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Research Article

CLINIC, DIAGNOSIS AND TREATMENT OF OCCLUSIVE HYDROCEPHALUS IN CHILDREN

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Saidumarov Dilshod Mirzaakhmatovich
Doctor Neurosurgeon, Fergana Branch Of Republican Scientific Center Of Emergency Medical Care, Fergana, Uzbekistan

Saidumarova Marguba Tulanovna
Assistant, Department Of Pathological Physiology, Fergana Medical Institute Of Public Health, Fergana, Uzbekistan

ABSTRACT

This article deals with the clinic, diagnosis and methods of treatment of occlusive hydrocephalus in children. The authors of the article consider that surgical intervention in the treatment of occlusive hydrocephalus in children is the only method to combat the disease. In most cases, medication can only slow down the course of the disease, but does not eliminate the underlying cause of the disease. In the case of a successful operation, almost complete recovery and a return to a normal life is possible.

KEYWORDS

Hydrocephalus, head, brain, treatment, surgery, neurosurgery, neurological and mental disorders, hematoma, intraventricular tumour.

INTRODUCTION

Occlusive hydrocephalus is one of the most common neurosurgical diseases of childhood [6; 7; 11]. According to different authors, the incidence of this disease is 3-4 cases per 1000 newborns [5; 10]. In most cases, hypertension-hydrocephalus syndrome is the main cause of patient decompensation, leading to

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severe, sometimes irreversible neurological and mental disorders, which lead to persistent disability of patients [3; 8], therefore, the search for effective treatments for this disease is an urgent medical and social task.

METHODS

Hydrocephalus was rarely described in ancient medical literature, although its existence and symptoms were well known. Hippocrates, the father of medicine, is considered to be the first physician to attempt to document the treatment of hydrocephalus. He recommended trepanation for the treatment of epilepsy, blindness and possibly hydrocephalus. The Greeks reportedly treated hydrocephalus by wrapping the bark around the patient's head and inserting it into the trepanation orifices.

Evacuation of superficial intracranial fluid in children with hydrocephalus was first described in detail by Ibn Sina. Due to a poor understanding of the pathophysiology of hydrocephalus, initial attempts at therapy were sporadic and usually resulted in failure. Many practitioners relied on conservative therapy. Treatment attempts included many medications, laxatives such as rhubarb, calomel and oil as well as various diuretics, head wraps, bloodletting and cranial trepanation [2].

In neurology, the term "hydrocephalus" currently refers to the increased accumulation of cerebrospinal fluid (cerebrospinal fluid) in the cranial cavity. If the cause is an occlusion (blockage, compression) of the cerebrospinal fluid pathways, hydrocephalus is called occlusive hydrocephalus. A synonym for this condition is closed hydrocephalus, as occlusion leads to the occlusion of the cerebrospinal fluid system and the accumulation of cerebrospinal fluid. In the ICD-10, occlusive hydrocephalus is listed under the name 'obstructive'. The disorder is secondary and always has a causal pathology. Occlusive hydrocephalus occurs at all ages, in children and adults, and may be congenital. In some cases there is an acute occlusion that requires immediate medical attention.

The cerebrospinal fluid system consists of four ventricles: the paired lateral ventricles and the unpaired third and fourth ventricles. Cerebrospinal fluid from the lateral ventricle enters ventricle III through the interventricular foramen of Monro, then through the aqueduct of Sylvius into ventricle IV, from which it drains through the Luschka and Magendie orifices into the cerebral and spinal subautery cisterns. Occlusive hydrocephalus develops when there is an obstruction in any part of the described cerebrospinal circulation system. Factors of occlusion may include:

- Abnormal development of the brain. Congenital stenosis, underdevelopment of the aqueduct of Sylvius, Dandy-Walker syndrome, and Arnold-Chiari anomaly are genetically determined or formed under conditions of intrauterine infection, fetal hypoxia, and teratogenic influences. Anatomical changes in these malformations lead to the development of hydrocephalus in the antenatal period or shortly after birth.

- Cerebral tumors. Ventricular neoplasms reduce ventricular volume and cause occlusion of the communicating orifices. Cerebral cistern tumours prevent the flow of cerebrospinal fluid from the ventricles into the cisterns. Pericentricular tumors, neoplasms of the trunk, cerebellum squeeze the cerebrospinal fluid pathways as they grow. The result of these processes is an accumulation of cerebrospinal fluid in the ventricles.

- Intracerebral hematoma. Formed due to craniocerebral trauma (including intracranial birth

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trauma), haemorrhagic stroke. Occlusive hydrocephalus is caused by the occlusion of the cerebrospinal fluid outflow pathways by the hematoma that has formed.

- III ventricular colloidal cyst is a benign neotumoral entity. The cyst increases in size and blocks the entrance to the aqueduct of Sylvius. The outflow tract is blocked and hydrocephalus develops.

- Hemorrhage in the ventricles of the brain. Occurs with trauma, rupture of the vessels of the arteriovenous malformation, hematoma bursting into the ventricles. The blood in the ventricular cavity coagulates to form clots that occlude the Monro, Luschka, Magendie orifices and the narrow canal of the cerebral aqueduct.

Obstruction, compression of the liquor-bearing tracts leads to a disturbance of the outflow and accumulation of cerebrospinal fluid in the ventricles of the brain. The ventricular volume increases, and with the intracranial space closed, this leads to an increase in pressure inside the cranial cavity. The rate of increase in intracranial hypertension depends on the extent and mechanism of the occlusion. Monroe orifice occlusion leads to lateral ventricular enlargement, occlusion at the level of the aqueduct of Sylvius leads to dilation of the III and both lateral ventricles, and at the level of the Magendie and Luschka orifices leads to total dilatation of the ventricular system.

Occlusive hydrocephalus of tumour origin forms gradually, with post-traumatic haematoma within a few days. Occlusion by a blood clot, part of an intraventricular tumour occurs suddenly, leading to acute hydrocephalus. Intraventricular neoplasms, colloidal cysts can cause occlusive crises, a transient blockage of the cerebrocirculation that occurs when the mass is displaced. Significant intracranial hypertension causes compression of brain tissue, feeding vessels. Hypoxia and dysmetabolic changes occur, leading to neuronal death. The continuing increase in pressure causes displacement of cerebral structures (mass effect), leading to severe complications.

In clinical practice, occlusive hydrocephalus is divided into etiological and anatomotopographic classifications. Both classifications have implications for the choice of the most appropriate treatment modality. According to the etiological principle, a distinction is made:

- the congenital form - formed during the intrauterine period due to malformations of the spinal cord system, cerebral anomalies (e.g. brain cysts) that squeeze the liquor-bearing tracts. Appears from the first days of life.

- acquired form - occurs during life, caused by trauma and CNS diseases. Appears in conjunction with other symptoms of the causal pathology.

According to anatomotopographic classification, occlusive hydrocephalus is subdivided into:

- monoventricular - one lateral ventricle is dilated. The cause is a blockage of the interventricular orifice.

- biventricular - the volume of the two lateral ventricles is enlarged. Occlusion at the level of the third ventricle.

- triventricular - enlargement encompasses three ventricles. Obstruction of the liquor flow is located in the area of the cerebral aqueduct.

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- tetraventricular - all ventricles are dilated. The cerebrospinal circulation is impaired at the level of the outflow pathways from the IV ventricle.

Regardless of the level of cerebrospinal block, the clinical picture is dominated by signs of intracranial hypertension. Patients present with intense headache (cephalgia), non-meal related nausea, vomiting, and frequent nosebleeds. Cephalgia causes a forced position of the head and is accompanied by a feeling of pressure on the eyeballs. The onset of symptoms is acute or subacute, with the background of the clinical picture of the underlying pathology. In some cases, occlusive hydrocephalus is the first sign of malignancy.

The cochleovestibular and optic nerves are most commonly affected by intracranial hypertension. Vestibular ataxia, tinnitus, hearing loss, visual acuity and visual field defects are observed. Epileptic paroxysms are not uncommon. Associated focal deficits depend on the causal pathology and are represented by paresis, paralysis, sensory disturbances, cognitive deficits and cerebellar syndrome. The cerebral block at the level of the third ventricle is characterized by diencephalic symptoms: pulse palpitations, blood pressure fluctuations, hyperhidrosis, discoloration of the skin (pallor, hyperemia). Obturation of the Sylvian aqueduct is accompanied by dissociation of the pupillary response to light, impaired convergence and gaze paresis. Block in the fourth ventricle is accompanied by cerebellar ataxia.

Occlusive hydrocephalus in young children is manifested by an increase in the size of the skull, divergence of the cranial sutures, enlarged and swollen fontanelles. Typical features of congenital hydrocephalus are an enlarged globe-shaped head, a relatively small torso, deep eye sockets and swollen scalp veins. The children are retarded in their psychophysical development. The severity of intellectual disability depends on the age of onset, duration, severity of intracranial hypertension.

Occlusive hydrocephalus can be accompanied by an acute and almost complete block of cerebrospinal fluid flow - occlusive hydrocephalus crisis. The attack is accompanied by acute intense cephalgia, repeated vomiting, facial hyperaemia followed by pallor, oculomotor disturbances, depression of consciousness and autonomic symptoms. The most severe complication of hydrocephalus is the mass effect. Displacement of brain tissue in the direction of the greater occipital foramen leads to compression of the medulla oblongata, which are located vital centres regulating cardiovascular and respiratory activity. A violation of the functions of the latter is fatal.

Diagnostic activities begin with the collection of anamnesis: determining the time of the onset of the intracranial pressure increase symptoms, the nature of their development, the presence of a diagnosis of brain disease, the fact of head trauma, etc. A further diagnostic algorithm includes:

- neurological examination. It allows the neurologist to identify objective symptoms of intracranial hypertension, existing focal deficits. The findings allow a topical diagnosis to be made.

- consultation with an ophthalmologist. It includes ophthalmoscopy, perimetry, visometry. Examination of the ocular fundus determines congested optic discs, with prolonged hydrocephalus - signs of optic atrophy. Examination of the visual fields reveals narrowing, loss of certain areas, and visometry reveals a decrease in visual acuity.

- echoencephalography. Due to its ease of execution it can serve as a screening method. Allows

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diagnosis of increased intracranial pressure, ventricular dilatation, and displacement of cerebral tissues.

- neuroimaging. Infants are examined by neurosonography through the fontanelle, while others are examined by MRI of the brain. The examination makes it possible to diagnose malformations, localize a cerebral block, and determine its cause. MSCT and CT scan of the brain are performed in complex diagnostic cases in addition to MRI, if there are contraindications to MRI examinations.

It is necessary to differentiate occlusive hydrocephalus from subarachnoid haemorrhage and other forms of hydrocephalus. Differential diagnosis is also made among the possible causes of occlusion. In children in the first months of life, hydrocephalus must be differentiated from macrocranial, which is mostly familial, without symptoms of hypertension or developmental delay.

The only effective treatment is neurosurgery

Since the 1950s, the standard treatment for any form of hydrocephalus has been bypass surgery to restore the movement of the cerebrospinal fluid. After cranial trepanation, one end of the shunt, ending with a radiopaque catheter, is inserted into the dilated ventricular cavity. The intermediate, longest part, made of silicone, is placed subcutaneously. The distal end, which also has a catheter, opens into the abdominal or thoracic cavity to allow drainage. The shunt is fitted with a pump that automatically regulates the pressure of the cerebrospinal fluid [1; 9]. Since the mid-1980s, endoscopic surgery has played a significant role in the treatment of hydrocephalus.

The treatment of occlusive hydrocephalus by shunt is quite effective, however, according to various sources, complications during this operation amount to 40-60% of cases. Depending on the cause of the dysfunction, all or parts of the shunt must be replaced. Experience shows that the most frequent complications that require shunt revision occur between six months and one year after surgery. Most patients who undergo bypass surgery have to undergo several surgical interventions during their lifetime. In any case, at least two or more revisions should be expected - after all, the child is growing. After bypass surgery, the patient becomes shunt dependent, meaning that the rest of his or her life will depend on the operation of the shunt.

Complications of bypass surgery:

- occlusion (blockage) in both the ventricles of the brain and the abdomen;
- infection of the shunt, ventricles, cerebral membranes;
- mechanical damage to the shunt;
- hyperdrainage (rapid discharge of cerebrospinal fluid from the ventricles) is often accompanied by rupture of the convexital veins and formation of hematomas;
- hypodrainage (slow ventricular outflow tract) - surgery is then ineffective;
- development of epileptic syndrome, abdominal decubitus, etc.

Endoscopic treatment of hydrocephalus

At present, endoscopic treatment of hydrocephalus is a priority in the world practice of neurosurgery. It is worth mentioning the effectiveness of endoscopic ventriculocysternostomy of the floor of the third ventricle in treating occlusive hydrocephalus. This operation is widely used and accounts for about 80% of neuroendoscopic surgeries. The aim of the operation is to create outflow pathways from the ventricular system of the brain (ventricle III) into the cisterns of

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the brain, through which fluid is reabsorbed (absorbed) as in a healthy person.

Indications for surgery:

- primary operation in occlusive hydrocephalus with the level of occlusion from the posterior third ventricle and beyond;
- alternative surgery for complications of bypass surgery with removal of the previously installed bypass system (instead of "bypass revision" surgery);
- post-traumatic hydrocephalus;
- mixed hydrocephalus (internal and external);
- operation of choice when removing the shunt system to achieve shunt-independence;

Advantages of the operation compared to classic shunts:

- the operation restores the physiological (as in a healthy person) liquor flow from the ventricular system of the brain to the basal cisterns;
- no foreign body (shunt system) is implanted into the body, thus avoiding related problems (infection, malfunction, need for revisions);
- a much lower risk of hyperdrainage and related complications (subdural hematomas, hydromas, etc.);
- the operation is less traumatic;
- the operation is more cost-effective for the hospitals;
- improved quality of life.

In addition to the above-mentioned treatments for occlusive hydrocephalus, the correction of the pathology of the liquor ducts (the plasticization of the aqueduct of Sylvius lends itself to this method) as well as the elimination of the blocking cause must also be mentioned. If the volume of the mass is large, this type of intervention is too traumatic. It can be performed in case of hematomas, tumours. In an emergency, external ventricular drainage of one of the lateral ventricles is performed.

CONCLUSIONS

Surgical intervention is virtually the only method of combating the disease. In the majority of cases, medication is only able to slow down the progression of the disease, but does not eliminate the underlying cause. If the operation is successful, a nearly complete recovery and a return to a normal life is possible.

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