

rupture, haemorrhage and infarction that might mask the original disease.

2. The oculomotor nerve (the IIIrd) and the abducent nerve (the VIth) are the anatomically most vulnerable nerves, and this is shown by **the paralysis of the eye bulbs**. The abducent nerve is vulnerable due to its long course in the subarachnoidal space. It is commonly injured on the contralateral side (the paradoxical sign) of the causative lesion.
3. An expansion that leads to intracranial hypertension might compress the midbrain, that can lead to the narrowing or even the obstruction of the aqueductus Sylvii. The cerebrospinal fluid circulation will be disturbed and the CSF will cumulate in the lateral ventricles and the IIIrd ventricle. An increased pressure leads to tissue damage in the area of the ventricular systems. The distension of the ventricular system worsens the intracranial hypertension.
4. A longlasting increase of the intracranial blood pressure leads to **the erosion of the brain bony cover**, which could be seen on the X ray. In children with intracranial hypertension the skull is deformed in shape, and especially if the fontanels are still open. Widening of the bony sutures of the skull, the pulsation of the fontanels and the prolonged time of fontanel ossification are other mechanisms, that can partially compensate for the process that leads to intracranial hypertension.

whereas the resorption is relatively unchanged. Yet the overproduction of the CSF is not the cause of hydrocephaly. We have to look for the cause of the overproduction of the CSF.

2. **CSF flow obstruction** (the most common cause), that disturbs the circulation of the CSF in the intracerebral or the extracerebral space. According to the level of obstruction we divide hydrocephalus into the non communicating – closed (obstruction above the level of the IVth ventricle e.g. aqueductus Sylvii, foramina Megendie and Luschke – the CSF doesn't reach the subarachnoidal space) or the communicating (the CSF circulation is obstructed in the subarachnoidal space.).

3. **Disturbance of the CSF absorption** (a rare abnormality). The absorption usually takes place in the venous sinuses of the arachnoid. This abnormality can result from an inflammatory process, bleeding, or the obliteration of the areas of resorption. The obstruction might be temporary upon taking some positions, or upon some head movements – this is known as **a ventral hydrocephaly**. It can develop with some tumor types, that grow from the stalk into the ventricular system of the brain.

The causes of hydrocephaly are divided into two groups:

1. congenital (developmental) abnormality,
2. acquired hydrocephaly

The congenital or the developmental abnormalities that might cause hydrocephalus can be divided into many groups:

1. **Arnold-Chiari malformation**, in which due to the increased intracranial pressure the cerebellum and medulla oblongata are pushed into the foramen magnum, upon which there will be CSF flow obstruction. The intracranial pressure increases and this might injure the respiratory centers in the medulla oblongata.
2. **Congenital stenosis or atresia of aqueductus Sylvii**, that is manifested by an excessive dilatation of the IIIrd ventricle and the lateral ventricles with a following increased intracranial pressure.

6.9 Hydrocephaly

In hydrocephaly the volume of the cerebrospinal fluid (CSF) is increased and the ventricles are dilated. In most of the cases the intracranial pressure is increased as well. Generally 3 possible mechanisms are accepted to be the cause of the development of hydrocephaly:

1. **An excessive production of CSF.** Where the choroid plexus produces more CSF, than normal

3. **Atresia of foramen Magendie and Luschke** that leads to the dilatation of the IVth ventricle, the IIIrd ventricle, and the lateral ventricles.

Acquired hydrocephalus can be caused by many factors. The usual causes are brain tumors (primary or secondary), haemorrhage or meningitis, upon which the resorption capacity of the arachnoidea is decreased.

A unique type of hydrocephaly is the so called **secondary hydrocephaly**, that in contrast with the former mentioned types, **is not manifested with high intracranial pressure**. This condition follows an over-committed brain infarction or a generalized brain atrophy, where the ventricular system dilatation comes secondary and this space is then filled with the CSF.

As an outcome of hydrocephaly in the newborn and in children with still opened fontanels there will be an expansion of the brainy part of the skull, and a prominent network of cerebral vessels will appear, the child's eyes will look very much like a sun set, and the percussion of the head will reveal a sound that is similar to the sound of a cracked ceramic pot. The brain is dilated, the CT or sonography reveals a massively dilated ventricular system in the cut sections, the brain tissue and according to the severity of hydrocephaly forms a thicker or a thinner layer on the margins. The brain groves become flattened, and the gray mater might often form only a layer of few mm. Some mental disorders will appear, as well as disturbances of vision, fine motor movements, and other functions. In the adults or in children with closed cranial sutures upon hydrocephaly there will be a raise in the intracranial pressure and the dominant signs and symptoms arise from this syndrom.

6.10 Demyelination disease

The group of demyelination diseases includes diseases, in which there will be a local or a diffused demyelination of the central or the peripheral nervous system. **Demyelination occurs most commonly in the white mater**, and only rarely in the gray mater of the CNS. The loss of myelin sheath can be primary or secondary. Upon myelin sheath destruction

the neurons and axons remain intact. The affection of their function is secondary. It is usually not connected to specific neuronal pathways or tracts. There are two types of demyelination disease:

1. **The myelin-classical type** – where the myelin is normal until adulthood, and its degradation occurs later on.
2. **Dysmyelin type** – the myelin is structurally abnormal from the early childhood.

The pathomechanism is yet unknown. We assume that there is **an autoimmune process, viral infection**, or the presence of both processes.

The secondary type might follow a previous injury to the neuron itself, or its axon. This type usually affects certain nerve tracts and hence it is possible to localize the precise anatomical position of the injury. This type of degeneration is typical for more disease groups. As a typical example we may take the degeneration of the neuronal tract as a consequence of a regional infarction.

Demyelination loci create neuritic plaques. The size of these plaques is variable. Histochemically it is a lipid degeneration of the myelin sheaths. The remainder of the degenerated myelin sheaths is removed by macrophages, that together with lymphocytes are gathered around the locus. The older loci are gray and sclerotic (hard). There is no cavitation what so ever. The consequences of the pathoanatomical changes are very similar in both cases (demyelination, the removal of the lipid particles by the phagocytic cells, gliosis).

Not only the covers of the neurons but even the axons themselves are injured by demyelination. Demyelination causes block of the neuronal conduction of stimuli.

Depending on the recent notes about the pathogenesis some of this disease group are classified under the so known neuroimmunological diseases. A relatively common disease such as multiple sclerosis belongs to this group.

6.10.1 Multiple sclerosis

Multiple sclerosis (MS) is a chronic, primarily **demyelination disease that involves the central nervous system**, and only very rarely the peripheral nervous system. It usually affects the young adults, more women than men (ratio 2:1). The beginning of the