

Pediatric Cardiac Masses: Clinical Characteristics and Long-Term Outcomes From a Two-Decade Single-Center Experience

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Conflict of Interest

The authors declare that they have no competing interests.

Ethics Approval

The study was conducted in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments. Ethical approval was obtained from the local Ethics Committee. Due to the retrospective nature of the study, the requirement for informed consent was waived.

(Etik kurul numarasını ekleyebilirsiniz.)

Consent to Participate

The requirement for informed consent was waived because of the retrospective design of the study.

Consent for Publication

Not applicable.

Data Availability Statement

The datasets generated and/or analyzed during the current study are available from the corresponding author on reasonable request.

Abstract

Background

Pediatric cardiac masses are rare and heterogeneous conditions with diverse clinical presentations, natural histories, and clinical courses. Although cardiac rhabdomyomas represent the most common pediatric cardiac tumor and are strongly associated with tuberous sclerosis complex, long-term data encompassing the broad spectrum of pediatric cardiac masses remain limited. We aimed to describe the clinical characteristics, treatment approaches, and long-term outcomes of pediatric cardiac masses encountered during two decades at a tertiary pediatric cardiology center.

Methods

This retrospective single-center study included pediatric patients diagnosed with cardiac masses between January 2006 and January 2026. Demographic characteristics, tumor subtype, anatomical localization, imaging findings, arrhythmias, treatment approaches, tumor behavior during follow-up, and clinical outcomes were retrospectively reviewed. Continuous variables were expressed as median values with interquartile ranges (IQR), whereas categorical variables were summarized as counts and percentages. Comparisons between cardiac rhabdomyomas and non-rhabdomyoma cardiac masses were performed using nonparametric statistical methods.

Results

Twenty-two pediatric patients were included, comprising 14 patients with cardiac rhabdomyomas and 8 patients with non-rhabdomyoma cardiac masses. Rhabdomyomas were diagnosed at a significantly younger age than non-rhabdomyoma lesions and were frequently associated with tuberous sclerosis complex. Arrhythmias occurred exclusively among patients with rhabdomyomas, whereas tumor regression was significantly more common in this group. Non-rhabdomyoma cardiac masses demonstrated greater pathological diversity and more frequently required surgical intervention or tissue diagnosis. Rare lesions encountered during the study period included pericardial tufted angioma, cardiac hydatid disease, and secondary cardiac involvement. Clinical management and long-term outcomes varied according to tumor subtype and individual patient characteristics.

Conclusion

Pediatric cardiac masses demonstrate substantial clinical and biological heterogeneity according to tumor subtype. Our two-decade single-center experience illustrates the diverse presentation, treatment approaches, and long-term outcomes of these uncommon conditions and highlights the value of individualized evaluation and longitudinal multidisciplinary follow-up.

Keywords: Pediatric cardiac tumors; cardiac rhabdomyoma; tuberous sclerosis complex; pediatric cardiac masses; arrhythmia; mTOR inhibitors.

Introduction

Pediatric cardiac masses are uncommon but clinically diverse conditions encompassing a broad spectrum of benign and malignant lesions with variable clinical presentation, biological behavior, and long-term outcomes. Although primary cardiac tumors are rare during childhood, advances in fetal and pediatric cardiac imaging have increased the frequency of incidental detection and facilitated earlier diagnosis [1,2].

Cardiac rhabdomyoma is the most common pediatric cardiac tumor and is strongly associated with tuberous sclerosis complex (TSC), with many lesions being identified during the fetal or neonatal period [2,3]. In contrast, non-rhabdomyoma cardiac masses comprise a heterogeneous group of entities including fibromas, teratomas, vascular tumors, parasitic lesions, and secondary cardiac involvement, each with distinct imaging characteristics, clinical manifestations, and natural histories [1,4,5]. Clinical presentation may range from asymptomatic incidental findings to arrhythmias, ventricular dysfunction, intracardiac obstruction, embolic events, or heart failure depending on tumor subtype, anatomical location, and relationship to surrounding cardiac structures [5,6].

Echocardiography remains the cornerstone of diagnosis and longitudinal follow-up, whereas cardiac magnetic resonance imaging and computed tomography may provide complementary information regarding tissue characteristics and anatomical extent in selected patients [7]. Management approaches vary considerably according to tumor subtype and clinical presentation. While many rhabdomyomas undergo spontaneous regression, other lesions may require medical treatment, surgical intervention, or prolonged surveillance because of hemodynamic compromise, arrhythmias, or diagnostic uncertainty [2,6,8].

Despite increasing recognition of pediatric cardiac masses, available evidence is largely derived from small retrospective series and isolated case reports, particularly for uncommon non-rhabdomyoma lesions [1,4]. Consequently, long-term single-center experiences encompassing the broad spectrum of pediatric cardiac masses remain valuable for illustrating the diversity of clinical presentation and follow-up encountered in routine practice.

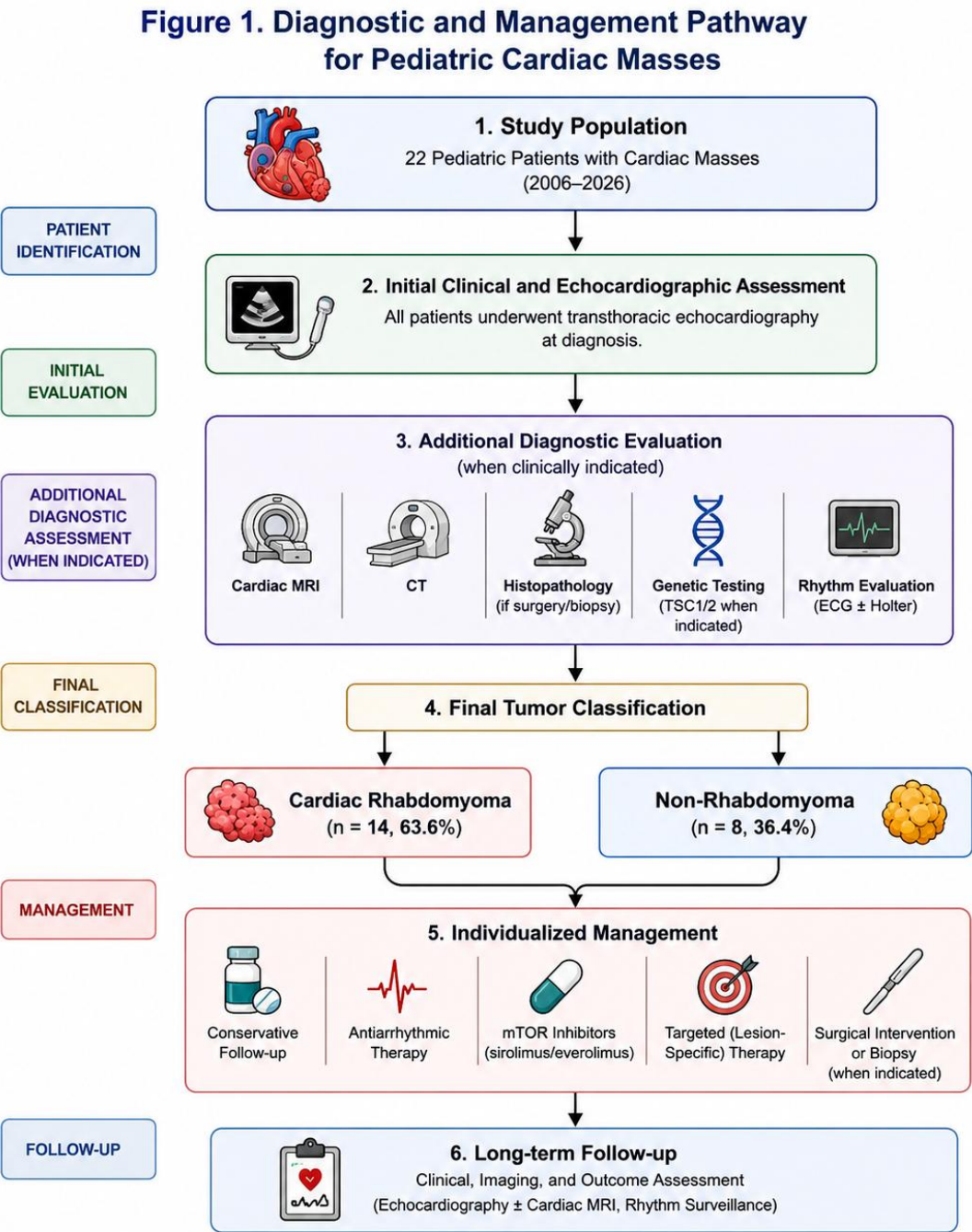
The aim of this study was to describe the clinical characteristics, imaging findings, treatment approaches, and long-term outcomes of pediatric cardiac masses diagnosed over a two-decade period at a tertiary pediatric cardiology center and to present our institutional experience within the context of the existing literature.

Materials and Methods

Study design and patient population

This retrospective single-center study included pediatric patients diagnosed with cardiac masses and followed at a tertiary pediatric cardiology center between January 2006 and January 2026. Patients younger than 18 years of age at diagnosis with echocardiographically confirmed cardiac masses were eligible for inclusion. Institutional medical records were retrospectively reviewed to identify consecutive eligible patients. Patients with insufficient clinical or imaging data preventing adequate assessment of diagnosis or follow-up were excluded. A total of 22 patients were included in the final analysis. The study was designed to

describe the clinical characteristics, imaging findings, clinical management, and long-term outcomes of pediatric cardiac masses encountered during the study period. The study flow and patient selection process are summarized in **Figure 1**.



Data collection

Demographic, clinical, imaging, treatment, and follow-up data were retrospectively collected from institutional electronic medical records and archived imaging databases. Data included patient characteristics, tumor morphology and anatomical localization, imaging findings, ventricular systolic function, arrhythmias, therapeutic interventions, histopathological diagnoses when available, and longitudinal tumor behavior during follow-up. Tumor behavior was classified as complete regression, partial regression, or no regression based on serial imaging findings. Complete regression was defined as complete disappearance of the lesion, whereas partial regression indicated a measurable reduction in tumor size without complete resolution.

Imaging assessment

All patients underwent transthoracic echocardiographic evaluation at diagnosis and throughout follow-up. Echocardiographic assessment included lesion size, multiplicity, anatomical localization, ventricular systolic function, valvular involvement, hemodynamic effects, and the presence of intracardiac obstruction. Additional imaging modalities, including cardiac magnetic resonance imaging and computed tomography, were performed when clinically indicated to further characterize lesion anatomy and extent. Cardiac magnetic resonance imaging was additionally used in selected patients for tissue characterization and longitudinal assessment when appropriate.

Clinical and genetic evaluation

Patients with tuberous sclerosis complex-associated cardiac rhabdomyomas underwent genetic evaluation, including TSC1 and TSC2 testing, together with multidisciplinary neurological follow-up. Standard 12-lead electrocardiography was performed in all patients at diagnosis, whereas ambulatory rhythm monitoring was obtained when clinically indicated. Arrhythmias were classified according to electrocardiographic findings and clinical documentation.

Clinical management

Clinical management was individualized according to tumor subtype, anatomical localization, clinical presentation, ventricular systolic function, arrhythmia burden, and hemodynamic significance. Patients without significant symptoms or hemodynamic compromise were generally managed with clinical and imaging follow-up. Medical treatment included antiarrhythmic agents, mTOR inhibitors, and lesion-specific targeted therapies when clinically indicated. The use of sirolimus or everolimus was determined through multidisciplinary clinical assessment and adapted according to individual patient characteristics and treatment response. Surgical intervention or tissue diagnosis was pursued according to individual clinical circumstances, including hemodynamic compromise, compressive effects, lesion progression, diagnostic uncertainty, refractory arrhythmias, or inadequate response to medical treatment. Histopathological confirmation was obtained in surgically treated or biopsied patients. In selected conservatively managed solitary fibromas, cardiac magnetic resonance imaging provided additional diagnostic support when histopathological confirmation was unavailable.

Statistical analysis

Continuous variables were expressed as median values with interquartile ranges (IQR), whereas categorical variables were summarized as counts and percentages. Given the small sample size, nonparametric methods were used for group comparisons. Comparisons between patients with cardiac rhabdomyomas and non-rhabdomyoma cardiac masses were performed using the Mann–Whitney U test for continuous variables and Fisher exact test for categorical variables, where appropriate. A two-sided p value of <0.05 was considered statistically significant.

Ethical approval

The study was conducted in accordance with the principles of the Declaration of Helsinki and was approved by the institutional ethics committee. Owing to the retrospective study design, the requirement for informed consent was waived.

Results

Between January 2006 and January 2026, 22 pediatric patients with cardiac masses were evaluated at our tertiary pediatric cardiology center. The cohort included a heterogeneous spectrum of cardiac tumors diagnosed during both the antenatal and postnatal periods, ranging from cardiac rhabdomyomas to rare non-rhabdomyoma lesions. Antenatal detection included tuberous sclerosis complex-associated rhabdomyomas as well as a rare case of pericardial tufted angioma. The baseline demographic and clinical characteristics of the study population are summarized in **Table 1**.

Table 1. Baseline demographic and clinical characteristics of the study population (n = 22)

Variable	Value
Total number of patients	22
Female sex, n (%)	9 (40.9%)
Male sex, n (%)	13 (59.1%)
Antenatal diagnosis, n (%)	5 (22.7%)
Postnatal diagnosis, n (%)	17 (77.3%)
Age at diagnosis (postnatal), months, median (IQR)	12 (2–84)
Body weight at diagnosis, kg, median (IQR)	5.9 (3.5–19.0)
Duration of follow-up, months, median (IQR)	36 (12–84)

Cardiac rhabdomyoma was the predominant tumor subtype, whereas the remaining patients comprised a diverse group of non-rhabdomyoma cardiac masses. Multiple lesions were mainly observed among patients with rhabdomyomas, while solitary lesions predominated in other tumor types. Cardiac masses involved various intracardiac structures, and several patients demonstrated involvement of more than one anatomical location. The distribution of tumor subtypes and anatomical characteristics are presented in **Table 2**.

Table 2. Tumor subtypes, lesion characteristics, and anatomical distribution

Characteristic	n (%)
Tumor type	
Cardiac rhabdomyoma	14 (63.6%)
Cardiac fibroma	4 (18.2%)
Hydatid cyst	1 (4.5%)
Teratoma	1 (4.5%)
Metastatic cardiac involvement	1 (4.5%)
Pericardial tufted angioma	1 (4.5%)
Number of lesions	
Single lesion	11 (50.0%)
Multiple lesions	11 (50.0%)
Tumor localization*	
Left ventricular free wall	13 (59.1%)
Right ventricular free wall	11 (50.0%)
Interventricular septum	8 (36.4%)
Mitral valve	4 (18.2%)
Tricuspid valve	4 (18.2%)
Right atrium	2 (9.1%)
Pericardial involvement	1 (4.5%)
Interatrial septum	1 (4.5%)

* Some patients demonstrated involvement of multiple anatomical locations.

Transthoracic echocardiography served as the primary diagnostic and follow-up imaging modality in all patients. Cross-sectional imaging with cardiac magnetic resonance imaging or computed tomography was selectively performed to further characterize tumor morphology, define the extent of involvement, and support clinical evaluation. Although ventricular systolic function was generally preserved, selected patients demonstrated clinically relevant findings including multicentric tumor involvement, ventricular dysfunction, valvular involvement, and intracardiac obstruction. The echocardiographic characteristics of the study population are summarized in **Table 3**.

Table 3. Echocardiographic and anatomical characteristics according to tumor subtype

Echocardiographic parameter	Rhabdomyoma (n = 14)	Non-rhabdomyoma (n = 8)	p value
Left ventricular ejection fraction (%), median (IQR)	68 (66.3–70.0)	68 (68.0–73.5)	NS
Left ventricular systolic dysfunction (EF <55%), n (%)	1 (7.1)	0 (0)	NS
Multicentric tumor involvement, n (%)	9 (64.3)	0 (0)	0.002
Left ventricular free wall involvement, n (%)	4 (28.6)	1 (12.5)	NS
Interventricular septum involvement, n (%)	8 (57.1)	2 (25.0)	NS
Interatrial septum involvement, n (%)	0 (0)	1 (12.5)	NS
Valve-related tumor involvement, n (%)	3 (21.4)	1 (12.5)	NS
Mild valvular regurgitation (any valve), n (%)	6 (42.9)	2 (25.0)	NS
Intracardiac obstruction, n (%)	2 (14.3)	1 (12.5)	NS

Abbreviations: EF, ejection fraction; IQR, interquartile range; NS, not significant

Clinically significant hemodynamic effects were observed in a limited number of patients. These included ventricular systolic dysfunction associated with a large left ventricular rhabdomyoma, left ventricular outflow tract obstruction related to an interventricular septal rhabdomyoma, and right ventricular outflow tract obstruction caused by a right ventricular teratoma. A rare case of pericardial tufted angioma was diagnosed after antenatal detection of pericardial effusion and was evaluated using multimodality imaging and histopathological examination, with regression observed during follow-up.

Cardiac arrhythmias represented the most frequent cardiac complication and were observed exclusively in patients with cardiac rhabdomyomas. Overall, six patients (27.3% of the cohort and 42.9% of the rhabdomyoma group) developed rhythm disturbances, whereas no arrhythmias occurred among patients with non-rhabdomyoma cardiac masses. Arrhythmias ranged from isolated ventricular ectopy to supraventricular and ventricular arrhythmias, including Wolff–Parkinson–White pattern. Clinical management was individualized according to arrhythmia burden, tumor location, ventricular function, and associated hemodynamic findings, with antiarrhythmic therapy administered when clinically indicated.

Longitudinal follow-up demonstrated substantial variability in tumor behavior. Cardiac rhabdomyomas frequently showed spontaneous regression, whereas non-rhabdomyoma lesions generally remained stable or required intervention because of diagnostic uncertainty or clinical indications. Complete tumor regression was observed in five patients and partial

regression in six patients, while eleven lesions remained stable without significant regression. Regression occurred predominantly among rhabdomyomas, whereas only one non-rhabdomyoma lesion demonstrated partial regression during follow-up.

Clinical management varied according to tumor subtype and individual clinical characteristics. Conservative follow-up was the predominant strategy, whereas antiarrhythmic therapy, mTOR inhibition, surgical intervention, diagnostic biopsy, and lesion-specific targeted treatments were implemented when clinically indicated. Overall management strategies and outcomes are summarized in Table 4.

Table 4. Overall management strategies and clinical outcomes of the study cohort

Variable	n (%)
Any documented arrhythmia	6 (27.3%)
Arrhythmia in rhabdomyoma group	6/14 (42.9%)
Arrhythmia in non-rhabdomyoma group	0/8 (0%)
Antiarrhythmic therapy required	6 (27.3%)
mTOR inhibitor therapy (sirolimus/everolimus)	9 (40.9%)
Surgical intervention or diagnostic biopsy	6 (27.3%)
Tumor regression	
Complete regression	5 (22.7%)
Partial regression	6 (27.3%)
No regression	11 (50.0%)

Comparative analysis between cardiac rhabdomyomas and non-rhabdomyoma cardiac masses demonstrated distinct clinical patterns. Rhabdomyomas were diagnosed at a significantly younger age and were more frequently associated with antenatal detection, multifocal involvement, arrhythmias, and spontaneous regression. In contrast, non-rhabdomyoma lesions more commonly required surgical intervention or tissue diagnosis because of their heterogeneous pathological characteristics and variable clinical behavior.

Overall, the cohort demonstrated substantial variability in clinical presentation, imaging findings, management strategies, and long-term outcomes, reflecting the heterogeneous nature of pediatric cardiac masses encountered during this two-decade single-center experience.

Discussion

Pediatric cardiac masses are rare and heterogeneous lesions with variable clinical presentation, anatomical distribution, biological behavior, and clinical course. In this two-decade single-center experience, cardiac rhabdomyoma was the most frequently encountered

diagnosis, consistent with previous pediatric series reporting rhabdomyoma as the predominant cardiac tumor during infancy and childhood [1,2]. Beyond rhabdomyomas, our cohort included several uncommon non-rhabdomyoma lesions, reflecting the broad diagnostic spectrum that may be encountered in tertiary pediatric cardiology practice. Secondary cardiac involvement, although generally more common than primary malignant cardiac tumors, remains infrequently represented in pediatric single-center cohorts and was also observed in our series [3].

Antenatal diagnosis was predominantly associated with rhabdomyomas and tuberous sclerosis complex, supporting the well-established relationship between fetal cardiac rhabdomyomas and TSC [4]. Multifocal tumor involvement was also mainly observed in this subgroup and may provide an important diagnostic clue in affected patients. In our experience, these children required coordinated multidisciplinary follow-up involving cardiology, neurology, and genetic evaluation, reflecting the multisystem nature of the disease rather than an isolated cardiac lesion.

Although ventricular systolic function was preserved in most patients, selected tumors were associated with clinically significant hemodynamic effects. Ventricular dysfunction, inflow or outflow tract obstruction, and valvular involvement were closely related to anatomical localization and the relationship of the lesion to adjacent cardiac structures rather than tumor subtype alone. These observations are in agreement with previous reports emphasizing the importance of tumor size, location, and myocardial involvement in determining clinical impact [5,6]. In selected patients, cardiac magnetic resonance imaging provided additional anatomical and tissue characterization, particularly when histopathological confirmation was unavailable or when further evaluation was required [7].

Arrhythmias represented an important clinical feature of rhabdomyomas in our cohort. Rhythm disturbances ranged from ventricular ectopy to more complex supraventricular and ventricular arrhythmias, including Wolff–Parkinson–White pattern. Previous studies have suggested that myocardial involvement and proximity to the conduction system may contribute to the arrhythmogenic potential of cardiac rhabdomyomas [8,9]. In our cohort, the management of arrhythmia-associated rhabdomyomas varied according to the clinical characteristics of individual patients. Some patients remained clinically stable with antiarrhythmic treatment and longitudinal follow-up, whereas mTOR inhibition was used in selected patients according to ventricular function, tumor burden, obstruction, and overall clinical status.

Tumor behavior during follow-up differed substantially according to pathological subtype. The tendency of rhabdomyomas to undergo spontaneous regression was consistent with their recognized natural history and previous reports in the literature [10]. In contrast, non-rhabdomyoma lesions represented a biologically heterogeneous group that included fibroma, teratoma, hydatid disease, vascular tumors, and secondary cardiac involvement. Consequently, comparisons between rhabdomyoma and non-rhabdomyoma groups should be interpreted as reflecting broad clinical phenotypes rather than biologically equivalent disease entities.

One particularly uncommon lesion encountered in our cohort was pericardial tufted angioma, a rare benign vascular tumor of infancy with exceptionally uncommon cardiac involvement [11,12]. In this patient, prenatal pericardial effusion preceded postnatal diagnosis, and multimodality imaging together with histopathological confirmation was essential for clinical

evaluation. The observed regression following sirolimus and propranolol treatment adds to the limited clinical experience with targeted medical therapy in selected vascular tumors, although available evidence remains limited and further studies are needed [13–16].

Cardiac hydatid disease represented another rare diagnosis in our series. Although cardiac involvement in *Echinococcus granulosus* infection is uncommon, it may carry serious risks including rupture, embolization, and anaphylaxis, and combined surgical and antiparasitic treatment has been described as the conventional approach in reported cases [17]. The presence of hydatid disease, metastatic involvement, and rare vascular pathology in our cohort further illustrates the broad differential diagnosis of pediatric cardiac masses and the variety of clinical scenarios encountered in tertiary referral practice.

Overall, our two-decade single-center experience illustrates the broad clinical spectrum of pediatric cardiac masses encountered in routine pediatric cardiology practice. Beyond the predominance of cardiac rhabdomyomas, our cohort included several uncommon lesions with distinct imaging characteristics, biological behavior, and clinical courses. The diversity of these conditions emphasizes that evaluation and follow-up should be adapted to the characteristics of the individual lesion and patient. We believe that sharing our institutional experience contributes to the existing literature and provides additional insight into the varied presentation, longitudinal behavior, and real-world clinical management of these rare conditions.

Conclusion

Pediatric cardiac masses represent a rare and heterogeneous group of conditions with diverse clinical presentations, imaging characteristics, and clinical courses. In our two-decade single-center experience, cardiac rhabdomyomas were the most frequently encountered lesions, whereas non-rhabdomyoma cardiac masses comprised a broad spectrum of uncommon entities with distinct biological behavior and clinical characteristics.

Our findings illustrate that the clinical course of pediatric cardiac masses is influenced not only by histopathological subtype but also by anatomical location, ventricular function, arrhythmia burden, and associated hemodynamic effects. The inclusion of rare vascular, parasitic, and secondary cardiac lesions further reflects the diversity of conditions encountered in tertiary pediatric cardiology practice.

This study presents our institutional experience with the evaluation, follow-up, and clinical management of a rare and heterogeneous patient population. We believe that sharing this two-decade experience contributes to the existing literature and provides additional insight into the varied presentation, longitudinal behavior, and real-world clinical practice associated with pediatric cardiac masses.

Limitations

This study has several limitations. First, its retrospective single-center design and relatively small sample size limit the generalizability of the findings. Second, the marked heterogeneity

of non-rhabdomyoma cardiac masses precludes meaningful subtype-specific statistical comparisons. Third, histopathological confirmation was not available for all conservatively managed lesions, with diagnosis in selected cases supported by multimodality imaging findings. Finally, changes in imaging techniques and management strategies over the two-decade study period may have influenced clinical practice and follow-up approaches.

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Figure Legends

Figure 1. Diagnostic and management pathway for pediatric cardiac masses.

Flow diagram illustrating the diagnostic evaluation, classification, individualized management, and long-term follow-up of pediatric patients with cardiac masses included in this two-decade single-center study. Additional diagnostic investigations, including cardiac magnetic resonance imaging (MRI), computed tomography (CT), histopathological evaluation, genetic testing, and rhythm assessment, were performed when clinically indicated.