

Cardiac Tumors and Associated Arrhythmias in Pediatric Patients, With Observations on Surgical Therapy for Ventricular Tachycardia

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- Objectives** The aim of this study was to describe a large experience with primary cardiac tumors in pediatric patients, characterize associated arrhythmias, and expand knowledge of natural history and treatment options.
- Background** Primary cardiac tumors in children are rare. The incidence of arrhythmias is not well-defined, and management plans vary widely.
- Methods** We employed a retrospective single-center review of patients ≤ 21 years of age diagnosed with a primary cardiac tumor between 1968 and 2010. Clinically significant arrhythmias were defined as: 1) sudden cardiac arrest; 2) nonsustained and sustained ventricular tachycardia (VT); 3) pre-excitation; and 4) sustained supraventricular tachycardia of any mechanism.
- Results** A total of 173 patients were identified: 106 rhabdomyoma, 25 fibroma, 14 myxoma, 6 vascular, 4 teratoma, 3 lipoma, and 15 other. Median age at diagnosis was 7 months (prenatal to 21 years). Of these, 42 (24%) had clinically significant arrhythmias. Patients with large fibromas were the highest-risk group, with VT occurring in 64%. Among rhabdomyoma patients, 10% had pre-excitation, and 6% had VT. Over a mean follow-up of 6 years (1 day to 34 years, median 4 years), surgical excision was performed in 62 cases, with rhythm treatment being 1 of the indications in 20. Post-operatively, clinically significant arrhythmias were eliminated in 18 of these 20, including all 13 fibroma patients.
- Conclusions** Clinically significant arrhythmias occurred in 24% of pediatric patients with cardiac tumors, VT being the most common type. Surgical excision for VT associated with rhabdomyomas and fibromas in selected patients is an important and effective management strategy in these patients. (J Am Coll Cardiol 2011;58:1903-9) © 2011 by the American College of Cardiology Foundation

Primary cardiac tumors in children are rare, with an incidence ranging between 0.03% and 0.32% (1-4). Although most are pathologically benign, they can be associated with serious cardiovascular complications, including intracavitary obstruction, coronary artery compromise, thromboembolic events, and rhythm disturbances (5-16). Data regarding associated arrhythmias and their treatment have been largely limited to case reports or small series (6,8,9,11-30). The most substantial study to date came from Beghetti et al. (20), who identified 56 pediatric patients with various tumor types and described arrhythmias in 11 cases. The purpose of

this study was to describe a relatively large experience with arrhythmias in pediatric patients with cardiac tumors and expand understanding of the natural history and treatment options.

Methods

This was a retrospective review describing all patients ≤ 21 years of age seen at Children's Hospital Boston between October 1968 and October 2010 with a primary cardiac tumor. Medical records were reviewed with permission of the hospital Committee on Clinical Investigation. Patients with secondary tumors due to metastatic disease were excluded. Patients with large non-neoplastic cysts involving the heart or pericardial space were included for review because of the known association of such masses with rhythm disturbances (31). The diagnosis of cardiac tumor was made by echocardiogram and/or magnetic resonance imaging in the current era or by angiography in years before noninvasive imaging capabilities.

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Abbreviations and Acronyms

- AV** = atrioventricular
- EAT** = ectopic atrial tachycardia
- ECG** = electrocardiogram
- LV** = left ventricle/ventricular
- SVT** = supraventricular tachycardia
- VF** = ventricular fibrillation
- VT** = ventricular tachycardia
- WPW** = Wolff-Parkinson-White syndrome

Records were examined for all information pertaining to rhythm status, including narrative history, electrocardiograms (ECG), Holter monitors, electrophysiological studies, and operative notes. Follow-up data were obtained from hospital records or, in some cases, by contact with the referring cardiologist.

Emphasis was placed on rhythm disorders that were judged to be “clinically significant,” defined a priori as: 1) sudden cardiac arrest with documented or suspected ventricular fibrillation (VF); 2) ventricular tachycardia (VT), either sustained (≥ 30 -s duration)

or nonsustained (≥ 3 beats, < 30 -s duration); 3) presence of manifest pre-excitation on ECG (Wolff-Parkinson-White syndrome [WPW]) whether or not there was a history of supraventricular tachycardia (SVT); and 4) SVT involving mechanisms other than WPW (e.g., ectopic atrial tachycardia [EAT], atrial flutter, or orthodromic re-entry via a “concealed” accessory pathway), but only if sustained. Note was also made of certain “low-grade” arrhythmias (e.g., frequent ventricular ectopic beats or couplets, brief nonsustained SVT), although these were considered clinically inconsequential for purposes of this study.

Results

Overview. Over a span of 42 years, 173 patients were seen with a primary cardiac tumor at Children’s Hospital Boston. Patient and tumor characteristics by tumor type are summarized in Table 1. Detection of cardiac tumors became much more frequent after introduction of high-quality echocardiography and magnetic resonance imaging, with nearly 80% of the cases being diagnosed during the most recent 2 decades of the review period.

Over a mean follow-up of 6 years (1 day to 34 years, median 4 years), 42 of the 173 study patients (24%) had at least 1 clinically significant rhythm disorder, as detailed in Table 2. There were 4 sudden cardiac arrests (2%). Three of these patients (2 with fibromas, and 1 with a Purkinje cell tumor) were successfully resuscitated and subsequently found to have malignant VT that required aggressive therapy. The fourth arrest patient, who had a vascular tumor (hemangioma) and an antecedent history of only isolated ventricular premature beats, expired with the event and was presumed to have suffered VF after no alternate cause was uncovered at autopsy.

A total of 27 patients (16%) had documented VT. This was the most common arrhythmia in the study group and was most prevalent among those with fibromas. Pre-excitation was present on ECG in 6% of the study group, although not all these patients experienced sustained SVT

Table 1 Patient and Tumor Characteristics

	All Tumors	Rhabdomyoma	Fibroma	Myxoma	Vascular	Teratoma	Lipoma	Other
n	173	106	25	14	6	4	3	15
Female	77 (45%)	57 (54%)	9 (36%)	8 (57%)	2 (33%)	3 (75%)	1 (33%)	5 (33%)
Median age at diagnosis, n	7 mo (PN-21 yrs)	2 mo (PN-18 yrs)	1 yr (PN-10 yrs)	9 yrs (3 wks-21 yrs)	11 yrs (PN-17 yrs)	20 days (PN-3 yrs)	13 yrs (1 day-16 yrs)	1 yr (1 day-16 yrs)
Hemodynamic changes	35 (20%)	18 (17%)	5 (20%)	6 (43%)	0	4 (100%)	0	2 (13%)
Significant arrhythmia	42 (24%)	17 (16%)	16 (64%)	1 (7%)	1 (17%)	0%	0%	7 (47%)
Asymptomatic	93 (54%)	76 (72%)	4 (16%)	5 (36%)	4 (66%)	0	3 (100%)	1 (7%)
Death due to tumor	6 (3%)	3 (3%)	0	0	2 (33%)	1 (25%)	0	0
Median follow-up, pt-yrs	4.4 yrs (0-34 yrs)	3.9 yrs (0-25 yrs)	4.1 yrs (7 days-34 yrs)	5.3 yrs (12 days-20 yrs)	4.3 yrs (7 mo-18 yrs)	5.0 yrs (4 days-15 yrs)	3.7 yrs (1-8 yrs)	5.2 yrs (4-21 yrs)
Total follow-up, pt-yrs	1,012	571	151	100	35	25	13	117

Values are n (%) or median (range). Hemodynamic changes are inflow/outflow tract obstruction or systolic dysfunction. Significant arrhythmia is sudden cardiac arrest/ventricular fibrillation, sustained or nonsustained ventricular tachycardia, pre-excitation (with or without supraventricular tachycardia), or sustained supraventricular tachycardia involving mechanisms other than Wolff-Parkinson-White syndrome. “Other” tumors: 1 foregut cyst, 2 papillary fibroelastoma, 1 inflammatory pseudotumor, 1 spindle cell sarcoma, 1 inflammatory myofibroblastic tumor, 1 plexiform neurofibroma, 1 pericardial cyst, 2 blood cysts, 1 Purkinje cell tumor, 1 paraganglioma, and 3 unknown or unclassified. mo = months; PN = prenatal; pt = patient; wks = weeks.

Table 2 Arrhythmias by Tumor Type

	All Tumors	Rhabdomyoma	Fibroma	Myxoma	Vascular	Teratoma	Lipoma	Other
Patients, n	173	106	25	14	6	4	3	15
Clinically significant arrhythmia	42 (24%)	17 (16%)	16 (64%)	1 (7%)	1 (17%)	0	0	7 (47%)
Cardiac arrest/VF	4 (2%)	—	2 (10%)	—	1 (17%)	—	—	1 (6%)
VT	27 (16%)	6 (6%)	16 (64%)	1 (7%)	—	—	—	4 (27%)
WPW/sustained SVT	2 (1%)	2 (2%)	—	—	—	—	—	—
WPW/no SVT	9 (5%)	8 (8%)	—	—	—	—	—	1 (7%)
Non-WPW sustained SVT	9 (5%)	5 (5%)	—	—	—	—	—	5 (33%)
Low-grade arrhythmia	15 (9%)	13 (12%)	1 (4%)	1 (7%)	—	—	—	—
Any arrhythmia (low-grade + clinically significant)	57 (33%)	30 (28%)	17 (68%)	2 (14%)	1 (17%)	0	0	7 (47%)

Values are n or n (%). Some patients might have had more than 1 arrhythmia. Clinically significant arrhythmias are subdivided by type and defined in text. Low-grade arrhythmias included frequent premature atrial beats, ventricular ectopic beats or couplets, and brief nonsustained supraventricular tachycardia (SVT) in patients without manifest pre-excitation.

VF = ventricular fibrillation; VT = ventricular tachycardia; WPW = Wolff-Parkinson-White syndrome.

during follow-up. Pre-excitation occurred most frequently in patients with rhabdomyomas. Sustained SVT involving mechanisms other than WPW was observed in 5% of the study group. Some patients had more than 1 clinically significant arrhythmia. Lower-grade arrhythmias that were considered of minimal clinical concern (although still abnormal for age) were observed in another 9% of the study group. Apart from 1 patient with surgically induced heart block after tumor resection, no other instance of bradycardia or atrioventricular (AV) block was observed. Roughly two-thirds of patients had no arrhythmia of any sort detected during follow-up.

There were a total of 10 patient deaths during follow-up, 6 of which could be directly attributed to the cardiac tumor or its treatment (Table 1). Complications due to attempted tumor excision in small infants accounted for 3 of the 6 deaths. The other 3 tumor-related deaths included 1 case of metastatic brain involvement in a patient with a vascular tumor (hemangioendothelioma), 1 case of low cardiac output in an infant with large obstructive rhabdomyomas not felt to be a candidate for surgery, and the 1 late sudden death in the aforementioned patient with a hemangioma.

Arrhythmias according to tumor type. RHABDOMYOMA. Rhabdomyomas were the most common tumors encountered in this study (106 of 173 cases, 61%). Most of these patients (78 of 106, 74%) carried a diagnosis of tuberous sclerosis. A majority of patients (42%) were diagnosed during a prenatal ultrasound or during evaluation for suspected tuberous sclerosis. Other presentations included murmur (16%), arrhythmia (13%), abnormal chest x-ray (11%), congestive heart failure (11%), incidental (11%), thromboembolism (2%), and other (8%). Reduction in both the size and number of tumors was common with increasing age (4), so that hemodynamic symptoms and arrhythmic events typically occurred early in life and often resolved over time.

Clinically significant arrhythmias were present at some point in 17 patients (16%). Six presented with VT, usually in infancy or early childhood. Two patients were treated effectively with antiarrhythmic medications (propranolol, sotalol) and ultimately had resolution of VT as tumor size

regressed. Three patients with VT underwent surgical excision of a large tumor at ages 3 weeks, 1 month, and 6 years. All 3 had successful elimination of VT. The final patient did not present with VT until age 17 years. She had undergone open surgical biopsy of a large left ventricular (LV) tumor early in life. Years later she developed recurrent episodes of rapid VT despite tumor regression, and antiarrhythmic drug trials failed. When mapped at electrophysiological study, VT seemed to relate to scarring at the old epicardial biopsy site but could not be ablated with either an endocardial or epicardial catheter approach (pericardial adhesions precluded epicardial access to target site). The patient ultimately underwent placement of an implantable cardioverter defibrillator, which discharged appropriately on several occasions for rapid VT. In this case, VT might have been caused by the biopsy scar rather than the tumor itself.

Ten patients had manifest pre-excitation, 2 with recurrent sustained SVT, and 8 who were asymptomatic. The 2 symptomatic WPW patients (2-week-old with a single right-sided accessory pathway, 5-year-old with multiple pathways) underwent successful radiofrequency catheter ablation procedures. The site(s) of successful ablation corresponded grossly to regions of tumor on echocardiogram. Of the patients with asymptomatic pre-excitation, 6 had spontaneous resolution of their delta waves with increasing age, although 2 still had pre-excitation persisting into teenage years. Sustained SVT caused by non-WPW mechanisms was present in 5 patients (2 “concealed” accessory pathways, 3 EAT). The EAT was initially suppressed with medications and later resolved completely as tumor regressed. Age at which resolution of a delta wave or EAT occurred varied widely between 1 and 12 years.

FIBROMA. A total of 25 patients were diagnosed with a cardiac fibroma. The most common presenting symptoms included arrhythmia (32%), murmur (20%), and abnormal chest x-ray (20%). Clinically significant arrhythmias were more common in fibroma patients than any other tumor group. Sixteen patients (64%) had documented VT, 2 of whom presented with VF arrest (age 3 weeks and 2 months). Baseline ECG in the majority of fibroma

patients demonstrated T-wave abnormalities, and VT morphologies on ECG were consistent with an origin near the tumor site (Fig. 1). Sustained VT with hemodynamic instability requiring urgent intervention was present in 8 of these 16 patients, whereas the other one-half presented in more stable condition with intermittent episodes of nonsustained VT.

Direct current cardioversion interrupted sustained VT in the 2 cases when it was attempted. Experience was too limited and varied to judge specific efficacy of drug therapy. Anecdotally, lidocaine did not seem to interrupt or suppress VT, although success was observed in some cases when it was used in combination with procainamide. Amiodarone suppressed nonsustained VT in some instances but only provided rate slowing when VT was sustained.

The mechanism for VT in these cases seemed consistent with re-entry, because episodes were usually monomorphic and regular in rate, and electrical cardioversion succeeded when tried. In selected patients undergoing preoperative electrophysiology studies, VT was inducible and terminable with pacing maneuvers. However, the rate and morphology of induced VT often differed from the clinical tachycardia. Furthermore, some patients demonstrated multiple VT morphologies over time, and some had polymorphic VT.

Although these observations could be explained by varied exit points from a re-entrant circuit, triggered automaticity cannot be dismissed.

Thirteen of the 16 VT patients underwent attempted fibroma excision. There were no surgical deaths, and VT was eliminated in all cases. Of the 3 VT patients who did not undergo resection (2 diagnosed in 1970s and felt to be inoperable, 1 parent choice), 2 continue to have nonsustained VT while receiving antiarrhythmic medication but are asymptomatic, and the third improved over a 35-year period to a pattern of low-grade ectopy off medications. The size of the fibroma in relation to cardiac mass decreased with somatic growth in unoperated patients, but unlike rhabdomyomas, fibromas never resolved completely over time.

MYXOMA. A total of 14 patients were diagnosed with myxoma, 3 of whom had associated Carney Syndrome. Patients were diagnosed due to murmur (n = 3), thromboembolism (n = 3), family history of Carney Syndrome (n = 2), arrhythmia (n = 1), congestive heart failure (n = 1), abnormal ECG (n = 1), and incidental finding (n = 2). Tumor locations included 6 left atrium, 2 right atrium, 1 atrial septum, 2 right ventricle, 1 LV, and 2 from the aortic valve. Only 1 patient had a clinically significant arrhythmia

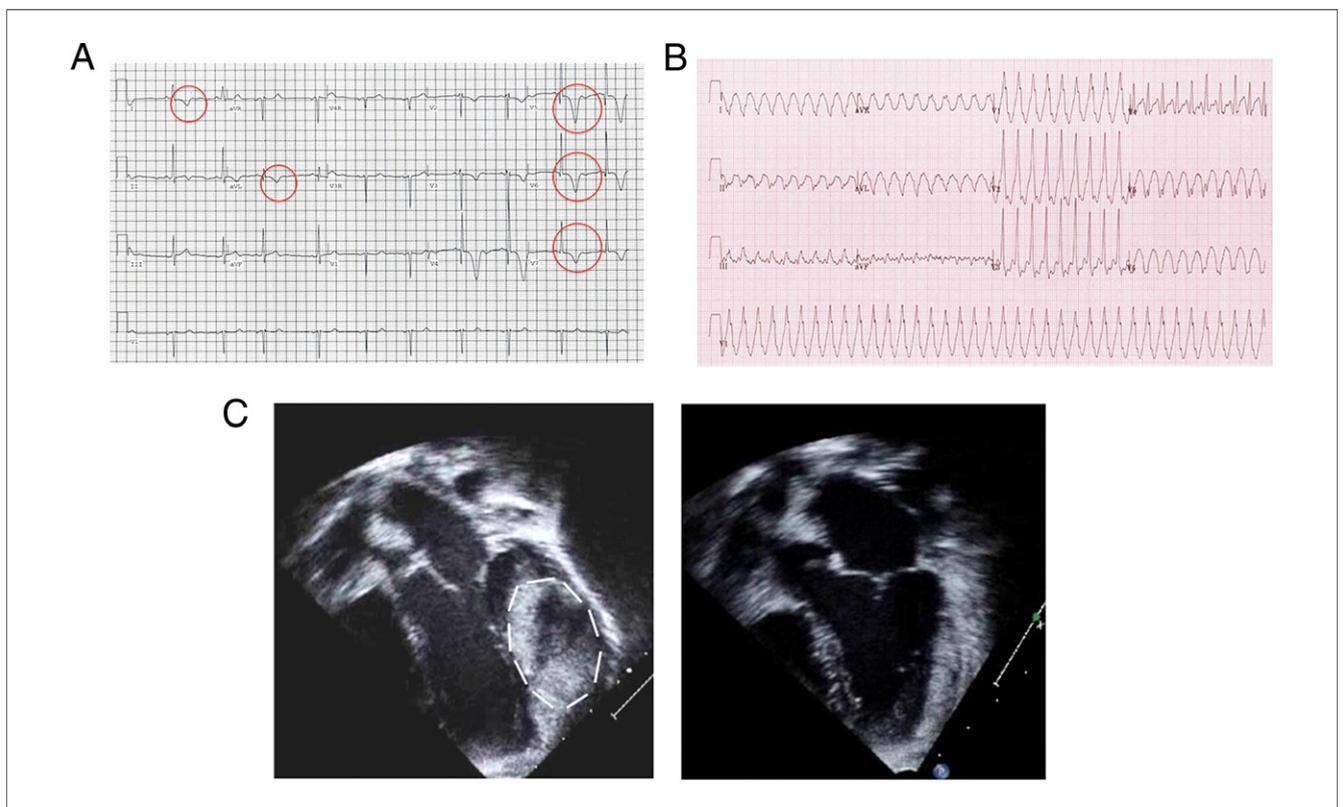


Figure 1 Patient With a Left Ventricular Fibroma

(A) Common electrocardiographic changes in this patient and a majority of patients with left ventricular fibromas demonstrating T-wave inversions in left lateral leads, (B) electrocardiogram of this patient in ventricular tachycardia, and (C) pre- and post-operative echocardiographic images of the same patient after surgical resection of a large left lateral ventricular fibroma.

in the form of nonsustained VT, which resolved after resection of a tumor from the left atrium.

VASCULAR TUMOR. Six patients were diagnosed with vascular tumors, including 5 with hemangiomas, and 1 with malignant hemangioendothelioma that developed into metastatic disease. Patients were diagnosed due to a variety of reasons, including arrhythmia, prenatal ultrasound, syncope, and chest pain. The only clinically significant rhythm event in this group involved the hemangioma patient who expired with sudden cardiac arrest.

TERATOMA. Four patients were diagnosed with teratomas (1 left atrial, 3 pericardial). Two patients were diagnosed due to abnormal chest x-rays, 1 by prenatal ultrasound, and 1 due to syncope. Tumors resulted in hemodynamic compromise in all 4 patients, but none had associated arrhythmias. One patient developed surgically induced heart block requiring a pacemaker after tumor resection.

LIPOMA. Three patients were diagnosed with primary cardiac lipomas—1 due to a murmur, and 2 incidentally. Tumors were located in the right AV groove, right ventricular apex, and central fibrous body. None of the patients had arrhythmias, hemodynamic compromise, or coronary involvement.

OTHER. A group of 15 patients had a variety of rare tumor types, as catalogued in the legend for Table 1. Clinically significant arrhythmias were present in 7 (47%). Arrhythmia was the most common presenting symptom. The most dramatic case involved an infant who was resuscitated from cardiac arrest and found to have intractable VT due to diffuse Purkinje cell tumor involving the apex and LV septum. Incidental pre-excitation but no SVT was also seen in this case. Despite surgical debulking of the apical portion of the tumor at age 13 months, VT promptly recurred. Three catheter ablation sessions with radiofrequency lesions along the LV septum were required in the weeks after surgery until VT could finally be controlled with medications. Residual ventricular arrhythmia then gradually decreased over several years. He is presently doing well on beta-blocker therapy alone more than 10 years later. Three other patients (1 with a foregut cyst, 2 with unclassified fatty tumors) had nonsustained VT that was managed medically.

Sustained SVT in the form of EAT was present in 3 patients with cystic masses (2 right atrial blood cysts, 1 foregut cyst within the triangle of Koch). Despite drainage of the blood cysts (1 surgically, 1 by transcatheter puncture), EAT persisted. Resection of the foregut cyst was deemed inadvisable because of its location near the AV node. All 3 patients had persistent EAT over many years of follow-up, with only partial control on medical therapy. In fact, the patient with the surgically drained blood cyst developed tachycardia-induced myopathy from nearly incessant EAT 19 years after the operation. Diffusely abnormal right atrial tissue with multiple arrhythmia foci was mapped during an unsuccessful catheter ablation attempt. The EAT was ulti-

mately controlled with a combination of medications, and ventricular function normalized. The final patient with SVT had both a “concealed” accessory pathway and atrial flutter in the setting of an inflammatory pseudotumor occupying 50% of the right atrium. After surgical excision, the patient has had no subsequent arrhythmias and is taking no antiarrhythmic medications.

Surgical outcomes. A total of 69 patients were referred to surgery; 5 for biopsy alone, and 64 (37%) for intended tumor resection. Resection attempts were abandoned in 2 cases when the tumor was found to involve critical structures upon direct inspection. Total or subtotal resection was attempted in the remaining 62 patients.

Indications for surgery varied according to tumor type. A subgroup of 21 patients were referred for rhythm indications, including 3 with rhabdomyoma (3 VT), 13 with fibroma (13 VT), 1 with myxoma (VT), and 4 with other rare tumors (1 Purkinje cell tumor with VT, 1 blood cyst with SVT, 1 inflammatory pseudotumor with SVT, 1 foregut cyst with SVT). In the patient with the foregut cyst, the tumor was found to be unresectable, due to its location within the triangle of Koch. Clinically significant arrhythmias were eliminated in 18 of the remaining 20 patients (Fig. 2). For the 18 VT patients, 17 cases had resolution of ventricular arrhythmias, with the only failure being the patient with diffuse Purkinje cell tumor (Fig. 2). Five continued receiving empiric treatment with beta-blocker, whereas the others received no antiarrhythmic medications. Seven recent patients underwent post-operative electrophysiology studies with negative ventricular stimulation. Of the 3 SVT cases, 1 was deemed unresectable, and 1 had a successful outcome after removal of an atrial pseudotumor, but EAT persisted in the patient with a right atrial blood cyst. The tumor itself served as a suitable surgical target in most cases, so that intraoperative arrhythmia mapping was only employed on 4 occasions. Surgical technique involved complete resection when possible, although subtotal resection was performed in 8 cases when critical structures were deemed at risk. Surgical cryoablation lines extending beyond the tumor region to interrupt specific arrhythmia foci or conduction corridors were only added in 3 cases.

A single patient with a fibroma in the arrhythmia group required reoperation due to residual subaortic obstruction. There were no surgical or late deaths. Long-term complications included the appearance of a ventricular wall aneurysm at the site of tumor resection in 3 infants. Two are otherwise doing well with normal global ventricular function, but 1 patient required surgical reoperation to reduce the aneurysm size and is left with a moderate degree of global ventricular dysfunction. Two other infants, both with concomitant congenital heart disease (1 with an atypical cleft mitral valve and mitral regurgitation, 1 with a subaortic membrane) have mild-to-moderate global dysfunction without aneurysms after surgical excision of large LV fibromas.

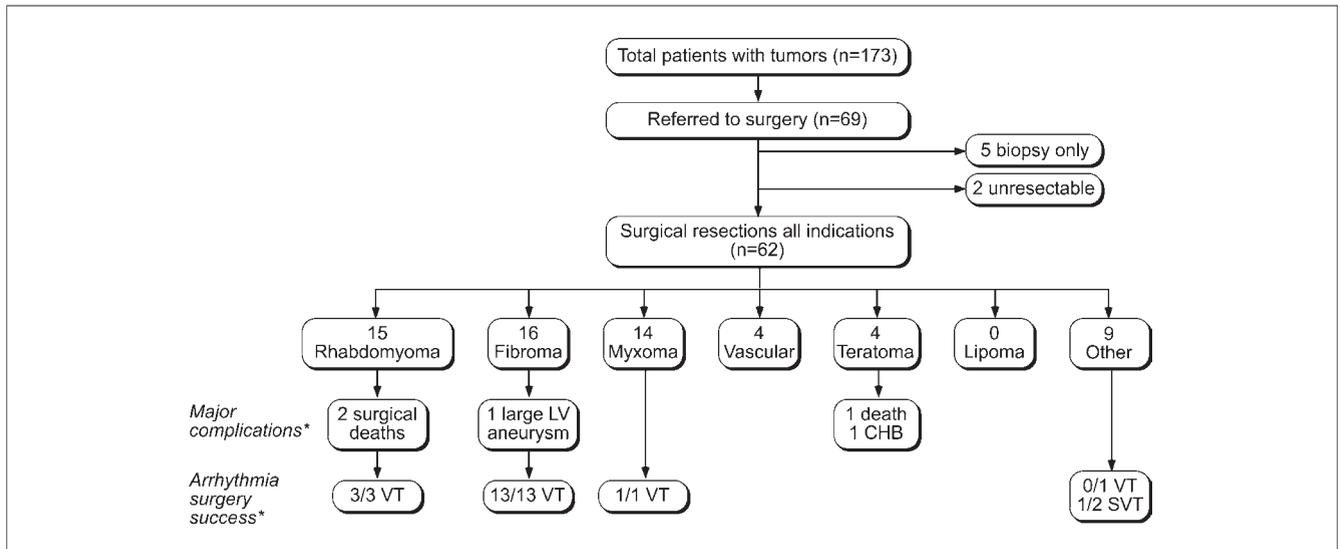


Figure 2 Surgical Patients: Flow Chart of All Patients Who Underwent Attempted Surgical Excision

A breakdown by tumor type and the number of patients who had successful elimination of arrhythmias among the 62 patients who underwent resection. Post-operative complications including death, aneurysm, and myocardial dysfunction are listed by tumor type. *See text for details of complications and arrhythmia surgery outcomes. CHB = complete heart block; LV = left ventricular; SVT = supraventricular tachycardia; VT = ventricular tachycardia.

Surgical resection was attempted for non-arrhythmia indications in 44 patients. One was found to have a nonresectable tumor (1 plexiform neurofibroma wrapping around the mitral valve apparatus). Tumor recurrence was noted after resection in 1 myxoma patient with Carney Syndrome. There were 3 surgical deaths in the non-arrhythmia subgroup—2 infants with large rhabdomyomas (coronary artery injury and mitral valve injury), and 1 infant with a teratoma (coronary artery injury). Other complications included phrenic nerve injury in 1 patient, 1 case of surgically induced heart block requiring a pacemaker, and 1 post-operative stroke. One patient in the non-arrhythmia surgical group exhibited mild-to-moderate global dysfunction but no aneurysm. Patients who developed post-operative aneurysms and/or ventricular dysfunction in this series were all infants notable for having the largest tumor volumes when corrected for body surface area.

Discussion

Optimal management strategy for pediatric patients with cardiac tumors remains unclear, particularly when serious arrhythmias are present. This large descriptive series of 173 patients expands the data available on this topic and provides new observations on the role of surgical intervention. Approximately one-half of the patients had 1 or more tumor-related symptoms due to hemodynamic obstruction, thromboembolic events, or rhythm disturbances, and surgery ultimately had to be performed in two-thirds of these symptomatic cases (46 cases).

Clinically significant arrhythmias were documented in 24% of the study group. The highest arrhythmia burden was seen in the fibroma group, who were strongly predisposed to

VT that was often life-threatening and difficult to control. Multiple prior reports of sudden cardiac arrest in these patients (6,9,12-14,18) attest to the gravity of this condition. Clinical observations from this study support re-entry as the most likely VT mechanism, although triggered automaticity cannot be excluded. Management can include antiarrhythmic medications, placement of an implantable cardioverter defibrillator, surgical excision, and even heart transplantation. Our experience indicates that tumor resection can be a very effective option, a view supported by multiple prior reports of successful VT surgery in this setting (6,21,22,29,30). The VT was eliminated without recurrence in all 13 of our fibroma patients undergoing operation. It was notable that success was achieved with both total and subtotal resection, suggesting that significant debulking of the tumor mass might be sufficient to reverse the arrhythmogenic substrate.

Rhabdomyoma patients presented a more diverse assortment of arrhythmias, but pre-excitation stood out as relatively common in this group, at least at younger ages before these tumors begin to regress. The majority of the rhabdomyoma patients with WPW were asymptomatic. When necessary, catheter ablation can successfully eliminate the accessory pathway, as it did in 2 of our highly symptomatic cases, but pre-excitation quite often will resolve spontaneously over time. Improvement in rhythm status with tumor regression was also seen in rhabdomyoma patients with VT and non-WPW forms of SVT. It seems reasonable to recommend that, whenever possible, medical therapy be used as a temporizing measure in rhabdomyoma patients, allowing tumors to regress before resorting to invasive options. However, in acutely unsta-

ble patients, both surgery and catheter ablation can play effective roles.

Surgery for resection of a large tumor mass is not without risks. There were 3 surgical deaths (5%), although these cases all involved ill infants with large tumors for whom transplant might have been the only alternative. There were no deaths among the 21 patients undergoing surgery for arrhythmia indications. The appearance of post-operative ventricular wall aneurysms and global systolic dysfunction in infants is an issue that will deserve long-term attention. All infants who undergo tumor resection should be followed carefully with serial echocardiograms to screen for these potential complications.

Conclusions

Cardiac tumors in children can be associated with serious arrhythmias in approximately one-quarter of cases. Patients with large fibromas seem to be the most concerning group, with a VT risk exceeding 50%. Surgical excision of such tumors can be a highly effective management option.

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