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Pericardial Teratoma: Prenatal Diagnosis and Course

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Key Words
Pericardial tumor · Fetal echocardiography · Pericardiocentesis

Abstract
We report on a case of primary pericardial teratoma detected in a 29-week-old fetus. Due to cardiac decompensation, pericardiocentesis was performed at 33 weeks of gestation, and surgical excision of the tumor was indicated shortly after birth. The present report draws attention to the impact of fetal echocardiography on perinatal management.

Intrapericardial teratomas are rare primary cardiac tumors. They are usually benign, but may cause cardiac decompensation due to pericardial effusion and cardiac compression. Fetal echocardiography makes diagnosis feasible even in prenatal life. Cardiac tamponade and thus intrauterine death can be averted by fetal pericardiocentesis.

Case Report

A 29-year-old woman, gravida III, para II, was referred to our hospital after detection of a large thoracic mass in her 29-week-old fetus on the occasion of a routine ultrasound screening. Fetal echocardiography revealed a pericardial effusion. Adherent to the right atrium, but definitively outside of the cardiac cavities (fig. 1), there was a huge oval echogenic mass including some small cystic areas. The heart was structurally and functionally normal and showed a regular rhythm.

During the following 4 weeks the effusion increased in size and fetal ascites was identified as a sign of cardiac decompensation. Therefore, pericardiocentesis was performed under sonographic guidance. 45 ml of a serous-sanguinous fluid was withdrawn; detailed analysis showed no malignant cells.

Within the following days, the pericardial effusion developed again and the heart, together with the solid mass and the effusion, comprised more than half of the fetal thorax. Besides ascites, pleural effusions were detected. Cardiotocography showed signs of fetal distress and the fetus was considered to be at high risk for hydrops fetalis.

By consequence a cesarean section was performed at 36 weeks of gestation. The baby was immediately intubated (Apgar 5/8/9) and transferred to the intensive care unit. Two-dimensional echocardiography confirmed the diagnosis of pericardial effusion (maximal diameter 1.5 cm) and of a 5 cm × 5 cm × 5 cm solid multicystic mass attached to the right atrium (fig. 2, 3).

Due to increasing signs of cardiac decompensation cardiac surgery was performed within the next hours. After pericardiotomy and drainage of 30 ml pericardial effusion a large cystic tumor attached to

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the right atrium, the right ventricle and the ascending aorta presented. Each manipulation of the mass provoked bradycardia or hypotension under cardiopulmonary bypass. The tumor was completely excised without damage to the great vessels or other structures. Pathomorphological examination revealed endodermic, mesodermic and neuroectodermic germinal layers, corresponding to a benign immature teratoma grade I [1]. The postoperative course was uneventful.

At 3 years of age the baby is alive and well, neither echocardiographic and magnetic resonance imaging nor tumor markers (like α-fetoprotein and human chorionic gonadotropin) showed evidence of recurrence.

Discussion

Primary cardiac tumors are rare in childhood, with an incidence ranging from 0.06 to 0.32% according to publications in the eighties and the nineties, respectively [2]. Data from pediatric autopsy series before the introduction of echocardiography reveal much lower numbers. The changes in diagnostic and imaging practice over the years, therefore, seem to have increased the frequency of pediatric cardiac tumors.

Primary pericardial teratomas are even rarer. They are usually of benign origin, and, as extragonadal teratomas, are histopathologically classified and graded according to Gonzalez-Crussi [1].

Echocardiography is a reliable tool to identify cardiac teratomas even in infants and children: typical diagnostic

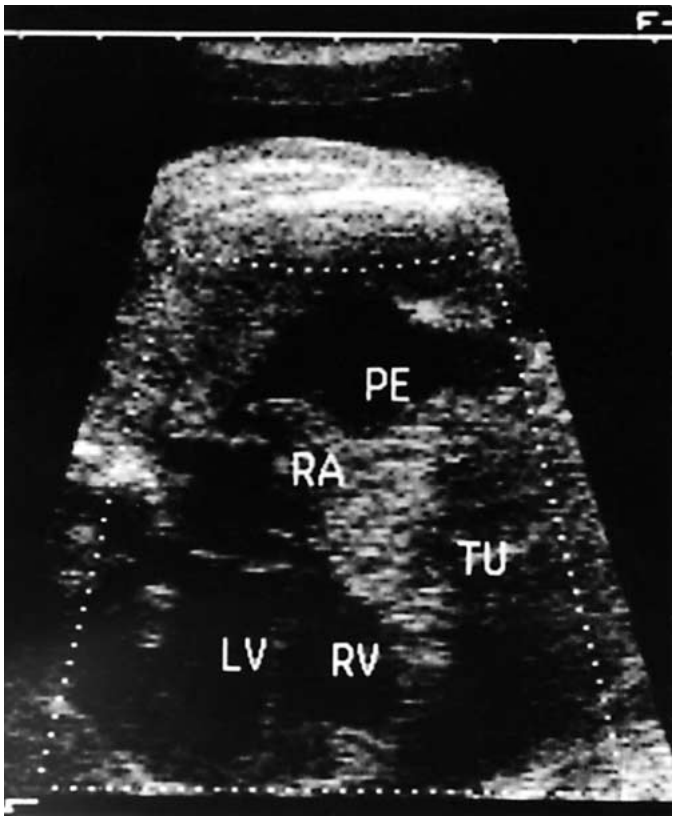

Fig. 1. Fetal scan at 29 weeks of gestation. Transverse thoracic section. A large tumorous mass (TU) seems to be adherent to the right atrium and right ventricle (RV). Huge pericardial effusion (PE). LV = Left ventricle.

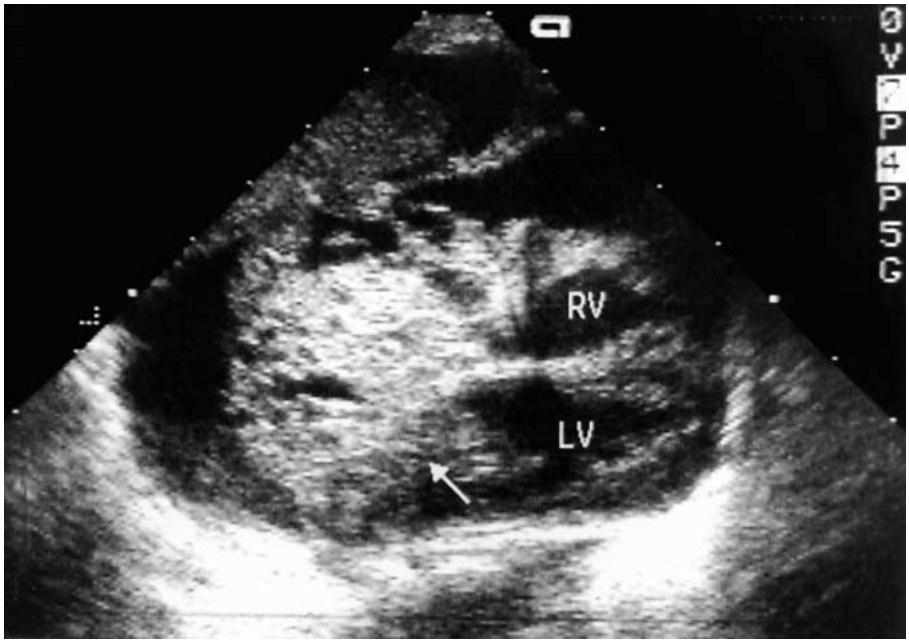

Fig. 2. Echocardiographic subxiphoidal four-chamber view immediately after birth. The heart is surrounded by pericardial effusion. The tumor (arrow) arises from the atria and reaches the atrioventricular level (RV = right ventricle, LV = left ventricle).

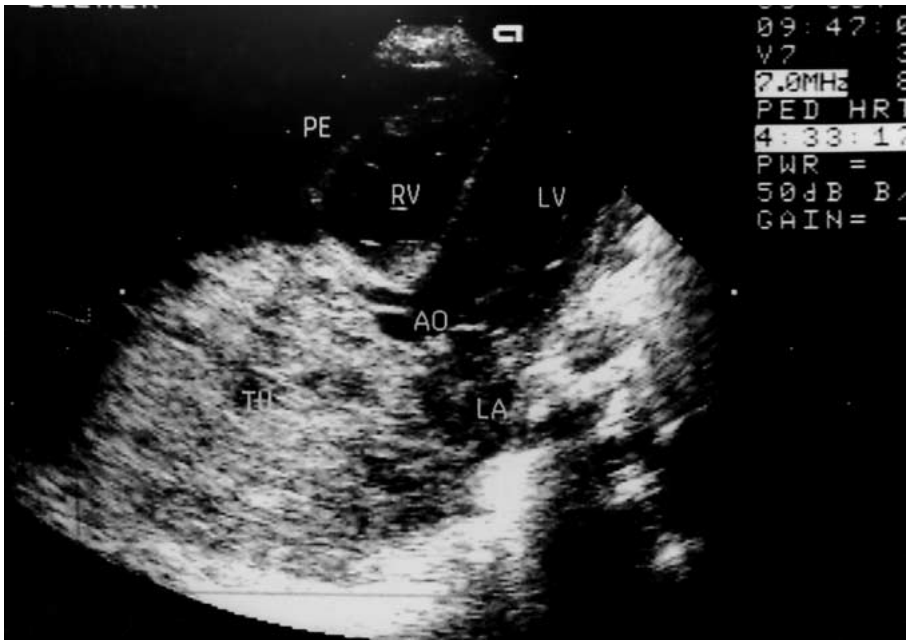

Fig. 3. Five-chamber view after birth: the tumor (TU) is also adherent to the ascending aorta (Ao). PE = Pericardial effusion; RV = right ventricle; LV = left ventricle; LA = left atrium.

ultrasonic findings are its multicystic nature, frequent location on the right anterior heart border, large size, and associated pericardial effusion [3]. The latter is presumed to be due to rupture of cystic areas or obstruction of cardiac and pericardial lymphatics [4]. Magnetic resonance imaging is particularly helpful to define the relationship of the tumor to normal structure, especially during the follow-up period.

Cardiac symptoms are more related to the size and location of the teratoma and the effusion than to the type of the tumor [2]. In the fetus, ascites, pleural effusions, right atrial and ventricular diastolic compression and finally generalized hydrops are characteristic signs of cardiac compromise.

Fetal echocardiography should be one of the first steps in these circumstances: cardiovascular malformations or tachyarrhythmias play an important role within the spectrum of nonimmunological hydrops fetalis. In case a fetal thoracic mass can be identified, differential diagnosis includes a variety of cardiac tumors (fibromas, hemangiomas, rhabdomyomas) and other thoracic malformations. Congenital cystic adenomatoid malformation and bronchogenic cysts must be considered if the tumor mass appears to originate from the lung [5]. Tollens et al. [6] reported a case of two independent primary teratomas of the thorax.

Of all reported cases of intrapericardial teratomas, to our knowledge, 15 cases [7] have been diagnosed during the fetal period and only 11 before the 30th week of gestation. Little is known about the exact time of appearance. In our case prenatal diagnosis was established in the 29th week of gestation after detection of a pericardial mass on the occasion of a routine ultrasound examination. One of the leading symptoms in this case was the pericardial effusion, leading to pericardiocentesis in the 33rd week of gestation after the size had increased and fetal hydrops was suspected.

Pericardiocentesis is a safe and effective method to remove cardiac compression, even in prenatal life. Since the first report of Benatar et al. [8] in a 34-week-old fetus 3 more cases of fetal pericardiocentesis in the setting of pericardial teratoma have been described [9–11], the earliest in a 24-week-old fetus [11]. In the latter, the measure had to be repeated 3 weeks later in order to deliver a nearterm baby at 35 weeks of gestation [11]. Thus, reaccumulation of the pericardial effusion is possible and can require serial pericardiocentesis to avoid prematurity with all its related complications. As in our case, there were no complications of pericardiocentesis in the reported fetuses and all infants had a favorable outcome after near-term delivery and early surgical intervention.

Surgical removal is the treatment of choice and is generally curative as the tumor is rarely malignant. Intracardial teratomas without the presence of severe heart failure or hydrops usually have an excellent prognosis [8]. How-

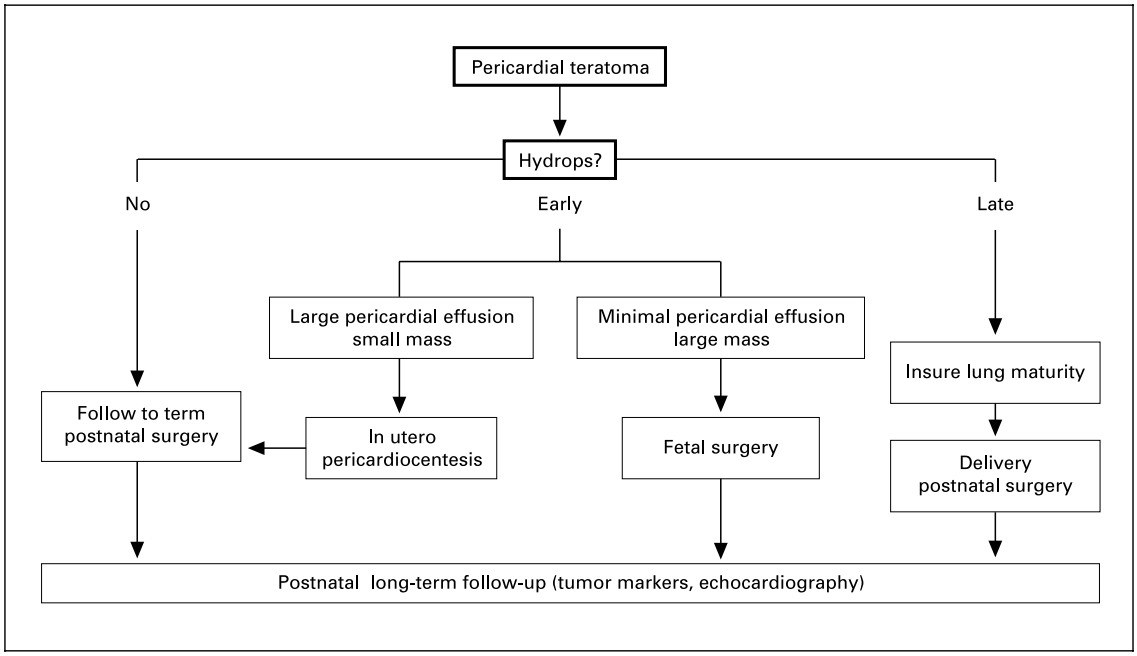

Fig. 4. Management algorithm for prenatally diagnosed pericardial teratoma [modified according to ref. 3].

ever, the overall outcome of prenatally diagnosed teratoma presenting with fetal hydrops is less favorable. Since the clinical course is strongly related to the development of cardiac decompensation and fetal hydrops, Bruch et al. [9] suggested a management algorithm (fig. 4) which should help to improve the outcome. Depending on gestational age and the development of fetal hydrops different strategies are suggested. If hydrops develops early in pregnancy, management depends on the size of the pericardial effusion and the tumor. For those with large pericardial effusion and small tumor mass pericardiocentesis is the therapeutic option. In case of minimal pericardial effusion but large tumor mass fetal surgery must be considered. If there is no hydrops or hydrops appears late in pregnancy, management depends on the maturity of the fetus.

According to this flow sheet, our management led to delivery of a nonhydropic near-term baby, which is well after successful operation in the neonatal period. Less well known, however, is the oncologic point of view: these infants deserve close follow-up. In case of incomplete tumor resection, which may be evident on histological examination only, the risk of malignancy is up to 50% [12]. Therefore, we recommend to expand the excellent flow sheet of Bruch et al. [9] into the postnatal period and to include this important point.

Fetal surgical intervention in the presence of intrapericardial teratoma, to our knowledge, has not been published yet. However, other conditions leading to nonimmune hydrops like sacrococcygeal teratoma, have already successfully undergone fetal surgery [13]. Even if there is a different pathomechanism in this setting, where a high cardiac output failure causes fetal cardiac compromise, the field of fetal surgery is likely to expand and may also be helpful in reducing the compressing effect of thoracic tumors.

In conclusion, fetal echocardiography allows very early detection of cardiac tumors and associated cardiac dysfunction. Serial echocardiographic assessments should be performed in order to plan obstetric, perinatal and operative management. Fetal pericardiocentesis is a therapeutic option which may help preventing fetal demise and premature delivery.

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