REVIEW ARTICLE

Current challenges in the diagnosis and treatment of cardiac myxoma

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KEY WORDS
cardiac myxoma, diagnosis, outcomes, symptoms, treatment

ABSTRACT
Cardiac myxoma is the most common benign cardiac tumor. It is located in the left atrium and typically arises from the foramen ovale in approximately 75% of the general patient population, in the right atrium in 23%, and in the ventricles in only 2%. Symptoms depend on its size, mobility, and relation to surrounding cardiac structures. Neurological complications resulting from cardiac myxoma are seen in 20% to 25% of patients. Molecular genetic studies show that the condition can be inherited in Carney complex due to mutations of the *PRKAR1A* gene. Cardiac myxoma resection is a cardiac surgery with a low complication rate and the 30-day mortality of up to 10%. Recurrence may be observed months or years after surgery, and its rate is approximately 5%. Long-term follow-up with transthoracic echocardiography is needed in all patients after tumor resection. This review summarizes the available data on cardiac myxoma and, in particular, issues relating to diagnosis and treatment.

Introduction Cardiac myxoma (CM) is the most common benign cardiac tumor. As the number of patients with CM is increasing, transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are widely used in routine cardiology practice to diagnose and monitor this cardiac disease. Of note, patients with permanent or temporary neurological disorders are at high risk of developing CM. A diagnostic approach to patients with CM is based on medical history, clinical examination, blood test results, and radiographic or echocardiographic findings. Computed tomography (CT) of the chest and magnetic resonance imaging (MRI) of the chest or the heart help with the differential diagnosis, which includes cardiac tumors and masses. Compared with classic coronary angiography, computed tomography coronary angiography reduces the complication risk during the preoperative assessment of young patients. Surgical excision remains the treatment of choice in patients with CM. Recurrence of CM after surgical excision is usually observed in individuals with familial and complex forms of the condition. A genetic analysis of patients with CM gives a new opportunity to identify occult CM in asymptomatic patients and in patients with a family history of CM.

Methods This review is based on research of the current literature regarding the epidemiology of CM, its clinical presentation, diagnosis, and treatment. The PubMed database was searched for eligible studies and the search was restricted to the years 2000 to 2019. The search term was "cardiac myxoma" and we included observational or retrospective studies with large samples of patients who were treated for CM only. In addition, the data of interest to the present review were long-term follow-up, the recurrence rate during follow-up, and the survival rate after CM resection. Additionally, we looked for studies on novel surgical techniques for CM resection and those describing unusual CM location or clinical presentation. The primary source for data extraction were 24 articles from 15 countries, which reported on a total of 2205 patients with CM (TABLE 1).[1-24] We included studies from Europe, North and South America,

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Middle East, and Asia. The median (minimum–maximum) study period was 20.6 (6–57) years, and the median (minimum–maximum) number of studied patients was 68 (23–403) (TABLE 1).[13,21]

Epidemiology Cardiac myxoma is a rare benign cardiac tumor, and its incidence is observed in approximately 0.5–1 cases per 1 million people per year.[1,4,11,24] The condition may develop in all ages, more frequently in women than in men. The demographic characteristics of patients included in this review are listed in TABLE 2. Recent large studies of patients with CM showed a higher prevalence of the disease in women (range, 53%–77.4% of patients).[2,16] Cardiac myxoma was observed in all age groups, and the mean range of age was 42 to 66 years.[17-18] It was rarely reported in younger patients compared with those older, and usually as a familial form of CM. Most commonly, CM was located in the left atrium, whereas unusual locations were detected in the right atrium in 0.7% to 7.5% of patients, in the right ventricle in 0.7% to 2.5% of patients, in the left ventricle in 0.7% to 3.6% of patients, and in a heart valve (aortic, mitral) in fewer than 1% of patients.[1,4,9,11,13,19,20,24] Most studies reported that CM was identified in the left atrium in about 72% to 92% patients.[2,5,15] The fossa ovalis in the interatrial septum was the most common site to which CM, with or without a broad base, was attached. Pedunculated or large CM may prolapse into a heart valve and this may occur regardless of tumor size. Unfortunately, the symptoms of CM (if present) usually appear when the tumor grows, and the diagnosis is established based on clinical manifestations. Numerous studies with large patient samples reported that the mean range of CM size was 2.7 to 5.8 cm.[7,16] Giant CM may occupy cardiac chambers and be difficult to remove surgically (FIGURE 1). In the past, when echocardiography was not widely used in everyday clinical practice, CM was mostly revealed after patient's sudden death or on autopsy. Nowadays, the use of TTE and TEE increases the rate of patients diagnosed with CM.

TABLE 1 Studies reporting on patients who underwent cardiac myxoma resection

Study	Country	Years	Study period, y	Patients, n (total n = 2205)	Operated patients per year, n
Pinede et al[1]	France	1959–1998	40	122	3.05
Tasoglou et al[2]	Turkey	1990–2006	17	67	3.9
Patil et al[3]	India	2000–2009	10	62	6.2
Wu et al[4]	China	1988–2010	23	112	4.9
Lee et al[5]	South Korea	2000–2011	12	59	4.9
Vaideeswar et al[6]	India	1995–2009	15	84	5.6
Garrati et al[7]	Italy	1990–2007	18	98	5.4
Bordalo et al[8]	Portugal	2003–2010	8	40	5
Obrenović-Kirćanski et al[9]	Serbia	1981–2010	30	74	2.5
Vroomen et al[10]	Netherlands	1990–2012	21	82	3.9
He et al[11]	China	1998–2014	17	162	9.5
Aval et al[12]	Iran	1994–2014	21	42	2
Shah et al[13]	United States	1955–2011	57	194	3.4
Lin et al[14]	China	1996–2012	17	68	4
Anvari et al[15]	Iran	2004–2014	11	73	6.6
Yóksel et al[16]	Turkey	1990–2014	25	43	1.7
Bainchi et al[17]	Italy	2006–2017	12	30	2.5
Abu Abeeleh et al[18]	Jordan	1984–2016	36	27	0.75
Lee et al[19]	South Korea	1986–2015	30	93	3.1
Karabinis et al[20]	Greece	1993–2017	25	153	6.1
Nehaj et al[21]	Slovakia	2011–2016	6	41	6.8
Gür et al[22]	Turkey	2010–2017	8	23	2.9
Cianciulli et al[23]	Argentina	1993–2013	20	53	2.7
Jiang et al[24]	China	2002–2016	15	403	26.9

TABLE 2 Demographic characteristics of patients with cardiac myxoma

Study	Patients, n	Age, y	Female sex, %	CM of the left atrium, %	Tumor size, cm
Pinede et al[1]	122	Median, 54	64	100	Range, 1–15
Tasoglou et al[2]	67	Median, 46.3	77.4	72	Median, 5.7
Patil et al[3]	62	Median, 38	63	75	–
Wu et al[4]	112	Mean, 48.8	58.9	85.8	–
Lee et al[5]	59	Mean, 57.5	64	93.2	Mean, 4.8
Vaideeswar et al[6]	84	Mean, 40.8	76.2	–	Mean, 5.2
Garrati et al[7]	98	Mean, 60	56	86	Mean, 2.7
Bordalo et al[8]	40	Mean, 64.2	65	92.5	Mean, 4.6
Obrenović-Kirćanski et al[9]	74	Mean, 46.7	68	89.1	Median, 5.7
Vroomen et al[10]	82	Mean, 61.3	58.5	92	Mean, 5
He et al[11]	162	Mean, 54	65	84.6	–
Aval et al[12]	42	Mean, 50.6	59.2	85.6	Mean, 5.2
Shah et al[13]	194	Mean,57.2	62	80	Mean, 4.3
Lin et al[14]	68	Mean, 50.9	72	88	Median, 5.3
Anvari et al[15]	73	Mean, 54.5	56.2	93.2	Mean, 5.3
Yóksel et al[16]	43	Mean, 51.7	53	86	Mean, 5.8
Bainchi et al[17]	30	Mean, 66	67	96.7	Mean, 4.3
Abu Abeeleh et al[18]	27	Mean, 42	48	77.7	Range, 1–5
Lee et al[19]	93	Mean, 54.7	67.7	92.5	Mean, 4.7
Karabinis et al[20]	153	Mean, 59	68	82.4	Mean, 4.5
Nehaj et al[21]	41	Mean, 61.7	60.9	90	Mean, 3.27
Gür et al[22]	23	Mean, 42.1	72.3	82.6	Mean, 3.5
Cianciulli et al[23]	53	Mean, 53	62.3	77.4	Mean, 4.7
Jiang et al[24]	403	Mean, 54.7	68.2	92.8	Mean, 4.0

Abbreviations: CM, cardiac myxoma

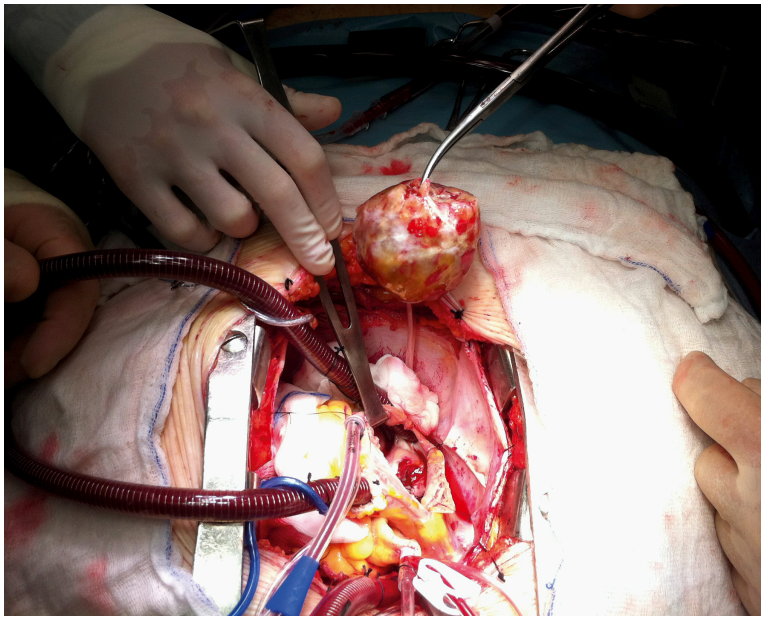

FIGURE 1 Intraoperative view of a giant cardiac myxoma

Clinical presentation Clinical presentation and signs observed in patients with CM depend on the location and mobility of the tumor. The clinical manifestations of CM are divided into 3 groups: hemodynamic consequences, systemic or pulmonary embolization, and systemic or constitutional manifestations.[1,4,7,10,20] However, approximately 3.2% to 46.4% of patients with CM are asymptomatic.[3,5,11,19,20] The clinical manifestations of patients included in this review are shown in TABLE 3. Usually, the symptoms become present when the tumor prolapses into the heart valve (aortic, mitral, or tricuspid) and obstructs a heart valve orifice. Temporary and permanent neurological disorders may occur due to embolization of the cerebral artery caused by the fragments of CM or as a consequence of heart valve obstruction. Several authors reported on asymptomatic patients with CM, in whom tumors were detected by TTE during a routine checkup. The hemodynamic consequences of CM include dyspnea, palpitations, atrial fibrillation, episodes

TABLE 3 Clinical manifestations in patients with cardiac myxoma

Study	Patients, n	Dyspnea, %	Systemic embolization, %	Systemic or constitutional manifestations, %
Pinede et al[1]	122	67	29	34
Tasoglou et al[2]	67	76.1	17.9	17
Patil et al[3]	62	62.9	10	23
Wu et al[4]	112	50	13	27
Lee et al[5]	59	62.7	22	16.9
Vaideeswar et al[6]	84	84.4	15.6	4.7
Garrati et al[7]	98	68	40	22
Bordalo et al[8]	40	35	15	23
Obrenović-Kirćanski et al[9]	74	79.3	20.2	8
Vroomen et al[10]	82	70	15.9	–
He et al[11]	162	48.8	–	–
Aval et al[12]	42	88	7.1	21.4
Shah et al[13]	194	–	–	–
Lin et al[14]	68	57	25	33
Anvari et al[15]	73	40	21.9	–
Yóksel et al[16]	43	51.4	21.6	32.4
Bainchi et al[17]	30	–	6.7	–
Abu Abeeleh et al[18]	27	30	33	37
Lee et al[19]	93	58	10.8	2.2
Karabinis et al[20]	153	47.7	4.6	0.7
Nehaj et al[21]	41	50	20	4.9
Gür et al[22]	23	53	–	43.4
Cianciulli et al[23]	53	56	24.5	26.4
Jiang et al[24]	403	13.3	14.9	–

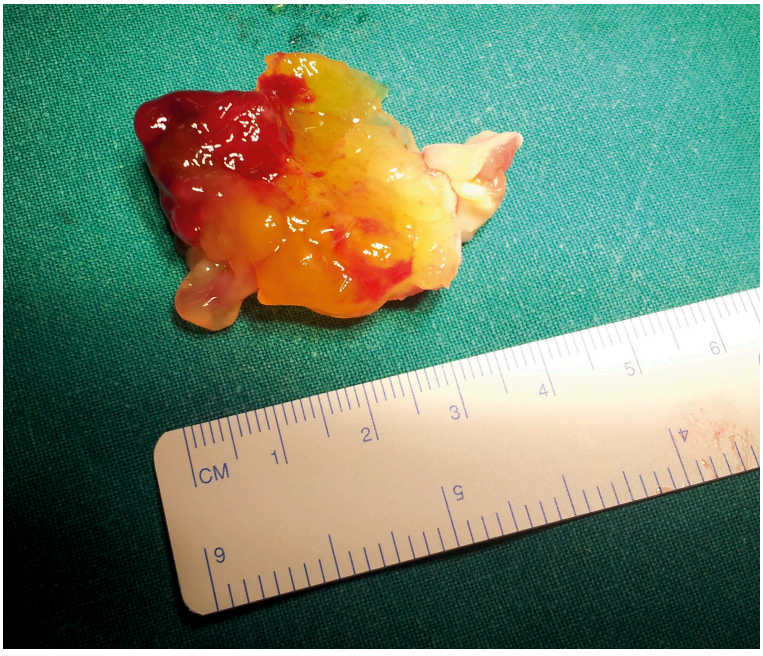

FIGURE 2 Resected cardiac myxoma

of syncope, tachycardia, and sudden death. Nonspecific clinical symptoms due to hemodynamic disturbances may be confusing for clinicians establishing a differential diagnosis of cardiac tumors and structural or ischemic cardiac disease. Cardiac myxoma should be suspected particularly in patients admitted to an emergency department. Changes in patient position may cause occasional loss of consciousness or syncope due to obstruction of the mitral or tricuspid valve. Large mobile CM in the left or right ventricle may lead to sudden death, particularly when CM prolapsed through the mitral valve into the left ventricle. Systemic or pulmonary embolization may be the first symptom of CM. Upper and lower extremity ischemia, pulmonary embolism, mesenteric ischemia with acute abdomen, and acute coronary syndrome are the most common clinical syndromes, which occur in patients with CM due to embolization.[11] He et al[11] analyzed 162 patients and confirmed that friability of CM may cause systemic embolization.[11] The uneven surface structure and gelatinous consistency of CM

predispose to peripheral embolization (FIGURE 2). Although systemic and pulmonary embolization is a rare complication of CM, acute coronary syndrome may lead to a life-threatening emergency. In addition, a permanent neurological disorder due to systemic embolization caused by CM may affect patients' quality of life. Constitutional manifestations (anemia, fever, weight loss, fatigue, arthralgia, myalgia, and Raynaud phenomenon) are rare findings reported in approximately 16.9% to 32.4% of patients with CM.[3-5,7,16] These symptoms are nonspecific and coexist with possible proinflammatory and chronic inflammatory reactions. Interleukin-6 is a commonly known inflammatory mediator found in patients with CM.[25,26] A few studies showed decreased interleukin-6 levels in peripheral blood of patients who underwent CM resection.[27,28] On the other hand, the tumor is also said to produce inflammatory mediators itself.[25] To confirm this hypothesis, we need further studies on interleukin-6 levels, which would examine larger patient samples during a long-term follow-up after CM resection.

Diagnosis Cardiac myxoma is diagnosed based on clinical examination, electrocardiography, TTE, TEE, chest CT, and chest or cardiac MRI. There is no evidence suggesting a specific blood test useful in the diagnosis of CM. A genetic analysis may provide valuable information in patients with the familial and Carney complex forms of CM.

The clinical examination of a patient includes heart auscultation for possible systolic or diastolic heart murmur, which can be heard if a myxoma has prolapsed or obstructed a heart valve orifice. Heart murmur is not a specific finding indicative of CM. In addition, concomitant structural heart diseases (affecting the mitral, tricuspid, or aortic valve) may be confused with CM. There are no criteria for the diagnosis of CM by electrocardiography, because heart rhythm disturbances, such as atrial fibrillation and sinus tachycardia, are commonly found in other cardiac diseases.[9,14,21] However, TTE and TEE show a 90% to 96% accuracy in diagnosing CM. Transthoracic echocardiography is the simplest and most useful examination in patients with cardiac diseases, particularly in those with cardiac tumors. It is used in all hospitals, including emergency departments. In routine clinical practice, it is a quick and safe tool, which provides detailed data regarding the structure and location of a cardiac tumor. If TTE does not provide additional information on the nature, structure, and mobility of CM, TEE may accurately assess tumor characteristics. It shows a sensitivity for the diagnosis of CM, which is higher than 90%. The most common characteristic of CM found on TEE is a mass attached to the interatrial septum, with or without a broad base.[13,19,24] Occasionally, CM may affect heart valves (mitral and tricuspid) and physicians should be aware of this rare pathology. The mobility of the tumor and its prolapse into the mitral or tricuspid valve during diastole are specific characteristics of CM (FIGURE 3). A pedunculated tumor attached to the interatrial septum is the most frequent characteristic of atrial CM (FIGURE 4). A smooth tumor, which does not affect the adjacent structures, is also indicative of CM (FIGURE 5).

Before surgery, a differential diagnosis should be established to exclude other benign or malignant (primary or metastatic) cardiac tumors, thrombi, and vegetations. Chest CT or MRI may help to detect any other coexisting conditions within the thorax (lung tumor, pulmonary embolism, esophageal pathology, and lymphoma). In patients with CM, brain and abdominal CT should be performed for cardiac tumor staging. If malignancy is suspected, further diagnostic workup should be considered. Positron emission tomography may confirm malignancy or benign

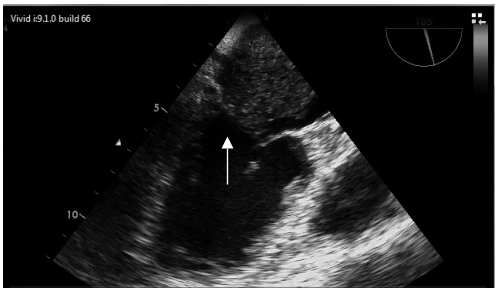

FIGURE 3 Transesophageal echocardiography showing cardiac myxoma prolapse into the mitral valve (arrow)

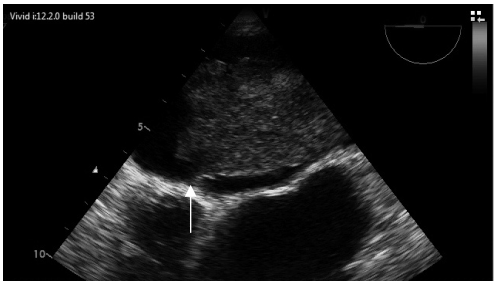

FIGURE 4 Cardiac myxoma (arrow) attached to the interatrial septum seen on transesophageal echocardiography

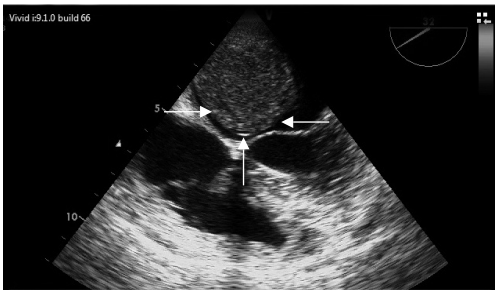

FIGURE 5 Cardiac myxoma (arrows) in the heart chamber seen on transesophageal echocardiography

features of a cardiac tumor with a 90% sensitivity.[29] Coronary angiography is performed in all patients with cardiac tumors, including CM. Computed tomography coronary angiography may be preferred in patients below 40 years of age, also in those at cardiovascular risk, whereas patients older than 40 years, including those at cardiovascular risk, should underwent conventional coronary angiography.[7,11,20]

A genetic analysis of patients with familial CM and Carney complex with concomitant CM may detect occult CM. Familial CM is an unusual form of CM, occurring in 5% of patients with CM. Multiple myxomas and recurrence in patients with familial CM were associated with a mutation of the *PRKAR1A* gene. A recent study suggested that the *PRKAR1A* gene was related to Carney complex, which is a condition manifested by skin pigmentation and multiple neoplasia affecting the heart and both nonendocrine and endocrine glands.[30] A mutation of the *PRKAR1A* gene was observed in patients with familial CM in Carney complex more frequently than in those with its sporadic form.[31] This observation may further facilitate the diagnosis of patients with occult and familial CM and, therefore, reduce the morbidity, mortality, and postsurgical recurrence rates in this population.

Treatment The first CM resection was described by the Swedish cardiac surgeon Clarence Crafoord and his team in 1954.[32] Since 1954, a lot of patients have been operated on for this rare cardiac tumor worldwide. The number of patients diagnosed with CM has increased due to the wide use of TTE and TEE. The radical resection of CM with surrounding structures invaded by the tumor remains the treatment of choice, which helps to avoid possible recurrence during long-term follow-up.

Surgical excision should be performed as soon as CM is diagnosed and without delay, as the behavior of the tumor cannot be predicted and it does not depend on how large and fragile it is. Although evidence for favorable outcomes of an emergency or urgent operation compared with the elective one has not been reported so far, the preoperative patient status may affect the time when an operation is conducted. Preoperative stroke, pulmonary embolism, and peripheral ischemia are additional risk factors for cardiac surgery, which may delay the procedure. Of note, heart valve obstruction caused by CM may lead to catastrophic consequences, including sudden death. A fragment of the tumor can embolize the left or right main coronary artery ostium and result in acute coronary syndrome. For these reasons, the preoperative assessment of patients with CM should be performed, and a systematic approach to these patients is the key to successful postoperative outcomes. Identifying structural heart disease and other comorbidities in patients with CM is a crucial point of the preoperative patient examination. Conventional coronary angiography or CT coronary angiography (in patients below 40 years of age) is recommended in all patients before surgery.[12,21] With its high accuracy, TEE may help to evaluate a heart valve affected by CM or determine the exact location of CM (in the interatrial septum, mitral, aortic, or tricuspid valve, left or right ventricular wall, or interventricular septum). The presence of heart valve disease and coronary artery disease affects surgical treatment and impacts postoperative outcomes. Cardiac myxoma is a benign cardiac tumor but the preoperative evaluation of patient's brain, chest, and abdomen using CT can contribute to detailed examination of other systems and target organs. For example, occult cerebral damage on preoperative CT may explain neurological disorders observed after surgery. Chest CT can detect calcification of the thoracic aorta, which is a prognostic factor for postoperative stroke. The risk of embolization of abdominal organs (spleen, kidneys, and abdominal aorta) should be considered before surgery. Infected thrombi resulting from CM may cause abscess formation in the target organs (brain, spleen, liver, and kidneys). Preoperative blood culture and antibiotic therapy (depending on the microorganism detected) may be needed before surgery to prevent infective endocarditis.

Diverse techniques have been proposed for CM resection with good short- and long-term results. During surgery, cardiopulmonary bypass and aortic cross clamps are established in all patients. In addition, combined operations such as coronary artery bypass grafting, heart valve surgery, ascending aorta surgery, and other procedures are performed due to patients' advanced age. Such operations account for 5% to 25% of all procedures conducted in patients with CM. Median sternotomy with distal ascending aortic and bicaval cannulation is the most common approach used during CM resection. Minimizing manipulation of the heart before the aortic cross clamp is placed as well as that of the tumor prevents systemic and pulmonary embolization. In all cases, CM should be removed first, prior to any other cardiac surgery. Initially, retrograde catheter placement should be avoided. Antegrade cardioplegia may be considered a preferable technique to achieve heart arrest during operation. If CM is located in the interatrial septum or left atrium, the biatrial approach (vertical incision) is the safest and most suitable technique for CM resection. Inspecting 4 chambers of the heart (left and right atria, left and right ventricles) is an advantage of this approach. Manipulation through the right atrium is enough to complete the resection of CM located in this chamber. A rare location of CM in the right or left ventricle or in the interventricular septum can be

accessed through the tricuspid or aortic valve or incision in the anterior wall of the right ventricle. Heart defects within the interatrial septum, left or right atrial wall, or interventricular septum, which are created after CM resection, should be repaired with autologous or bovine pericardium or a synthetic patch. All repairs should be evaluated by intraoperative TEE in the operating room and before weaning the patient from bypass. If a heart valve is repaired or replaced, TEE should be performed to assess the function of a prosthetic valve and detect possible paravalvular leak.

Minimally invasive or robotic CM resection was reported in small patient samples in the last years.[17,33-35] The scope of minimally invasive approach is the same as that of conventional open surgery, namely, the radical resection of CM with the affected surrounding structures. A double-lumen endotracheal tube is used for patient intubation. The heart is accessed through a small incision (approximately 3 to 5 cm) in the right lateral thoracic wall and the procedure is monitored by a camera. Cardiopulmonary bypass is established with cannulation of the common femoral artery and the common femoral vein. The operators use special minimally invasive instruments for cardiac surgery. Unfortunately, the existing data are not exhaustive enough to confirm the benefits of this technique. Another problem is that more and more patients with CM underwent combined cardiac surgery. For these reasons, a limited number of patients with isolated CM underwent minimally invasive resection.

Cardiac myxoma resection is associated with a low rate of postoperative complications, and the postoperative period is usually uneventful, with minor complications reported in most cases. Rhythm disturbances, mainly atrial fibrillation, are the most common complications after CM resection. Neurological disorders, hemorrhage, myocardial infarction, and other minor complications are observed rarely after surgery. The 30-day mortality rate after CM excision was reported to range between 0% and 10%, and the recurrence rate during long-term follow-up was 0% to 7.4% (TABLE 4).[4-6,8,11,22,23] The number of patients operated in different studies may explain the wide ranges of 30-day mortality and recurrence rates during follow-up. Most studies on small patient samples have higher mortality

TABLE 4 Thirty-day mortality, long-term survival, and recurrence during follow-up in patients after cardiac myxoma resection

Study	Patients, n	30-day mortality, %	Long-term survival		Patients with recurrence, %
			Time	Patients, %	
Pinede et al[1]	122	3.5	Median, 3 y	96	5
Tasoglou et al[2]	67	4.5	10 y	96.8	4.4
Patil et al[3]	62	3.2	10 y	95	3.2
Wu et al[4]	112	0	15 y	89.5	2.7
Lee et al[5]	59	0	Range, 2 mo to 11 y	84.7	0
Vaideeswar et al[6]	84	–	–	–	4.7
Garrati et al[7]	98	3	15 y	89	1
Bordalo et al[8]	40	10	Mean, 30 mo	100	0
Obrenović-Kirćanski et al[9]	74	0	Mean, 12.2 y	81	0
Vroomen et al[10]	82	1.2	20 y	75	0
Aval et al[12]	42	2.4	Mean, 48.8 mo	–	–
Shah et al[13]	194	0.5	15 y	70	5.6
Lin et al[14]	68	0	10 y	96	3
Yóksel et al[16]	43	2.3	15 y	78	2.3
Bainchi et al[17]	30	0	12 y	88	0
Abu Abeeleh et al[18]	27	0	–	–	7.4
Lee et al[19]	93	3.2	30 y	75	2.1
Karabinis et al[20]	153	0.7	–	–	3.3
Nehaj et al[21]	41	2.4	Median, 4.6 y	85.3	0
Gür et al[22]	23	0	–	–	–
Cianciulli et al[23]	53	0	10 y	87.9	1.9
Jiang et al[24]	403	0.7	Median, 4.5 y	94.7	1.5

and recurrence rates compared with those involving a large number of patients. This could be elucidated by the fact that the operations were performed in low-volume cardiac surgery centers or on a small number of patients treated for CM or a cardiac tumor over a long time period. However, this limitation is acceptable, as CM is a rare tumor accounting for a small number of conditions managed with cardiac surgery. Of note, recurrence is observed months and years after surgery and many patients are lost to long-term follow-up, which is another limitation in identifying tumor recurrence. Recurrence was observed in familial CM and the Carney complex form, and recurrence of sporadic CM was rare. However, authors of all studies concluded that long-term survival rates after CM resection are very high. The survival rates range between 85% and 96%, which confirms the hypothesis that CM resection provides very good short- and long-term postoperative outcomes and remains the treatment of choice in patients with this rare cardiac disease.[2-4,7,13]

Conclusions Cardiac myxoma is a rare heart disease observed in a small proportion of the general population. In most patients, clinical manifestations of CM are nonspecific, and dyspnea is the most frequently reported symptom. Cardiac myxoma should be suspected in patients with chronic dyspnea, a history of peripheral or systemic embolization, an acute neurological disorder, and a family history of CM. These patients are at high risk of developing CM and TTE should be considered a basic diagnostic test in this population. Furthermore, CM can be diagnosed using TEE with a sensitivity higher than 90%. The differential diagnosis should include other benign and malignant (primary or metastatic) cardiac tumors, thrombi, and infective vegetations, which can be visualized by CT and MRI. A mutation in the *PRKAR1A* gene is observed in familial CM and in CM associated with Carney complex. Surgical excision of CM, which results in very good short-, mid-, and long-term outcomes, remains the treatment of choice for CM. The recurrence rate of CM is approximately 5% and may be observed months or years after surgery. Long-term follow-up with TTE is needed in all patients to improve their quality of life and decrease morbidity and mortality rates.

ARTICLE INFORMATION

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CONFLICT OF INTEREST None declared.

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