Primary cardiac tumours: when is surgery necessary?

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Abstract

Objective: Primary cardiac tumours are rare. The literature predominantly contains series on myxomas in adults and only a few long-term series that involve the very different primary cardiac tumours in early childhood. As foetal ultrasonography has continued to improve, cardiac tumours are increasingly detected early before significant symptoms develop. It is a challenge for paediatric cardiologists and surgeons to ascertain which patients need surgery and which will benefit from conservative follow-up. Methods: A retrospective review of a 10-year period revealed 51 tumours in 26 children (median age: 1 month). Analysis was by presentation, location, associated findings, interventions, histological findings, and clinical course. Results: The most common tumours were rhabdomyomas (29), fibromas (nine), teratomas (two), and haemangiomas (two). The tumour location was the right ventricle in 24 and the left ventricle in 22 patients. The symptoms varied between abnormal heart murmur (20), arrhythmia and conduction abnormalities (ten), obstruction of the outflow tract >30 mmHg (nine), severe cyanosis (three) and congestive heart failure (two). Fourteen children with haemodynamic compromises underwent surgery. There was one post-operative death and one heart transplantation after bridging with an assist device. There was no tumour recurrence even when resection was incomplete. Nine of 13 children with rhabdomyomas had spontaneous tumour regression without intervention. Conclusions: Most of the cardiac tumours in children are benign. Spontaneous regression is possible not only in rhabdomyoma. Surgical intervention is only required for children who develop relevant clinical symptoms. Total resection of the tumour is not the only therapeutic aim; more important is the restoration of the best possible heart function. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Cardiac tumour; Children; Rhabdomyoma; Fibroma; Teratoma

1. Introduction

Cardiac tumours are rare at all ages and are even less common in infants and children. The literature mostly contains myxoma series in adult patients and only a few long-term observations that involve the different primary cardiac tumours in children [1–3]. The tumours differ in symptoms, histology, and therapy in infancy compared with adulthood. The symptoms depend upon the location and are usually due to obstruction or compression of cardiac cavities [4,5]. However, an increased incidence of diagnosed primary cardiac tumours has been reported since the improvement of non-invasive imaging modalities [6]. As echocardiographic techniques, such as foetal ultrasonography, have continued to improve, cardiac tumours are increasingly detected early, before significant symptoms develop, challenging paediatric cardiologists and surgeons to ascertain which patients need surgery and which will benefit from conservative follow-up [7–10]. Most knowledge is based on collections of case reports. So the objective of this study is to review a large collective of primary cardiac tumours over the past 10 years and stress the changes in management and diagnostics. We also want to emphasize that pediatric cardiac tumours differ entirely from adult myxomas. Last not least we want to describe several remarkable cases mimicking other cardiac or pulmonary diseases.

2. Patients and methods

We reviewed 51 cardiac neoplasms in 26 children (15 boys/11 girls), between 1991 and 2000. Patients with mediastinal tumour and adults with myxomas were excluded. The ages ranged from 1 day to 17 years (median: 1 month).
There were four asymptomatic patients, four cases were diagnosed by echocardiography after routine postnatal auscultation, and six children were diagnosed prenatally.

Six patients underwent cardiac catheterization, which included one radiofrequency ablation because of arrhythmias. In two cases we found abnormal coronary arteries, one narrowing of the left coronary artery because of stretching of the artery, and one tumour-related stenosis of the circumflex artery. One child with a haemangioma at the posterior leaflet of the tricuspid valve had lived near Chernobyl during the nuclear catastrophe.

### 3. Results

Fifty-one cardiac tumours were found in 26 patients. The location of these tumours was 67% in the right ventricle, 43% in the left ventricle, 8% in the right and 2% in the left atrium.

Rhabdomyomas were the most common tumour (29 tumours in 13 patients), followed by fibromas (eight single and one patient with two fibromas). A total of 14/26 patients underwent surgical treatment. They all had clinical symptoms, as there were severe obstruction of the outflow tracts \( (n = 7) \), inflow tract obstruction with cyanosis \( (n = 2) \), rhythm disorder \( (n = 2) \), pericardial effusion \( (n = 1) \) and two cases of teratoma (Table 1). Ten patients had complete and four partial tumour resection. In one patient with congenital aortic regurgitation aortic homograft implantation and mitral valve reconstruction was performed. In another child VSD closure, in one VSD and ASD closure and in three patients PFO closure was performed during surgery. An immediate, symptom-free status was achieved in all surgical patients. Post-operative complications included bleeding in one patient, one transient low output state and one failure of weaning from bypass in a 5-month-old girl (case 3, Table 1) with bridging to transplantation with an assist device. There was one perioperative death in a 6-month-old boy with 3 \( \times \) 4 cm fibroma with subcomplete obstruction of the right ventricle filling the cavium of the right ventricle and leading to compression of the right coronary artery. The child died a few hours after weaning from bypass because of severe right heart failure.

Twelve out of 26 patients have not undergone surgery to date. They are free from tumour-related symptoms. In 11 patients the tumours were classified as rhabdomyomas, tuberous sclerosis was diagnosed in six of them. In one patient with neurofibromatosis two solide tumours were classified as fibromas. Follow-up echocardiography showed a diminishing tumour mass in nine and a constant mass in three of these children.

The follow-up period was 4.1 years and ranged from 5 months to 10 years. One child was lost to follow-up. One child with severe regurgitation required re-replacement of a bioprosthetic aortic valve and replacement of the mitral valve. One infant received medication for Wolff–Parkinson–White syndrome. All survivors were free of tumour related symptoms and tumour recurrence or progression, even when the resection was incomplete. Eight rhabdomyomas vanished totally without surgery; one fibroma vanished completely 1 year after subtotal resection.

We would like to emphasise three remarkable cases in our series:

1. In a 2-month-old boy (case 2, Table 1) a fibroma mimicked pulmonary disease or cyanotic heart disease by recurrent attacks of cyanosis. Pulmonary disease was excluded. Echocardiography showed a 3 \( \times \) 2.5 cm pediculated tumour in the right atrium with intermittent obstruction of the tricuspid valve. Because of an atrial septal defect, there was a variable right-to-left shunt with a transcutaneous oxygen saturation of between 55 and 100%, which normalised after corrective surgery.

2. The second patient (case 6, Table 1) was admitted to our hospital because of medically uncontrollable ventricular tachyarrhythmia. Catheterization for electrophysiological study with radiofrequency ablation had been undertaken without benefit. Coronary angiography showed a tumour-related stenosis of the ramus circumflex, which stretched the artery so that there was no systolic blood flow and a reduced diastolic blood flow that may explain the severe ventricular arrhythmias. After an uneventful partial resection of the fibroma from the left lateral wall of the ventricle, the arrhythmias immediately disappeared.

3. The third patient (case 3, Table 1) presented a few days after birth with a heart murmur. A large mass that infiltrated most of the left ventricular wall was found by echocardiography. The child was asymptomatic. During the first weeks the tumour became somewhat smaller, but then the fibroma grew and developed calcifications and necrotic foci (Fig. 1). At the age of 5 months the girl with a bodyweight of 5.6 kg developed left heart failure and underwent cardiac surgery. The tumour occupied more than 60% of the left ventricle and replaced the functional muscle mass. Due to the large size of the fibroma, its complete removal was impossible. After subtotal removal, weaning from bypass failed. She was listed for transplantation. Heart transplantation with an uneventful course followed after 17 days of bridging on a biventricular assist device (Berlin Heart®, Germany). The child was discharged home 3 weeks later. Follow-up has been uneventful so far for 20 months.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at diagnosis</th>
<th>Sex</th>
<th>Weight (kg)</th>
<th>Tumour subtype</th>
<th>No. of tumors</th>
<th>Localisation</th>
<th>Size (cm)</th>
<th>Symptoms</th>
<th>ECG</th>
<th>Vitium cordis</th>
<th>Surgery</th>
<th>Outcome after surgery</th>
<th>Follow-up</th>
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<td>1</td>
<td>1 month</td>
<td>M</td>
<td>3</td>
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<td>1</td>
<td>LV</td>
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<td>VES</td>
<td>–</td>
<td>Septal resection</td>
<td>Alive</td>
<td>TU vanished AVR + MVR</td>
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<tr>
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<td>Cyanosis</td>
<td>–</td>
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<td>No symptoms 1 year</td>
</tr>
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<td>–</td>
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<td>Uneventful</td>
<td>20 month</td>
</tr>
<tr>
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<td>6</td>
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<td>1</td>
<td>RV</td>
<td>3 x 3.5</td>
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<td>–</td>
<td>–</td>
<td>Resection</td>
<td>Alive</td>
<td>Lost to follow-up</td>
</tr>
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<td>7</td>
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<td>1</td>
<td>RV</td>
<td>3 x 4</td>
<td>Cyanosis</td>
<td>Repol. disorder, –</td>
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<td>Died</td>
<td>–</td>
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<td>VT, resuscitation, VT, VES</td>
<td>–</td>
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<td>TU totally vanished</td>
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<td>8 x 8</td>
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<td>SVT</td>
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</tr>
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<td>–</td>
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<td>3</td>
<td>LA, RA</td>
<td>1.4 x 1.3</td>
<td>SVT</td>
<td>WPW</td>
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<td>–</td>
<td>–</td>
<td>2 vanished, 1 smaller</td>
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<td>LVOT</td>
<td>1 x 1</td>
<td>LVOTO</td>
<td>–</td>
<td>–</td>
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<td>–</td>
<td>TU smaller, no symptoms</td>
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<td>Tubulous sclerosis</td>
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<td>–</td>
<td>–</td>
<td>–</td>
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<td>–</td>
<td>–</td>
<td>TU same size, no symptoms</td>
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<td>–</td>
<td>–</td>
<td>–</td>
<td>3 vanished, 2 smaller</td>
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<td>1</td>
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<td>1.3 x 1.0</td>
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<td>–</td>
<td>PFO</td>
<td>–</td>
<td>–</td>
<td>TU smaller, no symptoms</td>
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<td>–</td>
<td>–</td>
<td>–</td>
<td>3 vanished, 1 smaller</td>
</tr>
<tr>
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<td>LV, RV</td>
<td>0.5 x 1.2</td>
<td>Tubulous sclerosis, VES, SVES</td>
<td>PFO</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>TU smaller, no symptom</td>
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<tr>
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<td>0.9 x 1.4</td>
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<td>–</td>
<td>–</td>
<td>–</td>
<td>TU same size, no symptoms</td>
</tr>
<tr>
<td>21</td>
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<td>3</td>
<td>Rhabdomyoma</td>
<td>1</td>
<td>RV</td>
<td>1.2 x 1</td>
<td>No symptoms</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
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<td>3 weeks</td>
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<td>4</td>
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<td>LV</td>
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<td>Tubulous sclerosis, SVES</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Regression 3 years</td>
</tr>
<tr>
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<td>RA</td>
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<td>CHF</td>
<td>–</td>
<td>ASD, VSD</td>
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<td>Pericardial effusion</td>
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<td>–</td>
<td>–</td>
<td>–</td>
<td>No symptoms 9 years</td>
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<td>11 months</td>
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<td>RA</td>
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<td>VSD-murmur</td>
<td>–</td>
<td>VSD</td>
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<td>No symptoms 2 years</td>
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<td>M</td>
<td>42</td>
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<td>1</td>
<td>RV</td>
<td>3.5 x 3</td>
<td>Pain, left arm and thorax</td>
<td>Repol. disorder, RBBB</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>No symptoms 3 years</td>
</tr>
</tbody>
</table>

4. Discussion

The differences between primary cardiac tumours in adults and infants are important. In both age groups the vast majority of the tumours are benign, but in adults 95–98% of the tumours are myxomas, predominantly (≥90%) located in the left atrium [1,10]. Apparently cardiac tumours in children are also benign, however, in contrast to those in adults these tumours in children are rarely myxomas; none in our series. Also the location appears different, being observed in both ventricles and atria in infants and children. Clinically important is the observation, that in children tumour presentation is often variable and unusual and the tumours are frequently hemodynamically relevant due to obstruction of the right or left ventricular inflow or outflow tract or coronary arteries (Table 1). Even ventricular arrhythmias can sometimes be caused by cardiac tumours, being an indication for surgery, as it is described in case 6 (Table 1). In childhood, the tumour subtypes show a wide variability with rhabdomyoma as the most common type [11–13].

In our collective half of the tumours were rhabdomyomas. Compared to the cases in the literature, the frequency of rhabdomyoma seems to be even higher [14]. This may be due to the fact that we did not see those patients with small rhabdomyoma and tuberous sclerosis in our surgical unit.

There are different diagnostic tools which are helpful to characterise the tumour in order to determine which will need surgical removal and which may regress spontaneously. Our electrocardiographic findings were non-specific and only reflected haemodynamic alterations caused by the tumour. Chest films revealed enlargement of the heart, whereas echocardiography provided the best evidence to make a diagnosis [3]. One important advantage of echocardiography is the chance for prenatal diagnosis of in- or outflow tract obstructions or other severe haemodynamic alterations [7]. Catheterization was not necessary in every case, although it provided important information in children with large tumours that compromised the coronary arteries. Supplemental diagnostic imaging included computed tomography and nuclear magnetic resonance. They provided sectional views of cardiac, mediastinal, pulmonary, and thoracic structures without superposition in any plane. These two techniques distinguish tissue composition and allow the identification of solid, liquid, fatty, haemorrhagic or calcified tumours.

Rhabdomyomas occur in association with tuberous sclerosis in more than half of the cases [15,16]. A typical characteristic of these tumours is their multiple occurrence, instead of single. As we saw in our children (Table 1), a peculiar feature of cardiac rhabdomyomas is their spontaneous regression, particularly of smaller lesions, followed by a resolution of symptoms [6,17]. The exact mechanism of apoptosis in rhabdomyoma has not been defined up to now. Based on the data of Black, who described 30 children with rhabdomyomas, spontaneous involution was seen in most of the cases without any deaths among the 23 children who did not undergo surgery. Only 7/30 children required surgical extirpation [18].

Fibromas are mostly found in the ventricles. As in one of our patients (case 3, Table 1), they can interdigitate with ventricular muscle at the tumour border and replace functioning muscle mass. In addition, they can expand into the ventricular conduction system and cause arrhythmias [19–21]. Interior calcification of the tumour seems to be pathognomonic for fibroma. Soler described a calcified intramural fibroma of the left ventricle with a large calcification visible on the chest film [21]. In four of our seven patients with fibromas the large tumours were resected subtotally and one child died perioperatively due to cardiac failure because of the enlarged replacement and interdigitation of the muscular fibres. According to the literature large fibromas are often treated by subtotal resection or primary transplantation after biopsy [20,21].

Large teratomas, as found in two of our children (cases 25, 26 Table 1), were easy to identify. Both contained elements from all three germ layers with predominantly cystic elements. After total resection the patients remained free of recurrence or symptoms for 2 and 3 years, respectively. Close follow-up is necessary, because, as Ali et al.

![Fig. 1. (a) Left ventricular fibroma in a 5-month-old girl (patient 3, Table 1). In magnetic resonance image (MRI) seen here in a coronal view, the tumour (arrow) appears inhomogenous with extensive involvement of the left ventricle. The upper part of the left lung is compromised with secondary atelectasis. (b) In axial T1 weighted MRI the tumour (arrow) demonstrates the massive size (5 × 5.5 cm), in comparison to the remainder of the ventricular cavity.](image)
described, intracardiac teratoma may recur even 3 years after the initial surgery [22].

Although the majority of the cardiac tumours in infancy are benign, they may be life-threatening because of their position. They can mimic not only every structural cardiac disease but also dysrhythmia and severe intermittent cyanosis. Mair reported a newborn with multiple cardiac rhabdomyomas that mimicked a hypoplastic left-heart syndrome [23]. Cardiac tumours must, therefore, be considered in the differential diagnosis of valvular heart disease, cardiac insufficiency, cardiomegaly, disturbances of ventricular and supraventricular rhythm, syncope, and systemic or pulmonary embolism. The symptoms depend on the size, mobility and location of the tumour.

Therapy should be individualized. In adult myxomas there is consensus that surgical resection is the therapy of choice [1,10]. In paediatric primary cardiac tumours, the indication for surgery is differentiated according to the severity of the clinical picture [11,12,14,24]. Asymptomatic rhabdomyomas, often multiple, with a tendency for spontaneous regression should be carefully followed-up by echocardiography [18]. In our series, patient 13 (Table 1) presented 3 days after birth with a tumour-related obstruction of the left ventricular outflow tract with a 60 mmHg gradient. One month later this gradient was less than 10 mmHg and the tumour had almost vanished after 4 months. This tumour seems to have been a rhabdomyoma, as suggested by size, location and regression. In such cases there is no need for anticoagulation or surgery.

During the last decade, the surgical intervention rate for primary cardiac tumours in children has ranged from 32 to 95% in tertiary care centres [11,25] but most authors agree that resection is only recommended when there is associated haemodynamic or respiratory compromise, severe arrhythmia, of a significant risk of systemic embolisation [11,17,18,25].

In conclusion, our study illustrates that despite the benign histology of most primary paediatric cardiac tumours, there may be significant associated morbidity and occasional mortality. Clinicians should keep in mind that although these tumours are rare they have a wide and unusual spectrum of presentation. Individualised surgery allows early and safe treatment of symptomatic tumours. Total resection is not the only therapeutic aim. Most important is the restoration of the best possible haemodynamic heart function.

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References


