Gait disturbance with frontal ataxia can also occur, especially in tumors located in the frontocallosal region (Fig. 4.21). Motor aphasia presumes involvement of the dominant frontal lobe.

2. Tumors of the Rolandic area (the area in front and behind the central (Rolandic) sulcus)

The neoplasms which develop in this area often trigger Jacksonian motor and sensory seizures affecting the contralateral half of the body. The seizures are usually followed by resultant contralateral hemiparesis which is often transient but gradually becomes permanent. Focal neurological deficit (motor and sensory) occurs early and follows rapid evolution (Fig. 4.21).

3. Temporal tumors

Epileptic seizures are common (60%). There can be psychomotor seizures with hallucinations and secondary

generalization often preceded by olfactory or gustatory “aura”. Raised ICP symptoms are observed in 60% of cases and can be a debut of the disease when the tumor affects the non-dominant hemisphere. In the affected hemisphere the dominated area has speech violations. The motor deficit is discrete and occurs relatively late while visual disturbances (quadrantanopia) are more frequent (Fig. 4.23).

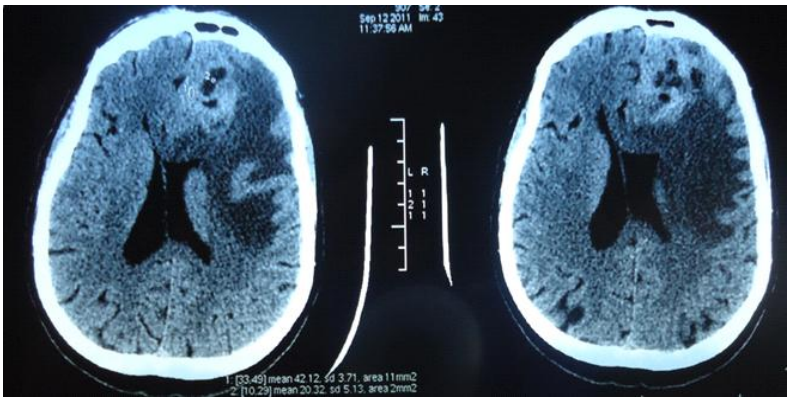

Fig. 4.21. CT image of a left frontal meningioma, compressing the frontal horns of the lateral ventricles

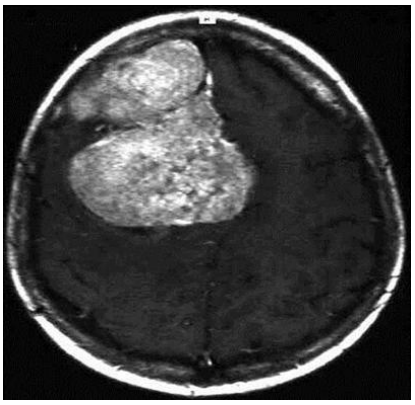

Fig. 4.22. MRI image of meningioma reaching the Rolandic area

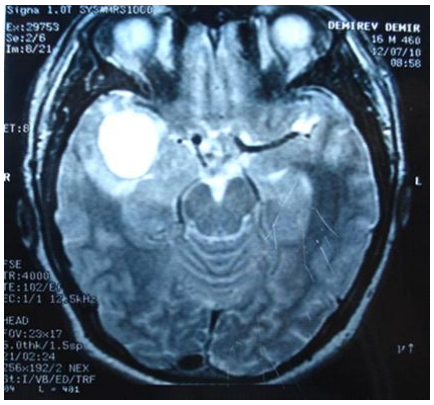

Fig. 4.23. MRI image of low-grade astrocytoma in the right temporal lobe

4. Parieto-occipital tumors

Intracranial hypertension is common (60%). There are typical epileptic seizures with visual hallucinations which can generalize. Visual deficits are frequent. Gerstmann syndrome may develop following lesions affecting the left parietal lobe in the region of the angular gyrus. It includes agraphia, acalculia, finger agnosia. Sensory aphasia develops as a result of damage to the Wernicke area (Fig. 4.24).

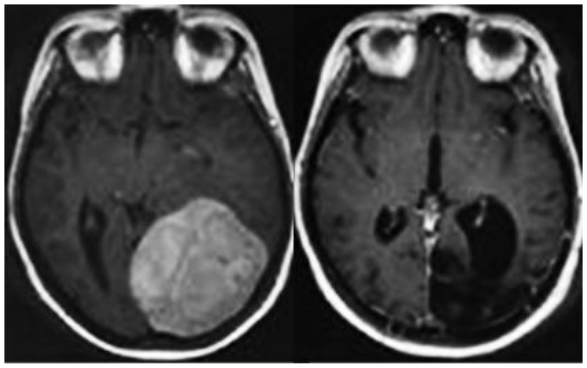

Fig. 4.24. MRI image of occipital meningioma before and after surgery

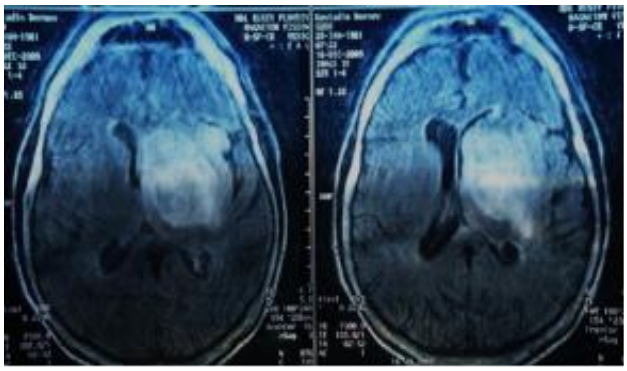

Fig. 4.25. MRI image of glioma affecting the left basal ganglia

Treatment of hemispherical tumors

Treatment of hemispherical tumors is surgical. Tumor histology influences the extent of tumor resection. An attempt should be made to achieve total tumor removal. Meningiomas are well demarcated tumors and should be totally excised. Metastases are also amenable to gross-total resection. Surgery is contraindicated in patients with multiple metastases and advanced systemic disease.

Gliomas are infiltrative tumors that include a broad spectrum of histological types and require individual surgical strategy for each case. For example, low-grade gliomas are indicated for total tumor removal (Fig. 4.23). In high-grade gliomas total cytoreduction is not possible. Therefore, the surgical treatment should be followed by adjuvant radiotherapy and in certain cases – chemotherapy. If there are general contraindications for open surgery, stereotactic biopsy is considered an option to obtain histological diagnosis (Fig. 4.25). Antiedematous treatment (steroids, mannitol) is initiated preoperatively in all cases with symptoms of intracranial hypertension. The presence of obstructive hydrocephalus requires preoperative CSF shunting.

Deep hemispherical tumors (thalamic and basal ganglia)

In these tumors, intracranial hypertension occurs early in the course of the disease while neurological deficits (sensory or motor) are discrete and occur relatively late. Extrapyramidal symptoms are observed in cases with basal ganglia involvement. Tumors in this area are mostly of glial origin (Fig. 4.25).

Intraventricular tumors

- **tumors of the lateral ventricles.** This group includes choroid plexus papilloma, meningioma, ependymoma, etc. These types

of tumors often cause hydrocephalus which presents with signs of elevated ICP (Fig. 4.26 and Fig. 4.27);

- **tumors of the 3rd ventricle**
  - **tumors of the anterior part of the 3rd ventricle.** Colloid cysts are relatively rare intracranial lesions located in the rostral aspect of the third ventricle. They may produce acute hydrocephalus, brain herniation, and lead to death (Fig. 3.28). Most reported cases occur in the third to fifth decades of life. Their incidence is 0.6 - 1.5% of all brain tumors. The clinical picture of colloid cysts typically develops in two directions:
    - ✓ **presentation with paroxysmal crises** - sudden blockage of CSF flow (obstruction of the foramina of Monroe) can provoke sevre diffuse headache with vomiting, decreased tone of the lower extremities for 5-10 min, impaired thermo-regulation, vasomotor and psychiatric distur-bances. These symptoms are transient but occasionally may lead to sudden death;
    - ✓ **presentation without paroxysmal crises** - includes symptoms of progressive intracranial hypertension, psychiatric and endocrine disorders (amenorrhea and precocious puberty).
- **tumors of the posterior part of the 3rd ventricle and pineal region**. They present 1-2% of all intracranial tumors and approximately 11% of tumors in childhood. There is a wide variety of histological types:
  - germinoma – 50% of tumors in the pineal region. They are eight times more common in males, occur mostly from 15 to 25 years;
  - embryonic carcinoma - 5%;
  - choriocarcinoma - 5%;
  - teratoma (with varying degrees of maturity) - 18-20%, mainly in children and adolescent males (Fig. 4.29);
  - endodermal sinus tumors - 7%;
  - pinealoma and pineoblastoma – 20%. These specific tumors of the pineal gland are found in children before the age of 10 in both male and female.

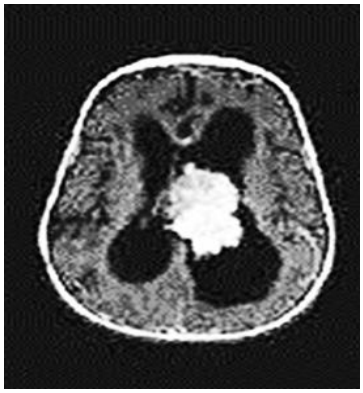

Fig. 4.26. CT image of choroid plexus papilloma causing hydrocephalus

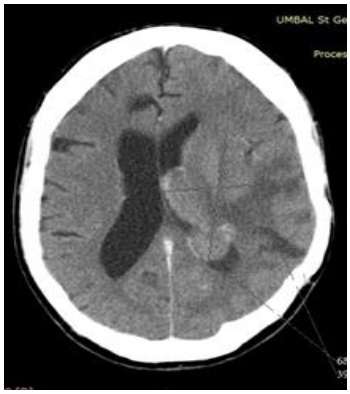

Fig. 4.27. CT image of ependimoblastoma

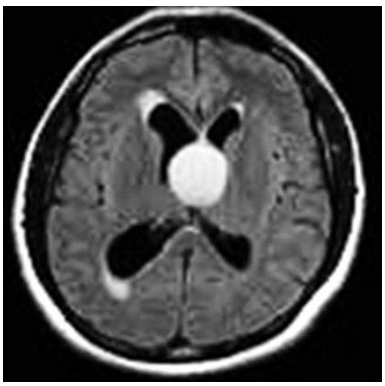

Fig. 4.28. CT image of colloid cyst

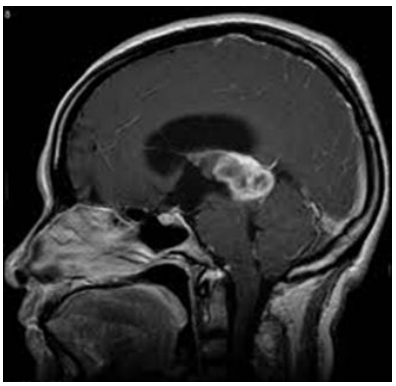

Fig. 4.29. MRI image teratoma in the pineal region

Clinical presentation:

- hydrocephalus and intracranial hypertension in 85-95% due to blockage of CSF pathways;
- oculomotor disorders - Parinaud's syndrome in 50%;
- endocrine disorders: diabetes insipidus (15%), precocious puberty, hypogonadism (7%);
- neuropsychiatric disorders: cerebrovestibular syndrome (20%), pyramid symptoms when capsula interna is affected (10%), mood disorders, somnolence, etc.

II. Skull base tumors

1. **Basal meningiomas** (located supratentorially - 35-45%)
   - **olfactory groove meningiomas** originate from the dura covering lamina cribrosa of the ethmoidal bone. They are relatively rare (approx. 10%), grow slowly and reach large size (Fig. 4.30). The clinical presentation is dominated by anosmia (or hyposmia), psychiatric symptoms typical of the frontal lobes;

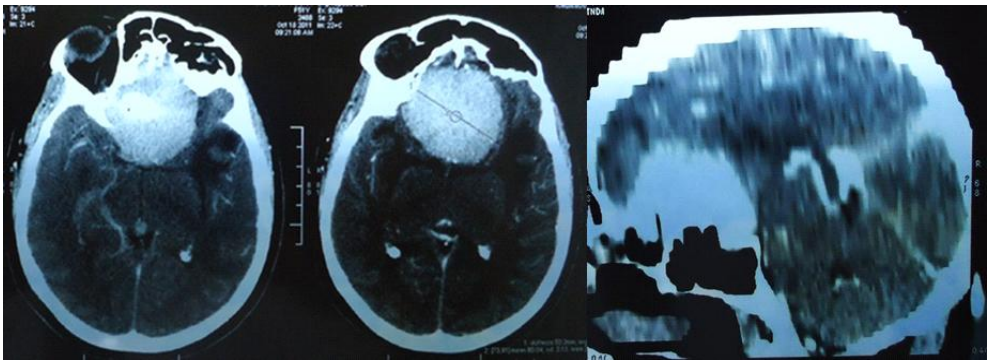

Fig. 4.30. CT image of olfactory meningioma (axial and sagittal reconstruction)

- **tubercullum sellae meningioma –** 8-9%. They present with visual disturbances due to compression of the optic chiasm. Larger tumors are more difficult to be removed due to the numerous vessels adherent to the tumor capsule;
- **sphenoid wing meningiomas – medial and lateral**. These tumors are located around the sphenoid ridge from the anterior clinoid process medially to the pterion laterally (Fig. 4.31, Fig.4.32 and Fig. 4.33);

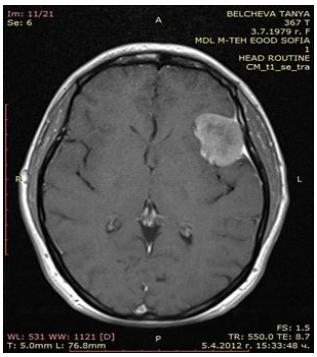

Fig. 4.31. MRI of meningioma of the greater wing of the sphenoid bone

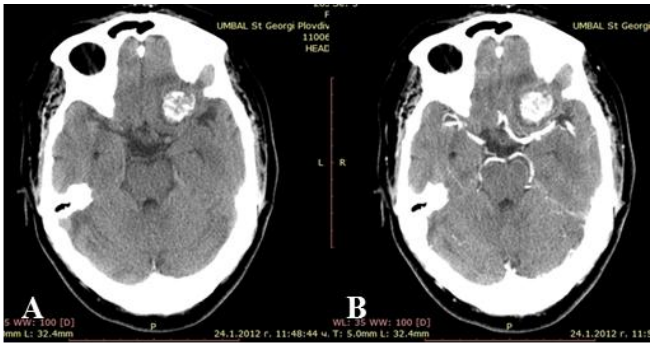

Fig. 4.32. CT image before (A) and after (B) contrast enhancement of meningioma of the lesser wing of the sphenoid bone**.**

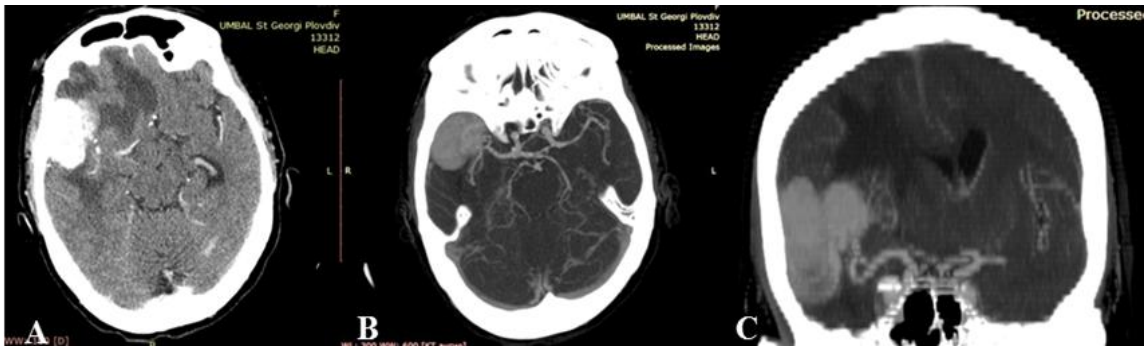

Fig. 4.33. CT image of meningioma of the greater wing of the sphenoid bone: A) axial image with contrast enhancement; B and C) CT angiography demonstrating tumor's blood supply

- **meningiomas of the cavernous sinus – 0.5 - 1%**;
- **temporobasal meningiomas** (4%). They originate from the meninges covering the region of the Gasserian ganglion and the apical part of the pyramid of the temporal bone (Fig. 4.34);
- **tentorial meningiomas** – approx 1% (Fig. 4.35).

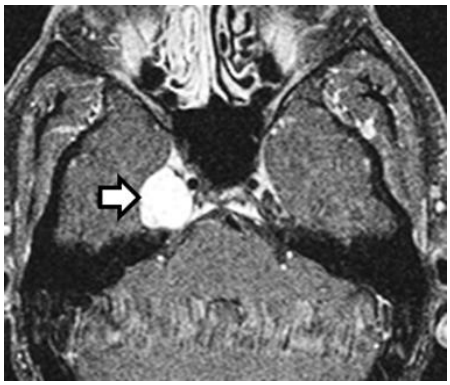

Fig. 4.34. MRI of cavernous sinus meningioma

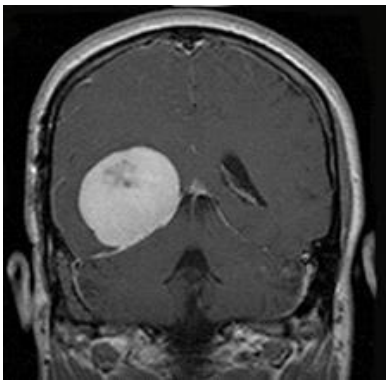

Fig. 4.35. MRI of meningioma of tentorium cerebelli.

III. Tumors of the pituitary gland

According to their size pituitary ademomas can be classified as follows:

- microadenoma – up to 1 cm;
- mesoadenoma – bigger than 1 cm, bulges the sellar diaphragm but remains intracellar;
- macroadenoma – with intra- and supracellular extension (Fig. 4.36);
- giant adenoma – with parasellar and suprasellar tumor invasion to the 3rd ventricle (Fig. 4.37).

Classification of pituitary adenomas in relation to their endocrine activity:

A. Functional (hormone-producing) adenomas:

- prolactinoma (galactorrhea, amenorrhea);
- growth hormone-producing (acromegaly, gigantism);
- ACTH-producing (Morbus Cusching, Nelson);
- TSH-producing (rare);
- gonadotropin (FSH or LH)-producing (rare).

B. Nonfunctional (non-producing) tumors (macro and giant adenomas).

Clinical features of the different forms of pituitary tumors:

- **tumor syndrome** – occurs after extracellar tumor expansion:
- **headache** – initially, it is due to stretching of the diaphragma sellae but alleviates after its rupture. It irradiates bilaterally to the temporal regions. Giant tumors cause headache as a result of intracranial hypertension;
- **visual disorders** – bitemporal hemianopsia is a typical sign of tumor enlargement which causes compression of the optic chiasm. Eventually, it results in blindness due to secondary optic nerve atrophy;
- **increased intracranial pressure** – giant tumors may obstruct the foramina of Monroe and cause hydrocephalus;

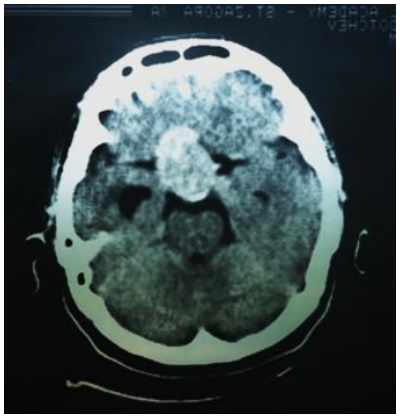

Fig. 3.36. CT image of pituitary intra- and supraselar adenoma

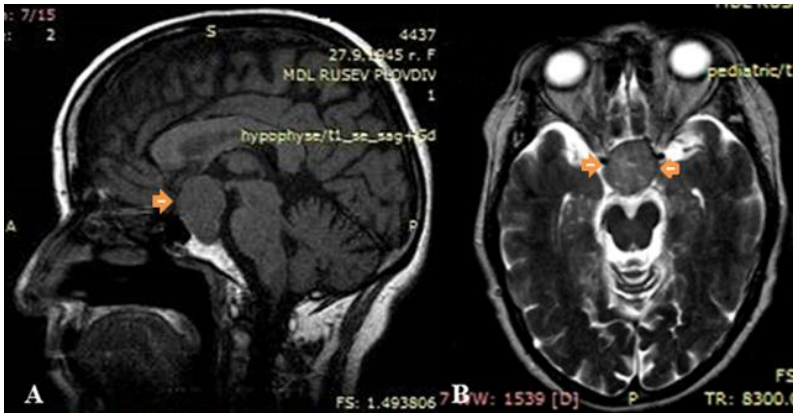

Fig. 3.37. MRI image of giant pituitary adenoma: **A/** sagittal image; **B/** axial image

- **pituitary apoplexy syndrome** - due to acute necrosis and/or hemorrhage within the tumor. Clinical onset is rapid - with

headache, impaired visual acuity and cranial nerve palsy;

- **endocrine syndrome**

Hypersecretion syndrome:

1. **Prolactinoma** (Fig. 4.38). It presents the most common pituitary tumor (30-40%)
   - **clinical features in females** (peak incidence between 28-30 years):
     - amenorrhea, dysmenorrhea;
     - galactorrhea (85%);
     - weight gain more than 5 kg (45%);
     - infertility in 30-40% of cases.
   - **clinical features in males** (15% of cases with an average age of 35):
     - tumor syndrome in 50-60% of cases;
     - decrease in libido, impotence, gynecomastia (30%).
   - **diagnosis of prolactinoma**:
     - increased serum prolactin levels over 150 ng/ml (normally 3-25 ng/ml). During pregnancy and lactation prolactin levels can reach normally 150-200 ng/ml;
     - skull X-rays – may demonstrate destruction of the sella turcica by macro- and giant adenomas;
     - CT with contrast enhancement – can diagnose macroadenomas and giant adenomas;
     - MRI with contrast enhacement is the method of choice – detects microadenomas, provides superior details of the tumor expansion and individual anatomy (Fig. 4.38).

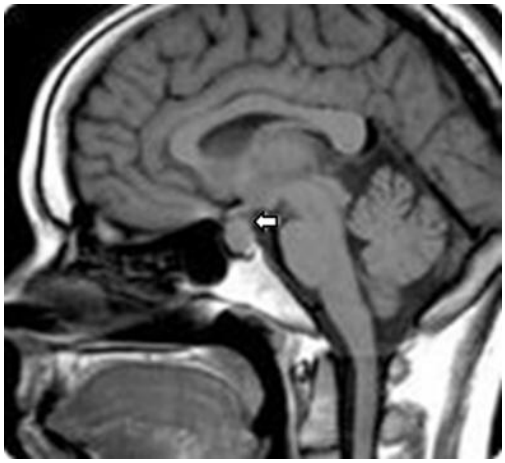

Fig. 4.38. MRI image of prolactinoma

2. **Growth hormone-producing adenoma**
   - causes acromegaly in adults and gigantism in children;
   - diagnosis:
     - serum levels of the growth hormone are increased (normal levels in children - 10ng/ml; adults - 5ng/ml;
     - skull X-rays, CT and MRI – similar to prolactinoma.
3. **ACTH-producing adenoma**
   - it causes Cushing's disease and has a frequency of about 5%. It is more often in females between 30 and 40 years;
   - **clinical presentation**:
     - weight gain and fatty tissue deposits, particularly around the midsection and upper back, in the face (moon face), and between the shoulders (buffalo hump);
     - pink or purple stretch marks (striae) on the skin of the abdomen, thighs, breasts and arms;
     - thinning, fragile skin that bruises easily;
     - slow healing of cuts, insect bites and infections;
     - acne;
     - generalized osteoporosis with osteopathic pain;
     - high blood pressure;
     - thicker or more visible body and facial hair (hirsutism) in females;
     - reduced potency in males;
     - psychiatric disorders – in case of Nelson syndrome;
     - melanodermia, asthenia, etc.
   - **diagnosis**:
     - MRI is the method of choice - detects microadenomas in 80% of the cases. It is necessary to exclude a tumor of the suprarenal glands;
     - laboratory examination reveales an increased serum levels of ACTH, cortisol and increased urinary levels of cortisol.

Craniopharyngioma

This is a benign brain tumor with slow evolution. It originates from embryonic cellular remains of the Rathke's pouch. In adults, the incidence varies between 2 and 4.5%, in children from 8 to 13% of all tumors in the sellar region. According to their location these tumors can be divided into:

- **intrasellar craniopharyngioma** - manifested by headache and pituitary hormone deficiency. Occasionally, the tumor may expand into the shenoid sinius;
- **intra and suprasellar craniopharyngioma** (Fig. 4.39);
- **suprasellar craniopharyngioma** - these tumors arise from the area of the pituitary stalk and infundibulum of the 3rd ventricle.

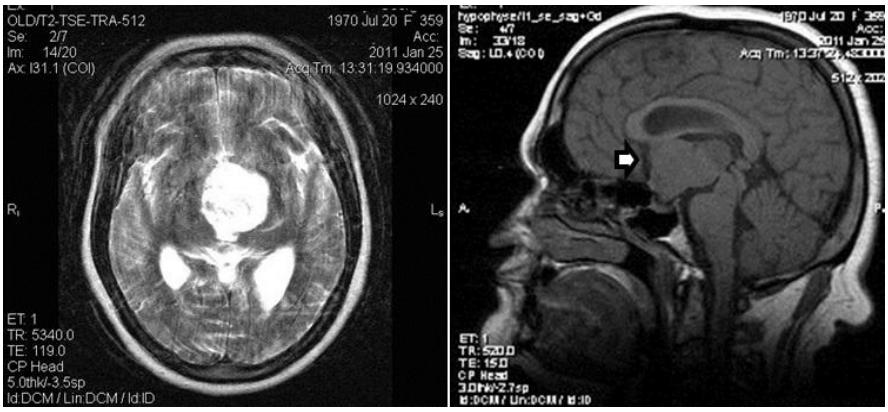

Fig. 4.39. MRI image (axial and sagittal) of intra- and suprasellar craniopharyngioma

Clinical presentation

- ophthalmic disorders (80-90%) – unilateral or bilateral loss of vision. Long-lasting compression of the optic chiasm may cause papilledema leading to secondary optic atrophy and blindness;
- endocrine disorders - are found in children in 65-90% and may lead to:
  - pituitary hormonal insufficiency;
  - genital hypoplasia;
  - adiposogenital syndrome.
- these may lead to:
  - **in children:** loss of growth in children (hormonal nanism), delayed puberty, disturbances in mental development;
  - **in adolescents:** hypogonadism;
  - **in females:** amenorrhea or irregular periods;
  - **in males:** reduced libido, impotence.
- neurologic complications:
  - intracranial hypertension in up to 60-70% of paediatric cases;
  - neurological deficit with loss of vision;

- psychiatric disorders (behavioral changes and Korsakoff syndrome).

- diagnosis:
  - skull X-rays – may detect calcifications of varying size and density;
  - CT with contrast enhancement - visualization the contours of the tumor and its relationship to surrounding structures;
  - MRI with contrast enhancement – method of choice, superior to CT.

IV. Subtentorial tumors

They represent 70% of neoplasms in children and can be classified as follows:

- tumors of the cerebellum and 4th ventricle;
- brainstem tumors;
- cerebelopontine angle tumors.

A/ Tumors of the cerebellum and 4th ventricle

1. Medulloblastoma

It originates from the vermis cerebelli and represents 4% of all intracranial tumors but 30% of neoplasms in the posterior cranial fossa. Approximately 75% of medulloblastomas occur up to 15 years with a peak incidence between 5 and 7 years. The male/female ratio is 2/1. The clinical presentation is dominated by symptoms of intracranial hypertension, commonly due to hydrocephalus. Vomiting is frequent and intense. Cerebellar symptoms such as ataxia and macrographia are also typical. This tumor can metastasize along the CSF system (Fig. 4.41). Diagnosis is made by contrast CT and MRI which demonstrate large rounded parenchymal tumor with necrotic areas, filling the 4^{th} ventricle (Fig. 4.40). Treatment includes surgery followed by radiation therapy (including the spinal axis) and chemotherapy. Small children (younger than 3 years) receive chemotherapy alone. If hydrocephalus is present, it should be shunted.

2. Ependynoma and ependimoblastoma of the 4^{th} ventricle

It is most often found in children (15%). It develops on a broad base from the roof of the 4^{th} ventricle and infiltrates the adjacent cerebellar tissue. This tumor also disseminates along the CSF pathways.

- clinical symptoms vary with age:
  - infants may develop acute hydrocephalus;
  - in older children and adults, tenacious vomiting can be

provoked by movements of the head, also known as Bruns syndrome. At a later stage, symptoms of intracranial hypertension, nystagmus and cerebellar symptoms may occur. If the tumor invades the brainstem, it may cause pyramid symptoms and damage to the cranial nerves.

- diagnosis is made by CT and MRI which visualize a tumor with heterogeneous appearance within the 4th ventricle. Obstructive hydrocephalus is often present (Fig. 4.42);
- treatment consists of surgical tumor removal followed by radiation therapy (except for children younger than 3 years).

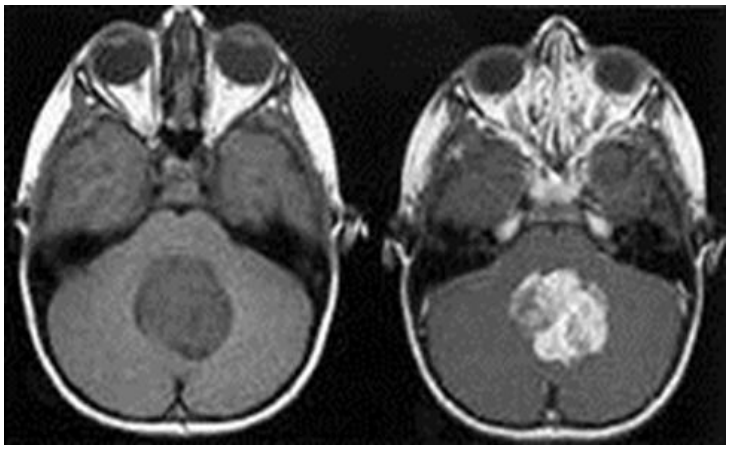

Fig. 4.40. MRI image of medulloblastoma

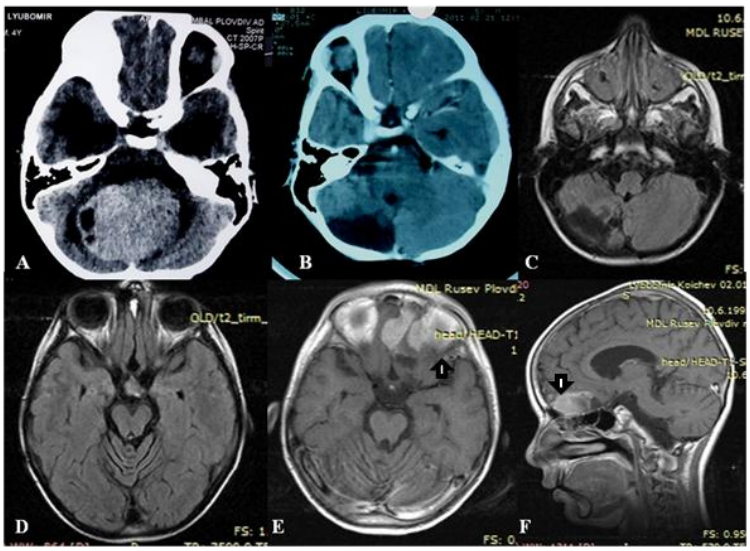

Fig. 4.41. Medulloblastoma: **A/** Preoperative CT image; **B/** Postoperative CT image – gross-total removal; **C**/ MRI at 6th postop. month – no evidence of recurrence; **D/** MRI at 1st postop. year – no evidence of CSF dessemination; **E** и **F/** MRI at 2nd postop. year (axial and sagittal images) – visible fronto-basal dissemination (histology: metastasis from medulloblastoma)

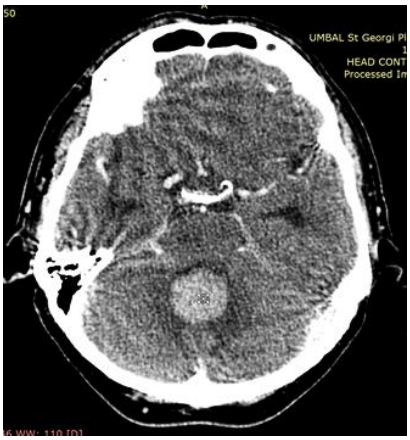

Fig. 4.42. CT with contrast enhancement
ependymoma of the 4th ventricle

3. **Tumors of the cerebellar hemispheres** can be diagnosed at any age:
   a) **Astrocytomas** present about 30-35% of all subtentorial tumors in children and are most common between 5 and 10 years;
      - the clinical evolution is slow and presents with local cerebellar symptoms which precede the symptoms of increased intracranial pressure;
      - diagnosis is made by CT and MRI which demonstrate parenchymal cerebellar tumor which can be cystic, solitary or mixed;
      - treatment is surgical. Gross-total removal of juvenile pilocytic astrocytoma in children results in definitive cure. If resection is not total, surgery should be followed by radiotherapy.
   b) **Hemangioblastoma** is a benign tumor originating from capillary endothelium. They are 1-2% of all intracranial tumors, nearly 90% of them are located subtentorially. 72% are located in the cerebellar hemispheres, 16% - in the vermis region and 2-3% in the region of the tonsils. The rest of the cases affect the brainstem (floor of the 4th ventricle). Average age is between 30-35 years, males are predominantly affected. In 15-20% these tumors can be hereditary within von Hippel-Lindau disease.
      - the clinical presentation is dominated by symptoms of increased ICP with acute evolution;
      - diagnosis is made by CT and MRI with contrast enhancement which shows cystic tumor of hyperdense structure attached to its wall (Fig. 4.43).

- treatment includes surgical removal of the tumor.

c) Cerebellar metastases. In adults, metastases originate mainly from lung (in males) and breast (in females) cancer (Fig. 4.44) They can be single or multiple (Fig. 4.45):

- the clinical presentation follows rapid evolution with signs of intracranial hypertension and cerebellar symptoms;
- diagnosis is made by CT and MRI with contrast enhancement;
- treatment is surgical.

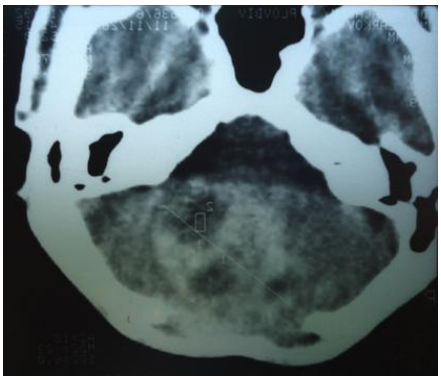

Fig. 4.43. CT image of hemangioblastoma

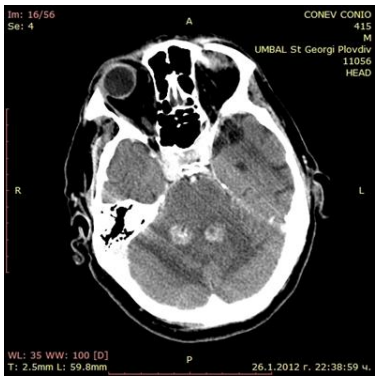

Fig. 4.44. CT image of multiple metastases

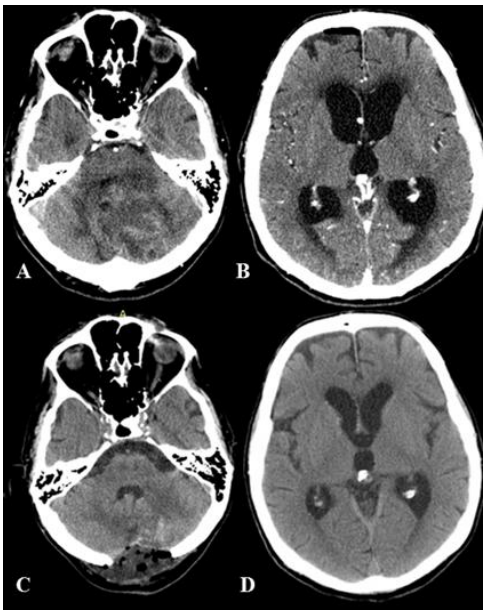

Fig. 4.45. CT image of cerebellar metastasis: **A/** tumor formation affecting the vermis and the cerebellar hemispheres, predominantly on the left; the 4th ventricle and posterior fossa subaracnoid spaces are compressed; **B/** CT image of obstructive hydrocephalus in the same patient; **C/** Postoperative CT demonstrates total tumor removal with decompression of the 4th ventricle and posterior fossa subaracnoid spaces; **D/** considerable reduction of the hydrocephalus

B/ Brainstem tumors.

The brainstem is usually affected by infiltrative tumors of glial origin. Peak incidence in adults is between 30-35 years. These lesions comprise 12-17% of all paediatric tumors. Their evolution depends on the tumor malignancy. Low-grade gliomas are more common in childhood whereas high-grade gliomas are typical of adulthood.

- **the clinical symptoms** progress slowly and have insidious onset. Gradually, one or more cranial nerves are affected, often bilaterally. The pyramid and sensory pathways are also involved (60%). Cerebellar symptoms develop at a later stage. Intracranial hypertension is rare because only 10% of these tumors propagate to the 4th ventricle and cause CSF obstruction;
- **diagnosis**: Modern imaging diagnosis is achieved by MRI with contrast enhancement (Fig. 4.46). Generally, the pons harbors high-grade lesions while the mesencephalon and medulla oblongata are affected by low-grade lesions;
- **treatment**: Modern surgery allows substantial activity, especially, in cases with benign tumors, but the surgeon should properly weigh "risks versus benefits". Stereotactic biopsy is successfully used with lower risks, usually, followed by chemotherapy and radiation therapy. Encouraging results are achieved with the application of modern radiosurgery by means of gamma-knife or cyber-knife for lesions with diameter of less than 2 cm.

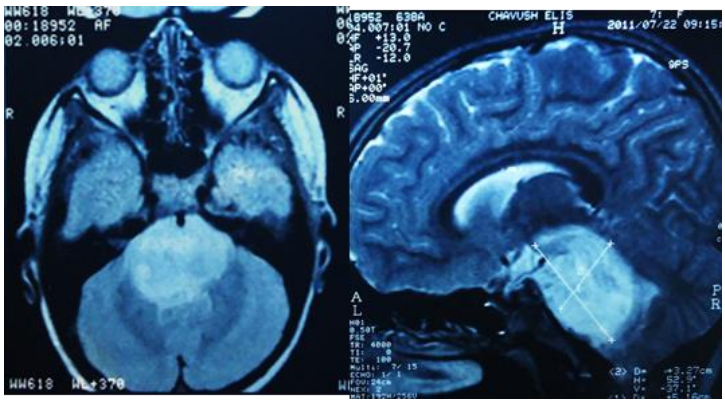

Fig. 4.46. MRI images (axial and sagittal) of glial brainstem tumor

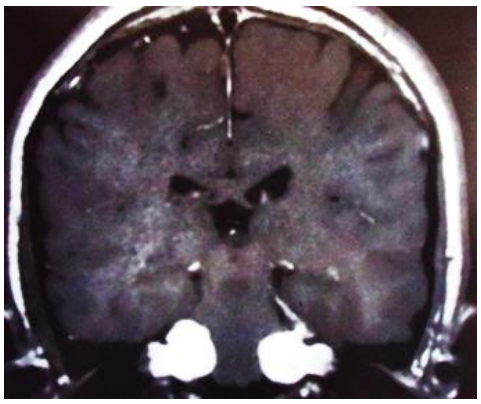

Fig. 4.47. MRI image bilateral vestibular neuroma

C/ Cerebelopontine angle tumors

1. Neuroma of the VIIIth cranial nerve (Acoustic neuroma (AN) or vestibular schwannoma). This type of tumor is often called acoiustic neuroma, despite it is actually schwannoma arising from the vestibular subdivision of the statoacoustic nerve at the site of ganglion vestibuli (Scarpi). The incidence is 8% of all intracranial tumors and 25% of all posterior fossa tumors. In 5% of cases AN are bilateral which is typical of neurofibromatosis type 2 (Fig. 4.47). AN is most common between 40 and 50 years with female/male ratio 3/2. AN is benign slow-growin encapsulated tumor which initially develops within the internal acoustic meatus. The Koos classification of AN is based on the tumor size:

- **stage I** – intracanalicular neuroma;
- **stage II** – the tumor has grown to cerebellopontine cistern;
- **stage III** – the tumor is in contact with the brainstem;
- **stage IV** – the tumor compresses and dislocates the brainstem.

- the clinical presentation includes the following stages:
  - **otoneurological (early) stage**: it begins with excitation of the acoustic nerve which creates unilateral noise in the ear called ‘tinnitus’. Subsequently, there is gradual loss of hearing in the ear (hypacusis to anacusis) (90%). Nausea, dizziness and imbalance are rarely observed in this stage;
  - **stage of neurological deficit:** this stage may include development of vestibular syndrome, involvement of the trigeminal nerve (facial paresthesia and numbness, decreased corneal reflex, rarely - trigeminal neuralgia), facial palsy. In larger tumors, cerebellar symptoms and caudal cranial nerve (IX, X, XI and XII) damage may be present. If the brainstem is compressed, pyramid symptoms may develop;
  - **stage of intracranial hypertension with hydrocephalus** – observed in the advanced stage of the disease (Fig. 4.48).
- **diagnostic procedures**:
  - **audiogram** (auditory and vocal) establishes retrocochlear conductive hearing loss;
  - **auditory evoked potentials** detect increase in the intervals I - II and II – IV;
  - **skull X-ray (Stenvers projection)** shows expansion of

the internal acoustic meatus on the affected side;

- **computed tomography and magnetic resonance imaging**. CT provides data about the volume and the structure of the tumor, its relationship to surrounding structures and the possible presence of bone changes (Fig. 4.48). MRI reveales the precise relationship of the nerves with neighboring brain structures, which is essential for surgery (Fig. 4.49).

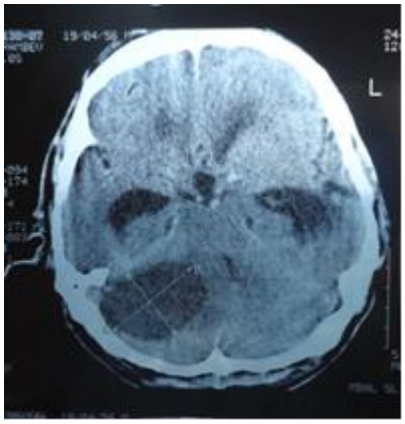

Fig. 4.48. CT image of acoustic neuroma with hydrocephalus

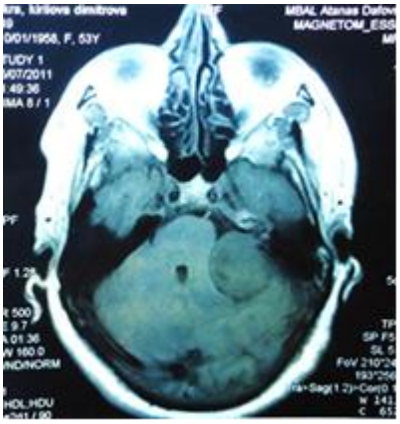

Fig. 4.49. MRI image of acoustic neuroma

Treatment aims at total tumor removal with hearing and facial nerve preservation in stages I and II. In stage III and IV this is not always possible. Despite the advance of modern imaging, only one of three cases is diagnosed early in the course of the disease (Stage I - 11%, Stage II - 23%).

2. **Other cerebellopontine angle tumors:**
   a) Cerebellopontine angle meningiomas (10%) (Fig. 4.50);

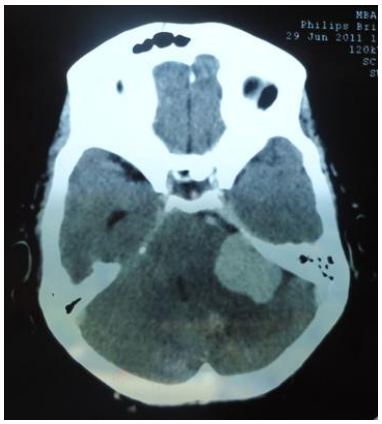

Fig. 4.50. CT image of meningioma of left cerebellopontine angle

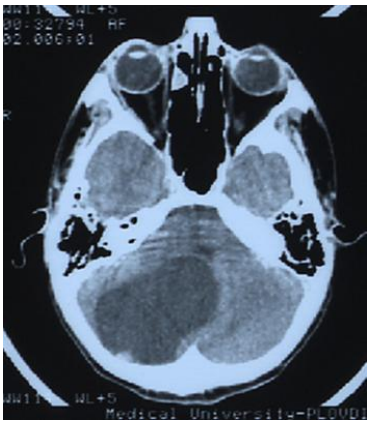

Fig. 4.51. Cholesteatoma in the right cerebellopontine angle

b) **Epidermoid cyst (cholesteatoma)** (3%): manifested by discrete cerebellopontine and cerebellar symptoms, trigeminal neuralgia (Fig. 4.51);
c) **Trigeminal neuroma** (2%): It can originate from the site of the nerve entry zone or Gasserian ganglion (Fig. 4.52). Clinical presentation may include pain followed by numbness in the nerve's distribution area;

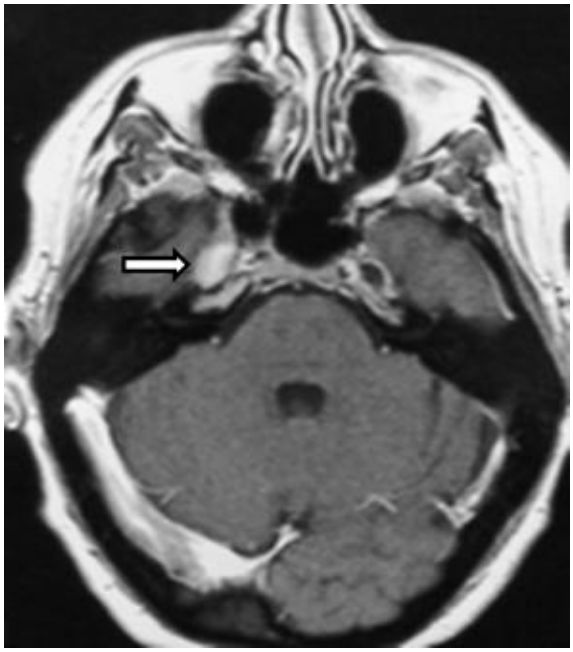

Fig. 4.52. MRI of neuroma
of the Gasserian ganglion

d) **Facial nerve neuroma** (1%): rare tumors that present with hemifacial spasm or facial palsy. Subsequently, loss of hearing may develop on the affected side as a result of damage to the acoustic nerve.

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CHAPTER 5 *I. Koev, M. Staykova, I. Batakliev*

TUMORS OF THE SPINE AND SPINAL CORD

Tumors of the spine and spinal cord present with syndrome of spinal cord compression. This syndrome can also be caused by other pathological conditions such as spinal abscess, hematomas, parasitic disease, disc herniation and spinal injuries. In this chapter we will focus on the compression syndrome caused by spinal oncological diseases. Narrowing of the spinal canal results in compression of the spinal cord, its vessels (arterial and venous) and nerve roots. For this purpose, it is necessary to recall the functional organization of the spinal cord in order to properly understand the clinical presentation of spinal cord compression which depends on the localization of the pathological process (Fig. 5.1).

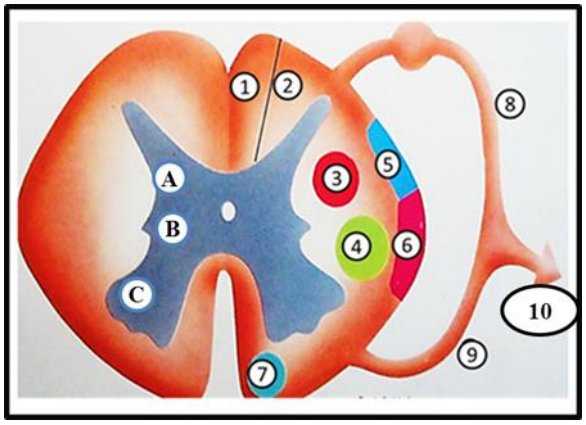

Fig. 5.1. Functional distribution of the spinal cord:
1. and 2. Goll and Burdach tracts (proprioception); 3. Pyramid tract; 4. Spinothalamic tract (sensation of pain and temperature); 5. and 6. Spinocerebellar tract; 7. Anterior spinothalamic tract; 8. Dorsal (sensory) nerve root; 9. Ventral (motor) nerve root; 10. Spinal nerve;
A/ Dorsal horn **B/** Lateral horn **C/** Ventral horn

Syndrome of spinal cord compression with slow evolution, caused by space-occupying process

Spinal cord damage results from mechanical compression and/or ischemia due to compression of its feeding and draining vessels. If the neurological deficit is partial, surgical decompression may lead to good or excellent recovery. If the neurological deficit is complete (total loss of motor and sensory functions), the prognosis for postoperative recovery is dismal.

Clinical presentation:

1. **Regarding the spine:**
   - pain along the spine axis which increases during strain (coughing, defecation) and does not alleviate at rest;
   - paravertebral muscle spasm in the region of the affected segment;
   - palpatory pain provoked by pressing the spinous process overlying the pathological process.
2. **Regarding the spinal nerve roots:**

A/ Sensory root involvement:

- **excitatory stage** – radicular pain and/or paresthesias which show dermatomal irradiation and depend on the level of impairment (cervico-brachialgia, intercostal neuralgia, lumboradiculalgia). The pain is constant. It does not improve at rest and can be provoked by strain. The pain follows progressive course and can be the only disease symptom for a long period of time;

In order to achieve optimal surgical results, the diagnosis should be made at this stage.

- **stage of sensory deficit** – there is sensory deficit (hypoesthesia or anesthesia) of dermatomal type.

B/ Motor root involvement

- is manifested by a partial or total paralysis of a particular muscle or muscle group innervated by the affected nerve root (areflexia, hypotrophy, paresis or plegia).

3. **Regarding the spinal cord**: Generally, the clinical presentation depends on the site of spinal cord compression (Fig. 5.2):
   - if the tumor is dorsally located, it presents with radicular pain, loss of proprioception and vibratory sensation. Subsequently, motor deficit can occur below the level of compression;
   - tumors, located ventrally to the cord, present with motor defict;
   - tumors, located laterally to the cord, may lead to Brown-Sequard syndrome.

Approximately 66% of spinal tumors are benign and 34% -

malignant. They are found along the entire spinal axis, but with different distribution across its regions:

- cervical - 18%;
- thoracic - 66%;
- lumbar - 15%;
- sacrum - 1%;

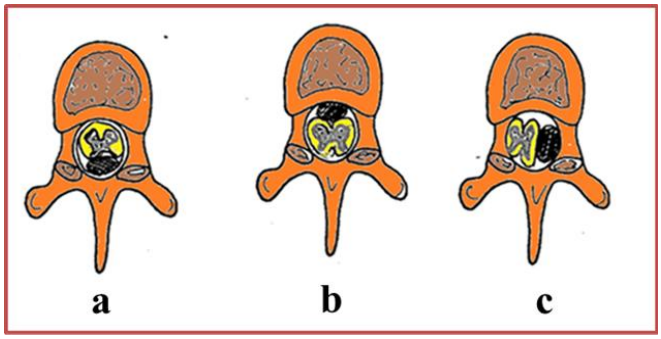

Fig. 5.2. Location of the tumor in relation to the spinal cord:
a/ dorsal; **b/** ventral; **c/** lateral

Spinal tumors can be classified as follows: extradural, intradural -extra-medullary and intramedullary tumors (Fig. 5.3).

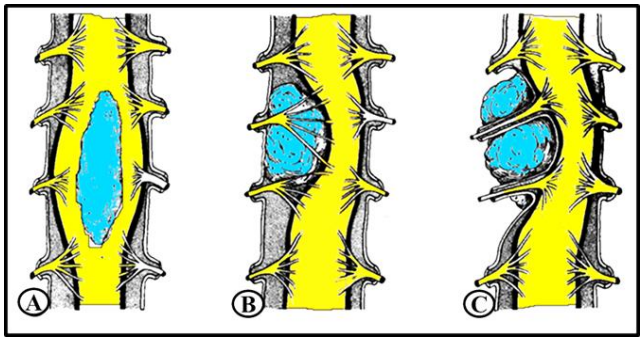

Fig. 5.3. Types of spinal cord tumors:
A/ intramedullary; **B/** intradural-extramedullary; **C/** extradural

I. Intramedullary spinal cord tumors

- approximately 5% of all CNS tumors and 20% of all spinal tumors;
- **clinical presentation:** Intramedullary neoplasms are infiltrative lesions which grow slowly. The pain is the most common initial symptom. It is usually diffuse and irradiates across the spinal axis. Patients often feel it as an electrical current that runs down the back and into the limbs (Lhermitte

symptom). Most intramedullary tumors originate from the gray matter of the spinal cord and resemble the symptoms of syringomyelia (progressive weakness in the arms and legs, stiffness in the back, shoulders, arms, or legs, and chronic, severe pain). Loss of conductive functions develops later. Movement disorders are initially manifested by atrophy and fasciculation of individual muscle groups. As the disease progresess, patients develop paretic symptoms with spasticity distal to the damaged level. If the neoplasm originates from the conus medullaris, it presents early with bowel and urinary disturbances.

Types of intramedullary tumors – over 80% of these tumors are astrocytomas and ependymomas:

1. Intramedullary astrocytoma. In 80-90% of cases, it is located in the cervicothoracic region, extending across 5-6 segments. It contains cystic and solid part with unclear infiltrative borders. Astrocytomas occur equally in both sexes, the average age of patients is 30 years.

2. Intramedullary ependymoma. It originates from ependymal cells covering canalis centralis of the spinal cord. 50% of the tumors are located in the region of the conus medullaris. These tumors usually extend across several spinal segments. Ependymomas are well demarcated, thus gross total resection is possible. Malignant forms are rare (Fig. 5.4).

3. Hemangioblastoma (10%). Most of these tumors are cystic and may propagate outside the spinal cord (Fig. 5.5).

4. Intramedullary metastases are extremely rare.

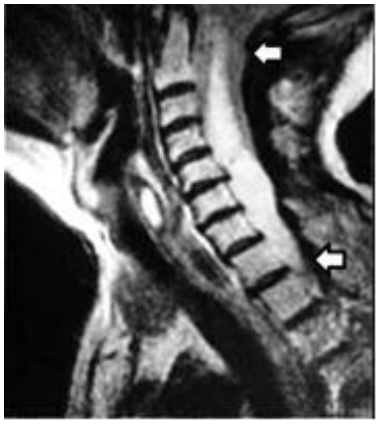

Fig. 5.4. MRI image of intramedullary ependymoma from C_2 to Th_2 level

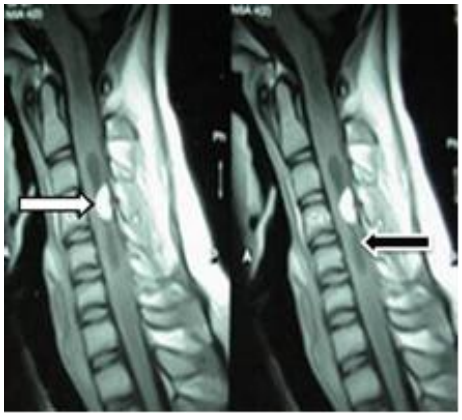

Fig. 5.5. MRI image of intramedullary hemangioblastoma

II. Intradural-extramedullary spinal tumors

- **clinical presentation:** the clinical symptoms are determined by their localization in relation to the spinal cord segment and cross section of the medulla. The pain is a common initial symptom which increases with strain. Spinal schwannomas (neuromas) cause radicular pain across the nerve distribution area followed by radicular (motor and sensory) deficits. Conductive sensory and motor deficits occur later and evolve slowly.

High cervical, conus medullaris and cauda equina tumors demonstrate specific manifestation:

1. Tumors with high cervical location (above C_4 segment) may damage caudal cranial nerves, caudal part of medulla oblongata and upper cervical segments of the spinal cord. This leads to a variety of clinical symptoms that mimic multiple sclerosis. High cervical tumors should be suspected if there is evidence of fasciculation and weakness of the tongue, weakness and dissociated sensory deficits in the limbs. Frequently, these complaints are accompanied by hyperacusis, nystagmus and ataxia.

2. Conus medullaris tumors may cause symmetrical paraparesis and hypesthesia and early bowel and bladder disturbances.

3. Cauda eqiuna tumors may cause radicular pain irradiating toward legs. Subsequently, bilateral dermatomal sensory loss and isolated muscle paralysis develops. Urination can also be affected (Fig. 5.6).

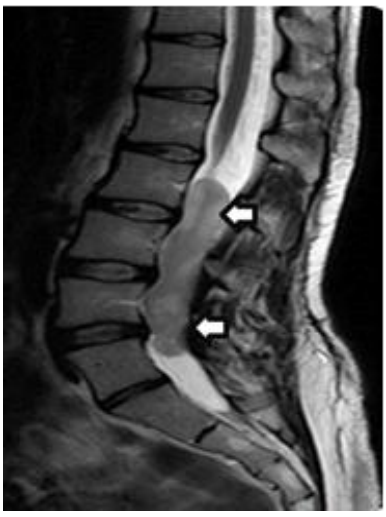

Fig. 5.6. MRI of ependymoma of cauda equina L_3-S_1

Types of intradural-extramedullary spinal tumors:

1. Meningioma – 20% of intradural tumors. In 80% of cases this occurs in women over 55 to 60 years. Their most common location is the upper thoracic region, followed by the cervical and lower thoracic region (distal to Th_9). They may have dorsolateral, ventrolateral, pure ventral and dorsal location in relation to the medulla (Fig. 5.18). Radicular pain is not typical of meningiomas.

2. Neuroma (schwannoma) – comprise about 25% of intradural tumors with benign nature and slow evolution. Frequently, they arise from the dorsal (sensory) nerve roots and are usually located in the thoracic and thoracolumbar region. Some neuromas, called "dumbbell schwannomas", may extend through the neural foramen to the paravertebral region (Fig. 5.21). The average age of the patients is between 30 and 40 years without gender predilection.

3. Other intradural-extramedullary tumors: These neoplasms are rare. Some of them are located in the subarachnoid space. This group includes epidermoid cysts, lipomas (Fig. 5.22) ependymomas (Fig. 5.23) and metastases.

III. Extradural spinal tumors

This group includes tumors with pure extradural location as well as those originating from the spine (primary and secondary) which expand to the epidural space.

1. Tumors with pure epidural location without involvement of the bones of the spine are very rare (Non-Hodgkin's lymphoma - 0.8% to 1.2%). Usually, they grow within the dorsal epidural space affecting 2-3 segments, typical of males between 50-60 years. Pure epidural metastases are rare.

2. Primary tumors of the spine:

- **Osteidosteoma** is a benign tumor which in 10% of the cases can affect the spine, most often in males in the third decade. It consists of bone nest surrounded by fibrovascular tissue and marked bone reaction (Fig. 5.7). It is commonly located in the cervical and lumbar region and involves the vertebral arch, joint and spinous process. The most frequent clinical sign is pain (70%) while symptoms of neural compression are rare;
- **Osteoblastoma** is a benign tumor which in 40% of cases is located in the spine. Its appearance resembles osteidosteoma, but with larger size and higher frequency of compression of neural structures (33%);

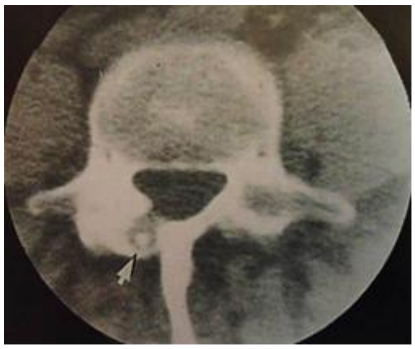

Fig. 5.7. CT image of osteidosteoma

- **Giant cell tumors** affect the spine in 5-8%, especially, the sacral region (Fig. 5.8). Although they have benign histological features, they follow invasive and destructive evolution. Clinically, they manifest with rachialgia and symptoms of compression of the cord or cauda equina. Malignant transformation is rare but possible;
- **Hemangiomas** are the most common primary tumors of the spine. They often affect more than one vertebra (33%) and are located mainly in the thoracic region in adults. Clinical manifestations are rachialgia, less frequently radicular and myelopathic symptoms (Fig. 5.9);

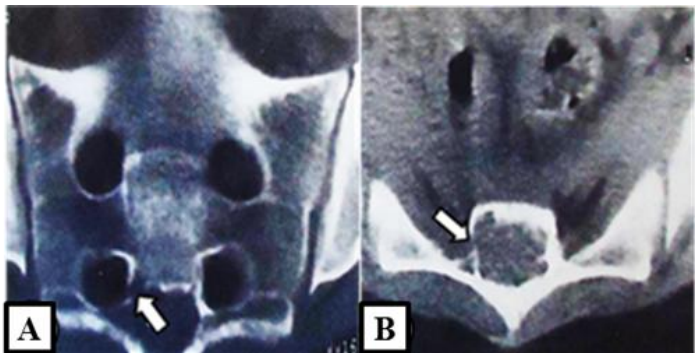

Fig. 5.8. CT image of giant cell tumor of the sacrum (coronal and transverse projections)

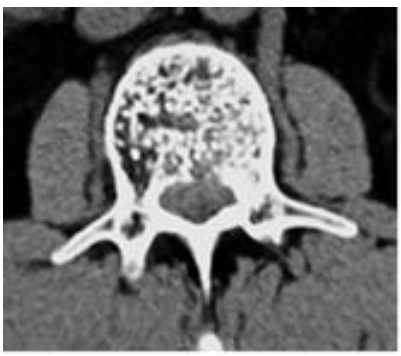

Fig. 5.9. CT image of hemangioma

- **Eosinophilic granuloma –** rare benign tumor of the vertebral body in children and adolescents. There is a possibility of a spontaneous cure. The most common locations are the sacrococcigeal region and the bodies of the lower lumbar vertebrae (15%). Pain in the lumbosacral region is common.

Perianal pain and urinary distarbances are typical of advanced cases. This tumor can invade the pelvic cavity;

- **Osteosarcoma –** rare malignant tumor of the spine (6% of all malignant tumors of the spine). Apart from being primary, these tumors can also metastasize from other parts of the body (metastatic osteosarcoma). Its clinical manifestation includes rahialgia and compression syndrome;
- **Chondrosarcoma –** rare malignant tumor of the spine. In 25-30% of cases, it originates from osteochondroma as a result of malignant transformation. It presents with rahialgia and compression syndrome. Tumor recurrence is common (Fig. 5.10);

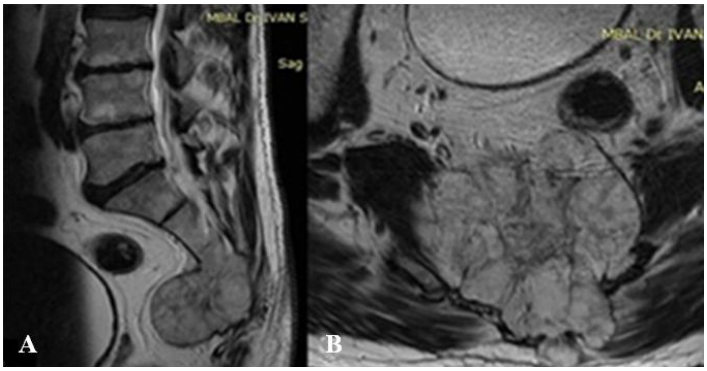

Fig. 5.10. MRI images of chondrosarcoma at S_3-S_5 level:
A/ sagittal view; B/ axial view

- **Plasmocytoma:** These tumors affect frequently the thoracic and sacral region of the spine in the 4^{th} and 5^{th} decade (Fig. 5.11 and Fig. 5.12). In the absence of neurological symptoms radiotherapy alone is sufficient. In the presence of neurological symptoms, surgical removal and decompression is needed, followed by radiotherapy.

3. Secondary (metastatic) tumors of the spine

They represent 70% of all spinal tumors. The spine is the third most common site for metastatic cancer after the lung and the liver (Fig. 5.13, Fig. 5.14, Fig. 5.15, Fig. 5.16, Fig. 5.17). Metastasis to the spine can be the intial manifestestation of unknown systemic cancer. The most common location of spinal metastases are the lumbar and thoracic vertebrae. Methastases can disseminate by means of the following mechanisms:

- arterial – through the feeding vessels of the vertebrae (most

common);

- venous – through venous reflux in the vertebral and epidural veins from tumors of the abdomen;
- per continuitatem – direct invasion by tumors located in close proximity to the spine.

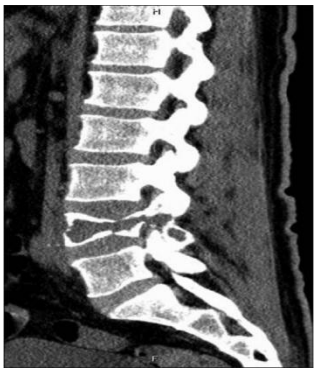

Fig. 5.11. CT reconstruction image of plasmocytoma affecting L_4 vertebra

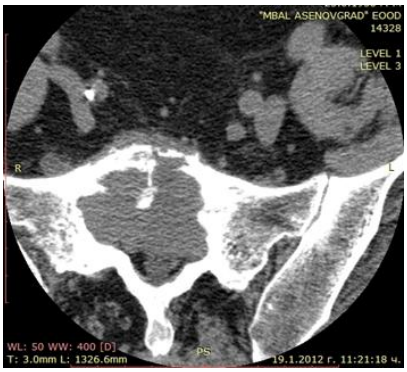

Fig. 5.12. CT image of plasmocytoma affecting S_1 vertebra

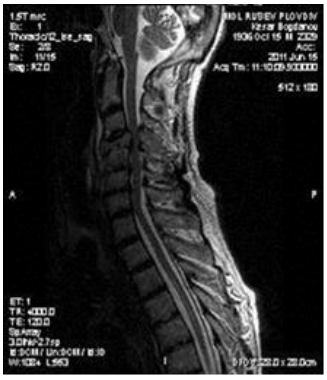

Fig. 5.13. MRI image of metastasis of C_4-C_5 segment from lung cancer

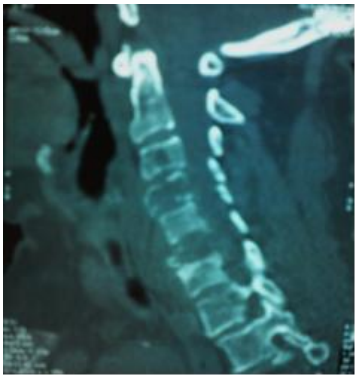

Fig. 5.14. CT-reconstuction of metastasis from lymphoma at C_4-C_6 level

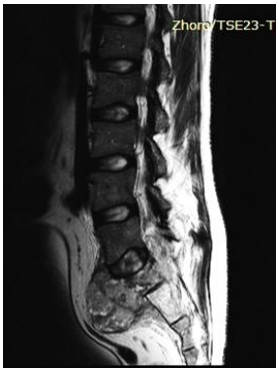

Fig. 5.15. MRI image of sacral metastasis from embryonal cell testicular carcinoma

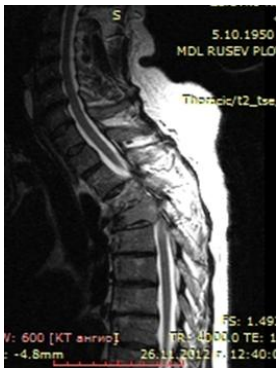

Fig. 5.16. MRI image of metastasis involving $Th_{5\text{-}6}$ segment

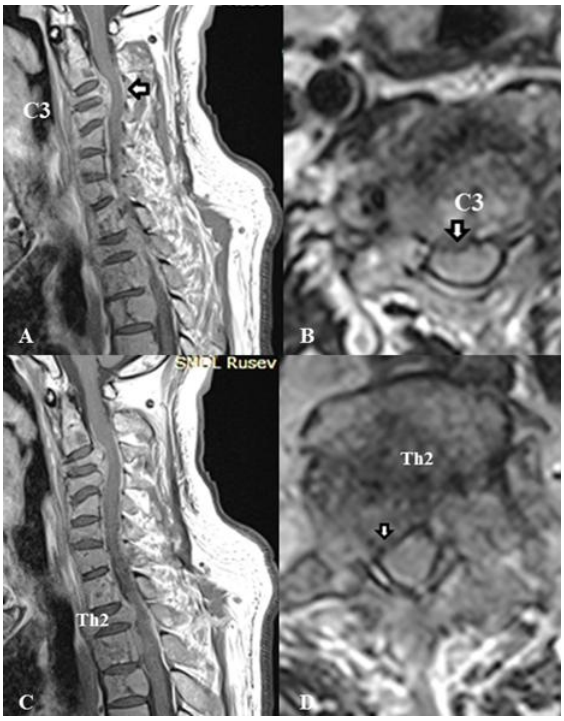

Fig. 5.17. MRI images of multiple spinal metastases:
A и **C**/ sagittal view; **B**/ axial view at C_3 level; **D**/ axial view at Th_2 level

The clinical presentation of the spinal metastases includes rachialgia (pain in the back) and compressive syndrome. Frequently, spinal metastases lead to pathological vertebral fractures, causing acute and severe compression of the spinal cord. In these cases, it is crucial to diagnose whether the fracture is stable or not. Spinal instability requires surgical stabilization of the involved segment.

Diagnosis of spinal tumors

- **spondylography (AP and lateral) -** provides information about the vertebral bodies and discs: bone destruction, bone exostoses, osteosclerosis, enlargement of the interpeduncular distance (Elsberg sign), enlargement of foramen intervertebrale (Fig. 5.18);
- **CSF examination** is important for the diagnosis of spinal compression. It establishes changes in CSF colour (xanthocromia), increased protein levels and cell count. **Queckenstedt** and **Stookey tests** reveal altered CSF circulation;
- **myelography** – in the presence of a compression, it demonstrates "filling defect" or "stop" of the contrast medium. The latter differs between different tumors (Fig. 5.19, Fig.

5.20, Fig. 5.21);

- c**omputed tomography (CT) –** suitable for visualization of of bone and soft tissue structures (normal and pathological). Contrast CT-assissted myelography can also be performed after intrathecal application of contrast medium (Fig. 5.22);

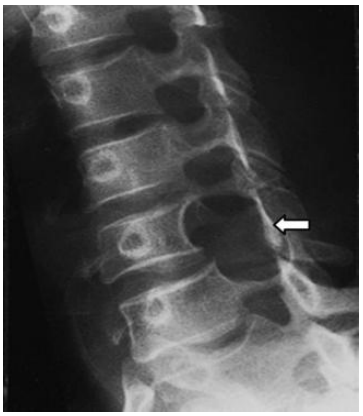

Fig. 5.18. Lateral spondylography: enlargement of the intervertebral foramen due to spinal neuroma (schwannoma)

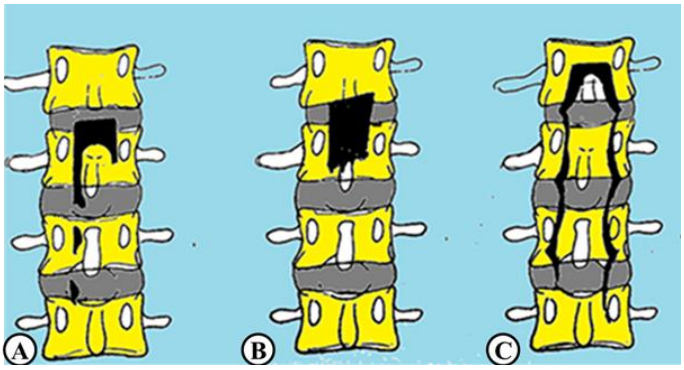

Fig. 5.19. Typical myelographic findings in spinal tumors: **A/** intradural; **B/** extradural; **C/** intramedullary

Fig. 5.20. Myelography-filling defect in intradural meningioma

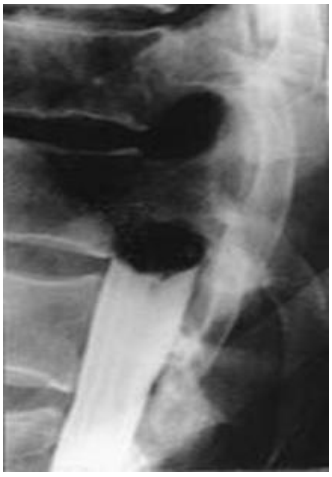

Fig. 5.21. Myelography - complete "stop" in intradural neuroma

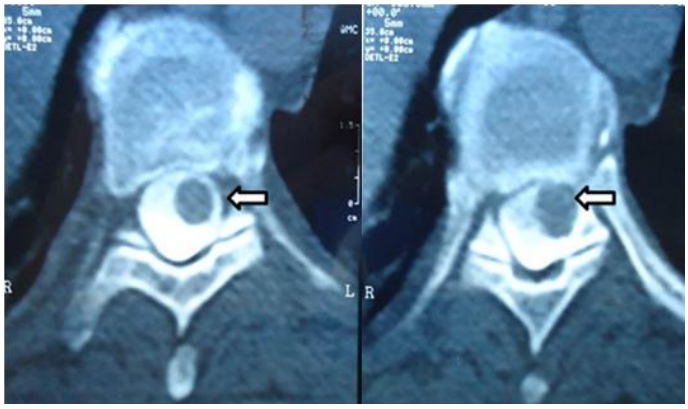

Fig. 5.22. CT-assissted myelography image of meningioma

- **magnetic resonance imaging (MRI)** – method of choice for spine and spinal cord tumors. It provides superior details about specific tumor anatomy and its relationship to surrounding neural structures (Fig. 5.23, Fig. 5.24, Fig. 5.25, Fig. 5.26, Fig. 5.27, Fig. 5.28, Fig. 5.29, Fig. 5.30 and Fig. 5.31).

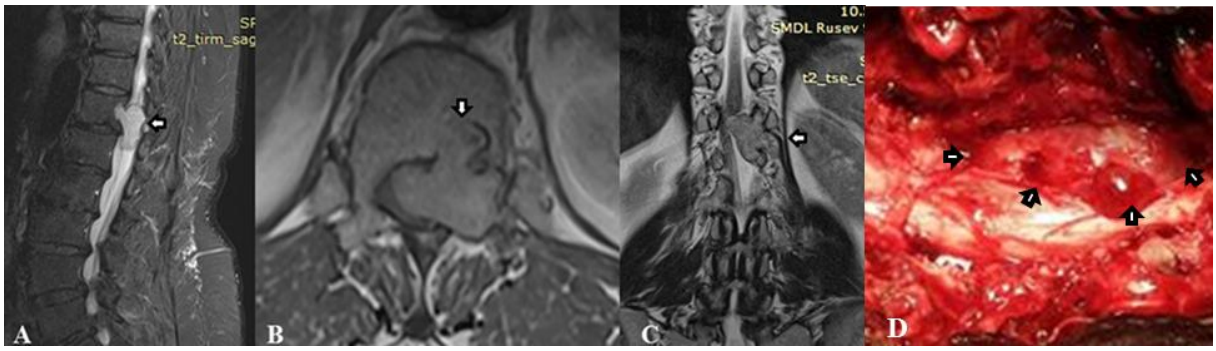

Fig. 5.23. MRI image of extradural schwannoma: A/ sagittal view; B/ axial view; C/ coronal view and D/ intraoperative view

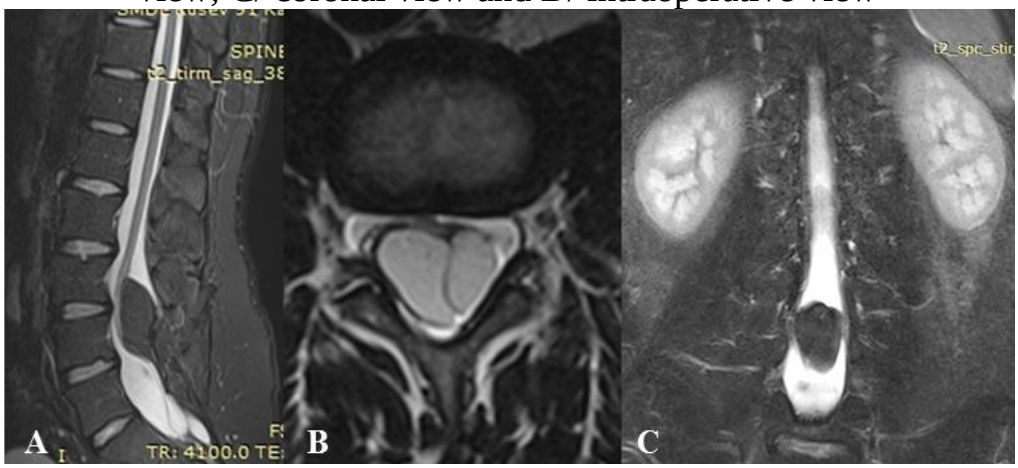

Fig. 5.24. MRI image of intradural lipoma at L_3 - L_4 level:
A/ sagital view; **B/** axial view; **C/** coronal view

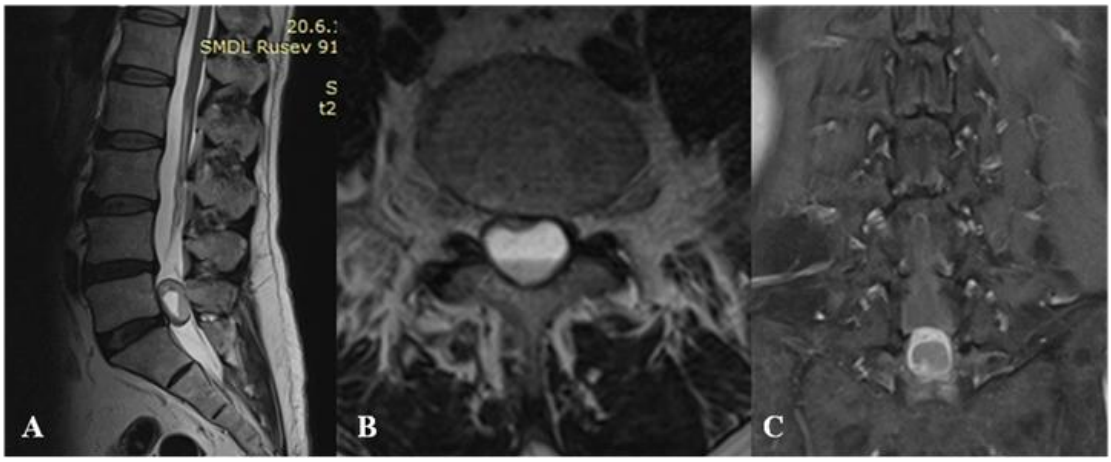

Fig. 5.25. MRI image of filum terminale myxopapillary ependymoma: A/ sagittal view; B/ axial view; C/ coronal view

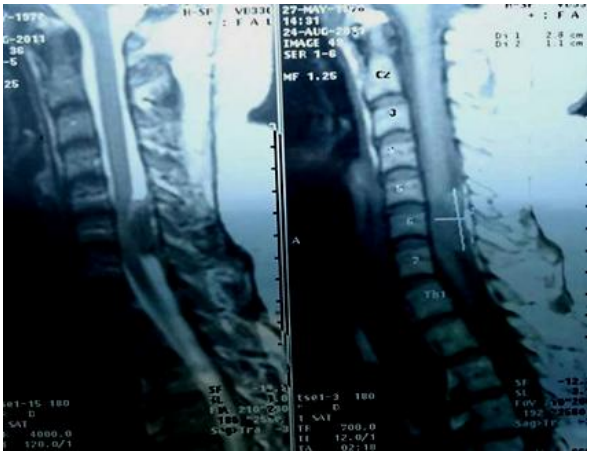

Fig.5.26. MRI image of intradural tumor (meningioma)

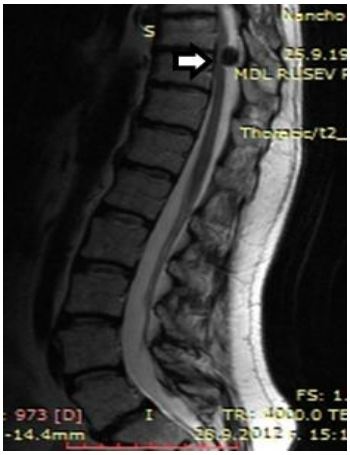

Fig.5.27. MRI image of dorsolateral meningioma

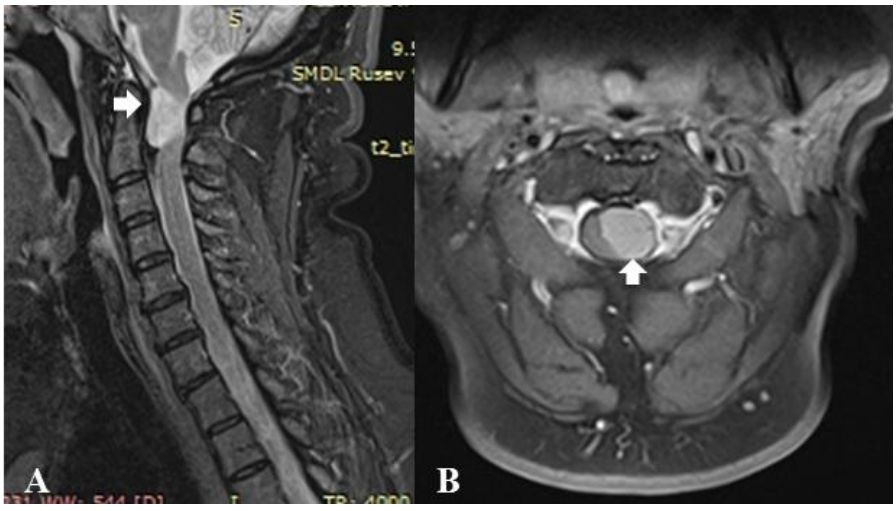

Fig. 5.28. MRI image of ventrolateral meningioma at the craniospinal junction

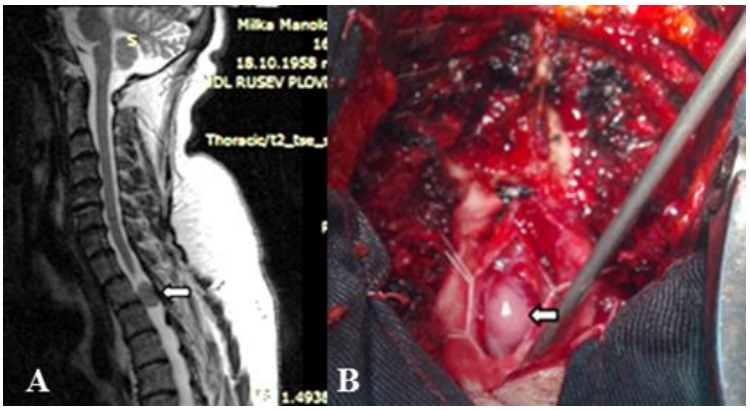

Fig. 5.29. A/ MRI image of dorsolateral meningioma at level Th_{2-3} ;
B/ intraoperative view

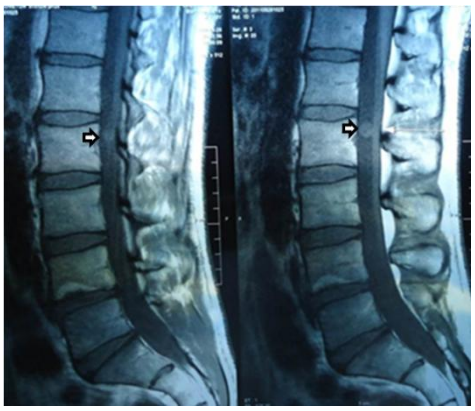

Fig. 5.30. MRI image of intradural tumor (neuroma)

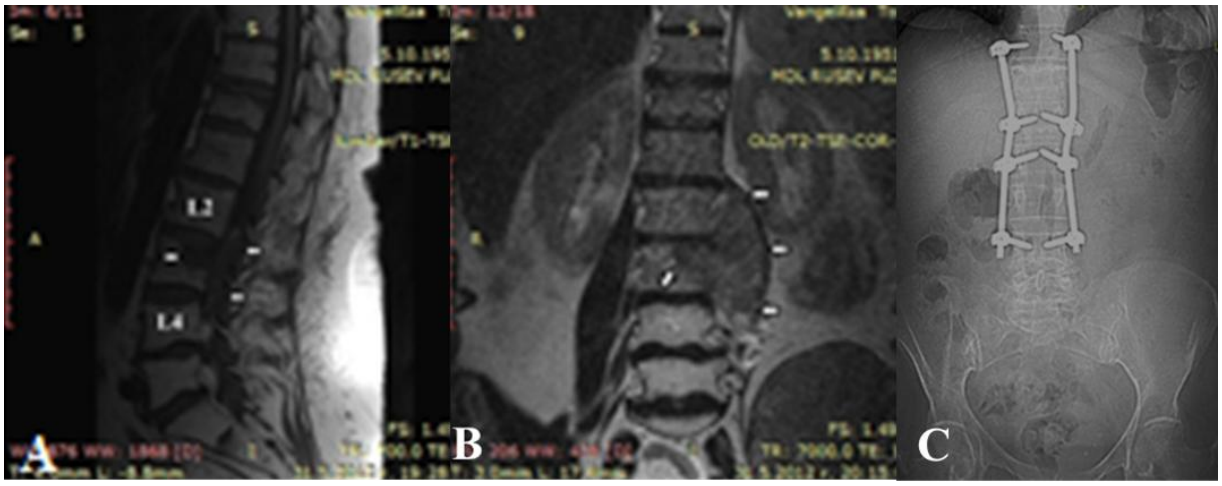

Fig. 5.31. MRI of the paravertebral myeloma affecting L_2-L_3 level which compresses the nerve roots of cauda equina: **A/** sagittal projection; **B/** transverse projection; **C/** postoperative spondylography - posterior pedicle screw fixation and stabilization of the involved segment

Treatment of spinal tumors: The goal of surgery is maximal but safe tumor resection and neural decompression. If spinal stability is compromised, it is necessary to stabilize the affected segment. Malignant tumors, which are usually infiltrative lesions, are not always amenable to gross total resection. In these cases, subsequent radiotherapy and/or chemotherapy should be considered.

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CHAPTER 6 *B. Kitov, H. Zheliazkov, I. Kehayov*

VASCULAR DISEASES OF THE CNS

From neurosurgical point of view, the most common cerebrovascular diseases that require surgery are spontaneous intracerebral haematomas, intracranial aneurysms and arteriovenous malformations. In most cases, these diseases manifest clinically as brain hemorrhage.

Spontaneous intracerebral haematomas (SIH)

SIH are non-traumatic intraparenchymal hemorrhages. The hemorrhages from aneurysms, arteriovenous malformations and hemorrhage within brain tumors should be excluded from this group. SIH are more common than the hemorrhages in the other organs. This is due to:

- the morphology of the cerebral arteries (thinner vascular wall, tunica media is represented mainly by internal elastic membrane and the smooth muscle fibres are greatly reduced);
- the specific features of the cerebral blood flow (the brain makes up about 2% of the body weight, but its oxygen consumption is about 20% of the total needs).

Characteristics of SIH

They comprise about 10% of the cerebrovascular accidents (strokes), but are related to higher rate of mortality and morbidity (50% of lethal strokes are hemorrhagic).

1. Gender and age: Approximately 2/3 of the cases with SIH appear in men and their incidence progressively rises over the age of 50 - 55 (85% of the cases).

2. Etiology: The exact cause for SIH can be determined by cerebral arteriography. The most common cause, however, is arterial hypertonia (65%). Rarely, do other etiologic factors occur (15%) - hematologic diseases, anticoagulant therapy, acute liver failure, etc. The cause for SIH can not be established in about 20%. In these cases, a doctor's attention should be focused on searching for vascular malformation. The vital prognosis of spontaneous intracerebral hematomas depends on:

- the patient's condition (age, blood pressure, presence of renal and heart diseases);
- location, size and intensity of the subsequent brain edema.

The influence of SIH on brain structures can be explained with the occurence of two types of phenomena:

- **local** – direct impact of the hemorrhage on the adjacent brain tissue (destruction, compression and modification of the local blood flow);
- **general cerebral** – high intracranial pressure due to the presence of the hematoma and the severity of the cerebral edema.

Local phenomenon

The destruction of the nervous tissue due to hemorrhage has different sequelae depending on the location and the volume of the hemorrhage (Fig. 6.1). In cases of affected deep brain structures (hypothalamus, subthalamic areas and brainstem), prognosis is usually poor. A severe neurological deficit develops when the pyramidal tracts in capsula interna are affected. The brain compression caused by the SIH together with the perifocal edema can affect essential vital and/or eloquent brain areas (Fig. 6.2).

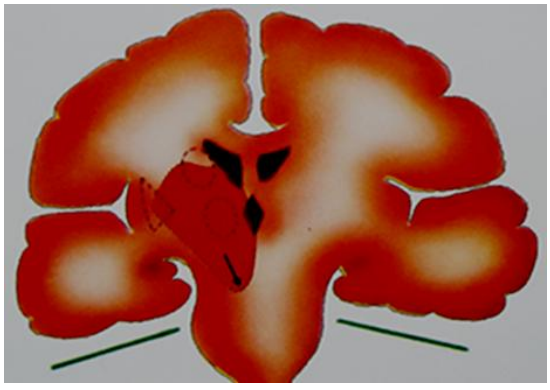

Fig. 6.1. Destruction: SIH in the basal ganglia with extension into brainstem

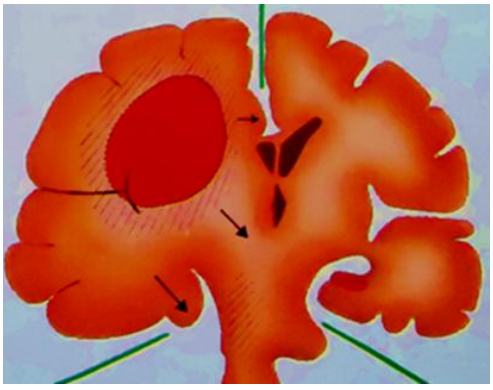

Fig. 6.2. Compression: SIH and perifocal edema cause midline shift and brain herniation

General cerebral phenomenon

The syndrome of increased intracranial pressure (ICP) as far as it is present in each clinical case depends on:

- the size of the hematoma;

- the compliance of the brain parenchyma;
- the severity of the cerebral edema;
- the presence of acute hydrocephalus as a consequence of blocked CSF circulation due to SIH.

In its final stage, the intracranial hypertension causes brain herniation that leads to severe damage to vital areas located in the brainstem which drastically increases the mortality rate.

Clinical presentation of SIH

- the disease has acute onset, in otherwise healthy individual, and presents with intractable headache, which radiates to the entire head, but it can also be lateralized on the side of the hemorrhage. Vomiting is also a common symptom;
- transient neurological deficits are missing (they are specific for ischemic events);
- sudden change in the level of consciousness;
- rapid progression of focal neurological deficit;
- rapid development of intracranial hypertension;
- nuchal rigidity (when blood enters the subarachnoid space).

Clinical symptoms specific for SIH depending on the location

1. **Hematomas affecting putamen and capsulla interna.** They are the most common type of SIH (45%). The onset of the disease can be gradual or acute, and it usually includes:
   - headache and mental confusion;
   - contralateral hemiparesis and hemisensory deficit;
   - speech disorder (when the dominant hemisphere is affected);
   - hemianopsia with conjugate deviation of the eyes towards the side of the lesion.

Small putaminal hematomas with gradual evolution have relatively good prognosis (Fig. 6.3 and Fig. 6.4). Usually, they spontaneously resorb with time and surgery is not indicated. Prognosis is poor in cases with massive hemorrhages that have acute onset and rapid progression.

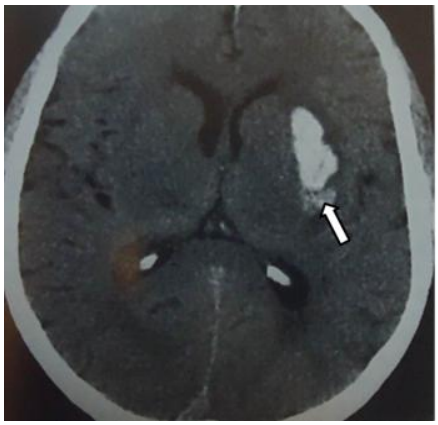

Fig. 6.3. CT image of putaminal hematoma

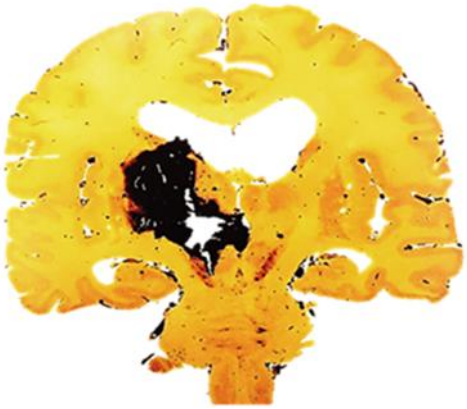

Fig. 6.4. Pathological specimen of capsulo-lenticular hematoma

2. Spontaneous intracerebral hematomas in the centrum ovale and the cerebral hemispheres (25 – 30%). The course of these hemorrhages often includes the 2 classical stages of clinical presentation of hemorrhagic stroke:

- **1st stage** - headache with vomiting and somnolence;
- **2nd stage** - delayed progressive development of neurological deficit

A. **Frontal hematoma** is characterized with:
   - changes in behaviour;
   - contralateral hemiparesis;
   - motor aphasia due to lesion in the dominant hemisphere.

B. **Temporal hematoma** (Fig. 6.5 and Fig. 6.6) usually presents with:
   - contralateral motor deficit (mainly for the upper extremity and face);
   - amnestic disorders;
   - aphasia when the dominant hemisphere is affected.

C. **Occipital hematomas** (Fig. 6.8) include:
   - eye disorders (reduced visual cortex, as well as visual agnosia).

D. **Hematomas in the temporo-parieto-occipital region** (Fig. 6.7):
   - lateral homonymous hemianopsia;
   - disorders of the praxis and gnosis, Wernicke's aphasia depending on the side of the lesion.

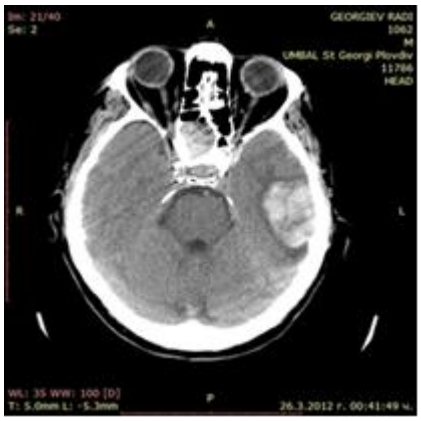

Fig. 6.5. CT image of temporal hematoma

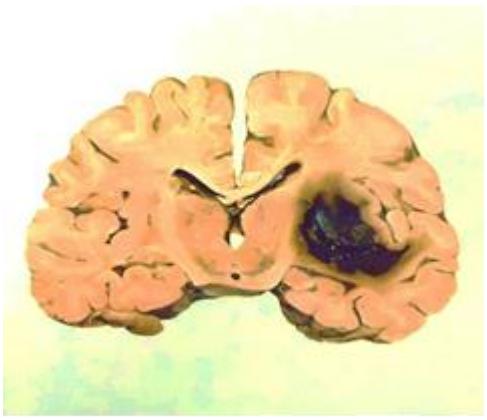

Fig. 6.6. Pathological specimen of temporal intracerebral hematoma

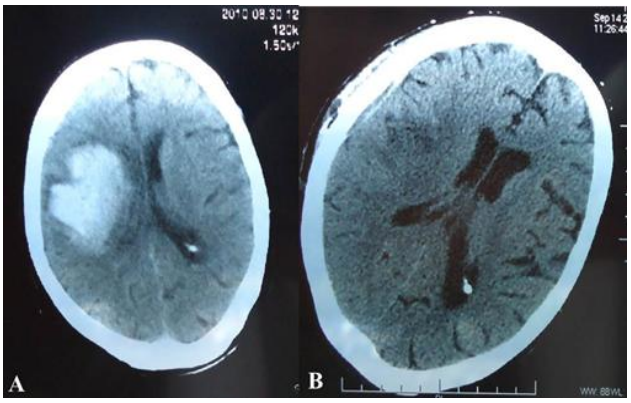

Fig. 6.7. SIH in the parietal region: **A/** before and **B/** after surgery

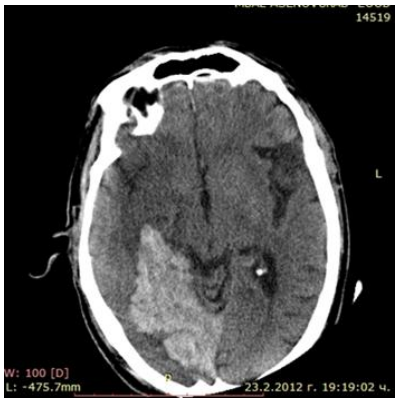

Fig. 6.8. CT image of occipital hematoma

E. Thalamic hematomas. They occur in about 10% of the cases. The most common clinical signs of thalamic hematomas are:

- headache and vomiting;
- marked contralateral hemihypesthesia with mild hemiparesis;
- eye movement disorders (vertical gaze palsy, convergence insufficiency, downward eye deviation, homolateral miosis, nonreactive pupils);
- homonymous hemianopsia;
- agnosia.

The prognosis of the restricted thalamic SIH is relatively good, that of the thalamocapsular SIH depends on its size. Prognosis of subthalamo-mesencephalic SIH is poor. Surgery is rarely indicated for thalamic hematomas.

F. Brainstem (pontine) hematomas (Fig. 6.9). They are 5% of all SIH. They can be three types:

- massive type – affect relatively large part of the pons;
- baso-tegmental type (unilateral or bilateral);
- tegmental type;
- the most common sypmtoms are:
  - headache and vomiting;
  - depressed consciousness;
  - hemi- or quadriparesis;
  - eye movement disorders (unilateral or bilateral constricted or dilated pupils irresponsive to light stimulation, conjugate deviation of the eyes, etc.);
  - cardiac and respiratory rhythm disorders.

The prognosis of brainstem hematomas is poor. They are rarely indicated for surgery.

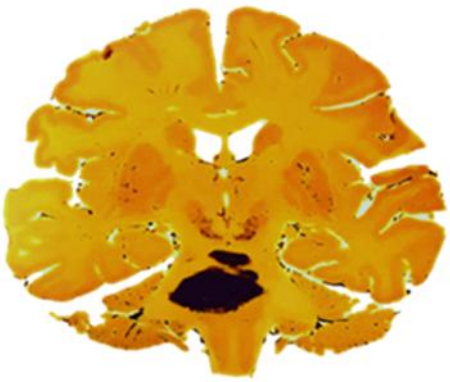

Fig. 6.9. Pathological specimen of pontine hematoma

G. Subtentorial hematomas (15%)

- in these cases, the clinical presentation includes:
  - nausea, vomiting and headache;
  - dizziness and truncal ataxia;
  - intractable hiccup, dysarthria and dysphagia;
  - depressed level of consciousness: mild form (stage 1) and severe form (stages 3 - 4).
- treatment of SIH with subtentorial location
  - conservative approach is recommended for:
    - ✓ patients who have preservation of alertness;
    - ✓ hematomas < 2,7 cm in diameter;

- ✓normal aspect of cisterna ambiens.
- surgery is performed in patients with:
  - ✓depressed level of consciousness;
  - ✓hematomas > 2,7 cm in size;
  - ✓progressive neurological deficit;
  - ✓compressed cisterna ambiens.

Diagnosis of spontaneous intracerebral hematomas

- **computed tomography (CT)** is a method of choice in the diagnosis of SIH. The examination should be performed before and after contrast enhancement, immediately after the occurrence of first symptoms. If CT angiography is available, it should also be applied (Fig.6.10 and Fig.6.11). CT scan confirms the hemorrhagic nature of the stroke, its localization (supratentorial in 85%), volume and stage. In addition, CT scan may demonstrate penetration of the SIH into the ventricles, presence of acute hydrocephalus, concomitant cerebral edema and incipient brain herniation;
- **magnetic resonance tomography (MRI) –** gives more accurate information about the localization of the hemorrhage and its relation to the adjacent brain structures and the extent of cerebral edema;
- **arteriography.** It should be performed if the cause of hemorrhage cannot be determined and vascular anomaly is suspected.

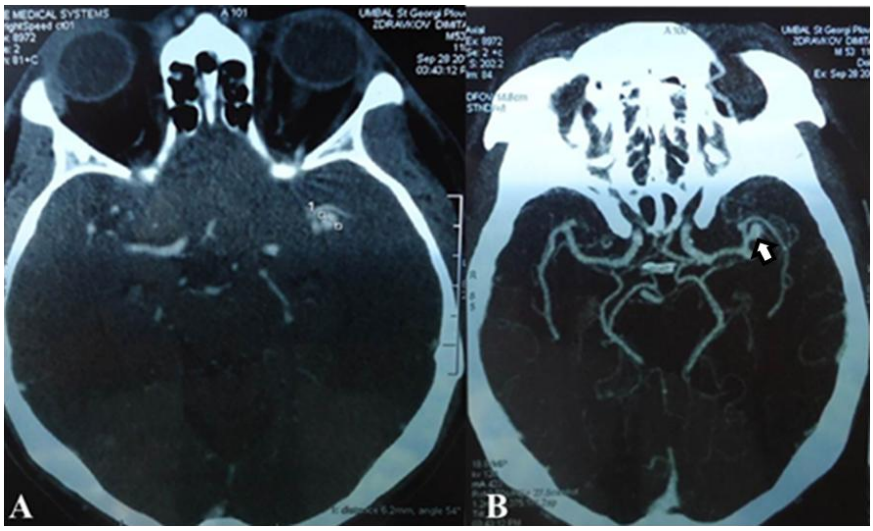

Fig. 6.10. A/ CT image of small spontaneous temporal hematoma;
B/ CT angiography shows an aneurysm of the middle cerebral artery

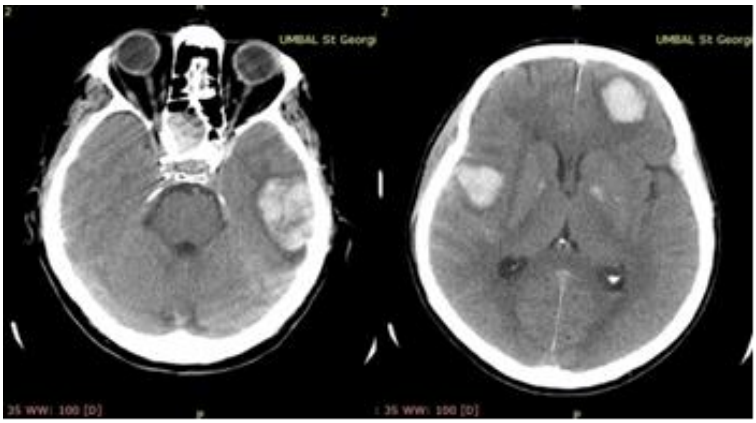

Fig. 6.11. CT image of multiple bilateral SIH due to anticoagulant therapy

Treatment of spontaneous intracerebral hematomas

Treatment of SIH should be performed at specialized clinical units that can provide reanimation support. Conservative therapy should be focused on:

- managing arterial blood pressure deviations;
- reducing intracranial hypertention (osmotic diuretics, steroids, hyperventilation, etc.).

Indications for surgery of SIH

The indications for surgical treatment depend on the localization and size of the hematoma, the age, general and neurological status of the patient. Satisfactory results have been observed after surgical evacuation of suporatentorial hemispheric capsullo-lateral hematomas and subtentorial cerebellar hematomas.

Contraindication for surgical treatment may include advanced age, severe coagulopathy, poor somatic status, deep coma (GCS = 3 to 5 points) with signs of brainstem dysfunction.

Intracranial aneurysms

Intracranial aneurysms (IAs) are saccular or rarely fusiform outpouchings of the vessels of the circle of Willis or their branches, which are located in the subarachnoid space (cisternal segments).

Epidemiology of IAs

- **Incidence**. Autopsy data show that their incidence varies betwen 0.6 - 1% as it increases with age. The incidence rate is highest between the age of 50 and 60.
- **Localization**. Aneurysms are more likely to occur in the anterior part of the circle of Willis (85%) (Fig. 6.12).

Regarding their lateralization, there is no significant difference.

- **Gender**. Female-to-male ratio is 1,6:1 and after the fifth decade increasing they affect predominantly males. In males, IAs are located predominantly in the territory of the anterior cerebral artery and anterior communicating artery, while in females – in the territory of the internal carotid artery and middle cerebral artery.
- **Size and multiplicity**. Generally, intracranial aneurysms are divided into three groups:
  - small IAs - up to 1cm.
  - large IAs– from 1 to 2.5 cm.
  - giant IAs– over 2.5 cm.

  Small IAs can become symptomatic in 70-80% of cases, large IAs-in 15-20% and giant IAs-in 1-2%. More than one aneurysm may be present in 20-30% of the cases (Fig.6.13).
- **Etiology**. The development of intracranial aneurysms is due to congenital defects in the wall of the arterial vessels of the brain (defect in tunica media) in combination with damage to the elastic membrane by hemodynamic factors (anomalies and variations of the vessels of the Willis' circle - aplasia, hypoplasia, asymmetry, etc.) (Fig. 6.14).

  There is evidence that genetic predisposition can play a role in the development of intracranial aneurysms:
  - they occur more often in patients with connective tissue disorders, Marfan syndrome, polycystic kidney disease, Ehlers-Danlos syndrome, etc.;
  - family history - 10-20% of the patients with ruptured aneurysms have first or second-degree relatives with confirmed presense of aneurysms. Smoking and arterial hypertonia are another predisposing factors.
- **Neuropathology.** Saccular IAs appear as a round outpouching which consists of fundus, body and neck. In contrast, fusiform IAs represent a spindle-shaped widening of a large segment of the arterial vessel due to atherosclerosis and arterial hypertonia (Fig. 6.15). The wall of the cerebral aneurysm is composed of adventitia, intima and remnants of tunica media lying inbetween (Fig. 6.14);

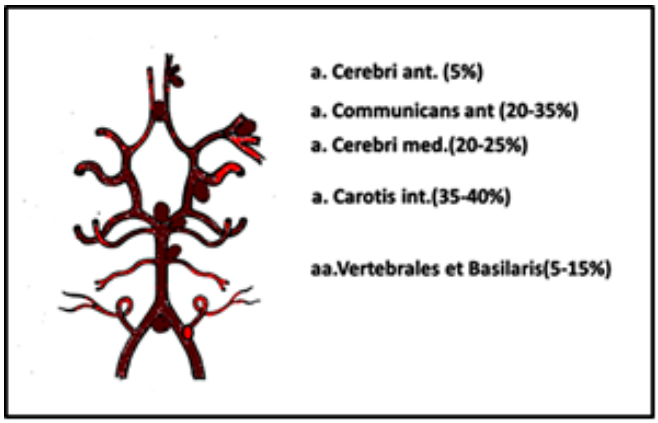

Fig. 6.12. Localization of IAs

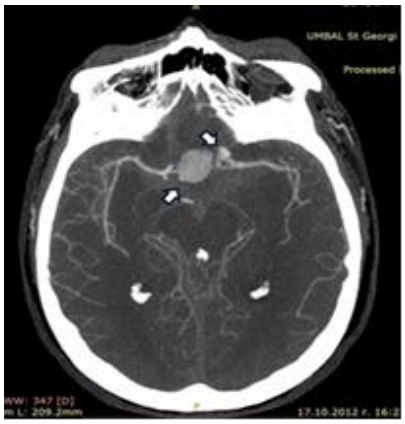

Fig. 6.13. CT image of aneurysms of the anterior communicating artery and anterior cerebral artery

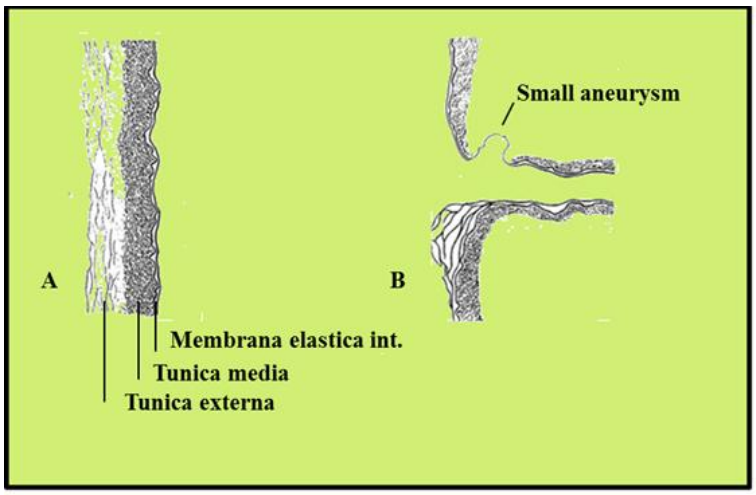

Fig. 6.14. Structure of a blood vessel of the brain: **A/** a normal wall; **B/** a wall of small aneurysm (by K.Johansen, 1982)

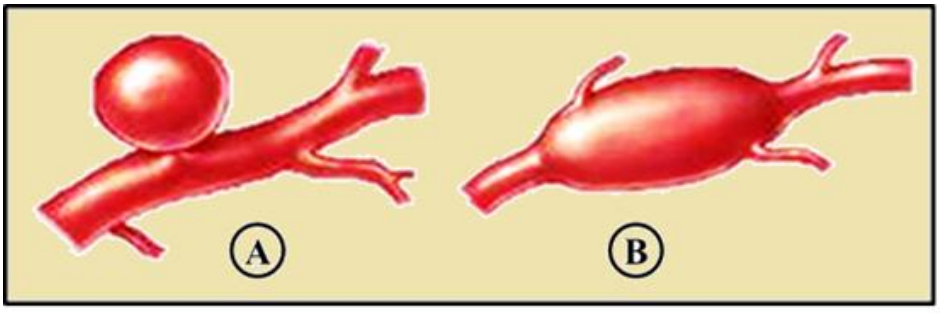

Fig. 6.15. Types of aneurysms: **A/** saccular; **B/** fusiform

Physiology of ruptured Ias. According to the type of hemorrhage:

- subarachnoid hemorrhage – the most common type;
- intracerebral hematoma – pia mater tear contributes to blood penetration into the brain parenchyma (20%);
- intraventricular hemorrhage – the blood enters the ventricles (20%);
- subdural hematoma - the fundus of the aneurysm is adherent to the arachnoid matter (1-2%).

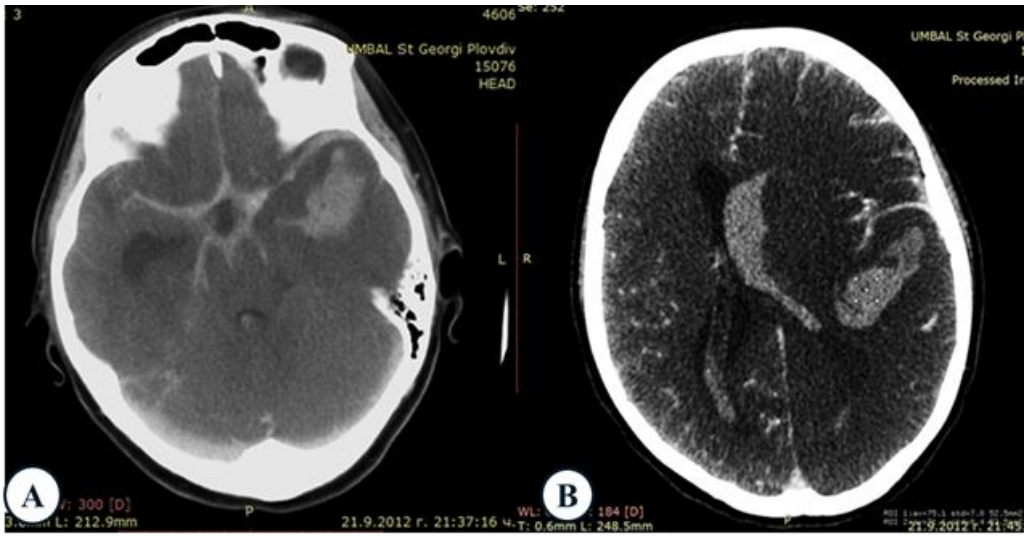

Fig. 6.14. CT image of hemorrhage due to a ruptured aneurysm:,
A/ subarachnoid and intracerebral hemorrhage;
B/ intracerebral and intraventrical localization

Clinical presentation

The clinical manifestation of ruptured IAs depends on the type of the subsequent hemorrhage:

Subarachnoid hemorrhage (SAH)

The incidence of aneurysmal ruptures varies between 0.1-0.2% annually. The onset of SAH is acute and is associated with significant mortality rate (10-40%) and severe disability in patients (30-34%). Various cognitive disorders can be observed in some patients with normal neurological status.

Clinical presentation of SAH

- **Headache**. It is almost a constant symptom with high intesity. Patients describe it as “the worst in their life” as if “they have suffered from a hit with a dagger in the head”. Initially, the headache is localized but it becomes diffuse and may radiate

down the neck and spine. Nausea and vomiting are present in 25-30% of the cases;

- **Symptoms of meningeal irritation**. Include neck pain, nuchal rigidity, Kerning and Brudzinski signs, photophobia, etc. Polyradicular pain, most common in the cervical region, can be observed in some patients. It is due to nerve root irritation caused by the hemorrhage;
- **Focal neurological deficit**. In cases of isolated SAH, focal neurological deficit is rarely observed in the beginning. It may appear later, between the 3rd and 21st day from the onset, as a result of cerebral ischemia due to arterial vasospasm and impaired cerebral blood supply. If intracerebral hematoma is present, the focal neurological symptoms occur at the very onset of the disease;
- **Alterations of consciousness** may vary from full alertness to deep coma depending on the severity and localization of the hemorrhage. Transient disorders of consciousness are observed in 40-45% of the patients. Qualitative alterations of consciousness can also be present (hallucinations, delirium-like states, etc.);
- **Epileptic seizures** are observed in 5-25% of cases. Frequently, these are generalized "grand mal" seizures. Their occurrence at a later stage of the disease may be due to a recurrent bleeding;
- **Ophtalmic symptoms.** Preretinal and subhyaloid hemorrhages can be found at ophthalmoscopy. They occur in about 20% of the cases with severe SAH and result from burst of small venous vessels as a consequence of the increased intracranial pressure. Terson's syndrome – hemorrhage inside the vitreous body, is rarely observed. The presence of eye movement disorders (oculomotor nerve palsy) refers to a ruptured aneurysm of the internal carotid artery.

Clinical presentation of giant aneurysms

In contrast to small aneurysms that cause symptoms only after rupture, the giant IAs rarely result in hemorrhage. The clinical symptoms of giant IAs result from direct compression of the surrounding brain structures, especially the cranial nerves passing near the lesion. In rare cases, CSF pathways may also be blocked which leads to hydrocephalus (Fig. 6.15).

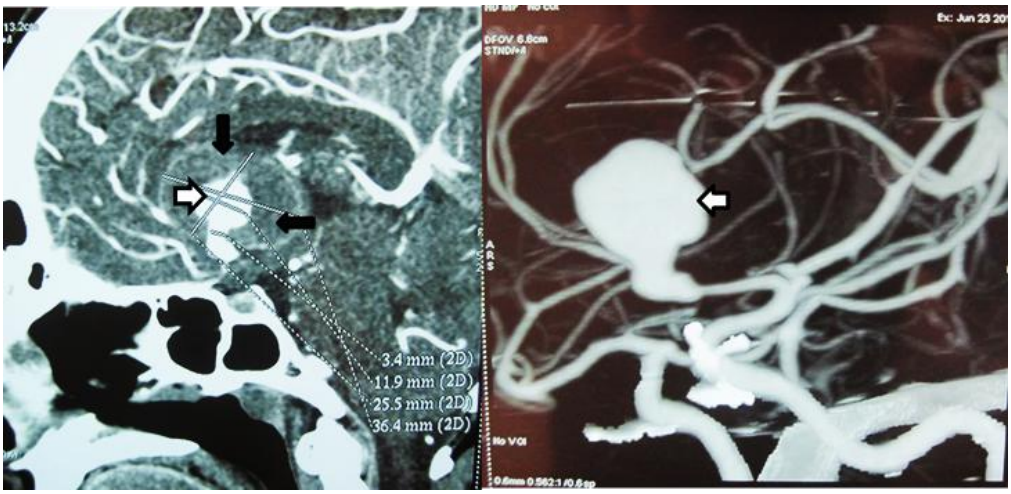

Fig. 6.15. CT angiography – a giant aneurysm of the anterior communicating artery that causes hydrocephalus: (⇨) a non-thrombosed part; (➡) a thrombosed part

The severity of the SAH can be assessed by the Hunt&Hess scale and the WFNS scale (World Federation of Neurological Surgeons; Drake, 1988) (Table 6.1 and 6.2).

Table 6.1. The Hunt and Hess scale

Grade	Clinical features
1	A conscious patient experiencing mild headache with or without symptoms of meningeal irritation
2	A conscious patient experiencing severe headache and symptoms of meningeal irritation without focal neurological deficits, except for cranial nerve palsies
3	A somnolent patient with moderate neurological deficit (usually motor deficit)
4	A patient in sopor with an apparent neurological deficit and symptoms of decerebration
5	A patient in deep coma, decerebrate rigidity, moribund appearance

Table 6.2. The WFNS grading scale

Grade	Total score of GCS points
1	A maximum score of 15 points
2	A score of 13-14 points
3	A score of 13-14 points plus motor deficit
4	A score of 7–12 points (present or absent motor deficit)
5	A score of 3– 6 points (present or absent motor deficit)

Diagnosis of ruptured IAs

- **computed tomography (CT)** can demonstrate the presence of:
  - subarachnoid hemorrhage – local or diffuse (Fig. 6.14 and Fig. 6.16). The localization of the hemorrhage can define the bleeding point. For example, hemorrhage within the interhemispheric and lamina terminalis cisterns suggests bleeding from ruptured aneurysm in the territory of the anterior cerebral and anterior communicating arteries; within the Sylvian cistern – aneurysm of the middle cerebral artery; within the carotid cistern – aneurysm of the internal carotid arteries;
  - intracerebral hematoma (Fig. 6.14A and B);
  - intraventricular hemorrhage (Fig. 6.14B);
  - subdural hematoma;
  - hydrocephalus;
  - brain ischemia (presence of hypodense areas);
  - a combination of these;
  - CT angiography allows visualization of the aneurym itself and the aneurysmal anatomy (Fig. 6.10B, Fig. 6.13 and Fig. 6.15).
- **CSF analysis.** Lumbar puncture is indicated when the CT or the MRI scan does not provide any evidence of subarachnoid hemorrhage.

The lumbar puncture is contraindicated in cases of raised intracranial pressure in order to avoid brain herniation

- **cerebral digital subtraction angiogrpahy**. This procedure is still "the golden standard" in the diagnosis of IAs. It allows visualization of the ruptured aneurysm (or multiple aneurysms), the presence of other vascular malformations (AVM, variations of the vessels of the circle of Willis, occlusions and stenoses), evidence of cerebral compression, hydrocephalus, etc. (Fig. 6.17, Fig. 6.18 and Fig. 6.19). Negative angiography does not rule out the presence of IAs. The examination should be repeated in one month after the period of cerebral vasospasm.

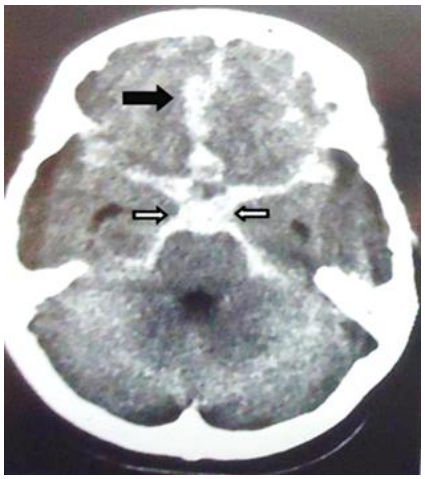

Fig. 6.16. CT image of diffuse SAH: (➡) blood in the hemispheric cistern; (➡) blood in the suprasellar cisterns

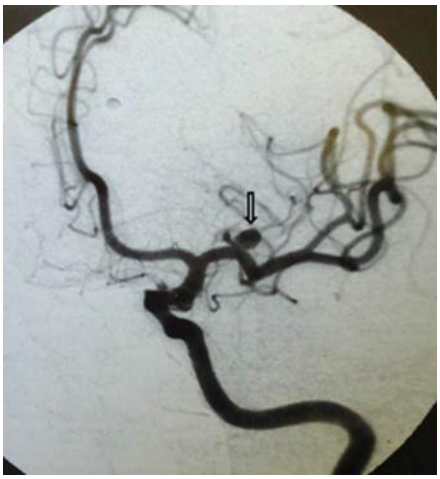

Fig. 6.17. Angiography of aneurysm of the middle cerebral artery

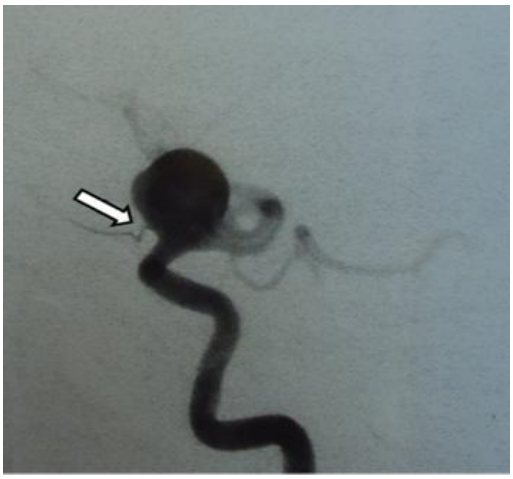

Fig. 6.18. Angiography of aneurysm at the bifurcation of the internal carotid artery and the ophtalmic artery (➡)

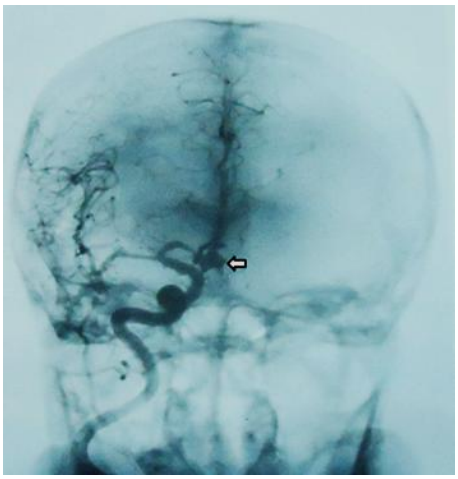

Fig. 6.19. Angiography of aneurysm of the anterior communicating artery

Treatment

- **indications for surgical treatment.** The most common and effective method of treatment is surgery. The aim of surgery is to exclude the ruptured aneurysm from the cerebral blood circulation and to remove any present intracranial hematomas (Fig. 6.20). This creates favourable conditions for aggressive treatment of the cerebral vasospasm.

If possible, multiple IAs should be excluded from the cerebral circulation through the same surgical approach.

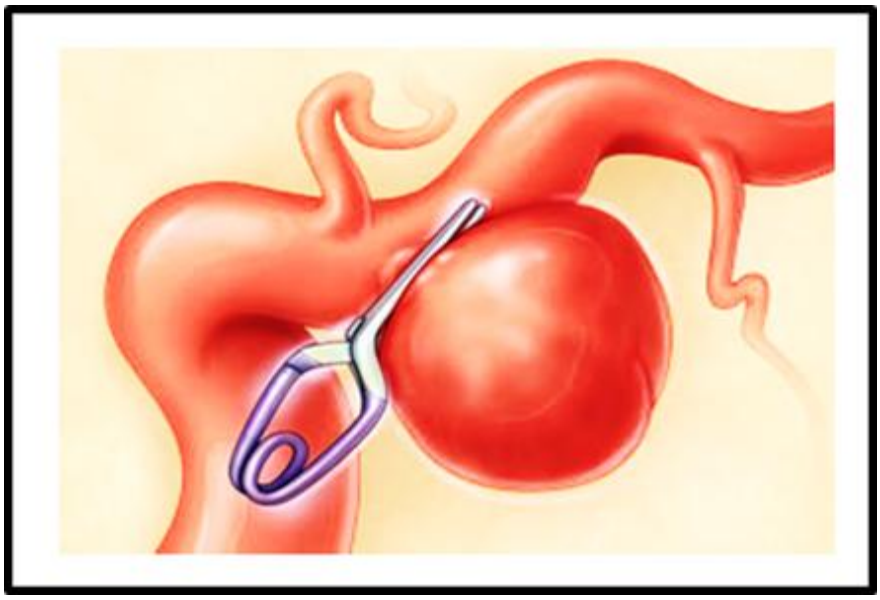

Fig. 6.20. Exclusion of an aneurysm from the cerebral circulation by clipping

Contraindications for surgical treatment. Surgery is not indicated for patients over the age of 80, with severe metabolic disorders and systemic diseases, as well as in patients with grade 4 and 5 on the Hunt&Hess scale.

Alternative treatment options for brain aneurysms

- **Endoscopic interventions**. The major disadvantage of this method is that it can not deal with intraoperative hemorrhage, if such occurs during the procedure.
- **Endovascular techniques.** They have been recently used for the treatment of IAs. The method includes use of arterial microcatheters to insert platinum spirals into the lumen of the aneurysm which stimulates thrombus formation and obliteration of the aneurysmal sac (Fig. 6.21). This approach can be also used for patients in critical condition (Grade 4 and 5).

Fig. 6.21. Stages of endovascular coiling and embolization

Preoperative care of patients with ruptured aneurysms. It should include measures to prevent the patient from recurrent bleeding: mild sedatives, calcium antagonists (Nimodipine), arterial blood pressure monitoring and control.

Postoperative

- follow-up of the patient's level of consciousness and the dynamics of the neurological deficit;
- continuous monitoring and control of the arterial blood pressure;
- CT monitoring and repeated angiography (if necessary);
- prophylaxis of infectious complications.

Cerebral arteriovenous malformations (AVMs)

Arteriovenous malformations are due to congenital defects of the embryonal vasogenesis that lead to abnormal connection between cerebral arteries and veins which bypass the capillary system.

- **Incidence and age**: AVMs affect 1% of the population. They are the second most common cause of non-hypertensive intracerebral hemorrhages, following intracranial aneurysms, and the most common cause of intracranial hemorrhage in children. They are most frequently observed in males between the age of 20 and 30;
- **Localization:** AVMs are localized mainly in the supratentorial space (94%). AVMs located within eloquent brain areas (Rolandic, speech, basal ganglia, thalamus or brainstem) are difficult to treat. In these areas, any hemorrhage resulting from this disorder may lead to severe complications, which makes them challenging for surgical removal.

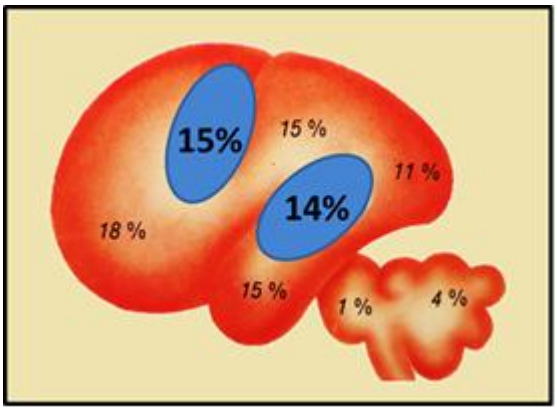

Fig. 6.22. Localization of AVM (blue) - eloquent brain areas (B. Pertuiset,1993)

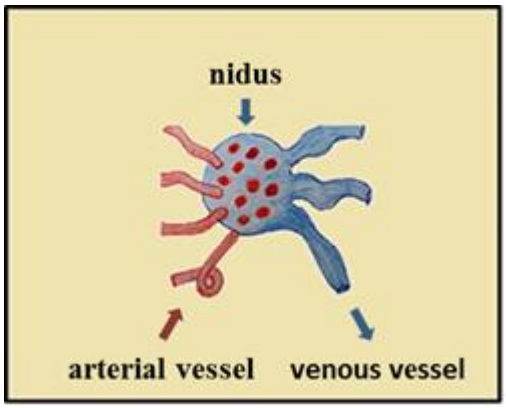

Fig. 6.23. Scheme of AVM

- **Neuropathology and neurophysiology**: AVM is an abnormal collection of blood vessels. It usually consists of central part, called „nidus”– a nest of numerous abnormal feeding arteries and draining veins, and peripheral zone of gliosis (Fig. 6.22). To a certain extent, AVMs cause change in the cerebral blood flow by „stealing" blood from the adjacent brain tissue. The resistence in the area between arteries and veins is significantly reduced due to the lack of capillary bed. It leads to an increase in the velocity and volume of blood flow in the AVM on account of the remaining brain tissue, which is in state of constant hypoxia. This may provoke epileptic seizures, intelectual and/or neurological deficit. Due to the increased pressure in the vessels of the AVM, arteries are the first to dilate, then veins and finally, the collateral vessels.

As a result from the hemodynamic stress, the walls of the dilated veins form pseudoaneurysmal sacks which are the sites of future rupture and bleeding.

Clinical presentation

Spontaneous hemorrhages. They occur in 30–75% of the cases. They are usually intraparenchymal, but they can penetrate to the subarachnoid space or the ventricles. The clinical features depend on the localization of the hematoma.

- **epileptic seizures**. Jacksonian seizures (60%) can be observed in parietal localizations while generalized seizures may be present in frontal and temporal localizations (40%);
- **neurological deficit (30– 55%)**. Neurological manifestations are usually mild and they gradually progress;
- **headache (20–45%)**. It is migrenous, located on the side of the malformation. Hypertensive headache is rarely observed as a result of the reduced reabsorption of CSF in the dural sinuses due to the increased venous pressure;
- **a throbbing noise in the head** occurs in about half of the patients. It is perceived as subjective unpleasant sensation which is synchronous with the cardiac pulsations and can be auscultated (especially in children);

- **evolution of arteriovenous malformations**. The mortality after the first hemorrhage is about 10%, but in subsequent reccurent bleedings it can reach up to 30-40%.

Generally, half of the patients with AVM experience hemorrhage within a 20-year period.

Diagnotics

- **Transcranial Doppler (TCD) sonography**. It's a screening method for detecting AVMs. It is used for examining the velocity of the blood flow through the AVM;
- **CT**. It can detect intracranial hematomas following AVM rupture (Fig. 6.24) which cause neurological symptoms (headache, epileptic seizures, motor deficit). When hemorrhage is not present, AVMs are seen as heterodense areas, accompanied by ischemic (hypodense) areas without dislocation (Fig. 6.25). When a contrast agent is administered, the folded draining veins can be visualized (Fig. 6.25 and Fig. 6.30);
- **MRI.** The role of this examination is significant because it allows more detailed visibility of the AVM in three planes, its feeding and draining vessels and their relation to the surrounding brain parenchyma (Fig. 6.26 A, B and C);

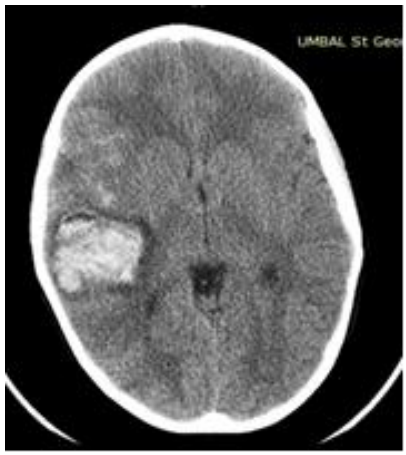

Fig 6.24. CT image of parenchymal hematoma from ruptured AVM

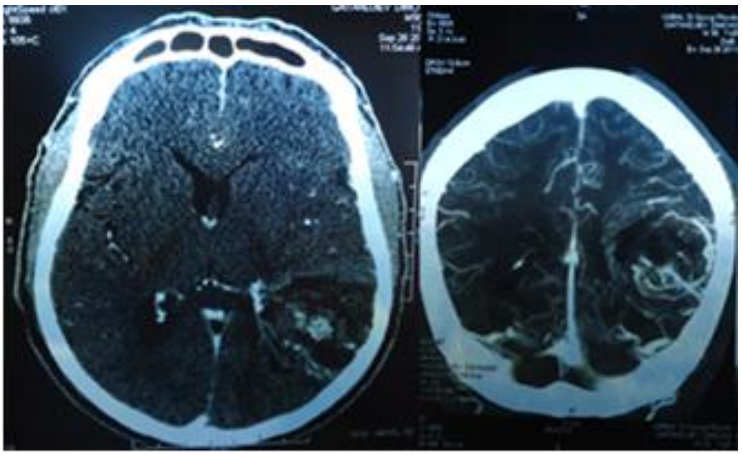

Fig. 6.25. CT image (axial and transverse) of non-ruptured AVM

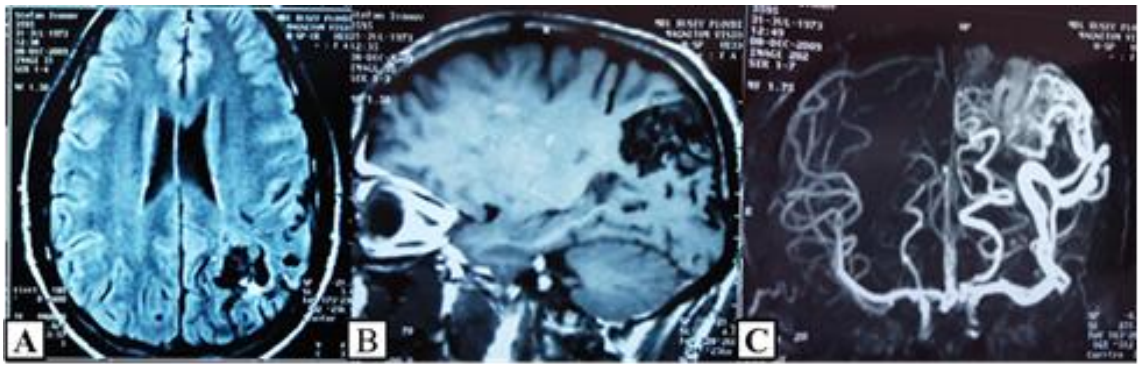

Fig. 6.26. MRI images of AVM:
A/ axial view; **B/** sagittal view; **C/** MRI angiography

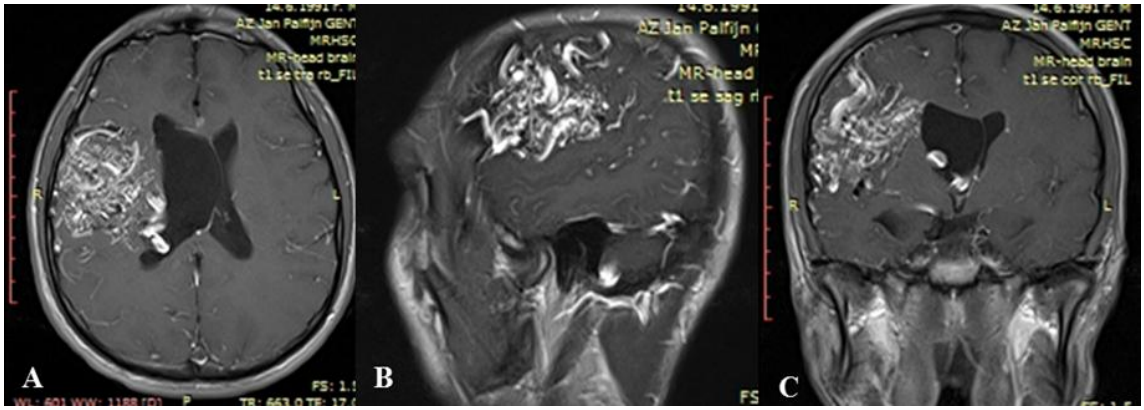

Fig. 6.27. MRI images of AVM:
A/ axial view; **B/** sagittal view; **C/** transversal view

- **Cerebral digital subtraction angiography**. This is the method of choice. It provides superior details of the AVM antomy - nidus, feeding arteries and draining veins (number, size, shape, etc.) (Fig. 6.28 and Fig. 6.29).

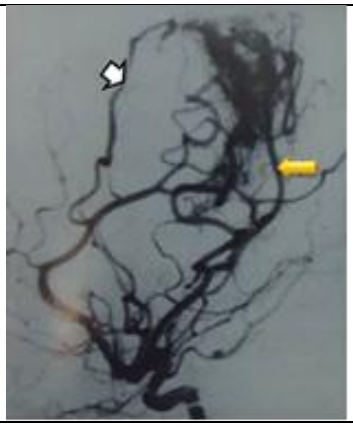

Fig. 6.28. Angiographic image of AVM: (➧) right anterior cerebral artery (⬅) right middle cerebral artery

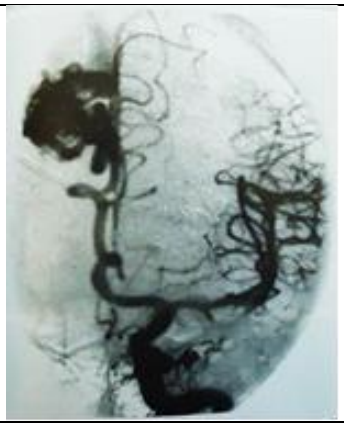

Fig. 6.29. AVM in the right hemisphere fed by the left pericallosal artery

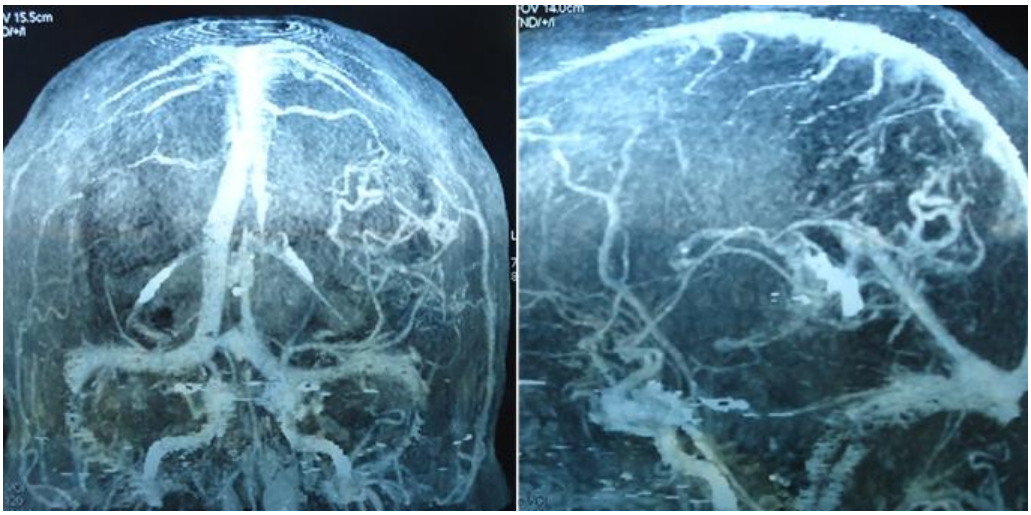

Fig. 6.30. CT angiography (venous phase) demonstrating dilated veins draining into the dural sinuses

Treatment

The surgeon should weigh "the risks versus benefits" when choosing the appropriate treatment. An exact individual evaluation of the AVM is essetial. Today, the Spetzler&Martin Scale is used for operative risk assessment of AVMs (Table 6.3).

Table 6.3. Factors for operative risk assessment of cerebral AVMs

Factor	Points
SIZE	
Small < 3 cm	1
Middle 3–6 cm	2
Large >6 cm	3
LOCALIZATION	
Non-eloquent brain area	0
Eloquent brain area	1
VENOUS DRAINAGE	
Superficial	0
Deep	1

The total sum evaluates the severity of the disease and the surgical risk. Total sum of 1or 2 points indicates low mortality and postoperative complication risk, total sum of 3 and 4 points indicates considerable operative risk. Total sum of 5 points indicates fatal outcome. If there is a large AVM in the deep brain structures (basal

ganglia, thalamus, hypothalamus or brainstem) surgery is doomed to be unsuccessful.

Treatment options for AVMs

- **Conservative therapy**. It is symptomatic and aims to relieve epileptic seizures.
- **Endovascular embolization**. It does not eliminate the AVM and only interrupts its blood flow.
- **Surgical treatment**. The ideal treatment includes complete surgical removal of malformation without damaging surroinding brain structures. It includes interruption of the feeding arteries and draining veins, followed by excision of the nidus.
- **Radiosurgery** refers to single high dose localised irradiation, using either Gamma-knife, or a linear accelerator, or Cyber-Knife. It aims to destroy blood vessels of the AVM nidus, while avoiding injury to normal brain and this is achieved by focusing radiation onto the lesion. The effectiveness of radiosurgery is generally measured in terms of disappearance of abnormal blood vessels on angiography. After radiosurgery, the rate of angiographic obliteration increases with time and is size and radiation dose dependent. The reported obliteration rates at 2 years are in the range of 80% to 90% for small lesions (<2 cm diameter) and 40% to 60% for larger lesions. AVMs indicated for radiosurgery should be 3 or less cm in diameter.

Radiosurgery may provide AVM obliteration within 2 years which does not exclude the risk of hemorrhage during this period

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CHAPTER 7 *I. Kehayov, A. Davarski, P. Miryanov*

INFLAMMATORY DISEASES OF THE CNS

The intracranial or intraspinal inflammatory processes result from hematogenous spread of purulent focuses inside the human body or purulent sources located in the structures surrounding the cranial cavity or the spinal canal. The conditions that contribute to these types of diseases are fractures of the cranial base, congenital CNS defects (dermal sinuses), dysraphic conditions (encephalocele and meningocele). Cerebrospinal posttraumatic or postoperative fistulas also contribute to the penetration of infection inside the CNS. Once, it has penetrated the intracranial or intraspinal space, the infection spreads via the CSF pathways and may cause inflammations of the cerebral meninges – meningitis, or penetrate the brain parenchyma – encephalitis or cerebral abscess.

Purulent meningitis

Meningitis is an inflammatory condition that affects the leptomeninges (arachnoid and pia matter). During this process the brain tissue is also involved, therefore, the condition should be considered as meningoencephalitis. However the cerebral meninges are more affected than the parenchyma.

- **Clinical features:**
  - **infectious syndrome** – acute onset, high fever, muscle pain, headache, nausea, fatigue;
  - **symptoms of meningeal irritation** – rigidity (increased muscle tonus) of the cervical and the dorsal paravertebral musculature. The most severe degree of hypertonus is called the so called “opisthotonus” which manifests as a severe hyperextension and spasticity in which an individual's head, neck, and spinal column enter into a complete "bridging" or "arching" position. Photophobia is also observed. The Kernig and Brudzinski tests are positive;
  - **focal neurological symptoms** – epileptic seizures which result from excitation of the cerebral cortex. Cranial nerve palsies can also be observed;
  - **symptoms of increased intracranial pressure.**

- **Diagnosis**:
  - **lumbar puncture**: The CSF is opalescent or turbid. It may also have purulent appearance and flows out under high pressure. The examination shows an increased number of cells – polynucleosis. The glucose level is decreased while the protein levels are slightly increased. Immediately after lumbar puncture, the CSF sample should be send for microbiological testing and culture;
  - **Treatment**. Purulent meningitis is treated conservatively with antibiotics, as the appropriate agent should be selected according to the results from the microbiological culture. In order to provide maximum CSF concentration, the antibiotics can be injected intrathecally.

Cerebral abscess

Cerebral abscess is focal inflammatory cerebral lesion. Nowadays, this type of disease tends to be limited (3-5/1 000 000) due to the wide-spread use of broad-spectrum antibiotics. Nevertheless, this condition still cause severe neurological disorders and is associated with high mortality rates.

Pathology: The cerebral abscess consists of ternary capsule and purulent center. The internal layer of the capsule is composed of vascularized tissue; the middle layer contains collagen fibers, and the external layer is composed of gliotic tissue with mononuclear infiltration.

Four stages, with distinct pathological and radiological features, are recognized:

- early cerebritis;
- late cerebritis;
- early capsule;
- late capsule.

Etiology and pathogenic mechanism of the cerebral abscess

I. Otogenic and sinogenic abscesses

1. Otogenic abscess – 60% of all cases, as 90-95% of them are located in the temporal lobe and the cerebellum. They are formed by the following mechanisms:

- direct penetration to the temporal lobe after osteitis of the roof of the cavum tympani and the area of the mastoid antrum;
- infectious thrombophlebitis of the veins which drain blood from the brain to the middle ear, including the inferior petrous

sinus and the transverse petrous sinus – retrograde spread of the infection;
- spread of the infection from the internal acoustic meatus to the cerebellum.

2. Sinogenic abscess – 10-15% of the cases. This type most commonly develops after frontal sinusitis. The route of infection penetration is similar to that of otogenic abscess:
- after previous development of osteomyelitis of the posterior wall of the frontal sinus;
- thrombophlebitis of the diploic veins which drain the brain tissue and the paranasal sinuses to the sagittal sinus.

II. Hematogenous abscesses

They are caused as a result of bacteremia and arterial embolism by inflammatory processes in the organism (lungs - 20%, soft tissue infections, kidneys, endocarditis, etc.). The tetralogy of Fallot is associated with cerebral abscess in 5% of the cases. The hematogenous cerebral abscesses are usually developed in the brain territory supplied by the medial cerebral artery. These abscesses are often multiple.

- **clinical features**: the clinical features of the cerebral abscess are not specific and mimic a space-occupying process. The full extent of the clinical features is most commonly manifested with:
  - **infectious syndrome** – fever (50%), meningeal irritation symptoms (40%);
  - **focal neurological deficit** – presents in 60-70% of the cases. Epileptic seizures are observed in 1/3 of the cases;
  - **symptoms of increased intracranial pressure** – they are observed in almost all the cases – headache (90%), nausea (50-60%).

Usually, the clinical manifestations are prolonged and the clinical presentation of the condition is observed within 1-2 months from its onset. Cases with hyperacute evolution were also described.

- **Diagnosis**:
  - **laboratory examination.** It shows a presence of infectious syndrome – peripheral leukocytosis and an increased erythrocyte sedimentation rate (ESR);

The lumbar puncture is contraindicated due to the increased intracranial pressure because it may cause brain herniation

- **computed tomography (with contrast enhancement).** The lesion is visualized as a central hypodense zone surrounded by a relatively smooth hyperdense ring and perifocal hypodense (edemic) zone with variable size. It demonstrates the localization of the abscess and the presence of sacculations, multiple abscesses, as well as about dislocation phenomena (Fig. 7.1, Fig. 7.2, Fig. 7.3 and Fig. 7.4).

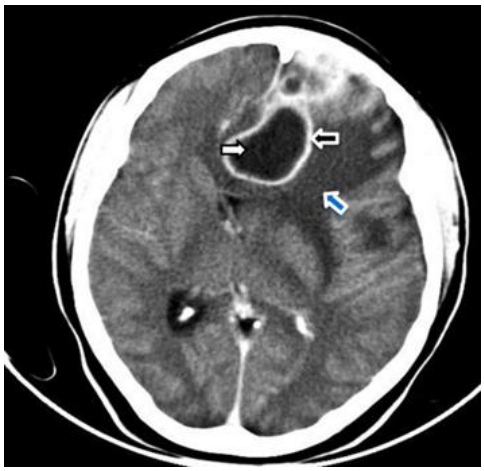

Fig. 7.1. CT image of cerebral abscess: (⇨)purulent center, (⇨) capsule (⇨) perifocal edema

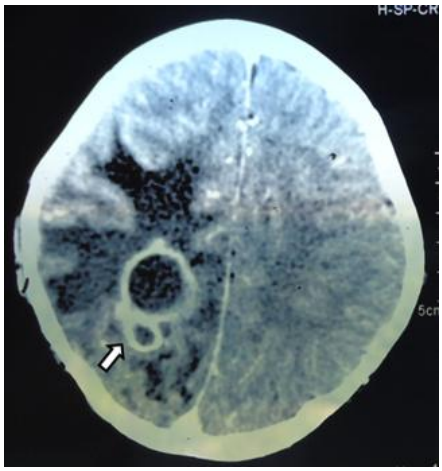

Fig. 7.1. CT image of cerebral abscess with sacculations(⇨)

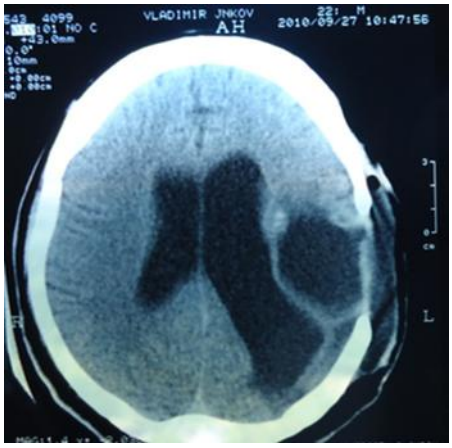

Fig. 7.3. CT image of postoperative brain abscess and assymetric hydrocephalus

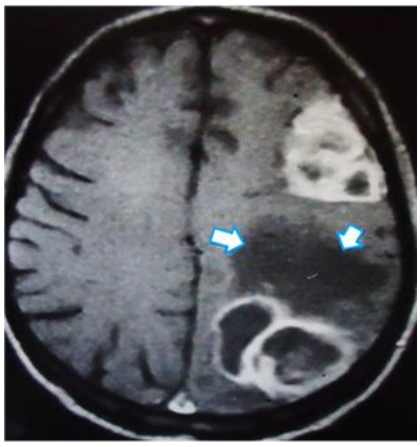

Fig. 7.4. CT image of multiple brain abscesses

- **Treatment:**
  - **conservative treatment** is applied in the following cases:
    - ✓small size: 1-2 centimeters in diameter;
    - ✓lack of dislocation phenomena;
    - ✓multiple abscesses without dislocation;
    - ✓known infectious focus and source;
    - ✓available frequent CT control.

The choice of the appropriate antibiotic is of paramount importance. It should be selected according to the sensitivity obtained from the antibiogram of the isolated agent from the primary focus. If the bacterial cultures are negative, a combination of the following agents should be applied: new generations of cephalosporins (Cefotaxime, Ceftriaxone); aminoglycosides (Amikacin), Metronidazol (in case of a possible anaerobic infection) in maximum dosages and adequate duration of the treatment, on average 5-6 weeks.

  - **surgical treatment:**
    - ✓**excision** – This intervention considerably shortens the duration of treatment. Surgical candidates are the cases with:
      - complete capsule formation;
      - multiloculate abscesses;
      - abscesses which show fluid-air level seen on the imaging studies which suggests the presence of gas collections;
      - abscesses formed as a result of penetrating injuries accompanied by presence of foreign bodies, bone fragments, etc.
    - ✓**puncture of the abscess and aspiration of the purulent content.** This intervention is recommended in cases of:
      - critical condition of the patient (sopor or coma) and a necessity of short intervention;
      - localization of the lesion in eloquent brain areas (Rolandic area);
      - localization in the area of the basal ganglia or the brainstem;
      - multiple abscesses.

Usually, after aspiration, the abscess cavity is being drained for a couple of days in order to evacuate the contents and to apply antibiotics locally. It is necessary to keep sterility in order to avoid another infection. CT follow-up is essential.

Subdural and epidural abscess

Osteomyelitis of the cranial bones can spread to the epidural or the subdural space. Similar localization of the abscess can also be caused by sinusitis or mastoiditis.

- **clinical features**:
  - headache is almost a compulsory symptom. It is commonly localized within the area affected by the process. Epileptic seizures, motor weakness symptoms and raised intracranial pressure are not rare.
- **diagnosis**
  - **skull X-rays** can demonstrate bone abnormalities that are specific for osteomyelitis or sinusitis;
  - **computer tomography** visualizes an area with different density from the suurounding brain adjacent to the cranial bones (Fig. 7.5).
- **treatment**. The subdural and the epidural abscesses have to be evacuated and followed by thorough irrigation and antiseptic processing of the cavity. If a capsule is present it should be excised. The affected bone should also be removed. It is advisory to insert drainage which can be used for postoperative lavage with antibiotics. Broad-spectrum antibiotic treatment is prescribed for 2-3 weeks after the surgical treatment.

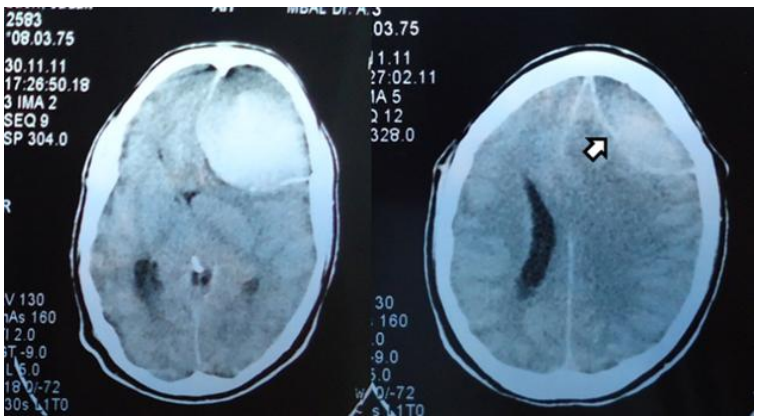

Fig. 7.5. CT image of epidural abscess: (➪) capsule

Inflammatory diseases of the spine and the spinal cord

The inflammatory diseases of the vertebrae, the intervertebral discs and the spinal epidural space lead to compression of the spinal cord and/or cauda equina. These conditions are relatively rare.

- **Etiology** – depending on the type of the bacterial agent:
  - **specific** – Mycobacterium tuberculosis;
  - **non-specific** – caused by a variety of bacteria. Staphilococcus aureus causes 55-60% of the cases of epidural abscesses. The most commonly isolated gram-negative bacteria are E. coli or Enterobacter, Proteus, Pseudomonas.
- **Pathophysiology**. Nonspecific vertebral osteomyelitis is caused by hematogenous spread of infection located in the urinary system, lungs, teeth, paravertebral tissues, bacterial endocarditis or thrombophlebitis. Thrombophlebitis of the paravertebral venous sinuses is also present. Necrosis with formed purulent collection and sequesters as well as leukocytic infiltration are observed. Initially, the purulent process affects the spongiosis of the vertebral bodies followed by inflammation of the intervertebral discs, the epidural space and the paravertebral tissues.
- **Clinical presentation**
  - begins with acute local axial pain, later accompanied by radicular or polyradicular pain;
  - the movements of the affected segment are severely restricted and painful;
  - toxo-infectious syndrome is present in 50% of the cases;
  - myelopathic and/or radicular deficits are observed in 70-80% of the cases.
- **Diagnosis:**
  - **laboratory examination.** Leukocytosis and increased erythrocyte sedimentation rate (more than 100 mm) are observed;
  - **imaging studies (spine X-rays, myelography, CT and MRI)**. These examinations can demonstrate;
  - collapse of the affected disc (Fig. 7.7);

- destruction of the impaired vertebral bodies (Fig. 7.6 and Fig. 7.7);
- affected paravertebral tissues (pathologic shadows) (Fig . 7.6);
- segmental deformity (usually kyphotic).

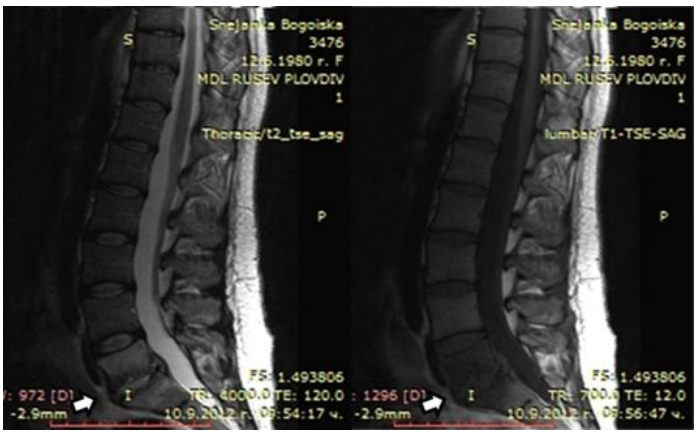

Fig. 7.6. MRI image of spinal epidural abscess at S_1–S_2 level

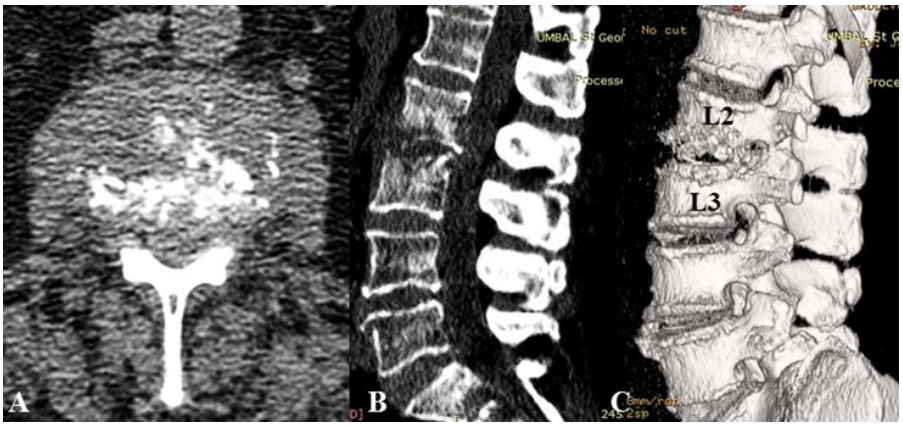

Fig. 7.7. CT images of spondylodiscitis at L_2–L_3 level:
A/ axial view of L4; **B/** sagtal CT reconstruction;
C/ 3D CT reconstruction

Treatment

A. **Conservative treatment** is applied in cases without neurological symptoms.
   - **antibiotic therapy**. An intensive parenteral antibiotic therapy is prescribed for a period of 6-8 weeks.

Isolation of the bacterial agent is essential in order to prescribe the appropriate antibiotic. This can be achieved by punch biopsy of the affected segment followed by microbiological culture.

- **immobilization** – the affected area should be immobilized for 3-4 months.

B. Surgical treatment is indicated for cases with neurological symptoms, instability of the affected segment and presence of spinal epidural abscess.

The aim of surgery is to achieve decompression of the neural structures, stabilization of the affected segment in order to avoid future spinal deformity vitiation of the functional recovery.

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CHAPTER 8 *B. Kitov, E. Hristov, S. Raykov*

PARASITIC DISEASES OF THE CNS

I. Echinococcosis

Echinococcosis is a helminthic disease with chronic evolution. The liver and the lungs are most commonly affected. CNS echinococcosis is the third most common form which affects 3-5% of the cases.

Etiology and pathogenesis

The disease is caused by the larval form of Echinococcus granulosus. The adult forms live inside the intestines of carnivores (dogs, wolves, jackals, etc.). The human, as an intermediate host, develops the disease after eating food contaminated with the eggs of the parasite. The consumed eggs release embryos which pass through the intestinal mucosa and enter the venous and the lymphatic system and from there spread to the parenchymal organs – liver and lungs. If the barrier function of the affected organ is impaired, the embryos enter the brain. The usual location in the brain is hemispheric, in the territory supplied by the middle cerebral artery, especially in the parietal lobes. The echinococcal cyst of the brain is usually single and rarely – multiple (Fig. 8.2). The size varies from several millimeters to 10-15 cm, as the bigger cysts are more often located subcortically.

The echinococcal cysts can also be found in the spine, the spinal canal and rarely – inside the spinal cord. The spinal location is characterized with deformation of the vertebral bodies and compression of the medulla without involvement of the intervertebral discs.

Clinical features are not specific.

The condition should be suspected in children and young patients who have frequent contact with animals and demonstrate slowly evolving signs of intracranial hypertension and subtle development of focal neurological deficit. Epileptic seizures are seen in 10-15% of the patients.

Diagnosis of cerebral echinococcosis

- **laboratory examination** – eosinophilia. The Casoni test is usually positive;

- **skull X-ray.** In 2-3% of the cases, a calcified parasitic cyst can be seen resembling egg shell;
- **computed tomography**. The location, size and number of the parasitic cysts can be established. They are visualized as purely rounded hypodense lesions without perifocal brain edema (Fig. 8.1 and Fig. 8.2);
- **MRI** – the most detailed examination.

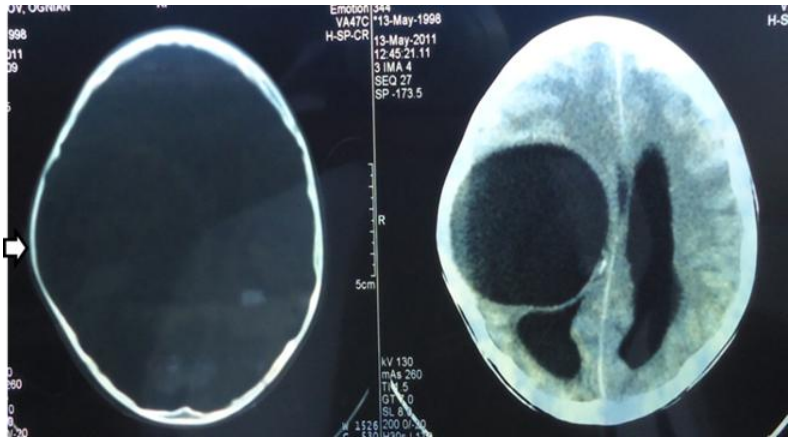

Fig. 8.1. CT image of cerebral echinococcosis in the parietal region, causing thinning and deformation of the overlying bone

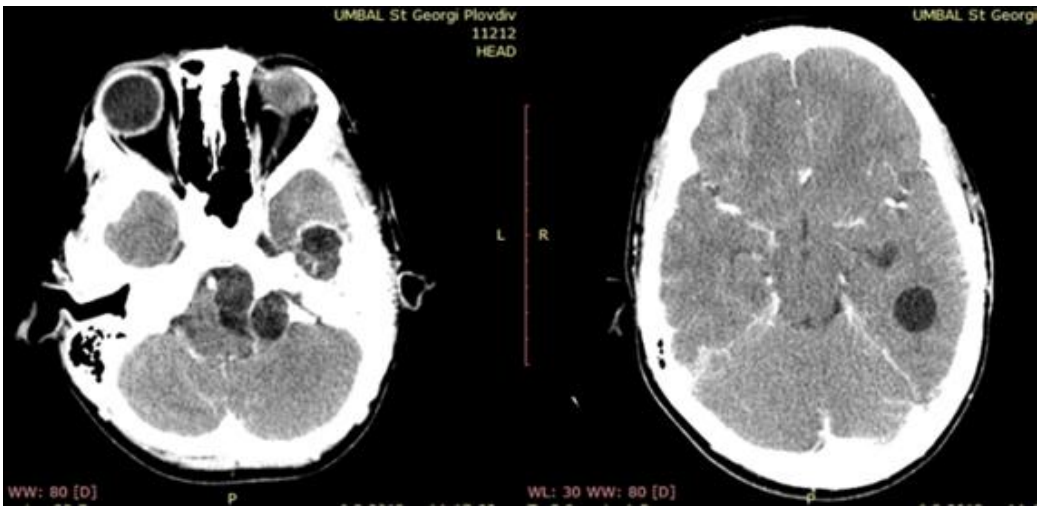

Fig. 8.2. CT image of multiple supra-infratentorial hydatid cysts

Diagnosis of spinal echinococcosis

- **laboratory examination –** similar to cerebral echinococcosis;
- **lumbar puncture –** provides information about the spinal compression and impaired circulation of the cerebrospinal fluid;

- **myelography.** In case of compression of the spinal cord or cauda equina, a filling defect or 'stop' of the contrast agent may be established (Fig. 8.3AB);
- **computed tomography –** visualizes changes in the vertebral bodies, the presence of cysts in the paravertebral tissues and the epidural space (Fig. 8.3C, Fig. 8.4 and Fig. 8.5);
- **MRI –** the most informative examination, especially for soft tissue damage (Fig. 8.6);

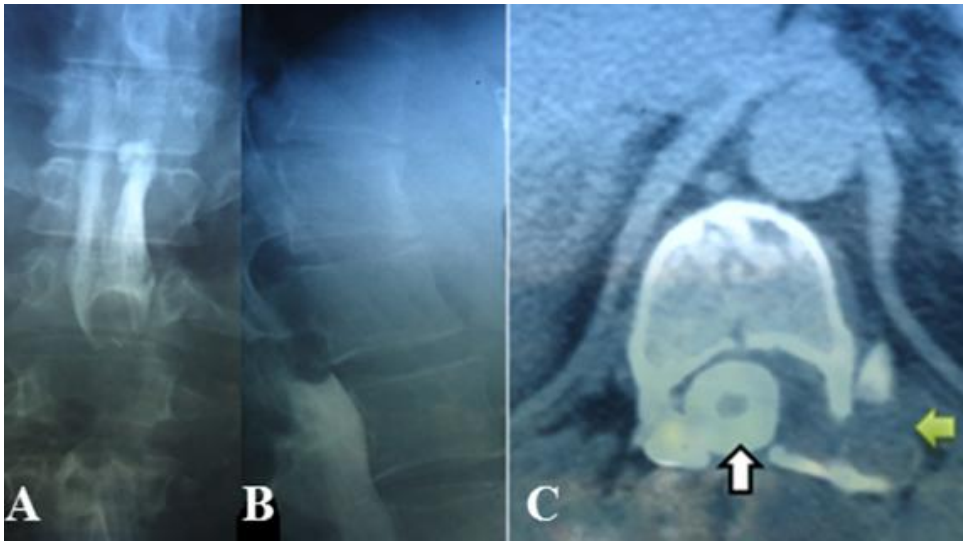

Fig. 8.3. Myelography demonstrating spinal echinococcosis in the thoracic region: A/ AP projection; B/ Lateral projection – "stop" of the contrast agent; C/ CT-assissted myelography – compressed spinal cord (⇨) and hydatid cyst (➡)

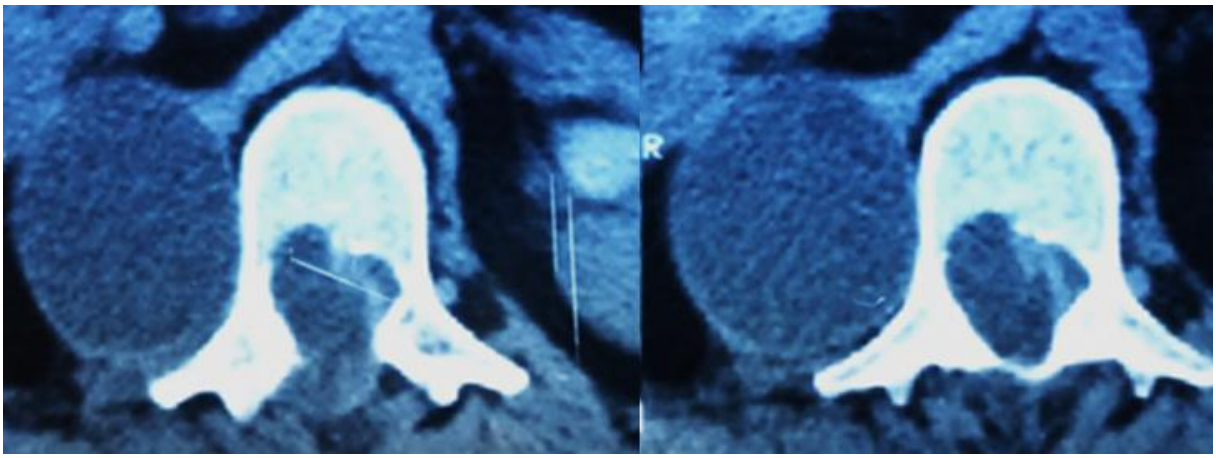

Fig. 8.4. CT image of spinal echinococcosis – paravertebral epidural extension with spinal cord compression

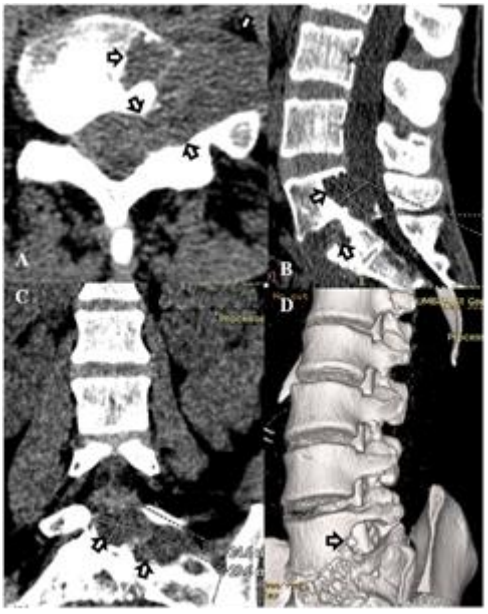

Fig. 8.5. CT image of spinal echinococcosis at L_5 - S_1 level:
A/ axial view – the cyst invades the spinal canal through the left neuroforamen and causes destruction of the vertebral body;
B/ sagittal reconstruction - shows lesion of the upper sacrum;
C/ coronal view; **D/** 3D reconstruction - widened neuroforamen

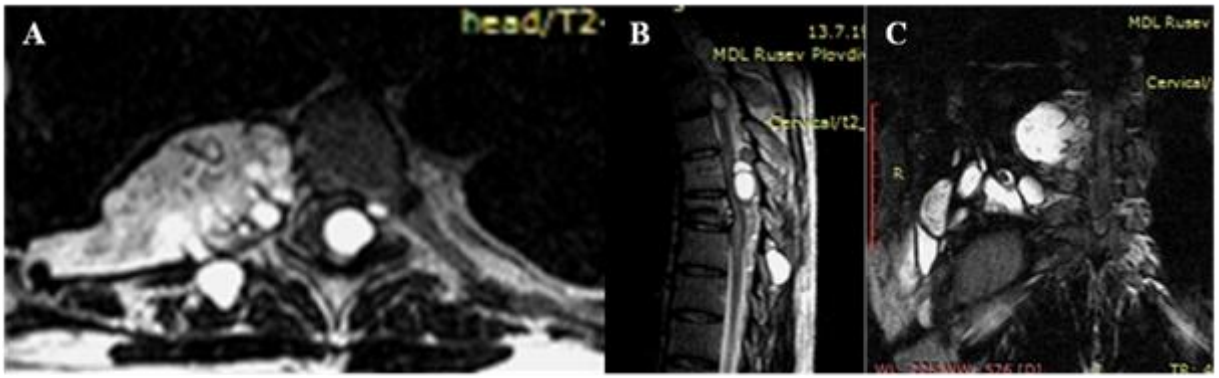

Fig. 8.6. MRI images of spinal echinococcosis in the thoracic region:
A/ axial view; **B/** sagittal view; **C/** coronal view. All images demonstrate the involvement of the spinal canal and the paraveretebral region

Treatment: The main treatment is surgical removal of the cyst. Perforation of the cyst and contamination of the brain parenchyma should be avoided in order to prevent the individual from developing anaphylactic shock and dissemination of the process.

II. Cysticercosis

Cysticercosis is a helminth disease caused by the larval form of the parasite Taenia solium (pork tapeworm). The actual causer is

cysticercus cellulosae which is a cyst with size of 5-20 mm, containing transparent liquid and a scolex. The consumed eggs release an embryo which penetrates the intestinal capillaries and through the blood stream reaches the organs. The most common locations are the muscles, the brain, the eyes and the parenchymal organs. For about 2-3 months the embryos become a cysticercus.

Depending on the location of the cysticercus in the brain, the following forms are distinguished:

- ventricular;
- cysternal;
- parenchymal.
- Mixed

Clinical features

- increased intracranial pressure (50-60%) with papilledema which can lead to atrophy of the optic nerves and blindness;
- epileptic seizures (40%);
- behavioral changes (20-30%);
- focal neurological deficit (10-20%);
- syndrome of meningeal irritation (10%);
- affected cranial nerves (5%);
- coordination disorders (10%).

Diagnosis

- **laboratory examination**
  - blood tests – peripheral leukocytosis and eosinophilia;
  - CSF analysis – moderate pleocytosis, lymphocytosis, eosinophilia, positive serological tests and an increased protein levels.
- **computed tomography** – cystic lesions are evident as hypodense zones; ependymitis, hydrocephalus and calcifications are also present:
  - single or multiple cystic formations – 30-40%;
  - calcifications and cystic formations – 20%;
  - calcifications and hydrocephalus – 15-20%;
  - hydrocephalus alone – 10-15%;
  - negative examination – 10-15%.

Treatment of cysticercosis

A. **Conservative treatment.** The drug Praziquantel (50mg/kg) is commonly used. In order to limit the inflammatory reaction and brain edema dexamethasone may also be used.
B. **Surgical treatment –** indications:
   - presence of hydrocephalus caused by obstruction of CSF pathways requires shunt surgery;
   - presence of parenchymal, ventricular or cysternal forms which cause compressions of the cerebral structures;
   - patients with intracranial hypertension.

The prognosis of this condition is dismal despite the advance in conservative and surgical methods. The mortality rate reaches 35-40%, as half of the lethal cases are in patients with basal cysticercal meningitis.

III. Toxoplasmosis

Toxoplasmosis is a parasitic disease caused by Toxoplasma gondii. The parasite can be established in many mammals but the most common host is the cat. The parasite exists in three forms: trophozoite, cyst and oocyst. The human is infected by consumption of oocysts from contaminated food with feline excrement. A congenital infection 'in utero' is also possible. The consumed cysts are decomposed in the digestive system and the released organisms spread through hematogenous pathways. Principally, the cysts are formed in the brain, myocardium and skeletal muscles. The frequency of toxoplasmosis considerably increases in patients with immune deficiency (AIDS).

Clinical features

- **Congenital form** occurs when the mother suffers from toxoplasmosis during pregnancy. This form affects only the brain and presents with epileptic seizures, hydrocephalus, chorioretinitis, etc. The contamination of the infant takes place during delivery and leads to generalized infection with hemorrhagic rash, hemolysis and hepatosplenomegaly.
- **Acquired form** begins as fever complicated by lympha-denopathy, meningoencephalitis, peripheral mononucleosis. Three neurological forms are known:

- diffuse encephalitis;
- meningoencephalitis;
- single or multiple focal lesions.

Diagnosis

- **laboratory examination** –serological tests are the basic diagnostic means. They include Sabin-Feldman test and the indirect immunofluorescent antibody test.
- **CT and MRT** – demonstrate multiple focal lesions located bilaterally in vicinity to the basal ganglia. The lesions are hypodense and enhance on contrast imaging.

Treatment of toxoplasmosis

A. Conservative treatment – combination of Pyrimethamine and Sulfadiazine is used.

B. Surgical treatment – it is indicated for cases with single and, rarely, multiple lesions which compress surrounding brain structures.

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CHAPTER 9 *H. Zheliazkov, M. Manova, B. Kalnev*

SPONDYLOGENIC MYELOPATHY AND RADICULOPATHY

Spondylogenic myelopathies and radiculopathies is a general term which includes a variety of diseases usually occurring as a result of constant repeted tension and micro traumas to the spine. It should be emphasized that some congenital anomalies and variations in the structure of the spine may potentiate the damage of the spinal cord and the roots. These diseases present considerable social issue because they are the second most common cause for temporary disability.

Disc Desease

The disc pathology can occur at all levels of the spine but it most frequently affects the lower lumbar and lower cervical region. In 90-95% of the cases, the disc pathology occurs in the lumbar area at L_4–L_5 and L_5–S_1 levels, 4-5% at L_3-L_4 and only 1-2% at L_1-L_2 and L_2-L_3. Usually, the affected segments in the cervical area are C_5-C_6 and C_6-C_7 levels. Thoracic disc herniations are rare. However, they often cause considerable damages to the spinal cord.

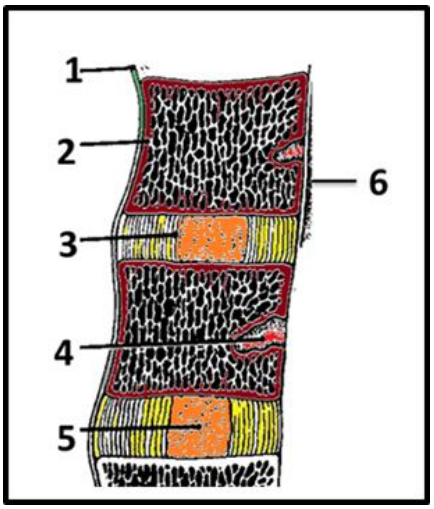

Fig. 9.1. Anatomy of the intervertebral discs: 1. anterior longitudinal ligament; 2. vertebral body; 3. anulus fibrosus; 4. intracorporal vein; 5. nucleus pulposus; 6. posterior longitudinal ligament

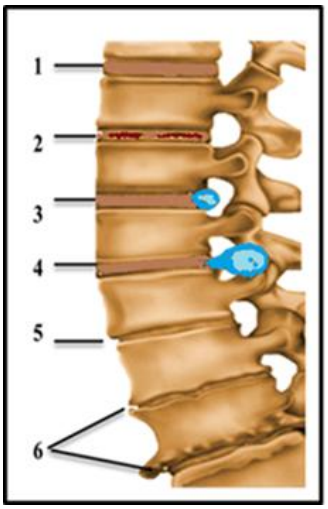

Fig. 9.2. Degenerative spondylosis: 1. normal disc; 2. degenerative disc; 3. bulging disc; 4. disc herniation; 5. narrowed disc; 6. osteophytes (bone spurs)

The diagnostics and treatment of the disc disease is an interdisciplinary problem, involving neurologists, physiotherapists, orthopedists and neurosurgeons.

The intervertebral discs are elastics buffers between the neighbouring vertebral bodies which consist of pulpous nucleus surrounded by a fibrous ring located beneath cartilage end plates, which cover the horizontal plates of the vertebrae (Fig. 9.1). The longitudinal ligaments, which run along the anterior and posterior surface of the vertebrae, are tightly fused with the fibrous ring. The latter consists of 10-12 concentric layers of collagenous fibers that support the entire structure. The fibrous ring is weaker in its lateral aspect because these parts are not covered with ligaments.

In young individuals, the pulpous nucleus is jellylike and very elastic due to the high contents of mucopolysaccharides and water (exceeding 80%). With aging, the constant tension and microtraumas cause degenerative changes in the pulpous nucleus and the fibrous ring. The pulpous nucleus loses its water saturation and fibers from the inner part of the fibrous ring break apart. Thus, the disc undergoes fibrous transformation and defects in the fibrous ring occur. Fragments of the pulpous nucleus can protrude through these newly-formed microcracks and exert pressure on the ligamentous apparatus of the spine. The latter contains oteoblasts which cause excessive bone proliferation at the edges of the vertebral bodies called osteophytes. The sum of these processes, called degenerative spondylosis, leads to hypertrophy, fibrosis and calcification of the intervertebral joints and ligaments and, in the long run, narrows the size of the neural foramina and the spinal canal that harbor the neural structures – spinal cord, cauda equina and nerve roots (Fig. 9.2).

Bulging disc, disc protrusion and disc herniation

1. **Bulging disc** – this is an initial stage of the disc disease. Degenerated nucleus pulposus presses the fibrous ring, thus, the latter bulges and compresses the nerve roots (Fig. 9.3). At this stage, the nucleus pulposus can return to its initial state which is associated with amelioration of the clinical symptoms.

2. Disc protrusion – this is the next stage of the disease which is characterized by breaking of the fibrous ring and migration of a fragment of the nucleus pulposus outside the fibrous ring. In particular cases, with the help of physiotherapeutic or balneotherapeutic procedures, the reversion of the migrated fragment is still possible (Fig. 9.4)

3. Disc herniation – In this stage, the fragment of the nucleus pulposus protrudes and migrates outside the ruptured fibrous ring In this case the reverse reposition is impossible. The tear of the posterior longitudinal ligament and the free migration of fragments of the ruptured disc to the spinal canal is called exteriorizated disc herniation. There are several types of disc herniation depending on its direction:

- median disc herniation – compresses the spinal cord in the cervical and thoracic segment or the cauda equina in the lumbar segment;
- paramedian disc herniation – compresses nerve root;
- lateral disc herniation (foraminal and extraforaminal) – compresses the nerve root.

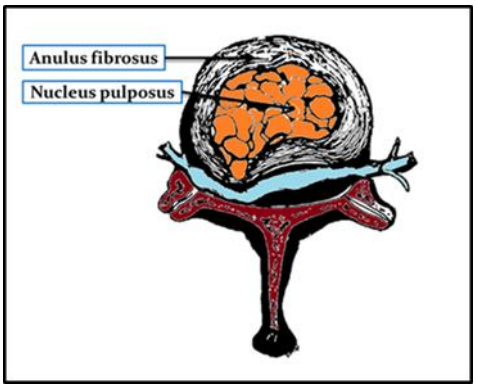

Fig. 9.3. Bulging disc

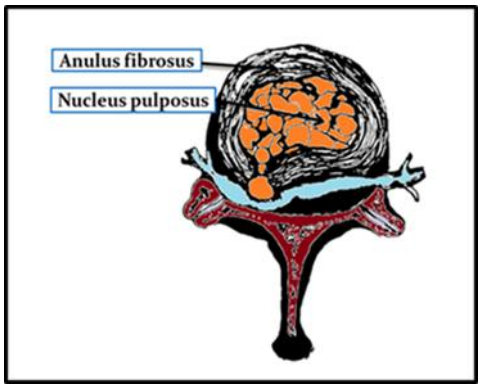

Fig. 9.4. Disc protrusion

Clinical presentation of the disc disease in the lumbar area

The symptomatology goes through the following stages:

1. Low back pain syndrome (Lumbalgia) – pain of variable duration and intensity located in the lumbar area which causes vertebral syndrome (restricted painful movements in the lower back, flattened lumbar lordosis, tense and rigid paravertebral muscles). In order to alleviate the pain, the patient keeps a specific posture - the

body is slightly bent forward and to the one side. The direction depends on site of nerve root compression (medial or lateral) – antalgic posture (Fig. 9.5).

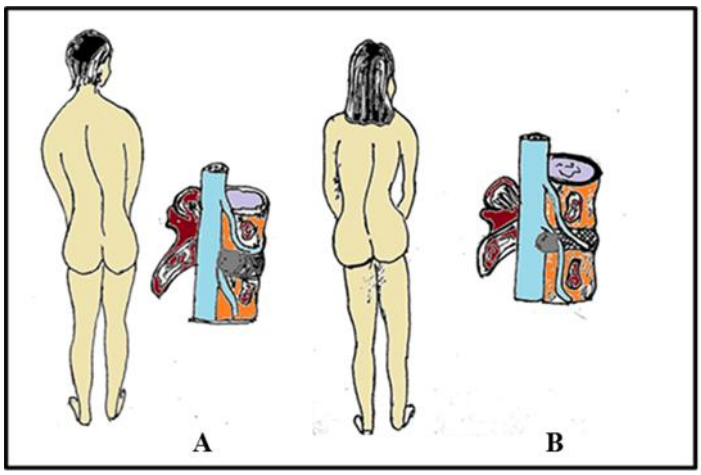

Fig. 9.5. The antalgic posture and its direction: **A/** contralateral scoliosis in case of lateral compression of the nerve root; **B/** ipsilateral scoliosis in case of medial compression of the nerve root

2. Lumboischialgic syndrome – the back pain radiates to the left or right leg. It is sharp with distinct localization along the dermatome (sensory distribution) of the compressed nerve root. The pain intensifies when standing up, bending down, walking, coughing, exertion, etc. The straight leg raise test is positive (Lasséque sign).

3. Lumbar radiculopathy syndrome. The long-standing compression of the nerve root leads to secondary neurological symptomatology – hypesthesia, hyporeflexia/areflexia, and muscle weakness. In cases of large herniatoions, the autonomic S_2-S_5 nerve roots can be affected which leads to the so called "cauda equina syndrome". The latter is characterized by perianal hypoesthesia and loss of bowel and bladder control. This syndrome is indication for emergency surgery within 24 hours.

I. Disc disease in the lumbar region

1. Disc herniation at L_3-L_4 level: the incidence varies between 5%-10% of all lumbar disc herniations. L_4 nerve root is compressed, which causes reduced sensitivity on the antero-lateral surface of the thigh, the popliteal region and the medial surface of the shank. The knee-jerk reflex is reduced. In severe cases knee extension can be

impaired. The femoral stretch test is positive – with the patient prone, the examiner grasps the patient's ankle on the symptomatic (ipsilateral) side and facilitates ipsilateral knee flexion; reproduction of typical lower extremity pain constitutes a positive test.

2. Disc herniation at L_4-L_5 level – L5 nerve root is affected (Fig. 9.6 and Fig. 9.7). Hypoaesthesia spreads on the lateral surface of the thigh and the shank reaching the great toe of the foot. The tendon reflexes are usually intact. In advanced cases, there can be weakness of the dorsal flexion of the great toe and/or the foot as well as hypotrophy of the gluteal muscles – the patient cannot walk on heels.

3. Disc herniation at L_5-S_1 level – S_1 nerve root is affected (Fig 9.8. and Fig. 9.9.) It is accompanied by reduced sensitivity along the postero-lateral surface of the thigh, the calf, heel and the lateral side of foot to the 5^{th} toe. The Achilles reflex is weakened. In advanced cases, there can be weakness of the plantar flexion of the great toe and/or the foot as well as hypotrophy of the calf – the patient cannot walk on toes.

4. Cauda equina syndrome: Large medial disc herniations can cause severe compression of the dural sac which contains the nerve roots of cauda equina (Fig. 9.10 and Fig. 9.11). The condition is manifested with sphincter disorders, saddle hypesthesia and isolated paralysis of different muscle groups of the legs.

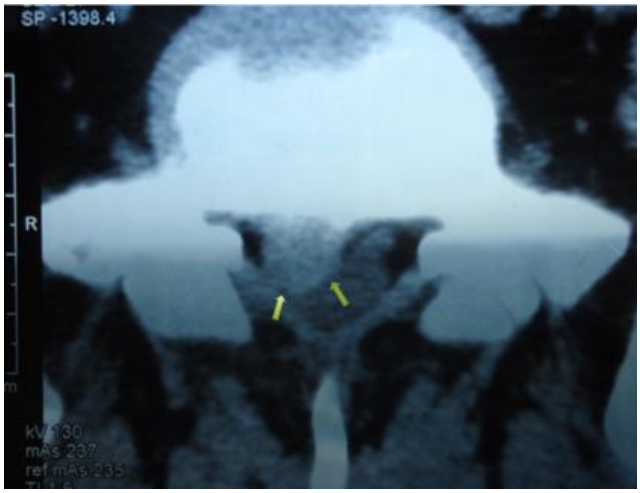

Fig. 9.6. CT image of disc herniation at L_4-L_5 level

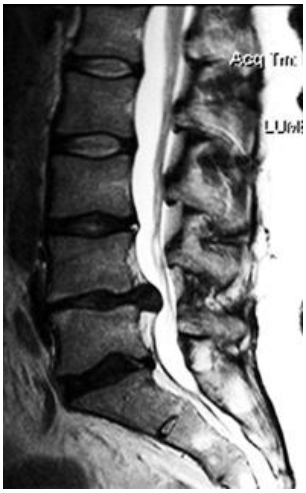

Fig. 9.7. MRI image of disc herniation at L_4-L_5 level

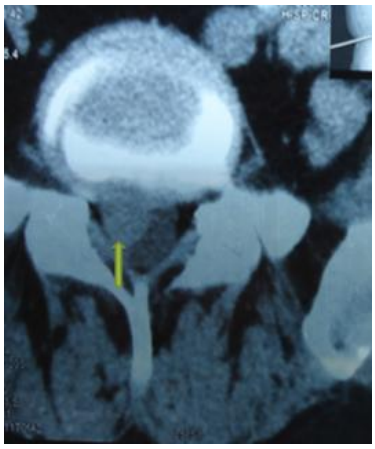

Fig. 9.8.CT image of disc herniation at L_5-S_1 level

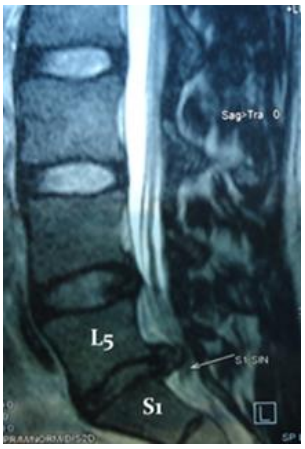

Fig. 9.9. MRI image of disc herniation at L_5-S_1 level

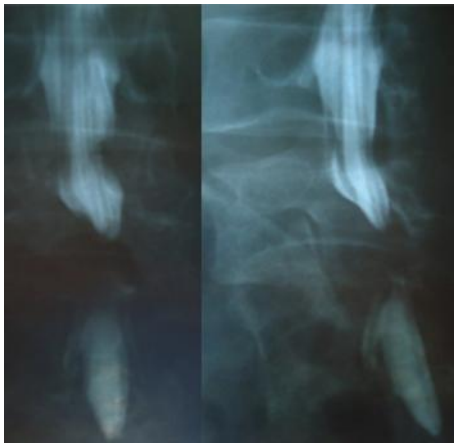

Fig. 9.10. Myelography demonstrating median disc herniation causing cauda equina syndrome

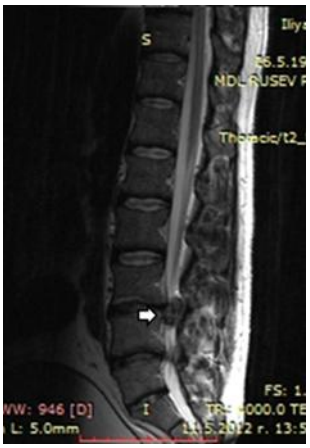

Fig. 9.11. MRI image of $L_{4\text{-}5}$ disc herniation leading to cauda equina syndrome

II. Disc disease in the cervical region

Initially, the clinical manifestation presents with pain in the back of the neck, which radiates between the scapulae and towards the left or right arm. Usually, the complaints begin after physical overloading. The cervical paravertebral muscles are rigid and painful. The head takes compensatory position. Neck and arm pain intensifies upon exertion.

1. Disc herniation at C_4-C_5 level - C_5 nerve root is affected: pain, hypesthesia and paraesthesia in the base of the neck, the shoulder and the anterior brachial surface, reduced biceps reflex, weakness of the deltoid muscle (Fig. 9.12).

2. Disc herniation at C_5-C_6 level - C_6 nerve root is compressed and this is manifested by: pain, hypesthesia and paraesthesia along the

lateral brachial and antebrachial surface reaching the thumb and the forefinger, reduced reflex and weakness of the biceps (Fig. 9.12).

3. Disc herniation at C_6-C_7 level - C_7 nerve root is affected: pain and hypesthesia along the anterolateral surface of the arm to the middle finger, reduced triceps reflex, weakness of the triceps and the extensors of the wrist (Fig. 9.13).

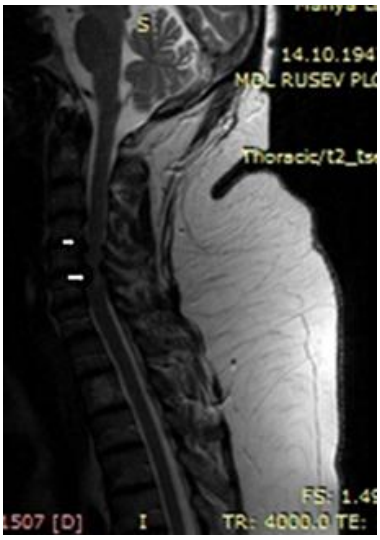

Fig. 9.12. MRI image of disc herniation at C_{4-5} и C_{5-6} levels

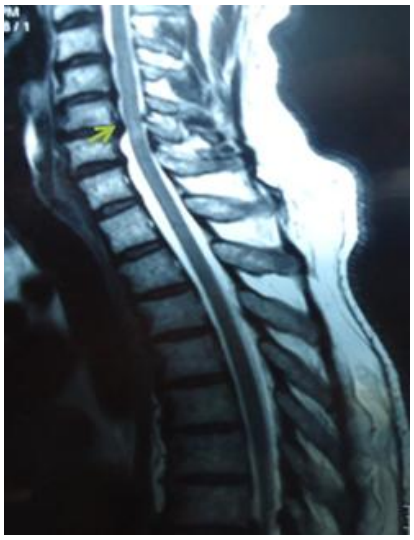

Fig. 9.13. MRI image of disc herniation at C_6-C_7 level

4. Disc herniation at C_7-Th_1 level - C_8 nerve root is affected. The clinical features include pain, hypesthesia and paraesthesia along the posterior surface of the arm, the palm, the 4^{th} and 5^{th} finger. Weakness and hypotrophy of the palmar muscles can also be observed.

5. Median cervical disc herniations – they cause acute or gradual but progressive compression of the spinal cord. This can result in spastic quadriparesis with pathological reflexes and conductive hypesthesia distally from the level of damage as well as bowel and bladder retention.

III. Disc disease in the thoracic region

1. Thoracic disc herniations - radicular symptoms are minor. However, when present, they manifest with symptoms of intercostal neuralgia. Thoracic disc herniation causes myelopathic symptoms – inferior paraparesis, conductive hypesthesia below the level of the herniation and bowel and bladder dysfunction (Fig. 9.14).

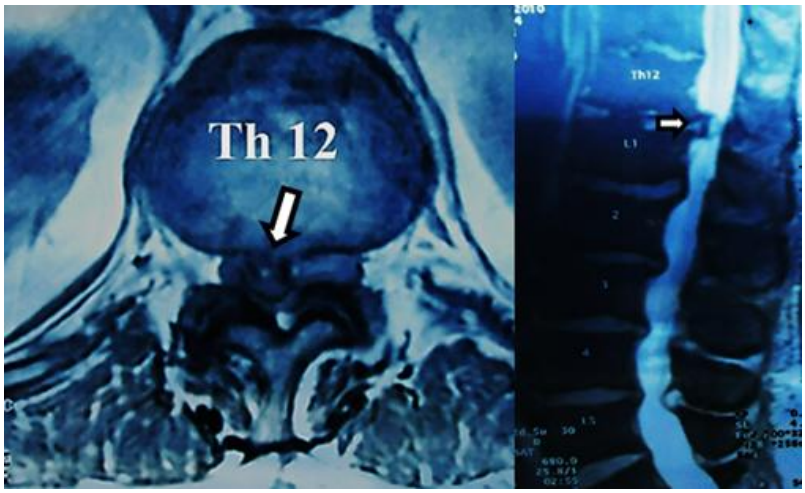

Fig. 9.14. Axial and sagittal MRI – thoracic disc herniation at Th_{12}-L_1

Differential diagnosis: This includes all conditions which cause myelopathies and radiculopathies (spinal cord tumors, inflammatory diseases - tuberculosis, spondylodiscitis, damages to the brachial plexus, syringomyelia, etc.

Diagnosis

- **spondylography** – the different projections may detect degenerative changes (narrowed disc space, osteophytes, flattening of the physiological lordosis) spondylolisthesis, osteolytic changes, infectious changes of the disc, etc.;
- **myelography** - this examination includes the injection of water-soluble contrast agent into the subarachnoid space. In the case of compression of the nerve structures, a "filling defect" or "complete stop" of the contrast agent can be observed (Fig. 9.10 and Fig. 9.15);
- **computed tomography** - demonstrates the degenerative bone alterations, calcifications of the longitudinal ligaments, disc herniation, vertebral dislocations, etc. (Fig. 9.6. and Fig. 9.8);
- **magnetic resonance imaging (MRI**) – this is the most sensitive imaging method for examination of spinal pathology. It provides superior details about the degenerative process in the intervertebral discs and the spine as well as their relation to the adjacent neural structures (Fig. 9.7, Fig. 9.9, Fig. 9.11, Fig. 9.12, Fig. 9.13 and Fig. 9.14).

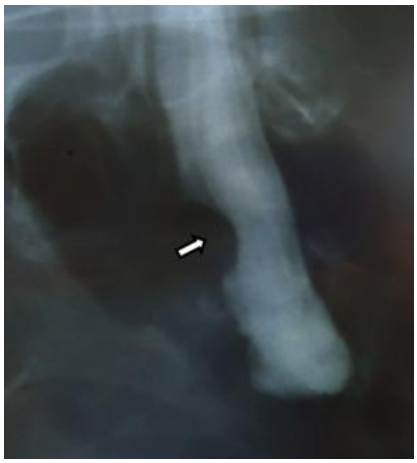

Fig. 9.15. Myelography demonstrating disc herniation at L5-S1 level

Treatment of the disc disease

A. **Conservative treatment.** It should be applied intensively for a sufficient period of time depending on the severity of the clinical symptoms. In general, it includes:
   - rest on a hard flat bed;
   - analgetics (including local paravertebral application);
   - anti-inflammatory non-steroid or steroid or drugs;
   - physiotherapeutic procedures (including manual therapy and extension procedures);
   - balneotherapy.

The conservative treatment should be applied for a period of 15-20 days. In 90% of the cases, especially those with bulging disc or disc protrusions, the treatment leads to alleviation of symptoms. This should be followed by a regime of physical activity which aims at strenghtening of the paravertebral musculature.

If the neurological symptoms persist or progress, imaging study should be performed in order to diagnose lesions which may require surgical treatment.

B. **Surgical treatment** – indications:
   - **urgent indications** – this includes acute onset of neurological deficit (paresis or paralysis, cauda equina syndrome with loss of bowel and bladder control, etc);
   - **elective indications:**

- intractable pain which cannot be alleviated by conservative treatment;.
- progressive neurological deficit (motor and sensory);
- persisting axial pain with compensatory scoliosis .

The surgical intervention in the case of cauda equina syndrome has to be performed as soon as possible (within 24 hrs) because any delay leads to irreversible neural damage and may compromise the functional recovery.

Spinal stenosis

The term 'spinal stenosis' was first introduced by the Dutch neurosurgeon Henk Verbiest in 1954. This term indicates narrowing of the spinal canal and foramina which harbor the neural structures. Spinal stenosis can affect:

- the cervical spine – more frequent in males;
- the thoracic spine – more frequent in males;
- the lumbar spine – more frequent in females.

Types of spinal stenosis

1. Congenital. It is observed among young individuals and is due to diffuse skeletal dysplasia – achondroplasia, spondylo-epiphyseal dysplasia, severe malformations of the spine and spinal cord such as spina bifida, meningocele, vertebral dysgenesia, etc. Some authors call it developmental stenosis in order not to underestimate and disregard the postnatal factors of growth and development.

2. Acquired. It can result from a variety of factors: degenerative spondylosis, hypertrophy and ossification of ligaments and joint capsules, posttraumatic, postoperative, metabolic-endocrinal (acromegaly), hypoparathyroidism, renal osteodystrophy, etc.

The narrowing is due to osteophytic proliferation. The osteophytes can be situated anteriorly to the dura matter when they occur on the posterior edges of the vertebrae; lateral osteophytes – when they occur at the pedicules (Fig. 9.17) and postero-lateral – when they originate from the intervertebral facets (Fig. 9.16). Each of the three types can cause compression of nerve roots in the area of the lateral recess and/or the intervertebral foramina.

The degenerative spondylosis is the most common cause for spinal stenosis.

Degenerative spondylolisthesis is the anterior or posterior displacement of a vertebra or the vertebral column in relation to the vertebrae below. It is frequently caused by degenerative alterations in the tendons which lead to their loosening and subluxations. Thus, there is narrowing of the spinal canal between the vertebral arch of the upper vertebra and the posterior upper edge of the body of the lower vertebra. The dural sac and the nerve roots are compressed. The degenerative spondylolisthesis is observed after the age of 50, predominantly in women at L_4-L_5 level (Fig. 9.18). It is a result of osteoarthritic hypertrophy of the intervertebral joints. In quantitative terms, the evaluation of spondylolisthesis is based on the degree of displacement of the vertebral body (Fig. 9.19). The lower percentage of displacement (10%-50%) is associated with lower risk of progression of the deformity.

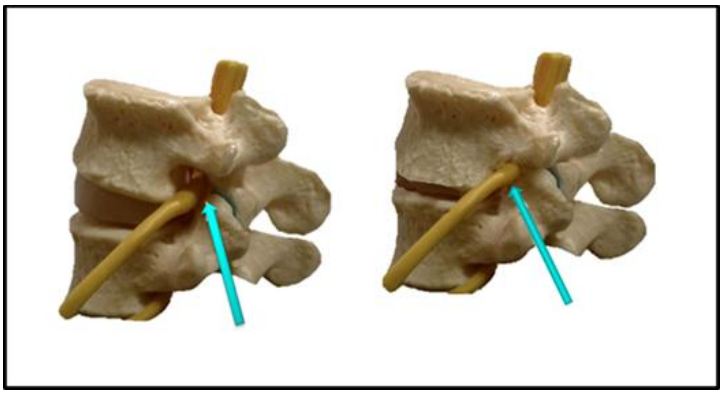

Fig. 9.16. Postero-lateral stenosis due to osteophytosis of the joints leading to narrowing of the nerve root foramina

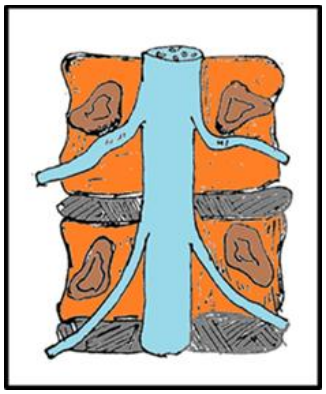

Fig. 9.17. Degenerative stenosis due to pedicular hypertrophy

Clinical features of the spinal stenosis

1. **Clinical presentation of the spinal stenosis in the cerival and the thoracic region**: It is manifested by symptoms of myelopathy (central paresis, sensory impairments due to damage of the posterior and lateral spinal cord columns, loss of bowel and bladder control). In addition, mono- or polyradiculopathy can overlap the former symptoms.
2. **Clinical presentation of the lumbar spinal stenosis**:
   - **lumboischialgia** presents with pain in the lower back radiating to one or both legs. It intensifies when standing up or walking and alleviates while at rest or when the lumbar spine is flexed; The lateral stenosis is similar in its presentation to disc herniation but frequently the Lasséque sign is negative;

- **lumboradiculopathy** (unilateral or bilateral) – in 40%-45% of the cases;
- **neurogenic claudication** is observed in 60-70% of the cases with central lumbar spinal stenosis. This term was introduced by J.G. Evans in 1964. It results from ischemia of the nerve roots of cauda equina. In contrast to the vascular intermittent claudication, here the arterial pulsations of the lower limbs are preserved. It is characterized by the presence of unilateral or bilateral paraesthesia, numbness and weakness during ambulation. It forces the patient to stop walking and rest. Flexion in the lumbar area alleviates the pain in the legs;
- **sphincter disoreders** are rare.

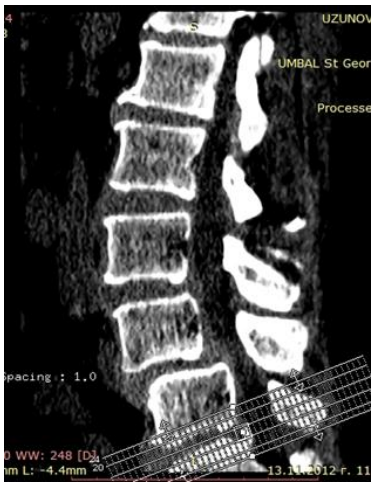

Fig. 9.18. CT reconstruction of degenerative spondylolisthesis at L_4-L_5 level

Fig. 9.19. Degrees of spondylolisthesis: I degree - below 25%; II degree - 25%-50%; III degree - 50%-75%; IV degree - over 75%.

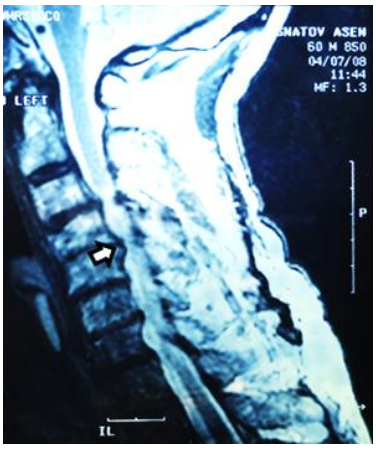

Fig. 9.20. MRI image of cervical stenosis at C_4–C_5 level

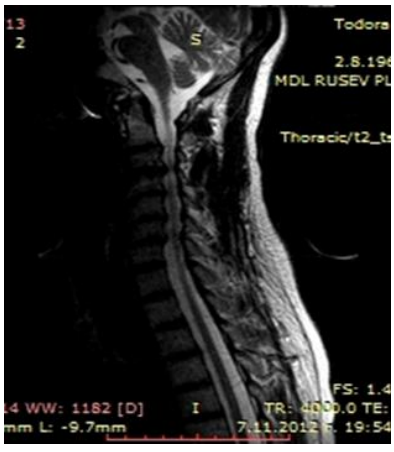

Fig. 9.21. MRI image of multilevel cervical stenosis (C_3 - C_6)

Diagnosis of spinal stenosis

- **spondylography** can suspect the presence of spinal stenosis. According to H. Verbiest (1955), the shortening of the sagittal diameter of the canal under 12 mm indicates relative central stenosis and under 10 mm - absolute central stenosis. Spondylolisthesis can be easily seen on plain spinal X-rays. In these cases, it is better to obtain dynamic spondylographies (in flexion and extension) in order to identify segmental spinal instability (Fig. 9.22);
- **radiculosaccography –** allows the identification of a filling defect resulting from the stenotic compression;

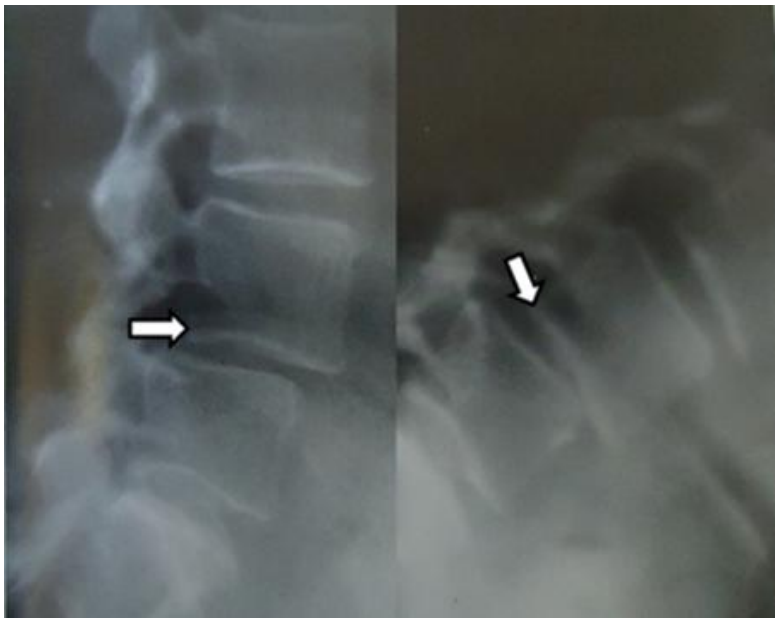

Fig. 9.22. Dynamic spondylography- increase in listhesis is not identified

- **computed tomography** – visualizes the degenerative changes in the bone structures which comprise the canal (Fig. 9.23). It also allows the identification of the type of the spinal stenosis (central, lateral or mixed) and its relationship to the adjacent neural structures (Fig. 9.24).
- **magnetic resonance imaging** is a particularly valuable study in cases with cervical and thoracic stenosis. It is superior to CT in the detection of any alterations in the neural structures and soft tissues (Fig. 9.20, Fig. 9.21 and Fig. 9.25).

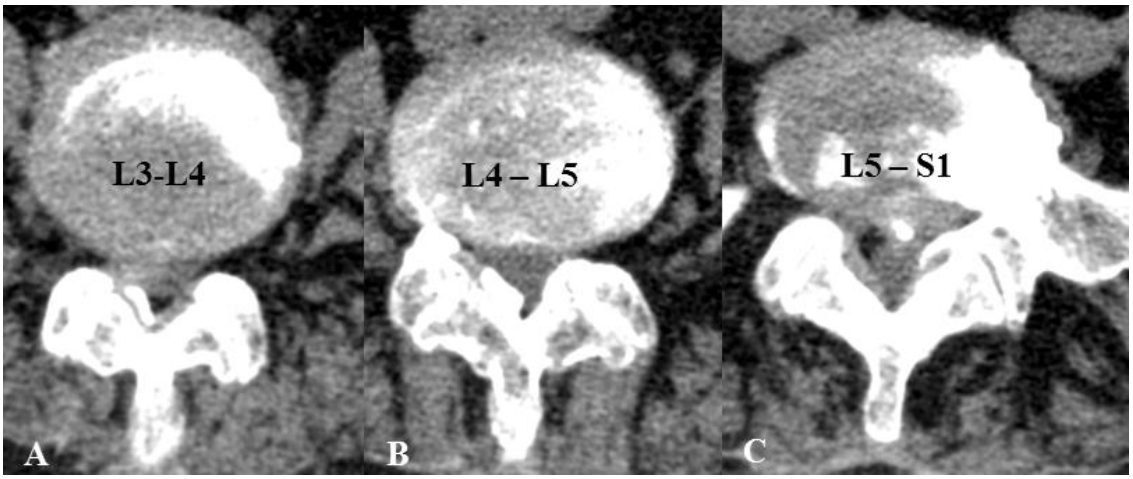

Fig. 9.23. CT images in a patient with: **A/** right paramedian L_3-L_4 disc herniation; **B/** right lateral stenosis at L_4-L_5 level; **C/** median L_5-S_1 disc herniation

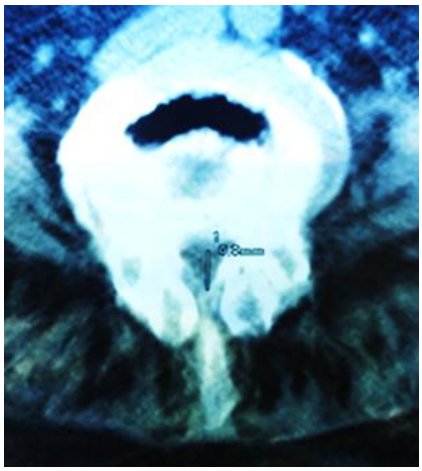

Fig. 9.24. CT- narrowed neural foramen with pronounced hypertrophy of the apophyseal joints

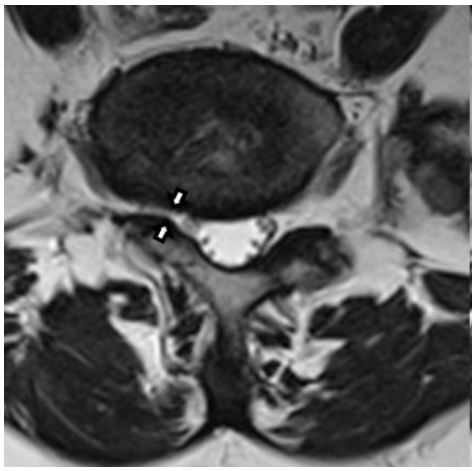

Fig. 9.25. MRI image of right foraminal stenosis at L_5 - S_1 level

Treatment of spinal stenosis

The indications for surgical treatment are similar to those of disc herniations – lack of effect from the conservative treatment in case of intractable pain and/or progressive neurological deficit and sphincter disorders. The precise diagnosis for identification of the type of stenosis (central, lateral or mixed), is of paramount importance for the preoperative planning of the extent of the surgical intervention.

The aim is to decompress the affected neural structures without causing instability of the affected segment.

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CHAPTER 10 *T. Kitova, B. Kalnev, A. Davarski*

HYDROCEPHALUS

Hydrocephalus is an enlargement of the ventricles of the brain due to excessive accumulation of cerebrospinal fluid (CSF). It is a pathology of the ventricular system which is due to impairments in the production, circulation or resorbtion of CSF. This is the so called "internal hydrocephalus" which differs from the "external hydrocephalus" which is characterized by an enlargement of the subarachnoid space. The hydrocephalus is a congenital condition. It refers to the malformations of the CNS.

Congenital hydrocephalus should be differentiated from the hydrocephalic syndrome which is caused by a variety of etiologies.

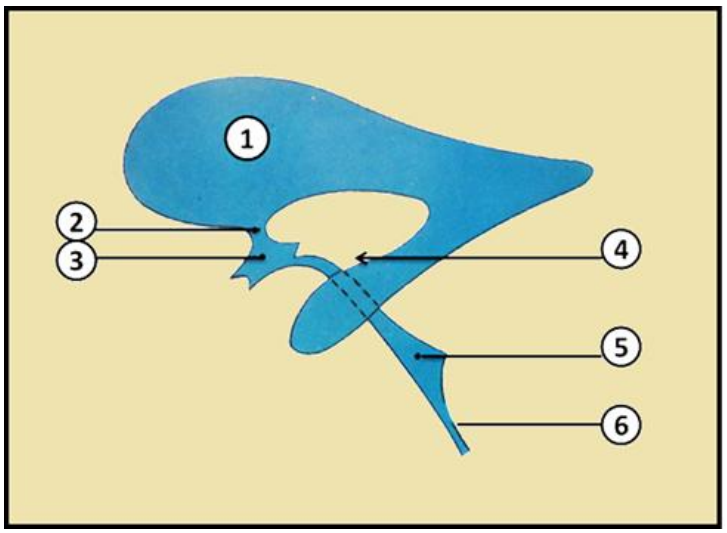


Fig. 10.1. Ventricular system:
1. Lateral ventricle; 2. Foramen of Monroe; 3. Third ventricle;
4. Aqueduct of Sylvius; 5. Forth ventricle; 6. Foramen of Magendie

The incidence of congenital hydrocephalus varies from 0.1% to 0.4% as it increases if the cases of hydrocephalic syndrome are included.

Etiopathogenesis

The etiopathogenesis of the congenital hydrocephalus is complex and depends on numerous factors that influence on the intrauterine development of the fetus and the infant.

Types of hydrocephalus

1. **Communicating hydrocephalus** – the CSF circulates freely in the ventricular system but its resorbtion is impaired.

2. **Obstructive hydrocephalus** results from obstruction of the CSF pathways from one ventricle to another or from the ventricular system to the subarachnoid space. According to the location of obstruction, there are several forms of this condition:

- atresia of the interventricular foramina (foramina of Monroe) (Fig. 10.2);
- stenosis of the aqueduct of Sylvius (Fig. 10.3);
- occlusion of the foramina of Magendie and Luschka.

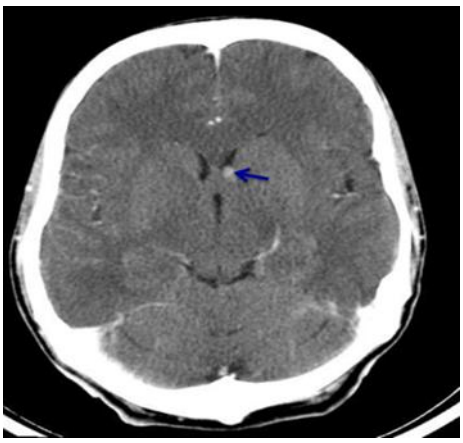


Fig. 10.2. CT image of obstruction of the foramen of Monroe due to tuberous sclerosis

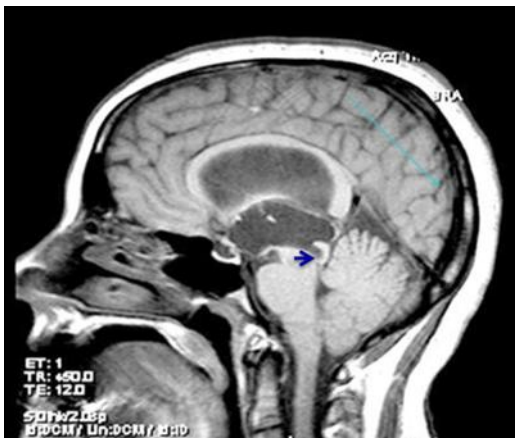


Fig. 10.3. CT image of stenosis of the aqueduct of Sylvius

Aqueductal stenosis is the most common cause of congenital hydrocephalus. It can result from:

- neonatal infections – ventriculitis which leads to stenosis of the aqueduct of Sylvius. Subependimal glial hypertrophy, which may be caused by variety of factors (toxic or inflammatory), can also lead to aqueductal stenosis;
- hypovitaminosis or hypervitaminosis “A”;
- intoxications or ionizing radiation;
- mixoviruses which cause specific infection of the ependyma leading to aqueductal stenosis and obstructive hydrocephalus.

Diseases that may lead to a pathological enlargement of the ventricular system:

- previuos infection (meningitis) may cause hydrocephalus due to the formation of adhesions and membranes in the ventricles, the subarachnoid cisterns and the openings of the 4th ventricle;
- subarachnoid hemorrhage may lead to arachnoiditis which impedes the circulation and resorbtion of the CSF that may result in hydrocephalus;
- CNS malformations that are etiopathologically related to hydrocephalus – Arnold-Chiari malformation, Dandy-Walker syndrome, basilar impression, etc.

Pathophysiological mechanisms of paediatric hydrocephalus

It results from impaired balance between the CSF and the venous systems due to agenesia or congenital defect of the arachnoid villi that impede the CSF resorbtion. This leads to an increased CSF volume, elevated intracranial pressure and, eventually, widening of the ventricles. The opened cranial sutures and the relatively elastic infant skull predispose to excessive enlargement of the head circumference, thus, preventing cerebral veins and brain parenchyma from further damage.

Once these compensatory mechanisms deplete, the increased ventricular pressure prevails, thus, causing collapse of the brain vessels, leading to ischemia and damage to the brain.

A. **Clinical features of hydrocephalus in infants**

The disease manifests in the first weeks after birth. The hydrocephalic babies have typical appearance: enlarged head with preserved facial skull size. The roof of the orbits protrudes forward and downward. The sunset eye sign (also known as the “setting sun phenomenon”) is a clinical phenomenon encountered in infants and young children with raised intracranial pressures (seen in up to 40% of children with obstructive hydrocephalus and 13% of children with shunt dysfunction). It consists of an up-gaze paresis with the eyes appearing driven downward. The lower portion of the pupil may be covered by the lower eyelid, and sclera may be seen between the upper eyelid and the iris (Fig. 10.4). It is presumably related to Parinaud's syndrome and results from compression of the peri-aqueductal structures. The skin of the head is thinned; the veins of the

forehead and the scalp are dilated with numerous collaterals (Fig. 10.5). Strabismus and/or ophthalmoparesis can also be present. A "cracked pot sound" can be heard during skull percussion. Subsequently, if left untreated, hydrocephalus leads to delayed psychomotor development accompanied by pyramidal and cranial nerve symptoms. Fundoscopy reveals papilledema or, at advanced stage, secondary optic athrophy resulting in blindness.

This disease may also follow subacute or chronic progression with remission periods of normal development.

B. **Hydrocephalus in children and adults**

During this period, the hydrocephalic syndrome predominates due to CSF pathways obstruction. As a result of the closure of the cranial sutures and lack of compensatory head enlargement, the clinical presentation follows acute progression, dominated by symptoms of intracranial hypertension. At fundoscopy, papilledema with secondary optic atrophy may be observed. With disease progression, oculomotor and abducens nerve palsies, Parinaud's syndrome, pyramidal syndrome, neuroendocrine symptoms can also develop.

In adolescents and adults urgent shunting procedure should be performed in order to avoid brain herniation and death.

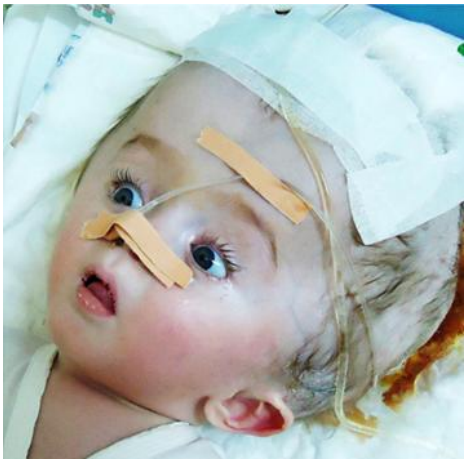


Fig. 10.4. The setting-sun phenomenon

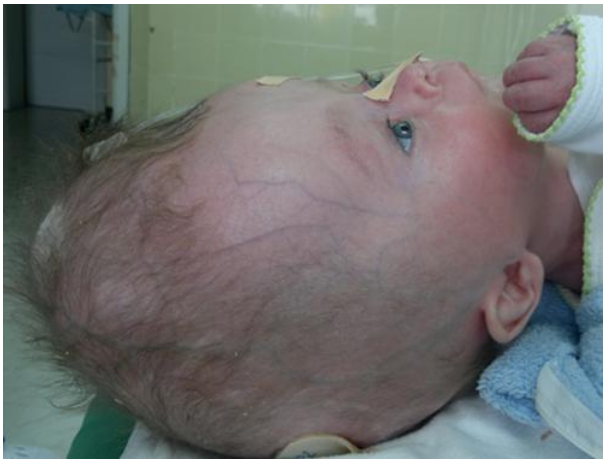


Fig. 10.5. Dilated scalp veins with numerous collaterals

C. **Normal pressure hydrocephalus (NPH)**

In 1965 Adams was the first to describe hydrocephalus in adult patients that were believed to have normal ventricular pressure. The disease is presented by the triad: dementia, gait disturbance and incontinence. The condition develops after previous subarachnoid hemorrhage, meningoencephalitis, head injury, etc. The CSF resorbtion is impaired. The increased size of the ventricles causes changes in the brain parenchyma which eventually leads to its atrophy. The compressed brain vessels impede the normal cerebral perfusion which leads to hypoxia and further damage to the brain structures, thus worsening the neurological deficit.

Diagnosis

- **CT and MRI** are the methods of choice in the diagnosis of hydrocephalus. They provide detailed information about the size of the ventricles, the changes of the subarachnoid space, the presence and site of CSF obstruction and transependimal resorbtion. (Fig. 10.6, Fig. 10.7 and Fig. 10.8);
- **ultrasound diagnosis** is used to diagnose congenital intrauterine hydrocephalus. It can also be used for transcranial examination of the ventricular size in newborns and infants. (Fig. 10.9);
- **radioisotope diagnosis** examines the CSF circulation impairments and the functional integrity of the shunting systems. The isotope is injected in the reservoir of the valve and the CSF flow is observed by gamma-camera.

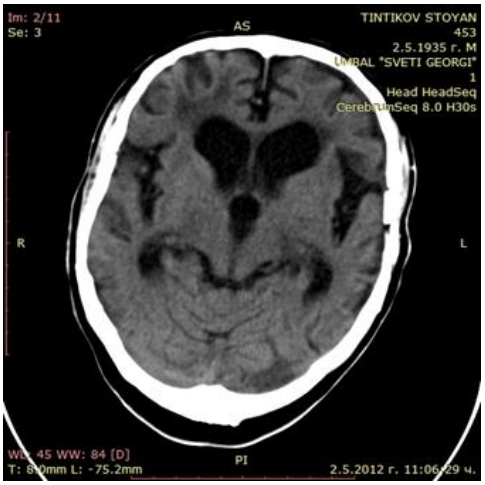


Fig. 10.6. Normal pressure hydrocephalus

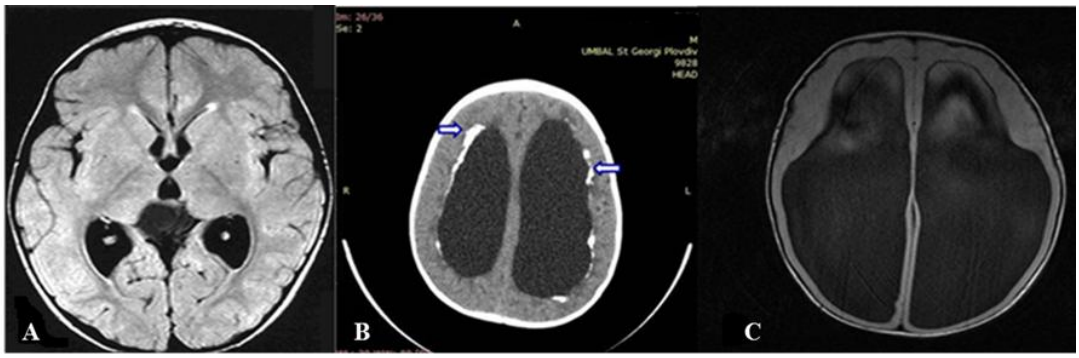


Fig. 10.7. CT images: A/ normal brain ; B/ severe hyfrocephalus with transependimal resorbtion (⇨) ; C/ extreme hydrocephalus

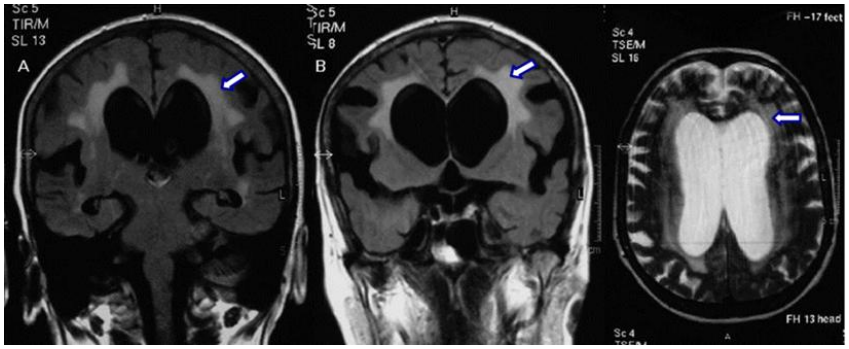


Fig. 10.8. MRI image of hydrocephalus with transependimal CSF resorbtion (⇦)

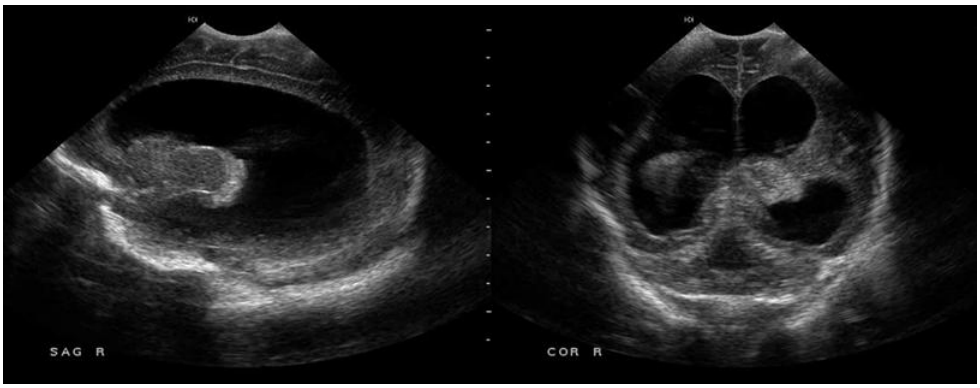


Fig. 10.9. Fetal ultrasound examination demonstrating intrauterine hydrocephalus

Treatment

The treatment of hydrocephalus is surgical. The following surgical interventions are possible:

- eliminating the obstruction by removing a tumor or another pathology;
- endoscopic CSF diversion by means of interventricular or ventriculo-cisternal anastomoses: the most common procedure

is the endoscopic third ventriculostomy which is efficient in cases of aqueductal stenosis (Fig. 10.10);

- CSF shunting can be performed by a variety of valves: ventriculo-peritoneal, ventriculo-atrial or lumbo-peritoneal. Ventriculo-peritoneal shunting is the most common procedure for the treatment of hydrocephalus. Good results can be achieved in 80-85% of the cases at low mortality rates (less than 0.5%). The main problem when performing shunt surgery is to choose a system with an appropriate pressure. This problem is being currently overcome by the introduction of adjustable pressure.

The complication rate of valve shunts is high and reaches 35-40% among different series**.** The most common causes are:

- hypodrainage or hyperdrainage of the shunt system;
- infectious cimplications;
- cardiac complications, peritonitis, ileus;
- subdural hematoma;
- valves (Fig. 10.11 and 10.12).

The surgical treatment of hydrocephalus provides the opportunity for normal psychomotor development of children. In case of acute hydrocephalus, the shunting is a life-saving procedure.

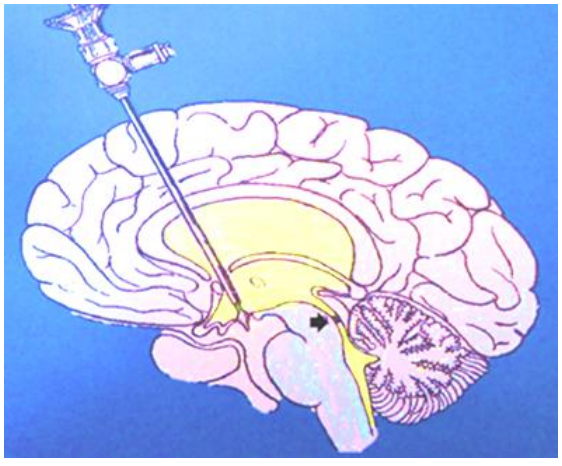
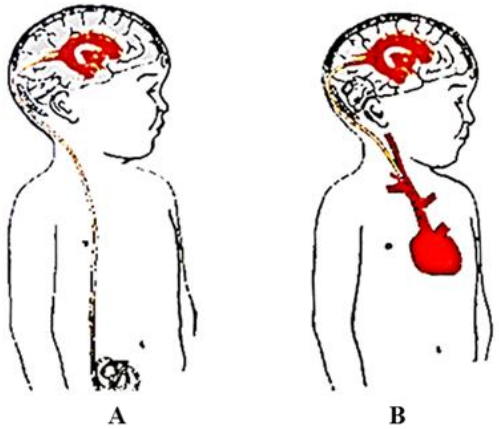


Fig. 10.10. Third ventriculostomy in aqueductal stenosis (➡)



Фиг. 10.11. Shunting procedures: **A/** ventriculo - peritoneal; **B/** ventriculo-atrial

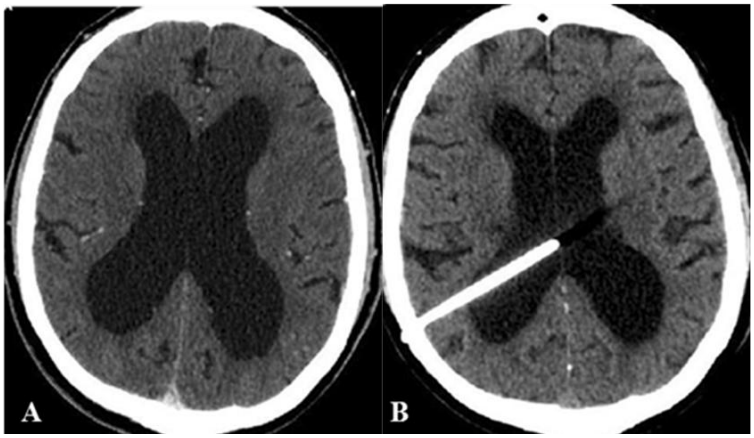


Fig. 10.12. CT – hydrocephalus :
A/ before shunting; **B/** after shunting

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CHAPTER 11 *A. Masmoudi, B. Kalnev, H. Zheliazkov*

MALFORMATIONS OF THE NERVOUS SYSTEM

Craniosynostosis

Craniosynostosis (sometimes called craniostenosis) is a disorder in which there is early fusion of one or more sutures of the skull in childhood. It produces an abnormally shaped head and, at times, abnormal features of the face. The deformity varies significantly depending on the suture or sutures involved. This condition can be hereditary and is often associated with other malformations that affect the facial skull (cranio-facial dysostosis – Crouzon syndrome), or the fingers (Apert syndrome, Carpenter syndrome).

Incidence. It occurs predominantly in males and varies from 0.4 to 0.6/1000 newborns.

Pathoanatomy

Normally after birth, there is fibrous tissue between the cranial bones, which gardually ossifies. The development of the skull includes two stages:

- stage I (infancy) is characterized by a rapid growth of the head mainly on account of the cranial sutures;
- stage II (from the 1^{st} year to 6^{th} year) – relatively slow and diffuse enlargement of the cranial bones by increase of the size of lamina externa and reduction of the size of lamina interna.

In case of premature fusion of one or more cranial sutures, the enlargement of the head becomes asymmetric and expands in the direction of the unclosed sutures. Craniostenosis is an active synostosis that differs from microcephaly in which the synostosis is passive due to lack of brain development (Fig. 11.1).

Etiopathogenesis. It is not completely understood. A variety of factors during pregnancy can contribute to premature fusion of the sutures: intrauterine infections, metabolic disorders, etc. Nowadays, it is considered that genetic factors, disturbed metabolism of mucopolysacharides and calcium, hyperthyroidism or hypercalciemia can play a role in the development of this disease.

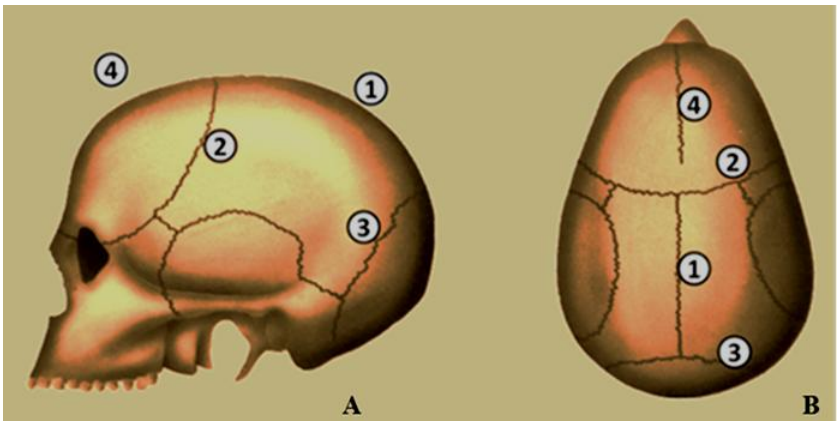

Fig. 11.1. Cranial sutures:
1. Sagittal; 2. Coronal; 3. Lambdoid; 4. Metopic

Types of craniosynostosis:

1. Scaphocephaly presents premature fusion of the sagittal suture which leads to elongation of the antero-posterior diameter of the head (Fig. 11.2A).

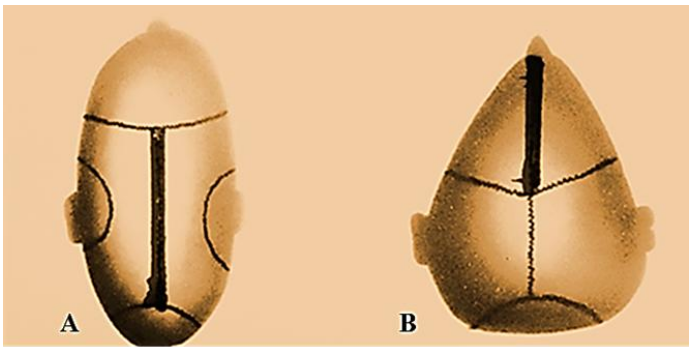

Fig. 11.2. Craniosynostosis: **A/** scaphocephaly; **B/** trigonocephaly

2. Trigonocephaly is caused by an early ossification of the metopic suture (between the frontal squamas) which leads to a triangular deformity of the forehead and hypertelorism on account of compensatory enlargement of the parietal lobes (Fig. 11.2B).

3. Brachycephaly presents a premature fusion of both coronal sutures. The head grows to the lateral and superior direction (Fig. 11.3A).

4. Plagiocephaly – in this case, there is unilateral ossification of the coronal sutures. At the side of fusion, the head grows superiorly and laterally but not anteriorly (Fig. 11.3B).

5. Oxycephaly presents premature fusion of more than two cranial sutures (coronal, sagittal, lambdoid). The configuration of the head depends on the moment of occurrence and the number of the affected sutures (Fig. 11.4).

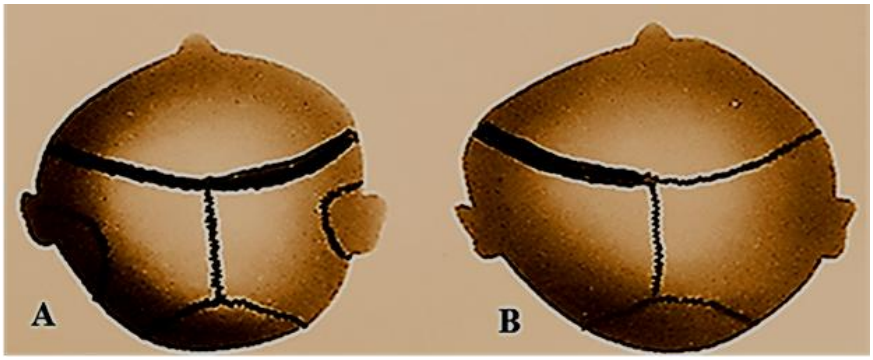

Fig. 11.3. Craniosynostosis: **A/** brachycephaly; **B/** plagiocephaly

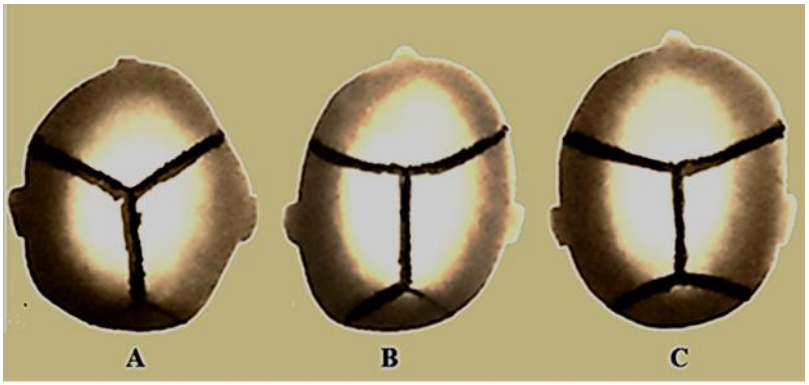

Fig. 11.4. Oxycephaly – synostosis of: **A/** coronal and sagittal sutures; **B/** coronal, sagittal and lambdoid sutures; **C/** total synostosis

Clinical features: There are two forms of clinical evolution: latent (50% of the cases) and acute (40% of the cases). In the first case, the deformity of the skull develops gradually without symptoms of increased intracranial pressure and neuropsychological disorders. In the acute form, the disease develops rapidly and causes symptoms of intracranial hypertension and mental retardation. Epileptic seizures can sometimes be present. Ophthalmological disorders include exophthalmos, hypertelorism, strabismus, etc. They are related to the type of craniostenosis and the deformity of the orbits. The stenosis of the optic canal causes atrophy of the optic nerve. Endocrine disorders are frequent and manifest with hypotrophy and delayed puberty.

Diagnosis

1. **Skull X-rays (craniography)** allow detection of the prematurely ossified sutures, the type of cranostenosis and the presence of associated malformation. Long-standing intracranial hypertension may appear as enlargement of impressiones digitatae of the skull.
2. **Computer tomography** specifies the extent of ossification of the cranial sutures; allows measurement of the different cranial diameters and reveals pathological changes in the brain structures.

Treatment

The surgical treatment aims at cosmetic correction of the cranial deformity and decompression of the brain and optic nerves. This is achieved by bone resections of the ossified suture or sutures and creation of specific bone flaps. The presence of intracranial hypertension is an absolute indication for surgical treatment.

The most favorable period for this intervention is before the 9th month after birth.

Cranial dysostosis

1. **Crouzon syndrome**. This condition is inherited and the most common manifestations are brachycephaly, small orbits, hypertelorism and exophtalmos. Divergent strabismus, nystagmus and deformation of the nose could also be present. In some cases, there is total synostosis and stenosis of the optic canal which causes secondary atrophy of the optic nerves. The condition often occurs along with other malformations (heart defects, spina bifida, etc.)
2. **Acrocephalosyndactyly (Apert syndrome)**. It is a combination between bilateral coronal synostosis (brachycephaly), syndactyly and enlargement of the distal phalanx of the thumb. The other malformations are similar to those typical of Crouzon disease.
3. **Carpenter syndrome**. Besides brachycephaly and syndactyly, there is lateral displacement of the joints of the small fingers.

Dysraphic malformations

The dysraphic malformations are 60-70% of all CNS anomalies. They occur as a result of defect during the embryonic development caused by impaired embryogenesis of the mesoderm and the ectoderm

leading to incomplete fusion of the structures in the midline – the formation of the neural tube is disturbed.

Cranial dysraphism

1. **Cranial meningocele** This malformation is characterized by prolapse of the meninges through the available cranial bone defect. This cystic formation is filled with CSF and communicates with the intracranial subarachnoid space. The most common localization is in the occipital region (60-70%) (Fig. 11.5). Surgical treatment is required. The defect is resected and closed. Thus, intracranial infection is avoided.

Fig. 11.5. Occipital meningocele:
A/ before surgery; **B/** after surgery

2. **Encephalocele** In this case, the meningeal sac contains neural structures such as abnormal brain tissue and parts of the ventricular system. This malformation can be found along the entire midline:
   - **Frontal encephalocele** results from a defect of the anterior part of the neural tube and is localized on a relatively wide base in the inferior part of the frontal bone (at the base of the nose) (Fig. 11.6A and Fig. 11.7). At times, the sac is separated in two because of the division of the bone defect by crista gali. In other cases, there is propagation of the sac to the orbits.
     - **clinical features** include deformation of the face and widening of the basis of the nose, hypertelorism, strabismus,

loss of olfaction, impaired nasal breathing. Focal neurological deficit is rare. When the bone defect is located in the base of the skull, the sac can enter the oral or the nasal cavity through the sphenoid or the ethmoid sinuses – basal encephalocele;

- **diagnosis:**
  - ✓skull x-rays may demonstrate the bone defect.
  - ✓CT and MRI can visualize the neural structures inside the sac and concurrent malformations (Fig. 11.7 and Fig. 11.7).
- **the surgical treatment** aims at excision of the sac along with its contents and correction of the facial deformity.

- **Occipital encephalocele –** the skull defect is in the occipital bone. The most frequent location is between the external occipital protuberance and the foramen magnum. Usually, the sac contains parts of the cerebrum or the cerebellum (Fig. 11.6B).
  - **clinical features**: there is cystic formation of variable size located in the occipital region. The skin can be intact or lacerated. The neurological deficit can vary from pyramid signs and epileptic seizures to bulbar symptoms and hypertensive-hydrocephalic syndrome.
  - **the diagnosis and treatment** are the same as in case of frontal encephalocele.

Fig. 11.6. Fetuses with encephalocele: **A/** frontal; **B/** occipital

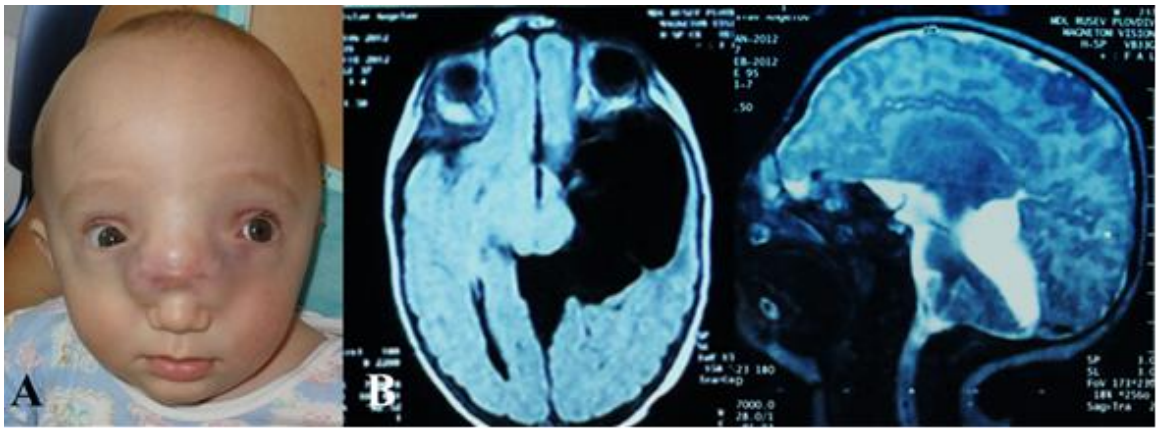

Fig. 11.7. Patient with: **A/** frontal encephalocele and hypertelorism; **B/** MRI image of the same case demonstrates the sac which contains fluid and altered brain tissue

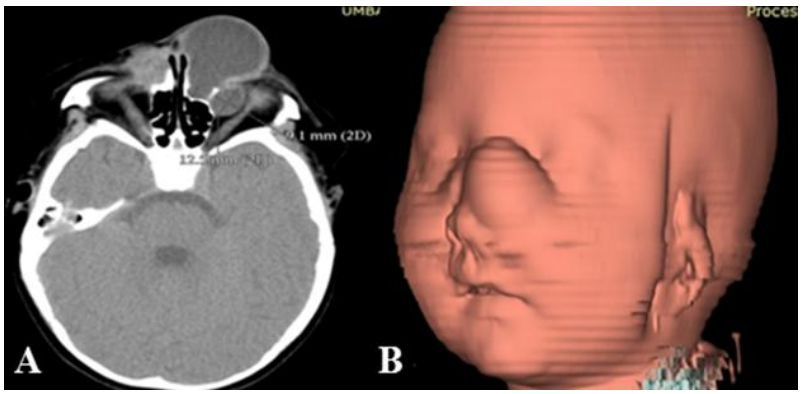

Fig. 11.8. CT of frontal encephalocele: **A/** axial image; **B/** 3D reconstruction

The CNS anomalies of cerebral hemisphere development also fall into this category:

- agenesia of corpus callosum (ACC) (Fig. 11.9);
- congenital cyst of the septum pellucidum ("5th ventricle");
- cavum Vergae ("6th ventricle");
- holoprosencephaly (a defect in the division of the two cerebral hemispheres and formation of one large ventricle) (Fig. 11.10 and Fig. 11.11);
- anencephaly (an absence of cerebral hemispheres) (Fig. 11.12 and Fig. 11.13);
- exencephaly (a prolapse of the brain and meninges though a large skull defect) (Fig. 11.14);
- inencephaly (a combination between anencephaly and rachischisis) (Fig. 11.15).

Holoprosencephaly, anencephaly, exencephaly and inencephaly are not compatible with life and lead to death.

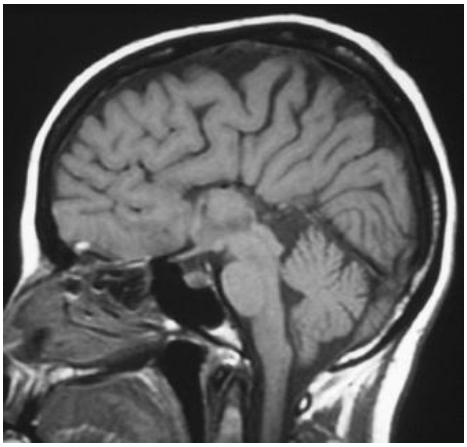

Fig. 11.9. MRI image of agenesia of corpus callosum

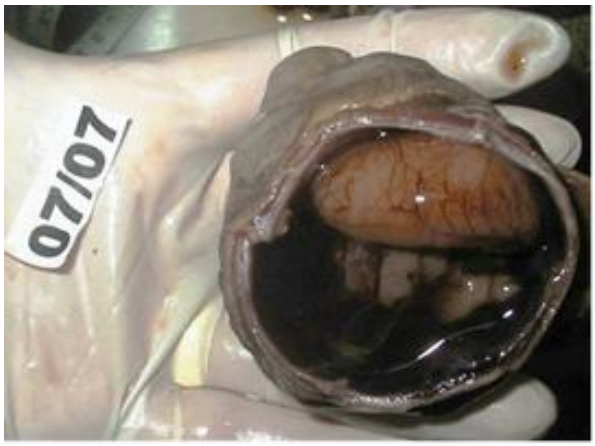

Fig. 11.10. Alobar holoprosencephaly

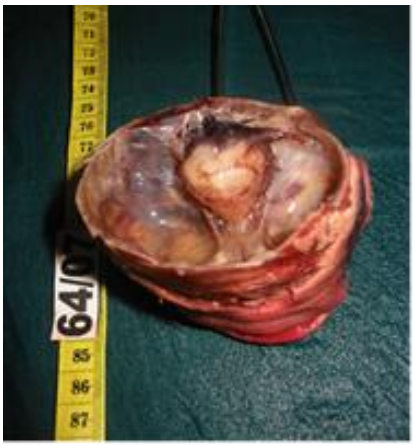

Fig. 11.11. Aprosencephaly

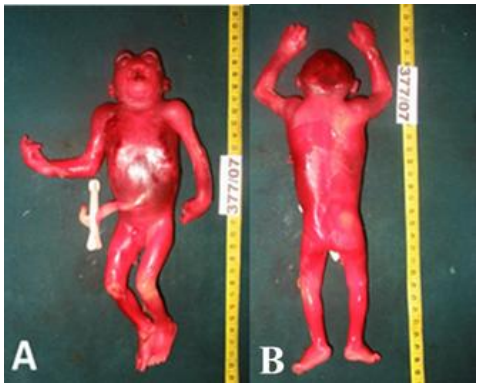

Fig. 11.12. Fetus with anencephaly: **A/**anterior view; **B/** posterior view

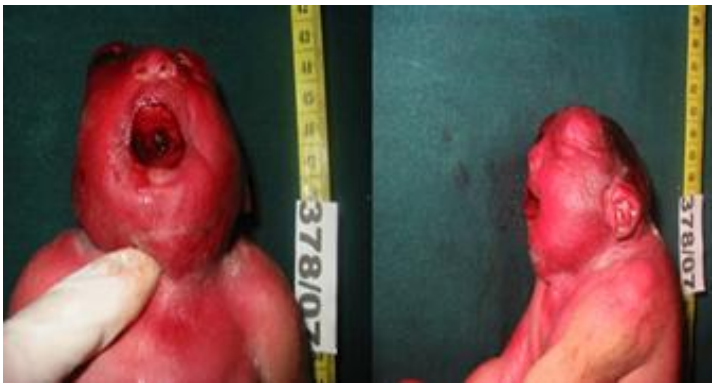

Fig. 11.13. Fetus with anencephaly - AP and lateral view

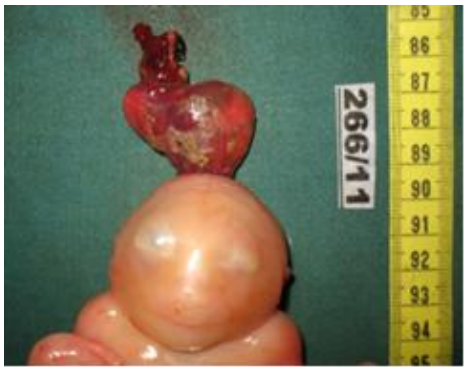

Fig. 11.14. Fetus with exencephaly

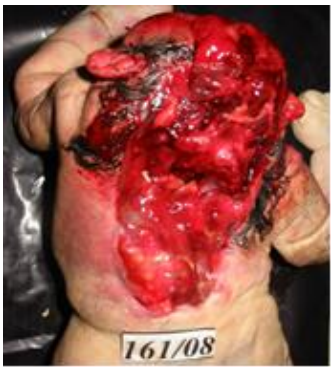

Fig. 11.15. Fetus with inencephaly

Congenital intracranial cysts

Congenital intracranial cysts are expansions of the subarachnoid space combined with agenesia of different parts of the cerebral hemispheres. There are arachnoid and ependymal cysts which depend on the histology obtained from the cystic wall.

1. **Arachnoid cyst** – the walls of the cyst are covered with altered and degenerated arachnoid mater. The usual location of the cyst is in the area of the Sylvian fissure (the lateral sulcus) (Fig. 11.16). The location in the suprasellar area between the two hemispheres is rare. The cyst grows slowly by accumulating CSF based on valve mechanism.

The neurological symptoms results from compression of the adjacent brain structures.

Diagnosis is achieved by CT and MRI.

The surgical intervention is indicated for cases with raised intracranial pressure, focal neurological deficit or epileptic seizures. The aim is to create communication between the cyst and the basal subarachnoid spaces. Sometimes shunting procedure is necessary.

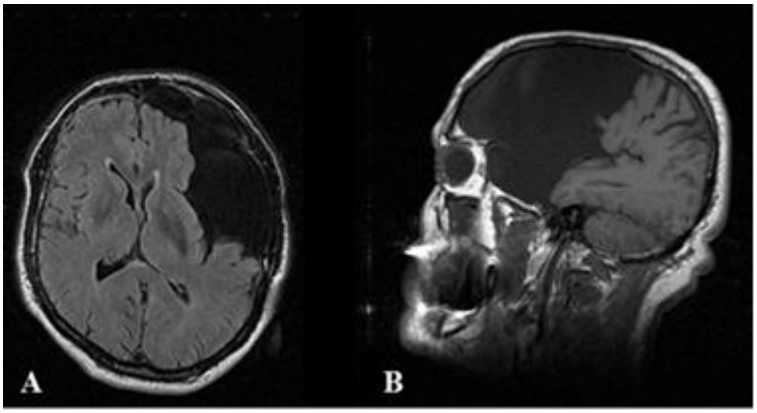

Fig. 11.16. MRI of arachnoid cyst in the left Sylvian fissure:
A/ axial image; **B** /sagittal image

2. **Ependymal cyst** walls are covered with ependyma of embryonic choroid cells. The ependymal cysts are less common and located in the brain parenchyma in close vicinity to the lateral ventricles. Because of their secretory features, these cysts grow rapidly and compress the cerebral structures.

Diagnosis and treatment are the same as those of arachnoid cyst.

3. Dandy-Walker cyst. This type of congenital anomaly includes atresia of the foramina of Magendie and Luschla, cystic dilation of the 4^{th} ventricle and hypoplasia of the cerebellar hemispheres (Fig. 11.17). The malformation is commonly associated with other anomalies such as agenesia of corpus callosum, atresia of the Sylvian aqueduct, etc.

The clinical presentation includes hydrocephalus which results from the obstruction of CSF pathways. Because of the occurrence of hydrocephalus, this malformation is described as Dandy-Walker syndrome.

Diagnosis is based on the modern imaging methods – CT and MRI.

Treatment In case of communication between the cyst and the ventricular system, shunt surgery is sufficient. If there is atresia of the cerebral aqueduct, the cyst itself should be shunted.

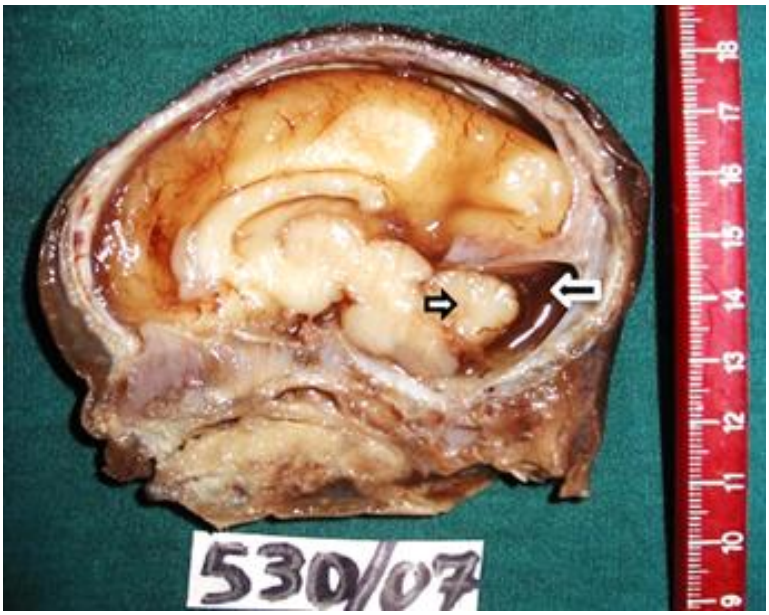

Fig. 11.17. Anatomical specimen of fetus with Dandy-Walker cyst (⇨) cerebellar hypoplasia; (⇨) cyst

Spinal dysraphism

This group of malformations is caused by defects in the normal embryonic closure of the neural tube. The defects in the embryonic development of the fetus can affect all spinal structures – vertebrae, arches, the spinal cord or its meninges, the nerve roots.

Spina Bifida

It is caused by the lack of fusion between the two halves of the arches of one or more vertebrae. The neural structures (spinal cord and its meninges, the nerve roots) protrude through the defect, thus forming a bulge. Usually, the medulla is fixed to the wall of the cyst and forms the so called "medullary zone". The medullary zone could be covered with a layer of epidermis which gradually traverses into the adjacent skin – the so called "dermal zone".

Spina bifida is commonly associated with other malformations of the CNS – hydrocephalus, Arnold-Chiari, malformation, Dandy-Walker syndrome, etc.

Location and incidence – usually, spina bifida occurs in the lumbar region (40%) and the sacral region (30%). The incidence varies from 1-9.5/1000 newborns, predominantly in females.

Pathoanatomy – depending on the extent of neural damage, there are several types of spina bifida:

1. **Spina bifida occulta.** There is malfusion of the arches of 1-2 veretebrae without hernation of neural structures. The skin overlying the defect may appear completely intact. In some cases, there is hyperpigmentation and focal hairy area.
2. **Meningocele** is a hernation of the meninges of the spinal cord through the vertebral defect. There is visible fluctuating sac at back of the baby (Fig. 11.18 and Fig. 11.19).
3. **Myelomeningocele** is a prolapse of all neural structures (meninges, spinal cord and nerve roots). The deformed medulla is seen on the surface as the medullary zone. Frequently, there is large lacerated area (Fig. 11.20, Fig. 11.21 and Fig. 11.22).

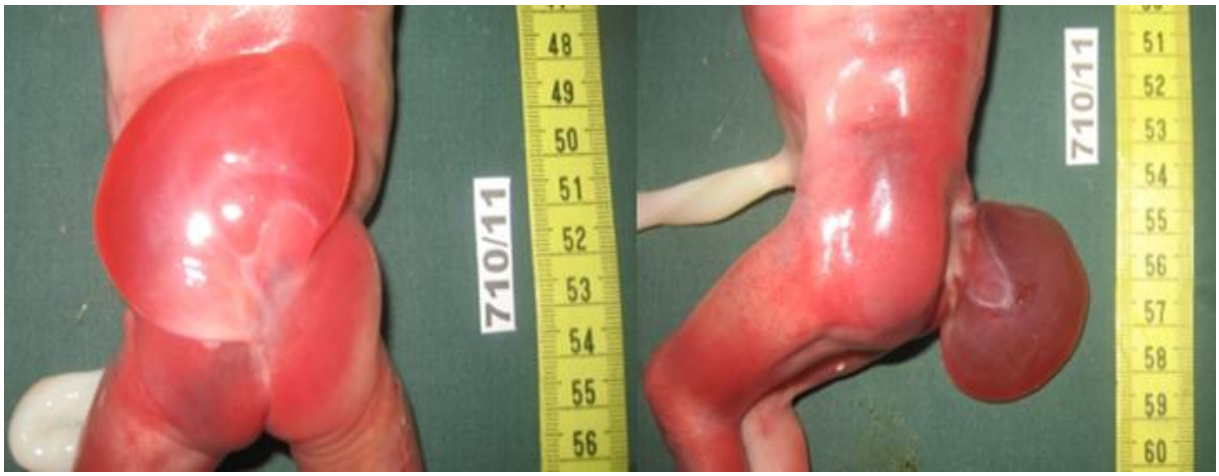

Fig. 11.18. Fetus with lumbo-sacral meningocele

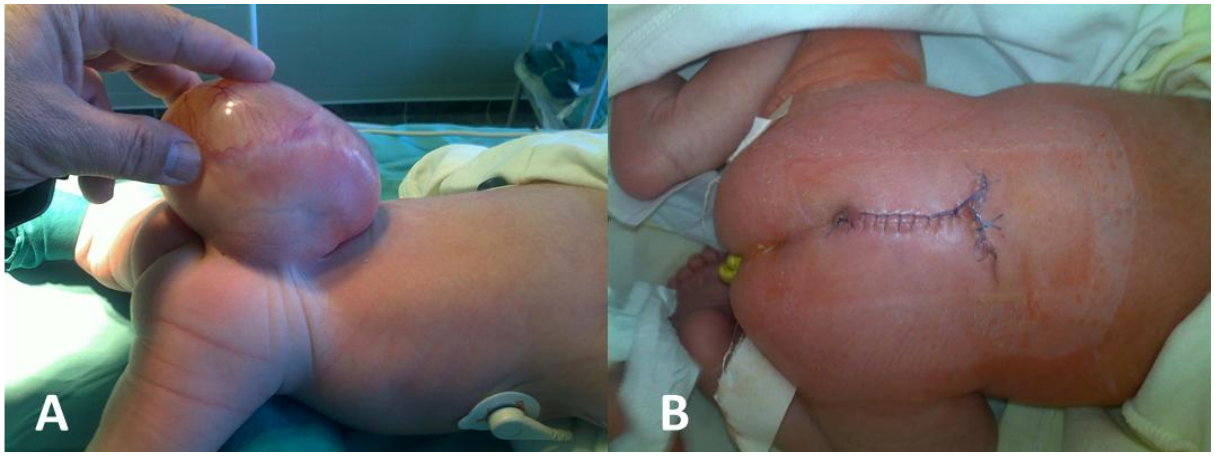

Fig. 11.19. Infant with lumbo-sacral meningocele:
A/ before surgery; **B/** after surgery

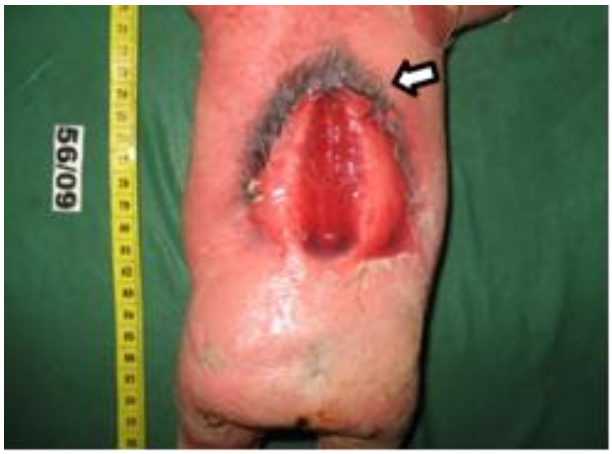

Fig. 11.20. Fetus with thoracolumbar myelomeningocele (➪) abnormal hairiness

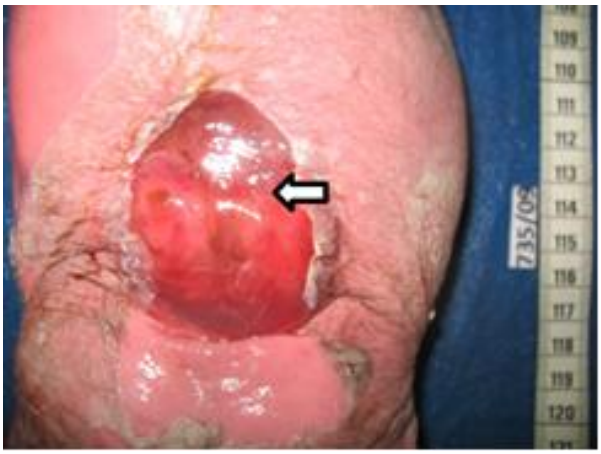

Fig. 11.21. Fetus with lumbosacral myelomeningocele (➪) thin epidermal layer

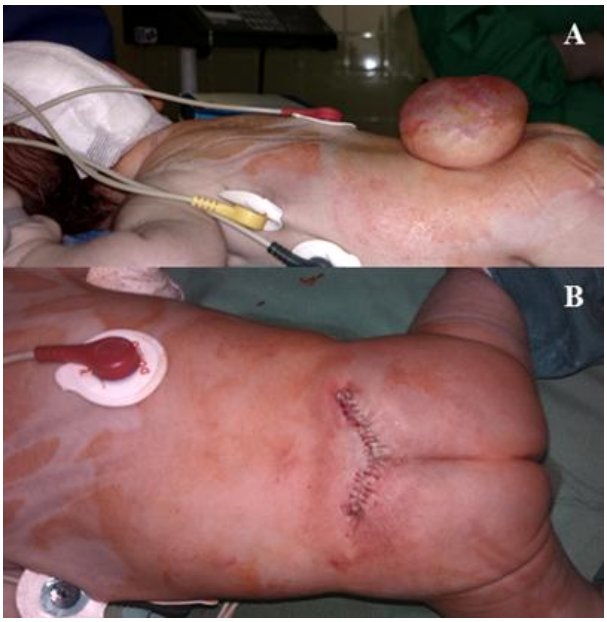

Fig. 11.22. Images of an infant with lumbo-sacral myelomeningocele: **A/** before surgery; **B/** after surgery

4. Rachischisis and craniorachischisis. It is a severe lethal anomaly. A large area of the spinal column is dysraphic with or without herniation of neural structures (Fig. 11.23). In case of craniorachischisis the skull remains opened (Fig. 11.24). In general, this malformation is associated with anencephaly.

5. Lipomeningocele. The meningo-medullary malformation is associated with lipoma which is large and tightly adherent to the nerve roots.

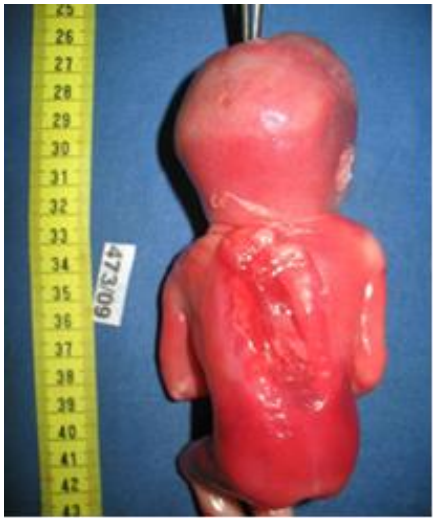

Fig. 11.23. Fetus with rachischisis

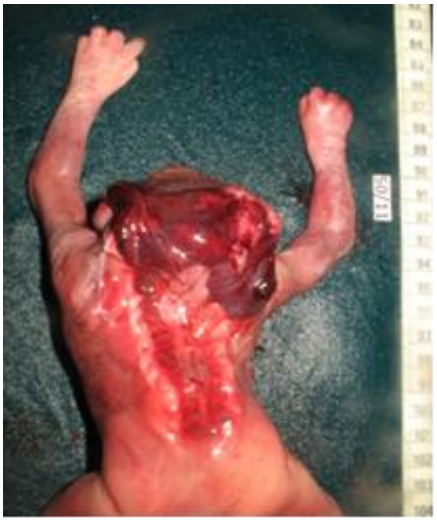

Fig. 11.24. Fetus with craniorachischisis

Clinical features – generally, the spina bifida occulta and the meningocele are not associated with focal neurological deficit and mental retardation. 70-80% of the cases with myelomeningocele and lipomeningocele manifest with radicular motor, sensory and/or sphincter disorders that predispose to urinary infections. The skin overlying the malformation is hyperpigmented with pathological hairiness. The feet are hypotrophic and deformed.

The meningocele is associated with certain complications:

- ***Infection (meningitis) – the skin overlying the malformation is thinned and cannot adequately protect the individual against invasion of bacterial agents.***
- ***Hydrocephalus often develops as a result of impaired CSF circulation.***

Diagnosis

1. **Spondylography** demonstrates vertebral arch defect, expansion of the spinal canal, enlarged interpedicular distance in the area of the malformation and spinal deformity.
2. **CT and MRI** provide information about the presence of herniated neural structures, lipoma, teratoma etc.

Treatment: The aim of the surgical intervention is to restore the normal relationship of the neural structures and to correct the skin defect. Children in stable general status, preserved neurological function and correctable defects are indicated for surgical treatment.

Diastematomyelia

It is a type of spinal dysraphism in which the spinal canal is divided into two halves by a bony or cartilaginous septum originating from the body of the vertebra. The usual location of the malformation is between Th_4 and L_4 level (Fig. 11.25 and Fig. 11.26).

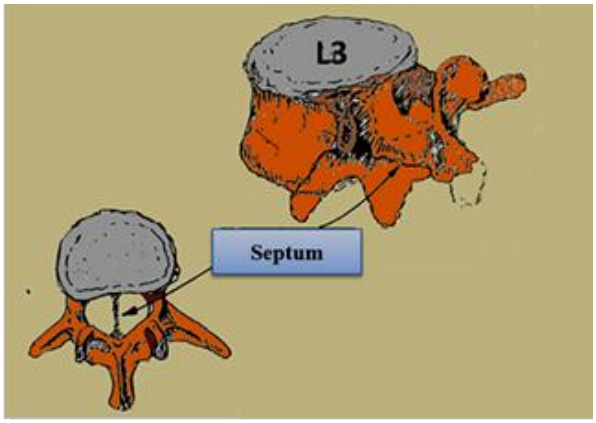

Fig. 11.25. Scheme of diastematomyelia at L_3 level

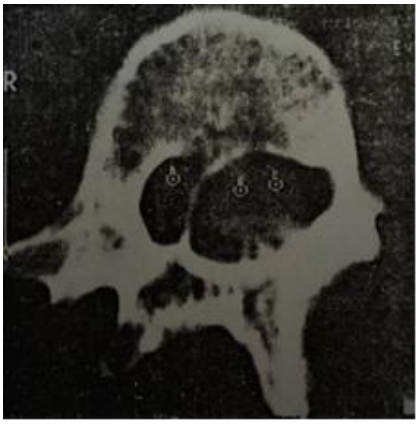

Fig. 11.26. CT of diastematomielia at L_3 level

The pathoanatomal features include widening of the spinal canal, malfusion of several vertebral arches; the spinal cord is divided into two halves that have separate duro-arachnoid sacs.

The clinical symptoms result from the extension of the spinal cord. Initially, there are sphincter disorders followed by gradual development of motor deficit (paraparesis). The disease follows slow but progressive evolution.

Diagnosis is achieved by X-rays, CT and MRI.

The surgical intervention aims at removing the abnormal bony and dural septum, thus restoring the dural cover of the spinal cord.

Tethered cord syndrome

This is an abnormal fixation of the filum terminale to the lower parts of the spinal canal during the embryonal development. This impedes the spinal cord ascent to its usual level (L_1-L_2) during ontogenesis. This malformation is frequently associated with spina bifida and presents with similar symptoms: hypertrichosis, hyperpigmentation of the skin, deformation of the feet, sphincter disorders, radiculalgia, mild motor and/or sensory deficit. The disease has a slow progression.

Diagnosis includes X-rays, CT and MRI.

The purpose of surgery is to cut the filum terminale in order to release the spinal cord.

Congenital dermal sinus

It is an ectodermal invagination which connects the neural structures with the overlying skin surface (Fig. 11.27). The most common location is the lumbar or occipital region.

Fig. 11.27. Intraoperative view of dermal sinus (➪)

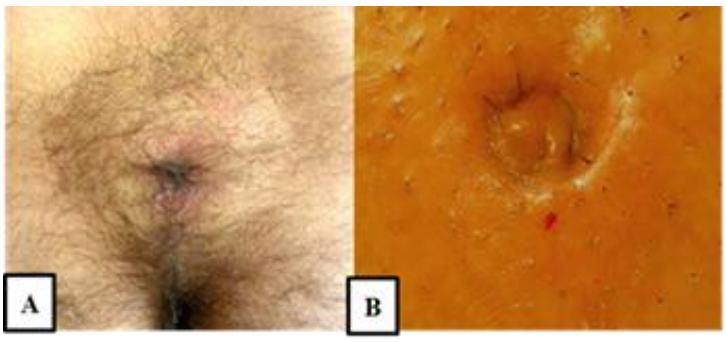

Fig. 11.28. Congenital dermal sinus: **A/** hypertrichosis; **B/** invagination

Pathoanatomy – usually, the dermal sinus is covered with squamous epithelium and connects the dermal structures with a dermoid cyst located amongst the nerve roots of cauda equina. Occasionally, congenital dermal sinus is associated with embryonic tumors (teratoma) (Fig. 11.31). Vertebral arches remain opened at the level of the malformation.

Clinical features. Frequently, the area of the dermal invagination is with hypertrichosis (Fig. 11.28). Because of the direct communication with the surface of the skin, infectious complications are common. The dermoid cyst compresses the nerve roots and can cause radiculalgia, radiculopathy or cauda equina syndrome.

The diagnosis is based on the clinical symptoms, CT and MRI findings (Fig 11.29 and Fig. 11.30).

The surgical treatment aims to remove the dermoid cyst, its communication with the skin and any concurrent tumor (Фиг. 11.31).

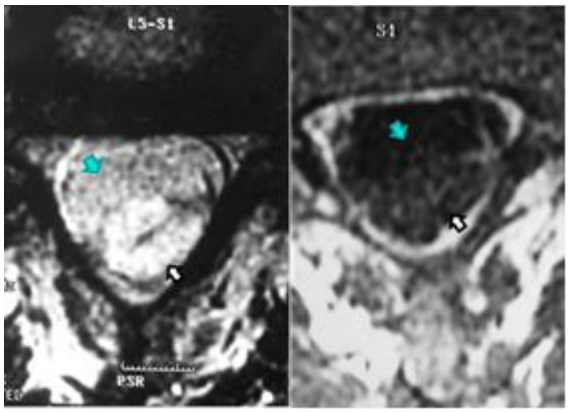

Fig. 11.29. MRI of congenital dermal sinus – axial image at L_5 – S_1 level (➪) tumor; (➧) cystic formation

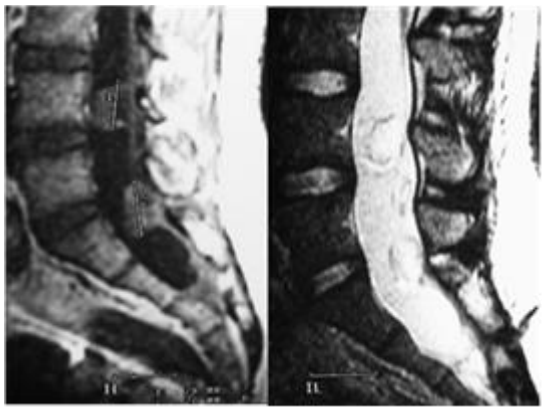

Fig. 11.30. MRI of congenital dermal sinus with dermoid cyst at L_4–S_2 level sagittal T1 and T2 images

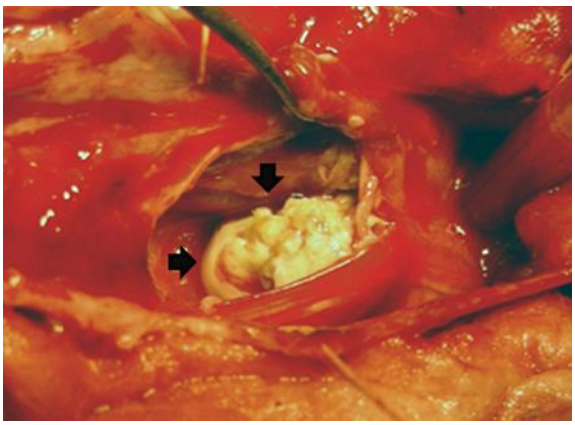

Fig. 11.31. Intraoperative view of teratoma located inside the cyst (➧)

Arnold-Chiari Malformation

It consists of downward displacement of the lower parts of the cerebellum (the lower vermis and the cerebellar tonsils) and, occasionally, the inferior portion of medulla oblongata, below the level of foramen magnum. The malformation is associated with hydrocephalus and spina bifida. There are three forms:

- **Arnold-Chiari I** is herniation of the tonsils below the level of foramen magnum in the absence of spinal dysraphism and hydrocephalus (Fig. 11.32A and Fig. 11.33);
- **Arnold-Chiari II** is herniation of the tonsils and the lower vermis below the level of foramen magnum. There is deformation of the 4th ventricle and the lower part of brainstem associated with hydrocephalus and dysraphism (spina bifida) (Fig. 11.32B);
- **Arnold-Chiari III** is characterized by extensive dysraphism of the upper cervical spine through which large part of the cerebellum prolapses. Hydrocephalus and syringomyelia are present.

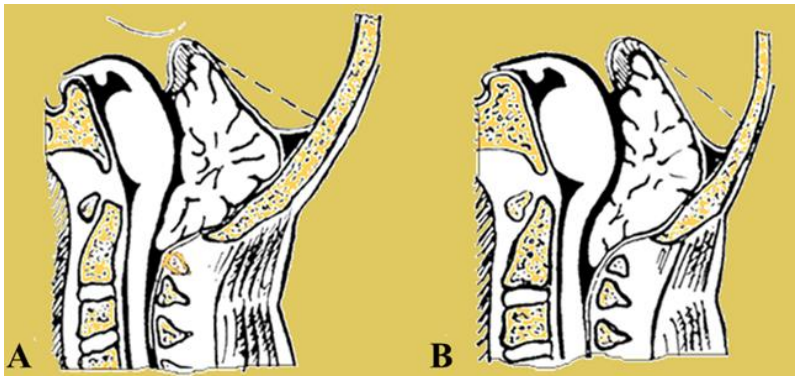

Fig. 11.32. Schematic drawing of Arnold – Chiari malformation:
A/ Type I; B/ Type II

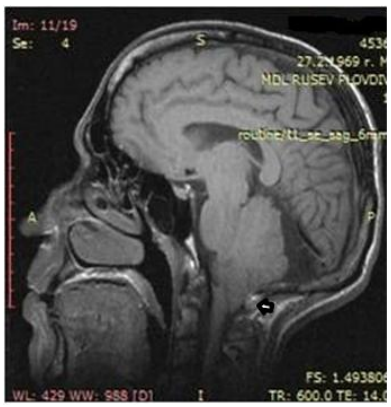

Fig. 11.33. MRI image of Arnold-Chiari malformation type I

Clinical features. The onset of the disease is manifested with restricted movements and pain in the neck, dizziness and vomiting, occasionally associated with torticollis. At advanced stage, cerebellar, pyramid and bulbar symptoms can be added.

The diagnosis is based on the clinical symptoms, CT and MRI findings.

The surgical intervention aims to achieve bony, dural and parenchymal decompression of the posterior cranial fossa and the upper cervical spine.

Syringomyelia

This is a malformation with a progressive course which is characterized by the formation of intramedullary cyst filled with fluid. The intramedullary cavity often spreads across several spinal cord segments. It is usually located in the cervical region (the cervical intumescence) or medulla oblongata (syringobulbia). Occasionally, the malformation is located in the thoracic region. The syrinx is located in the gray matter of the spinal cord behind the central canal. The spinal cord is widened and has fusiform shape (Fig. 11.34 and 11.35). The syringomyelia is frequently associated with the malformations of Arnold-Chiari, Dandy-Walker, spina bifida, etc.

Pathogenesis

1. **Primary form –** few theories exist:
   - dysraphic theory – a defect in the closure of the neural tube;
   - hydrodynamic theories – defects in the foramina of Magendie and Luchka which lead to an impaired CSF outflow from the 4^{th} ventricle and affect the cerebellum and the brainstem;
   - Abulker's theory – CSF circulation at the level of the cerebellomedullary cistern is impaired and the fluid infiltrates the parenchyma of the spinal cord.
2. **Secondary form –** this form is rare and results from:
   - spinal cord injury;
   - intramedullary hemorrhage or ischemia;
   - postinflammatory complications of the spinal cord.

Clinical presentation The evolution of the disease is insidious and presents with typical symptoms such as cervicobrachialgia, specific sensory loss (anesthesia for pain and temperature and preserved sensation to touch and vibration), muscular atrophy in the

distal portions of the upper limbs. At later stages, conductive motor and sensory impairment, affecting the lower limb, can be observed. Caudal cranial nerve palsies can develop in case of syringobulbia.

Diagnosis by MRI is the contemporary procedure of choice. It provides detailed information about the location, size and type of the syrinx.

The treatment is surgical and indicated for cases with progressive neurological deficit. The type of the surgical intervention depends on the presence or absence of associated CNS malformations. Generally, the surgical intervention aims to drain the syrinx towards the pleural cavity, the peritoneal cavity or the subarachnoid space.

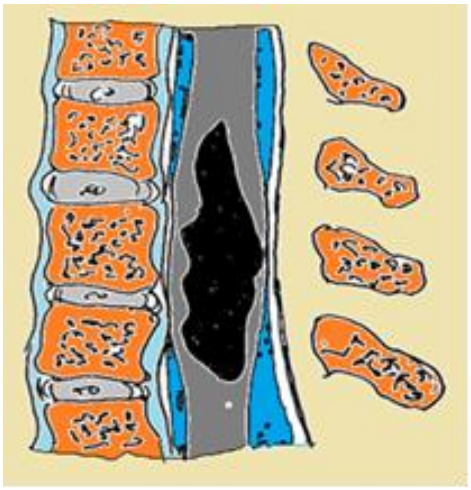

Fig. 11.34. Schematic drawing of syrinx

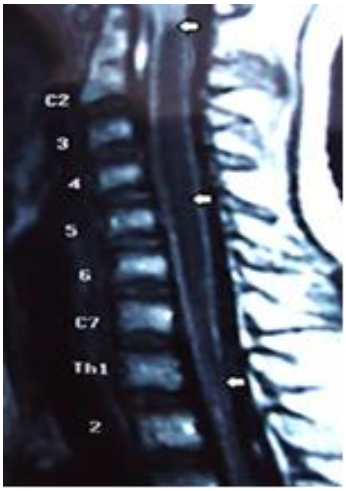

Fig. 11.35. MRI of cyringomyelia affecting C_1–Th_1 spinal segment

The contemporary management: CNS anomalies put an emphasis on prenatal diagnosis and prevention. For example, the increased maternal and amniotic levels of α-fetoproteins after the 14th gestational week indicate presence of spina bifida in 70-90% of the cases. Another prognostic marker is the presence of acetylcholinesterase in the amniotic fluid. Therefore, the fetal sonographic investigation is of paramount importance for the detection of these malformations. Currently, the interruption of the pregnancy due to medical reasons after patient's informed consent is the most secure method of management of CNS anomalies, bearing in mind the great social and personal burden that parents have to face.

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CHAPTER 12 *T. Kitova, B. Kitov, E. Hristov*

SURGICAL TREATMENT OF PAIN

The aim of this field of neurosurgery is to influence specific neurophysiological functions by destruction of certain zones and nuclei or by interruption of different conduction systems.

The pain is a warning signal sent to the brain from different parts of the human body, indicating that these parts are being exposed to pathological stimulation. Perception of pain is subjective and varies between individuals. It is dependent upon the individual excitatory threshold.

Anatomophysiology and biochemistry of pain

1. There are no specific stimulants that cause pain. All intensive, repeated stimuli (mechanical, thermal or chemical) can cause pain.

2. There are three types of somatosensory receptors – mechanoreceptors (transmit impulses of touch, compression, vibration and position of the different parts of the body); nociceptors (transmit pain impulses) and thermoreceptors (transmit impulses about temperature changes). The effects on the terminal receptors in the epidermis and dermis cause the so-called "primary pain". The painful stimulus leads to a local release of great amount of biochemical substances such as kinins, prostaglandins, histamine, serotonin, etc. These substances affect the terminal receptors, thus causing secondary pain.

3. The nociceptive impulse originating from the terminal receptors is transmitted to the spinal cord by neurofibers with small caliber (myelinated A-delta and amyelinated C). The myelinated A-beta fibers transmit proprioceptive and epicritic signals. Classification of neurofibers is based on the thickness of the fiber and the speed of impulse conduction (Table 12.1). The differences in the size and the conduction speed between thick fibers (for touch) and thinner fibers (for pain) allow selective stimulation of one of these groups. It is a specific phenomenon that underlies transcutaneous electrostimulation of peripheral nerves – a method of relieving chronic pain. Current studies indicate that there is some difference between the so-called slow and fast pain. Fast pain (pin-prick) is conducted by phylogenetically new pathway – the neospinothalamic tract whereas slow pain is conducted by phylogenetically old pathway - paleospinothalamic tract (or the spino-reticulo-thalamic tract).

Table 12.1. Classification of sensory fibers

Classification	Myelinization	Diameter (Microns)	Conduction speed (m/s)	Type of sensory information
I A alfa	Yes	12 - 20	75 - 120	Position of limbs
II A beta	Yes	6 - 12	30 - 75	Touch, pressure and vibration
III A delta	Yes	1 - 6	5 - 30	Fast pain and cold
IV C	No	< 1,5	0,5 - 2	Slow pain and cold

4. The first neurons conducting pain sensation enter the spinal cord through the most lateral parts of the dorsal nerve roots forming the dorso-lateral fasciculus or the Lissauer's tract.

This anatomical feature underlies the neurosurgical procedure known as DREZ (Dorsal Root Entry Zone) rhizotomy, which is performed in cases with chronic cancerogenic pain and pain caused by extraction of spinal nerve roots.

5. From the dorsal horn of the spinal cord, the A-delta and C fibers cross the midline in the white matter at the level of the anterior commissure and connect with the neurons of the spino-reticulo-thalamic tract, which is situated in the ventro-lateral quadrant of the medulla and ends in the reticular formation and the thalamus as the medial lemniscus, responsible for perception of non-localized pain (Fig. 12.1 and Fig. 12.2).

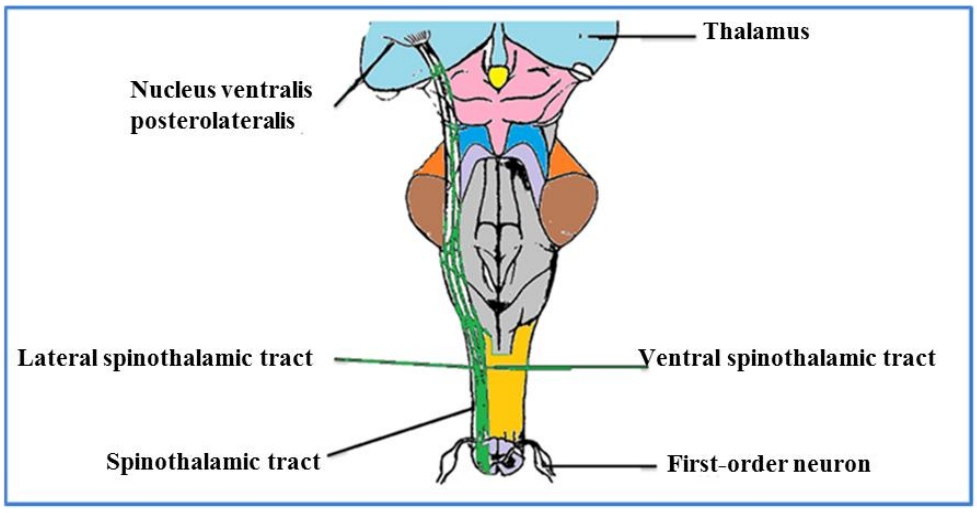

Fig. 12.1. Epicritic sensation pathways

The position of the second neuron of the epicritic sensation in the spinal medulla underlies the neurosurgical procedures called commissurotomy (interrupts segmental pain conduction) and antero-lateral chordotomy (blocks pain perception below the level of the intervention).

The tracts of the epicritic and the proprioreceptive sensation reach the brain stem through the tracts of Goll and Burdach. which further form the medial lemniscus that ends in the thalamus (the nucleus for specific sensation). Along the thalamic radiations the information reaches the somatosensory area in the parietal cortex (Fig. 12.2).

6. Phenomena of control over the pain transmission: the synaptic connection between the A-delta neurons and C neurons of the spino-thalamic path is inhibited by special interneurons located in the gelatinous substance of the dorsal horn of the spinal cord that control the neurons transmitting secondary pain by inhibition of their excitement and stop pain conduction before reaching the relevant brain center. A group of neurons located in the ventral part of the mesencephalon and the diencephalon (analgesia center) provoke reduction of pain sensation. They connect with the reticular formation in the brainstem and by the reticulo-spinal tract reach the neurons located in gelatinous substance that inhibit the ascending pain impulses.

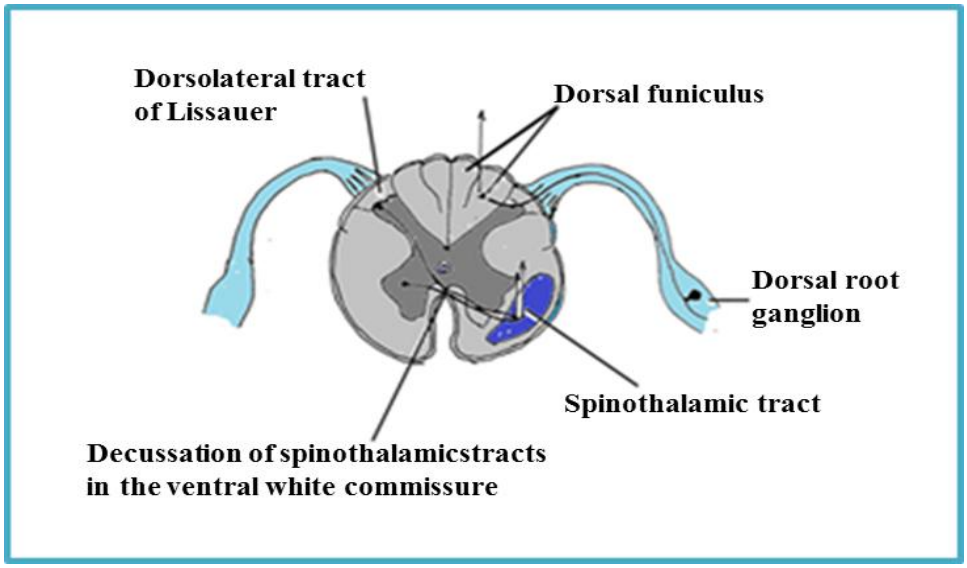

Fig. 12.2. Transverse section of the spinal cord showing epicritic and proprioceptive sensation pathways

This mechanism of inhibition of the ascending pain impulses is utilized during the implantation of electro-stimulating electrodes in the so-called analgesia center in order to relieve chronic pain.

Mechanism of pain perception.

Generally, the perception of pain is due to:

- excessive nociceptive stimuli (hyperstimulation of the terminal receptors caused by neoplasms, inflammatory processes, traumas, etc.);
- reduction or loss of inhibitors of pain conduction.

Drug treatment of pain aims at reducing the sensitivity of the peripheral receptors or increasing the threshold of pain perception. Surgical treatment is indicated for patients with intractable pain due to structural lesions that cannot be controlled by the help of drugs. The choice of appropriate surgical treatment depends on:

- the etiology of pain – cancerous or non-cancerous pain;
- type of the pain:
  - neuropathic (excruciating pain in the area of neurological deficit);
  - somatic (nociceptive) – in the case of tissue or organ damage.
- duration of pain – acute or chronic.

Surgical interventions

- **stimulating procedures –** the nerve fibers are stimulated with variable impulses which induces inhibition of the nociceptive perceptions. The different types of surgical procedures are shown on Table 12.2;

Table 12.2. Types of surgical procedures for treatment of chronic pain

Peripheral nerves	Spinal cord	Brain
Neurotomy	Chordotomy	Mesencephalotomy
DREZ rhizotomy	Median myelotomy	Brain stimulation
Ganglionectomy	Spinal stimulation	Hypophysectomy
Transcutaneous electrostimulation	Intrathecal morphine infusion	Intraventricular infusion of morphine

- **destructive procedures –** these procedures have temporary success (1-2 years) and are usually performed in case of acute or cancerous pain when the expected survival period is short.

Etiology of pain syndromes

1. **Cancerous pain –** depending on the location of the neoplasm the most frequent are:
   - **pain caused by cervical and head carcinoma** - the neoplastic process can induce intractable pain in the regions innervated by the trigeminal and the glossopharyngeal nerve. When the tumor penetrates the cervix the pain is caused by irritation of the upper sensory (dorsal) cervical nerve roots;
   - **pain caused by thoracic and abdominal cancer -** the thoraco-mediastinal cancer can cause pain syndromes due to involvement of the thoracic vertebrae or direct invasion of the brachial plexus. The intra-abdominal neoplasms cause pain by irritation of the lumbosacral plexus and peripheral nerves;
   - **pain caused by pelvic cancer -** due to irritation of the peripheral nerves in the pelvic region.
2. **Chronic non-cancerous pain.** Apart from cancerous pain, there are other pain syndromes with variable etiology. Surgical treatment is indicated for the cases with established pathophysiological mechanisms.
3. **Peripheral nerve pain syndromes**
   - **anesthesia dolorosa.** Persistent pain in the region of the anesthesia due to an injury of a peripheral nerve that has an undefined character. It is treated with electro-stimulation;
   - **phantom pain**. Sensations are described as perceptions that an individual experiences relating to a limb or an organ that is not physically part of the body. Sensations are recorded most frequently following the amputation of an arm or a leg, but may also occur following the removal of a breast or an internal organ. Phantom limb pain is the feeling of pain in an absent limb or a portion of a limb. It intensifies during emotional stress;
   - **causalgic pain.** Occurs in patients that have sustained traumatic injury to peripheral nerves that contain numerous automatic nerve fibers (the ulnar nerve, the median nerve, the

sciatic nerve). The pain is of burning character accompanied by vasomotor and trophic disturbance in the nerve distribution area. The pain sensations begin shortly after the trauma. Usually, the sensory and motor functions are preserved.

Trigeminal neuralgia

This disorder is characterized by episodes of intense facial pain that last from a few seconds to several minutes or hours. The episodes of intense pain may occur paroxysmally in the distribution area of one or more branches of the trigeminal nerve. To describe the pain sensation, patients may describe a trigger area on the face so sensitive that touching or even air currents can trigger an episode; however, in many patients the pain is generated spontaneously without any apparent stimulation.

Etiopathogenesis. It is considered that the trigeminal neuralgia is due to compression of the trigeminal nerve entry zone into the brainstem caused by an aberrant arterial loop or adjacent venous vessel that causes chronic irritation and subsequent demyelinization of the axons. The condition is more frequent after the age of 60 with male:female ratio 2:1. The right half of the face is more often affected than the left half (3:2). In 5-10% of the cases, the cause of this condition is a tumor or vascular malformation, and in 2-3% - multiple sclerosis.

Clinical features. Acute burning pain is present along the distribution area of the branches of the trigeminal nerve (most frequently the maxillary and the mandibular nerve). The duration of pain varies from seconds to minutes. During the interictal periods it is absent or mild. It can be triggered by temperature changes, chewing, speech or touch of the cutaneous distribution area of the nerve in which sensory deficit may be present. If there is constant pain with pronounced sensory deficit, it is compulsory to rule out tumor or blood vessel anomaly in the area of the posterior cranial fossa.

Treatment

A. **Conservative treatment** starts with drug therapy (Carbamazepin or Phenytoin) which can be ineffective.
B. **Surgical treatment** is performed in cases of allergy to the medications, lack of therapeutic effect and intractable pain syndrome. The surgical procedures include:

- **percutaneous trigeminal rhizotomy** (by glycerol rhizolysis or radiofrequency thermoablation of the Gasserian ganglion) – preferred for elderly patients.
- **microvascular decompression (Janetta procedure) –** in cases with proven neuro-vascular conflict (by MRI), the aberrant vessel that compresses the trigeminal nerve is dissected free off the nerve and displaced by microsurgical technique. Then the nerve is isolated from the aberrant vessel by a teflon sponge (Fig. 12.1).

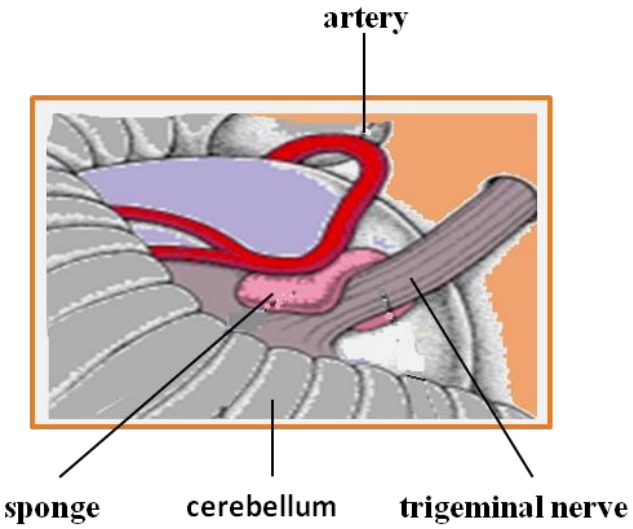

Fig. 12.1. Schematic drawing of the Janetta procedure

- **partial retrograde rhizotomy –** if there is absence of vascular compression of the trigeminal nerve, the sensory branch is cut while the motor fibers are preserved.
- **injection of absolute alcohol in the branches of the trigeminal nerve (supraorbital and infraorbital nerves) –** the effect of these procedures is insignificant and temporary.

Glossopharyngeal neuralgia

The glossopharyngeal neuralgia is similar to the trigeminal neuralgia but the pain affects the area of the posterior portion of the tongue and the lateral aspect of the pharynx.

Treatment: Microvascular decompression of IX and X cranial nerves is performed. The compression of the nerve is usually caused by ectopic vertebral artery or the aberrant loop of the posterior inferior cerebellar artery.

Neuralgia of the greater occipital nerve

The occipital neuralgia is manifested by unilateral pain in the occipital region, limited movements of the neck, dizziness and visual disorders. The condition usually occurs after cervical trauma without any evident roentgenographic changes.

Treatment includes microsurgical decompression of C_2 and C_3 nerve roots and spinal ganglions.

Types of neurosurgical procedures for pain relief

1. Intraspinal or intraventricular injection of morphine or other strong analgesics. The advantage of these manipulations is that the pain control can be achieved with lower drug dosages. They include the implantation of intraventricular or intraspinal (epidural or subarachnoid) catheter which is connected to a subcutaneous reservoir that can be refilled and used for a long period of time (Ommaya, Infusid, Medtronic, Spinalgesic). Absolute alcohol (3-4 ml) may be injected by lumbar puncture in the subarachnoid space of cauda equina.

The intraventricular injection is indicated for patients with neoplasms of the head and the neck, as well as in the case of diffuse pain. The intraspinal injection is indicated for pain localized in the legs and lesser pelvis in patients with paraplegia, bowel and bladder disturbances.

2. Chordotomy includes interruption of the lateral spino-thalamic tract performed by:

- percutaneous radiofrequency thermocoagulation (after lateral C_1-C_2 puncture);
- after laminectomy at Th_{2-4} levels and anatomical interruption of the tract.

Chordotomy is indicated for unilateral cancerous pain.

3. Commissural myelotomy includes laminectomy at Th_{9-10} levels and median myelotomy (in depth of 5-6mm).

It is indicated for bilateral pain syndrome below the cervical region.

4. **Mesencephalic tractotomy (thalamotomy) –** stereotactic radiofrequency thermocoagulation is performed by insertion of special electrode 5mm postero-inferior to the cerebral aqueduct and 9mm lateral from the midline. Mesencephalic tractotomy aims at destruction of the centromedian nucleus, which is part of the pathway of unlocalized pain.

It is indicated for patients suffering from head or neck cancer who have sustained unsuccessful trigeminal rhizotomy.

5. **DREZ (Dorsal Root Entry Zone) rhizotomy** includes destruction of the Lissauer's tract (the posterolateral tract) achieved by thermocoagulation, laser, bipolar coagulation or sharp incision.

It is indicated for chronic pain syndrome following traumatic injury to the brachial and lumbosacral plexus, phantom pain, postherpetic neuralgia, etc.

6. Spinal cord stimulation includes electrode implantation by percutaneous or open surgery techniques.

It is indicated for postherpetic neuralgia, posttraumatic phantom pain, etc.

7. Deep brain stimulation of the analgesia center which inhibits pain sensations.

It is a terminal procedure for patients with intractable pain (phantom pain, cancerous pain, brachial plexus avulsion, etc.

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CHAPTER 13 *I. Kehayov, T. Kitova, B. Kitov, K. Kilova*

SURGICAL TREATMENT OF EPILEPSY

Epilepsy is a disease known since antiquity. It is manifested by paroxysmal and recurrent seizures which represent as self-limited change in behaviour associated with excessive electrical discharge from the CNS. There are many types of seizures with predominant motor, sensory or psychic disorders. Epilepsy is defined as a condition of recurrent seizures and medical intractability as recurrent seizures persist despite optimal treatment under the direction of an experienced neurologist over a two to three year period. The incidence varies between 0.5-1% of the population. In the majority of patients with epilepsy, seizures can be well controlled with appropriate medication. However, current estimates indicate that 20-30% of patients with epilepsy are refractory to all forms of medical therapy. These medically intractable patients are candidates for surgical treatment in an attempt to achieve better seizure control. Another group of patients who might benefit are those whose seizures may be relatively well controlled but who have certain characteristic presentations or lesions that strongly suggest surgical intervention might be curative. Overall, the single most important determinant of a successful surgical outcome is patient selection. This requires detailed presurgical evaluation to characterize seizure type, frequency, site of onset, psychosocial functioning and degree of disability in order to select the most appropriate treatment from a variety of surgical options. This type of evaluation is best carried out at a multi-disciplinary center experienced in the investigation and treatment of epilepsy.

Classification of epileptic seizures

1. **Generalized seizures (there is loss of consciousness)**

1.1. Absence (petit-mal):

- only with change in consciousness;
- with mild clonic manifestations;
- with atonic manifestations;
- with tonic manifestations;
- with automated movements;
- with autonomic manifestations.

1.2. Atypical absence;
1.3. Myoclonic seizures;
1.4. Tonic seizures;
1.5. Clonic seizures;
1.6. Tonic-clonic seizures (grand-mal);
1.7. Tonic seizures.

2. Partial (focal) seizures

2.1. Simple partial seizures without change in consciousness:

- with motor manifestations;
- with sensory manifestations;
- with autonomic manifestations;
- with psychic manifestations.

2.2. Complex partial seizures with change in consciousness
2.3. Partial seizures with secondary generalization

3. Unclassified epileptic seizures

4. Epileptic status – constantly recurrent seizures without lucid periods which continue for more than 20-30 minutes.

Patients with medically intractable epilepsy require thorough examination and selection of appropriate cases amenable to surgical treatment.

Presurgical evaluation

The goal of epilepsy surgery is to identify an abnormal area of cortex from which the seizures originate and remove it without causing any significant functional impairment. The primary components of the presurgical evaluation include a detailed clinical history and physical examination, advanced neuro-imaging, video-EEG monitoring, neuropsychological testing and assessment of psychosocial functioning. The major surgical questions one hopes to answer with this evaluation are: 1) are the seizures focal or generalized?; 2) if focal, are they temporal or extratemporal in origin?; 3) is there a lesion associated with the seizures? and 4) if surgery is undertaken what functional deficits, if any, might be anticipated?

Clinical Features

The presurgical evaluation of a patient with medically intractable epilepsy begins with a complete history and physical examination. One attempts to classify the different kinds of seizures as well as the

frequency, severity and duration of each type. The clinical semiology of these events can yield important localizing information to the experienced clinician. It is also important to determine the age of onset, response to treatment and familial tendency to seizures. The pregnancy and delivery history is helpful in assessing congenital or early acquired abnormalities. Other past medical history of significance would include a history of febrile seizures, head injury or intracranial infection. An assessment of the adequacy of medication trials must also be made to ensure that the patient is truly refractory to medical therapy.

On examination, the clinician looks for obvious asymmetries of development compatible with an early structural CNS lesion and focal neurologic or cognitive abnormalities suggestive of acquired disease. In the great majority of patients, however, the neurological examination is completely normal.

Neuro-imaging

Modern neuroimaging is crucial to surgical decision making. In the past, skull X-rays, ventriculograms, pneumoencephalography and computerized tomography (CT) scans demonstrated indirect evidence of cerebral pathology in the form of focal or diffuse atrophy or space-occupying lesions. Recently, magnetic resonance imaging (MRI) has replaced CT scanning as the imaging study of choice to evaluate patients with epilepsy. MRI is an extremely sensitive tool that can detect abnormalities of the brain with exceptional anatomical detail. This has been especially true for detecting focal atrophy (e.g. hippocampal atrophy), indolent gliomas, cortical dysplasias, cerebral gliosis and small structural lesions of the neocortex. Functional imaging attempts to visualize alterations in cerebral metabolism using Positron Emission Tomography (PET) and Single Photon Emission Computerized Tomography (SPECT). These studies reveal epileptic areas as hypometabolic between seizures and hypermetabolic during seizures. Although they lack the spatial resolution of MRI, PET and SPECT can play an important role in the localization of abnormal cortex. Ictal SPECT studies can be obtained if injection of an appropriate radioisotope is performed within seconds of a seizure onset. The isotope is concentrated in the region of seizure onset and imaging studies can be obtained up to several hours after injection to

demonstrate the area of ictal onset. These studies have been useful in many patients with occult epileptic foci.

Electroencephalographic (EEG) Investigation

Electroencephalographic (EEG) investigation remains the most important aspect of the presurgical evaluation. Analysis of unselected EEG activity between events (interictal) or of specific activity during events (ictal) can provide evidence of focal electrical dysfunction. While certain interictal EEG abnormalities (spike and slow wave complexes) can be of localizing value, it is considered extremely important to record the EEG with concommitant videotape during the spontaneous occurrence of the patient's events. Video/EEG monitoring can continuously record the EEG over a 24 hour period which allows for careful inspection of the record during any symptomatic event. Sophisticated computer hardware and software also allow for automatic detection of spontaneous interictal epileptiform transients and electrographic seizures that otherwise might have gone unrecognized. It is the EEG activity at the very beginning of the seizure before spread to adjacent areas that is most important in terms of localization, and if a specific cortical area is involved consistently at the onset then that area is likely to be the site of seizure origin. Patients are often hospitalized with reduction in anti-seizure medications and may be recorded for up to 7-14 days in order to capture 3-5 of their habitual seizures.

Neuropsychological Testing

Detailed neuropsychological testing is carried out to reveal specific focal or multifocal cognitive deficits that might be correlated with the neuroimaging and EEG. This testing may help in localizing an abnormal area of the brain but also serves as a comparison for post-surgical evaluation. An intracarotid amobarbital test is generally done as a prelude to surgery in order to lateralize language and memory function and to avoid neurocognitive deficits.

Psychosocial Assessment

Psychosocial evaluation is also extremely important to assess current level of functioning and to ensure realistic goals and attitudes are engendered in both the patient and their family prior to surgery.

Diagnostic surgical options

When a primary epileptogenic region or seizure focus is suspected but remains obscure despite appropriate neuro-imaging and

scalp (non-invasive) video/EEG recordings, some form of implanted (invasive) electrodes may be indicated. Intracranial electrodes can be placed in areas not readily sampled by routine surface electrodes and can give more precise EEG information because of their proximity to discharging areas of the brain and the lack of movement/muscle artifact on the recordings. They have the disadvantage, however, of sampling from a relatively small area of cerebrum surrounding the contact points and the fact that they are accompanied by a surgical risk. They should only be undertaken after appropriate noninvasive monitoring has been completed so that a hypothesis of seizure onset has been formulated and a clear goal of the investigation has been defined. The diagnostic surgical options of implanted electrodes include epidural, subdural and intracerebral or depth electrodes.

Epidural electrodes

Epidural electrodes are used infrequently and generally only for lateralization and approximate localization of seizure onset. These electrodes are placed through tiny openings in the skull with the electrode contact resting on the dura to provide a high amplitude EEG signal without muscle or movement artifact. Because they do not penetrate the dura the risk of infection is minor. These electrodes can only record from the lateral convexity of the cerebral hemispheres and therefore are limited in their spatial resolution.

Subdural electrodes

These electrodes are placed subdurally on the surface of the brain in the form of rectangular grids or linear strips with flat metal contact points mounted in flexible plastic. The grids require a craniotomy for placement and therefore are limited to unilateral application. The strip electrodes can be placed through burr holes over the lateral convexity or under the frontal or temporal lobes. It is difficult to place them in the interhemispheric fissure to record from parasaggital regions because of technical risks associated with large cortical veins. The major advantage of subdural electrodes is that they do not penetrate cerebral tissue and can record from a relatively wide area of the cortical surface. They can also be used for extraoperative cortical stimulation to map out specific areas of cortical function. Unfortunately, subdural electrodes cannot record directly from the deep cerebral structures (i.e. amygdala, hippocampus and cingulum) which are characteristically involved in many medically refractory

partial epilepsies. They also have a small but real risk of intracranial infection and hemorrhage estimated to be approximately 4%.

Intracerebral depth electrodes

Intracerebral depth electrodes can be placed stereotactically into deep cerebral structures with the aid of CT, MR and angiography. Most centers employ flexible electrodes with multiple contact points that are placed through small holes in the skull and secured with some form of cranial fixation. Electrodes are usually targeted towards the amygdala, hippocampus, orbital-frontal and cingulate regions and may be inserted via a lateral or vertex approach. Using a lateral approach, stereotactic cerebral angiography must be utilized to avoid major blood vessels during placement of the depth electrodes. Depth electrodes may be used in combination with scalp or subdural electrodes for more extensive coverage. Depth electrode investigation is generally indicated for patients with bitemporal, bifrontal of frontal temporal seizures and can localize a focal area of seizure onset not possible with scalp recordings. The major complications of depth electrodes include hemorrhage and infection with mortality and morbidity rates between 1-4%. It should be noted that the intracranial monitoring incurs greater risk than resective surgery itself and is also considerably more expensive than a noninvasive evaluation and therefore should be used only when necessary. With modern neuro-imaging, the use of invasive intracranial monitoring has declined from about 40-50% of patients in most centers to 10-20%.

EEG remains the most important and compulsory investigation of the brain electrical bioactivity.

Patient selection and surgical decision making

If the information obtained during the noninvasive presurgical evaluation consistently points towards a single area of the brain as being the site of seizure onset, then the patient may be taken directly to surgery for resection of that area. If neuro-imaging demonstrates a well-characterized lesion (i.e. unilateral hippocampal atrophy, cavernous angioma, focal cortical dysplasia, etc.) and is supported by the clinical features of the seizures then surgery may be reasonable without the general requirement for ictal EEG data. However, if the

data gathered from the clinical examination, imaging studies and noninvasive EEG evaluation are conflicting or disparities arise in the presumed localization of the seizure, then invasive intracranial monitoring is warranted. This is especially true in the extra-temporal epilepsies where EEG localization is notoriously difficult. If a localized area of seizure onset is confirmed then these patients too can undergo resective surgery.

Important indications for surgery:
1. Sufficient duration of epilepsy (at least 2 years)
2. Accurate topical diagnosis of the epileptic focus
3. Accurate determination of the seizure type
4. Mental retardation in children and intellectual decline in adults

Therapeutic surgical options

Epilepsy surgery began as removal of gross structural lesions of the brain. With the addition of EEG data from preoperative and intraoperative recordings, areas of removal expanded to include tissue that was grossly normal in appearance but known to give rise to epileptiform activity. Small areas of resection were soon replaced by partial lobectomies and more extensive cortical resection. While resection techniques (lesionectomy, lobectomy, hemispherectomy, corticectomy) generally yield the best surgical results, disconnection (callosotomy, subpial transection) and augmentation (cerebellar and vagal stimulation) techniques remain worthwhile considerations.

Lesionectomy

With the advent of MRI, many patients with recurrent seizures are now discovered to have small, previously unrecognized lesions such as cavernous angiomas, low grade astrocytomas, cortical dysplasias and areas of focal atrophy that are clearly the cause of their seizures. In general, if these are located in extratemporal sites, removal of the lesion and a small rim of surrounding cortex is often successful in controlling seizures. Removal of significant perilesional cortex may be necessary to achieve optimal seizure control in some patients. In many instances, if only a portion of the lesion is removed, the surgical result is suboptimal. If lesions are located within the temporal lobe, lesionectomy along with temporal lobectomy is carried out including the mesial temporal structures in order to yield good

results in 80% of cases. Overall, lesionectomy is associated with excellent results with success rates that are generally better than with surgery performed in patients without discrete lesions.

Temporal resections

The majority of resections involve the temporal lobe and initially consisted of the classical anterior temporal lobectomy. This was either carried out "en-bloc" under general anesthesia or using a more tailored resection with electrocorticography and cortical mapping under local anesthesia. The majority of temporal lobectomies, whether in the dominant or nondominant hemisphere, can now be safely performed under general anesthesia with or without electrocorticography. In the dominant hemisphere, temporal lobe removals usually extend back 3.5-4 cm behind the temporal tip or to the level of the central sulcus. In the non-dominant hemisphere, temporal lobectomies can extend beyond 7 or 8 cm but will result in a contralateral superior quadrantanopsia because of encroachment upon the optic radiation. It is important that the mesial temporal structures are included in the removal because most neurosurgeons believe that the hippocampus is intimately involved in seizure propagation or amplification. Studies also indicate that recurrent seizures are more likely following temporal lobectomy when the hippocampus is not removed.

Since almost 80% of temporal lobe seizures originate in the mesial structures, several operative approaches have been designed to reduce the amount of temporal neocortex removed but still resect the amygdala and hippocampus. The so-called antero-medial temporal lobectomy with amygdalo-hippocampectomy is a modification of the classical temporal lobectomy by reducing the amount of cortical removal and extending the hippocampal resection. Selective amygdalo-hippocampectomy removes the mesial temporal structures via either trans-sylvian, transcortical or trans-sulcal microsurgical approach with the goal of sparing temporal neocortex and reducing any possible neuropsychological deficits. Some cortical injury and white matter disruption does occur with this technique and it is only applicable to patients with clear cut mesiobasal temporal lobe epilepsy. No matter which procedure is advocated, if patient selection is appropriate, surgery in the temporal lobe offers good to excellent results in 75 - 85% of the cases. With modern imaging techniques, seizure free rates are now approaching 90% with febrile seizures,

hippocampal atrophy and mesial temporal sclerosis being positive predictors of a good outcome.

Morbidity and mortality figures for cortical excisions are quite low, less than 0.2% in one large series with over 2000 patients. The incidence of hemiparesis was 0% following temporal lobectomies and 0.5% for hemiparesis and/or dysphasia following frontal lobectomies. In dominant hemisphere removals, however, there is often a temporary speech deficit. Specific cognitive testing may detect permanent subtle deficits consistent with the site of removal but generally these are nonspecific. An upper quadrantanopsia may occur with larger temporal removals in the nondominant hemisphere. This may be acceptable if required for seizure control since it is usually unnoticed by the patient and does not interfere with normal daily living. Memory impairment has occurred with unilateral temporal removals in rare cases but this complication is avoided by preoperative testing of speech and memory function during the intracarotid amytal test. If memory is affected by amytal injection ipsilateral to the proposed side of the temporal removal, temporal excision may be designed to spare the hippocampus and medial structures but this approach may reduce operative success rates.

Extra-temporal resections

Extra-temporal resections are much less commonly performed with the majority being carried out in the frontal lobe. En bloc standardized resections are not generally suitable and most surgeons guide their resections by detailed electrocorticography, both intra- and extra-operatively along with detailed cortical mapping. Frontal resections range from localized topectomies to complete frontal lobectomies and must be carefully individualized. Identification of the primary motor cortex is essential to avoid motor deficits and anterior language cortex to avoid speech difficulties. Parietal and occipital resections are rarely carried out but may be gratifying in patients with clear structural lesions.

The results of cortical excision for extratemporal epilepsy are variable depending upon patient selection and method of presurgical evaluation. Outcome statistics are not as impressive for extra-temporal resections as they are for temporal removals. Nevertheless, extra-temporal resections including the frontoparietal and occipital regions can give excellent results. Patients with epileptic discharge

limited to the lobe of resection obviously tend to do better than those with more widespread discharges. In addition, some patients have more wide spread epileptogenic zones that require multilobar resections. In the largest cumulative series 64% of patients were improved, 36% being seizure free. With advances in neuro-imaging and other aspects of the presurgical evaluation, it is hoped that surgical success rates can improve in the future.

Hemispherectomy

Hemispherectomy is another form of cortical excision that is limited to patients with congenital hemiplegia, chronic encephalitis, hemi-megalencephaly or Sturge-Weber syndrome. These patients tend to have severe epilepsy with wide spread independent epileptic discharges that often extend to the contralateral (normal) hemisphere. It is only performed on patients who have a dense hemianopsia and are already hemiplegic with no fine motor activity on the affected side. The acute surgical risk is that some crude movement or sensation on the opposite side of the body would be adversely affected. A chronic complication was recognized to occur approximately 8 to 10 years after gross total hemispherectomy. This condition called superficial cerebral hemosiderosis resulted from chronic leaking of blood into the resection cavity producing recurrent seizures, sensori-neural deafness and hydrocephalus. It occurred in approximately 25% of patients by ten years and mandated a modification of the procedure. This complication is now avoided by performing anatomically subtotal but functionally complete hemispherectomy in which the frontal and occipital poles are left in place with their blood supply but all neural connections are transected. Residual cerebral tissue either decreases the risk of hemorrhage into the resection cavity or alternatively absorbs any blood that might leak in. Alternatives to anatomical hemispherectomy include hemispherotomy, cerebral hemicorticectomy, dural plication and ventriculoperitoneal shunting. All of these modifications attempt to reduce the risk of superficial cerebral hemosiderosis by minimizing cortical resection while maintaining complete functional disconnection.

Functional hemispherectomy or any of its variants, is one of the most successful surgical procedures for epilepsy with over 85% markedly improved and about 60% seizure free. Many patients also

demonstrate behavioral improvement probably on the basis of a better attention span and cognitive functioning.

Corpus callosotomy

Corpus callosotomy has been offered as an alternative to hemispherectomy in epileptic patients with congenital hemiplegia but the results are not as good as with hemispherectomy. It is indicated when the patient has a severely damaged hemisphere but motor, sensory or visual function would be valuable to preserve. In general, however, corpus callosotomy is most useful for those patients with generalized seizure disorders and bilateral independent epileptic areas in the frontal region. The seizures that respond best to callosotomy are sudden falls or "drop attacks" with injury to the patient. Some patients with additional focal seizures may experience an improvement or overall reduction in these partial seizures but about 20% of patients will have an increase in the number of focal seizures. The generalized seizures and drop attacks tend to improve markedly although a complete cure of seizures is extremely rare. Early surgical experience included deaths and severe morbidity but the risks have become extremely low with modern microsurgical techniques. The current practice is to section the anterior 2/3 of the corpus callosum on the first procedure. The posterior 1/3 may be sectioned at a second procedure if the results of anterior section are not satisfactory. Transient abulia is common following anterior callosotomy but other disconnection effects are fortunately mild and uncommon. In patients with complete callosotomy, disconnection symptoms are more frequent. There is often some difficulty in bimanual tasks and apraxia for commands directed to the nondominant extremity. Visual presentation to the hemifield opposite to the dominant hemisphere cannot be comprehended or described by language modalities and there is often significant difficulty writing with a nondominant hand. Fortunately, most of these functional deficits are not noticeable in normal daily living and are balanced by the improved seizure control.

Multiple Subpial Transections

In patients with seizure onset or epileptic zones located in eloquent cortex, multiple vertical subpial transections have been recommended as an alternative to cortical resection. This technique leaves the vertical columnar arrangement of the cortex intact thereby preserving function but prevents spreading of the seizure discharge in

the horizontal plane to reduce seizures. Some neurological deficits appear postoperatively but these generally resolve over several weeks with satisfactory improvement in seizure control in 70 % of patients. Experience with this technique is still rather limited but it does provide a surgical option in patients with seizures arising in cortex that has been previously considered inoperable.

Stereotactic ablations

Stereotactic lesions of deep cerebral structures have been carried out for a variety of generalized and focal forms of epilepsy in the past. Bilateral cingulotomies, amygdalotomies, lesions in the Field of Forel and thalamic lesions have all been tried. Results are scattered and too few for any conclusions to be made although generally they are unimpressive. While some lesions may have an initial good result, seizures tend to recur in virtually all patients and stereotactic ablations of subcortical structures are no longer in use.

Vagus nerve stimulation

More recently, a number of patients with both focal and generalized intractable seizures have undergone implantation of a nerve stimulator around the left vagus nerve. Less than half experienced a >50% reduction in seizure frequency and only the rare patient became seizure free.

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