RHABDOMYOMA OF THE HEART IN CHILDREN

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Annotation. Rhabdomyoma of the heart refers to benign neoplasm, often diagnosed in the first year of life, can cause serious disorders in the work of the heart. In some cases, rhabdomyoma of the heart is observed with tuberoussclerosis e, in this regard, it is necessary to conduct a full examination of the child and promptly prescribe treatment and regular monitoring.Early detectionof rhabdomyoma of the heart and дprenatal observation of a cardiologist and cardiac surgeon with symptomatic treatment will allow you to choose the right treatment strategy, since in some cases there isa regression of rhabdomyoma.

Keywords:rhabdomyoma of the heart, tuberous sclerosis.

Heart tumors are observed infrequently. According to various authors [1], they make up 0.05% of tumors of various localities, so the disease is not a social problem. The relevance of the issue is that the clinical diagnosis of heart tumors is very complex. Clinically, they are detected only in 5-10 % of cases [1, 2].

Heart tumors are a rare pathology of childhood. Among benign primary heart tumors, rhabdomyoma is the most common in children [3,4,5].

Rhabdomyoma of the heart is a benign neoplasm that usually develops from striated muscle tissue of the heart. It can be localized in any part of the organ, except for the valves, but it is most often found in the ventricle cavity.

Rhabdomyoma in most cases develops in the fetus, much less often after the birth of a child. There is evidence that the tumor progresses most actively in the second half of pregnancy. In some cases, after the birth of a child, the tumor mayspontaneously regress, up to complete absorption within a few years [6].

Rhabdomyomacan be diagnosed from the first months of life. It is formed from embryonic muscle cells as a result of early dysembryogenesis disorders [7].

The causes of the development of the disease are not fully understood. Among the possible factors that provoke the development of pathology are called:

- genetic predisposition;
- adverse environmental conditions;
- viral and bacterial infections of the mother;
- hormonal disorders in the mother's body;
- a tense psychological situation, severe stress;
- the use of certain medications, etc.

There are no specific signs of the disease, usually the tumor grows slowly and does not cause any inconvenience. If the neoplasm is small in size, it can remain undetected for many years.If the tumor is large, it can seriously interfere with the heart and negatively affect the development of the child. Symptoms such as heart failure, arrhythmia, tachycardia or bradycardia may occur, and atrioventricular block and ventricular extrasystole may develop. Further progression of the neoplasm can cause serious violations in the work of the heart up to its sudden stop.

Rhabdomyomatous formations can be in the form of a single node or multiple. They are more often localized in the ventricles and have mixed intra- / extra-mural growth. Children in combination with MVP and LVAC may have complaints of “aching” pain in the heart, palpitations, cephalalgia and dizziness, increased fatigue, a feeling of “chilliness” and cold hands at room temperature [18]. In rare cases, rhabdomyomascan be localized in the Atria, based on the atrial septum. Depending on the location, rhabdomyoma of the heart can have a malignant clinical course, leading to critical obstruction of the valve or ventricular chambers' exit tract, damaging the conducting system of the heart, leading to a fatal outcome, including sudden death [8,9,10, 22, 23, 24].

Rhabdomyoma of the heart with a highfrequency (up to 50-80%) it is associated with tuberous sclerosis (TS) (Bourneville's disease), which is a genetically determined disease and is characterized bythedevelopment ofmultiple

benign tumors invarious organs, with a progressive course [8,9,10,11,14-17]. In this case, rhabdomyoma may be the first sign of the disease, and subsequent examination of the patient reveals a symptom complex of tuberous sclerosis [12, 13, 20,21], the diagnostic criteria of which are presented in table 1.

Table 1.

Diagnostic criteria for tuberous sclerosis (E.S.Roachetal, 1999)

Main (large) criteria for TS	Additional (small) criteria for TS
— angiofibroma the face (cheeks, bridge of the nose) or the area of the forehead in patches (plaques) — Nontraumaticsubungual fibroma Genesis — Three or more spots of hypo pigmentation Areas in the form of shagreen plaques — Multiple tumor nodules in the retina — Tubercles in the cortex — Subependymal nodules —Subependymal giant cell astrocytoma — Rhabdomyomas hearts (single or multiple) — angiomyolipoma Renal or pulmonary lymphangiomyomatosis	Multiple random way distributed depressions (pits) on tooth enamel — Hamartoma rectal polyps — Bone cysts — Migration of the white matter of the brain in the form of lines beam — gingival Fibromatosis — Non-renal hamartomas — Uncolored (achromatic) spot on the retina — Cutaneous manifestations in the form of confetti (small round spots) —Multiplerenalcysts

The diagnosis of tuberous sclerosis is considered reliable if the patient has two large or one large and two small criteria [9, 10].

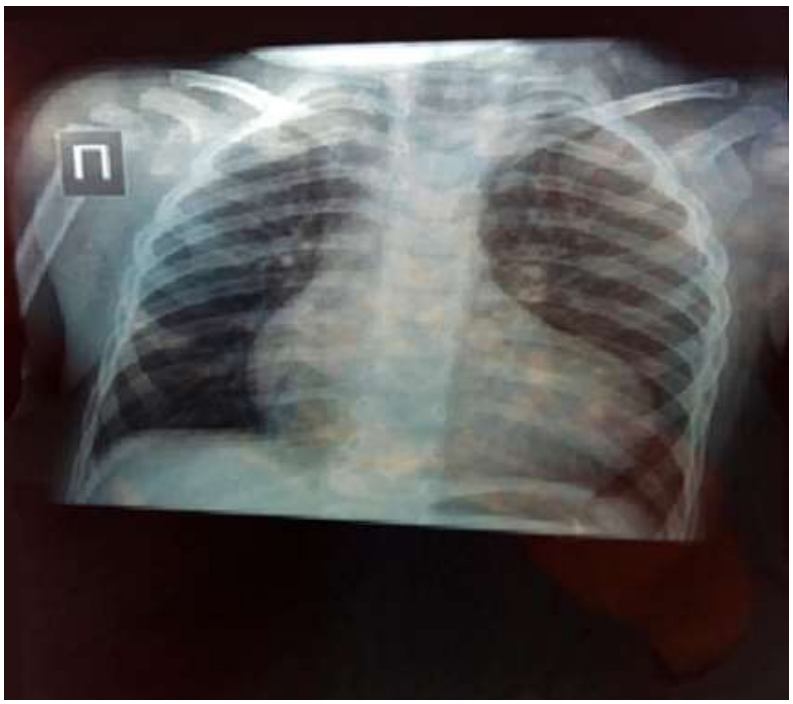
As a rule, nextunderstudiesare conducted:

- MRI of the head at least every 3 years to detect intracranial complications;
- Kidney ultrasound or abdominal MRI every 3 years for school-age children and every 1-2 years for adults to detect kidney tumors;
- Girls over 18 years of age should be screened annually for shortness of breath during physical activity and at rest, as well as high-resolution CT scans every 5-10 years;
- Periodic neuropsychological testing and behavioral screening of children to provide assistance and support in school and behavioral measures.

The prognosis depends on the severity of symptoms. All patients should be regularly examined to detect possible complications of TSC in a timely manner. Currently, the treatment of tuberous sclerosis is symptomatic [9,21].

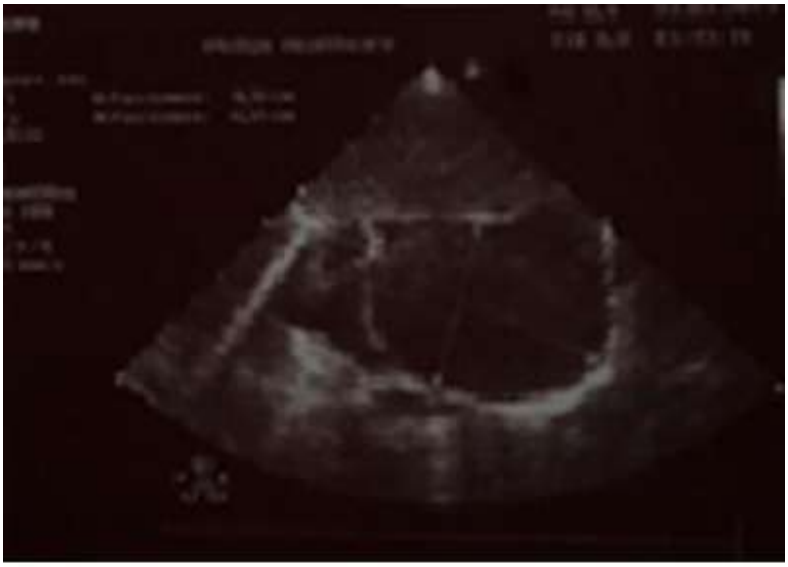
In most cases, rhabdomyomais first diagnosed, and tuberous sclerosis must be excluded in the dynamics of the examination.

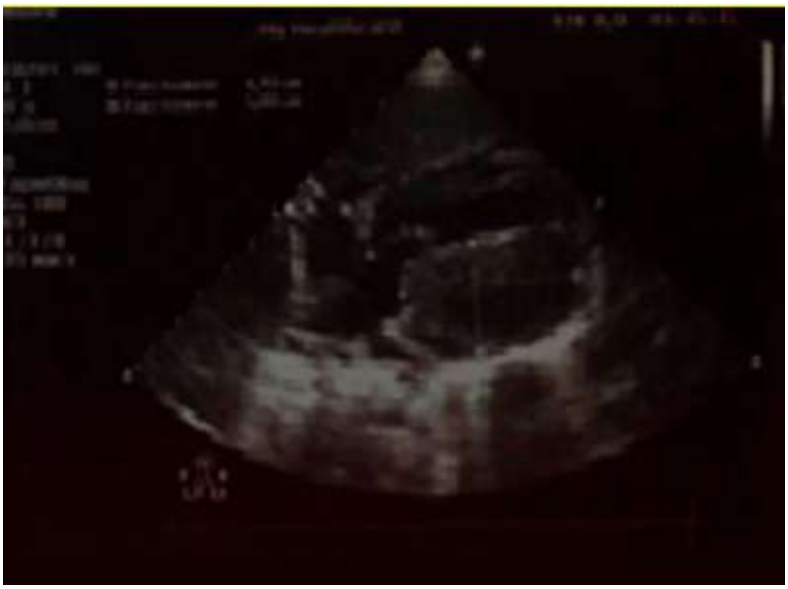
Here is a case from our practice, when a childborn in 2014 in the first year of life was admittedto a hospital with clinical signs of acute respiratory disease. In the anamnesis, the heredity is not burdened. There are two older children in the family, aged 7 and 4 years, and the children are healthy. Pregnancy of this child took place against thebackground of repeated respiratory infections in the mother,received treatment. These deliveries were on time, but delayed. The child was born in a state of asphyxia, to the breast was attached on the 2nd day. The newborn period proceeded smoothly, was regularly examined by a pediatrician and neurologist, and grew and developed according to age. At the age of 10 months, he was admitted to a hospital, where he was diagnosed with acute bronchitis, which was confirmed byR-gram of the chest, when an increase and changes in the heart were detected.



Chest radiographycan detect changes and increases in the size of the heart, stagnation in the small circle of blood circulation, but does not give complete information.

To clarify the diagnosis, the child underwent an echocardiographic study (EchoCG).





Echocardiogram:Acute formation in the left ventricle cavity:48x35mm (rhabdomyoma). The outline is even. The left ventricular cavity is reduced and the paradoxical movement of the interventricular septum.

Conclusion:

1. Formation in the cavity of the left ventricle (rhabdomyoma). To exclude tuberous sclerosis, the babywas examined in the Republican cancer centerwith ultrasound diagnostics.

On echotomograms-liver OVD of the right lobe-81mm, CCD of the left kidney-48mm, the contours are even. The parenchyma is homogeneous, fine-grained. Intraparenchymal formations are not visualized. The intrahepatic bile ducts are not dilated. The diameter of the liver veins is 2 mm. The portal vein is 6 mm. The gallbladder is reduced.The pancreas is not visualized due to gases.Spleen dimensions-54x22mm, smooth contours. The parenchyma is homogeneous, fine-grained.The right kidney is53x22mm, the topography is not changed, the contours are smooth and clear. Parenchyma of normal echogenicity (0 degree), 8-9 mm thick, without deformation, not expanded. There are no stones. Kidney function is normal. The parenchyma has not been changed. The right ureter is not dilated.The left kidney is 53x24mm, the topography is not changed. The contours are clear and even. Parenchyma of normal echogenicity(0 degree), 8-9 mm thick, without deformation, not expanded. There are no stones, the tour is normal. The parenchyma has not been changed. The left ureter is not dilated.The adrenal glands are not visualized.Paraaortic and paracaval lymph nodes are not visualized due to pneumatics in the intestine.

In the projection of the anterior mediastinum (thymus gland projection)an additional "echo shadow" with dimensions of 37x45x38 mm is visualized.

2. Mediastinal Tumor?

- The child isconsulted with a dermatologist, neuropathologist, and oculist for the necessary examinations.

- A previous study ruled out the presence of signs of tuberous sclerosis in the child.

- Repeat holdingEchoCGconfirmed the established diagnosis: rhabdomyoma of the heart.

- The child is referred for consultation and observation by cardiac surgeon with regular EchoCG every 3 to 6 months to monitor the growth of the tumor and determine for surgical treatment.

EchoCG allows you to determine the size, shape, clarify the intramuscular or intracavitary localization of the tumor, identify relationships with the valvular heart apparatus, monitor the dynamics of tumor growth, and determine indications for surgical treatment.

In the dynamics of follow-up for 3years by a cardiologist and cardiac surgeon, signs of regression of rhabdomyomawere noted.

3. Timely conduct of such research methods as EchoCG, if necessary MRI and CT,allow you to make a timely diagnosis of heart pathology. Early detectionofrhabdomyomaof the heart and дprenatal observation of a cardiologist and cardiac surgeon with symptomatic treatment will allow you to choose the right treatment strategy, since in some cases there isa regression of rhabdomyoma.

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