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a normal or slightly enlarged heart. Pericardial calcification is most common in tuberculous pericarditis. Pericardial calcification may, however, occur in the absence of constriction, and constriction may occur without calcification.

Inasmuch as the common physical signs of cardiac disease (mur murs, cardiac enlargement) may be inconspicuous or absent in chronic constrictive pericarditis, hepatic enlargement and dysfunction asso ciated with jaundice and intractable ascites may lead to a mistaken diagnosis of hepatic cirrhosis. This error can be avoided if the neck veins are inspected and found to be distended with the characteristic waveform features. PART 6 Disorders of the Cardiovascular System The transthoracic echocardiogram often shows pericardial thicken ing, dilation of the inferior vena cava and hepatic veins, and a sharp halt to rapid left ventricular filling in early diastole, with normal ventricular systolic function and flattening of the left ventricular posterior wall. There is a distinctive pattern of transvalvular flow velocity on Doppler echocardiography (Fig. 281-4). During inspiration, there is an exagger ated reduction in blood flow velocity in the pulmonary veins and across the mitral valve, and a leftward shift of the ventricular septum; the opposite occurs during expiration. Diastolic flow velocity in the inferior vena cava into the right atrium and across the tricuspid valve increases in an exaggerated manner during inspiration and declines during expiration. However, echocardiography cannot definitively establish or exclude the diagnosis of constrictive pericarditis; CT and MRI are more accurate, with the latter useful in evaluating myocardial involvement. ■ ■DIFFERENTIAL DIAGNOSIS As with chronic constrictive pericarditis, cor pulmonale (Chap. 264) may be associated with marked systemic venous hypertension, little pulmonary congestion, a (left) heart that is not enlarged, and a para doxical pulse. However, in cor pulmonale, advanced parenchymal pulmonary disease is usually apparent and venous pressure falls dur ing inspiration (i.e., Kussmaul’s sign is negative). Tricuspid stenosis (Chap. 277) may also simulate chronic constrictive pericarditis with congestive hepatomegaly, splenomegaly, ascites, and venous distention. However, the characteristic murmur and that of accompanying mitral stenosis are usually present. Because it can be corrected surgically, it is important to distin guish chronic constrictive pericarditis from restrictive cardiomyopathy (Chap. 266), which has a similar pathophysiologic underpinning (i.e., restriction of ventricular filling). The differentiating features are sum marized in Table 281-2. When a patient has progressive, disabling,

and unresponsive congestive heart failure and displays any of the features of constrictive heart disease, Doppler echocardiography to record respira tory effects on transvalvular flow (Fig. 281-4) should be performed and an MRI or CT scan should be obtained to detect or exclude constrictive pericarditis because the latter is usually correctable. TREATMENT Constrictive Pericarditis Pericardial resection is the only definitive treatment of constric tive pericarditis and should be as complete as possible. Coronary arteriography should be carried out preoperatively in patients aged

> 50 years to exclude accompanying coronary artery disease. The benefits derived from cardiac decortication are usually progressive over a period of months. The risk of this operation depends on the extent of penetration of the myocardium by the fibrotic and calcific process, the severity of myocardial atrophy, the extent of second ary impairment of hepatic and/or renal function, and the patient's general condition. Operative mortality is in the range of 5–10% even in experienced centers; the patients with the most severe dis ease, especially secondary to radiation therapy, are at highest risk. Therefore, surgical treatment should, if possible, be carried out as early as possible. Subacute Effusive-Constrictive Pericarditis This form of pericardial disease is characterized by the combination of a tense effu sion in the pericardial space and constriction of the heart by thickened

pericardium. As such, it shares a number of features with both chronic pericardial effusion producing cardiac compression and with pericar dial constriction. It may be caused by tuberculosis (see below), multiple attacks of acute idiopathic pericarditis, radiation, traumatic pericardi tis, renal failure, scleroderma, and neoplasms. The heart is generally enlarged, and a paradoxical pulse is usually present. After pericardio centesis, the physiologic findings may change from those of cardiac tamponade to those of pericardial constriction. Furthermore, the intra pericardial pressure and the central venous pressure may decline, but not to normal. The diagnosis can be established by pericardiocentesis followed by pericardial biopsy. Wide excision of both the visceral and parietal pericardium is usually effective therapy. Tuberculous Pericardial Disease This chronic infection is a common cause of chronic pericardial effusion, especially in the developing world where active tuberculosis and HIV are endemic. Tuberculous pericarditis may present as pericardial effusion, chronic constrictive pericarditis, or subacute effusive-constrictive pericarditis (see above). The clinical picture is that of a chronic, systemic illness in a patient with pericardial effusion. It is important to consider this diagnosis in a patient with known tuberculosis, with HIV, and with fever, chest pain, weight loss, and enlargement of the cardiac silhouette of undetermined origin. If the etiology of chronic pericardial effusion remains obscure despite detailed analysis including culture of the peri cardial fluid, a pericardial biopsy, preferably by a limited thoracotomy, should be performed. If definitive evidence is still lacking but the speci men shows granulomas with caseation, antituberculous chemotherapy (Chap. 183) is indicated. If the biopsy specimen shows a thickened pericardium after 2–4 weeks of antituberculous therapy, pericardiectomy should be per formed to prevent the development of constriction. Tubercular cardiac constriction should be treated surgically while the patient is receiving antituberculous chemotherapy. ■ ■FURTHER READING Arula S, Madsen N: Management of acute pericarditis. Curr Opin Caradiol 38:364, 2023. Bayes-Coenis A et al: Colchicine in pericarditis. Eur Heart J 38:1706, 2017. Garcia MJ: Constrictive

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Atrial Myxoma and

Other Cardiac Tumors Cardiac tumors can be broadly classified into those that arise primar ily in the heart and those that reflect metastatic disease from a distant primary source. Primary cardiac tumors can be further divided into those that are pathologically benign and those that are malignant.

TABLE 282-1 Imaging Modalities and Their Utility in the Evaluation of Cardiac Tumors MODALITY UTILITY IN CARDIAC TUMOR EVALUATION Transthoracic

echocardiography (TTE) (including two-dimensional, three-dimensional, and contrast) Assessment of tumor location and size and its impact on adjacent structures (e.g., valves, pericardium). Transesophageal echocardiography (TEE) Improved tumor characterization and spatial resolution compared with TTE. May aid in determining surgical approach. Cardiac magnetic resonance imaging (MRI) with gadolinium contrast Improved tissue characterization, definition of tumor size, and identification of local invasion when compared with TTE or TEE. May differentiate tumor from thrombus. Gated cardiac computed tomography (CT) Provides anatomic assessment and tissue characterization of the tumor. Useful when patients cannot tolerate MRI or when MRI is not feasible (e.g., patients with implantable cardiac devices). Allows for better assessment of calcified lesions and evaluation of extracardiac tumor involvement. Nuclear imaging (including 18F-fluorodeoxyglucose positron emission tomography [FDG-PET]) Definition of extracardiac disease. May be useful in diagnosis of certain cardiac tumors (e.g., neuroendocrine tumors), but assessment of smaller tumors may be limited by surrounding myocardial FDG uptake. Overall, primary cardiac tumors are relatively uncommon, whereas secondary involvement of the heart or pericardium occurs in as many as 20% of patients with end-stage metastatic cancer. While patients with cardiac tumors may present with a variety of symptoms, many patients are asymptomatic at the time of diagnosis, with the tumor being identified incidentally on imaging studies performed for other reasons. Cardiac tumors need to be differentiated from other cardiac masses such as vegetation, thrombus, inflammatory myofibroblastic tumors, or myocardial hypertrophy. Echocardiography is usually the initial imaging modality used to evaluate cardiac tumors; however, a variety of imaging modalities are now available, and a multimodality approach is often necessary for accurate diagnosis and clarification of treatment options (Table 282-1). When diagnosis is uncertain after echocardiography, cardiac magnetic resonance imaging (MRI) is often useful to help further differentiate lesions. Imaging characteristics, including size, location, presence of myocardial infiltration, and first pass perfusion of gadolinium contrast agents, can help differentiate benign from malignant tumors. However, tissue histology remains the gold standard for diagnosis of cardiac tumors. ■ ■PRIMARY TUMORS Primary tumors of the heart are rare, occurring in ~1 in

2000 patients in autopsy series. Approximately three-quarters are histologically benign, the majority of which are myxomas. Malignant tumors, almost all of which are sarcomas, account for 25% of primary cardiac tumors. All cardiac tumors, regardless of pathologic type, have the potential to cause life-threatening complications. Many tumors are now surgically curable; thus, early diagnosis is imperative. Clinical Presentation Cardiac tumors may present with a wide array of cardiac and noncardiac manifestations. These manifestations, which depend in large part on the location and size of the tumor as well as its impact on surrounding cardiac structures, are often nonspecific features of more common forms of heart disease, and include chest pain, syncope, congestive heart failure (CHF), murmurs, arrhythmias, conduction disturbances, pericardial effusion, and pericardial tampon ade. Additionally, embolic phenomena and constitutional symptoms may occur. Myxoma Myxomas are the most common type of primary cardiac tumor in adults, accounting for one-third to one-half of all cases at postmortem examination, and approximately three-quarters of the tumors treated surgically. They occur at all ages most commonly in the

third through sixth decades, with a female predilection. Approximately 90% of myxomas are sporadic; the remainder are familial with autoso mal dominant transmission. The familial variety often occurs as part of a syndrome complex (Carney complex) that includes (1) myxomas (cardiac, skin, and/or breast), (2) lentigines and/or pigmented nevi, and (3) endocrine overactivity (primary nodular adrenal cortical dis ease with or without Cushing’s syndrome, testicular tumors, and/or pituitary adenomas with gigantism or acromegaly). The genetic basis of this complex has not been elucidated completely; however, inactivating mutations in the tumor-suppressor gene PRKAR1A, which encodes the protein kinase A type I-α regulatory subunit, have been identified in ~70% of patients with Carney complex.

CHAPTER 282 Atrial Myxoma and Other Cardiac Tumors Pathologically, myxomas are gelatinous structures that consist of myxoma cells embedded in a stroma rich in glycosaminoglycans. Most sporadic tumors are solitary, arise from the interatrial septum in the vicinity of the fossa ovalis (particularly in the left atrium), and are often pedunculated on a fibrovascular stalk. In contrast, familial or syndromic tumors tend to occur in younger individuals, are often multiple, may be ventricular in location, and are more likely to recur after initial resection. Myxomas commonly present with obstructive signs and symptoms. The most common clinical presentation mimics that of mitral valve dis ease: either stenosis owing to tumor prolapse into the mitral orifice or regurgitation resulting from tumor-induced valvular trauma or distor tion. Ventricular myxomas may cause outflow tract obstruction similar to that caused by subaortic or subpulmonic stenosis. The symptoms and signs of myxoma may be sudden in onset or positional in nature, owing to the effects of gravity on tumor position. A characteristic lowpitched sound, a “tumor plop,” may be appreciated on auscultation during early or mid-diastole and is thought to result from the impact of the tumor against the mitral valve or ventricular wall. Myxomas also may present with peripheral or pulmonary embolic phenomenon (resulting from embolization of tumor fragments or tumor-associated thrombus) or with constitutional signs and symptoms, including fever, weight loss, cachexia, malaise, arthralgias, rash, digital clubbing, and Raynaud’s phenomenon. These constitutional symptoms are likely the result of cytokines (e.g., interleukin 6) secreted by the myxoma. Labo ratory abnormalities, such as hypergammaglobulinemia, anemia, poly cythemia, leukocytosis, thrombocytopenia or thrombocytosis, elevated erythrocyte sedimentation rate, and elevated C-reactive protein level are often present. These features account for the frequent

misdiagnosis of patients with myxomas as having endocarditis, collagen vascular disease, or a paraneoplastic syndrome. Two-dimensional and three-dimensional transthoracic and/or transesophageal echocardiography are useful in the diagnosis of cardiac myxoma and allow for assessment of tumor size and deter mination of the site of tumor attachment, both of which are impor tant considerations in the planning of surgical excision (Fig. 282-1). A B FIGURE 282-1 Transthoracic echocardiogram demonstrating a large atrial myxoma. The myxoma (Myx) fills the entire left atrium in systole (A) and prolapses across the mitral valve and into the left ventricle (LV) during diastole (B). RA, right atrium; RV, right ventricle. (Courtesy of Dr. Michael Tsang; with permission.)

PART 6 Disorders of the Cardiovascular System FIGURE 282-2 Cardiac magnetic resonance imaging demonstrating a rounded mass (M) within the left atrium (LA). Pathologic evaluation at the time of surgery revealed it to be an atrial myxoma. LV, left ventricle; RA, right atrium; RV, right ventricle. Computed tomography (CT) and MRI may provide important addi tional information regarding size, shape, composition, and surface characteristics of the tumor (Fig. 282-2). Although cardiac catheterization and angiography were previously performed routinely before tumor resection, they no longer are con sidered mandatory when adequate noninvasive information is avail able and other cardiac disorders (e.g., coronary artery disease) are not considered likely. Additionally, catheterization of the chamber from which the tumor arises carries the risk of tumor embolization. Because myxomas may be familial, echocardiographic screening of first-degree relatives is appropriate, particularly if the patient is young and has multiple tumors or features of a myxoma syndrome. TREATMENT Myxoma Surgical excision using cardiopulmonary bypass is indicated regard less of tumor size and is generally curative. Myxomas recur in 12–22% of familial cases but in only 1–2% of sporadic cases. Tumor recurrence most likely results from multifocal lesions in the former setting and incomplete tumor resection in the latter. Other Benign Tumors Cardiac lipomas, although relatively com mon, are usually incidental findings at postmortem examination; however, they may grow as large as 15 cm, may present as an abnor mality of the cardiac silhouette on chest x-ray, and should be resected if they produce symptoms owing to mechanical interference with cardiac function, arrhythmias, or conduction disturbances. Papillary fibroelastomas are friable tumors with frond-like projections that are usually solitary and are the most common tumors of the cardiac valves. Although usually clinically silent, they can cause valve dysfunction and may embolize distally, resulting in transient ischemic attacks, stroke, or myocardial infarction. In general, these tumors should be resected even when asymptomatic, although a more conservative approach may be considered for small, right-sided lesions. Rhabdomyomas and fibro mas are the most common cardiac tumors in infants and children and usually occur in the ventricles, where they may produce mechanical obstruction to blood flow, thereby mimicking valvular stenosis, CHF, restrictive or hypertrophic cardiomyopathy, or pericardial constriction. Rhabdomyomas are probably hamartomatous growths, are multiple in

RA RV T T LV T T FIGURE 282-3 Transthoracic echocardiogram revealing multiple tumors (T) consistent with rhabdomyomas in a 1-day-old infant. The largest tumor (arrows) was located in the left antrioventricular groove and measured 2 cm × 2 cm. LV, left ventricle; RA, right atrium; RV, right ventricle. 90% of cases, occur in ~50% of children with tuberous sclerosis, and are associated with mutations in the tumor-suppressor genes TSC1 and TSC2 (Fig. 282-3). These tumors have a tendency to regress completely or partially; only tumors that cause obstruction require surgical resec tion. Fibromas are usually single, universally ventricular in location, often calcified, and may be associated with mutations in the tumorsuppressor gene PTCH1. Fibromas tend to grow and

cause arrhythmias and obstructive symptoms and should be completely resected when possible. Paragangliomas are rare chromaffin cell tumors that represent extra-adrenal pheochromocytomas. Most are located in the roof of the left atrium and can be identified on cardiac CT or MRI or with nuclear scanning using 131I-metaiodobenzylguanidine. They are highly vascu lar and may be hormonally active, resulting in uncontrolled hyperten sion. Extensive surgical resection is usually required. Hemangiomas and mesotheliomas are generally small tumors, most often intramyo cardial in location, and may cause atrioventricular (AV) conduction disturbances and even sudden death as a result of their propensity to develop in the region of the AV node. Other benign tumors arising from the heart include teratoma, chemodectoma, neurilemoma, and granular cell myoblastoma. Malignant Tumors Almost all malignant primary cardiac tumors are sarcomas, which may be of several histologic types; angiosarcomas are the most common type in adults, whereas rhabdomyosarcomas are the most common type in children. In general, sarcomas are characterized by rapid progression that culminates in the patient’s death within weeks to months from the time of presentation as a result of hemodynamic compromise, local invasion, or distant metastases. Almost one-third are metastatic at the time of initial diagnosis, usually involving the lungs. Sarcomas commonly involve the right side of the heart, are rap idly growing, frequently invade the pericardial space, and may obstruct the cardiac chambers or venae cavae. Sarcomas also may occur on the left side of the heart and may be mistaken for myxomas. Isolated cardiac lymphomas have been rarely described but more commonly occur in the context of systemic disease. They are more common in men and in the elderly; usually involve the right heart; may present with arrhythmias, syncope, CHF, or constitutional symptoms; and are usually of the large B-cell type. TREATMENT Malignant Tumors The optimal therapy for cardiac sarcoma is complete resection, often with neoadjuvant and postoperative chemotherapy; however, at the time of presentation, many of these tumors have spread too extensively to allow for surgical excision. Although there are

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