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*Case Report*

# Large Intrathoracic Teratomas in Neonates: Surgical Considerations

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**Abstract**

We report the case of 3 mediastinal teratomas, 1 intrapericardial immature and 2 mediastinal grade 0, diagnosed in the 38th week of gestation (intrapericardial immature) in the last stage of pregnancy (mature mediastinal). The surgical resection of the masses was successfully undertaken shortly after birth. The histological description of tumor after excision has reported immature multicystic teratoma Grade 2 of 3, with immature neural tissue and without yolk sac tumor cells (for the intrapericardial case) and mature tissue grade 0 large masses, with all tissues represented at histology (for the extrapericardial case). All neonates showed a favorable postoperative course after excision.

**Keywords:** Teratoma, neonate, heart, cardiothoracic surgery.

## Introduction

Cardiac intrapericardial teratomas are rare and usually benign tumors. They originate from within the pericardium, usually attached to the great vessels as pedunculated, solid and cystic masses. This kind of disease can be associated with non-immune hydrops fetalis, and a pericardial effusion is resent in almost all patients and should alert the clinicians to the possibility of a tumor[(10)]; it may lead to a life-threatening tamponade, and to symptoms related to the compression of the mass itself on the heart. We report a three case series of neonatal teratomas:

**1st Patient**: A case of large intrapericardial teratoma, compressing the heart and associated with pericardial effusion diagnosed in utero with ecocardiography, but asymptomatic. The neonate has successfully undergone surgical excision after birth.

**2nd Patient**: A newborn with large mediastinal teratoma diagnosed in utero in the last stage of pregnancy and operated on the 15th days of life, successfully.

**3rd Patient**: A 14 day old neonate of life, without prenatal diagnosis, undergone an operation for large mediastinal mass causing severe respiratory distress, without adverse events after operation, but with bilateral pleural effusion treated with 2 thoracentesis and with no sequelae after the drainage of pleural fluid in the next days of recovery.

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**Case Series**

Patient **n. 1** was referred to our department from Neonatology Intensive Care Unit with a huge extracardiac mass, diagnosed by echocardiography during a prenatal exam in the 38th week of the intrauterine life ***(image 1).***

The tumor was associated with pericardial effusion, mainly anterior. The foetus was absolutely asymptomatic. A female baby was delivered by a Cesarean section at the end of pregnancy. A two dimensional echocardiography showed a 38 x 39 mm intrapericardial multicystic roundshaped mass heart-diameter equivalent, and a large pericardial effusion. The tumor was attached to the anterior portion of the ascending aorta and compressed the right atrium, right ventricle and superior vena cava. The MRI confirmed the presence of a 38 mm diameter intrapericardial mass attached to the ascending aorta, compressing the superior cava vein and the right ventricle, and a large pericardial effusion as well. At the age of 7 days (3,2 Kgs weight), a complete surgical excision was successfully performed. Afterperforming median sternotomy and pericardiotomy, the tumor showed a base of implant of 1,5 $cm^2$ on the antero-lateral wall of the ascending aorta; it layed on the right atrium and right ventricle. The pedicle of the tumor was clamped and the tumor was completely resected. The aortic adventitia surrounding the pedicle was also resected ***(image 2)***. The fine histological study on the mass after excision has shown a great variety of tissues, including not only immature neural, but also mature mesodermal and endodermal, and confirmed the diagnosis of immature teratoma. The clinical outcome postoperatively was regular. The patient was estubated on the first day after the operation, and remained symptom-free through the period of in - hospital recovery. The one-month follow up confirmed the freedom from signs of clinical relevance in the patient. After three months of life, the growth of the baby has been regular and symptom-free.

Patient **n. 2** was a neonate born from a Cesarean cut at the end of pregnancy, with an intrauterine diagnosis of a mediastinal large extrapericardial mass by echography, and with a CT scan pattern **(image 3 and 4**) to be likely a teratoma. The patient was asymptomatic and without haemodynamic adverse events before and after birth. Surgical excision has been performed on day 15 of life, in cervico–sternotomy without any mechanical assistance, and with complete success. The patient has been discharged from hospital 7 days after the operation. The histological exam has revealed a mature multicystic teratoma grade 0 (**image 7**).

Patient **n. 3** has been referred to our centre without a prenatal diagnosis of intramediastinal extrapericardial mass, and with a severe pulmonary distress immediately after birth (termed neonate without Cesarean cut). A mechanical mandatory ventilation following recovery in neonatal I.C.U was necessary, and an urgent surgical approach was planned after a chest X- ray test (**image 5**). The surgical approach was the same, in cervico-sternotomy. Surgical excision was completed and with no collateral events: a chilotorax was observed in both pleural spaces and was drained during the operation (**image 6**). The histological examination has showed a mature multicystic teratoma grade 0 (**image 8**). In the first 6 days after the operation, a bilateral pleural effusion was observed in the patient and was treated with 2 thoracentesis. Discharge of the patient from hospital was completed in 15 days after operation, without any other adverse event.

**Comment**

Cardiac (mainly intrapericardial) teratomas are a rare form of congenital tumor, are often benign and occur more frequently than extracardiac mediastinal forms. Teratoma with a malignant somatic component (TMSC) is rare but described in adults, whereas information on pediatric presentation is sparse[12]. Malignant forms make 15%, according to Dehner, in the pediatric age

group of mediastinal teratomas. Teratomas with yolk sac tumor are rarely documented, and its presence in histological samples worsen the prognosis of this kind of disease[10] . During the intrauterine life, their diagnosis is established by two- dimensional echocardiography, necessary to delineate the tumor, reveal pericardial effusion and point out hydrops fetalis[1-3]. They usually appear like calcificated and multicystic masses that could compress the heart, leading to congestive failure, whilst the pericardial effusion could cause cardiac tamponade. In case n. 1, diagnosis was performed in the 38th week of intrauterine life. The foetus was absolutely free of symptoms and was delivered with a Cesarean section. In our personal experience, the resection of the **intrapericardial immature** tumor without extracorporeal circulation, despite a gross dislocation of the right chambers and superior cava vein, together with a strict connection to the ascending aorta, couldn't be achieved more easily.

Later, a fine histological analysis on the mass showed that the tumor was encapsulated and the cut surface was predominantly solid with small cysts. The solid areas were soft, fleshy with focal haemorrhage and necrosis. Neural immature tissue occupied large areas and consisted not only of glial elements and coryoid plexus-like structures, but also neuroepithelium with tubules, rosettes and retinal anlage. A mature tissue of mesodermal and endodermal origin was also present. Immature teratomas are classically graded 1 to 3 on the base of the increasing amount of immature, usually neural, tissue. This case is classified as grade 2 over 3 immature teratoma, with absence of yolk sac tumor [10], since a moderate quantity of immature tissues was present in fewer than three low power field (LPF) per slide. In conclusion, in case n. 1, we present an asymptomatic neonate despite the presence of a large intrapericardial teratoma compressing the right atrium and the right ventricle, pedunculated to the ascending aorta. Prenatal diagnosis seems to be essential in the planning of fetal clinical management, delivery and surgical decision. Altough prenatal diagnosis has been delayed to the 38th week, compared with other early cases, the clinical and surgical management has been easy due to favorable hemodynamics and absence of hydrops fetalis[6-7-8]. It remains unclear, in our opinion, the reasons why a diagnosis is made too late at the end of pregnancy; for our statement, it could be better to effort the surgical strategy, to get a stronger relation between the perinatal echocardiographist and neonatology unit, to let the baby undergo the operation earlier than in our experience, regardless of symptoms and haemodynamycs conditions[11].

Once excised, intrapericardial mediastinal teratomas seldom recur; if resectable, extracardiac forms are 90% cured (s. 97% of intracardiac forms)[6-10]. So, in our case, prognosis probably seems to be excellent.

In cases 2 and 3, tumoral lesions were benign, and both grade 0, with an excellent prognosis in the long term. Surgical procedure was safe and easy to drive, being teratoma extrapericardial, despite the fact that in case 2 the respiratory distress has complicated the course of clinical algorithm in the early hours of extrauterine life. Even in cases 2 and 3, a median sternotomy has been the surgical access of choice, enlarged to cervicotomy due to the enormous dimension of tumors[11]. Only in case 3, dislocation of lungs and thracoea has been a trigger for the respiratory distress syndrome observed afterbirth, meanwhile in case 2 the compression of the heart was asymptomatic and the extrapericardial location was safer, compared with case 1, because the lower risk of bleeding and the "no" relation with gross vessel like the aorta complicates the surgical procedure.

In conclusion, the experience of our pediatric centre with this kind of tumoral disease has been successful. In all three cases the surgical procedure has been safe, complete and fast[12]; mechanical circulatory support by extracorporeal circulation has been delayed, especially in case 1, where compression of the right heart and aorta was evident, and the teratoma itself originated from the ascending aorta by a pedicle which probably supported the circulation and growing of the mass as well. All 3 patients have been discharged within 20 days after procedure without symptoms, in very good

condition and with excellent prognosis, especially in cases 2 and 3.

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