TUMORS OF THE HEART IN CHILDREN

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Abstract. *Rhabdomyoma of the heart refers to benign neoplasms, often diagnosed in the first year of life, can cause serious disorders in the heart. In some cases, rhabdomyoma of the heart is observed in tuberous sclerosis, in this regard, it is necessary to conduct a full examination of the child and timely prescribe treatment and regular monitoring. Early detection of rhabdomyoma of the heart and dynamic observation of a cardiologist and cardiac surgeon with symptomatic treatment will allow you to choose the right treatment strategy, since in some cases there is a regression of rhabdomyoma.*

Keywords: *rhabdomyoma of the heart, tuberous sclerosis*

Heart tumors are infrequent. According to sectional studies, heart tumors are detected in 0.0017-0.28% of all autopsies. The largest group of heart tumors is represented by benign, which occurs in 75% of cases of primary tumors, these are rhabdomyomas, fibroids, teratomas and myxomas. Malignant tumors are 3 times less common. These are usually sarcomas, of which angiosarcomas are more often recorded, mainly affecting boys, quickly and massively metastasizing through the blood vessels.

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].

We have our own data on the successful diagnosis cardiac rhabdomyoma, which is presented in this paper.

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 is most often found in the ventricular 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 may regress spontaneously, up to complete resorption within a few years [6] .

Rhabdomyoma can be diagnosed from the first months of life. It is formed from embryonic muscle cells as a result of early disembryogenesis 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:

- genetic predisposition,
- unfavorable 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.

The disease has no specific signs, usually the tumor grows slowly and does not cause any inconvenience. If the neoplasm is small in size, it may remain undetected for many years. If the tumor is large, it can seriously hinder the work of the heart and negatively affect the development

of the child. Symptoms such as heart failure, arrhythmia, tachycardia or bradycardia may occur, atrioventricular blockages and ventricular extrasystole may develop. Further progression of the neoplasm can cause serious disturbances 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/extramural growth. In rare cases, rhabdomyomas can be located in the atria, starting from the atrial septum. Rhabdomyoma of the heart, depending on the location, can have a malignant clinical course, leading to critical obstruction of the valve or ventricular outlet tract, damaging the cardiac conduction system, leading to death, including sudden death [8 – 10, 21 - 23].

Cardiac rhabdomyoma with a high frequency (up to 50-80%) is associated with tuberous sclerosis (TS) (Bourneville's disease), which is a genetically determined disease and is characterized by the development of multiplex benignx tumors in various organs, with a progressive course [8,9,10,11,9,10,11, 14-1–18]. In this case, rhabdomyoma may be the first sign of the disease, and a subsequent examination of the patient reveals a symptom complex of tuberous sclerosis. [12, 13, 19, 20], the diagnostic criteria of which are presented in Table 1.

Table 1. Diagnostic criteria for tuberous sclerosis (E. S. Roach et al., 1999)

Main (large) TS criteria	Additional (small) TS criteria
- Angiofibromatosis of the face (cheeks, back of the nose) or forehead in the form of spots (plaques — - Subungual fibroids of non-traumatic origin -Three or more hypopigmentation spots - Areas in the form of shagreen plaques -Multiple tumor-like nodules on the retina -Bumps in the cerebral cortex — Subependimal nodules — Subependimal giant cell astrocytoma — Rhabdomyoma of the heart (single or multiple) — Renal angiomyolipomas or pulmonary lymphangiomyomatosis	— Multiple randomly dispersed depressions (pits) on the enamel of teeth — Hamartoma polyps of the rectum — Bone cysts — Migration of white matter of the brain in the form of ray lines — Fibromatosis of the gums - Non-renal hamartomas - Unpainted (achromatic) spots on the retina - Skin manifestations in the form of confetti (small round spots — - Multiple kidney cysts

The diagnosis of tuberous sclerosis is considered reliable if the patient has two large or one large and two small criteria [9, 10,1-8].

As a rule, the following studies are carried out:

MRI of the head at least every 3 years to detect intracranial complications;

Ultrasound of the kidneys or MRI of the abdominal cavity every 3 years for school-age children and every 1-2 years for adults to detect kidney tumors

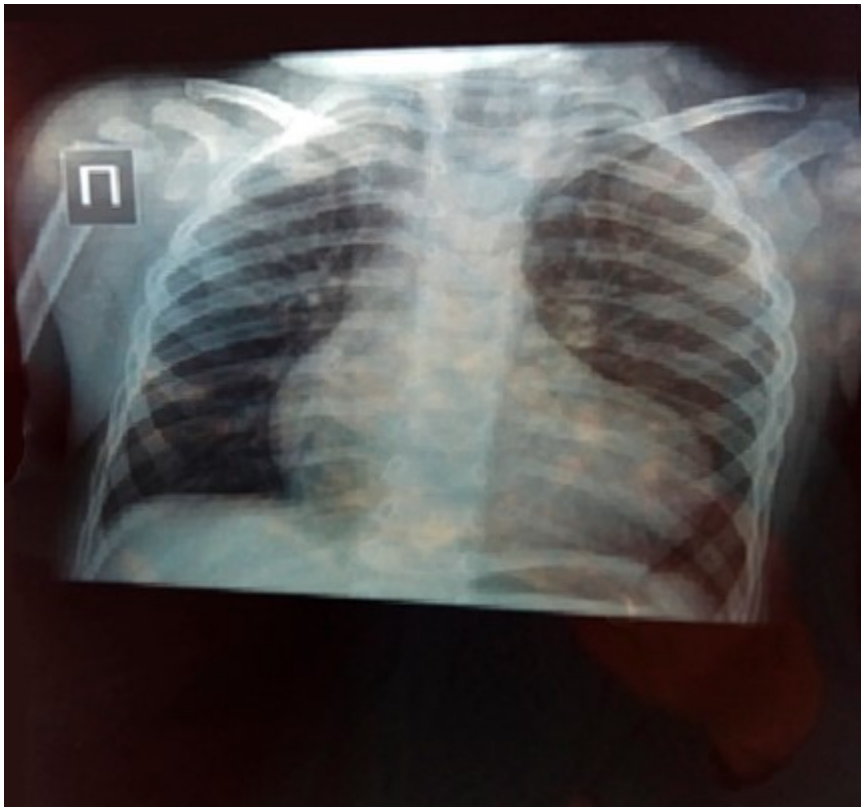
Girls ≥ 18 years of age should be screened annually for shortness of breath during exercise and at rest, as well as a high-resolution CT scan every 5-10 years

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 the symptoms. All patients should be examined regularly to detect possible complications of TSC in a timely manner. Currently, the treatment of tuberous sclerosis is symptomatic [9**,** 20**].**

In most cases, rhabdomyoma is initially diagnosed, and tuberous sclerosis should be excluded in the course of the examination.

Here is a case from our practice, when a child born in 2014 in the first year of life was admitted to the hospital with clinical signs of acute respiratory disease. In the anamnesis of heredity is not burdened. The family has two older children aged 7 and 4 years, the children are healthy. Pregnancy with this child occurred against the background of repeated respiratory infections in the mother, received treatment. These deliveries were on time, but prolonged. The child was born in a state of asphyxia, and was applied to the chest 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 by an R-gram of the chest, when an increase and changes in the heart were detected.



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

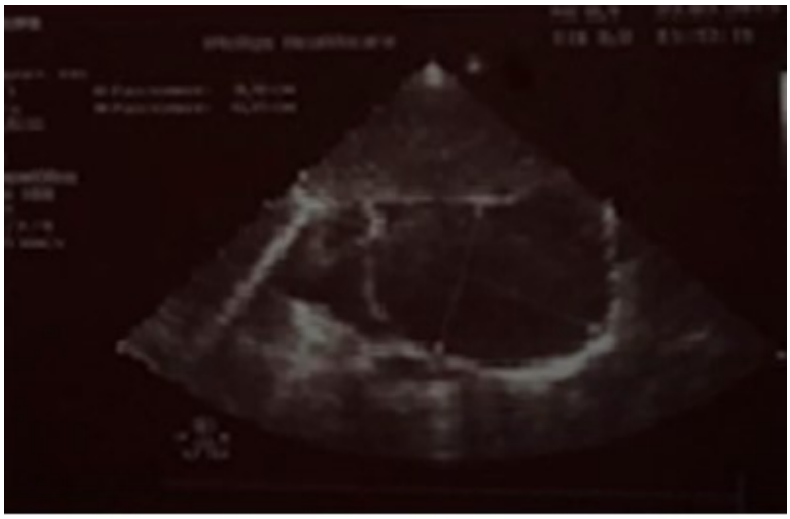
To clarify the diagnosis, the child underwent an echocardiographic examination (EchoCG).

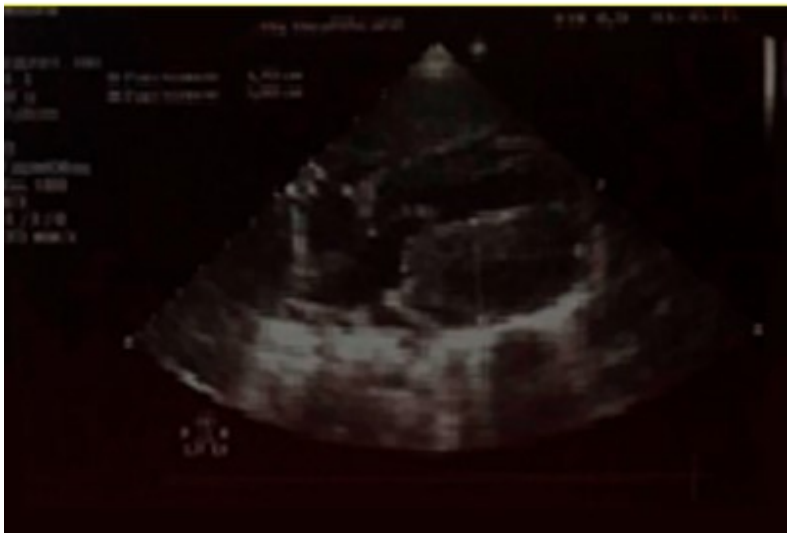
Echocardiography: Oral formation in the left ventricular cavity:48x35mm (rhabdomyoma). The contours are smooth. The left ventricular cavity is reduced and paradoxical movement of the interventricular septum.

Conclusion: Left ventricular cavity formation (rhabdomyoma)

To exclude tuberous sclerosis, the calf was examined at the Republican Cancer Center with ultrasound diagnostics.

On echotomograms-liver OVS of the right lobe-81mm, CCS of the left kidney-48mm, the contours are even. The parenchyma is homogeneous, dandpoechogenic, fine-grained. Intraparenchymal formations are not visualized. Intrahepatic bile ducts are not dilated. The diameter of the hepatic veins is 2 mm. Portal vein – 6 mm. The gallbladder is contracted.





The pancreas is not visible due to gas.

Spleen dimensions- 54x22mm, smooth contours. Parenchyma is homogeneous, fine-grained.

The right kidney is 53x22mm, the topography is not changed, the contours are smooth and clear. The parenchyma of normal echogenicity (grade 0), 8-9 mm thick, without deformation, is not expanded. There are no rocks. Kidney excursion is normal. Paranephritis is not changed. The visualization of the right ureter is not expanded.

The left kidney is 53x24mm, the topography has not been changed. The contours are clear, and even. The parenchyma of normal echogenicity (grade 0), 8-9 mm thick, without deformation, is not expanded. There are no stones, the tour is normal. Parenchyma is not changed. The upper left ureter is not dilated.

The adrenal glands are not visualized

Para-aortic, and paracaval lymph nodes are not visualized due to pneumatosis in the intestine.

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

Conclusion: Tumor Mediastinal tumor?

The child is consulted with a dermatologist, neurologist, or optometrist to conduct the necessary research.

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

Repeated echocardiography confirmed the established diagnosis: Cardiac rhabdomyoma.

The child is referred for consultation and follow-up by a cardiac surgeon with regular echocardiography every 3 to 6 months to monitor tumor growth and determine whether to perform surgical treatment.

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

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

Conclusion. Timely implementation of such research methods as echocardiography, if necessary, MRI and CT, allow you to make a timely diagnosis of heart pathology. Early detection of cardiac rhabdomyoma and follow-up by a cardiologist and cardiac surgeon with symptomatic treatment will allow you to choose the right treatment strategy, since in some cases there is a regression of rhabdomyoma.

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