

# SURGICAL MANAGEMENT AND OUTCOME IN PRIMARY CARDIAC TUMORS

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## REZUMAT

**Introducere:** Tumorile cardiace primitive sunt entități foarte rare. Majoritatea sunt tumori benigne, iar dintre acestea mixoamele reprezintă grupul cel mai frecvent. **Material și metode:** S-au studiat retrospectiv dosarele a 3325 pacienți consecutivi operați în perioada 1998-2004 în Institutul de Boli Cardiovasculare Timișoara, găsindu-se 21 de cazuri de tumori cardiace primitive. Majoritatea au fost tumori benigne (71,5%) la pacienți de sex feminin (76,1%). Tratamentul a fost chirurgical indiferent de etiologie, constând în rezecția tumorii, cu concursul circulației extracorporeale și refacerea structurilor afectate. **Rezultate:** În cazul celor 15 tumori benigne operate, rezultatele au fost excelente, fără mortalitate precoce și pe termen lung și fără recidivă locală. Pacienții cu tumori maligne au decedat toți, fie în cursul spitalizării, fie la distanță de intervenția chirurgicală. **Concluzii:** Tratamentul chirurgical este metoda de elecție pentru tratamentul tumorilor cardiace indiferent de etiologie. Rezultatele sunt foarte bune în cazul tumorilor benigne care, neoperate, expun pacientii la complicații și deces prin emboli sau obstrucții mecanice. În cazul tumorilor maligne rezultatele sunt sumbre, chiar și transplantul cardiac neputând prelungi pe termen lung viața. Cu toate acestea, efectul paliativ, de rezecție, asociată cu terapie adjuvantă, poate avea efecte benefice pe termen scurt și mediu.

## ABSTRACT

**Introduction:** Primary cardiac tumors are very rare. The majority of these tumors are benign and myxomas are the most frequent. **Material and methods:** We reviewed the records of 3325 consecutive open-heart procedures performed in the Institute of Cardiovascular Medicine, Timisoara, between 1998 and 2004 and found 21 primary cardiac tumors. Most were benign tumors (71.5%) in female patients (76.1%). The treatment of choice was surgical removal, regardless the etiology, under cardiopulmonary bypass and reconstruction of the structures involved. **Results:** For the benign tumors, the early and late results were excellent, without any mortality or recurrence. However, all patients with cardiac neoplasms died, either during hospitalization, or in the following months. **Conclusion:** Surgical intervention is the management of choice regardless of the origin for primary cardiac tumors. Benign tumors carry the risk of embolic complications or mechanical obstruction; however the surgical outcome is excellent. Cardiac neoplasms are associated with high mortality, either early or late after the diagnosis.

## INTRODUCTION

Primary cardiac tumors are very rare entities with an incidence between 0.0017-0.19% in general unselected necropsy series. Primary cardiac tumors are uncommon and represent only 5-10% of this pathology.<sup>1,2</sup> The majority of the tumors are benign with myxomas being the most frequent.<sup>3</sup> We reviewed our medical records for patients diagnosed with primary cardiac tumors.

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## PATIENTS AND METHODS

Between 1998 and 2004, 3325 consecutive patients underwent open-heart procedures in the Institute of Cardiovascular Medicine, Timisoara. Twenty-one patients (0.63%) had primary cardiac tumors. The majority were benign tumors (15, 71.5%) and most were found in female patients (76.1%). The age range was between 3 months and 75 years.

The diagnosis was made late in the course of the disease because of unspecific clinical symptoms, however it was established beyond any doubt by transthoracic and transesophageal echocardiography. Whenever the suspicion of a malign tumor arises, a full-body CT-scan or MRI is recommended for detecting the metastatic extension. We systematically performed angiocoronarography for all patients over 45 years of age irrespective of clinical symptoms for detecting occult coronary involvement.

The treatment of choice was surgical removal, regardless the etiology, under cardiopulmonary bypass and reconstruction of the structures involved (walls, valves). The local extension of the tumor was established intraoperatively. All the resected specimens were referred at the same histopathology laboratory for the exact typing of tumors. (Table 1)

**Table 1.** Tumor characteristics and location in heart.

Tumor	No	LA	RA	LV	RV	MV	TV	Great Vessel
<b>BENIGN</b>	15							
Myxomas	12	12	-	-	-	-	-	-
Fibroelastoma	1	-	-	-	-	1	-	-
Rhabdomyoma	1	-	1	-	-	-	-	-
Lipomas	1	-	-	-	-	-	-	-
<b>MALIGN</b>	6							
Angiosarcoma	1	-	1	-	-	-	-	-
Rhabdomyosarcoma	2	-	y	-	yes	-	yes	-
Lymphoma	1	-	es	-	yes	-	-	yes
Fibrosarcoma	1	ye	-	-	-	yes	-	-
Mesothelioma	1	s	-	-	yes	-	-	yes
		-	-					

LA = left atrium, RA = right atrium, LV = left ventricle, RV = right ventricle, MV = mitral valve, TV = tricuspid valve

For the left atrial tumors, suspected to be myxomas on the basis of preoperative exams, the approach was through the interatrial groove but with minimal manipulation before the aortic crossclamp was applied, in order to avoid systemic embolism. In one case a special aortic cannula with Embolex filter was used.

Whenever the tumor appearance was suggestive of neoplastic disease, complete resection was attempted. There were no intraoperative deaths. (Table 2)

## RESULTS

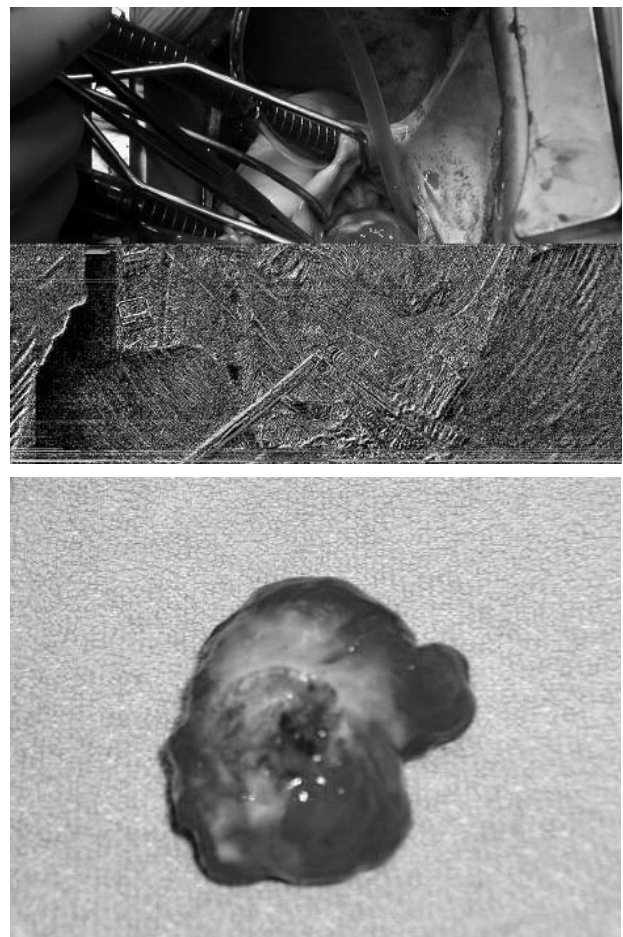
There were no intraoperative deaths, but two patients with malignant tumors died during hospitalization. The surgical result was excellent for benign tumors, without early or late mortality and without recurrence at the latest follow-up.

Benign tumors occurred more often, most in the form of atrial myxomas, and are more prevalent in women. Figure 1 shows a typical case of left atrial myxoma in a young female patient. (Fig. 1) If the tumor appears in elderly patients, it can be associated with coronary artery disease, as was the case in one of our patients. (Fig. 2) The treatment involves tumor resection and revascularization of the affected myocardium.

**Table 2.** Surgical procedures performed for primary cardiac tumors.

Tumor	No.	Procedure Performed
<b>BENIGN</b>	15	
	11	Simple Resection
	1	Resection + Fosa Ovalis - suture
	1	Resection + Ostium Secundum closure +TV DeVega plasty
	1	Resection + CABG
	1	Resection AV Junction + AVR + MVR
<b>MALIGN</b>	6	
	1	RV + RA Resection - Patch Reconst. + TVR Sorin Pericarbon
	1	LA Resection + MV Repair; Redo 2 mo later - exitus 1 year later
	1	RA resection - Bovine pericard repair - exitus 6 mo later
	1	RA+ TV+ RV Resection, TVR- Omnicarbon - exitus 3 mo later
	1	Exitus during angiography
	1	CABG x 1- off pump - exitus some hours later

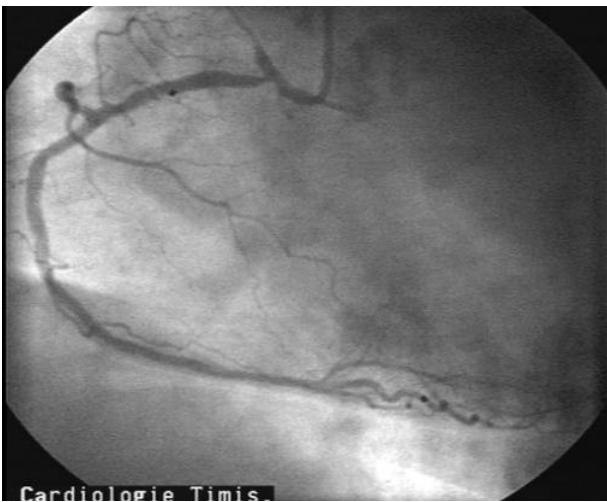
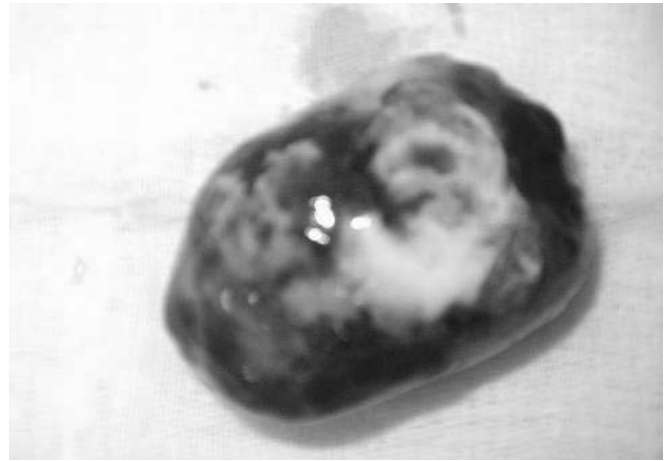
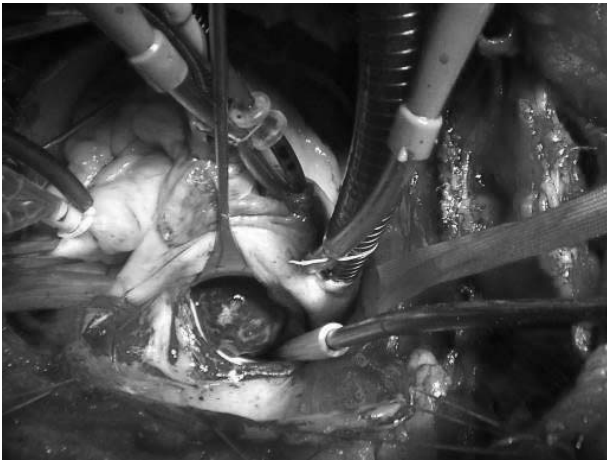
LA = left atrium, RA = right atrium, LV= left ventricle, RV= right ventricle, MV = mitral valve, TV = tricuspid valve, CABG= Coronary artery bypass grafting



**Figure 1.** A 47 years-old female with myxoma of LA diagnosed very late, after two years of diffuse complains.

Malignant tumors are very rare. Our patients with this pathology presented with unspecific complaints and most tumors were intraoperative discoveries.

Our first patient presented with a pericardial effusion and the tumor was discovered incidentally while draining the fluid. A biopsy was taken and the

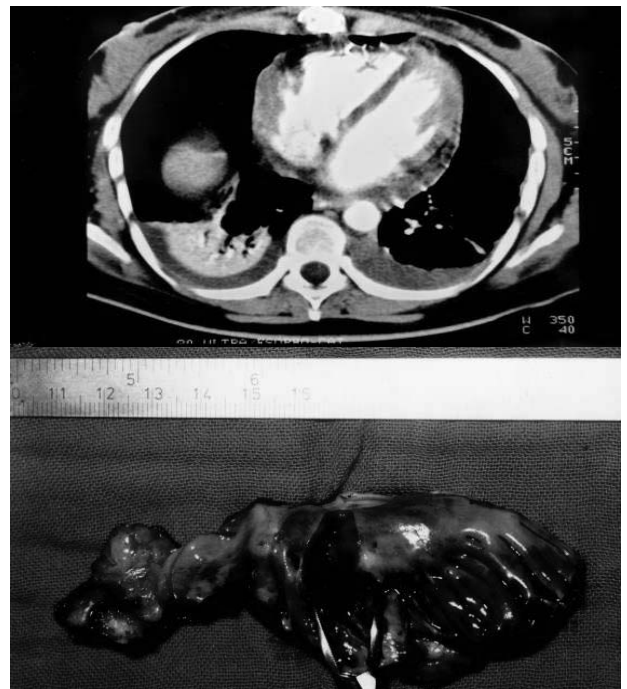


**Figure 2.** Severe ostial stenosis of RCA on LAO incidence and normal left coronary arteries. The myxoma was excised and a venous graft performed on the RCA (a 56 year-old female with left atrial myxoma and coronary artery disease, 2004, Timisoara).

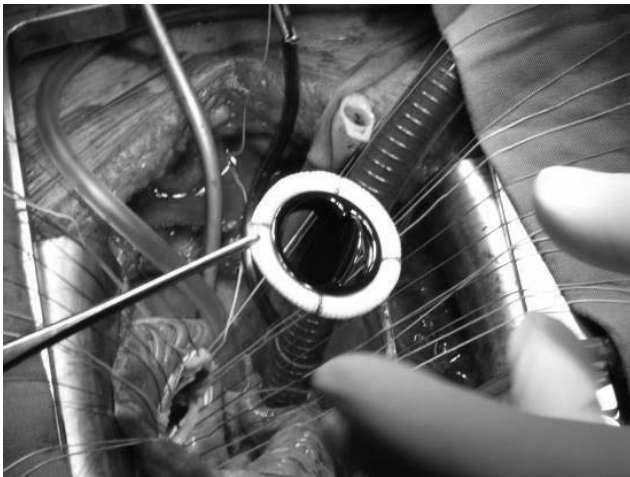
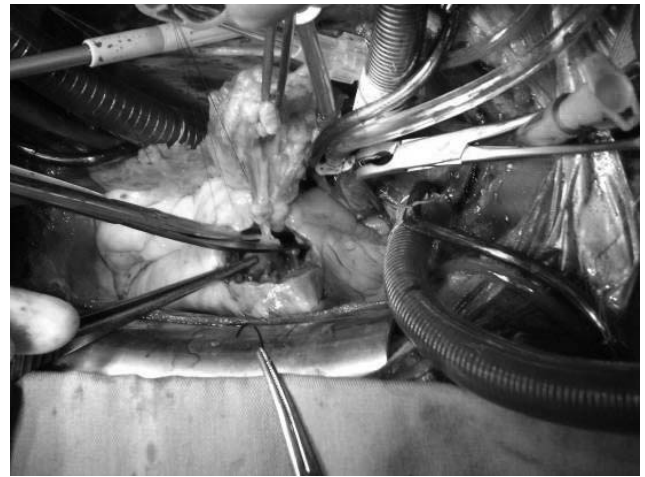
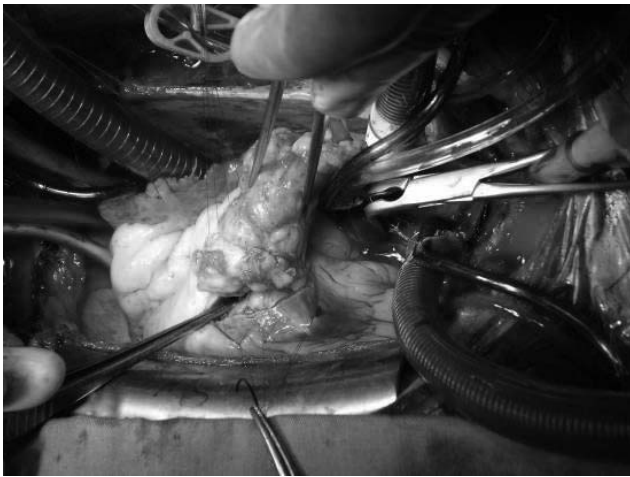
diagnosis of right atrial angiosarcoma established. She was subsequently operated upon and the tumor was fully resected, reconstructing the right atrium with a bovine pericardial patch, however she died of lung metastases six months later. (Fig. 3)

Our second patient presented with clinical signs and symptoms of a pulmonary embolism. A rhabdomyosarcoma invading the right atrium, tricuspid valve and parts of the right ventricle was discovered. Total resection with tricuspid valve replacement by a mechanical prosthesis and reconstruction of the atrial and ventricular walls by synthetic patches was undertaken. The primary evolution was very good, the patient being discharged, however she expired 6 months later due to local relapse. (Fig. 4)

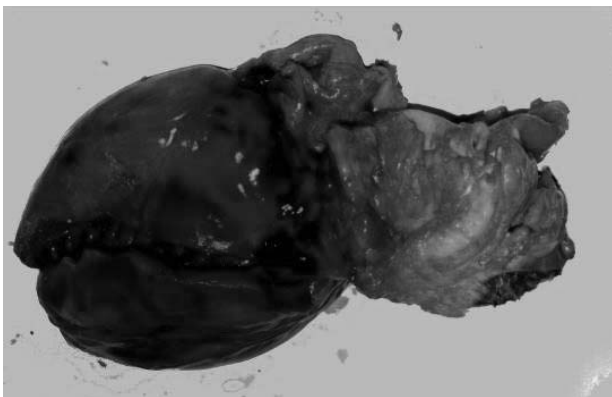
The third patient was a 30-year old male who presented with typical chest pain. An angiographic exam was performed during which he presented sudden cardiac arrest with electro-mechanical dissociation during the exam. At autopsy we found a massive mediastinal lymphoma, invading and occluding the aorta and right ventricular outflow tract. (Fig. 5)



**Figure 3.** RA angiosarcoma operated by resection and bovine pericardium reconstruction, died six months after operation by lung metastasis (Prof. Ernst Wolner - Assoc. Prof. Marian Gaspar, Vienna).



**Figure 4.** Right atrium, tricuspid valve and right ventricle obstruction by a massive rhabdomyosarcoma. Tricuspid valve replacement. (Prof. Gunther Laufer, Assoc Prof. Marian Gaspar, Timisoara, 2004)



**Figure 5.** Great vessel invasion by a lymphoma in a young 30-year old male patient. He died during the cardiac catheter exam.



**Figure 6.** The right ventricle tumor in a 60 years old, female scheduled for a CABG.

Our fourth patient was a 60-year-old female patient with coronary artery disease. Upon surgery we found a tumor invading the pulmonary artery and right ventricle. Off-pump left internal mammary artery to left anterior descending bypass was performed, however she presented severe low-cardiac output and died hours after the procedure. Tumor biopsy revealed a cardiac mesothelioma. (Fig. 6)

There were two redo procedures for a local relapse. One patient expired on the first postoperative day while the second survived another year. Chemotherapy was delivered in our first two patients, but without an effective result, both died six months later from lung metastases or local relapse, respectively.

## **DISCUSSION**

Primary cardiac tumors are rare. With the widespread use of modern diagnostic techniques, such as transthoracic and transesophageal cardiac ultrasonography, contrast enhanced computed tomography and magnetic resonance imaging, the relative frequency seems to increase due to better screening. The exact pathological type can be determined preoperatively only for right-sided tumors by using a transvenous biopsy device, however, this rarely changes the therapeutic attitude.<sup>4,6</sup>

The exact incidence of this pathology is largely unknown. Approximately 70 -80% of these are benign, the majority of which are represented by myxomas. These are diagnosed by transthoracic ultrasonography on the basis of unspecific clinical signs (dyspnoea, fatigue).<sup>2</sup> Angiogramography should be performed in patients at risk for coronary artery disease or who are symptomatic at rest or during stress testing.<sup>7</sup>

Surgical resection is the treatment of choice for all cardiac tumors, and is indicated even for benign tumors for the risk of systemic embolization and valve obstruction. The approach depends on the location of the tumor and its local extension.<sup>8,9</sup>

Myxomas are the most frequent primary cardiac tumors, and are located within the left atrium in the vast majority of patients. They are pedicled, inserting on the interatrial septum, and their resection should include their base, as there is a risk of recurrence.

Angiosarcomas and rhabdomyosarcomas are the most frequent malignant primary cardiac tumors. If the valve tumor is malignant, complete resection of the tumor and replacement of the involved valve is the best choice.<sup>4,5</sup> Surgical treatment is not curative and incomplete for these tumor types, as the local extension often precludes a reasonable safe resection. Nevertheless, in some cases palliation of symptoms and extension of life may be achieved by aggressive adjuvant therapy. The major role of surgery in such cases is to establish a diagnosis in order to exclude the possibility of a curable benign tumor.<sup>9</sup>

Clinical manifestations are common to all tumor types, and include shortness of breath, edema, hepatomegaly, ascites, syncope, from obstruction of flow or interference with valvular function, cardiac dysrhythmias and sudden death.

Survival up to three years has been reported following: partial resection, associated with chemo- and radiotherapy. Orthotopic cardiac transplantation

not indicated in the current era of donor shortage for unresectable malignant tumors, as they tend to progress very quickly.<sup>10</sup>

Some success in palliation of symptoms has been reported following chemotherapy, but not in our two cases. Lymphosarcoma of the heart frequently responds to chemotherapy, radiation therapy, or both. Unfortunately, many other reports indicate a failure to alter the course of cardiac sarcomas despite various combinations of surgery, chemotherapy and radiation therapy.<sup>4,6,8,10</sup>

## **CONCLUSION**

Surgical intervention is the management of choice regardless of the origin of all primary cardiac tumors. The results for benign tumors are excellent, with survival similar to that in the general population and a low risk of recurrence. Malignant tumors have a dim outlook, in spite of the advances in chemo- and radiotherapy, as these are frequently aggressive, with extensive local invasion and distant metastases.

In the absence of randomized clinical trials it is not clear whether adjuvant chemotherapy may be beneficial in patients in whom “curative” surgery has been performed. The role of radiation therapy is less well known. However, effective palliation with resection and adjuvant therapy may prolong life.

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