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CONGENITAL ANOMALIES OF THE BRAIN

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Abstract. Abnormalities of brain development are the result of abnormalities in the formation of individual cerebral structures or the brain as a whole occurring in the intrauterine period. They often have non-specific clinical symptoms: predominantly epileptic syndrome, mental and intellectual retardation. The severity of the clinic directly correlates with the degree of brain damage. Diagnosed antenatally by obstetric ultrasound, after birth by EEG, neurosonography and MRI of the brain. Treatment is symptomatic: antiepileptic, dehydration, metabolic, psychocorrective.

Key words: brain, brain anomalies, intrauterine period

Introduction. Anomalies of brain development are malformations consisting in abnormal changes in the anatomical structure of cerebral structures. The severity of neurological symptoms accompanying cerebral anomalies varies considerably. In severe cases, malformations are the cause of antenatal fetal death, they account for up to 75% of intrauterine deaths. In addition, severe cerebral anomalies cause about 40% of neonatal deaths. The time of manifestation of clinical symptoms can vary. In most cases, cerebral anomalies manifest themselves in the first months after birth. However, since brain formation lasts until the age of 8 years, a number of malformations are combined with malformations of somatic organs. Prenatal detection of cerebral anomalies is an urgent task of practical gynaecology and obstetrics, and their postnatal diagnosis and treatment are priority issues of modern neurology, neonatology, paediatrics and neurosurgery.

Abnormalities of brain development

Causes

The most significant cause of failures of intrauterine development is the effect on the body of the pregnant woman and on the fetus, various harmful factors that have a teratogenic effect. The occurrence of anomalies as a result of monogenic inheritance occurs in only 1% of cases. The most influential cause of brain malformations is considered an exogenous factor. Teratogenic effect have many active chemical compounds, radioactive contamination, some biological factors. Of no small importance here is the problem of pollution of the human environment, which causes the entry of toxic chemicals into the body of the pregnant woman.

Various embryotoxic effects may be associated with the lifestyle of the pregnant woman herself: for example, smoking, alcoholism, drug addiction. Dysmetabolic disorders in the pregnant woman, such as diabetes mellitus, hyperthyroidism, etc., may also cause fetal cerebral anomalies.

Many medications that a woman may take early in pregnancy, unaware of the processes occurring in her body, also have teratogenic effects. A powerful teratogenic effect has a powerful teratogenic effect infections carried by the pregnant woman, or intrauterine infections of the foetus. The most dangerous are cytomegaly, listeriosis, rubella, toxoplasmosis.

Pathogenesis

The construction of the nervous system of the foetus begins literally from the first week of pregnancy. Already by the 23rd day of gestation, the formation of the neural tube is completed, incomplete closure of the anterior end of which entails serious cerebral anomalies. Approximately by the 28th day of pregnancy, the anterior cerebral bubble is formed, subsequently dividing into 2 lateral ones, which lie at the base of the cerebral hemispheres. Further, the cerebral cortex, its gyrus, corpus callosum, basal structures, etc. are formed.

Differentiation of neuroblasts (germinal nerve cells) leads to the formation of neurons, which form the grey matter, and glial cells, which make up the white matter. The grey matter is responsible for the higher processes of nervous activity. In the white matter are various conductive pathways that link cerebral structures into a single functioning mechanism. Born in time, a newborn has the same number of neurons as an adult. But the development of his brain continues, especially intensively in the first 3 months of life. There is an increase in glial cells, branching of neuronal outgrowths and their myelination.

Failures can occur at different stages of brain formation. If they occur in the first 6 months of pregnancy, they can lead to a decrease in the number of formed neurons, various disorders in differentiation, hypoplasia of various parts of the brain. In later periods, lesions and death of normally formed cerebral substance may occur.

Purpose of work: to study brain anomalies, to familiarise with the frequency of occurrence of these congenital pathologies.

Material and methods of research: review of literature sources. Anomalies of brain development (malformations) are the result of abnormalities in the formation of individual cerebral structures or the brain as a whole occurring in the intrauterine period. Anencephaly is a rare congenital malformation characterised by the complete absence of the cerebral hemispheres, the bones of the skull vault and the soft tissues covering it. The brain matter is usually replaced by connective tissue, rich in blood vessels, with cystic cavities. Anencephaly occurs at an incidence of approximately 1 in 10,000 newborns. In a study of the incidence of this malformation, the incidence of anencephaly was found to be significantly higher in low birth weight babies (less than 3kg) and in girls. The rate of birth of children with this anomaly is higher in young women (under 19 years of age) and in women over 35 years of age compared to the group of women 20-24 years of age .If a child with anencephaly is born alive, his life expectancy is not more than 10 days, which is associated with severe impairment of vital functions.

Microcephaly (Giacomini syndrome) is an underdevelopment of the brain manifested at birth by a decrease in its weight and size. Microcephaly is usually combined with a reduced head circumference (at least 5 cm from the average indicators) and further lagging growth of the cerebral skull (microcrania), and its sutures may remain open for a long time. Its morphological sign is the underdevelopment and irregular structure of the large hemispheres with a relatively normal architectonics of the cerebellum and brain stem. This is one of the most frequent anomalies in patients with severe mental retardation - up to 20% of all cases of oligophrenia are due to this pathology. [2, c.55]. Occurs rarely, on average in 1 case per 6-8 thousand births Primary microcephaly - a consequence of a genetic defect inherited by autosomal recessive type or arising in connection with chromosomal abnormalities. Secondary microcephaly can be caused by intrauterine infection (rubella, cytomegalovirus encephalitis, toxoplasmosis), intoxication or asphyxia, brain injury. In secondary microcephaly in the brain may be cystic cavities, foci of haemorrhage and calcification (improper distribution of calcium). Macrocephaly is an increase in the weight and volume of the brain and, with it, the brain skull at birth. Macrocephaly affects up to 5% of the paediatric

population. In most cases, it is accompanied by a disruption of the location of the cerebral gyrus, changes in the cytoarchitectonics of the cortex, foci of heterotopia in the white matter, and usually manifestations of oligophrenia (mental retardation) are noted, a seizure syndrome is possible. The causes of macrocephaly have not been identified. On craniography (radiographs of the skull bones), the bony sutures are not dilated, and the brain ventricles are normal or nearly normal in size. Macrocephaly should be differentiated from hydrocephalus. Hydrocephalus is a definite symptom complex, which is based on a pathology of the liquor system and is characterised by an increase in the amount of fluid in the cranial cavity, e.g. due to the absence of Mohandy's and Luschka's foramen or narrowing of the Monroe's or Sylvian ducts. In a newborn child, the total amount of cerebrospinal fluid is 15-20 ml, and in hydrocephalus can increase to 1-1.5 litres. [3, c.14]. An important role belongs to damage to the nervous system in the first half of intrauterine development and various diseases of the mother (toxoplasmosis) in the second half of pregnancy, disrupting the development of the fetal brain. Ultrasound screening of congenital hydrocephalus is performed at 16-20 weeks of pregnancy. At this time, the lateral ventricles are easily seen. Diverse neurological symptomatology is a consequence of increased intracranial pressure with the development of atrophic and degenerative processes in the brain and cranial nerves. Persistent increase in intracranial pressure leads to compression of brain capillaries and, as a consequence, to atrophy of nervous tissue. The incidence is 0.5 cases per 1000 births.

RESULTS: Structural brain abnormalities are relatively common and can be detected both intrauterine and postnatally. Problems with brain formation in the foetus can lead to serious consequences, including death before or after birth. Most often, these abnormalities are caused by harmful factors affecting the mother and baby during pregnancy. Hereditary factors play a minor role in the development of brain malformations, accounting for only 1% of cases. The main cause is considered to be the influence of external factors such as: toxic substances, infections, nutritional deficiencies, and the use of medications. **Conclusions:** Severe brain anomalies can lead to the death of the foetus in the womb or the newborn. Even less severe malformations can have long-term consequences for the child's health and development./Congenital malformations of the central nervous system in children are one of the topical issues of modern medicine, accounting for about 25% of all CHD in children

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