

fractionation of sequestered blood allowed salvage of platelets and clotting factors.

Exchange transfusion is a mainstay of CPB in patients with sickle cell disease. Most reports detail management of pediatric patients with HbSS and acyanotic disease [5]. Law and colleagues [6] reported a child with tricuspid atresia who survived a bidirectional Glenn procedure with exchange transfusion to reduce HbS and HbC levels to 3% each; however, simple exchange does not address the dilutional coagulopathy induced by exchange transfusion. In children and adults, reports of exchange transfusion followed by component separation and retransfusion of platelets and plasma suggest an advantage in reducing or avoiding non-red-cell transfusion in the perioperative period [7, 8].

Our experience indicates that careful planning and attention to management during the entire perioperative period allow CPB to be safely performed in a child with cyanotic disease and HbSC hemoglobinopathy as an alternative to an off-pump repair.

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Uncommon Multicystic Lesion of the Interventricular Septum in a 7-Year-Old Boy: Unusual Presentation of an Intracardiac Teratoma

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Intracardiac teratomas are very rare primary cardiac tumors; only a few cases have been reported. We present the case of a 7-year-old boy who early in life showed

pulmonary stenosis and needed percutaneous and surgical procedures, including sectioning of the right ventricular bands and reconstruction of the right ventricular outflow tract. At the age of 7 years the patient received a diagnosis of a multilobular cystic mass in the right ventricle adherent to the interventricular septum, which was not present at birth. Successful surgical resection was performed. Histologic examination revealed a mature teratoma. We emphasize the differential diagnosis of teratomas in cystic lesions of the interventricular septum.

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Multicystic lesions of the heart are rare, and the differential diagnosis usually includes hydatid disease, hemangioma, aneurysms of the membranous septum, endocarditis, bronchogenic cysts, and cardiac tumors [1]. Cardiac teratomas are very rare primary cardiac tumors, and within this group, teratomas of the pericardium are far more frequent than true intracardiac teratomas, with only a few cases reported in the literature [1, 2]. Clinical presentations can be variable and depend on the actual location of the tumor, but they usually include one or more symptoms of the classic triad (intracardiac obstruction, systemic embolization, and systemic or constitutional symptoms) [3]. Sudden death in asymptomatic patients is rare [4]. Benign intracardiac teratomas are multicystic masses with a tendency to originate from the right interventricular septum. The only effective treatment is surgical excision [1, 3, 5].

A 7-year-old boy had evidenced pulmonary stenosis 3 days after his birth, when a percutaneous dilation of the pulmonary valve was performed in our hospital. Because of continuous cyanosis, a modified Blalock-Taussig Shunt (MBTS) with the use of a polytetrafluoroethylene tube 4 mm in diameter was performed on the right side. In a second operation, when the child was 1 year old, sectioning of the right ventricular bands and reconstruction of the right ventricular outflow tract (RVOT) with a pericardium patch was successfully performed. The branches of the pulmonary artery were of normal caliber. The MBTS was ligated.

Until the child was 6 years old, no significant issues were reported, but then, owing to continuous fatigue, the patient returned to Portugal, when transthoracic echocardiography was performed and showed a multilobular cystic mass measuring 38 × 43 mm in the right ventricle, adherent to the interventricular septum, causing obstruction from the right ventricular entrance to the RVOT. Echocardiographic imaging suggested a diagnosis of cardiac hydatid disease. An available echocardiographic control examination when the child

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was 4 years old had revealed normal results without suspicion of intracardiac cysts. Computed tomography (CT) and magnetic resonance imaging (MRI) were then performed and confirmed a multisepted cystic formation situated superior and anterior to the interventricular sulcus (6×3 cm), with calcifications at the marginal borders occupying the lumen of the right ventricle with adherence to the interventricular septum (Fig 1). The CT and MRI findings also suggested intracardiac hydatid disease. Cranial, thoracic, and abdominal CT showed no other cystic formations. The results of hemogram, electrophoresis, and clinical chemistry (including C-reactive protein) were normal. The results of antiechinococcal antibodies titer and of serologic analysis for hepatitis B and C and human immunodeficiency virus 1 and 2 were all negative.

In a medical-surgical meeting, the decision was made to start antiparasitic therapy with albendazole (200 mg twice a day) and prednisolone (50 mg once a day) for 1 month and then schedule an elective operation. Elective reoperation through a median sternotomy and with the patient under extracorporeal circulation with central aortic and bicaval cannulation was performed. For cystic mass removal, the heart was arrested and the pericardium patch over the RVOT was opened. Careful dissection from the intracardiac structures with electrocautery and scissors allowed total resection of the mass (Fig 2). The interventricular septum, tricuspid valve, and RVOT were left intact. The pericardial patch incision was closed with a continuous suture. The postoperative course in the intensive care unit was uneventful. Histopathologic examination revealed a mature teratoma containing elements of all three germ layers (endoderm, mesoderm, and ectoderm) with predominantly cystic elements. The levels of α -fetoprotein and β -human chorionic gonadotropin 3 weeks after the operation were within normal range.

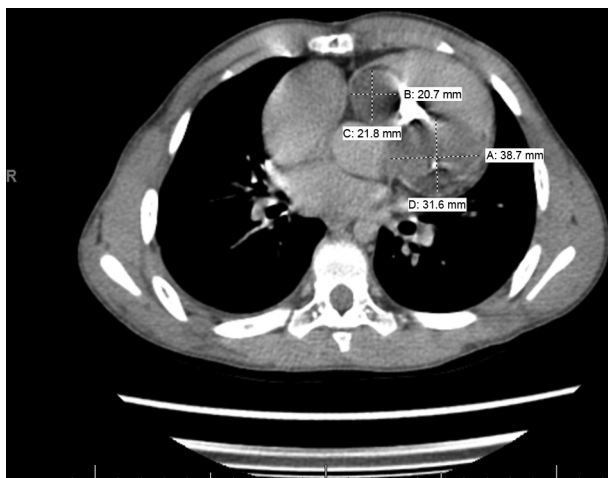


Fig 1. Axial view of computed tomographic image of the chest showing the dimensions of a multicyclic mass in the right ventricle.

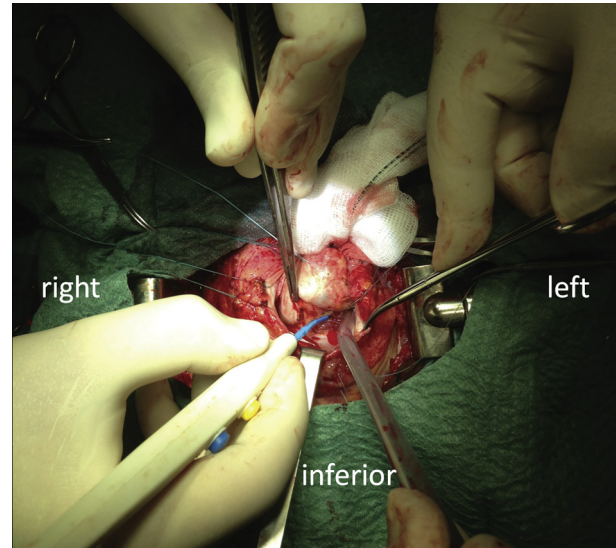


Fig 2. Intraoperative dissection of the tumor. The intracardiac multilobular cystic teratoma was exposed through the pericardium patch and dissected carefully from the interventricular septum with electrocautery and scissors.

Comment

Considering the previous operation and the patient's history and background, the most probable diagnosis was hydatid disease. Echocardiography with additional computed tomography and magnetic resonance imaging usually confirms the diagnosis. In studies, antiparasitic therapy with albendazole for 4 to 8 weeks before operation has been shown to reduce metastatic spread before an elective operation [6].

In other cases, intracardiac teratomas usually present in neonates or infants with RVOT obstruction. We describe a very rare case of an intracardiac teratoma that was not present at birth or at least not detectable by echocardiography. The initial diagnosis after birth was pulmonary stenosis with the necessity for various procedures. When the child was 1 year old, the right ventricle and RVOT were inspected in a second operation but no cystic lesions were present. Follow-up studies with echocardiographic control examinations were then regularly performed and never showed any suggestive structures along the interventricular septum (not visible in the last control examination when the child was 4 years old). The histologic examination showed a mature teratoma, which must have started growing to a detectable size only after the child was 4 years old. The teratoma then probably grew quickly to its immense size, which has also been described in the literature [1].

Although intracardiac teratomas are very rare, they should be considered in the diagnosis of a cystic mass in the right cardiac cavities, even in older individuals. Curative treatment can be achieved by total surgical excision. Long-term follow-up with regular cardiac imaging is strongly recommended, because

recurrence of intracardiac teratomas has been occasionally reported [7].

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Crossed Pulmonary Arteries in a Patient With Persistent Truncus Arteriosus

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We report a 14-month-old child with persistent truncus arteriosus and crossed pulmonary arteries. The potential advantage of crossed pulmonary artery arrangement in achieving surgical correction is discussed.

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Persistent truncus arteriosus (PTA) is a rare anomaly accounting for 2% to 3% of all congenital heart defects. It is commonly associated with ventricular septal defect (VSD), truncal valve dysplasia or dysfunction, and aortic arch anomalies. Crossed pulmonary arteries (PAs) are a rare form of congenital anomaly wherein both PAs cross each other while coursing toward their respective lungs. This association of PTA and crossed PAs is extremely rare. We present

the morphologic findings in 1 such patient with details of surgical management.

A 14-month-old infant girl weighing 4.5 kg presented with recurrent episodes of lower respiratory tract infections and rapid breathing while feeding. The pertinent clinical findings included a displaced apical impulse in the left sixth intercostal space lateral to the midclavicular line, single second heart sound, and a grade II/VI systolic murmur in the left upper sternal border.

A chest roentgenogram showed cardiomegaly with a cardiothoracic ratio of 0.70 and a left ventricular type of apex. A transthoracic echocardiogram confirmed type II PTA, a large malaligned subtruncal VSD, and severe pulmonary arterial hypertension. Although the PAs were seen arising separately, the anatomy of the left PA (LPA) was not clear. The truncal valve was tricuspid without stenosis or regurgitation. Computed tomographic (CT) angiography showed both PAs originating separately from the left side of the PTA. The right PA (RPA) arose inferior relative to the LPA and ran leftward before running toward the right lung. The LPA thus crossed the origin of the RPA (Fig 1).

The operation was performed through a standard median sternotomy. After the pericardium was opened, a PTA was noted giving rise to the aortic arch and its branches. The RPA was around 12 mm in diameter, arose from the left lateral aspect of the PTA just above the sinotubular junction, and was seen coursing leftward before running behind the ascending aorta to reach the right lung. The LPA, which was around 8 mm in diameter arose separately from the PTA just above the origin of the RPA (Fig 2).

The aorta, LPA, and RPA were dissected and mobilized. Standard cardiopulmonary bypass (CPB) was established with aortobicaval cannulation and core cooling to 28°C. Vascular loops were tightened around the RPA and LPA. A C clamp was placed on the LPA close to its origin. It was then divided, and the truncal end was oversewn. Similarly, another clamp was placed on the RPA close to its origin, it was divided, and the truncal end was oversewn. The LPA was now implanted on the greater curvature of the RPA in end-to-side fashion. A 22 mm-pulmonary homograft was then downsized to a 14-mm bicuspid conduit. The distal end of this homograft was anastomosed to the divided end of the RPA. After the conduit had been anastomosed to the RPA, the aorta was cross-clamped and cold blood cardioplegia was delivered into the aortic root. A right ventriculotomy was made, and the large subtruncal VSD was closed, followed by anastomosis of the proximal end of the pulmonary homograft to the right ventriculotomy. The remainder of the operation was completed in the usual fashion, and the patient was uneventfully weaned from CPB receiving elective inotropic support of dobutamine 10 µg/kg/min and milrinone 0.5 µg/kg/min. The aortic cross-clamp time was 53 minutes, and the CPB time was 131 minutes. Sildenafil was administered through the nasogastric tube postoperatively in view of the patient's delayed presentation and elevated

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