

The Echocardiographic Evaluation of Intracardiac Masses: A Review

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Echocardiography is an invaluable procedure for the evaluation of intracardiac masses, and can reliably identify mass location, attachment, shape, size, and mobility, while defining the presence and extent of any consequent hemodynamic derangement. With careful attention to mass location and morphology, and appropriate application of clinical information, echocardiography can usually distinguish between the 3 principal

intracardiac masses: tumor, thrombus, and vegetation. Transesophageal imaging frequently adds additional important information to the assessment of mass lesions and should always be considered when image quality is inadequate or pertinent clinical questions remain unanswered with surface imaging. This review will focus on primary and metastatic tumors of the heart. (J Am Soc Echocardiogr 2006;19:230-240.)

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Echocardiography provides a dynamic evaluation of intracardiac masses, and can identify and evaluate associated abnormalities. In addition, unlike other imaging modalities, a complete echocardiographic study can be performed at the bedside. However, echocardiographic image quality can be suboptimal, and ultrasound artifact can occasionally be mistaken for an anatomic mass. In addition, echocardiography provides a relatively narrow field of view compared with computed tomography and magnetic resonance imaging. Importantly, current echocardiographic techniques cannot characterize tissue, although contrast enhancement can demonstrate mass vascularity.

Careful identification of a mass lesion throughout the cardiac cycle in more than one imaging plane with appropriate machine settings and transducers (eg, high frequency and short focal length for the evaluation of suggested thrombus at the left ventricular [LV] apex) is an important first step in evaluating a suggested mass, and will decrease the likelihood of misinterpreting artifact as pathology. A thorough understanding of normal anatomy, of normal variants and embryologic remnants, and of the structural changes seen with certain operative and interventional procedures is crucial and will further avoid misdiagnosis. Lastly, it is important that pertinent clinical and historic information be available and thoughtfully applied to the final echocardiographic interpretation.

NORMAL VARIANTS AND MIMICKERS

Numerous normal anatomic variants exist that can easily be confused with primary mass lesions. In the LV, webs and heartstrings, prominent or calcified papillary muscles, and dense mitral annular calcification¹ can mimic abnormal pathology (note: some LV cords may contain conduction fibers of the left bundle).^{2,3} Prominent apical trabeculations, true ventricular noncompaction, and the apical form of hypertrophic cardiomyopathy can also be confused with tumors.⁴⁻⁷ (Ventricular noncompaction and apical hypertrophic cardiomyopathy are significant and distinct disease processes, not normal variants. Administration of contrast agents can help to distinguish these important entities from mass lesions.) In the left atrium (LA), beam-width artifact can cause interpretive confusion, as can the suture lines associated with cardiac transplantation. LA cords are rare but well-recognized findings (at least in the pathology suite). These cords typically originate at the

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atrial septum and insert into the atrial surface of the mitral leaflets; no clinical significance has ever been described, although there is some speculation that these strings may be the cause of vibratory murmurs (these cords should not be confused with true obstructive cor triatriatum.) The most notable description of an atrial cord came from Goforth⁸ in 1926: a 12-cm cord passed from the right atrium (RA) through the interatrial septum, LA, and LV, looping round the anterior mitral cusp, transfixing the anterior aortic cusp, and finally inserting into the aorta nearby. An interatrial septal aneurysm can be quite provocative and may appear as a cystic mass bulging into either atrium.⁹⁻¹² A dilated coronary sinus can mimic a LA mass in the parasternal long-axis view, as can a prominent descending aorta from the apex. A left arm injection of agitated saline will define a persistent left superior vena cava draining into the coronary sinus, which is the most common structural anomaly associated with dilatation of the coronary sinus. Occasionally multiple lobes of the LA appendage can be visualized with transthoracic imaging and can be confused with mass lesions in the main pulmonary artery; the transverse sinus can become prominent in this position as well when a large accumulation of pericardial fluid is present. Pectinate muscles in the appendage can mimic thrombus. An inverted LA appendage (typically a postoperative finding) can also be confused with tumor.^{13,14} A hiatal hernia can impinge on both atria and can be confirmed after ingestion of carbonated beverage.

A prominent moderator band or tricuspid papillary muscles can create diagnostic confusion in the right ventricle (RV). A fatty tricuspid annulus can be quite prominent. Monitoring lines and pacemaker wires create significant reverberation artifact (but do occasionally harbor thrombus). The RA can host a substantial number of congenital remnants, all of which can create confusion for echocardiographers. The crista terminalis is a dense muscle ridge that extends between the right sides of the superior and inferior caval orifices and continues cephalad to open into the RA appendage. It can be quite prominent, but is commonly seen in the same image plane as the superior vena cava, which should be a clue to its identity.¹⁵ Well known to echocardiographers, the eustachian valve is an incompetent valve flap of variable fullness and mobility that guards the orifice of the inferior vena cava (IVC) (interestingly, the superior vena cava has no valve). The Chiari network is a network of both coarse and fine fibers with attachments extending from the region of the crista terminalis to the eustachian valve or floor of the RA. Although not an unusual echocardiographic finding, and generally of no clinical significance, these fibers may become the site of thrombus formation. Entrapment of a catheter in the

network has been described, requiring thoracotomy for retrieval; several recent reports describe entanglement of an atrial septal defect closure device and septal ablation guidewire in the network.¹⁶⁻¹⁸ The network itself is generally quite mobile, but typically does not prolapse through the tricuspid orifice.

Pectinate muscles can be seen to layer the interior of the RA appendage and the free wall of the RA and, when prominent, can mimic thrombus. Venous varices (clumps of veins) of the heart, rare lesions of unknown incidence and with no distinct or specific echocardiographic characteristics, are surprisingly well described in the pathology literature.¹⁹ They almost always occur in the RA at the posteroinferior border of the fossa ovalis. They are clumpy, dilated venous channels with irregular borders. Reports of the echocardiographic diagnosis of these lesions suggest they closely resemble myxomas.²⁰ As noted previously, a hiatal hernia can mimic a mass lesion compressing the atria, but the "mass" should opacify when the patient drinks a carbonated beverage. Monitoring lines and pacemaker wires will also be evident in the RA and can occasionally become coiled, simulating a mass.

Fatty infiltrate of the atrial septum ("lipomatous hypertrophy") is preferential to the RA. Even though it can occasionally be quite prominent (≤ 6 cm) and is a true cardiac mass, it should be distinguishable from a tumor by typical location and shape. Fat typically spares the fossa ovalis producing a characteristic dumbbell appearance, with infiltrate in the superior and inferior portions of the atrial septum, with the preponderance of fat typically in the superior (cephalad) portion of the septum. When the atrial septum is massively infiltrated by fat, the amount of adipose tissue in other parts of the heart is always increased, particularly the RV epicardial surface. In the series described by Shirani and Roberts,²¹ hearts with widely distributed, massive deposits of fat actually floated in water. When there is diagnostic uncertainty regarding the cause of the infiltrate in the atrial septum, (as might be true if there is concern about metastatic disease), magnetic resonance imaging can easily distinguish fat from thrombus and neoplasm.

Certain valvular structures can be confused with mass lesions as well. The nodules of Arantius and lunulae, and the threadlike Lambl's excrescences (Figure 1), which are commonly found on the aortic valve in patients older than 60 years, can be confused with vegetations. Redundant supporting apparatus and redundant leaflet tissue of the mitral valve may mimic vegetations as well. Thoughtful application of clinical information is necessary to distinguish degenerative processes from infectious mechanisms. Excess epicardial fat may be confused with tumor contained in the pericardium, as can fibrinous debris in free pericardial fluid. Atelectatic segments

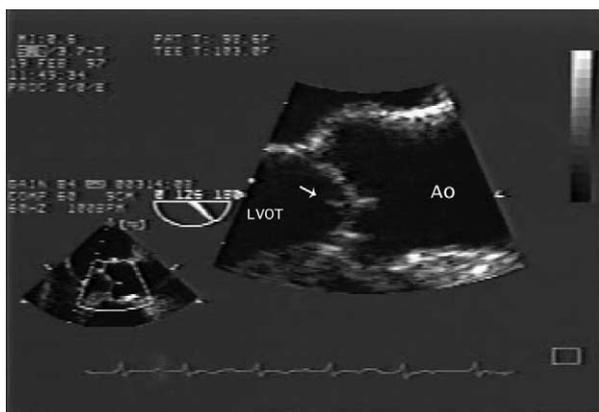


Figure 1 Transesophageal long-axis view of left ventricular outflow tract (LVOT) and aortic (AO) valve, demonstrating threadlike Lambli's excrescence (arrow) prolapsing through valve in diastole.

of lung can be misinterpreted as primary masses in the pleura.

PRIMARY TUMORS

The first description of an intracardiac mass was published in 1559. Before 1960, cardiac tumors were rarely diagnosed before death. An accurate premortem diagnosis was not made until 1934, when a primary cardiac sarcoma was identified. The first successful resection of a cardiac myxoma was in 1954.²²⁻²⁶ Today, although still rare, cardiac tumors represent an important group of cardiovascular abnormalities that, at least in the case of myxoma, have the potential for complete cure with timely diagnosis and surgical excision.

Primary tumors of the heart and pericardium are difficult to diagnose because they are so uncommon and because their clinical presentation is so variable. The incidence of primary tumors at autopsy ranges from 0.002% to 0.3%.^{27,28} Approximately 75% of primary cardiac tumors are benign. In adults the most common cardiac tumor is the myxoma; in children younger than 15 years, the most common tumor is the rhabdomyoma.²⁸ Rhabdomyomas are strongly associated with tuberous sclerosis. Of the primary malignant tumors, sarcomas are by far the most common, including angiosarcoma, rhabdomyosarcoma, and fibrosarcoma. Mesotheliomas and primary intracardiac lymphomas comprise approximately 6% of the malignant tumors of the heart.²⁹ Primary malignant cardiac tumors are rare in children.

The most common cardiac tumor is the myxoma, accounting for 25% of all cardiac neoplasms and 50% of the benign tumors of the heart in adults (Figure 2).



Figure 2 Transthoracic apical 4-chamber view demonstrating typical left atrial myxoma (arrow). LV, Left ventricle.

Approximately 75% of myxomas are found in the LA, with 20% in the RA and the remaining 5% equally distributed between the LV and RV.²⁹⁻³³ Valvular origin of these tumors is uncommon. Typically myxomas are pedunculated (approximately 10% are sessile) and attached in the region of the fossa ovalis. Depending on the size, the tumor may prolapse through the mitral orifice in diastole, occasionally producing a tumor "plop" and accounting for the common misdiagnosis of mitral stenosis. Myxomas are usually globular and irregular in shape (grape cluster), and appear heterogeneous on echocardiography, occasionally demonstrating cavitations and protruding frondlike extensions. Color flow and spectral Doppler are useful to evaluate functional obstruction to LA emptying. Work by Malekzadeh and Roberts³⁴ suggests that myxomas grow on average 1.8 cm or 14 g each year.

Myxomas usually present between the ages of 30 and 60 years, and can be sporadic, familial, or complex (syndrome myxoma). The sporadic variety is by far the most common. The familial variety constitutes approximately 7% of all myxomas, presents earlier in life, may be part of a syndrome (Carney's complex, NAME, LAMB), frequently includes multiple tumors in several chambers, and has a high rate of tumor recurrence.²⁷ If the patient is young with multiple tumors, screening of first-degree relatives is indicated. The complex variety, also known as Carney's complex, may include a combination of the following: (1) multiple pigmented skin lesions (lentigines); (2) breast adenomas; (3) skin myxomas; (4) endocrine overactivity (eg, pituitary adenomas); and (5) cardiac myxomas, often multiple.^{35,36} Syndrome myxoma is also occasionally referred to as the NAME syndrome (nevi, atrial myxoma, myxoid neurofibroma, ephelides) or the LAMB syndrome (lentigines, atrial myxoma, and blue nevi).³⁶ Transesophageal imaging is usually indicated when a

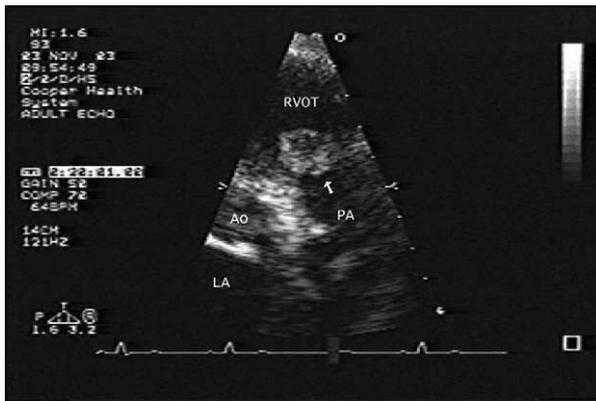


Figure 3 Transthoracic basal short-axis view demonstrating fibroelastoma of pulmonary valve (*arrow*). *Ao*, Aorta; *LA*, left atrium; *PA*, pulmonary artery; *RVOT*, right ventricular outflow tract.

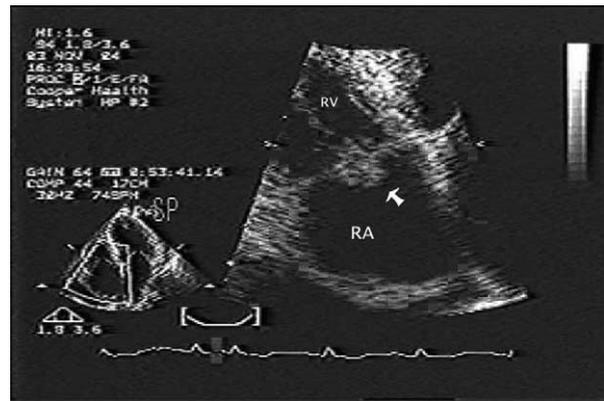


Figure 4 Transthoracic 4-chamber image demonstrating fibroelastoma of tricuspid valve (*arrow*). *RA*, Right atrium; *RV*, right ventricle.

myxoma is suggested to characterize the morphology and extent of the mass, and to exclude the presence of additional tumors. Surgical removal for all varieties of myxomas is the treatment of choice and intraoperative transesophageal echocardiography post-tumor excision is indicated to identify any residual tumor fragments. Careful follow-up is also indicated in patients with familial or syndrome myxoma as the tumor recurrence rate for these patients may be as high as 21%.³⁷

Papillary fibroelastomas are uncommon cardiac neoplasms of unknown prevalence.²⁹ They are generally small (<1 cm), usually single, almost always attached to valve surfaces, and may be pedunculated with some mobility (Figures 3 and 4). Recently, Sun et al³⁸ of the Cleveland Clinic published a comprehensive review of 162 patients with fibroelastomas identified during a period of 16 years, 141 of whom underwent echocardiography. Approximately 45% of the tumors were on the aortic valve (24 on the right coronary cusp (RCC), 6 on the left coronary cusp (LCC), and 19 on the non-coronary cusp (NCC) with 40 tumors on the aortic side of the valve. A total of 40 tumors were identified on the mitral valve (23 on the anterior and 17 on the posterior leaflet), with 32 on the atrial surface of the leaflets. These tumors can also attach to the supporting apparatus of the mitral valve. Typical echocardiographic features of fibroelastomas were identified, including: (1) the tumor is round, oval, or irregular in appearance with well-demarcated borders and a homogeneous texture; (2) most are small (99% < 20 mm); (3) many have stalks, and those with stalks are mobile; and (4) fibroelastomas may be single or multiple and are most often associated with cardiac valve disease, although in no patient in this series was the fibroelastoma believed to be responsible for the valve disease. Although fibroelastomas are often diagnosed incidentally, cerebral embolization, an-

gina, sudden death, acute myocardial infarction, pulmonary emboli, and retinal artery emboli related to fibroelastomas have been described; in the Cleveland series, 23 of 26 patients with prospectively diagnosed tumors had symptoms that could be attributable to embolization.³⁸⁻⁴³ Echocardiography is a reliable means of evaluating the extent and anatomic attachment of these very small tumors, but many go undetected. In the Cleveland series, 141 patients had echocardiograms, and 48 patients had fibroelastomas confirmed by pathologic examination that were not visible on echocardiography because they were less than .2 cm. The Cleveland authors reasoned that the tumors went unidentified because: (1) the tumor was masked by an associated lesion; (2) the tumor was too small; (3) the examination was not done carefully with a significant index of suspicion; or (4) there were no significant characteristics that enabled differentiation of fibroelastoma from degenerative valve disease. Surgical excision may be indicated in patients with large, mobile, left-sided tumors.

Rhabdomyomas are the most common cardiac tumor of infants and young children, are commonly multiple, and occur with equal frequency in the LV and RV.^{44,45} As previously noted, rhabdomyomas have a strong association with tuberous sclerosis, a familial neurologic syndrome characterized by hamartomas, epilepsy, mental retardation, and skin lesions. One study has indicated that 80% of patients with cardiac rhabdomyomas have tuberous sclerosis, and 60% of patients with tuberous sclerosis younger than 18 years have cardiac rhabdomyomas.⁴⁵ When associated with mechanical complications such as outflow tract obstruction, surgical excision may be indicated. Spontaneous resolution of these tumors is well described, however, and surgical intervention is usually not necessary in the asymptomatic patient.^{46,47}

sudden death. Angiomas are vascular tumors, and myocardial contrast echocardiography has been particularly useful in determining their vascular nature.⁵²⁻⁵⁴ Teratomas are usually found in the mediastinum. Only rarely are they intracardiac, and are usually within the pericardial space. These tumors most commonly occur in children. Teratomas have elements of all 3 germ cell layers, and can have skin, hair, and muscle. Large bloody pericardial effusions causing hemodynamic compromise are a well-described presentation of this uncommon tumor.^{55,56} Fetal echocardiography has been useful in identifying mediastinal masses causing cardiac compression, with associated accumulations of pericardial fluid.

About one fourth of all primary cardiac tumors are malignant, and nearly all are sarcomas.²⁹ Sarcomas most commonly occur in patients between the ages of 30 and 50 years, are unusual in children, are typically found in the right heart chambers, and occur equally in men and women.²⁷ There are several unsettling reports of sarcomas developing around surgically implanted Dacron grafts or prosthetic valves, both in the heart and in other peripheral vascular sites.⁵⁷⁻⁶⁰ Apparently, as reported by Oppenheimer et al,⁶¹ the common denominator of tumor-producing materials is a long-chain polymer structure, such as Dacron. Cardiac sarcomas are characteristically very aggressive tumors and, once diagnosed, are associated with a precipitous downhill course. They grow rapidly, and death is usually the result of widespread infiltration of the myocardium or extensive distant metastasis. Sarcomas may cause right heart failure as a result of obstruction anywhere in the right heart inflow/outflow tract; penetration into the pericardial space and subsequent pericardial effusion may occur. Dysrhythmias are common. Cardiac findings are determined by the location of the tumor. A recent report by Plana et al⁶² described a patient with angiosarcoma compressing the left upper pulmonary vein and LA appendage, invading the anteroseptal surface of the LA with extensive involvement of the RA as well. This patient presented with invasive hemodynamics consistent with mitral stenosis (pulmonary wedge-LV gradient) and effusive-constrictive pericarditis. Echo characteristics of sarcoma are not specific, but, less like a myxoma, sarcomas may attach at any site in the chamber, and many are sessile. A heterogeneous mass lesion in the right heart associated with dysrhythmia or conduction disturbance should include sarcoma in the differential. Although not commonly considered because these tumors metastasize so aggressively, cardiac extirpation, transplantation, and extensive right heart reconstruction have been performed with modest success.^{63,64}



Figure 8 Subcostal view of hypernephroma (*) invading right atrium (RA) by dilated inferior vena cava (IVC). Tumor is heterogeneous in appearance and abuts superior border of IVC.

METASTATIC TUMORS

Whereas primary tumors of the heart are rare, cardiac metastases have been described in up to 20% of patients with malignancies of other organ systems, and are up to 40 times more common than primary tumors.²⁹ No malignant tumor preferentially metastasizes to the heart, with the possible exception of malignant melanoma, which involves the heart in up to 50% of patients.⁶⁵ Cardiac metastases are encountered typically in patients with widespread systemic tumor dissemination; even in this setting, the heart may still be spared tumor deposition because of vigorous cardiac contractility and rapid coronary blood flow.

Tumors spread to the heart by lymphatic channels, direct/venous extension, and hematogenous spread.⁶⁶ The tumors that most commonly manifest cardiac metastasis are lung, breast, ovarian, kidney, leukemia, lymphoma, esophageal, and, as noted, melanoma.^{27,29} Although solid intracardiac metastasis from melanoma is well described, most commonly cardiac extension of melanoma is subclinical and manifests as “charcoal” heart, with tumor studing the pericardial surface.⁶⁷ Both benign and malignant tumors can extend to the heart through the IVC. The most commonly reported benign tumor with IVC intracardiac extension is intravascular leiomyomatosis of pelvic or uterine origin. Of all tumors that metastasize to the heart by the IVC, renal cell carcinoma (hypernephroma) is the most common, with up to 43% of patients with this tumor demonstrating RA involvement⁶⁸ (Figures 8 and 9). Wilms’ tumor (common in children), uterine leiomyosarcoma, and hepatomas may also metastasize to the heart by the IVC. Their point of origin and extension into the IVC usually can distinguish these

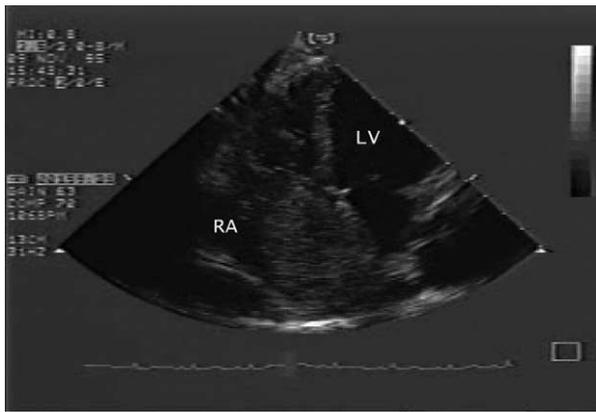


Figure 9 Four-chamber view of hypernephroma nearly filling dilated right atrium (RA). LV, Left ventricle.

metastases from the typical myxoma, and this attachment is best imaged in the subcostal plane. Carcinomas of the lung and breast commonly invade the heart, either by direct extension or by lymphatic channels, given the proximity of the heart to mediastinal lymphatic channels²⁷ (Figure 10).

Pericardial metastasis occurs more often than myocardial invasion, and manifestations include pericarditis, pericardial effusion with and without tamponade, and atrial dysrhythmias.²⁷ Pericardial effusion is overall the most common echocardiographic finding in metastatic disease. The effusion may contain solid material adherent to the visceral or parietal pericardium; these masses may be tumor or may be clotted blood. As is true of every mass lesion identified in the heart or pericardium, cytologic examination of pericardial fluid is essential, as not all pericardial effusions in the setting of other primary organ malignancy yield malignant cells. Unfortunately, routine cytologic examination of the fluid is associated with a false-negative rate of perhaps as high as 20%.⁶⁹ When large effusions associated with tamponade and adherent masses are discovered, constrictive physiology may be operant after pericardiocentesis. Recurrent effusions in this setting are common, and pericardial window may be necessary. Solid pericardial metastasis that extends into cardiac chambers can be very aggressive, with the tumor expanding rapidly and causing significant hemodynamic derangement, including obstruction to cavity emptying and filling. Because of the aggressive nature of metastatic tumors, patients with intracavitary extension are likely to have symptoms directly related to the tumor. Chemotherapy and tumor resection or debulking may alleviate symptoms and prolong survival.

The carcinoid syndrome (flushing, gastrointestinal hypermotility with secretory diarrhea, bronchospasm associated with wheezing, and carcinoid heart disease) results from circulating

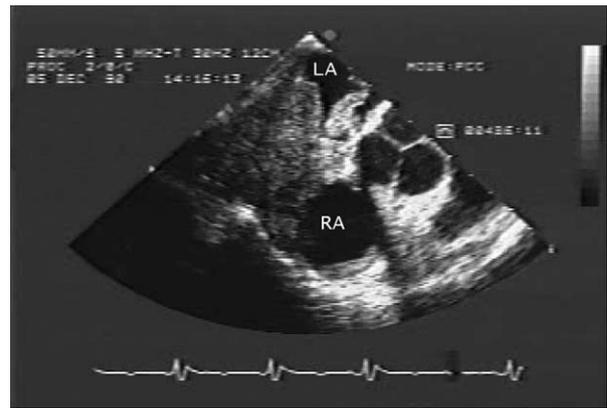


Figure 10 Transesophageal image at level of aortic valve, demonstrating large mass lesion that has eroded into both atria and originates outside heart. Patient presented with emboli to both upper extremities, and was subsequently determined to have metastatic lung cancer. LA, Left atrium; RA, right atrium.

humoral substances secreted by the tumor.⁷⁰ The diarrhea probably results from circulating serotonin, which is secreted in large amounts by carcinoid tumors; the flushing and bronchospasm are related to the release of kinin peptides. Patients with primary carcinoid tumors of the ileum are those who develop carcinoid syndrome; only those patients in that group who have liver metastasis develop the distinctive lesions of the heart. These lesions are always located on the right side of the heart and occasionally on the left as well. When the primary carcinoid tumor is of a pulmonary bronchus, the carcinoid valvular lesions may be limited to the left-sided valves. In this setting, the liver may be tumor free.⁷¹ Carcinoid valve lesions are characterized by plaque-like, fibrous endocardial thickening that causes retraction and fixation of the tricuspid and pulmonary valve leaflets. Tricuspid regurgitation (TR) is a nearly universal finding; tricuspid stenosis, pulmonary regurgitation, and pulmonary stenosis may also occur.⁷⁰ In an extensive review of 76 patients with carcinoid syndrome, Pellikka et al⁷² from the Mayo Clinic identified 5 patients with left-sided valve involvement. Carcinoid tumor involved the lung in one of those 5 patients; in 3, a patent foramen ovale with right-to-left shunting was identified with contrast echocardiography. One of those 5 patients had neither lung involvement nor shunting on contrast echocardiography, but had been symptomatic for 5 years before the diagnosis of carcinoid syndrome was made. Carcinoid involvement of all 4 valves was identified, and the patient underwent replacement of 4 valves. These 5 patients all had typical right-sided valvular lesions as well.

As noted, the principal cardiac finding of carcinoid involvement of the heart is TR. The mechanism

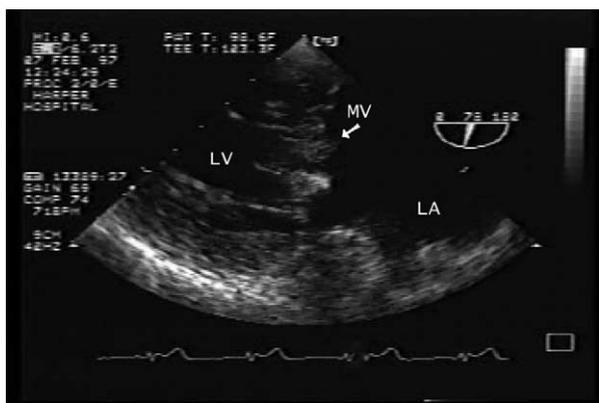


Figure 11 Two-chamber transgastric transesophageal echocardiographic image. *LA*, Left atrium; *LV*, left ventricle; *MV*, mitral valve.

of TR is well described in a review by Roberts.⁷⁰ The plaquing occurs nearly entirely on the downstream (ventricular) side of the septal and posterior tricuspid leaflets. On the anterior tricuspid leaflet the deposits can occur on both sides, which results in adherence of the leaflet to the underlying mural endocardium and in significant valvular incompetence and occasionally some degree of stenosis as well. The dominant pulmonic valve lesion tends to be stenosis. Carcinoid is the only condition in which both right-sided valves are uniformly involved, and the lesions are pulmonary stenosis and TR. The coexistence of these lesions is particularly unfavorable because the pulmonic stenosis increases the TR. As noted in the review of Pellikka et al,⁷² the spectral Doppler display of the TR was characterized by a fairly low velocity (2.5 M) dagger-shaped profile, with early peaking and rapid fall-off, consistent with rapid equalization of RA and RV pressures. The typical valve morphology was of rigid leaflets fixed in a semi-open position. Doming of the tricuspid leaflets was not noted in any patients in this series. Surgical replacement of dysfunctional valves is described, but the mortality appears to be fairly high (30-day mortality of 35% in one series and 56% in another).^{73,74} In addition, carcinoid plaquing on bioprostheses has been observed at autopsy.⁷⁵

Although carcinoid syndrome with valvular plaque deposition is the most common cardiac consequence of carcinoid tumor, solid tumor metastasis may occasionally occur. In a series of 11 patients with pathologically confirmed myocardial metastasis, 9 intracavitary tumors were detected by echocardiography in 6 of the 11 patients.⁷⁶ The mean tumor size was 2.4 cm. Of the 9 tumors, 4 were located in the LV myocardium, 4 in the RV myocardium, and one in the interventricular septum. The tumors were homogeneous and none were pedunculated or mobile. Six macroscopic tumors were noted at autopsy, with

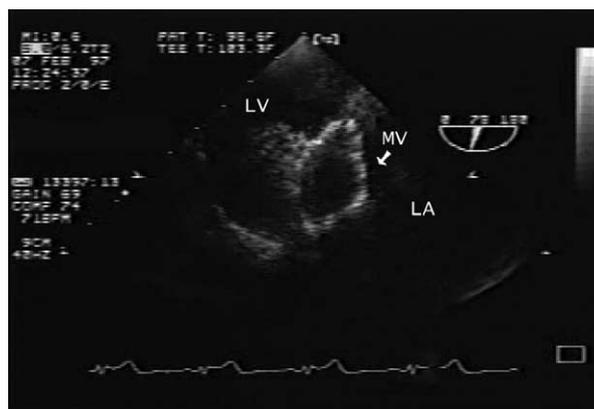


Figure 12 Same patient as in Figure 11, in same transgastric transesophageal echocardiographic plane, demonstrating well-circumscribed, echolucent structure (*arrow*) involving mitral valve leaflets with extension into supporting apparatus. At operation, lesion was blood cyst. *LA*, Left atrium; *LV*, left ventricle; *MV*, mitral valve.

a mean size of .35 cm. None of those tumors were detected by echocardiography, even on retrospective review. Interestingly, although all 11 patients had carcinoid syndrome, the typical valve lesions were present in only 8 patients.

Although not true neoplasms, cysts are occasionally found within the heart and pericardium. The most common cysts that have described echocardiographic characteristics are pericardial cysts, echinococcal (hydatid) cysts, and blood cysts. *Echinococcus* is a parasite endemic to Greece and Turkey, and intracardiac cysts result from the larval stage of the parasite. Intracardiac cysts account for less than 2% of all hydatid disease, and invade the myocardium by the coronary circulation. They are found in the LV free wall, the interventricular septum, the RV, and the pericardium. Transesophageal imaging is indicated for the evaluation of these lesions. They are usually septated and can occasionally calcify. Rupture, dysrhythmia, and emboli are common and can result in death. Operation is the treatment of choice.⁷⁷⁻⁸¹ Pericardial cysts are rounded echolucent structures typically adjacent to the RA. The diagnosis can be made on chest radiograph by the identification of a rounded mass along the right heart border. Pericardial cysts are usually asymptomatic, but can become quite large and cause compression of the RA and RV, and the surrounding mediastinal structures, including the bronchus and esophagus. Operation is indicated when significant symptoms dominate the clinical picture.⁸²⁻⁸⁵ Blood cysts are presumed congenital cysts typically found on the lines of closure of valvular endocardium (Figures 11 and 12). They appear as well-circumscribed masses with thin walls and an echolucent core. Common findings at infant autopsy, these cysts are rare in adults, and only a few

reports of prospective echocardiographic diagnosis are available. There is no consensus as to the origin of blood cysts, but the cysts may form during valve development as a result of blood pressed and trapped in crevices that later seal. Because blood cysts are so rare, consensus is also lacking concerning appropriate treatment. Careful echocardiographic monitoring of the cysts for change in size and for the assessment of change in cardiac function may be appropriate, and operation may be indicated when the cysts are noted to cause cardiac dysfunction.⁸⁶⁻⁸⁹

Because echocardiography is the procedure of choice for the evaluation of intracardiac masses, techniques must be meticulously applied and data cautiously interpreted. The mandate for the echocardiographer is appropriate knowledge of and careful attention to cardiac anatomy, use of multiple scan planes, and sound application of clinical information.

REFERENCES

- Harpaz D, Auerbach I, Vered Z, Motro M, Tobar A, Rosenblatt S. Caseous calcification of the mitral annulus: a neglected, unrecognized diagnosis. *J Am Soc Echocardiogr* 2001;14:825-31.
- Tamborini G, Pepi M, Celeste F, Muratori M, Susini F, Maltagliati A, et al. Incidence and characteristics of left ventricular false tendons and trabeculations in the normal and pathologic heart by second harmonic echocardiography. *J Am Soc Echocardiogr* 2004;17:367-74.
- Asmussen I, Ornvold K. Strings containing left bundle branch conductive tissue in the left ventricle. *Acta Pathol Microbiol Immunol Scand (A)* 1985;93:345-9.
- Agmon Y, Connolly H, Olson L, Khanderia B, Seward J. Noncompaction of the ventricular myocardium. *J Am Soc Echocardiogr* 1999;12:859-63.
- Lowery MH, Martel JA, Zambrano JP, Ferreira A, Eco L, Gallagher A. Noncompaction of the ventricular myocardium: the use of contrast-enhanced echocardiography in the diagnosis. *J Am Soc Echocardiogr* 2003;16:94-6.
- Stollberger C, Finsterer J. Left ventricular hypertrabeculation/noncompaction. *J Am Soc Echocardiogr* 2004;17:91-100.
- Ward RP, Weinert L, Spencer KT, Furlong KT, Bednarz J, DeCara J, et al. Quantitative diagnosis of apical cardiomyopathy using contrast echocardiography. *J Am Soc Echocardiogr* 2002;15:316-22.
- Goforth JL. Unique heart anomaly: free fibrous cord passing through three heart chambers to the aorta. *JAMA* 1926;86:1612-3.
- Mugge A, Daniel WG, Angermann C, Spes C, Khanderia B, Kronzon I, et al. Atrial septal aneurysm in adult patients: a multicenter study using transthoracic and transesophageal echocardiography. *Circulation* 1995;91:2785-92.
- Malaterre HR, Cohen F, Kallee K, Deharo JC, Djiane P. Giant interatrial septal aneurysm mimicking a right atrial tumor. *Int J Card Imaging* 1998;14:163-6.
- Malaterre HR, Kallee K, Deharo JC, Djiane P. Right and left sided interatrial septal aneurysm mimicking atrial tumor and stroke. *J Am Soc Echocardiogr* 1998;11:829-31.
- Ginon I, Mestrallet C, Barthelet M, Robin J, Andre-Fouet X. A closed interatrial septal aneurysm, filled with blood, mimicking a tumor in the right atrium. *Eur J Echocardiogr* 2000;1:289-90.
- Minich LL, Hawkins JA, Tani LY, Judd VE, McGough EC. Inverted left atrial appendage presenting as an unusual left atrial mass. *J Am Soc Echocardiogr* 1995;8:328-30.
- Toma DM, Stewart RB, Miyake-Hull CY, Otto CM. Inverted left atrial appendage mimicking a left atrial mass during mitral valve repair. *J Am Soc Echocardiogr* 1995;8:557-9.
- Pharr J, West M, Kusomoto F, Figueredo V. Prominent crista terminalis appearing as a right atrial mass on transthoracic echocardiogram. *J Am Soc Echocardiogr* 2002;15:753-5.
- Goldschlager A, Goldschlager N, Brewster H. Catheter entrapment in a Chiari network involving an atrial septal defect. *Chest* 1972;62:345-6.
- Cooke J, Gelman J, Harper R. Chiari network entanglement and herniation into the left atrium by an atrial septal defect occluder device. *J Am Soc Echocardiogr* 1999;12:601-3.
- Shimoike E, Ueda N, Maruyama T, Kaji Y, Niho Y. Entrapment of a guide wire by the Chiari network in a patient with ablated idiopathic ventricular tachycardia. *J Interv Card Electrophysiol* 2001;5:219-22.
- Heggveit HA. Thrombosed varices of the heart [abstract]. *Am J Pathol* 1966;48:50a.
- Harrity P, Tazelaar H, Edwards W, Orszulak T, Freeman W. Intracardiac varices of the right atrium: a case report and review of the literature. *Int J Cardiol* 1995;48:177-81.
- Shirani J, Roberts WC. Clinical, electrocardiographic and morphologic features of massive fatty deposits ("lipomatous hypertrophy") in the atrial septum. *J Am Coll Cardiol* 1993;22:226-38.
- Mahaim I. Les Tumeurs et les Polypes de Coeur. Etude Anatomico-clinique. Paris: Masson; 1945.
- Barnes AR, Beaver DC, Snell AM. Primary sarcoma of the heart: report of a case with ECG and pathological studies. *Am Heart J* 1934;9:480-4.
- Goldberg HP, Glenn F, Dotter CT, Steinberg I. Myxoma of the left atrium: diagnosis made during life with operative and post-mortem findings. *Circulation* 1952;6:762-6.
- Crafoord CL. A case report. In: Lam CR, editor. Proceedings of international symposium on cardiovascular surgery. Philadelphia: WB Saunders Co; 1955. p. 202.
- Effert S, Domanig E. Diagnosis of intra-auricular tumors and large thrombi with the aid of ultrasonic echography. *Dtsch Med Wochenschr* 1959;84:6-8.
- Roberts WC. Primary and secondary neoplasms of the heart. *Am J Cardiol* 1997;80:671-82.
- Lam KY, Dickens P, Chan ACL. Tumors of the heart: a 20-year experience with review of 12,485 consecutive autopsies. *Arch Pathol Med* 1993;117:1027-31.
- Burke A, Virmani R. Atlas of tumor pathology. In: Tumors of the heart and great vessels. Third series, fascicle 16. Washington (DC): Armed Forces Institute of Pathology; 1996. p. 231.
- St John Sutton MG, Mercier LA, Guiliani ER, Lie JT. Atrial myxoma: a review of clinical experience in 40 patients. *Mayo Clin Proc* 1980;55:371-6.
- Markel ML, Waller BF, Armstrong WF. Cardiac myxoma: a review. *Medicine (Baltimore)* 1987;66:114-25.
- Burke AP, Virmani R. Cardiac myxomas: a clinicopathologic study. *Am J Clin Pathol* 1994;100:671-80.
- Keeling IM, Oberwalder P, Anelli-Monti M, Schuchlenz H, Demel U, Tilz GP, et al. Cardiac myxomas: 24 years of

- experience in 49 patients. *Eur J Cardiothorac Surg* 2002;22:971-7.
34. Malekzadeh S, Roberts WC. Growth rate of left atrial myxoma. *Am J Cardiol* 1989;64:1075-6.
 35. Carney JA, Gordon H, Carpenter PC, Shenoy BV, Go VL. The complex of myxomas, spotty pigmentation, and endocrine overactivity. *Medicine (Baltimore)* 1985;64:270-83.
 36. Mahilmaran A, Seshadri M, Nayar PG, Sudarsana G, Abraham KA. Familial cardiac myxoma; Carney's complex. *Tex Heart Inst J* 2003;30:80-2.
 37. Van Gelder M, O'Brien DJ, Staples ED, Alexander JA. Familial cardiac myxoma. *Ann Thorac Surg* 1992;53:419-24.
 38. Sun JP, Asher C, Yang XS, Cheng G, Scalia GM, Massed AG, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas. *Circulation* 2001;103:2687-701.
 39. Zell DN, Diamond M, Beringer D. Angina and sudden death resulting from papillary fibroelastoma of the aortic valve. *Ann Emerg Med* 1985;14:470-3.
 40. Kasarskis EJ, O'Connor W, Earle G. Embolic stroke from cardiac papillary fibroelastoma. *Stroke* 1988;19:1171-3.
 41. Topol EJ, Biern RO, Reitz BA. Cardiac papillary fibroelastoma and stroke: echocardiographic diagnosis and guide to excision. *Am J Cardiol* 1986;80:129-32.
 42. Waltenberger J, Thelin S. Papillary fibroelastoma as an unusual source of repeated pulmonary embolism. *Circulation* 1994;89:24-33.
 43. Zamora RL, Adelberg DA, Berger AS, et al. Branch retinal artery occlusion caused by a mitral valve papillary fibroelastoma. *Ophthalmology* 1995;119:325-9.
 44. Chan HSL, Sonley MJ, Moes CAF, Daneman A, Smith CR, Martin DJ. Primary and secondary tumors of the heart, pericardium, and great vessels: a report of 75 cases and review of the literature. *Cancer* 1985;56:825-36.
 45. Bass JL, Brenningstall GN, Swaiman KF. Echocardiographic incidence of cardiac rhabdomyoma in tuberous sclerosis. *Am J Cardiol* 1985;55:1379-82.
 46. Fesslova V, Villa L, Rizzuti T, Mastrangelo M, Mosca F. Natural history and long-term outcome of cardiac rhabdomyomas detected prenatally. *Prenat Diagn* 2004;24:241-8.
 47. Verhaaren HA, Vanakker O, DeWolf D, Suys B, Francois K, Matthys D. Left ventricular outflow obstruction in rhabdomyoma of infancy: meta-analysis of the literature. *J Pediatr* 2003;143:258-63.
 48. Cina SJ, Smialek JE, Burke AP, Virmani R, Hutchins GM. Primary cardiac tumors causing sudden death: a review of the literature. *Am J Forensic Med Pathol* 1996;17:271-81.
 49. Wong JA, Fishbein MC. Cardiac fibroma resulting in fatal ventricular arrhythmia. *Circulation* 2000;101:e168-70.
 50. Cho JM, Danielson G, Puga F, Dearani JA, McGregor CGA, Tazelaar HD, et al. Surgical resection of ventricular cardiac fibromas: early and late results. *Ann Thorac Surg* 2003;76:1929-34.
 51. Lang-Lazdunski L, Oroudji M, Pansand Y, Vissuzaine C, Hvass U. Successful resection of giant intrapericardial lipoma. *Ann Thorac Surg* 1994;58:238-40.
 52. Bednarz JE, Spencer KT, Weinert L, Sugeng L, Mor-avi V, Lang RM. Identification of cardiac masses and abnormal blood flow patterns with harmonic power Doppler contrast echocardiography. *J Am Soc Echocardiogr* 1999;12:871-5.
 53. Kirkpatrick JN, Wong T, Bednarz JE, Spencer KT, Sugeng L, Ward RP, et al. Differential diagnosis of cardiac masses using contrast echocardiographic perfusion imaging. *J Am Coll Cardiol* 2004;43:1412-9.
 54. Lepper W, Shivalkar B, Rinkevich D, Belcik T, Wei K. Assessment of the vascularity of a left ventricular mass using myocardial contrast echocardiography. *J Am Soc Echocardiogr* 2002;15:1419-22.
 55. Tollens M, Grab D, Lang D, Hess J, Oberhoffer R. Pericardial teratomas: prenatal diagnosis and course. *Fetal Diagn Ther* 2003;18:432-6.
 56. Tollens T, Casselman F, Devlieger H, Gewillig MH, Vandenberghe K, Lerut TE, et al. Fetal cardiac tamponade due to an intrapericardial teratomas. *Ann Thorac Surg* 1998;66:559-60.
 57. Okada M, Takeuchi E, Mori Y, Ichihara S, Usui A, Ueda Y. An autopsy case of angiosarcoma arising around a woven Dacron prosthesis after a Cabrol operation. *J Thorac Cardiovasc Surg* 2004;127:1843-5.
 58. Ben-Izhak O, Vlodavsky E, Ofer A, Engel A, Nitecky S, Hoffman A. Epithelioid angiosarcoma associated with a Dacron vascular graft. *Am J Surg Pathol* 1999;23:1418-23.
 59. Durrleman NM, El-Hamamsy I, Demaria RG, Carrier M, Perrault LP, Albat B. Cardiac lymphoma following mitral valve replacement. *Ann Thorac Surg* 2005;79:1040-2.
 60. Fyfe BS, Quintana CS, Kaneko M. Aortic sarcoma four years after Dacron graft insertion. *Ann Thorac Surg* 1994;58:1752-4.
 61. Oppenheimer BS, Oppenheimer ET, Stout AP, Willhite M, Danishefski I. The latent period in carcinogenesis by plastics in rats, and its relation to the presarcomatous stage. *Cancer* 1958;11:204.
 62. Plana JC, Iskander SS, Ostrowski M, Howell JF, Kurrelmeyer K, Raizner AE, et al. Cardiac angiosarcoma: an unusual presentation simulating mitral stenosis and constrictive-effusive pericarditis. *J Am Soc Echocardiogr* 2003;16:1331-3.
 63. Talbot SM, Taub RN, Keohan ML, Edwards N, Galantowicz ME, Schulman L. Combined heart and lung transplantation for unresectable primary cardiac sarcoma. *J Thorac Cardiovasc Surg* 2002;124:1145-8.
 64. Hoffmeier A, Deiters S, Schmidt C, Tjan TD, Schmid C, Drees G, et al. Radical resection of cardiac sarcoma. *Thorac Cardiovasc Surg* 2004;52:77-81.
 65. Mukai K, Shinkai T, Tominaga K, Shomosato Y. The incidence of secondary tumors of the heart and pericardium: a ten-year study. *Jpn N Clin Oncol* 1988;18:195-201.
 66. Colucci WS, Braunwald E. Primary tumors of the heart. In: Braunwald E, editor. *Heart disease: a textbook of cardiovascular medicine*. Philadelphia: WB Saunders; 1980. p. 1501-16.
 67. Waller BV, Gottdeiner JS, Virmani R, Roberts WC. The "charcoal heart": melanoma to the cor. *Chest* 1980;77:671-6.
 68. Almasi GH. Surgery for tumors with cavoatrial extension. *Semin Thorac Cardiovasc Surg* 2000;12:111-8.
 69. Warren WH. Malignancies involving the pericardium. *Semin Thorac Cardiovasc Surg* 2000;12:119-29.
 70. Roberts WC. A unique heart disease associated with a unique cancer: carcinoid heart disease. *Am J Cardiol* 1997;80:251-6.
 71. Kulke M, Mayer RJ. Medical progress: carcinoid tumors. *N Engl J Med* 1999;340:858-68.
 72. Pellikka PA, Tajik AJ, Khandheria BK, Seward JB, Callahan JA, Pitot HC, et al. Carcinoid heart disease: clinical and echocardiographic spectrum in 74 patients. *Circulation* 1993;87:1188-96.
 73. Connolly HM, Nishimura RA, Smith HC, Pellikka PA, Mullaney CJ, Kvols LK. Outcome of cardiac surgery for carcinoid heart disease. *J Am Coll Cardiol* 1995;25:410-6.
 74. Robiolio PA, Rigolin VH, Harrison JK, Lowe JE, Moore JO, Bashore TM, et al. Predictors of outcome of tricuspid valve replacement in carcinoid heart disease. *Am J Cardiol* 1995;75:485-8.

75. Ridker PM, Chartow GM, Karlson EW, Neish AS, Schoen FJ. Bioprosthetic tricuspid valve stenosis associated with extensive plaque deposition in carcinoid heart disease. *Am Heart J* 1991;121:1835-8.
76. Pandya U, Pellikka P, Enriquez-Sarano M, Edwards WD, Schaff H, Connelly H. Metastatic carcinoid tumor to the heart: echocardiographic-pathologic study of 11 patients. *J Am Coll Cardiol* 2002;40:1328-32.
77. Ben-Hamda K, Maatouk F, Ben-Farhat M, Betbout F, Gamra H, Addad F, et al. Eighteen year experience with Echinococcus of the heart: clinical and echocardiographic features in 14 patients. *Int J Cardiol* 2003;91:145-51.
78. Atilgan D, Kudar H, Tukek T, Ozcan M, Yildirim O, Elmaci TT, et al. Role of transesophageal echocardiography in diagnosis and management of cardiac hydatid cyst: report of three cases and review of the literature. *J Am Soc Echocardiogr* 2002;15:271-4.
79. Makaryus AN, Hametz C, Mieres J, Kort S, Carneglia J, Mangion J. Diagnosis of suspected cardiac echinococcosis with negative serologies: role of transthoracic, transesophageal, and contrast echocardiography. *Eur J Echocardiogr* 2004;5:223-7.
80. Hassine E, Kraoua S, Marniche K, Bousnina S, Lefi A, Kachboura S, et al. A dead and calcified right ventricular hydatid cyst: the limits of imaging. *Presse Med* 2003;32:1802-4.
81. Nisanoglu V, Erdil N, Isik B, Battaloglu B, Alat I. Acute abdominal aortic embolism caused by rupture of a cardiac hydatid cyst. *Ann Vasc Surg* 2004;18:484-6.
82. Patel J, Park C, Michaels J, Rosen S, Kort S. Pericardial cyst: case reports and a literature review. *Echocardiography* 2004;21:269-72.
83. Bava GL, Magliani L, Bertoli D, Gorrieri PF, Rimini A, Zaccagnini G, et al. Complicated pericardial cyst: atypical anatomy and clinical course. *Clin Cardiol* 1998;21:862-4.
84. Bandeira FC, de Sa VP, Moriguti JC, Rodrigues AJ, Jurca MC, Filho OC, et al. Cardiac tamponade: an unusual complication of pericardial cyst. *J Am Soc Echocardiogr* 1996;9:108-12.
85. Antonini-Canterin F, Piazza R, Ascione L, Pavan D, Nicolosi GL. Value of transesophageal echocardiography in the diagnosis of compressive, atypically located pericardial cysts. *J Am Soc Echocardiogr* 2002;15:192-4.
86. Pelikan HMP, Tsang TSM, Seward JB. Giant blood cyst of the mitral valve. *J Am Soc Echocardiogr* 1999;13:1005-7.
87. Nkomo V, Miller FA. Eustachian valve cyst. *J Am Soc Echocardiogr* 2001;14:1224-6.
88. Sim EKW, Wong ML, Tan KT, Sim SK. Blood cyst of the tricuspid valve. *Ann Thorac Surg* 1996;61:1012-3.
89. Prasad A, Callahan MJ, Malouf JF. Acquired right atrial blood cyst: a hitherto unrecognized complication of cardiac operation. *J Am Soc Echocardiogr* 2003;16:377-8.