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Hydrocephaly and its Clinical Course

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Abstract. This article provides information on the clinical manifestations, course, causes, and diagnosis of hydrocephalus in children and adolescents.

Keywords: Hydrocephalus, brain, bone, subarachnoid space, syndrome, pyramid, subarachnoid space.

Hydrocephalus is a disease characterized by an increase in the amount of cerebrospinal fluid in the cavity of the skull. A normal adult has 35-40 ml of fluid in the subarachnoid space and 75 ml in the subarachnoid space of the spinal cord, and 20-25 and 40-45 ml, respectively, in CKD. The amount of cerebrospinal fluid in a child with hydrocephalus is on average 1-2 l, in severe cases it can be 10 l or more. The amount of cerebrospinal fluid is determined by its production and absorption [1]. Classic experiments of Dandy (1927), repeated by many scientists, showed that cerebrospinal fluid is produced on the side of the epithelium of the vascular tangles of the cerebral ventricles. Clinical practice confirms the results of experimental studies. The participation of ependymal cells of cerebral ventricles in the production of cerebrospinal fluid in pathological conditions, in particular, in inflammatory processes, cannot be denied. Cerebrospinal fluid flows from the ventricles of the brain into the subarachnoid space of the brain and spinal cord. It is absorbed through the venous system. This explains the increase in the amount of cerebrospinal fluid in the head space, even if it is produced normally, as a result of the lack of flow of cerebrospinal fluid caused by the violation of venous blood circulation [1,2].

Hypersecretory, hypo- or aresorptive (resulting from impaired absorption) hydrocephalus, caused by excessive production of cerebrospinal fluid, is distinguished. Sometimes, both of these factors are involved. It is of great importance to determine the presence of general hydrocephalus with an increase in the amount of fluid in both the cerebral ventricles and the subarachnoid space, or internal obstruction as a result of the closure of the connection between the cerebral ventricles and the subarachnoid space. It should not be forgotten that hydrocephalus always develops ahead of the blockage [4]. External hydrocephalus is characterized by an increase in the amount of cerebrospinal fluid in the subarachnoid space and a normal amount in the ventricles of the brain, and it is caused by a decrease in the weight of the brain substance due to atrophy or swelling of the brain. It is less common than general or internal hydrocephalus. From a clinical point of view, hydrocephalus should be divided into congenital and acquired types according to the time of appearance. These forms of brain injury differ from each other in the course of clinical signs and consequences [3,5].

Congenital hydrocephalus. Etiology and pathology. Hydrocephalus can be caused by various harmful factors of pregnancy: infectious diseases of the mother, which only have a toxic effect on the fetal brain or are accompanied by infectious damage to the brain substance and membranes; various developmental anomalies and defects, traumas, etc. Nervous system damage in the first half of fetal development and various diseases of the mother in the second half of

pregnancy that disrupt the development of the fetal brain are of primary importance. In the past, trauma was given great importance in the origin of congenital hydrocephalus[6,7]. But the importance of toxoplasmosis is more important, one of its cardinal symptoms is hydrocephalus. Mild and uncomplicated viral infections in pregnant women (influenza, acute respiratory diseases, measles, etc.) can cause various damage to the fetal nervous system, including internal hydrocephalus. We present the classification of congenital hydrocephalus according to VAPurin and TPJukova (1976).

a) Simple hydrocephalus not associated with spina bifida.

1. Constitutional form (familial transient hydrocephalus)
2. Tumors - papilloma of vascular entanglement, liquor channels tumors that cause conduction disorders.
3. Due to the influence of unconscious factors on the fetus during the period of fetal development emerging hydrocephalus:

a) caused by a congenital injury

b) caused by congenital toxoplasmosis

c) caused by generalized cytomegaly

d) as a result of other effects on the fetus during pregnancy issued:

- Interrelated form
- occlusion of the cerebral aqueduct
- IV ventricular orifice occlusion.

4. In children who had intracranial trauma at birth hydrocephalus:

a) shape of conductor

b) occlusion of the cerebral aqueduct B Associated with spina bifida or cranial hernias hydrocephalus.

From the pathologoanatomic point of view, thinning of the brain substance is observed in various degrees. The white matter of the hemispheres is more damaged than the gray matter. The most affected parts are the axillary body, pyramid and other conduction systems. In a few patients, the subcortical nodules were reduced in size, thinned, and atrophied[8].

In severe cases, the cerebral hemispheres become fluid-filled thin-walled sacs. In moderately severe cases, the brain matter is preserved to one degree or another, the ventricles are enlarged, and the amount of fluid increases sharply. The thinned floor of the 3rd ventricle often becomes bulging, balloon-like, and may compress the chiasm and pituitary [9].

Congenital hydrocephalus is characterized by an increase in the size of the skull and a change in its shape. An enlarged head can be observed at birth, and often such children die during childbirth. Instead of 34 cm (norm), the head circumference can reach 50-70 or even 100 cm. In cases of congenital hydrocephalus, where the head size at birth is relatively normal, they increase rapidly in the first months of life, especially due to the sagittal size, the head enters the dolichocephalic form. But the head can be more spherical and brachycephalic. Forehead and crown bulge sharply, the forehead protrudes in relation to the face. The Turkish saddle is deformed, the bones of the skull are thinned. The sutures of the skull expand. In cases of acutely developed congenital hydrocephalus, when hydroanencephaly is observed, a vital diagnosis is made by means of transillumination or diaphanoscopy [9]. A child's head can be seen under the light of a 100-watt lamp in a dark room, and the house will light up. Changes in the eye socket are very characteristic: their upper wall is pressed down, the eyes are widely spaced and narrowed. The root of the nose is wide, the nostrils are directed forward and even upwards. The

child's gaze is directed downward according to the features of the structure of the eyeball, the upper eyelid does not completely cover the sclera, and the lower eyelid partially covers the iris. The bones of the skull are thinned, sometimes when pressed with a finger, they can sink in sharply. Thinning of the cranial bones causes high-pitched sounds on auscultation. The network of veins is sharply expanded, especially in the area of the nasal septum, temple and forehead, and sometimes in the area of the neck and upper chest. When the child screams or cries, the venous net bulges [9]. An enlarged and deformed head contrasts sharply with a small, triangular face. The base of the triangle is pointing up and the apex is pointing down. The face is pale, covered with thin skin. The hair on the head is sparse. In some patients, hydrocephalus is accompanied by microcephaly. In such cases, the size of the head decreases rather than increases. Teething is delayed, but general physical development is also delayed[7].

In addition to hydrocephaly, various anomalies and developmental defects are often observed: non-union of spina bifida arcs of different vertebrae, hydrocephaly, syringomyelia, albinism, dwarfism, early sexual development, adipozogenital dystrophy, etc. Spinal hernias with myeloschisis or myelocele are often accompanied by hydrocephalus.

The data of Leber(1964) which revealed hydrocephalus in 80% of children with spinal cord hernia in the first months of life, and 63% of hernia in other locations are of interest. The author notes that hydrocephalus in lower paraplegia is almost always accompanied by Arnold-Chiari syndrome in spinal cord hernia without paraplegia, and hydrocephalus also occurs in it[10]. The question of the pathogenesis of Arnold-Chiari syndrome is still complicated. In this case, there is a defect in the development of the brain stem. On the X-ray of the skull, the dimensions of the spinal cavity are reduced and the large occipital foramen is enlarged. The elongated brain, which has changed in size and structure, is surrounded by a dense fibrous tissue ring in the area of the cerebrum, which is connected to the soft and hard membranes of the brain and blocks the subarachnoid space. Neurological symptoms of congenital hydrocephalus are very polymorphic. Movement and mental disorders take the first place: spastic paralysis and paresis, often lower paraplegia, rarely hemiplegia with increased muscle tone, increased tendon reflexes, clonus of the kneecaps and palms of the feet, rarely clonus of the palms of the hands, hemiplegias with pathological bending and writing reflexes. Babinski's sign is almost always called, but it should not be forgotten that in children of the first year of life, Babinski's sign is physiological. Symptoms of pyramidal damage include cerebellar ataxia and tremor[1].

Damage to the pyramidal systems or their deficiency underlies the motor function delay. The patient is late for children to hold their head, sit, stand, and walk. In severe patients, it is not observed at all. In children who started walking very late, their head hangs due to their weight, their body is bent, their hands are lowered, their gait is spastic-atactic. A common symptom of hydrocephalus is nearsightedness or farsightedness caused by damage to the optic nerve. In most patients, nystagmus, pupillary reaction disorders, dimmed optic nerve papillae or their atrophy (primary or secondary) are evident. Cerebrospinal fluid is clear, colorless, and flows under high pressure. Most patients with hydrocephalus have a decreased amount of protein and cells in the cerebrospinal fluid. Wasserman's reaction is often negative, even in patients with a wound etiology. Spontaneous discharge of fluid is observed in some patients from the nose, in rare cases from the larynx and from the sutures of the skull. Speech development is delayed to varying degrees, and in some severe cases, children do not even begin to speak. The degree of mental retardation varies from mild retardation to profound retardation and idiocy. Even at a relatively high level of mental development, features characteristic of children with hydrocephalus are observed: alertness, tendency to rude jokes, use of loud, beautiful expressions when they are not appropriate. Behind this lies the mechanical repetition of phrases that are easily remembered. Actions are as automated as speech. Mechanical memory is well developed in children with hydrocephalus.

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