

Primary Cardiac Tumors in Children: A State-of-the-Art Review

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Cardiac tumors are benign or malignant neoplasms arising from the endocardium, myocardium, or the surrounding pericardium. They can be primary or metastatic. Primary tumors are rare, with a reported autopsy frequency of up to 0.30%. These tumors can occur in the neonatal or fetal period, with an incidence of 0.14% in the latter. Most pediatric cardiac tumors are benign, with approximately 10% being malignant. Rhabdomyoma is the most prevalent benign tumor in children and accounts for more than 50% of all primary cardiac tumors in this population. Multiple cardiac tumors noted during the fetal or neonatal period are commonly associated with tuberous sclerosis. Additional risk factors include family history and other genetic predispositions such as the Carney complex. When clinically suspected, echocardiography, computed tomography, and magnetic resonance imaging help with diagnosis. Treatment includes surgery, chemotherapy, and targeted therapy. This article provides a scoping review of cardiac tumors in children, their epidemiology, histological patterns, diagnosis, and treatment.

Keywords: Cardiac Tumors; Echocardiography; Rhabdomyoma; Atrial Fibrillation; Arrhythmias

Introduction

Cardiac tumors in children are rare, arising most commonly from the endocardium, myocardium, or the surrounding pericardium (1, 2). These can be primary or metastatic, with primary tumors accounting for a prevalence between 0.0017% and 0.28% in autopsy series (3). In the fetal period, the incidence of cardiac tumors has been reported to be up to 0.14% (3, 4). The overwhelming majority of primary cardiac neoplasms in children are benign, with

up to 10% being malignant (5). Secondary malignant tumors are much more prevalent than primary malignant neoplasms, and the type and frequency of cardiac tumors differ in adults and children (3-5).

In adults, 75% of tumors are benign, and 25% are malignant (3-6). Myxomas are the most common primary tumor in adults, and sarcomas constitute up to 75% of malignant masses (7-9). Rhabdomyoma is the most

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common cardiac neoplasm during childhood and fetal life, followed by fibroma and teratoma (Figure 1) (10, 11). Patients with cardiac masses may be asymptomatic or present with heart failure, cardiac murmurs, sudden cardiac death (SCD), or arrhythmias (12). Echocardiography is the primary diagnostic modality, and cardiac magnetic resonance imaging (cMRI) further characterizes the tumor and the extent of adjacent cardiac structure to support treatment planning (13, 14). Management varies from conservative to resection or transplantation. This article provides a scoping review of cardiac tumors in children, including their diagnosis and treatment.

Epidemiology

Primary cardiac tumors (PCTs) are scarce in the pediatric population, with a prevalence of up to 0.28% in various autopsy series (3). In addition, the incidence of cardiovascular neoplasms during fetal life has been reported to be approximately 0.14% in autopsy series (3, 4). Most PCTs are benign, with an estimated

10% malignant (3,4,15). Cardiac tumor types and frequencies differ between children and adults. In the adult cohort, benign primary tumors constitute up to 75% of cardiac neoplasms, while malignant tumors represent approximately 25% (16, 17).

Furthermore, myxomas are the most common primary neoplasms in adults, accounting for about 40% of benign tumors, in contrast with neonates and fetuses, where they are exceedingly rare (18). Rhabdomyoma is the most common PCT in childhood and fetal life, accounting for nearly 70% of primary neoplasms, followed by teratoma, fibroma, and hemangioma (19, 20). Similar to myxoma, rhabdomyomas are exceedingly rare in adults, and secondary malignant cardiac tumors are up to 20x more frequent than primary malignant tumors (19-21). Over the past several years, the incidence of PCTs has increased noticeably (22). Part of this trend may be attributed to advances in imaging techniques, with more sophisticated imaging being readily accessible.

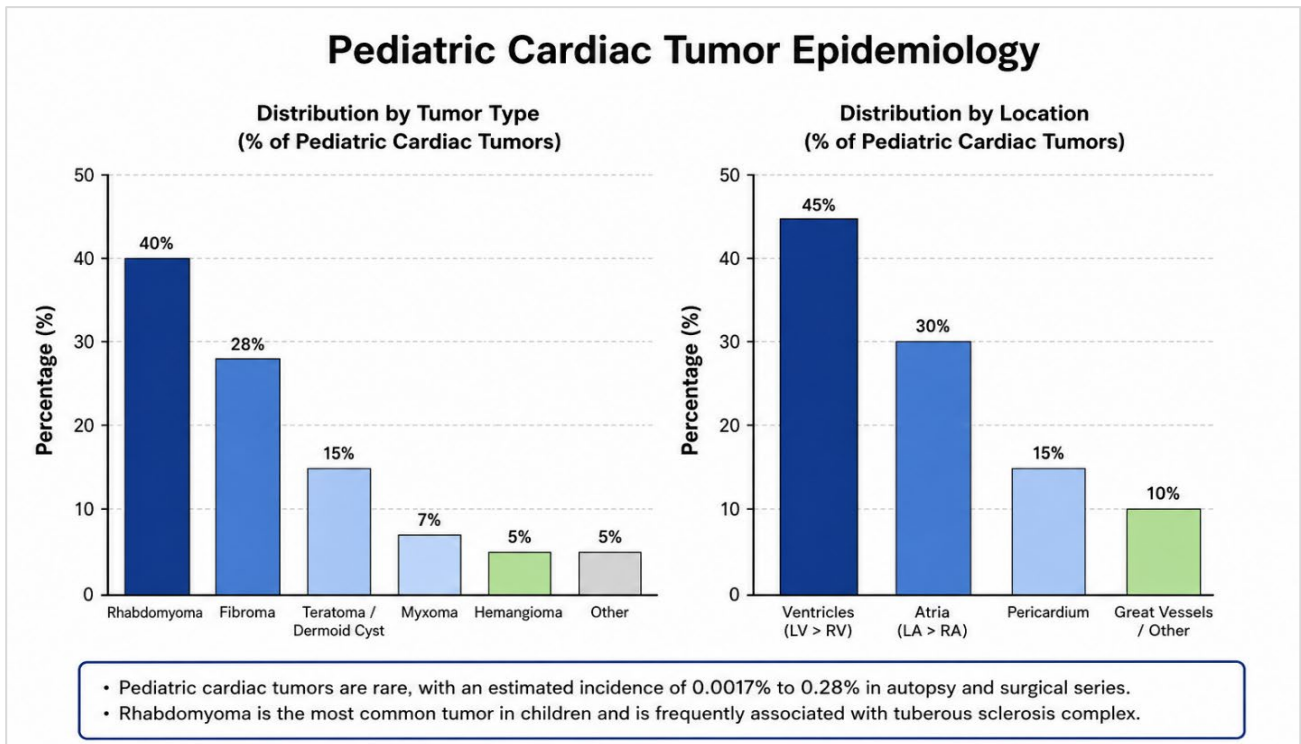


Figure 1. Bar diagram showing the epidemiology and location distribution of pediatric cardiac tumors. LV = Left ventricle; RV = Right ventricle; LA = Left Atrium; RA = Right Atrium

Clinical Manifestations

In general, cardiac tumors may appear in fetal or postnatal life. Some tumors may be symptomatic or found incidentally during evaluation for another reason. Symptoms usually relate to the tumor's cardiac location, although some may cause constitutional symptoms. The clinical manifestations of cardiac neoplasms in fetal life may include congestive heart failure (CHF), arrhythmias, and hydrops fetalis (23, 24). In postnatal life, tumors may present with myocardial dysfunction, respiratory distress, murmurs, cyanosis, arrhythmias, valvular insufficiency, and SCD due to in- or outflow obstructions (23-25). Ventricular tumors more commonly may obstruct either the left or right outflow tract, which may present as chest pain or syncope. Large atrial tumors can obstruct atrioventricular (AV) valvular flow and mimic valvular stenosis.

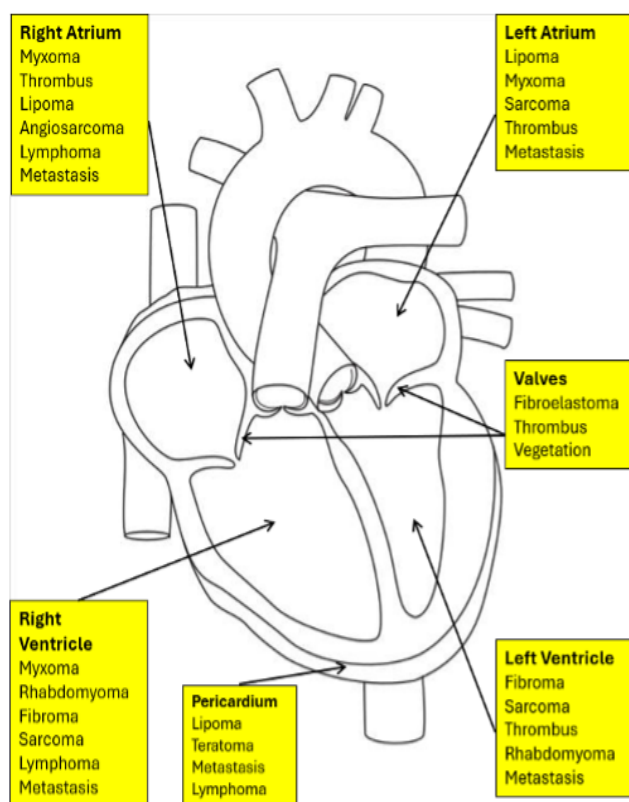


Figure 2. Illustrative diagram showing the most common locations of primary cardiac tumors in children.

From an arrhythmia standpoint, intracavity or intramyocardial neoplasm can affect cardiac rhythm through myocardial irritation or direct infiltration of conduction tissue, which may result in AV block and ventricular tachycardia. However, presentation may also be direct SCD. The presence of ventricular arrhythmias should always lead to a search for structural heart disease, where cardiac tumors are sometimes incidentally found (26, 27). Patients may also present with pulmonary and/or systemic thromboembolism from the tumor (28). Multiple minuscule emboli may mimic endocarditis or vasculitis, while larger fragments may lead to ischemic strokes when left-sided tumors are involved (28,29). Right-sided tumors classically mobilize to the lungs, which may produce pleuritic symptoms and possible right heart failure.

Lastly, patients may present with systemic symptoms such as fever, arthralgia, weight loss, and fatigue, as well as paraneoplastic syndromes (30, 31). Hematologic abnormalities, including polycythemia, anemia, leukocytosis, thrombocytosis, and elevated inflammatory markers (i.e., C-reactive protein and erythrocyte sedimentation rate), may also occur (30-32).

Diagnostic Work-up

The clinical presentation and prognosis of patients with cardiac tumors vary depending on lesion location and histologic type (Figure 2). Additionally, the setting provides diagnostic clues that help determine the lesion or tumor's etiology. For example, vegetation may be observed in patients with prosthetic or native valve disease who present with bacteremia and elevated inflammatory markers. In contrast, cardiac thrombi are more likely to be associated with arrhythmias, such as atrial fibrillation, or a left ventricular apical infarct with hypokinesis or akinesis. If a cardiac mass is ultimately found, determine its etiology by considering four factors: the patient's age, the tumor's location, tissue characterization based on cMRI, and the

epidemiological likelihood or clinical probability of the tumor (Tables 1-2) (33).

Primary Benign Cardiac Tumors

Rhabdomyoma

Cardiac rhabdomyoma is the most common primary cardiac tumor in children, accounting for more than 60% of PCTs (10,11,34). It is most commonly associated with tuberous sclerosis complex, an autosomal dominant disorder characterized by noncancerous tumors or lesions in the brain and other areas (35). Rhabdomyomas are typically located within the ventricles. On echocardiogram, they appear as homogeneous, well-circumscribed echogenic masses in the ventricular myocardium (36). Computed tomography (CT) or MRI can better delineate the relationship of the masses with the amount of involved myocardium and adjacent structures. These patients should also

undergo whole-body imaging to rule out other benign tumors and malignancies, such as renal cell carcinoma and breast and thyroid cancer, among others (37-39). Rhabdomyomas show pathognomonic spider cells with centrally located cytoplasm, with the nucleus and myofibrils radiating to the cell wall (Figure 3A) (40). Clinical presentation varies with the tumor's position, number, and size. Their presence at the AV junction can sometimes enable the tumors to act like an accessory pathway, which can cause tachyarrhythmias (41). Cardiac rhabdomyomas can lead to CHF and infant respiratory distress (10,11,40,41). Given their high rate of spontaneous regression after birth, they can be managed conservatively with echocardiography and electrocardiogram monitoring (42). Surgical intervention should be reserved for only hemodynamic compromise, intractable arrhythmias, or severe obstruction.

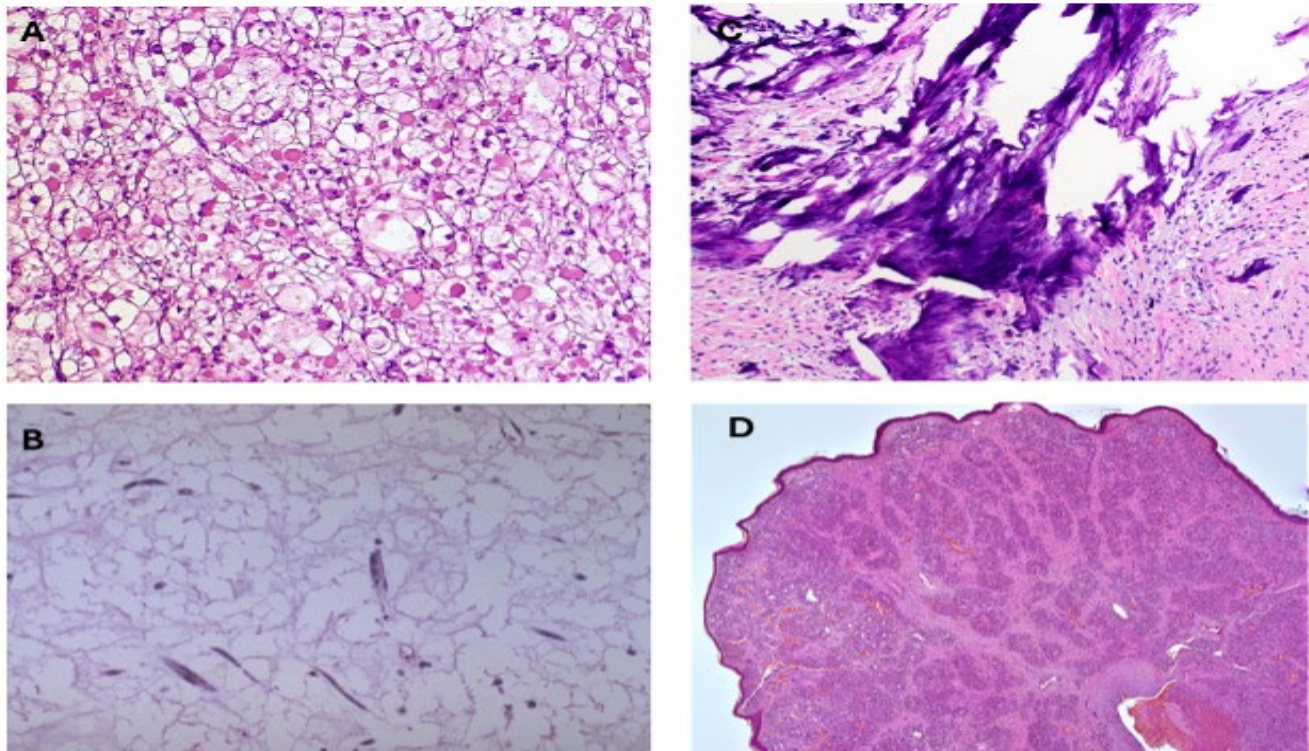


Figure 3. (A) Electron microscopy showing cardiac rhabdomyomas composed of large, vacuolated cells with abundant glycogen (spindle cells). (B) High-power microscopic appearance of cardiac myxoma shows minimal cellularity. There are scattered spindle cells with scant pink cytoplasm present in a loose myxoid stroma. (C) Dystrophic calcifications are sometimes seen in cardiac fibromas (Hematoxylin and Eosin stain, 20x). (D) Low-power view of a lobular capillary hemangioma with a prominent, thick-walled, large-caliber feeding vessel in the deep part of the lesion.

Myxoma

Cardiac myxomas are common benign PCTs most often found in the left atrium (Figure 3B) (43). Myxomas are sporadic; however, a familial incidence has been described in up to 7% of all cardiac myxomas, sometimes associated with Carney complex, a rare genetic disorder caused by mutations in the *PRKAR1A* gene, which causes pigmentation changes, benign tumors, and endocrine abnormalities (33,44,45). The tumors are pedunculated with a short, broad-based attachment to the atrial wall with a gelatinous consistency. On echocardiography, myxomas appear as a mobile mass attached to the endocardial surface by a stalk, usually originating from the fossa ovalis (46). More than 50% of patients with left atrial myxomas experience symptoms, which can include dyspnea and orthopnea from mitral valve stenosis or obstruction (47). Fragmentation of tumor may embolize and cause cerebrovascular events. Surgical excision remains the mainstay of treatment and leads to excellent long-term survival, with most patients having a good prognosis (48).

Fibroma

Cardiac fibromas are solitary neoplasms derived from connective tissue fibroblasts, which are mainly located within the ventricular septum (49). They are commonly seen in childhood and range from 1 to 10 cm (49, 50). On the macroscopic level, fibromas appear clearly demarcated from the myocardium. Microscopically, interdigitations surround the myocardium, and calcification of the central portion of the neoplasm is pathognomonic, reflecting poor blood supply to the tumor (Figure 3C) (51). On echocardiography, they appear as a large, well-circumscribed solitary mass in the ventricular free wall or septum (52). A solitary, large, homogeneous myocardial mass centered within the ventricles is typically evident on CT or MRI (52, 53).

Fibromas can invade the ventricular muscle, replace functional myocardium, and potentially cause refractory cyanosis or CHF (49-51). Occasionally, the tumor may extend into the ventricular conduction system, resulting in AV block or arrhythmias (50-54). These neoplasms commonly remain dormant, and spontaneous regression rarely occurs. As a result, surgical resection is usually recommended (55). The progressive loss of functional myocardial fibers and heart failure does not resolve following resection, which may lead to consideration of heart transplantation.

Papillary fibroelastoma

Papillary fibroelastoma is one of the most common PCTs (56). They are now increasingly recognized as imaging quality and techniques improve over the years. On gross inspection, they appear as small intracavity masses that are usually firm, with a short pedicle and multiple papillary fronds (57). These tumors are typically small and rarely present clinically with signs of obstruction. Cases treated with surgery occur in patients with symptoms related to cerebral ischemia (58).

Hemangioma

Angiomas are benign neoplasms composed predominantly of blood vessels (59). They are most frequently found in the right atrium and ventricular septum, and are most often discovered incidentally (59, 60). Multiple skin hemangiomas in neonates should prompt work-up for cardiac hemangiomas (60, 61). On histology, they appear as a proliferation of blood vessels lined by endothelial cells, which may be large, thin-walled vessels (such as cavernous hemangioma, the most common type), small capillaries (capillary hemangioma), or dysplastic malformation of veins and arteries (arteriovenous hemangioma) (Figure 3D) (62). Hemangiomas are benign tumors that typically regress spontaneously and have a good prognosis (60-62).

Lipoma

Lipomas are benign tumors predominantly composed of mature fat cells (63). They can appear at any age and occur with equal frequency in males and females. Lipomas may occur anywhere in the heart and present clinically depending on the tumor's location. MRI is commonly used to diagnose lipomas, which clearly demonstrate fatty tissue (64). These tumors are commonly silent, although they can sometimes cause arrhythmias and AV blocks (63, 64).

Primary Malignant Cardiac Tumors

Angiosarcoma

Angiosarcoma is an aggressive neoplasm that frequently originates in the right atrium and has a high likelihood of metastases at the time of presentation (65). Histopathology commonly shows a tumor that is highly vascularized with myocardial infiltration, pleomorphism, necrosis, and mitosis (66). Echocardiography usually reveals a nodular or lobulated mass in the right atrium near the inferior vena cava (67). CT or MRI shows arterial-phase enhancement, which improves diagnostic accuracy (68, 69). Clinical presentation is often nonspecific and may include symptoms of pericardial involvement or the right-sided heart failure (66, 67). Local pulmonary, pleural, and mediastinal metastases are also common (66-69). Treatment typically involves a combination of surgery and radiation, along with chemotherapy (68-70).

Leiomyosarcoma

Leiomyosarcoma is a primary sarcoma of smooth muscle cells typically originating in two sites (71). It can form in the left atrium, mimicking left atrial myxoma and its symptoms, or the pulmonary infundibulum and artery, mimicking a pulmonary embolism (72, 73). Dyspnea is the main complaint, but patients may sometimes present with pleuritic chest pain, hemoptysis, and palpitations from tachyarrhythmias such as atrial fibrillation. Treatment

includes surgical excision (74). Chemoradiation therapy may provide palliation (72-74).

Rhabdomyosarcoma

Cardiac rhabdomyosarcomas are the most common malignant cardiac tumors in children and often affect the valves (75). These are malignant tumors of striated muscle that account for approximately 4% to 7% of cardiac sarcomas, with a male prevalence (75, 76). On gross inspection, they appear mucoid or gelatinous in consistency. Histologically, the embryonal type shows rhabdomyoblasts with abundant glycogen and expression of desmin, myoglobin, and myogenin (75). Patients may present with symptoms of pericardial disease, pleural effusion, or embolic phenomena but could also manifest systemic illness, including fever, weight loss, and malaise. Compared with angiosarcoma, diffuse pericardial involvement is uncommon, and the neoplasm rarely invades beyond the parietal pericardium. The prognosis is generally poor (77).

Fibrosarcoma

Fibrosarcomas are mesenchymal tumors that originate from fibroblasts and are generally seen within the left or right heart chambers (78). Clinical presentation varies based on tumor location and size. Symptoms of mitral stenosis, pulmonary congestion, and pulmonary vein obstruction are common with left chamber involvement (79). The prognosis of cardiac fibrosarcoma is generally poor, with a median survival time of less than a year (80).

Undifferentiated pleomorphic sarcoma

Undifferentiated cardiac sarcomas are invasive and possess no specific histological pattern (81,82). They are most often seen in the left atrium, and most patients present with pulmonary congestion. The prognosis is generally poor despite surgery and palliative chemotherapy (81-83). Mean postoperative survival ranges from 5 to 18 months (81-84).

Table 1. Clinical features, incidence, gender distribution, associated genetic syndromes, histopathology, and required surveillance in childhood, infancy, and adults of primary benign cardiac tumors modified from Tyeally *et al* (33).

Primary benign cardiac tumors	Clinical features	Incidence in children (%)	Incidence in infancy (%)	Incidence in adults (%)	M/F	Associated syndromes	Histopathology	Required surveillance
Rhabdomyoma	Might result in cardiomegaly, CHF, arrhythmias, SCD, or still birth	42	62	1	n/a	Tuberous sclerosis complex	Spider cell (vacuolated enlarged cardiac myocyte with clear cytoplasm)	TTE every 1-3 years in asymptomatic pediatric patients until resolution of rhabdomyoma is documented. Serial echocardiography is not recommended in the first year of life in the absence of clinical symptoms
Cardiac myxoma	Embolic, obstructive cardiac, or systemic symptoms. Pulmonary edema, tachyarrhythmias, and progressive heart failure	17	0	52	1:2	Carney complex	Spindle or stellate cells, pseudo-vascular structure, myxoid matrix, hemorrhage Dystrophic calcification can be present	All myxomas: after surgery, TTE should document complete tumor resection and follow-up echo are recommended every 6 months for a minimum of 4 years. Carney complex: TTE should start during the first 6 months of life and be performed at least once a year thereafter
Cardiac fibroma	33% are asymptomatic. The others usually present with chest pain, arrhythmia, syncope, heart failure, cyanosis, and SCD	18	17	3	1:1	Nevoid basal cell carcinoma syndrome (i.e., Gorlin-Goltz)	Fibroblasts and collagen bundles, some elastic fibers, calcification is a common finding	Baseline TTE should be obtained at the time of diagnosis, ideally within the first 6 months
Papillary fibroelastoma	n/a	n/a	n/a	10	1:1	n/a	Endocardium-coated fronds with an avascular collagenous core containing mucopolysaccharide and elastin	n/a
Hemangioma	Arrhythmias, shortness of breath, and pericarditis	5	4	6	1:1	Kasabach-Merritt/mutation gene 9q21		Periodic TTE is recommended
Lipoma	Commonly asymptomatic. Symptoms vary based on location and size	Rare	n/a	16	1:1	n/a	Mature adipocyte, occasionally with entrapped myocytes at the periphery	n/a

Abbreviations. M: Male; F: Female; CHF: Congestive heart failure; SCD: Sudden cardiac death; n/a: Not applicable; TTE: Transthoracic echocardiogram.

Table 2. Clinical features, incidence, gender distribution, and histopathology in childhood, infancy, and adults of primary malignant cardiac tumors, modified from Tyebally *et al* (33).

Primary Malignant Cardiac Tumors	Clinical features	Incidence in children (%)	Incidence in infancy (%)	Incidence in adults (%)	M/F	Histopathology
Angiosarcoma	Non-specific symptoms	6	n/a	26	2:1	Highly vascularized, myocardial infiltration, pleomorphism, necrosis, and mitosis
Leiomyosarcoma	SOB, chest pain, right heart failure, valve stenosis, arrhythmias, and SCD	n/a	17	5	1:1	Compact bundles of spindled cells with blunt nuclei, regions of necrosis, and mitotic figures with epithelioid regions are often present
Rhabdomyosarcoma	Non-specific symptoms	41	50	11	n/a	Embryonal type with rhabdomyoblasts containing abundant glycogen and expressing desmin, myoglobin, and myogenin
Fibrosarcoma	Pulmonary congestion	18	17	8	n/a	n/a
Undifferentiated sarcoma	Pulmonary congestion	6	Rare	6	1:1	Consists of undifferentiated plump spindle cells with frequent mitotic activity and (often) nuclear pleomorphism

Abbreviations. M: Male; F: Female; SOB: Shortness of breath; SCD: Sudden cardiac death; n/a: Not applicable

Limitations of Review

The available literature on pediatric cardiac tumors is limited by their rarity and small sample sizes, with most published evidence comprising single-center case series, reviews, and multicenter collaborations that still enroll modest cohorts. Primary cardiac tumors occur in approximately 0.0017–0.28% of pediatric autopsy series and represent fewer than 0.1% of pediatric cardiac surgeries (3, 85). The largest multicenter cardiovascular magnetic resonance study to date included 213 confirmed pediatric cardiac masses across 31 international centers, yet even this substantial effort reflects the aggregation of rare cases over many years (86).

Single-institution surgical series typically report 16–120 patients accumulated over 2–3 decades, and malignant histotypes such as primary cardiac Ewing sarcoma are documented in fewer than 20 cases worldwide (85, 87–89). This data paucity limits the development of evidence-based management guidelines; the 2022 Society for Cardiovascular Magnetic Resonance/American Society of Echocardiography consensus notes that published cardiovascular magnetic resonance experience consists largely of "case reports and relatively small case series" (90). Consequently, diagnostic algorithms rely on expert consensus rather than prospective validation, surgical strategies are

extrapolated from benign tumor experience, and outcomes—particularly for rare malignant variants—remain poorly characterized (86, 91). Tumor heterogeneity (rhabdomyoma, fibroma, myxoma, teratoma, hemangioma, and various sarcomas) further fragments the available evidence, complicating efforts to establish histotype-specific prognostic models or standardized treatment protocols.

Conclusions

In summary, cardiac tumors are rare and can potentially lead to serious complications such as fatal arrhythmias and intracardiac obstruction, even when benign. Imaging techniques such as MRI and CT scans play a pivotal role in diagnosis, helping distinguish between tumor types and ultimately shortening the time from diagnosis to treatment. These neoplasms can be treated surgically with an acceptable incidence of morbidity and mortality. The strongest predictor of mortality is linked to patient comorbidities, hemodynamic status at presentation, and the percentage of myocardial tissue involvement. Evaluation of patients with pediatric cardiac tumors requires multidisciplinary evaluation and individualized management. Overall, benign tumors possess high survival rates, and patients should be monitored for recurrence following resection.

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