Online First

The European Research Journal 2026 Research Article

DOI: https://doi.org/10.18621/eurj.1220 *Pediatric Cardiology*

Long-Term Outcomes of Pediatric Intracardiac Masses: Clinical Course and Tumor Behavior in a Single-Center Experience

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Abstract:

Objective: We aimed to evaluate the clinical characteristics, diagnostic pathways, echocardiographic and Holter findings, treatment strategies, and long-term outcomes of children with intracardiac masses, and to explore factors associated with tumor regression.

Methods: This retrospective observational study included children with echocardiographically confirmed intracardiac masses followed at a tertiary pediatric cardiology center between January 2010 and January 2022. Demographic and clinical characteristics, tumor features, serial echocardiographic measurements, 24-hour Holter recordings, treatment strategies, and follow-up outcomes were analyzed.

Results: A total of 20 patients were included. Rhabdomyoma was the most common tumor type (60%, n=12), followed by fibroma (30%, n=6). Most patients were asymptomatic or mildly symptomatic at presentation (90%, n=18). Clinically significant arrhythmias were observed in 15% (n=3), predominantly in patients with fibromas. Tumor size reduction was observed in 40% (n=8) of patients. All patients diagnosed during the fetal period (15%, n=3) exhibited tumor size reduction; all had rhabdomyomas. Everolimus was administered in 25% (n=5) of patients, with tumor size reduction observed in 80% (n=4) of treated cases; however, this was not statistically significant (P=0.253). Left ventricular systolic function remained preserved in all patients during follow-up.

Conclusion: Pediatric intracardiac masses are predominantly benign but show heterogeneous clinical behavior. Tumor regression was more frequent in rhabdomyomas; however, this may reflect tumor type distribution. Arrhythmias were more frequent in fibromas, supporting rhythm surveillance. Everolimus may be considered in selected patients; however, findings should be interpreted with caution due to limited sample size and potential selection bias, and require confirmation in larger studies.

Keywords: Pediatric Cardiac Tumors, Rhabdomyoma, Fibroma, Arrhythmia, Echocardiography, Everolimus

Primary cardiac tumors in children are rare and predominantly benign, with rhabdomyoma and fibroma being the most common histological types. Despite their benign nature, these tumors may lead to clinically significant complications, including arrhythmias and inflow or outflow tract obstruction,

Submitted: March 8, 2026 **Accepted:** May 9, 2026 **Published Online:** May 12, 2026

How to cite this article: Arıcı Ş, Çırakoğlu AM, Akalın F. Long-Term Outcomes of Pediatric Intracardiac Masses: Clinical Course and Tumor Behavior in a Single-Center Experience. Eur Res J. 2026. doi: 10.18621/eurj.1220

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Available Online at https://www.eurj.org.tr

underscoring the importance of careful evaluation and follow-up [1, 2].

Rhabdomyomas are frequently associated with tuberous sclerosis complex and may be detected during fetal life owing to advances in prenatal imaging. Although spontaneous regression is commonly observed, particularly in asymptomatic patients, some tumors may cause hemodynamic compromise or arrhythmias, necessitating medical or surgical intervention [3, 4]. In contrast, cardiac fibromas are less likely to regress and are more commonly associated with clinically significant arrhythmias [5, 6].

Although the general clinical course of pediatric cardiac tumors is well described, variations in tumor behavior and clinical outcomes according to presentation remain less clearly defined. In particular, factors associated with tumor regression and the need for intervention during follow-up are not fully clarified.

The aim of this study was to evaluate the clinical characteristics, diagnostic pathways, echocardiographic and Holter findings, treatment strategies, and long-term outcomes of pediatric patients with intracardiac masses. In addition, we aimed to explore factors associated with tumor regression during follow-up, in order to provide further insight into tumor behavior and clinical management.

METHODS

Study Design and Population

This retrospective observational study included pediatric patients with echocardiographically confirmed intracardiac masses who were followed at a tertiary pediatric cardiology center between January 2010 and January 2022. Medical records were retrospectively reviewed to identify eligible cases with available clinical follow-up data. Demographic characteristics, clinical presentation, etiological distribution, diagnostic pathways, echocardiographic and 24-hour Holter findings, treatment strategies (medical and interventional/surgical), treatment duration and response, and follow-up outcomes were systematically evaluated.

Follow-up duration was defined as the time interval between the first evaluation at our center and the most recent follow-up visit.

Data Collection and Definitions

Clinical, demographic, and follow-up data were retrospectively extracted from patients' medical records. Retrieved variables included age at diagnosis, sex, presenting symptoms, referral indications, associated systemic conditions, and family history. Tumor-related characteristics were obtained from archived echocardiographic reports and included tumor type, size, number, and anatomical localization.

Tumor size was defined as the largest diameter reported on transthoracic echocardiographic examinations in any imaging plane, based on measurements obtained from standard examinations performed as part of routine clinical evaluation. Tumor number was categorized as single or multiple based on echocardiographic documentation. The presence of inflow or outflow tract obstruction was determined according to findings recorded in echocardiographic reports.

Echocardiographic parameters, including cardiac chamber dimensions, ventricular systolic and diastolic function indices, and Doppler flow measurements, were collected from baseline and follow-up echocardiographic records. When available, z-scores adjusted for body surface area were used as documented in the original reports.

Rhythm evaluation was based on the review of archived 24-hour ambulatory Holter electrocardiographic records obtained at the time of diagnosis and during follow-up when clinically indicated. Documented arrhythmias were categorized as supraventricular or ventricular ectopic activity and further classified as rare or frequent, as reported in the original Holter reports. Clinically significant arrhythmias were defined as ventricular tachycardia or rhythm disturbances documented in the medical records that required initiation of antiarrhythmic therapy during follow-up.

Tumor behavior during follow-up was assessed using serial echocardiographic records. Changes in tumor size and tumor number were categorized as decrease or non-decrease. The non-decrease category included tumors that remained unchanged, increased in size or number, or required surgical resection.

Tumor regression was defined as a documented reduction in the largest tumor diameter and/or tumor number on follow-up echocardiographic examinations compared with baseline findings. Regression assessment was based on the comparison between baseline and the most recent follow-up echocardiographic measurements. In patients with multiple masses, the largest lesion was used for comparison. No predefined percentage threshold was applied; any measurable reduction in the largest tumor diameter was considered regression.

This study was approved by the local Clinical Research Ethics Committee (Protocol No: 09.2022.462, approval date: March 4, 2022) and was conducted in accordance with the principles of the Declaration of Helsinki. Due to the retrospective study design, informed consent was waived.

Statistical Analysis

Statistical analyses were performed using SPSS version 15.0. Categorical variables are presented as frequencies and percentages. Continuous variables were assessed for distribution using visual methods and are presented as mean±standard deviation or median and interquartile range, as appropriate. Comparisons between baseline and follow-up echocardiographic parameters were performed using appropriate paired statistical tests. Correlation analyses were conducted using Pearson or Spearman correlation coefficients according to data distribution. Associations between tumor behavior and categorical variables were analyzed using Fisher's exact test. A two-sided P-value <0.05 was considered statistically significant.

RESULTS

Patient Characteristics and Diagnostic Pathways

A total of 20 pediatric patients diagnosed with cardiac masses were included in the study. Regarding tumor distribution, rhabdomyoma was the most frequently identified mass, accounting for 60.0% (n=12) of cases, followed by fibroma in 30.0% (n=6). Rare etiologies included an intracardiac hydatid cyst in one patient (5.0%) and intracardiac extension of Wilms tumor complicated by tumor-associated thrombosis in one patient (5.0%). The clinical characteristics, imaging findings, treatment strategies, and outcomes of the study cohort are summarized in Supplementary Table 1.

The demographic and baseline characteristics of the study population are summarized in Table 1. The cohort comprised 11 females (55.0%) and 9 males (45.0%). The mean age at diagnosis was 59.5±65.0 months (range: 0–170), and the mean follow-up duration was 46.7±48.1 months (range: 1–168). The mean body weight, height, and body surface area were 19.73±17.20 kg, 97.60±39.49 cm, and 0.71±0.46 m², respectively.

Evaluation of diagnostic pathways demonstrated that 20.0% of patients (n=4) were referred for cardiac-related reasons, whereas the majority, 80.0% (n=16), were referred due to non-cardiac clinical contexts. Cardiac-related referrals were limited to patients evaluated because of a cardiac murmur detected on physical examination (n=3) and exercise intolerance (n=1).

Non-cardiac referrals were most commonly related to systemic conditions requiring assessment for potential cardiac involvement, particularly tuberous sclerosis (n=5). Other frequent reasons included prenatal detection of a cardiac mass on obstetric ultrasonography (fetal diagnosis; n=3), neurological findings identified during evaluation (including seizures and café-au-lait spots; n=3), and routine screening (n=2). Less frequently, patients were referred following evaluations for oral abscess or abdominal mass by other specialties.

Clinical Features and Associated Conditions

Within the study cohort, associated conditions were identified in 80.0% (n=16) of patients. Tuberous sclerosis was the most frequently observed associated condition, present in 50.0% (n=10) of cases. Other associated conditions or diagnoses, each identified in a single patient (5.0%), included asthma, Gorlin syndrome, neurofibromatosis, intracardiac hydatid cyst, neurogenic bladder accompanied by chronic kidney disease, and Wilms tumor.

At the time of initial clinical evaluation, the majority of patients were either asymptomatic or exhibited mild symptoms. Based on functional status assessment, 90.0% (n=18) of the cohort were classified as New York Heart Association (NYHA)

SUPPLEMENTARY TABLE 1. Clinical Characteristics, Imaging Findings, Treatment, and Outcomes of Children Diagnosed with Intracardiac Masses (n=20)

Case	Age (months) / Sex	Follow-up duration (months)	Mass type	Location	Associated condition	Clinical presentation	Arrhythmia / holter findings	Key echocardiographic findings	Treatment	Outcome (Follow-up)
1	2/F	53	Fibroma	RV	None	Murmur	Rare VEA	Mass (19 mm) arising from the right ventricular septum, mild RVOT obstruction, normal left ventricular systolic function.	None	Tumor regression
2	126/F	48	Fibroma	RV	Gorlin syndrome	Systemic disease evaluation	Normal	Mass (12 mm) located on the right ventricular free wall, no outflow tract obstruction, normal left ventricular systolic function.	None	Tumor progression
3	82/F	125	Fibroma	IVS	Asthma	Murmur	Frequent VEA, VT	Large interventricular septal mass (32 mm), no outflow tract obstruction, preserved ventricular systolic function.	Aspirin + Propranolol + Flecainide + Everolimus	Tumor progression
4	155/M	16	Fibroma	LV	Neurogenic bladder, chronic kidney disease	Routine screening	Normal	Mass (29 mm) attached to the left ventricular free wall and papillary muscle, no obstruction, normal left ventricular systolic function	Aspirin	Tumor progression
5	28/F	3	Intracardiac Wilms tumor invasion with associated thrombus	RV, RA	Wilms tumor	Systemic disease evaluation	Normal	Large right atrial mass extending into the right ventricle (42 mm), no inflow or outflow obstruction, preserved ventricular systolic function.	Aspirin	Surgical resection
6	3/F	145	Rhabdomyoma	IVS	Tuberous sclerosis	Seizure	Frequent SVEA, Rare VEA	Small interventricular septal mass (4 mm), no obstruction, normal ventricular systolic function.	Propranolol	Tumor progression
7	14/M	34	Rhabdomyoma	IVS	Tuberous sclerosis	Seizure	Normal	Apical interventricular septal mass (6 mm), no obstruction, normal ventricular systolic function.	None	Tumor regression
8	110/F	1	Fibroma	LV	None	Asymptomatic	Normal	Subaortic mass (12 mm) causing mild left ventricular outflow tract obstruction, normal left ventricular systolic function.	None	Stable disease
9	158/M	20	Fibroma	IVS	Neurofibromatosis	Systemic disease evaluation	Normal	Multiple small masses involving the interventricular septum, no obstruction, preserved ventricular systolic function.	None	Tumor progression
10	1/M	24	Rhabdomyoma	RV, LV, IVS	Tuberous sclerosis	Murmur	Normal	Multiple small masses involving the interventricular septum, right ventricular free wall, and left ventricular papillary muscle, no obstruction, normal ventricular systolic function.	Aspirin	Tumor progression
11	151/M	1	Rhabdomyoma	IVS	Tuberous sclerosis	Systemic disease evaluation	Normal	Multiple interventricular septal masses (largest 7 mm), no obstruction, normal ventricular systolic function.	None	Stable disease
12	14/M	45	Rhabdomyoma	RV, IVS, IAS, RA, LA	Tuberous sclerosis	Systemic disease evaluation	Normal	Multiple small masses involving the interventricular septum, interatrial septum, right atrium, and right ventricle, no obstruction, preserved ventricular systolic function.	None	Tumor regression
13	60/M	168	Rhabdomyoma	LV, IVS	Tuberous sclerosis	Systemic disease evaluation	Normal	Multiple masses involving the interventricular septum and left ventricular free wall, no obstruction, normal ventricular systolic function.	None	Stable disease
14	6/F	60	Rhabdomyoma	LV	Tuberous sclerosis	Systemic disease evaluation	Rare SVEA, Frequent VEA	Small left ventricular mass (5 mm), no obstruction, normal left ventricular systolic function.	Sotalol	Stable disease
15	1/F	11	Rhabdomyoma	RV	None	Routine screening	Normal	Right ventricular outflow tract mass (21 mm) causing severe RVOT obstruction, preserved ventricular systolic function.	Everolimus	Tumor regression
16	0/M	28	Rhabdomyoma	LV, IVS	Tuberous sclerosis	Fetal diagnosis	Normal	Multiple masses involving the interventricular septum and left ventricular apex (largest 17 mm), no obstruction, normal ventricular systolic function.	Everolimus	Tumor regression
17	0/M	60	Rhabdomyoma	RV, LV, IVS	Tuberous sclerosis	Fetal diagnosis	Normal	Multiple masses involving both ventricles and the interventricular septum (largest 17 mm), no obstruction, normal ventricular systolic function.	Everolimus	Complete regression
18	5/F	24	Rhabdomyoma	RV	None	Fetal diagnosis	Normal	Multiple small masses on the tricuspid valve, no inflow obstruction, preserved ventricular systolic function.	None	Complete regression
19	104/F	67	Rhabdomyoma	IAS	Tuberous sclerosis	Systemic disease evaluation	Normal	Interatrial septal mass (9 mm), no obstruction, normal ventricular systolic function.	Everolimus	Complete regression
20	170/F	1	Hydatid cyst	RV	Hydatid cyst	Exercise intolerance	Normal	Right ventricular mass (21 mm), no outflow tract obstruction, normal ventricular systolic function.	Surgical resection + albendazole	Postoperative follow-up unavailable

RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; IVS, interventricular septum; IAS, interatrial septum; RVOT, right ventricular outflow tract; VEA, ventricular ectopic activity; SVEA, supraventricular ectopic activity; VT, ventricular tachycardia.

TABLE 1. Demographic and Baseline Characteristics of the Study Population

Variable	Data
Female sex, n (%)	11 (55.0)
Male sex, n (%)	9 (45.0)
Age at diagnosis (months)	59.5±65.0 (0–170) 21 (2.75–114)
Follow-up duration (months)	46.7 ± 48.1 (1–168) 31 (14.75–60)
Body weight (kg)	19.73±17.20 (3.3–64.5) 13.75 (6.05–30.25)
Height (cm)	97.60±39.49 (51–157) 86.50 (60.75–136.00)
BSA (m²)	0.71±0.46 (0.21–1.57) 0.55 (0.31–1.08)

Data are shown as mean±standard deviation (minimum–maximum), median (interquartile range) or n (%) where appropriate. BSA, body surface area.

functional class I, while 10.0% (n=2) were classified as NYHA class II. Palpitations were reported in only one patient (5.0%), and no episodes of syncope or chest pain were documented.

Family history assessment revealed a history of cardiac disease in 10.0% (n=2) of patients. There was no history of unexplained sudden cardiac death, documented arrhythmias, permanent pacemaker or implantable cardioverter–defibrillator implantation, or congenital deafness within the study cohort.

On physical examination, cardiac auscultation was normal in 65.0% (n=13) of patients, whereas 35.0% (n=7) demonstrated an audible cardiac murmur. The murmurs varied in intensity and timing, ranging from low-grade systolic murmurs to pansystolic murmurs; however, no distinctive auscultatory pattern associated with tumor type or localization was identified.

Echocardiographic Findings at Baseline and During Follow-up

At baseline echocardiographic evaluation, the mean size of cardiac masses was 13.3±10.8 mm (range: 2.1–42.3), with a median diameter of 8.9 mm (IQR: 5.9–18.5). Masses were most frequently localized to the interventricular septum (50.0%), followed by the right ventricle (45.0%) and left ventricle (35.0%). Less common locations included the interatrial septum (10.0%), right atrium (10.0%), and left atrium (5.0%). A single mass was identified in 60.0% of patients, whereas multiple masses were detected in 40.0%. Left or right ventricular outflow tract obstruction was present in 15.0% of cases (Figure 1). A detailed comparison of baseline and follow-up echocardiographic measurements is provided in Table 2.

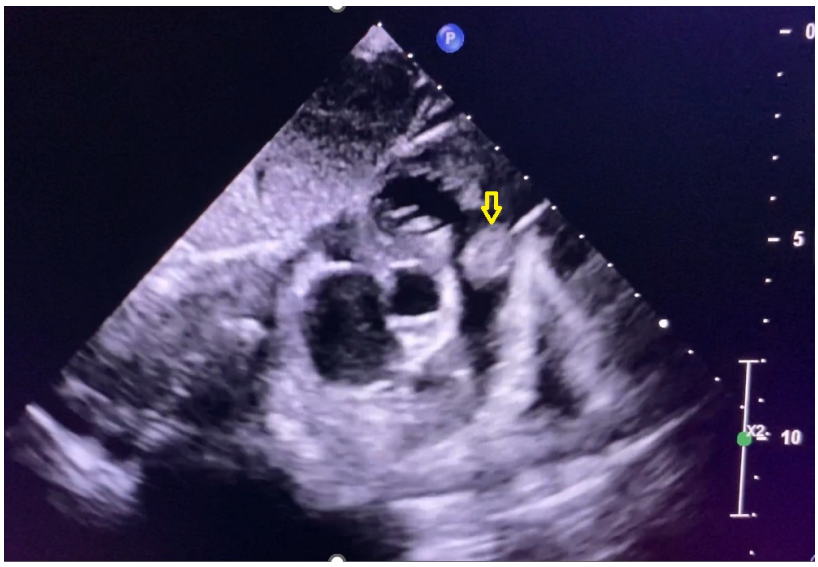

FIGURE 1. Representative transthoracic echocardiographic image from Case 1 obtained in the subcostal short-axis view, demonstrating an intracardiac fibroma (arrow) arising from the right ventricular septum and causing right ventricular outflow tract obstruction.

TABLE 2. Echocardiographic Findings at Baseline and Follow-up

Parameter	Baseline	Follow-up	P-value
IVSd (cm)	0.64±0.13 (0.43–0.89) 0.62 (0.56–0.75)	0.77±0.32 (0.42–1.83) 0.71 (0.61–0.83)	**0.028**
LVDd (cm)	3.01±0.91 (1.64–4.64) 2.95 (2.23–3.76)	3.57±0.72 (2.45–4.81) 3.52 (2.87–4.18)	**0.003**
LVDs (cm)	1.89±0.53 (1.15–2.87) 1.90 (1.34–2.27)	2.17±0.47 (1.44–2.87) 2.12 (1.81–2.70)	**0.009**
LVPWd (cm)	0.56±0.17 (0.36–0.90) 0.54 (0.43–0.68)	0.61±0.18 (0.30–1.00) 0.61 (0.45–0.70)	**0.022**
IVSd z-score	0.30±1.03 0.43 (–0.54–1.07)	0.48±2.64 –0.10 (–0.82–0.95)	0.481
LVDd z-score	–1.01±1.20 –1.55 (–1.94–0.12)	–1.07±0.89 –1.50 (–1.89–0.15)	0.763
LVDs z-score	–1.08±0.87 –1.58 (–1.79–0.30)	–1.15±0.67 –1.36 (–1.74–0.48)	0.968
LVPWd z-score	–0.23±1.02 –0.46 (–1.07–0.53)	–0.49±0.91 –0.52 (–1.24–0.04)	0.615
EF (%)	72.15±6.13 (64–86) 71.0 (67.3–77.3)	70.42±5.80 (61–83) 71.0 (65.5–73.5)	0.552
FS (%)	39.95±6.34 (30–53) 39.5 (35.3–45.0)	39.53±5.16 (31–49) 39.0 (35–42)	0.955
Ao diameter (cm)	1.60±0.39 (0.92–2.13) 1.64 (1.24–1.98)	1.92±0.42 (1.29–2.74) 1.90 (1.57–2.18)	**0.002**
Ao z-score	–0.59±1.04 –0.69 (–1.21–0.22)	–0.53±1.24 –0.88 (–1.68–0.46)	0.872
LAD (cm)	2.03±0.64 (1.08–3.04) 2.06 (1.40–2.61)	2.47±0.53 (1.46–3.32) 2.65 (2.00–2.87)	**0.002**
LAD z-score	–0.33±1.00 –0.44 (–1.17–0.62)	0.13±1.04 0.12 (–0.73–0.98)	0.198
Mitral E/A	1.43±0.28 (0.71–1.86) 1.43 (1.29–1.63)	1.57±0.21 (1.24–2.04) 1.54 (1.44–1.73)	**0.046**
DecT (ms)	95.4±28.2 (45–137) 103 (69–116)	127.7±35.3 (63–204) 135 (103–150)	**0.002**

Data are shown as mean±standard deviation (minimum–maximum) and median (interquartile range). IVSd, interventricular septal thickness at end-diastole; LVDd, left ventricular end-diastolic diameter; LVDs, left ventricular end-systolic diameter; LVPWd, left ventricular posterior wall thickness at end-diastole; EF, ejection fraction; FS, fractional shortening; Ao, aortic root diameter; LAD, left atrial diameter; DecT, deceleration time; IVRT, isovolumic relaxation time.

Statistically significant P-values are shown in bold.

Associated echocardiographic structural abnormalities were infrequent. Atrial septal defect was detected in 10.0% of patients, and mild tricuspid regurgitation in 5.0%, while 85.0% had no additional structural cardiac pathology. On telecardiographic evaluation, the mean cardiothoracic ratio was 0.50±0.05, and no statistically significant change in pulmonary vascular markings was observed during follow-up.

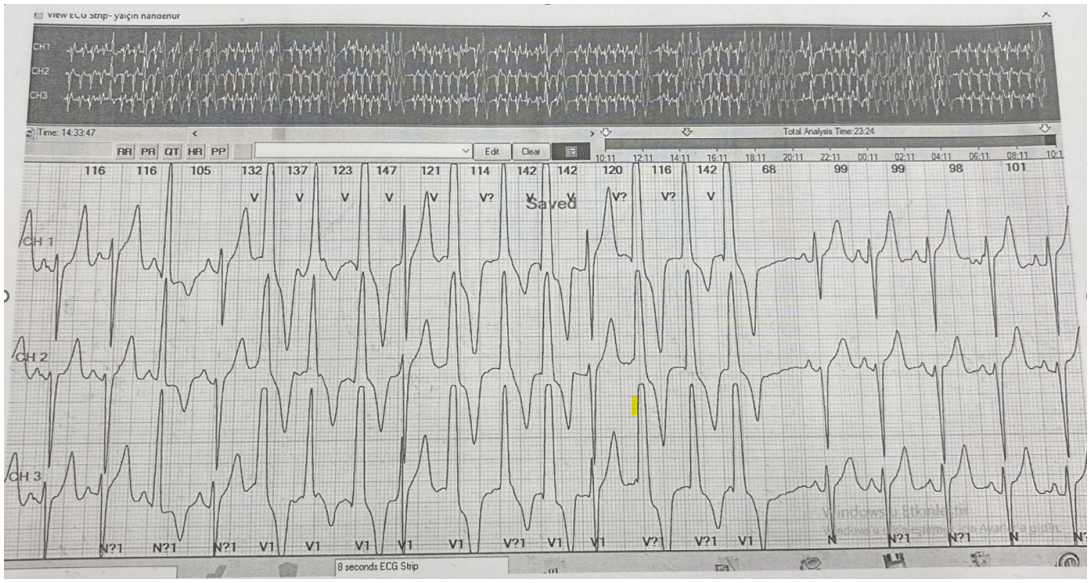

FIGURE 2. Representative 24-hour Holter ECG recording from Case 3 showing a non-sustained ventricular tachycardia episode in a patient with intracardiac fibroma.

Serial echocardiographic measurements obtained during follow-up demonstrated statistically significant changes in left ventricular dimensions and wall thicknesses. Interventricular septal thickness at end-diastole (IVSd) increased from 0.64±0.13 cm at baseline to 0.77±0.32 cm at follow-up (P=0.028). Similarly, left ventricular end-diastolic diameter (LVDd) increased from 3.01±0.91 cm to 3.57±0.72 cm (P=0.003), and left ventricular end-systolic diameter (LVDs) increased from 1.89±0.53 cm to 2.17±0.47 cm (P=0.009). Left ventricular posterior wall thickness at end-diastole (LVPWd) also showed a significant increase during follow-up (from 0.56±0.17 cm to 0.61±0.18 cm; p = 0.022). In contrast, no statistically

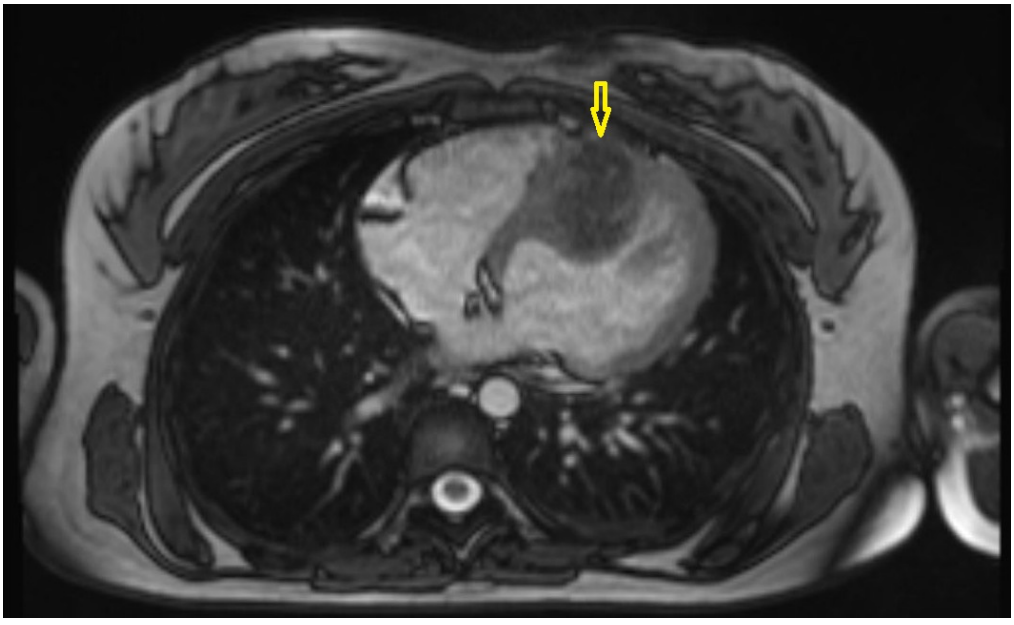

FIGURE 3. Representative cardiac magnetic resonance image from Case 3 obtained in the axial plane, demonstrating a large interventricular septal fibroma (arrow).

TABLE 3. Factors Associated with Tumor Regression During Follow-up

Variable	Category	Tumor size decrease n (%)	Tumor size non-decrease n (%)	P-value
Everolimus therapy	Yes (n=5)	4 (80.0)	1 (20.0)	0.253
	No (n=15)	4 (26.7)	11 (73.3)	
Timing of diagnosis	Fetal (n=3)	3 (100)	0 (0)	0.294
	Postnatal (n=17)	5 (29.4)	12 (70.6)	
		Tumor number decrease n (%)	**Tumor number non-decrease n (%)**	
Everolimus therapy	Yes (n=5)	3 (60.0)	2 (40.0)	0.188
	No (n=15)	2 (13.3)	13 (86.7)	
Timing of diagnosis	Fetal (n=3)	3 (100)	0 (0)	0.029
	Postnatal (n=17)	2 (11.8)	15 (88.2)	

Tumor size and tumor number changes were categorized as decrease versus non-decrease (unchanged, increased, or surgically resected). All patients with fetal diagnosis had rhabdomyomas. P values were calculated using Fisher's exact test.

significant changes were observed in the corresponding z-scores for these parameters over time (all P>0.05).

With regard to left ventricular systolic function, no statistically significant differences were observed between baseline and follow-up assessments. Fractional shortening was 39.95±6.34% at diagnosis and 39.53±5.16% at follow-up (P=0.955). Similarly, left ventricular ejection fraction did not differ significantly over time (72.15±6.13% vs. 70.42±5.80%; P=0.552).

During follow-up, significant increases were observed in the aortic root diameter (from 1.60±0.39 cm to 1.92±0.42 cm; P=0.002) and left atrial diameter (from 2.03±0.64 cm to 2.47±0.53 cm; P=0.002). However, no statistically significant differences were found in aortic root or left atrial z-scores between baseline and follow-up measurements (P=0.872 and P=0.198, respectively).

Assessment of diastolic function revealed significant increases in the mitral E/A ratio (from 1.43±0.28 to 1.57±0.21; P=0.046) and deceleration time (from 95.4±28.2 ms to 127.7±35.3 ms; P=0.002). Other diastolic parameters, including mitral E and A velocities, isovolumic relaxation time, and z-score–adjusted indices, showed no statistically significant changes during follow-up. Likewise, no significant differences were observed between baseline and follow-up measurements for aortic, pulmonary artery, and descending aortic flow velocities.

Arrhythmia Findings and Holter Monitoring

All patients underwent 24-hour Holter electrocardiographic monitoring for rhythm assessment. Supraventricular ectopic activity (SVEA) was detected in 10.0% (n=2) of patients; one patient exhibited rare SVEA, while one patient exhibited frequent SVEA. No supraventricular ectopic beats were observed in 90.0% (n=18) of the cohort. Supraventricular tachycardia (SVT) and sinus pauses were not detected in any patient.

Ventricular ectopic activity (VEA) was identified in 20.0% (n=4) of patients, with rare VEA observed in 10.0% (n=2) and frequent VEA in 10.0% (n=2). Ventricular tachycardia (VT) was documented in one patient (5.0%) (Figure 2). In this patient, cardiac magnetic resonance imaging demonstrated a large interventricular septal mass consistent with fibroma (Figure 3). Atrioventricular block was not observed in any patient.

Overall Holter evaluation revealed clinically significant arrhythmias in 15.0% (n=3) of patients, while 85.0% (n=17) demonstrated no clinically significant rhythm disturbances.

Treatment Strategies and Response

During follow-up, 50.0% (n=10) of patients received medical therapy directed at the cardiac mass, while the remaining 50.0% (n=10) were managed without mass-directed pharmacological treatment. Medications administered for mass-related indications included aspirin in 15.0% (n=3) of patients, everolimus in 25.0% (n=5), and albendazole in 5.0% (n=1). Everolimus was administered in selected patients in accordance with dosing regimens reported in the literature for pediatric cardiac rhabdomyoma, typically ranging from 1.5–2 mg/m²/day. No treatment discontinuation due to adverse effects was observed among patients receiving everolimus.

During follow-up, antiarrhythmic therapy due to clinically significant arrhythmia was required in 15.0% (n=3) of patients. In these patients, propranolol, sotalol, and flecainide were used. The remaining 85.0% (n=17) of patients did not require antiarrhythmic medication during follow-up.

Surgical intervention was required in two patients (10.0%) due to specific mass-related etiologies, including intracardiac Wilms tumor invasion with associated thrombosis and intracardiac hydatid cyst.

Factors Associated with Tumor Regression

Changes in Tumor Size and Number During Follow-up

Changes in tumor size during follow-up were categorized as unchanged, decreased, increased, or surgically resected. Overall, tumor size reduction was observed in 40.0% (n=8) of patients. Tumor size increase occurred in 30.0% (n=6), while 10.0% (n=2) of patients underwent surgical resection. In the remaining cases, tumor size remained unchanged during follow-up. The association between everolimus therapy and tumor size decrease did not reach statistical significance (P=0.253). Factors associated with changes in tumor size and number during follow-up are summarized in Table 3.

Among the five patients who received everolimus, tumor size reduction was observed in four patients (80.0%) during follow-up. Changes in tumor number were evaluated using the same categorical approach. Overall, tumor number reduction was observed in 25.0% (n=5) of patients, while tumor number increase occurred in 20.0% (n=4), and 10.0% (n=2) of patients underwent surgical resection. Among patients receiving everolimus, tumor number reduction was observed in three patients (60.0%); however, the distribution of tumor number changes was not statistically significant (P=0.188).

Impact of Fetal Diagnosis on Tumor Dynamics

Comparisons were performed between patients with prenatal (fetal) diagnosis and those diagnosed postnatally. Tumor size outcomes differed between the two groups; however, this difference did not reach statistical significance (P=0.294). While postnatally diagnosed patients demonstrated heterogeneous tumor size trajectories during follow-up, all patients with fetal diagnosis showed tumor size reduction over the observation period. All patients diagnosed in the fetal period had rhabdomyomas (n = 3).

In contrast, tumor number dynamics differed significantly according to the timing of diagnosis. Among patients without fetal diagnosis, tumor number remained unchanged in 52.9%, increased in 23.5%, and decreased in 11.8% of cases. In patients with fetal diagnosis, tumor number reduction was observed in all cases, resulting in a statistically significant difference between groups (P=0.029).

The frequency of everolimus use was higher among patients with fetal diagnosis; however, this difference did not reach statistical significance (66.7% vs. 17.6%, P=0.140). The requirement for antiarrhythmic therapy during follow-up was similar between patients with and without fetal diagnosis (P=1.000).

Relationships Between Mass Characteristics and Cardiac Parameters

Mass Location, Outflow Tract Obstruction, and Arrhythmia Development

The relationship between mass localization, outflow tract obstruction, and the development of clinically significant arrhythmias detected on Holter monitoring was evaluated. No statistically significant association was identified between arrhythmia occurrence and involvement of the right ventricle, left ventricle, interventricular septum, interatrial septum, right atrium, or left atrium (all P>0.05). Similarly, neither tumor multiplicity (single vs. multiple masses) nor the presence of right or left ventricular outflow tract obstruction was associated with an increased risk

of clinically significant arrhythmia development (P=0.242 and P=1.000, respectively).

Tumor Size and Number in Relation to Echocardiographic Parameters

Correlation analyses demonstrated no significant association between tumor size and most echocardiographic structural or functional parameters, including interventricular septal thickness, left ventricular dimensions, wall thicknesses, systolic function indices, or cardiac chamber dimensions (all P>0.05). Similarly, comparisons between patients with single and multiple tumors revealed no statistically significant differences in left ventricular geometry, systolic or diastolic function, chamber dimensions, or flow velocities (all P>0.05).

However, significant positive correlations were identified between tumor size and mitral A-wave velocity (r=0.536, P=0.015), aortic outflow velocity (r = 0.520, P=0.019), and descending aortic flow velocity (r=0.569, P=0.009). No significant associations were observed between tumor size or tumor number and other diastolic parameters, z-scores, or pulmonary artery flow velocity.

Additionally, analyses based on mass localization (presence vs. absence of right ventricular, left ventricular, and interventricular septal involvement) demonstrated no statistically significant differences in echocardiographic parameters between groups (all P>0.05).

DISCUSSION

In this retrospective cohort of pediatric patients with intracardiac masses, several important findings emerged. Rhabdomyoma was the most frequently observed tumor type and was commonly associated with tuberous sclerosis complex, consistent with previous pediatric series [1–3]. The majority of patients were asymptomatic or mildly symptomatic at presentation, and clinically significant arrhythmias were observed in a minority of cases, reflecting the generally benign but potentially unpredictable clinical course of these tumors [1, 4]. Tumor behavior during follow-up was heterogeneous; while spontaneous regression was frequently observed—particularly among patients with fetal diagnosis—tumor growth or persistence occurred in a subset of patients, underscoring the need for individualized follow-up strategies [3, 5]. In selected cases, tumor regression was observed in most patients receiving everolimus therapy without treatment-limiting adverse effects, although this finding did not reach statistical significance. Nevertheless, these observations may suggest a potential role for everolimus as a non-surgical therapeutic option in selected patients with symptomatic tumors or hemodynamic compromise, consistent with recent literature supporting the efficacy of mTOR inhibitors in pediatric cardiac rhabdomyomas [7-9]. Finally, despite structural cardiac involvement, left ventricular systolic function remained preserved throughout follow-up, suggesting a generally favorable functional prognosis in carefully monitored patients.

The predominance of rhabdomyoma in our cohort is consistent with previously published pediatric cardiac tumor series [1, 2, 10]. The frequent association with tuberous sclerosis complex reflects the well-known genetic background and natural history of these tumors [3, 4, 11]. Despite their predominantly benign histology, pediatric cardiac tumors may present with a broad clinical spectrum, ranging from incidental findings to symptoms such as murmurs, exercise intolerance, or arrhythmias. The high proportion of asymptomatic patients in our cohort further highlights the important role of echocardiography in both the diagnostic evaluation and incidental detection of intracardiac masses in children [1, 10].

Cardiac arrhythmias represent one of the most clinically relevant complications of pediatric cardiac tumors [12-15]. In our cohort, clinically significant arrhythmias were observed in a minority of patients; however, ventricular arrhythmias were more frequently associated with fibromas, particularly those involving the interventricular septum. This observation is in line with previous studies describing fibromas as highly arrhythmogenic tumors due to their intramyocardial location and potential involvement of the cardiac conduction system [5, 6, 13]. While supraventricular ectopic activity was occasionally detected, ventricular ectopic activity and ventricular tachycardia constituted the arrhythmias of greatest

clinical concern and required antiarrhythmic therapy in selected patients [12, 13, 15]. These findings highlight the importance of systematic rhythm surveillance, including Holter monitoring, even in patients without overt arrhythmic symptoms [13, 15].

Tumor behavior during follow-up demonstrated marked heterogeneity. Spontaneous regression was frequently observed among patients with rhabdomyomas, particularly in those diagnosed during the fetal period, in accordance with the well-established natural course of these tumors [3, 11]. It should be noted that all patients diagnosed in the fetal period in our cohort had rhabdomyomas, a tumor type known for its high likelihood of spontaneous regression. Notably, all patients with fetal diagnosis in our cohort exhibited tumor number reduction, supporting the importance of early postnatal surveillance. However, since all patients diagnosed during the fetal period had rhabdomyomas, the relative contribution of prenatal diagnosis and tumor type to these favorable tumor dynamics remains unclear [3, 16]. In addition, everolimus therapy was associated with tumor regression in most treated patients, without treatment-limiting adverse effects. Although the number of patients receiving everolimus was limited, these findings are consistent with previous reports and support its role as a therapeutic option in selected patients with symptomatic tumors or significant hemodynamic compromise [6-8].

Although absolute cardiac dimensions increased over time on serial echocardiographic evaluation, the stability of the corresponding z-scores suggests that these changes are most plausibly explained by normal somatic growth during childhood rather than pathological remodeling [1, 10, 17, 18]. Importantly, left ventricular systolic function remained preserved throughout follow-up, consistent with previous pediatric series reporting favorable functional outcomes despite structural cardiac involvement [14, 17, 19]. These results indicate that myocardial function can remain stable in the majority of patients when appropriate longitudinal monitoring is maintained. The absence of progressive systolic dysfunction supports conservative management strategies in asymptomatic or mildly symptomatic patients and highlights the value of serial echocardiographic follow-up using z-score–adjusted parameters [1, 10, 17–19].

Strengths and Limitations

Despite its retrospective design and limited sample size, this study has several strengths, including a relatively long follow-up period and a comprehensive evaluation based on serial echocardiography and 24-hour Holter monitoring. The inclusion of both fetal and postnatal diagnoses enabled assessment of tumor behavior according to the timing of detection. However, the single-center retrospective design, the limited number of patients receiving everolimus, and the heterogeneity of tumor types and treatment strategies should be considered when interpreting the findings. In addition, the fact that all patients diagnosed in the fetal period had rhabdomyomas limits the ability to disentangle the independent effects of tumor type and timing of diagnosis on tumor regression patterns. In addition, the retrospective nature of echocardiographic data collection over a long study period may have limited the uniform availability of standardized imaging parameters. The relatively small sample size (n=20) may have limited the statistical power, particularly in subgroup analyses such as everolimus-treated patients and fetal diagnosis cases. The small sample size and heterogeneity of the study population also precluded the use of multivariable analysis.

CONCLUSION

In conclusion, pediatric intracardiac masses are predominantly benign but may demonstrate heterogeneous clinical and biological behavior. Rhabdomyoma was the most common tumor type and frequently exhibited spontaneous regression. Although favorable tumor dynamics were observed in patients diagnosed during the fetal period, the relative contribution of prenatal diagnosis and tumor type remains unclear. Clinically significant arrhythmias were uncommon but occurred more frequently in patients with fibromas, highlighting the importance of systematic rhythm surveillance. Tumor regression was observed in selected patients receiving everolimus therapy without treatment-limiting adverse effects; however, these findings should be interpreted cautiously given the limited sample size. Overall, preserved ventricular systolic function and favorable outcomes in most patients underscore the value of

individualized management strategies and careful long-term follow-up.

Ethics Approval and Consent to Participate

This study was approved by the Marmara University Faculty of Medicine Clinical Research Ethics Committee. (Decision No: 09.2022.462; date: 04.03.2022). All procedures were conducted in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments. Informed consent was waived because of the retrospective nature of the study and the analysis used anonymous clinical data.

Data Availability

All data generated or analyzed during this study are included in this published article. The data that support the findings of this study are available on request from the corresponding author, upon reasonable request.

Authors' Contribution

Study Conception: ŞA, FA; Study Design: ŞA, FA; Supervision: FA; Funding: N/A; Materials: ŞA, AMÇ; Data Collection and/or Processing: ŞA, AMÇ; Statistical Analysis and/or Data Interpretation: ŞA, FA; Literature Review: ŞA, AMÇ; Manuscript Preparation: ŞA; and Critical Review: FA.

Conflict of Interest

The author(s) disclosed no conflict of interest during the preparation or publication of this manuscript.

Financing

The author(s) disclosed that they did not receive any grant during the conduction or writing of this study.

Acknowledgments

The authors have no acknowledgments to declare.

Generative Artificial Intelligence Statement

The author(s) declare that no artificial intelligence-based tools or applications were used during the preparation process of this manuscript. The all content of the study was produced by the author(s) in accordance with scientific research methods and academic ethical principles.

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