

## A Case of Giant Intrapericardial Teratoma of the Heart in an Infant with a Favorable Outcome

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### 1. Abstract

Primary cardiac tumors in newborns and young children are rare, but they affect significantly their health and can be the cause of sudden death. These tumors compromise the integrity and function of adjacent cardiac structures, causing often impaired blood flow, causing myocardial dysfunction, valve failure, arrhythmias, respiratory changes, and cyanosis. We present a clinical case of an oversized intrapericardial teratoma diagnosed in a child aged 19 months, which ended in recovery. Proper diagnosis and surgical treatment saved the child from lethal outcome even in a case of a large tumor.

### 2. Introduction

According to the literature available, the overall prevalence of cardiac tumors reaches 0.14% of all fetal cardiovascular anomalies, and pericardial teratomas account for 9.5%–19% of all fetal primary cardiac tumors [1-3]. Teratoma is a tumor that develops from pluripotent germ cells of different tissues, whose localization being not specific for any the organ and anatomical region [4]. In appearance, it is an encapsulated lobular cystic mass filled with serous fluid. Such a tumor can contain derivatives of all three germinal sheets - nerve, cartilage, bone, muscle (smooth and transverse striated) and glandular tissue. Teratoma occurs in 1/30,000-70,000 live births [5]. Teratomas of rare localization - maxillofacial region, or epignathus, thyroid teratomas of the neck have been described [6-8]. More than 72% of teratoid tumors are localized in the sacrococcygeal region and ovaries, and only 5-6% having been

found in the thoracic cavity. Thoracic cavity teratomas are most frequently located at the base of large vessels of which 90% are in the anterior mediastinum [9]. According to some authors, teratoid tumors comprise 15-19% of all cardiac tumors [3,10]. Intrapericardial teratomas are extremely rare and described mostly as isolated cases [10-12]. These tumors arise from pericardium and contain multicellular layers of ecto-, endo-, and mesoderm of different maturity level. Primary cardiac tumors in children, are extremely rare and have been described in pediatric autopsies with a their on size frequency of 0.027% to 0.08% [13,14]. Clinical manifestations of intrapericardial tumors depend on the degree of compression of adjacent organs, vessels and nerves. Large tumors are a significant predictor of lethal outcome [1].

There are many scientific explanations for the origin of teratomas (from the Greek teratos-("miracle")). According to one of them, during pregnancy the fetus has abnormal development and is located inside another embryonic sheet, developing up to a certain period of development, and then degenerative changes occur in it. Another hypothesis explains the occurrence of teratomas by parthenogenetic development of fetal cells. The third theory of the of teratomas origin of associates it with the "remnants" of polypotent cells "forgotten" during the process of embryogenesis, possibly due to the violation of mechanisms of cell differentiation.

Teratomas are predominantly found in young people (20 to 40 years), and can be of different sizes, causing compression of anterior and posterior mediastinal organs. They occur very rarely in

young children. The main method of mediastinal teratomas diagnosing is radiological and spiral computed tomography with intravenous contrast. The only rational treatment for teratomas is their radical removal.

### 3. Clinical Case

Child, boy 19 months old, born from the second pregnancy. Mother (25 years old) and father (26 years old), are healthy. The first child – a girl (5 years old) is healthy. No hereditary diseases were observed in the family. There was no obstetric-gynecological anamnesis. Physiological delivery, the child scored 7 according to the Apgar scale at 1 minute and 8 at 5 minutes. The boy was discharged from the hospital on the 4th day. The child was on natural feeding. His neuro-psychological and physical development was adequate to his age. During the first year he had twice rhinopharyngitis. At the age of 19 months the child had a cough, confirmed as bronchitis. Prescribed treatment for bronchitis had no effect. The child was examined by physicians of different specialties and are recommended prescribed chest X-ray and echocardiography are recommended. Hydropericardium was detected, so the child was sent to the cardiac surgical department. On admission, the child's condition was severe, lethargic. Moderate dyspnea (up to 40 per minute), arterial blood oxygen saturation by pulse oximeter reached 93%, with heart rate 170 beats/min, blood pressure in the upper and lower extremities 85/45 and 90/53 mm Hg, respectively, were recorded. The lower edge of the liver protruded from under the edge of the rib arch by 4 cm along the midclavicular line. A chest X-ray showed a significant dilation of the mediastinum beyond the right edge heart shadow (CTI - 0.68) (Figure 1).



**Figure 1:** Chest radiography. Preoperative hydropericardium.

Transthoracic EchoCG revealed a volumetric mass of round shape with cystic elements, with no signs of blood flow, with the size 55x45x49 mm. The mass was located along the right heart border, extended to the vascular bundle area and enveloped the aorta, pushing the heart to the left. The pericardial sheets could not be clearly visualized. A large amount of fluid was detected in the pericardial cavity. The child was operated. Surgeons found giant teratoma and 300 ml of fluid in the pericardium. The teratoma was intimately connected with the great vessels, in particular, it squeezed the right ventricle of the heart (Figure 2).



**Figure 2:** Teratoma compressing the right ventricle of the cardiac. It is intimately connected to great vessels, in particular to the pulmonary artery.

The teratoma was successfully excised. When it was measured, it turned out to be quite large, 60x49x50 mm, and matched almost the echocardiography dimensions. Histological examination of the mass removed confirmed the diagnosis: pericardial cyst, teratoma (Figure 3). On the 15th day after surgery, the boy was discharged in satisfactory condition under the supervision of a cardiologist and a pediatrician. Signs of dilated cardiac contour and its compression on radiography disappeared. At present, three years after surgery, the child's physical and neuropsychiatric development is adequate to age (Figure 4).



**Figure 3:** Hollowed out teratoma.



**Figure 4:** Chest radiography. Twelve days after surgery.

#### 4. Discussion

Clinical manifestations of thoracic tumors are determined by their type of neoplasm, localization, and size. The majority of children with mediastinal teratomas are seen by a pediatrician for suspected pneumonia, bronchitis, thymomegaly [12]. Intrapericardial teratomas are very rare. A teratoma is a tumor that grows rapidly and causes almost always pericardial effusion. The first report of mediastinal teratoma in the literature, according to E. Arciniegas appeared in 1890 (by V.J.Joel) [11]. Excision of a cardiac tumor in a newborn with a body weight of 4010g at the 3rd week of life dates back to 1982. To date, more than 100 clinical cases of surgical treatment of intrapericardial teratoma have been described worldwide. The incidence of this localization is extremely rare and ranges from 0.0017 to 0.003% among alive born infants [10]. Intrapericardial teratomas are often missed during routine scanning of the second trimester of pregnancy because they tend to grow rapidly during the third trimester. Intrapericardial teratomas are somewhat more common in male than in female fetuses. This type

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of tumor is visualized during ultrasound examination as a large heterogeneous mass with dense hyper echogenic and cystic hypo echogenic areas [6].

Cardiac tumors can cause a variety of symptoms through the following four mechanisms:

1. The mass of the tumor interferes with intra cardiac blood flow or impairs valve function;
2. Intramural tumor localization can lead to arrhythmias or pericardial effusion with tamponade;
3. Emboles may detach from the tumor, causing systemic deficits if the tumor is located on the left side of the heart;
4. Tumors can cause general symptoms [10,13].

The peculiarity of our observation is that the clinical manifestations appeared at the age of 1 year 7 months and debuted with the coughing. Giant teratoma and pericardial effusion of 300 ml suggested cardiac tamponade. Successful surgical treatment made it possible to save the boy's life. We can assume that the child was born with an intrapericardial teratoma, which was apparently small and did not disturb hemodynamics. The increase in teratoma size was accompanied by fluid production in the pericardium. According to the literature available, immediately after the birth of a child with intrapericardial teratoma hemodynamic disorders are clinically manifested and the life can be saved very rarely, most often such cases end lethally. With a significant increase in the tumor size, the lethal outcome is inevitable [15]. In this case, successful surgical treatment made it possible to save the boy's life due to the high professionalism of cardiac surgeons.

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