Paraganglioma originating at the aortic root and extending along the anterior wall of the left ventricle in the atrioventricular sulcus.
We Welcome Your Questions and Comments

Inquiries, letters to the editor and original contributions can be directed to MDCVJ@tmhs.org.
Tumors of the heart are uncommon, yet with the advent of diagnostic procedures such as angiography, echocardiography, computed tomography and magnetic resonance imaging, they are more frequently diagnosed and amenable to cure by surgery. Prior to the development of cardiopulmonary bypass, excision was rarely possible. Currently, with precise anatomic diagnosis and open-heart techniques, complete excision and cure is possible, especially for benign lesions. Myxoma is the most frequent primary tumor in most reported series, and results of surgery are usually successful. Other benign tumors are also mostly removable without complications. Most primary malignancies may not enjoy such a favorable prognosis.

Since the first reported cardiac transplantation in 1967, the technique for cardiac replacement has become standardized, and is now proved to be therapeutic for complex cardiac disease. In advanced neoplastic disease of the heart or adjacent organs or tissues, access to the posterior aspect of the heart and left atrium may not be technically possible. Thus, removal of the heart may be necessary to expose and remove the tumor. In 1984, such a situation was encountered in a patient at my hospital, and excision of the tumor and cardiac repair was performed. Subsequent bleeding from the highly vascularized mediastinum prevented survival, but the technique of autotransplantation was thus introduced. My assistant at that operation was Dr. Michael J. Reardon, who was inspired to apply the technique later in many such challenges. He has become an authority for the treatment of cardiac neoplasms, with impressive results. I congratulate him and his dedicated team.
I would like to thank Dr. Winters for allowing me to serve as organizer of this issue of the *Methodist DeBakey Cardiovascular Journal* on primary tumors of the heart. Cardiac tumors are classified as primary (tumors that arise in the heart itself) secondary or metastatic tumors. Secondary tumors are 40 to 100 times more common than primary tumors of the heart. Metastatic tumors of the heart are generally not considered candidates for surgical resection, although there are rare cases in which resection with reasonable risk and good long-term outcome can be achieved. These will not be discussed in this issue of the journal.

Primary tumors of the heart arise from cellular elements of the heart itself and are broadly divided into benign and malignant. In our experience at the Methodist DeBakey Heart & Vascular Center, we have found 80% of our tumors to be benign and 20% to be malignant. Almost all of our malignant cases are sarcoma due to our referral network for these cases, but a review of the literature shows that 75% are sarcoma and the rest are a variety of tumors, with lymphoma leading the pack. Benign tumors tend, to have an excellent outcome at a low surgical risk except for certain rare and complex benign lesions such as cardiac paraganglioma. The most common benign tumors — myxoma (>50%), papillary fibroelastoma and lipoma — all can be cured with complete surgical resection and low risk. Our current interest in benign tumors involves an active interest in better diagnosis and minimally invasive approaches to these tumors. Cardiac paragangliomas are rare, highly vascular, usually very large and technically difficult to treat surgically. Our group is currently reviewing our cardiac paraganglioma work to define surgical approaches best suited for this difficult problem.

The use of MRI for imaging cardiac tumors will be discussed by Dr. Dipan Shah of the Methodist DeBakey Heart & Vascular Center, and echocardiography for the imaging of cardiac tumors will be discussed by Dr. Juan Carlos Plana from The University of Texas MD Anderson Cancer Center. From the cardiovascular surgery group at the Methodist DeBakey Heart & Vascular Center, Dr. Brian Bruckner will discuss our experience with benign cardiac tumors in detail, and Dr. Mahesh Ramchandani will discuss our inroads in the use of minimally invasive approaches to treat benign cardiac tumors.

Malignant primary tumors of the heart are rare. I developed a strong interest in cardiac tumors while serving as a cardiothoracic resident with Dr. Denton Cooley. Dr. Cooley was treating a patient sent from Italy with a large left atrial paraganglioma. Surgery had been attempted in Italy and was unsuccessful due to the size, location and vascularity of the tumor. To solve the issue of poor exposure of the left atrium that contains a large tumor, Dr. Cooley explanted the heart, resected the tumor, and rebuilt the left atrium without the heart in the way. He then reimplanted the heart and introduced cardiac autotransplant as a technique for cardiac tumors, igniting my lifelong interest in cardiac tumors and especially malignant primary cardiac tumors.

Primary cardiac sarcoma treated without surgery has about a 10% survival at 1 year. The patients we see with primary cardiac sarcoma are often young, with families that they are trying to raise. They have often been given this dismal prognosis and told that nothing realistic can be done. Although this remains a difficult and deadly disease, significant progress has been made; our 2-year survival for cardiac sarcoma is now 61%, and we have patients alive at 10 years. The goal of our group as expressed in this issue of the *Methodist DeBakey Cardiovascular Journal* is to define the proper classification system, define treatment protocols, and define and enhance survival of these patients.

Dr. Shanda Blackmon of our thoracic surgery group at The Methodist Hospital will join me in exploring our approach and results in treating pulmonary artery sarcoma, and Dr. David Rice of The University of Texas MD Anderson Cancer Center will do the same with regard to left heart sarcoma. Dr. Ara Vaporciyan of
The University of Texas MD Anderson Cancer Center will join me in discussing our treatment of right heart sarcoma. Finally, Dr. Bob Benjamin, the head of the Department of Sarcoma Medical Oncology at The University of Texas MD Anderson Cancer Center, will discuss future direction in the biologic treatment of cardiac sarcoma. Our approach to these difficult tumors has been to cast a wide net in bringing together a multidisciplinary team within and across institutions to tackle this formidable problem for the benefit of our patients. This collaboration has resulted in numerous publications,2-23 2 IRB protocols at The Methodist Hospital, and an international registry. Our entire group looks to the future with great hope of continuing progress in our battle to defeat this disease.

References

Pseudomasses

It is important to recognize that there are a number of normal structures or abnormal lesions that are not true masses, but can mimic a cardiac or paracardiac mass. The most common of these is a right atrial pseudotumor produced by a prominent crista terminalis, which can appear as a right atrial mass on echocardiography. A prominent Chiari network or eustacian valve can also be mistaken as a right atrial mass; these can be easily visualized by CMR, and a true mass can be excluded (Figure 1A).

Extracardiac structures that can simulate cardiac pathology include a large hiatal hernia that can produce significant displacement of the atria. Additionally, the heterogeneous internal architecture in a hiatal hernia can lead to significant confusion on echocardiography. CMR is readily able to discern a hiatal hernia from a true extracardiac mass (Figure 1B).

Tissue Characterization by CMR

Potentially the most robust feature of CMR in the evaluation of cardiac or paracardiac masses is its ability to characterize the composition of abnormal tissue better than any other imaging modality. This is accomplished by imaging the mass using a variety of different MRI pulse sequences that are designed to allow examination for a vast number of different biological properties (e.g., T1 weighted, T2 weighted, perfusion or delayed contrast enhancement). These biologic properties can be analyzed to help narrow down a differential diagnosis as to the etiology of the mass, as shown in Table 1. One will notice that there may be significant overlap between biologic properties of different neoplasms, and therefore, decisions regarding chemotherapy or radiation treatment can generally not be made purely on the basis of CMR findings, but typically require a tissue diagnosis. There are, however,
Figure 1. Typical cardiac pseudomasses that are easily identified on CMR. Frame A represents a 4-chamber view displaying a prominent crista terminalis (red arrow) — a normal structure in the right atrium that can appear as a cardiac mass on echocardiography. Frame B represents an axial image in the chest that demonstrates a large hiatal hernia (red arrows), which is compressing upon the left atrium. This can create the appearance of a left atrial mass on echocardiography. RV: right ventricle; RA: right atrium; LV: left ventricle; LA: left atrium.

Table 1. CMR characteristics of cardiac masses

<table>
<thead>
<tr>
<th>Cardiac Mass</th>
<th>T1 Weighted</th>
<th>T2 Weighted</th>
<th>Post Contrast</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myxoma</td>
<td>Isointense, heterogeneous</td>
<td>Hyperintense, heterogeneous</td>
<td>Heterogeneous enhancement</td>
</tr>
<tr>
<td>Papillary fibroelastoma</td>
<td>Isointense</td>
<td>Hyperintense</td>
<td>Hyperintense</td>
</tr>
<tr>
<td>Rhabdomyoma</td>
<td>Iso- or hyperintense</td>
<td>Slightly hyperintense</td>
<td>Hyperintense</td>
</tr>
<tr>
<td>Fibroma</td>
<td>Iso- or hyperintense</td>
<td>Hypointense</td>
<td>Hyperintense</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>Isointense</td>
<td>Hyperintense, heterogeneous</td>
<td>Hyperintense or heterogeneous</td>
</tr>
<tr>
<td>Paragangioma</td>
<td>Iso- or hypointense</td>
<td>Hyperintense</td>
<td>Hyperintense</td>
</tr>
<tr>
<td>Intravenous leiomyomatosis</td>
<td>Isointense</td>
<td>Isointense</td>
<td>Heterogeneous</td>
</tr>
<tr>
<td>Bronchogenic cyst</td>
<td>Hypointense</td>
<td>Hyperintense</td>
<td>None</td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>Isointense, with hyperintense areas</td>
<td>Iso- or hyperintense</td>
<td>Hyperintense</td>
</tr>
<tr>
<td>Undifferentiated sarcoma</td>
<td>Isointense</td>
<td>Isointense</td>
<td>Nonspecific</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>Isointense</td>
<td>Isointense, heterogeneous</td>
<td>Central nonenhancing areas</td>
</tr>
<tr>
<td>Osteosarcoma</td>
<td>Hyperintense</td>
<td>Hyperintense</td>
<td>Nonspecific</td>
</tr>
<tr>
<td>Malignant fibrous histiocytoma</td>
<td>Isointense</td>
<td>Hyperintense, heterogeneous</td>
<td>Nonspecific</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>Isointense</td>
<td>Hyperintense</td>
<td>Nonspecific</td>
</tr>
<tr>
<td>Fibrosarcoma</td>
<td>Isointense, heterogeneous</td>
<td>Hyperintense</td>
<td>Central nonenhancing areas</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Hypo- or isointense</td>
<td>Hyperintense</td>
<td>Variable</td>
</tr>
</tbody>
</table>
a number of cardiac masses that can be definitively diagnosed by CMR and management decisions made without the need for an invasive procedure. The most commonly encountered of these are intracardiac thrombus, benign lipomatous hypertrophy of the interatrial septum, and benign intracardiac lipoma.

**Thrombus.** Although not a neoplasm, thrombus is actually the most common intracardiac mass.\(^3\),\(^6\) It tends to occur most commonly in the left atrial appendage, but can occur within any cardiac chamber. Left atrial appendage thrombus is most commonly associated with atrial fibrillation, atrial dilation and mitral valve disease.\(^2\) Ventricular thrombus usually occurs in the setting of cardiomyopathy, most commonly ischemic in nature. The formation of a ventricular aneurysm, or simply decreased contractility predisposed to sluggish blood flow, and can be a substrate for thrombus formation. Denudation of the endothelium from prior infarction is also a likely contributor to thrombus formation. Right atrial thrombi can develop adjacent to long-standing central venous catheters, or be “thrombi in transit” from embolization of lower-extremity thrombi.

CMR is significantly more sensitive and specific than echocardiography for detecting ventricular or atrial thrombi, with studies demonstrating an approximately twofold increase in sensitivity compared to echocardiography.\(^3\) This sensitivity is significantly improved by the administration of intravenous contrast material.

Pre-contrast cine imaging often will fail to detect ventricular thrombi that clearly are seen as low-signal intensity foci, following the administration of contrast. In addition, post-contrast delayed-enhancement inversion recovery images with a long inversion time are exquisitely sensitive for the detection of even small thrombi.\(^7\) In this instance, the long inversion time allows recovery of signal by virtually all tissues except thrombus, which remains low in signal intensity and therefore dark on imaging (Figure 2). At the Methodist DeBakey Heart & Vascular Center, CMR diagnosis of an intracardiac mass as thrombus mitigates the need for additional invasive procedures, and patients are simply followed with serial imaging after an appropriate period of anticoagulation.

**Lipomatous Hypertrophy of the Interatrial Septum.** Lipomatous hypertrophy of the interatrial septum is not a true neoplasm, and it is not truly hypertrophy of the adipocytes. Rather, it represents a nonencapsulated hyperplasia of otherwise normal fatty cells within the interatrial septum. This diagnosis is based on the finding of fatty deposits in the interatrial septum, resulting in a diameter exceeding 2 cm in transverse dimension.

The exact etiology is unknown, but it appears to be associated with obesity and advanced age. The average age at diagnosis is approximately 69 years, and there appears to be a slight male predominance.\(^3\) It is said to be associated with tachyarrhythmias, predominantly atrial in origin. The exact incidence of this disorder is

---

**Figure 2.** Identification of a cardiac mass as left ventricular thrombus. Frame A (left side) displays the short- and long-axis cine-CMR imaging in an individual referred for evaluation of a left ventricular mass (red arrows). On the right side of frame A are the corresponding short and long axis contrast CMR images performed 10 to 15 minutes after administration of gadolinium, using a long inversion time (TI). This demonstrates no delayed contrast update within the cardiac mass (red arrows). This finding is highly specific for thrombus. The patient was treated with oral anticoagulation and returned for follow-up imaging in approximately 9 months. On follow-up cine CMR imaging (frame B), the mass is no longer present (red arrows).
difficult to discern, as some series do not separate this disorder from lipomas. However, it increasingly is recognized based on echocardiographic imaging and CMR.

CMR is quite specific in this disorder. Thickening of the interatrial septum to a diameter greater than 2 cm is noted, and sparing of the fossa ovalis is apparent. This often results in a dumbbell- or barbell-like appearance (Figure 3). The fatty hyperplasia results in high signal on T1-weighted images through the interatrial septum. The addition of fat saturation to the imaging sequences results in signal dropout, confirming the fatty nature of these lesions (Figure 3). In addition, cine imaging with SSFP sequences results in a characteristic chemical shift artifact at the interface between the fatty portions of the septum and the remainder of the myocardium. As CMR is highly specific for diagnosis and since the condition is benign, no further evaluation is generally necessary.

**Lipoma.** Cardiac lipomas are benign neoplasms composed of encapsulated mature adipose tissue, similar to extracardiac lipomas. There does not appear to be any sex predilection. They are usually discovered in adulthood but can occur at any age. Most originate along the epicardial surface of the heart, although myocardial or endocardial origins have been reported and can occasionally protrude into cardiac cavities.

Most lipomas do not cause any symptoms, but occasionally can lead to dyspnea if there is obstruction of blood flow, and/or to arrhythmias if there is involvement of the cardiac conduction system. In the absence of symptoms, most cardiac lipomas do not warrant surgical excision and, as such, appropriate non-invasive diagnosis is very important. Given their fatty nature, cardiac lipomas are high in signal intensity on T1-weighted sequences, and they show evidence of signal dropout on fat saturation sequences. CMR therefore provides a specific diagnosis of these lesions and can obviate the need for unnecessary cardiac surgery in patients with this condition.

**Other Primary Benign Cardiac Tumors.** Cardiac myxoma represents approximately 50% of all primary benign cardiac tumors. Myxomas occur predominantly in adulthood and do not tend to recur after complete excision, except when they are part of a familial myxoma syndrome. Myxomas are always intracavitary masses and are most commonly located in the left atrium, usually attached to the fossa ovalis; a small number originate in the right atrium. Ventricular origin for a myxoma is very rare (<2%). CMR features of myxoma are described in Table 1 and an example is shown in Figure 4.

Papillary fibroelastomas are benign avascular papillomas of the endocardium, and are similar to Lambli's excrescences both histologically and in their predilection for cardiac valves; they differ in that they tend to be larger in size and away from the site of valve closure (whereas Lambli's excrescences, are by definition, at the sites of valve closure). These benign lesions are usually detected incidentally by echocardiography that is performed for another indication. They rarely require excision except in instances where there are neurological symptoms, presumably on an emboli basis.

![Figure 3. Lipomatous hypertrophy of the interatrial septum. Tissue characteristics of lipomatous hypertrophy are highly specific for diagnosis of this condition (see text for details). FS: fat saturation.](image)

![Figure 4. Example of a right atrial myxoma. Tumor is attached to the atrial septum (cine systole) but prolapses through the tricuspid valve in diastole. The mass is isointense to myocardium on T1-weighted imaging, hyperintense to myocardium on T2-weighted imaging, and demonstrates heterogeneous uptake on post-contrast imaging.](image)
Cardiac fibromas are fairly uncommon tumors that are congenital in origin and represent a discrete focal mass of collagen and fibroblasts. Because of their congenital origin, they primarily occur in children. Approximately one-third of patients present with arrhythmias, one-third with heart failure or cyanosis, and one-third are detected incidentally. Their CMR features are described in Table 1; the key findings are that they demonstrate reduced signal on T2-weighted imaging (due to their limited water content) and demonstrate very high signal intensity on delayed enhancement imaging (due to their high collagen content).

Paragangliomas are tumors originating from neuroendocrine cells, and typically present with symptoms of catecholamine excess (e.g., hypertension, tachyarrhythmias or heart failure). Their point of origin is typically in the atria, along the atrioventricular sulcus, or at the root of the great vessels. Imaging features are described in Table 1; key findings are of high signal on T2-weighted imaging, heterogeneous delayed hyperenhancement, and high vascularity on perfusion imaging (Figure 6).

**Malignant Cardiac Tumors.** Malignant tumors comprise approximately 25% of primary cardiac neoplasms. Most of these are some form of sarcoma with the remainder being lymphomas. Imaging characteristics of malignant tumors are quite similar, with most lesions demonstrating invasion of surrounding...
structures and myocardium, poor border definition, and frequent coexisting pericardial effusion. While the exact etiology cannot be distinguished based on imaging characteristics, findings favoring malignancy usually can be detected.

Angiosarcoma is the most common form of cardiac sarcoma and accounts for approximately 40% of primary cardiac malignancies. Angiosarcoma has a predilection for the right atrium with more than 90% originating at this location. Location of origin is a key distinguishing feature different from most other forms of sarcomas (e.g., undifferentiated sarcomas, malignant fibrous histiocytoma, osteosarcoma, leiomyosarcoma, fibrosarcoma or rhabdomyosarcoma) that tend to arise in the left atrium. As the imaging features of different forms of sarcomas are similar, specific differentiation can only be made by histology. See Figure 7 for an example of a right atrial angiosarcoma.

Primary cardiac lymphomas are distinct from systemic lymphoma with cardiac involvement. These neoplasms have increased prevalence in immunocompromised patients, but also can occur in immunocompetent patients. Clinical features are typically of dyspnea, arrhythmia, superior vena cava obstruction or cardiac tamponade due to frequent involvement in the pericardium resulting in pericardial effusions. Imaging features include frequent epicardial surface or pericardial involvement. Tissue characteristics are described in Table 1 and an example case is shown in Figure 8.

Associated Findings

Even in patients with cardiac masses where a tissue diagnosis has been made, CMR plays a role in determining the full extent of tumor involvement and associated findings. This information is useful for the surgeon planning the excision procedure. In addition to determining which cardiac chambers are involved, CMR can provide several additional, useful pieces of information that are detailed in this section.

Pericardial Effusion. The presence of a pericardial effusion in association with a cardiac mass is highly suggestive of malignancy. In fact, pericardial effusion is probably the most common imaging manifestation of metastatic disease. Additionally, the presence of nodular implants on the pericardium should be viewed as highly suggestive of metastatic disease. Lastly, CMR imaging characteristics of pericardial effusion can be useful to ascertain whether an effusion is likely transudative, exudative or hemorrhagic.

Coronary Involvement. Cardiac masses that involve the epicardium of the heart can occasionally affect the coronary arteries, leading to direct coronary invasion that results in coronary stenosis and occasionally myocardial infarction. CMR is very useful in identify-
ing coronary involvement, and delayed-enhancement CMR of the myocardium can identify associated myocardial infarction.\textsuperscript{17} Identification of coronary involvement is useful in surgical planning, as these patients may require coronary excision and replacement with a free graft or an internal mammary graft.\textsuperscript{18}

\textbf{Cardiac Valve Involvement.} Tumors of the heart can occasionally infiltrate into the cardiac valves. This can lead to the need to replace the affected valve either with a mechanical valve (in which case the patient will require chronic anticoagulation therapy) or a biologic valve (in which case the patient may require reoperation in 10 to 12 years). CMR can readily identify tumor infiltration into the valve leaflets or valve annulus. Even in patients who do not have valve involvement directly from tumor, cine and phase-contrast, CMR can identify coincident valve stenosis or regurgitation\textsuperscript{19} that may warrant intervention at the time of cardiac surgery.

\textbf{Conclusion}

CMR has emerged as an extremely useful tool in evaluating known or suspected cardiac masses. In its ability to discriminate between true cardiac masses and pseudomasses, tissue characterization by CMR can distinguish a cardiac neoplasm (which generally will require excision) from other conditions, such as intracardiac thrombus, lipomatous hypertrophy or benign lipomas (which generally do not require excision). This can obviate the need for unnecessary surgical procedures. In the setting of biopsy-confirmed cardiac masses, CMR can assist with identifying the full extent of tumor involvement and associated features, such as pericardial effusion, coronary involvement, or cardiac valve involvement, and thus aid in surgical planning. This has led to its essential role in the evaluation and pre-surgical work up of cardiac masses.

\textbf{References}


THREE-DIMENSIONAL ECHOCARDIOGRAPHY IN THE ASSESSMENT OF CARDIAC TUMORS: THE ADDED VALUE OF THE EXTRA DIMENSION

Juan Carlos Plana, M.D.
The University of Texas MD Anderson Cancer Center, Houston, Texas

Introduction

Echocardiography is the most frequently used imaging modality in the assessment of cardiac tumors. Historically, this evaluation had been based on the analysis of 2-dimensional (2D) echocardiographic sectors of the heart. The information obtained from orthogonal tomographic planes from several acoustic windows was used in an attempt to mentally reconstruct a model of how the tumor would actually look in 3 dimensions and how it would relate to its adjacent structures.

New technology using matrix-array-transducers has permitted the development of real-time three-dimensional echocardiography (RT3DE), bringing cardiac imaging to a new dimension. Now it is possible to capture and analyze the entire volume of a cardiac tumor in a single cardiac cycle. This new imaging modality provides valuable clinical information that empowers echocardiographers with new levels of confidence in the diagnosis of heart disease.¹

This manuscript discusses the added value of this new technology in the echocardiographic assessment of cardiac tumors.

Echocardiographic Assessment of Cardiac Tumors

The complete echocardiographic evaluation of a cardiac tumor is summarized in Table 1. It starts with a thorough description of its location and relationship to adjacent structures. Tumors can be either intra- or extracardiac. The description should include the location and mechanism of implantation of the tumor to the heart (for instance, “pedunculated mass attached to the basal segment of the inferior wall through a long stalk”), the malignancy’s route of access to the heart (such as superior or inferior vena cava, pulmonary veins, direct access through a wall), and whether or not the tumor is primarily attached to the heart (Figure 1).

The characterization of the shape, longest dimensions, and ideal volume of the mass is essential. A description of the hemodynamic consequences of the mass should also be reported. The echocardiographer should then integrate all the information to generate a differential diagnosis.

The anatomic assessment of the tumor should be complemented with an accurate calculation of cardiac chamber dimensions, left ventricular volumes and ejection fraction (EF).

Once all this information is taken in consideration, a decision can be made as to the most appropriate treatment for the patient (chemotherapy, immune therapy, radiation or surgery).

The 3D Examination

Three acquisition modes are used with RT3DE in the evaluation of cardiac tumors: full volume, live 3D and 3D zoom (a smaller, magnified pyramidal data at a higher resolution).²

Unlike the other modes, the full-volume (wide angle) acquisition usually requires electrocardiographic gating. Depending on the vendor and the resolution desired, the gating requires somewhere between 2 to 6 cardiac cycles, as the dataset is compiled by merging the
narrower pyramidal scans during the consecutive heartbeats. To minimize reconstruction artifacts, data should be acquired during suspended respiration. Full-volume acquisitions can be obtained from the parasternal, apical 4-chamber, apical 2-chamber, and sub-costal views. However, the availability of a larger amount of data in these big pyramids of information comes at the expense of lower image resolution. Hence, imaging with narrow angles (live 3D) is recommended if high-resolution images of the cardiac mass are desired.

**Real Time 3D Evaluation of Cardiac Tumors**

In our institution, the evaluation of a cardiac tumor using transthoracic RT3DE includes, at the minimum, performing a full-volume acquisition in the parasternal long axis and in the apical 4-chamber views. If the mass in question is located in the right atrium, a full-volume acquisition and live 3D images are obtained from a modified right ventricular inflow view and the subcostal window.

Live 3D images of the right parasternal and supraclavicular windows have been used in the echocardiographic assessment of thrombus in the innominate vein and the superior vena cava (SVC). If the clinical question is not successfully answered by the transthoracic RT3DE, a real-time 3D matrix array transesophageal echocardiogram (3D-MTEE) is performed. In addition to the 2D images obtained following our lab protocol, a full-volume acquisition of the left ventricle (LV) is obtained. Depending upon the location of the mass in question, 3D-zoom images are captured from the mitral valve (MV), the tricuspid valve (TV), the left atrial appendage (LAA), the left upper pulmonary vein (LUPV), and the interatrial septum (IAS). Full-volume and live 3D acquisitions in the bicaval view are useful in characterizing masses in the SVC, inferior vena cava (IVC), inter-atrial septum, and right atrium.
Differential Diagnosis

The differential diagnosis of intracardiac masses includes tumors (benign, malignant or metastatic), normal structures or their variants, embryonic remnants, thrombi, masses associated with the presence of cardiomyopathy (apical hypertrophic cardiomyopathy, noncompaction cardiomyopathy and hypereosinophilic syndrome) and masses, or complications associated with device implantation (Table 1).

Primary tumors are far less common than metastatic tumors in the heart, occurring in at least 7 out of 10,000 people (Figure 2).4 Benign primary cardiac tumors occur more frequently than malignant ones. The most common cardiac tumor is a myxoma. In a large, single-institution series of primary cardiac tumors at the University of Minnesota, 42% were cardiac myxomas and 16% were malignant tumors (sarcomas).5

Added Value of RT3D Echocardiography in the Assessment of Intra-Cardiac Benign, Malignant, and Metastatic Cardiac Tumors

Unlimited slicing and cropping

Once a full-volume 3D data set is acquired, it can be sliced and cropped in many different ways. This allows manipulation of images in space, obtaining views and planes, and aligning structures in ways that were impossible to get with 2D imaging.

This feature is particularly useful in the way in which echocardiography is practiced in the United States, where images are captured by a sonographer and interpreted later by a physician. The availability of the full-volume acquisition allows the echocardiographer to slice and crop the heart in as many ways as required to obtain a comprehensive tomographic evaluation of the mass (Figure 3). The full volume allows for a detailed description of the location, shape, attaching interface and relationship to adjacent structures of the cardiac mass (Figure 4). In addition, it is well known that 2D echocardiography is very operator dependant. RT3DE, on the other hand, permits acquisition of the cardiac mass images with less dependence on the operator’s
skill level. It is also important to realize that the availability of all of the information in just one capture can significantly reduce the time needed to fully characterize the cardiac tumor.

**Evaluation of the size of the cardiac mass**

Echocardiography is the method of choice to establish the diagnosis and prognosis of cardiac masses, whether they are thrombi, vegetations or tumors. Maximum diameter measurements from 2D echocardiography are routinely used to determine mass size. The size of an intracardiac mass has important clinical implications in predicting embolic events, congestive heart failure, and death, and as an efficacy assessment after treatment (anticoagulation, antibiotics and chemotherapy).

However, most masses are irregularly shaped, making it difficult to accurately image or select the largest diameter. Nanda et al. reported that 2D measurements from a transthoracic or a transesophageal study underestimates the true maximum diameter of irregularly shaped structures. In the case of cardiac masses, this can lead to a misrepresentation of the patient’s prognosis. RT3DE images the entire volume of a mass, which allows for accurate measurements in multiple planes. Asch et al. conducted a study testing the hypothesis that measurements of the maximum diameter of a mass by RT3DE are larger than those obtained by 2D. He reported that 2D transthoracic and 2D transesophageal consistently underestimated the maximum diameter by 24.6% (P<0.001) and 19.8% (P=0.01), respectively, as compared to RT3DE. The measurements were fast and with excellent intraobserver and interobserver variability (better than 2D echocardiography). The authors suggested that RT3DE may be the technique of choice for the noninvasive evaluation of intracardiac mass size.

**Evaluating the composition of the tumor**

One of the critical aspects of characterizing a cardiac tumor is the evaluation of its composition (Figure 5). Mehmood et al. demonstrated in a preliminary study the superiority of live 3D over 2D transthoracic echocardiography (2D-TTE) in assessing left atrial (LA) tumors in patients with myxomas and hemangiomas. Because of the unique ability of live RT3DE to systematically section and view the contents of an intracardiac mass, LA myxomas in the patients studied could be more confidently diagnosed by noting isolated echoluent areas consistent with hemorrhage/necrosis in the tumor mass. In patients with hemangiomas, live RT3DE showed much more extensive and closely packed echolucencies with little solid tissue, as compared to a myxoma consistent with a highly vascularized tumor. Other studies have demonstrated that RT3DE provides a more comprehensive assessment of the inner structure of the mass that correlates better with pathologic findings (necrosis, hemorrhage, cystic areas or fibrotic bands).

**Unparalleled level of anatomic detail**

Recent advances in ultrasound transducer technology have lead to the miniaturization of matrix array transducers, allowing their incorporation into the tip of an esophageal probe and, in turn, the ability to perform 3D-MTEE.

Sugeng et al. recently published the initial experience of her group using 3D-MTEE. They tested the feasibility and clinical utility of this new technology in the imaging of different cardiac structures, including mitral, aortic, and tricuspid valves, interatrial septum, LA appendage, and pulmonic veins. The percentage of

---

**Figure 5.** (A and B) Orthogonal views from live 3D-MTEE obtained at 0 degrees from a 61-year-old male with a metastatic thymoma. The mass appears homogenously echodense in frame A. When the mass is cropped from the orthogonal plane, it appears to have a cystic component as well.
patients in whom each cardiac structure was assigned an optimal score of 2 was 85%–91% of all scallops for both mitral valve leaflets, 84% of the interatrial septum, 86% of the left atrial appendage, and 77% of the left ventricular endocardium.11

One of the biggest limitations of assessing cardiac masses with 2D echocardiography is the possibility of actually “missing” the mass during the evaluation. As mentioned before, orthogonal planes are used to evaluate cardiac masses; if the mass happens to be anatomically located in an area between the imaging planes, it would not become apparent during the examination.

3D-MTEE has allowed us to acquire and analyze the full volume of 3-dimensional structures with a new level of unparalleled anatomic detail (Figure 6). We are now able to really understand the 3D nature of the left atrial appendage and its unique anatomic relationship with the ridge and the left upper pulmonary vein. With this new technology, we are also able to visualize the foramen ovale of the interatrial septum from the left or the right atrium, taking advantage of its posterior anatomic location.

**Evaluation of associated abnormalities**

Real-time 3D echocardiography can also enhance the ability of the echocardiographer to detect associated abnormalities and conditions that predispose to the development of a mass, such as an LV apical aneurysm or rheumatic mitral valve disease.12-15

**Added value in the evaluation of embryonic remnants and normal variants**

Normal cardiac structures (moderator band or false chords) or their variants (accessory papillary muscles) can be confused with cardiac tumors in a 2D-TTE. In addressing these questions, we find it useful to obtain a full-volume acquisition and crop the structure to understand its 3D relationship. We also receive frequent referrals to our laboratory to characterize masses in the right atrium, where the differential diagnosis includes a normal structure (prominent IVC ridge or a crista terminalis), embryonic remnants (prominent eustachian valve or a Chiari network), a thrombus, a tumor or a tumor-thrombus arising from the IVC.16-18 The ability

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure6}
\caption{Cropped full-volume acquisition obtained from a 3D-MTEE at 90 degrees from the patient with metastatic thymoma mentioned in Figure 5. The tumor completely obliterates the superior vena cava (SVC) and extends through the right atrium into the tricuspid valve annulus. Please note the mass is heterogeneous. The mass appears uniformly echodense in the SVC and at the cavo-atrial junction, and appears cystic as it gets close to the tricuspid valve annulus.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure7}
\caption{(A) Full-volume acquisition obtained from the short axis view of the patient with osteosarcoma shown in Figures 2 and 3. The mass was not amenable for surgical resection as it involved the aortic root. (B) Full-volume acquisition obtained from the apical 4-chamber view of a 60-year-old male with metastatic squamous cell carcinoma of the penis. The mass was not thought to involve the tricuspid valve after 2D-TTE evaluation; however, follow-up RT3DE revealed involvement of the tricuspid valve.}
\end{figure}
to use the live-3D mode to see these 3D structures in motion has in many instances allowed us to answer questions without conducting more invasive tests such as a 3D-MTEE.

**Surgical planning**

Muller et al. conducted a study evaluating the value of 3D-MTEE as an adjunct to conventional 2D imaging in preoperative evaluation of cardiac masses. In 37% of the patients, 3D-MTEE revealed 1 or more items of additional information regarding type and site of attachment, surface features, and spatial relationship to surrounding structures. They estimated that in at least 18% of all intracardiac masses, 3D-MTEE can be expected to deliver supplementary information. In 6 of their patients, the additional findings led to decisions deviating from those made on the basis of 2D-TEE. The authors concluded that the information revealed by 3D imaging facilitated therapeutic decision-making, especially the choice of an optimal surgical access prior to removing the intracardiac mass (Figure 7).19

Sugeng et al. demonstrated that 3D-MTEE consistently provided excellent quality, volume-rendered images of mitral valve components, including anterior and posterior leaflets as well as the annulus and subvalvular structures. This finding suggests that 3D-MTEE may become one of the modalities of choice to assess this valve during preoperative planning of mitral valve surgery, including the resection of tumors from the valve.11,20 Le Tourneau et al. reported on the use of live 3D-MTEE in assessing tumors of the aortic valve (papillary fibroelastomas). In their opinion, the use of this technology improved their operative planning.21 However, the visualization of the aortic valve by 3D-MTEE appeared to be more challenging, as it is an anteriorly located cardiac structure; optimal visualization of the aorta from both the aorta and the LV perspectives was possible only in 18%–22% of the patients.11 3D-MTEE has also been used to assess neoplasms of the pulmonic valve.22

**Visualization of the true apex and calculation of LV volumes and ejection fraction**

Besides a thorough evaluation of cardiac tumors and their hemodynamic consequences, it is essential to be accurate in the reporting of LV ejection fraction. The mainstay in the treatment of cardiac sarcomas continues to be anthracycline-based regimens, which are known to cause cardiac toxicity in the form of systolic dysfunction. Hence, the accurate calculation of volumes and ejection fraction is essential in the initial and follow-up evaluations of these patients.

A variety of methods are available for calculation of EF using 2D echocardiography. Unfortunately, they all have 2 big limitations: they are based on geometrical models that have not considered the architecture of an abnormal sick heart, and they are strongly affected by foreshortening of the LV cavity by the tomographic plane of the 2D image. RT3DE has emerged as a solution to these problems. The ability to capture a full-volume acquisition of the LV allows for accurate identification of the true apex of the heart. An algorithm based on the detection of the endocardial border in turn allows for direct quantification of LV volumes without multiplane tracing or geometric modeling.

Jacobs et al. compared 2D and 3D echocardiography against cardiac magnetic resonance imaging (CMR) in their ability to accurately calculate end diastolic volume, end systolic volume and ejection fraction. RT3DE measurements correlated highly with similar measurements by CMR (r = 0.96, 0.97, and 0.93 for EDV, ESV, and EF respectively).23 The small underestimation of volumes, as compared to CMR, will hopefully be reduced as we gain experience with this new technique and learn to trace the endocardium underneath the trabeculations and not on top of them.1

More recently, contrast has been used to enhance RT3DE images. Contrast enhancement was found not only to improve the accuracy and reproducibility of LV volume measurements in patients with poor image quality, but also to enhance the assessment of regional wall motion from RT3DE datasets. The authors found that, with the use of selective dual triggering to minimize bubble destruction by ultrasound energy, contrast enhancement increased the accuracy of RT3DE-based analysis of regional LV function against CMR reference, and its reproducibility to levels similar to those noted in patients with optimal imaging quality.24 The improved accuracy and reproducibility of RT3DE-based LV volumes and EF measurements are of vital importance to patients with cardiac tumors, since clinical decision-making relies completely in this measurement. In the study mentioned above by Jacobs, there was evidence of a wider limit of agreement for EDV, ESV, and EF (29 mL, 24 mL, and 9.5%) for 2D-TTE, as compared to RT3DE (17mL, 16 mL and 6.4%).23 This means that when using 2D-TTE echocardiography, the ejection fraction can be potentially miscalculated by 9.5 points.

In our institution, anthracyclines are discontinued if patients have a symptomatic drop of more than 5% of their ejection fraction below 50% or an asymptomatic drop of more than 10% of their initial EF. The miscalculation of the ejection fraction by 2D-TTE can lead to the decision by the oncologist to erroneously stop the anthracycline-based regimen, due to concern for a toxic-
ity that is actually not occurring and is solely the result of the inherent limitations of the technology used.

**Conclusions**

Real-time 3D transthoracic and transophageal echocardiography have greatly improved the echocardiographic detection and evaluation of cardiac masses and tumors. A detailed characterization of the mass size, composition, location and relationship to adjacent structures, in conjunction with an accurate assessment of LV volumes and ejection fraction, empowers the echocardiographer with a new level of confidence in the diagnosis and surgical planning of the patient with a cardiac mass.

**References**


Introduction

Tumors of the heart are very uncommon and can occur as primary or secondary metastatic tumors. Metastatic tumors are over 40 times more common than primary cardiac tumors. Primary tumors of the heart are benign in 75% of cases and malignant in 25%.1, 2 We first reported our institutional experience with all primary cardiac tumors in 2003.3 Of the 85 patients seen, 17 had malignant tumors (20%) and 68 had benign tumors (80%). These benign tumors and our subsequent experience form the basis of this report.

Historical Review of Cardiac Tumors

In 1559, Realdo Colombo became the first physician to report a primary benign cardiac tumor.4-6 An atrial tumor that was thought to cause valvular obstruction was described in 1809 by Alden Allen Burns of England.7 The first series of cardiac tumors that matched what we now recognize as myxoma was noted in 1845 by King.8 In 1931, Yates reported on 9 cases of primary cardiac tumor and established a classification system similar to today’s.9 All of these tumors were postmortem reports, with the first antemortem diagnosis reported in 1934 of a cardiac sarcoma with abnormal electrocardiogram and a metastatic lymph node that allowed tissue diagnosis.10 Surgical intervention with excision of cardiac tumors was uncommon; the first reported success, described by Beck in 1936, involved removal of a teratoma external to the right ventricle.11 In 1951, Mauer reported the successful removal of a left ventricular lipoma.12

This paucity of surgical success changed because of 2 events: 1) the introduction of cardiopulmonary bypass (CPB) in 1953 by John Gibbon, allowing a safe and reproducible entry into the cardiac chambers, and 2) the introduction of echocardiography, allowing easy, noninvasive imaging of intracardiac structures and masses. Bhanson made an early attempt at removing an intracardiac right atrial myxoma in 1952 by using caval inflow occlusion, but the patient expired 24 days later.13 The first successful removal of an intracardiac tumor is generally credited to Crafoord of Sweden, who in 1954 removed a left atrial myxoma using CPB.14 Progress from this point was rapid; by 1960, 60 successful cases of atrial myxoma removal had been reported, with increasing use of echocardiography for detection and CPB for removal.15 Today, operation for benign cardiac tumors, although still uncommon, is not rare and is generally carried out with low morbidity and mortality. Even in our last reported series containing 68 patients with benign primary cardiac tumors, we

<table>
<thead>
<tr>
<th>No.</th>
<th>Type of benign primary cardiac tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>73</td>
<td>Myxomas</td>
</tr>
<tr>
<td>11</td>
<td>Fibroelastoma</td>
</tr>
<tr>
<td>6</td>
<td>Paragangliomas</td>
</tr>
<tr>
<td>3</td>
<td>Hemangioma</td>
</tr>
<tr>
<td>1</td>
<td>Lipoma</td>
</tr>
<tr>
<td>1</td>
<td>Fibroma</td>
</tr>
<tr>
<td>1</td>
<td>Castleman’s tumor</td>
</tr>
</tbody>
</table>

Table 1. Number and types of benign primary cardiac tumors treated at the Methodist DeBakey Heart & Vascular Center.
had no hospital deaths, local recurrence or late death from tumor. Benign primary cardiac tumors have been categorized in the Atlas of Tumor Pathology by McAllister, with myxoma being most common (49%), followed by lipoma (19%), and papillary fibroelastoma (17%). Table 1 describes our current institutional experience, which includes 73 myxomas, 11 fibroelastomas, 6 paragangliomas, 3 hemangiomas, 1 lipoma, 1 fibroma, and 1 Castleman's tumor.

Types of Benign Tumors

Myxoma

Myxoma is by far the most common benign primary cardiac tumor. The peak incidence for myxoma is between the third and sixth decades of life, and 94% of these tumors are solitary. Although they can occur in any of the cardiac chambers, about three-quarters of these myxomas occur in the left atrium, 10%–20% in the right atrium, and the rest equally divided between left and right ventricles. These tumors tend not to be associated with other abnormalities and rarely recur. There is a familial occurrence of myxoma in about 5% of patients that exhibits an autosomal dominance pattern of inheritance. These familial or complex myxomas tend to occur in younger patients, are more often multicentric (22%), and have a recurrence of 21%–67%. Carney's complex is an autosomal X-linked inheritance characterized by cardiac myxoma, cutaneous pigmented lentigines and primary pigmented adrenocortical disease with hypercortisolism.

Myxomas are usually round or oval tumors with a smooth or slightly lobulated surface that tend to be on short stalks and mobile. Myxomas arise from the endocardium and are derived from a subendocardial multipotential cell. The classic triad of clinical presentation is congestive heart failure (CHF) from intracardiac obstruction of blood flow, embolization, and constitutional symptoms of fever and weight loss. In our series, patients with benign tumors presented with CHF (29%), chest pain (16%), palpitations (15%), central nervous system embolus (12%), peripheral embolus (7%) and constitutional symptoms (13%). All tumors were diagnosed with transthoracic echocardiography. We routinely use intraoperative transesophageal echocardiography, which we believe eliminates the need for multicaardiac chamber exploration, to detect additional myxomas as has been suggested by some. We have had no hospital deaths or late-tumor recurrences in our series. Most myxomas can be approached through standard and often minimally invasive techniques, but some are so large or complex that more advanced approaches are necessary for success. One patient with a giant left atrial myxoma required cardiac autotransplant for removal because of its size and increased likelihood of malignancy (Figure 1A, intraoperative photo; Figure 1B, explanted tumor).

Another patient had a large left ventricular tumor that presented with right renal artery embolism and involved the ventricular side of the mitral valve and the left ventricular wall. This tumor also required a cardiac autotransplant technique for complete removal (Figure 2). Two months later, a right nephrectomy was performed, revealing viable myxoma tissue in the renal artery and infarcted kidney. Both of these patients did well and are currently free of disease.

Papillary Fibroelastoma

The next most common primary benign cardiac tumor is the papillary fibroelastoma (PF), which most often arises from the cardiac valves or the endocardium immediately adjacent to the valve. The most common location is the aortic valve, followed by the mitral valve. Atrioventricular and ventricular-arterial valves are equally involved. These tumors are usually asymptomatic but can embolize to the coronaries, causing a myocardial infarction, or to the brain, causing a stroke. It is difficult to accurately estimate the risk of embolism since the true denominator of patients with asymptomatic PF is not known. The classic description
of a papillary fibroelastoma is a tumor with frond-like projections that, when dropped into a basin of water, resembles a sea anemone. These tumors are generally single and solitary. Multiple tumors occur in about 7%–10% of cases. Cases with numerous multiple tumors numbering between 35 and 40 tumors in a single case have rarely been described.

Hypertrophic cardiomyopathy and previous cardiac surgery have been suggested to be associated with the occurrence of papillary fibroelastoma, yet the etiology of fibroelastoma is neither clear nor agreed upon. Some suggest that PF is a reactive lesion and not a true tumor; others have suggested a virus as the inciting factor in tumor growth, and still others suggest these may be hamartomas or organizing thrombi. It is our opinion that these represent true tumors, but their origin remains unclear. We have operated on 11 patients with PF with no hospital mortality. In these cases, all valves were spared, no valve replacement was necessary, and no patients have had a recurrence of tumor to our knowledge.

Most cases are approached through the wall of the atrium or across the ventricular arterial valve using standard techniques. One of our recent patients, who presented with repetitive transient ischemic attacks, had a PF in the intracavitary apex of his left ventricle (Figure 3A). Visualization and surgical approach to this tumor in the left ventricular apex was with thorascopic-assisted resection through the aortic valve orifice (Figure 3B). The patient did well and has had no further problems.

Cardiac paraganglioma

The next most common benign cardiac tumor in our experience has been the paraganglioma. We have operated on 6 cardiac paragangliomas, and this high incidence likely represents a referral pattern to our cardiac tumor center, rather than a true incidence of these uncommon tumors in the general population. Paragangliomas are rare tumors of neural crest origin. Cardiac paragangliomas account for only about 0.3% of all mediastinal neoplasms, and less than 50 cases were reported in the world literature as recently as 1992. Cardiac paragangliomas are highly vascular tumors that usually involve the roof of the left atrium but can occur around the intrapericardial aorta, or the right atrium and ventricle. They tend to parasitize the coronary blood flow, which gives them a soft, brownish appearance at surgery, and major hemorrhage is a substantial risk of biopsy or resection. The biologic activity that is usually seen in adrenal pheochromocytomas is rarely seen in cardiac paragangliomas. Clinical presentation can be incidental, or these tumors can present with heart failure due to obstruction of intracardiac or coronary blood flow. Coronary ostial obstruction can also lead to ischemia or conduction abnormalities, which also can occur if there is tumor encroachment on AV nodal tissue. “Hormonally active cardiac paraganglioma” is the terminology for biologically active cardiac paragangliomas rather than pheochromocytoma, which is confined to hormonally active adrenal paragangliomas. Of our 6 patients with cardiac paraganglioma, none of them had a hormonally active tumor. All tumors were very large and extremely vascular in nature, as seen in 1 patient’s angiogram depicted in Figure 4.

Overall in our series, 5 of the tumors occurred in the roof of the left atrium and involved most of the posterior and superior left atrial wall. One tumor involved the root of the aorta (Figure 5A, CT scan; Figure 5B,
Hemangioma

Cardiac hemangiomas are extremely rare primary tumors of the heart. They comprise 2%–5% of benign tumors and can arise from all layers of the heart, including endocardium, myocardium and epicardium. Most common locations in the heart include ventricles, atria and tricuspid valve. They can also arise from the pericardium, as we have described in 1 patient (TEE), (Figure 6). Although these tumors are extremely rare, they can cause outflow tract obstruction, valvular regurgitation, dysrhythmia and embolization. Diagnosis is made by TEE, CT scan, MRI or cardiac catheterization, which demonstrates a tumor blush. The natural history ranges from dormancy, accelerated growth or spontaneous regression. The mainstay of treatment includes operative intervention with resection or close observation, depending on symptomatology.

Lipoma

Lipoma is usually the third most common benign cardiac tumor seen, although we have only operated on 1 cardiac lipoma in our experience. Most of these do not cause symptoms, and have been resected in the past because of the uncertainty of the diagnosis and risk of leaving a malignant tumor. These tumors have a typical T1 and T2 signal on cardiac MRI that readily classifies them as a lipoma, and they are usually followed. If surgical resection is required (i.e., obstructive physiology), then routine cardiac surgical techniques can be employed with relatively low risk.

Fibroma

Fibromas are more common in children and are the second most common benign primary cardiac tumor in children, with most diagnosed by 2 years of age. Only about 100 of these unusual tumors have been reported in the literature. They are not inherited or associated with other disease. They are firm, nodular, gray-white tumors that tend to be large in size. When symptomatic, it is usually because the tumor impairs normal cardiac function by interfering with valve function or cardiac blood flow. Successful, complete surgical excision is curative, but can be difficult due to the large tumor size. Cardiac orthotopic transplantation has been used, but carries all the attendant problems and risks of heart transplantation.

Our 1 case was a 35-year-old female with a large left ventricular tumor that was resected, and the left ventricle was reconstructed with bovine pericardium (Figure 7). The patient did well for 4 days and then developed severe mitral regurgitation. She was...
returned to surgery and found to have disruption of her antero-lateral papillary muscle, which was close to the tumor resection margin. Mitral valve replacement was performed, and the patient has done well for the last 5 years without further problems.

**Castleman’s disease**

Castleman’s disease is a poorly understood, benign lymphoproliferative disease that may behave in a malignant fashion, depending on the structures it invades. Our 1 case was a patient whose tumor invaded the left ventricle and obstructed the proximal left anterior descending coronary artery. A coronary stent was placed, but a fistula later developed in communication with the tumor, leading to a large anterior-wall myocardial infarction. Due to the patient’s progressive heart failure, heart transplant was considered but rejected by the outside institution due to the possibility of the tumor being malignant. The patient was referred to our institution for tumor removal and possible heart transplant. The patient underwent tumor removal, which required CPB and bypass to the distal left anterior descending artery (Figure 8A). The degree of LV dysfunction eventually required placement of a Thoratec LVAD, and the patient actually recovered (Figure 8B). He received a heart transplant 2 months later and fully recovered. He is now 3 years post transplant and doing well.39

**Conclusion**

Benign cardiac tumors will continue to be present and will require standard cardiac surgery resection techniques. With the advancement in imaging technologies including MRI, patients will benefit from more focused surgical and diagnostic interventions. Those tumors with unusually aggressive behavior and large size may need more invasive strategies, including cardiac autotransplantation techniques. In summary, our significant experience with benign cardiac tumors would suggest that this condition is highly treatable, and surgical intervention is often necessary for cure.
References


27. Straus R, Merliss S. Primary tumor of the heart. Arch Pathol. 1945;39:74-8


LESS INVASIVE SURGERY FOR CARDIAC TUMORS
Mahesh Ramchandani, M.D.
Methodist DeBakey Heart & Vascular Center, Houston, Texas

Introduction
Cardiac surgery has undergone remarkable changes in the last 2 decades. Percutaneous coronary interventions (PCI) were becoming increasingly sophisticated, and patients preferred these to the more durable but invasive option of surgery. Spurred by this, the evolution towards less invasive techniques in cardiac surgery was led by coronary bypass surgery, which of course involved operating on the surface of the heart.

The development of less invasive techniques for valve surgery began in the mid-1990s, with advances made simultaneously in the United States and Europe. It became possible to operate inside the heart using techniques that spared the sternum partially or completely. The ability to access all the chambers of the heart with these techniques made it possible to deal with cardiac tumors as well.

Less invasive advances in other surgical specialties preceded cardiac surgery and provided some of the ideas that were applied to the special problems of operating in the thorax and, in particular, on the heart. It was clear that there were 2 main groups of patients who would benefit from less invasive techniques in cardiac surgery: 1) patients who want it, including those who are the breadwinners or the main care providers in a family, and 2) patients who need it, mainly elderly, steroid dependent, debilitated, or deconditioned patients.

This article offers an overview of the development of less invasive techniques in cardiac surgery and how they have been adapted for cardiac tumors.

Evolution of Techniques
The goals of minimal-access techniques have been to minimize morbidity, enable early return to a normal lifestyle, and improve cosmesis (Figures 1 and 2). These need to be achieved without compromising the efficacy of the procedure as compared to traditional techniques. It is generally felt that preserving the bony integrity of the thorax is an important aspect of these approaches. Partial sternotomy (upper or lower) and right minithoracotomy approaches are used, with the latter being favored by most surgeons for mitral or tricuspid valve surgery. Right-sided approaches offer excellent access to the atria, often better than that achieved via sternotomy (Figure 3).

Figure 1. Minimally invasive incision for left atrial myxoma — 1 month later.
There has been a stepwise evolution in less invasive cardiac surgical techniques. This has required refinements in access incisions, thoracoscopic visualization, instrumentation, and perfusion technology and strategies. A critical element in the refinement of these techniques has been a better understanding of the teamwork that is needed. Roles need to be clearly defined, and good communication is key. Every member of the team is empowered to question strategy and offer suggestions.

**Access — A Stepwise Evolution**

*Direct Vision* — smaller incisions via partial sternotomy or mini thoracotomy. Specially designed longer instruments were necessary. These techniques were first reported in the mid-1990s from U.S. and European centers. They were used for aortic and mitral surgery, and low rates of mortality (1%–3%) and morbidity were reported (Figures 4 and 5).

*Video Assisted* — techniques for video-assisted surgery were well developed in general for urologic and gynecologic surgery. These almost always involved the removal of a structure. Their adoption was delayed in cardiac surgery because most procedures involve a reconstructive element; valve repair or replacement and coronary anastomoses are examples. The removal of an intracardiac tumor requires, at the least, closure of the involved chamber of the heart. Two-dimensional scope visualization is augmented by the ability to look in through the incision and use scope illumination within the chest. Long shafted instruments continue to be refined and are essential in this method. This remains the most popular less invasive technique for cardiac surgery (Figure 6).
Video Directed — these are done entirely with video-scoptic visualization. The surgery is performed through a small access incision with the surgeon looking at a monitor. The benefits of tactile feedback are preserved. Three-dimensional scope visualization has improved the ease of using this technique.\(^7,10\)

Robotic (Telemanipulation) — the Da Vinci surgical robot (Intuitive Surgical) allows true port access surgery with shafted instruments that have “wrists” with many degrees of freedom. The robot is used most frequently for mitral valve surgery and could potentially be used for the removal of cardiac tumors (Figure 7).\(^8\)

Perfusion and Myocardial Protection Strategies

Intracardiac surgery requires placing the patient on cardiopulmonary bypass and usually achieving cardioplegic arrest. In less invasive surgery, this usually requires peripheral cannulation techniques. Femoral arterial and venous cannulation are used most frequently (Figure 8). Femoral arterial cannulation has the potential for catastrophic complications such as retrograde dissection, embolic complications and limb ischemia. Patients who are being considered for this must be screened for the presence of significant aortoiliac disease. There is a growing tendency for axillary arterial cannulation to be used because of the antegrade perfusion achieved and the very low risk of arterial complications. All techniques of peripheral cannulation have been made safer and easier by the development of cannulae that are thin-walled and strong. These allow smaller diameters to be used while still achieving good flow rates on cardiopulmonary bypass.

Cross clamping of the aorta is done using modified clamps that can be introduced through separate incisions. Chitwood, Cosgrove and Cygnet clamps are examples of these (Figure 9). Endoclamping of the aorta was made feasible by Heartport technology in the late 1990s and may lead to a lower embolic risk. Most surgeons employ external aortic clamping because of its simplicity.

Cardioplegic arrest may be obtained with antegrade and retrograde cardioplegia using cannulae designed for less invasive techniques.

Transesophageal echocardiography is indispensable for cannula placement. In addition, it provides precise delineation of the extent of the tumor and confirms complete removal.

---

Figure 7. (A and B) Da Vinci robot setup for minimal access to left atrium.

Figure 8. Femoral cannulation for cardiopulmonary bypass.
Cardiac Tumors

The most commonly encountered tumors are benign. Half of these are myxoma and the rest are mainly lipoma, fibroelastoma, rhabdomyoma and fibroma. About 75% of myxomas occur in the left atrium, 10%–20% in the right atrium, and the remainder in the ventricles (Figures 10 and 11).¹ ²

Malignant cardiac tumors are rare and are probably not suitable for less invasive techniques because of the extensive resection and reconstruction that may be required.⁴

Less invasive techniques provide excellent access to the atra and, through the mitral and tricuspid valves, to the ventricles. Resection of atrial tumors frequently involves a partial resection of the septum (Figure 12). Primary or patch reconstruction is easily performed via a right chest mini access incision — typically submammary and through the fourth intercostal space. Benign tumors in the ventricle may be removed by going through the atioventricular valves. They are usually attached to the free wall in the right ventricle and the posterior papillary muscle in the left ventricle (Figure 13). Recurrence following resection is rare.³

Conclusion

A myriad of small improvements in a few years have brought less invasive cardiac surgical techniques into the mainstream, and it can now be offered to most patients with benign cardiac tumors. Early recovery, return to work, and superior cosmetic results are the rule following these techniques.
References

Patient presenting with midscapular pain. Contrast enhanced MR reveals a penetrating aortic ulcer resulting in formation of an aortic pseudoaneurysm. Image courtesy of Dipan J. Shah, M.D.
IN THE NEWS

MULTI-MODALITY CARDIOVASCULAR IMAGING FOR THE CLINICIAN:
Update in Echocardiography, Nuclear, CT & CMR

SATURDAY–SUNDAY, OCTOBER 2–3, 2010
J.W. Marriott | Houston, Texas

Course Director: William A. Zoghbi, M.D., FASE, FAHA, FACC
Course Co-Directors: John J. Mahmarian, M.D., FACC, FASNC
Miguel A. Quiñones, M.D., FACC
Dipan J. Shah, M.D., FACC

methodisthealth.com/CVImaging

In cosponsorship with

Accolades
Dr. Basel Ramlawi, cardiac surgeon in the Methodist DeBakey Heart & Vascular Center, was named to the Houston Business Journal’s 40 under 40 list, which identifies 40 young leaders “who excel in their industries, are respected business leaders and show dynamic leadership in their community.”
HEALTH CARE REFORM: A POETICAL PLEA
William L. Winters Jr., M.D.
Methodist DeBakey Heart & Vascular Center, Houston, Texas

Many people decreed it to be
That health care as anyone could see
Needed reforming
A sort of reborning
To avoid a financial melee.

Major problems were quite visible
That made most people miserable
Like cost and access
Tricky insurance and malpractice
But each issue quite addressable.

Then there’s the matter of waste
Not curable just by cut and paste
So look for the source
Not obvious of course
That may require patience not haste.

Because some politicians just don’t get it
Their constituents to their great credit
Rose up en masse
Like weeds in the grass
To head off a worsening debit.

So as more people saw through it all
And tea parties slowed it to a stall
Let’s think this thing through
So as not to fall into
An uncontrolled financial freefall.

Please look at the problems out there
Try to show us you really do care
Take it step by step
With an eye on the debt
To find benefits that we all may share.

Rather than shake our foundations
Out of sheer and utter frustrations
Take a look at the parts
Readjust it with smarts
Then stand back to laud the creation.

With that thought in mind
Craft reforms of the kind
That look at the mix
To accomplish a fix
To solve this unending old bind.

As a doctor, I want you to know
I believe our profession would show
More trust in the end
If politicians would bend
And think like that sleuth, Hercule Poirot.

He achieved all manner of fame
By listening to all sides of the game
Then melding the ideas
For everyone to see as
A solution that all can acclaim.

So I think we all would agree
That such health care reform should be
An open forum for all
Young, old, short, and tall
And quite transparent for all to see.

It’s not necessary for all to agree
Expect different opinions to be
Stumbling blocks for some
A trial to overcome
In route to the game-winning tee.

I believe the best leaders today
Are those with attitudes that say
We’ll do it somehow
With grace and a bow
The consensus we’ll hail with hooray!

Having said all this with vigor and vim
Midst a strong wish for us all to win
To the parties in power
No more of those glowers
We will see if you sink or you swim.

If swimming were to happen
indeed
We’d be more than happy to read
That patients come first
Best news, not the worst
That our leaders did, in fact, take the lead.

It just goes to show you all
Common sense wins over a brawl
So it comes as a plea
From my colleagues and me
Make it work for the very long haul.
Overview

Primary cardiac tumors are very unusual, with an autopsy incidence of 0.001 to 0.003%. In practical terms, primary cardiac tumors are seen in about 1 in every 500 cardiac surgical cases. Of these tumors, about 75% are benign and 25% are malignant. Three-quarters (75%) of the malignant tumors are sarcomas, which will be the topic in our malignant primary cardiac tumor section.

This rarity leads to few institutions and even fewer individual surgeons having accumulated any substantial experience. Treated without surgical resection, the prognosis for primary cardiac sarcoma is dismal.

In a 2007 literature review spanning from 1973 to 2006, 117 cases of primary cardiac sarcoma were identified. In the most common group, angiosarcoma, the outcome without surgical resection and with medical therapy alone showed 90% of patients dead within 9 to 12 months. The Mayo Clinic published their 32-year experience, during which 34 patients had surgery for primary cardiac sarcomas with a median survival time of 12 months. A combined series from the Texas Heart Institute and the MD Anderson Cancer Center that spanned 26 years found 21 patients who had surgery for primary cardiac malignancy with a survival of 14% at 2 years. These institutions and almost all others in the literature have grouped all primary cardiac sarcomas together for analysis. We have also used this approach in our earlier analysis, where we looked at 27 surgical resections for primary cardiac sarcoma over a 16-year period using a multimodality approach, with a 1-year survival of 80.9% and a 2-year survival of 61.9%.

In this article, we separate the primary cardiac sarcomas into groups based on anatomic location rather than histology, but continue to analyze the data as a single group. Our current experience includes 83 primary cardiac sarcomas, of which 55 underwent surgical resection.

Our increasing referral pattern for primary cardiac sarcoma, combined with this background information from the medical literature, led our group working with our colleagues at MD Anderson Cancer Center to engage in a systematic study of primary cardiac sarcoma in an effort to better define this deadly and difficult disease, establish therapies, and continue to improve outcomes for these often very young patients. We proposed a classification system based on anatomic location rather than cell type, as is often done, because we have found that histology does not greatly affect treatment or prognosis, whereas anatomic location does. Our classification system divides primary cardiac sarcoma into right heart sarcomas, left heart sarcomas, and pulmonary artery sarcomas, and these are the categories we use in our discussions of these tumors to follow.

These tumors have no well-recognized predisposing factors, occur with an average presentation age of 40 years, and have no sex predilection. Symptoms depend on the anatomic location and extent of the tumor. Clinical presentation is generally due to intracardiac obstruction to blood flow with heart failure symptoms, local invasion with pericardial effusion or arrhythmia, embolism, or systemic symptoms of dyspnea, fever, malaise, and/or weight loss.

Tumors of vascular origin, angiosarcomas, are most commonly seen, but bone, muscle, neurogenic, and soft tissue cell lines occur as primary cardiac sarcomas. In the series reported by Putnam, angiosarcoma occurred in 37% of cases, malignant fibrous histiocytoma in 24%, leiomyosarcoma in 9%, rhabdomyosarcoma in 7%, unclassified in 7%, and in another category 16% of the time. Malignant fibrous histiocytoma is a diagnosis less often used by our pathologists today, and a recent pathologic review of 24 of our cases showed angiosarcoma in 10 (42%), unclassified in 9 (37%), 3 synovial cell sarcomas, and 2 leiomyosarcomas. We have found angiosarcoma to be the most frequent right
heart tumor, and unclassified, many of which were previously labeled malignant fibrous histiocytoma, to be the most frequent left heart sarcoma. Pulmonary artery sarcoma in our experience has a widely varied histology. Between 33%–80% of patients are reported to have metastatic disease on presentation.\textsuperscript{11, 12} Gender, age and histologic type do not appear to be correlated with prognosis.\textsuperscript{11, 13} Tumor grading, however, has generally been shown to correlate with prognosis. In all studies, surgical resection remains the mainstay of therapy and is vital for the hope of success.

Right heart sarcomas tend to metastasize early and are very bulky and infiltrative. Their bulk can occupy much of the right atrium but grows largely in an outward pattern, avoiding heart failure based on obstruction to flow until the very late stages of the disease. This usually allows the use of neoadjuvant chemotherapy with these tumors in an attempt to shrink the tumor bulk and sterilize the infiltrating edges, which increases the chance of obtaining a resection with microscopically negative margins for malignancy or an R0 resection for the patient. Because benign right heart tumors are less common than left-sided benign tumors and are often very large, we frequently see these tumors early before surgical resection has been attempted because of the suspicion of malignancy.

Left heart sarcomas tend to be more solid and less infiltrative than right heart sarcomas, and metastasize later in our experience. These are most often in the left atrium and tend to grow into the left atrium, obstructing blood flow and often presenting with significant and life-threatening heart failure. Neoadjuvant chemotherapy can be less often used because of this presentation. All left atrial sarcomas we have seen were first operated on elsewhere, with a presumed diagnosis of myxoma, and grew back rapidly, most likely from incomplete resection before referral to our institution.

Figure 1 shows a schematic view of the difference in survival from medical therapy alone,\textsuperscript{9} to the survival in the early surgical series of Putnam from MD Anderson Cancer Center and the Texas Heart Institute,\textsuperscript{5} to our last publication from the Methodist DeBakey Heart & Vascular Center and MD Anderson Cancer Center using a multimodality approach.\textsuperscript{6} Although this remains a
difficult and deadly disease, progress has been and is being made. In the following articles, we will discuss in detail our experience with this disease, including current outcomes and improvements over past approaches, current treatment protocols, and our vision for future improvements.

References

Introduction

Primary heart tumors are rare, and malignant primary heart tumors are only a small subset of these. Most primary malignant heart tumors are sarcomas arising from the cells of the structural elements of the heart such as blood vessels, muscle, connective tissue, fat and even bone. Unlike most malignancies, where cell type often dictates treatment choices and prognosis and is used for classification, the histology in primary cardiac sarcoma plays little role in determining therapeutic options or prognosis. We have found that anatomic location within the heart is the major determining factor in clinical presentation, treatment options and prognosis in cardiac sarcoma. Therefore, we accordingly classify primary cardiac sarcomas into right heart sarcomas, left heart sarcomas and pulmonary artery (PA) sarcomas.

Since the first autopsy report of a primary PA sarcoma in 1923, there have been fewer than 250 cases reported in the English literature. Most of these reports have been single autopsy or case reports, and patient prognosis has generally been dismal. Since few institutions and even fewer individual physicians acquire much exposure to this disease, the diagnostic and treatment approaches have remained unresolved. Our cardiac sarcoma group working at the Methodist DeBakey Heart & Vascular Center and the MD Anderson Cancer Center has undertaken a systematic study of this disease, and operated on 9 patients using a radical resection with curative intent and multimodality approach. Based on this work, we have suggested a diagnostic strategy, treatment approach and staging system for primary PA sarcoma. A substantial improvement in patient survival over historical controls has also been demonstrated and will be discussed in this review.

Discussion

Cardiac sarcomas are rare tumors, and primary PA sarcoma is an unusual subset of these already unusual entities. The first case of primary PA sarcoma was reported in an autopsy study in 1923,1 and fewer than 250 cases have been reported in English literature since then. We recently published our series and treatment approach to primary PA sarcoma, and reviewed the current literature to establish a treatment plan and examine our outcomes in this difficult group of patients.2 In this report, we did a literature search to identify cases of surgical resection of primary PA sarcoma for historical controls, and reviewed the databases of the MDHVC and the MD Anderson Cancer Center to identify all cases of primary PA sarcoma having surgical resection at our institutions. In our search, we selected all cases that adequately described the surgical technique and survival to allow analysis. This yielded 60 patients, with 25 having complete resections, 24 having debulking resections or endarterectomy, and 11 having a poorly reported resection technique but good survival data. We identified 8 patients who underwent surgery for PA sarcoma at our institutions, and we included an additional patient who recently was operated on by our group for primary PA sarcoma. These 9 cases serve as the basis for this review.

The clinical presentation of patients with PA sarcoma can be easily confused with other entities that affect pulmonary circulation. Symptoms almost always
included cough and dyspnea, with hemoptysis and chest pain occurring less frequently, in a picture that may mimic pulmonary embolus (PE). Constitutional symptoms were often present, including fever, anemia, and weight loss, and are more consistent with malignancy than PE. These mass lesions will not decrease in size with anticoagulation but will continue to grow. In our recently reported series, all 8 patients had cough and dyspnea, 7 of 8 had constitutional symptoms of fever and anemia, and only 1 of 8 had chest pain. A lack of history or findings of deep venous thrombosis should also help guide the clinician towards consideration of malignancy.

The difficulty in establishing the diagnosis of PA sarcoma is highlighted by the fact that the typical duration between symptom onset and diagnosis ranges from 3 to 12 months. The most common initial misdiagnosis is PE, but pulmonary hypertension, congenital pulmonary stenosis, fibrosing mediastinitis and lung tumor have all been confused with PA sarcoma. By the time the correct definitive diagnosis is achieved, about 50% of these patients will have metastatic lung involvement. These patients usually present to the clinician with right heart failure and pulmonary hypertension of often substantial levels that requires urgent treatment. Since this presentation is not infrequently confused with chronic pulmonary emboli, pulmonary artery thromboendarterectomy is often recommended. While tumor endarterectomy can be achieved once sarcoma is found rather than thrombosis, and will relieve pulmonary artery obstruction, it is associated with rapid tumor regrowth and poor survival outcomes. Therefore, establishing the correct diagnosis to allow appropriate planning for radical surgical resection is important if we are to increase patient survival in this disease.

Establishing the correct diagnosis relies on appropriate imaging. Imaging has proven crucial in our approach to these tumors, and we have used a variety of imaging techniques in these cases to ascertain the correct diagnosis, since tissue diagnosis prior to open surgery has been difficult to obtain. We have used computed tomography angiography (CTA), trans-thoracic echocardiography (TTE), transesophageal echocardiography (TEE), magnetic resonance imaging (MRI) and positron emission tomography (PET) scans to characterize these mass lesions. Since this entity mimics the much more common PE, a CT scan with PE protocol is often the initial diagnostic test chosen. All of our patients who had CT scans showed hyperdense lesions and actual distention of the artery lumen in areas where the tumor completely filled the artery (Figure 1). The extent of these tumors and dilatation of the artery itself should steer the clinician away from a diagnosis of chronic PE and towards 1 of PA sarcoma. MRI scan has been particularly useful in identifying PA sarcoma as the tumor enhances with gadolinium contrast more than thrombus. MRI scan is also helpful in the follow-up of these patients if new pulmonary artery masses appear after surgery, to differentiate from post-operative hematoma or clot. The degree of tumor differentiation, myxoid matrix content and thrombus content has been shown to correlate with the degree of gadolinium enhancement. The hyperactivity of these tumors make them PET avid and will help the clinician differentiate tumor from thrombus as well as look for extra cardiac disease (Figure 2).

Accurate preoperative imaging is crucial to allow appropriate surgical planning, since tumor debulking or endarterectomy is associated with poor survival and complete radical resection improves the patient’s prog-
nosis. Unfortunately, obtaining a tissue diagnosis is difficult prior to surgery and most often is not obtained until the time of surgery. We have attempted transvenous catheter biopsy of these tumors but have had limited success in obtaining the tissue needed. We currently employ a treatment algorithm shown in Figure 3 for these patients. Patients considered for operation are those with adequate cardiopulmonary reserve, adequate lung function if pneumonectomy is contemplated, disease limited to the main pulmonary artery system — or, if outside this, manageable with chemotherapy — and surgical judgment of resectability. Although preoperative imaging is helpful, we have found that the ability to completely resect the tumor cannot be fully assessed prior to surgery. We have also proposed our own staging system for PA sarcoma shown in Table 1.

Anatomically, these tumors tend to arise from the dorsal surface of the main pulmonary artery just beyond the pulmonary valve. They form from multipotential mesenchymal cells from the muscle remnant of the bulbus cordis in the intimal and subintimal surfaces. The tumor then tends to grow distally along the artery, rarely penetrating the actual wall of the artery but rather distending it, which is an important point in the surgical resection. Distal extension can go to the lung parenchyma itself as emboli, infarction or metastasis. Although some have shown survival differences based on cell histology in limited cases, that has not been our experience in cardiac sarcoma in general. This is further complicated by the wide variety of histology we have seen in these patients, with 1 unclassified sarcoma, 2 leiomyosarcomas, 2 angiosarcomas, 1 high-grade sarcoma and 1 spindle cell sarcoma being seen in our published series as well as angiosarcoma in our recently added patient.

Pulmonary artery sarcoma is a very aggressive disease, and survival after diagnosis is often discussed as weeks or months rather than years. The results of chemotherapy alone have been disappointing, with numerous studies reporting a lack of tumor response. The response to chemotherapy alone has been so poor that a recent paper published from Japan claimed to be the first case of PA sarcoma responding to chemotherapy. The role of radiation therapy also is unsettled, although more applicable, due to tumor location above the myocardium. Surgical resection remains

Table 1. Staging system for primary pulmonary artery sarcoma.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I</td>
<td>Tumor limited to main pulmonary artery</td>
</tr>
<tr>
<td>Stage II</td>
<td>Tumor involving 1 lung and main pulmonary artery</td>
</tr>
<tr>
<td>Stage III</td>
<td>Bilateral lung involvement</td>
</tr>
<tr>
<td>Stage IV</td>
<td>Extrathoracic spread</td>
</tr>
</tbody>
</table>
the primary method of treatment in patients with PA sarcoma, and the only method shown to increase survival.\textsuperscript{25}

Although surgical resection is the mainstay of therapy for these patients, the techniques used have varied from local debulking to tumor endarterectomy to complete radical resection. We believe only complete radical resection allows a reasonable increase in survival and plan our surgery accordingly. Unfortunately, the ability to completely resect the tumor cannot always be determined from preoperative imaging studies, and can only be definitively assessed at surgery. Since exposure of the right main pulmonary artery may require division of the aorta and possibly the superior vena cava (SVC), we plan surgical cannulation for cardiopulmonary bypass accordingly. We use dual venous cannulation with direct SVC cannulation and normal inferior vena cava (IVC) cannulation via the right atrium. Arterial cannulation via the ascending aorta is normal. Both SVC and IVC are looped with tourniquets to isolate the right heart. Cardiopulmonary arrest is achieved with cold potassium blood cardioplegia. Fortunately, these tumors rarely penetrate the pulmonary artery wall, allowing reasonably easy mobilization. The main pulmonary artery has always been involved in our experience, and the pulmonary valve is involved about 30\% of the time.\textsuperscript{14} An assessment is made of the distal extent of the tumor into each main pulmonary artery. These arteries can be relatively easy to resect out to their first branch points on each side, and we resect as far as is necessary to go beyond the tumor, up to this branch point.

In cases where 1 pulmonary artery is relatively free and the other involved is all the way out into the lung and completely obstructing blood flow, we have chosen to do a pneumonectomy to completely remove the disease. When a pneumonectomy is performed, the pulmonary veins and main bronchus are dissected and divided before cardiopulmonary bypass (CPB) is instituted to avoid bleeding while heparinized. The branch PAs and main PA are mobilized, and then CPB is instituted and the involved main PA divided. In these cases, the involved lung had no blood flow, and hemodynamics were actually improved by pneumonectomy and removing the obstruction of the contralateral artery. Once each PA has been divided — or the lung, in the case of pneumonectomy — and its main PA removed, the main PA trunk can be assessed for pulmonary valve involvement. If the pulmonary valve is involved, the entire PA trunk must be removed and replaced by a pulmonary allograft. Removal of the entire PA trunk and replacement by an allograft is similar to its mobi-

\begin{figure}
\centering
\includegraphics[width=\textwidth]{cumulative_survival_probabilities.png}
\caption{Five-year cumulative survival probability in the current case series (dotted line) vs. a combined series (solid line) of all historical controls since 1990.}
\end{figure}
lization and replacement for a Ross procedure, which we have previously described. If the resection of the right and/or left main PA is limited, then the allograft branches may surmise to span the defect. When the extension had been too extensive for this, we used an appropriately sized Gore-Tex graft to go from distal right PA resection point to distal left PA resection point, and then implanted the pulmonary allograft into the side of this graft. Despite the extensive nature of the resection, separation from CPB had not been difficult since the surgery relieves the patient of the severe PA obstruction that is present preoperatively.

We had no hospital deaths in our reported series or in our additional case. Survival rates of primary cardiac sarcoma have been historically dismal and generally reported for all cardiac sarcomas as a group, rather than with the anatomic classification our group has put forth. Survival without surgical resection has been reported to be about 10% at 9 to 12 months. The Mayo Clinic has reported its experience over 34 years with 32 patients undergoing surgical resection for cardiac sarcoma with a median survival of 12 months. The combined series of the Texas Heart Institute and MD Anderson Cancer Center looked at 21 patients over 26 years who had surgical resection of cardiac sarcoma to yield a survival of 14% at 2 years. To confine our analysis to PA sarcoma only, we chose to use historical controls from the literature representing only PA sarcoma resections. Historical controls in this study had a mean survival of 18 months, and our series yielded a median survival of 71 months. The survival curve for both is shown in Figure 4. This improved survival clearly suggests that the surgeon should strive for complete resection, and that limited procedures such as tumor endarterectomy are less effective. All patients are offered adjuvant chemotherapy, but not all patients in this series had postoperative chemotherapy. To continue our study of this disease, we have standardized our diagnostic and treatment approach included in our institutional database and established an international registry of PA sarcoma.

Conclusion

We believe that we have substantially improved the survival rate of patients with pulmonary artery sarcoma due to our use of aggressive radical resection with curative intent and multimodality therapy. Of the 9 patients we have currently treated, 5 are still alive and 3 have no evidence of disease. We have proposed a standard treatment protocol and a staging system for pulmonary artery sarcoma. The continued and expanded use of multimodality biologic therapy, in addition to surgical resection, represents the greatest area for potential advances in this difficult disease.

References


RIGHT HEART SARCOMAS
Ara Vaporciyan, M.D., Michael J. Reardon, M.D.
The University of Texas MD Anderson Cancer Center, Houston, Texas
Methodist DeBakey Heart & Vascular Center, Houston, Texas

Introduction
Primary cardiac tumors are unusual, and primary cardiac sarcomas constitute a rare subset of these. In cardiac sarcoma, unlike many malignancies, the histologic cell type does appear to affect the treatment options or prognosis in a significant way. The presenting symptoms, treatment options and, indeed, prognosis are largely controlled by the tumor’s anatomic location.

We have proposed a classification system based on anatomic location that divides cardiac sarcoma into left heart, right heart and pulmonary artery sarcomas. In our experience, right heart sarcoma tends to be bulky, grow in a more exophitic manner, be more infiltrative, and metastasize earlier than left heart or pulmonary artery sarcoma. Right heart sarcoma also presents less often in congestive heart failure or with compromised hemodynamic status than left heart and pulmonary artery sarcoma, which are usually highly symptomatic at presentation. The prognosis for right heart sarcoma without surgery is dismal. Complete surgical resection remains the goal of therapy and the only treatment modality shown to increase survival. Complete surgical resection is complicated both by the bulky infiltrative nature of right heart sarcoma and the high incidence of metastatic disease at presentation.

The current approach of our cardiac sarcoma group to right heart sarcoma has been to begin neoadjuvant chemotherapy once a definitive tissue diagnosis of sarcoma is achieved. After 4 to 6 rounds of chemotherapy, the patient is considered for surgical resection. This standardized treatment has been approved in our IRB protocol: A Clinical Trial to Assess the Safety and Efficacy of a Novel Radical Tumor Resection Procedure used in conjunction with Neoadjuvant Chemotherapy to treat Malignant Primary Right Heart Cardiac Tumors — the ESPERO trial. This protocol is designed to compare our existing 24 index cases of surgical resection of right heart sarcoma using a nonstandardized treatment plan, with routine neoadjuvant chemotherapy, and a standardized treatment plan to see if the rate of microscopically complete resection can be improved from its current level of 33% and if this will improve patient survival. In this review, we will discuss the experience with right heart sarcoma.

Discussion
Primary cardiac tumors are unusual, with about 75% being benign and the other 25% malignant. Of the malignant tumors, about 75% are sarcomas. With primary cardiac sarcoma, we previously found that histology did not significantly influence surgical approach, clinical outcomes or survival. Our findings therefore led us to classify primary cardiac sarcoma according to anatomic location, which is the main determinant of clinical presentation and surgical approach. In our experience, right heart sarcomas tend to be bulky and exophitic, and do not usually cause heart failure from obstruction of intracardiac blood flow, as is common in left heart and pulmonary artery sarcomas. They also tend to present with metastatic disease at a higher early rate than left heart sarcomas. These characteristics allow us to approach right heart sarcomas with
neoadjuvant chemotherapy and treatment planning that can differ from left heart and pulmonary artery sarcomas.

Right heart sarcoma occurs in a young patient population. Our series had a mean age of 40.7 years and a range of 17 to 61 years. The signs and symptoms in right heart sarcoma are often nonspecific and constitutional, and early diagnosis often eludes the clinician.6 Pericardial effusion associated with nonspecific chest pain has been a common initial presentation in our experience. Despite the often large size of these tumors, heart failure is usually not a prominent feature until very late for right heart sarcomas. Multiple lung lesions later found to be metastatic angiosarcoma have been common at presentation.

The nonspecific nature of presenting the symptoms and young ages of these patients has led to a number of diagnostic tests for initial assessment. The 3 most common modalities employed initially in patients referred to our group were transthoracic echocardiography (TTE), computer tomography (CT) scan of the chest and standard chest X-ray. When we suspect cardiac sarcoma in a patient referred to our group without diagnosis, our initial diagnostic test has been TTE. This diagnostic modality rarely misses these large tumors. CT scan of the chest and abdomen is obtained in all patients, allowing better characterization of the extent of local tumor involvement than TTE. This local tumor involvement often can extend to the superior vena cava (SVC), aorta, pulmonary artery and other upper mediastinal structures. CT scan also allows a screen for metastatic disease that is common at presentation for right heart sarcoma, especially pulmonary metastatic disease. PET/CT scan is obtained in all patients at presentation. The level of 18-fluorodeoxyglucose (FDG) uptake or the standardized uptake valve (SUV) in the primary tumor will allow us to follow the response to neoadjuvant chemotherapy and screen for distant metastatic disease.

We have found the right coronary artery to be involved in about one-third of our cases, and we routinely perform cardiac catheterization with coronary arteriography to define the right coronary anatomy in case resection and replacement is needed. These diagnostic tests usually allow good definition of a large right heart mass and a strong suspicion of malignancies, but do not allow a definitive diagnosis. Unlike left atrial masses that are usually benign myxomas even when large, the majority of large right atrial masses are malignant. Definitive tissue diagnosis is crucial in order to rule out other disease processes, such as lymphoma, and to establish histologic proof of sarcoma prior to initiating neoadjuvant chemotherapy. If metastatic disease is present and amenable to biopsy, this allows tissue diagnosis without approaching the heart. Percutaneous approaches to biopsy the primary cardiac mass have not been routinely successful in our experience, and pose risks that our radiology department prefers to avoid. A subxiphoid pericardial window and tumor biopsy can often establish the diagnosis. When a pericardial window is unsuccessful, right anterior thoracotomy and sometimes even median sternotomy may be required to allow safe biopsy for definitive diagnosis.

We currently treat all biopsy-proven right heart sarcomas on our IRB based protocol, the ESPERO trial. All patients without overt heart failure that might force an early surgery are started on neoadjuvant chemotherapy, doxorubicin hydrochloride (75 mg/m²) and ifosfamide (106 mg/m²), in an attempt to cytoreduce the tumor bulk and sterilize the infiltrative, microscopic fingers

**Figure 1.** CT scan of large angiosarcoma filling right atrium and right ventricle, causing severe heart failure.

**Figure 2.** Surgical specimen of tumor seen in Figure 1.
of disease that tend to invade the margins of even wide resections. This was introduced because only about 33% of our right heart sarcoma resections had microscopically negative margins (R0 resection) in our initial data analysis. After every second round of chemotherapy, the imaging is repeated to assess tumor response. Our goal is to consider surgical resection when appropriate after either the fourth or sixth round of chemotherapy, based on tumor response. Tumors considered for surgical resection at this point are tumors without extra cardiac extension that appear to be anatomical candidates for complete resection. Patients whose tumors presented with limited metastatic disease have a good chance of further resection after chemotherapy. Tumors not responding well to chemotherapy are not considered candidates for surgery. Patients with widely metastatic disease are not considered candidates unless severe symptoms warrant palliative surgery (Figures 1 and 2). All patients are offered continuation of their chemotherapy after recovery from surgery.

In patients treated without surgical resection, the survival is only about 10% at 12 months. Complete surgical resection with negative margins has been shown to extend survival, and an R0 resection is always the goal. The margins for surgical resection should be wide to encompass all diseases, but they are limited by anatomic constraints of what cannot be replaced or reconstructed after resection. Resection can include the SVC, a short segment of inferior vena cava (IVC), the entire right atrium, the tricuspid valve, the right coronary artery, and up to about 30% of the right ventricular muscle mass if right ventricular function is normal to begin with. These tumors can be quite large, and based on the anatomic extent of tumor and the margins needed for resection, venous cannulation for cardiopulmonary bypass must be carefully planned and individualized to each patient (Figure 3).

We have routinely cannulated directly into the high SVC for upper-body drainage. Usually we can cannulate directly into the IVC at the diaphragm, but occasionally we need to use percutaneous femoral venous cannulation to allow exposure for complete inferior resection. Aortic cannulation is a standard fashion as it is distant from the tumor. The right atrium can be completely resected and replaced with bovine pericardium. If the resection involves the SVC or IVC, we use a vascular stapler to recreate the vein segment and leave the rest of the pericardium open to replace the atrium (Figure 4). The 1 area of real danger to the surgeon is where the right atrium meets the root of the aorta; overzealous resection here will take the surgeon into the fibrous skeleton of the heart, and this can be very difficult to repair.

The most common reason we have seen for a failed complete resection in patients referred to us has been the surgeon’s reluctance to resect the right coronary artery that can be involved by the tumor. Incomplete resection that leaves gross disease will only lead to rapid regrowth of the tumor, and should be avoided whenever possible. When we suspect right coronary artery involvement, we mobilize the right internal mammary artery at the beginning of the operation. If needed, it is simple to either clip distally and use as a pedicle graft, or divide proximally and use as a free graft as needed. The right ventricular wall can be replaced with Bovine pericardium or, if it is the area

Figure 3. Surgical view of large right heart sarcoma.

Figure 4. Recreating superior vena cava and right atrium with bovine pericardium.
along the anterior tricuspid valve where the right coronary was resected, it is simply advanced forward to the prosthetic tricuspid valve used for valve replacement (Figures 5 and 6).

We have preliminary data on 24 patients that we have operated on with right heart sarcoma. Of these, 22 of 24 (92%) were right atrial, and 2 of 24 (8%) were right ventricular. We had an equal distribution of 12 males and 12 females. The age range was 17 to 61 years of age with an average age of 40.7 years. Angiosarcoma was found in 18 of 24 (75%), synovial cell sarcoma in 3 of 24 (13%),

Figure 5. Completed repair.

Figure 6. (A) Right atrial sarcoma; (B) extent of resection; (C) tricuspid valve replacement; (D) reconstruction with bovine pericardium and right internal mammary artery graft.

Figure 7. Cumulative survival probability for all right heart sarcomas with surgical resection.
rhabdomyosarcoma in 1 of 24 (4%), undifferentiated sarcoma in 1 of 24 (4%), and leiomyosarcoma in 1 of 24 (4%). This is the initial data used to write our current IRB protocol for treating these tumors. In 16 of 24 (66.6%) patients, we had microscopic disease at the margin (R1 resection), and achieved an R0 resection with negative margins in only 8 of 24 (33.3%) patients. Hospital mortality was 8%, with 1 being from right heart failure and 1 from bleeding in a patient with extensive radiotherapy to the heart and mediastinum. Overall survival for our preliminary data is seen in Figure 7 and survival for R0 and R1 resections is seen in Figure 8. Although this remains a difficult disease, the survival with surgical resection far exceeds that of medical therapy alone, and we now have survivors of almost 10 years and still living. The hypothesis of our current protocol is that standard use of neoadjuvant chemotherapy will lead to a higher rate of R0 resection and hence a higher long-term survival, compared with this historical cohort.

**Conclusion**

Right heart sarcomas are unusual tumors that continue to pose difficult therapeutic problems to the clinician. Our initial data suggests reasonable surgical risk for resection and survival of up to 10 years, but the ability to obtain microscopically negative margins occurs only about 33% of the time. We have initiated a protocol-driven diagnosis and treatment of right heart sarcomas that incorporate routine neoadjuvant chemotherapy, and we hope to include 25 patients over 5 years. We hypothesize that this approach will let us increase our rate of complete resection with negative margins and further improve survival in this patient group.

**References**

LEFT HEART SARCOMAS
David C. Rice, M.D.; Michael J. Reardon, M.D.
The University of Texas MD Anderson Cancer Center, Houston, Texas
Methodist DeBakey Heart & Vascular Center, Houston, Texas

Introduction
Primary tumors of the heart are uncommon, with roughly 75% benign and 25% malignant. Most of the malignant tumors are sarcomas and historically have had a very poor prognosis. These tumors tend to occur in young patients with a mean age of 40 years. Making a diagnosis of cardiac sarcoma can be difficult due to its rarity and the nature of the symptoms. For left heart sarcomas, almost all patients are symptomatic by the time the diagnosis has been made. Symptoms are dependent on the location and the extent of the tumor and are not related to tumor histology; similarly, histologic cell type has not been found to be related to prognosis in most studies. We therefore previously proposed a classification system for primary cardiac sarcoma based on anatomic location, dividing primary cardiac sarcoma into right heart, pulmonary artery and left heart sarcomas.

Left heart sarcoma presents a technical anatomic challenge: the left atrium, being the posterior heart chamber, allows somewhat limited access using routine surgical approaches. The role of chemotherapy or radiotherapy remains unclear and unproven, leaving complete surgical resection as the only mode of therapy with a proven survival benefit. Our review of the published literature showed frequent local recurrence and poor long-term survival in left heart sarcomas. Our hypothesis was that the left atrium had limited anatomic accessibility for large complex resections and reconstructions, and this led cardiac surgeons to do a more limited tumor removal with an increased chance of local recurrence and a detrimental effect on survival. To address this technical challenge, our group introduced the surgical technique of cardiac explantation, ex vivo tumor resection, cardiac reconstruction, and subsequent cardiac reimplantation or cardiac autotransplantation for left heart sarcoma in an attempt to improve the completeness of local resection, decrease local recurrence, and extend patient survival. This review discusses the approach of the cardiac sarcoma group at the Methodist DeBakey Heart & Vascular Center and the MD Anderson Cancer Center to the diagnosis and treatment of left heart sarcoma, as well as our current patient outcomes.

Discussion
Cardiac tumors occur either as primary tumors or metastatic tumors, which are far more common. Primary tumors of the heart are quite rare, with about 25% being malignant and most of those being sarcomas. The clinical presentation of patients with primary left heart sarcoma depends on the anatomic location and extent of the tumor, and is not influenced by histology. The most common presenting symptoms for left heart sarcoma, in our experience, are shortness of breath and dyspnea on exertion — both consistent with congestive heart failure (CHF), and arising from the obstruction of intracardiac blood flow. Over half of our patients had NYHA-FC III or IV CHF symptoms at presentation. Arrhythmia from local invasion, pericardial effusion and embolization occur, but are less common with left heart sarcomas than right heart sarcomas.
Constitutional symptoms such as fever, malaise and weight loss are also common.\(^2\) The mean age of presentation is reported to be 40 years of age\(^3\), and is 38.5 years in our current series, with a range of 20 to 57 years old.

Because most patients present with symptoms of CHF or other symptoms suggesting a cardiac problem, transthoracic echocardiography is the most common initial diagnostic test. Transthoracic echocardiography (TTE) is widely available, rapid and noninvasive, and has a high ability to identify left-sided intracardiac masses. Most primary left heart sarcomas are reported to occur in the left atrium, and this is supported by our experience in which 22 of 24 (92%) occurred in the left atrium and 2 of 24 (8%) occurred in the left ventricle. Myxoma is a far more common left-sided heart mass than sarcoma, but a number of findings on echocardiography should help the physician differentiate sarcoma from the more common myxoma. Nonseptal origin of the tumor, extension into a pulmonary vein, multiple masses, a broad attachment, and a solid or semi-solid consistency are all more common in sarcoma than myxoma. Unfortunately, this is still not always easy to interpret, and misdiagnosis of sarcoma as myxoma, only to be surprised at surgery, is relatively common. Most left atrial masses seen by cardiac surgeons are benign myxoma. Therefore, it is not too great a surprise that all cases of left atrial sarcoma referred to us had been previously operated on with a presumed diagnosis of left atrial myxoma, since few cardiac surgeons have been exposed to primary cardiac sarcoma cases. All of these cases had rapid reappearance of the left atrial tumor at the site of resection that was more consistent with regrowth of persistent, incompletely resected sarcoma than with recurrence. Intracavitary left ventricular tumors are very uncommon and usually presumed to be malignant by most physicians, and they also present a much greater technical challenge for resection. Not surprisingly, both of our cases of large intracavitary left ventricular tumors were referred without prior attempt at resection.

Once a left-sided mass suspected to be sarcoma is identified by TTE, we obtain a transesophageal echocardiogram because of its increased definition of left-sided masses. Chest and abdomen CT scan and cardiac MRI are obtained in all cases thought to be sarcoma, and are complimentary to echocardiography. CT scan allows assessment of myocardial infiltration, pericardial involvement, extra cardiac involvement, and mediastinal disease for the local tumor, as well as assessment for metastatic disease. Cardiac MRI allows tissue characterization, as well as the assessment of cardiac valvular function and intracardiac blood flow, in a dynamic fashion. This will be discussed further in a separate section by Dr. Dipan Shah (see page 4). We also obtain a PET/CT scan in all patients suspected of sarcoma to see the PET activity of the primary tumor, and look for metastatic disease that is unfortunately common with these tumors. Cardiac catheterization and coronary arteriography is individualized based on the risk of coronary involvement with atherosclerotic disease or tumor.

Although a tissue diagnosis would be helpful in planning an approach to these tumors, it is very difficult to obtain from these left-sided lesions unless there is metastatic disease that can be easily biopsied. Most left-sided intracavitary cardiac masses need surgical removal even when benign due to the risk of blood flow obstruction or embolization, and most of our patients have been highly symptomatic leading up to the final tissue diagnosis usually made at surgery.

Primary cardiac sarcoma is a disease with an often dismal prognosis. When treated without surgical resection, the survival at 9 to 12 months is only 10%.\(^4\) Most reports in the literature are either autopsy series or case reports.\(^5\) Several series have been published dealing with cardiac tumors in general and containing a small number of sarcomas. These tend to highlight the technical difficulty, with operative mortality usually exceeding 20% and mean survival around 12 months.\(^8\) Other series have been published that focused exclusively on primary cardiac sarcoma but did not group patients by anatomic location, as we propose. The Mayo Clinic reported 34 patients over 32 years with a median survival of 12 months.\(^11\) A combined series from the Texas Heart Institute and the MD Anderson Cancer Center reported an actuarial survival of 14% at 2 years in 21 patients over a 26-year period.\(^12\) We have previously used this approach and reported on our cardiac sarcomas as a combined entity using a multimodality approach; we found a median survival of 23.5 months in 27 patients over 16 years, with survival of 80.9% at 1 year and 61.9% at 2 years.\(^13\)

The greatest determinant of long-term survival in primary cardiac sarcoma is complete resection. Complete resection is greatly affected by the anatomic location of the tumor and the structures that it infiltrates. The left atrium and left ventricle present unique anatomic challenges for the exposure needed for complete resection and reconstruction due to their anatomic location, as well as proximity, to vital structures that cannot be easily resected and reconstructed. This is evidenced by the high rate of local recurrence and secondary resections often reported in the literature.\(^14,15\) We hypothesized that the high rate of local recurrence
was due to the inability of the surgeon to adequately visualize the structures that needed to be removed and reconstructed, causing an inadequate resection that lead to rapid regrowth of tumor that was not completely removed, and that this could result in both secondary surgery for local recurrence and decreased survival. Typically, left atrial tumors are approached through the interatrial groove, and left ventricular tumors through the aortic valve, the mitral valve or the ventricle wall itself. Surgical approach through the interatrial groove is generally adequate for benign tumors, but limits visualization for malignant tumors, that are often larger and require a more generous margin of resection for success. We have considered complete cardiectomy and orthotopic cardiac transplantation for complete removal of these tumors. Although feasible, this approach requires the availability of a donor and post-operative immunosuppression, both of which present potential problems. Additionally, series using orthotopic cardiac transplantation for this purpose have only shown a median survival of 12 months.

Left ventricular tumors can be approached through the aortic valve, through the mitral valve, or with a ventriculotomy through the ventricular wall. A trans-aortic valve approach works nicely for benign tumors, but is inadequate for malignant tumors due to their size and the amount of resection needed. A ventriculotomy through normal ventricular muscle is possible, but not preferred by the cardiac surgeon if other options exist. Our group adopted the approach of cardiac explantation, ex vivo tumor resection, and cardiac reconstruction and reimplantation of the heart — cardiac autotransplantation — to allow a radical tumor resection and accurate reconstruction in the challenging case of left heart sarcomas. Our hypothesis was that this approach would allow complete tumor resection, yielding deceased local recurrence and increased long-term survival.

The technique of cardiac autotransplantation was introduced for cardiac tumors by Cooley in 1985 to deal with a large left atrial pheochromocytoma. Although this case was not successful, it introduced the senior author, Michael J. Reardon, to the technique and its potential use for cardiac tumors. Our group did the first successful cardiac autotransplant for cardiac sarcoma in 1998. We have reported this for left atrial sarcoma and left ventricular sarcoma. Working closely with the sarcoma oncology group and thoracic surgery group at MD Anderson Cancer Center, we currently have done 28 cardiac autotransplants, with 23 of these for primary cardiac sarcoma. The surgical technique of cardiac autotransplantation has been previously addressed and will be reviewed here.

Cardiac Autotransplantation

Cardiac autotransplantation has several fundamental differences from standard orthotopic heart transplantation. In orthotopic heart transplantation, unless a domino procedure is being done, the explanted heart is not to be used and any damage to its structures is inconsequential. Therefore, the cardiectomy can be performed, leaving a wide margin of remaining tissue to use in tailoring the heart to be implanted, without regard to cutting critical structures such as the coronary sinus. Similarly, the donor heart can usually be harvested with extra tissue at its margins to be used to help tailor the implantation. In cardiac autotransplantation, the heart must be excised in a manner that does not damage any structures that cannot be repaired or replaced or are vital to cardiac function. Additionally, if the heart is simply excised and reimplemented, there is a loss of tissue to sewing, which makes reimplantation more challenging than orthotopic heart transplantation. It is therefore vital that the surgeon carefully plan the cardiac excision and reconstruction in a manner to avoid these pitfalls.

Since the heart is to be totally explanted, the first consideration is a cannulation technique that does not involve the heart itself in any way. The aorta can be cannulated in the distal ascending aorta with the standard technique used by the cardiac surgeon. The venous cannulation, however, must be directly into the superior vena cave (SVC) and inferior vena cava (ICV) just below its junction with the right atrial junction. This requires exposure and mobilization of both the SVC and IVC at the diaphragm. After commencing cardiopulmonary bypass (CPB), we further mobilize the SVC and IVC until completely free and surround each with an umbilical tape on a tourniquet. We widely mobilize the interatrial groove and circumferentially mobilize the ascending aorta and pulmonary artery. This extensive dissection facilitates both the accurate excision of the heart and its reimplantation. The ascending aorta is cross clamped, and antegrade cold blood potassium cardioplegia (10 cc/kg) is administered to achieve cardiac arrest. The left atrium is opened at the beginning of cardioplegia and a sump drain placed to decompress the heart. After cardioplegia and cardiac standstill, we widely open the left atrium to confirm pathology and the need for autotransplantation as a technique. We divide the SVC first just beyond the right atrial junction. This is followed by the IVC division, which actually has a transection line on the right atrium near its junction with the IVC. For each of these, it is important to note that the rim of tissue remaining on the body side will retract substantially towards the venous cannulae,
and a wide rim must be planned for during transection, or reimplantation can be exceedingly difficult, especially for the IVC. We then divide the ascending aorta about 1 cm distal to the sinotubular junction, and the pulmonary artery just proximal to its bifurcation. The left atrium transaction is then completed, dividing the atrium just anterior to the pulmonary veins, and on the left side, equal distance between the pulmonary veins and the mitral valve and left atrial appendage. This allows complete removal of the heart, which is placed into a basin on ice slush (Figure 1). The posterior left atrium is then inspected and any tumor widely excised (Figures 2 to 4). Reconstruction is with bovine pericardium, and the pulmonary veins may be individually reimplanted into openings cut in the ovine pericardium, or left as a cuff if pathology permits (Figure 5). The anterior left atrium can be removed in its entirety to include the mitral valve, leaving only a mitral annulus if necessary. Reconstruction occurs by cutting an opening in the sheet of bovine pericardium to match the mitral annulus opening. Mitral valve replacement is then done with pledgeted 2-0 Ticron sutures, with the pledgets on the left ventricular side of the annulus passing through the annulus, then through the bovine pericardium, and then through the prosthetic mitral valve. When complete, this fully seals the neo-atrial wall to the valve and annulus. The anterior and posterior bovine pericardium can then be tailored for reanastomosis (Figures 6 and 7). Reimplantation is similar to standard cardiac transplantation, with reanastomosis of the left atrium first. We then reattach the right atrium to the IVC, and then the right atrium to the SVC. At this point, the need for a wide margin of tissue remaining on the body side of the original transaction is very apparent. If either of these requires excess tension, an interposition graft of Gore-
Tex, Dacron or pericardial self-constructed tube graft can be used to bridge the defect successfully. The pulmonary artery and then the aorta are reanastomosed in a standard fashion, warm blood potassium cardioplegia given antegrade, and the cross clamp is removed.

The procedure for left ventricular tumors is similar, and has required, in both of our cases, mitral valve excision due to tumor involvement and partial excision of the intracavitary interventricular septum (Figures 8 and 9). The interventricular septum was reconstructed with bovine pericardium, and valve replacement is done with a tissue valve. Although these patients are younger than the age in which we typically use a tissue valve, we would prefer to avoid Coumadin. The aggressiveness of this disease makes the risk of structural valve deterioration small.

We last presented our work in 2007 and published this experience with left heart sarcomas in 2008. This work is currently done under our IRB-approved protocol, A Phase II Trial of Cardiac Tumor Resection Including Autotransplantation and Radical Resection of Cardiac Tumors. At that time, we had 20 patients who had undergone 21 cardiac autotransplants (1 also had a redo autotransplant) by the senior author. Three of these patients had complex benign disease, leaving 17 patients and 18 cardiac autotransplants for left heart sarcoma for analysis. There were 7 males and 10 females with an average age of 39.5 years and a range of 20 to 57 years. Pathology showed 7 malignant fibrous histiocytomas (MFH), 5 undifferentiated sarcomas, 3 leiomyosarcomas, 1 osteosarcoma and 1 myxoid...
sarcoma (Table 1). Most of the MFH cases were early in the series, and the diagnosis of undifferentiated sarcoma occurred later, suggesting a change in pathology nomenclature rather than a fundamental change in tumor types being seen. There were 11 cases that involved cardiac autotransplant, alone, and 6 that also required pneumonectomy due to extensive lung, as well as cardiac, involvement.

In the 11 cases of cardiac autotransplant alone, there were no hospital deaths and all patients were discharged to home doing well. In the 6 cases that required pneumonectomy, there were 3 hospital deaths. Each of these deaths was precipitated by severe post-operative coagulopathy requiring multiple transfusions. This volume overload led to severe unilateral pulmonary edema in the remaining lung, and subsequent right heart failure leading to death was the common pathway of loss in each of these. The survival curve for all patients with left heart sarcoma is shown in Figure 10, and the survival curve for patients with left heart sarcoma with and without pneumonectomy is shown in Figure 11. Subsequent to this publication, we have operated on 4 more isolated cardiac autotransplants for left heart sarcoma with no hospital mortality, and 1 more combined cardiac autotransplant with pneumonectomy that we lost to the same mechanism of right heart failure. We now consider the need for pneumonectomy, in addition to cardiac autotransplant, a contraindication to surgery in almost all cases. At the time of publication, 8 of 17 patients with left heart sarcomas were alive 3 to 50 months after surgery, with a mean of 22 months. All isolated cardiac autotransplants for left heart sarcomas that we operated on after this study are alive without disease. All patients are offered an adjuvant chemotherapy regimen of doxorubicin hydrochloride (75 mg/m²) and ifosfamide (106 mg/m²). All deaths have been related to distant metastatic disease. There have been only 2 cases of local recurrence. One was successfully treated with redo cardiac autotransplantation but died 12 months later from metastatic disease, and 1 is alive and continuing systemic chemotherapy.
Figure 11. Survival curve of left heart sarcomas with and without pneumonectomy.

Conclusion

Treatment of cardiac sarcoma is based upon complete surgical resection, and survival rates have been shown to be directly related to complete versus incomplete resection. Cardiac autotransplantation is a viable technique to achieve complete resection, despite the anatomic difficulties posed by a left heart anatomic site. This can be done with a reasonable mortality in an experienced center for cardiac autotransplantation alone. The addition of pneumonectomy to cardiac autotransplantation adds an unacceptable risk and should be avoided. If extensive pulmonary involvement would necessitate pneumonectomy in addition to cardiac resection, systemic biologic therapy alone should be considered. Survival with this approach has improved over historic controls, and further improvements will likely come from a multimodality approach and better biologic treatment options.

References

Introduction

Cardiac sarcomas create 2 risks: local problems and metastatic disease. Most frequently, the histologies are angiosarcoma and high-grade pleomorphic unclassified sarcoma (formerly called MFH or malignant fibrous histiocytoma). There is also a clinical-pathological entity without distinctive histological features of tumors that originate in the pulmonary artery and are referred to as pulmonary artery sarcomas or intimal sarcomas of the pulmonary artery. Conventional wisdom indicates that soft-tissue sarcomas are poorly responsive to chemotherapy. Luckily, that is not the case. Attempts to concentrate on the local problem only with therapies up to and including cardiac transplantation have been unsuccessful due to the high rate of fatal metastatic disease.

Cytotoxic Chemotherapy

There are 2 combinations of standard chemotherapy drugs that can be beneficial in treating cardiac sarcomas: Adriamycin (doxorubicin) and ifosfamide, and gemcitabine and docetaxel. Most of the histological types of sarcomas that originate in the heart can respond to either or both of these regimens. For angiosarcomas, there are additional options.

The key to effective use of Adriamycin and ifosfamide is dose intensity. For Adriamycin, a randomized dose-response study showed a doubling of response rate at a dose of 75 mg/m² compared with 45 mg/m².1 For ifosfamide, the data are weaker, but our data indicate higher responses at higher doses.2,3 Based on this information, the standard Adriamycin-ifosfamide regimen at MD Anderson is Adriamycin at 75 mg/m² and ifosfamide at 10 g/m², and for younger, relatively healthy patients otherwise, we usually use Adriamycin at 90 mg/m².4 Of course, we always use a granulocytic growth factor to minimize the period of severe neutropenia, and we always give the Adriamycin with a cardioprotective strategy, usually continuous infusion over 72 hours and less frequently by rapid infusion with dexrazoxane. When mucositis limits tolerance, the shorter infusion is somewhat protective. Alternatively, palifermin at 180 mcg/m² given 3 days prior to Adriamycin infusion is effective at minimizing the mucositis.5 Ifosfamide is given in divided doses over 3 hours daily for 4 to 5 days, in conjunction with alkaline fluids and mesna. Ifosfamide is limited by nephrotoxicity and neurotoxicity. Nephrotoxicity is minimized by adequate hydration and, in some cases, by renal doses of dopamine. Neurotoxicity is minimized by assuring alkalization (never let the CO₂ go below 26) and by higher levels of serum albumin; we administer albumin prior to ifosfamide for anyone who has a borderline albumin level, or when a prior course caused neurotoxicity. Fortunately, it is always reversible, so there is no need to stop therapy in the presence of neurotoxicity. About 60% of patients will have objective tumor shrinkage and an additional 20%-30% will have lesser shrinkage or stabilization of previously progressive disease.

The combination of gemcitabine and docetaxel has been demonstrated to have greater activity than gemcitabine alone in a study carried out by the Sarcoma Alliance for Research through Collaboration (SARC), designed as a phase III study for patients with progressive disease after standard chemotherapy. The study
was a Bayesian adaptively randomized study, and was ultimately published as a phase II study as an editorial decision by the journal.\textsuperscript{6} Taxanes have no activity as single agents in most sarcomas, but they are highly active in angiosarcomas of the scalp and have some, but much less, activity in primary angiosarcomas of other primary sites. Gemcitabine has single-agent activity in sarcomas, so the gemcitabine-docetaxel combination is particularly attractive for angiosarcomas.\textsuperscript{7,8} The histological type most sensitive to the gemcitabine-docetaxel combination in the salvage setting was MFH, so this combination is also attractive for cardiac sarcomas.

One of the limiting side effects of this combination is fluid retention from the docetaxel, and when that occurs, we recommend continuation of gemcitabine with increased dose if tolerated. Gemcitabine requires phosphorylation to be incorporated into DNA, and the phosphorylation is limited to 10 mg/m\textsuperscript{2}/min, so the relatively rapid infusion of gemcitabine when used according to the package insert results in excretion of a substantial portion of inactivated drug.\textsuperscript{6} For good-risk patients taking the gemcitabine-docetaxel combination, gemcitabine is given on days 1 and 8 at 900 mg/m\textsuperscript{2} over 90 minutes and docetaxel at 100 mg/m\textsuperscript{2} over 60 minutes. For poor-risk patients, gemcitabine is given at 675 mg/m\textsuperscript{2} over 70 minutes and docetaxel at 75 mg/m\textsuperscript{2} over 60 minutes.

**Targeted Therapy of Cardiac Angiosarcomas**

Since angiosarcomas are malignant tumors with functional and morphological features of normal endothelium, targeted agents that inhibit the process of new blood vessel growth have been a source of great interest in the treatment of these tumors. Angiogenesis, the process of endothelial proliferation and formation of microvessel sprouts, is physiologically brought about by the interaction of a host of angiogenic factors and inhibitors. Pathological angiogenesis, seen in tumors and some other diseases, continue to depend on tipping the balance of these factors in favor of new blood vessel formation. With the aid of drugs that inhibit angiogenesis, we hope that malignant proliferating endothelial cells can be inhibited and thereby arrest the growth of these tumors.

Cardiac angiosarcomas are extremely rare, and large studies examining the utility of targeted agents in treating these are unavailable. Most of the data comes from small trials that enroll a variety of sarcomas, including angiosarcomas. Agents that inhibit angiogenesis may have a role in the treatment of angiosarcomas, and may be broadly classified into 3 groups depending on the number of angiogenic proteins they inhibit. It is important to make the distinction between agents that inhibit only a single angiogenic protein from others that affect multiple target proteins, as it may be relevant to the efficacy of these agents and to the development of drug resistance. Most tumors can express multiple angiogenic proteins\textsuperscript{9} and tend to develop drug resistance more readily when treated with agents with activity towards 1 specific target. Another critical aspect to consider while choosing therapy is the rate of progression of cardiac angiosarcomas. It should be borne in mind that monotherapy with anti-angiogenic agents works best in slow-growing tumors, and is best avoided in rapidly progressive tumors. Cytotoxic chemotherapy, either alone or in combination with targeted agents, should be considered in those situations.

**Group I: Drugs that Inhibit 1 Main Angiogenic Protein**

Bevacizumab (Genentech, South San Francisco, California) is a humanized monoclonal anti-vascular endothelial growth factor (anti-VEGF) antibody that is currently widely used in the treatment of colorectal, lung and breast cancers in combination with cytotoxic chemotherapy. The agent appears to have limited activity as monotherapy, and the same is true for angiosarcomas. Combination therapy of bevacizumab with doxorubicin was examined in a phase II trial that enrolled 17 patients with sarcoma; unfortunately, no angiosarcomas were enrolled. This trial raised some toxicity concerns as a grade 2 decline in LVEF was observed in one-third of the patients. Cardiac toxicity was often reversible — of the 6 patients who had a decline in cardiac ejection fraction, 5 showed improvement over time.\textsuperscript{10} Since doxorubicin appears to have good activity in cardiac angiosarcomas, the role of the combination needs further study. Combination of gemcitabine and docetaxel with bevacizumab has been shown to have activity in angiosarcoma, and the authors reported 2 complete responses in patients with angiosarcoma.\textsuperscript{10} The role of bevacizumab in these responses is unknown since the chemotherapy combination is definitely active by itself, in our experience. Currently, there are 3 trials that are recruiting patients looking at the efficacy of bevacizumab in the treatment of angiosarcomas: the first is a monotherapy trial, the second is a combination of gemcitabine and docetaxel with or without bevacizumab, and the third is a combination of paclitaxel and bevacizumab. These trials are specific for angiosarcoma, and will have sufficient numbers of patients with angiosarcoma enrolled in them to derive useful conclusions about the utility of this agent in angiosarcoma therapy.
**Group II: Drugs that Inhibit 2 or 3 Main Angiogenic Proteins**

Sunitinib (Sutent, Pfizer Inc., New York, NY) is a multi-targeted tyrosine kinase inhibitor with activity against multiple targets, including VEGF-R2, platelet-derived growth factor (PDGF) receptor and c-KIT receptor. It also has activity against Flt-3, neurotrophic factor receptor and CSF-1, and blocks VEGF receptors 1 through 3.¹¹ Data on the utility of this agent in treating angiosarcomas is limited, as the only published study on the activity of this agent in non-GIST (gastrointestinal stromal tumor) sarcomas enrolled 2 patients with angiosarcoma, and both patients failed to show a response.¹²

Sorafenib (Nexavar, Onyx, Emeryville, California, and Bayer, Leverkusen, Germany) is a multi-targeted tyrosine kinase inhibitor with activity against Raf, PDGFR, VEGF-R2, VEGF-R3 and c-KIT. The activity of this agent was examined in a phase II trial that enrolled 37 patients with angiosarcoma, and 5 patients (14%) showed a response to sorafenib. The only patient to develop a complete response to treatment was an angiosarcoma patient; 4 other patients had a partial response.¹³ In another trial that enrolled 37 patients with sarcoma, 7 out of 9 patients with vascular tumors had stable disease with a median progression-free survival of 4.7 months.¹⁹ Clinical trials examining the combination of sorafenib with cytotoxic agents such as dacarbazine are currently underway.

**Group III: Drugs that Block a Wide Range of Angiogenic Proteins**

This group includes agents that have a broad spectrum of activity against a wide range of angiogenic regulators. Examples include endostatin and capillosstatin that downregulate VEGF, bFGF, bFGF receptor, HIF1α, EGF receptor, ID1 and neuropilin, and upregulate thrombospondin. Currently, there are no published data evaluating the role of these agents in cardiac angiosarcomas, but it is an area requiring investigation.

**Key Concepts for Effective Inhibition of Angiogenesis in Tumors**

While cytotoxic chemotherapy is intended to cause tumor cell death by a direct effect, anti-angiogenic therapy can exert its effect on endothelial proliferation either directly (by inhibiting the endothelial cells) or indirectly (by reducing the tumor’s production of angiogenic proteins). There are several differences between the cytotoxic strategy and the anti-angiogenic strategy that need to be considered while picking a treatment for particular clinical situation.

**Tumor growth rate.** Angiosarcomas can grow quite rapidly, and the rate of progression of the tumor needs to be considered while developing a therapeutic strategy. Cytotoxic chemotherapy is more effective on rapidly growing tumors, while anti-angiogenic therapy works best in slow-growing tumors.⁹ Consequently, monotherapy of rapidly growing angiosarcomas with anti-angiogenic agents may not be effective. In such situations, a cytotoxic agent alone or in combination with an anti-angiogenic agent may be a better choice. After several cycles of chemotherapy as the disease settles to a stable status, monotherapy with anti-angiogenic agents may be considered with close radiological monitoring.

**Relationship between dose and response.** Dose and response tend to have a more linear relationship with cytotoxic chemotherapy. Anti-angiogenic agents tend to have a biphasic, U-shaped dose efficacy curve, where blood levels that are too low or too high may be ineffective in the inhibition of angiogenesis. This biphasic dose efficacy of anti-angiogenic agents needs to be considered while using these drugs alone and in combination with cytotoxic chemotherapy. More is not necessarily better when it comes to anti-angiogenesis, and this has been demonstrated in interferon-α⁴ and endostatin.¹⁵

**Frequency of dosing.** Cytotoxic chemotherapy is often administered at the maximum tolerated dose with a long off-therapy interval to allow for bone marrow and mucosal recovery. While this might be optimal for producing cytotoxicity, anti-angiogenic therapy requires endothelial cell exposure to steady blood levels of the inhibitor.¹⁰ Hence, anti-angiogenic agents with a short half-life need to be dosed daily and without any breaks.⁹

**Adverse effects.** Side effects of anti-angiogenic therapy are different from those of cytotoxic agents. Unlike cytotoxic agents, myelosuppression, nausea and hair loss are all unusual with anti-angiogenic therapy, while a whole host of other side effects such as hypertension, bleeding, bowel perforation and thromboembolism can occur. Evaluation of the side-effect profile against the backdrop of the patient’s existing medical problems is an important step while choosing therapy. Combination of cytotoxic therapy with anti-angiogenic therapy may increase the risk of thromboembolism over monotherapy.¹⁷
References


The weekend after Dr. Michael E. DeBakey died, one medical historian described him as the greatest physician of the 20th century. I consider him to be one of the most influential physicians since Galen, a Greek physician born in the Roman Empire in the 2nd century A.D. Galen was so influential that his theories were accepted as dogma in western medical science for over 1,000 years. I would like to think that Dr. DeBakey will still be remembered a thousand years from now.

The son of Lebanese immigrants to the United States, Dr. DeBakey was an internationally renowned pioneer in the treatment and prevention of cardiovascular disease. His accomplishments in medical science would fill an entire book. His research was published in more than 1,000 papers. His contributions as a medical statesman are unparalleled — from helping to start the government-administered health insurance program (Medicare) in the United States, to promoting the establishment of the National Library of Medicine, to consulting on the heart operation of Boris Yeltsin — and the list goes on. I had the opportunity to travel many places with him, and whether it was Turkey, China, or Russia, he was approached by people who thanked him for saving or extending the life of a spouse, child, or loved one. Dr. DeBakey treated the rich, the famous, and the powerful, and he treated the poor and humble. He treated all of his patients with the same dedication to the relief of human pain and suffering.

His influence was truly global and continues to touch people, even in Qatar. A couple of years ago, he started the DeBakey High School for Health Professions in Houston, Texas. A couple of days after he died, I received a copy of the *Gulf Times*. It contained an advertisement for a new program started by the Qatar Foundation. The advertisement read, “DeBakey High School for Health Professions at Qatar is now accepting student applications.” I think it is amazing that his influence continues to span the globe in this way, and that it connects his Middle Eastern heritage with his illustrious career in the United States.

What set Dr. DeBakey apart from so many others in our field was his basic humanity. There was a period of years when Houston was the mecca of cardiovascular surgery. I remember when the Duke of Windsor said he came to Houston to see “the maestro.” I spoke recently with a prominent cardiologist in New York who would send all of his difficult surgical cases to Dr. DeBakey. Not a single time did Dr. DeBakey ever ask, “Can this patient pay? Does he have insurance? Is he on Medicare? Is he a VIP?” He would take one and all, regardless of whom they were. I observed him treat a poor patient with no source of funds with the same attention and respect that he gave to a head of state.

I would like to share a personal experience. One of my daughters was hospitalized scores of times during her high school years. When he was in town, Dr. DeBakey never missed a day visiting her in her room. And some days, this was the only time that her face would brighten up, and she would become animated. One day when he visited, my wife, Anita, was sewing a prom dress for one of our daughter’s friends whose mother was ill.

Dr. DeBakey said, “Anita, what are you doing?” My wife told him, and he said, “Let me see your stitches.” He examined the dress and said, “These are your basting stitches, aren’t they?”

She said, “No, Dr. DeBakey, these are my finishing stitches.”

“These are terrible,” he said. “Let me have your scissors!”

He ripped out every single stitch and said, “Let me have your needle and thread.” He proceeded to resew the dress in its entirety while my wife sat there in a state of astonishment.
Fast-forward 15 years later. Dr. DeBakey was having a New Year’s Eve dinner at our home in Houston. My wife cooked gumbo, a kind of stew from Louisiana that she knew he loved. And, of course, Dr. DeBakey never forgot anything. He told her, “Anita, you may not be able to sew, but you sure cook good gumbo.”

I cannot begin to describe the impact of Dr. DeBakey’s influence on my own career. When I told him I was moving to New York at age 60, he said, “No one will miss you more than I will, but you have to go where you can accomplish the most. You know you have another 35 or 40 years of work ahead of you.”

In summing up my feelings about Dr. DeBakey, I would include awe, admiration, inspiration and love. I will remember him as a man of great kindness and extraordinary ability.

---

Typical Day in the Life of a Surgeon

“I usually get up about 4:30 or five o’clock, and I work in my study for maybe a couple of hours, mostly studying data or writing, and then I come to the hospital. I get to the hospital between 6:30 and seven. And then I check on the cases I’ve got operating that morning. Usually by 7:30 we are in the operating room, starting operations. Depending upon what the load is, I may be through by three or four o’clock. Then, I will often take the necessary calls that have accumulated, try to get to some correspondence. And then I’ve got to see patients that are coming in as outpatients, and also patients that are in the hospital being prepared for an operation the next day. Or there may be a committee meeting I have to attend, or meet with my people in the research laboratory to go over certain things that they are doing, and sort of bringing up to date where we are, certain subjects that we are dealing with, things of that sort. So by eight or nine o’clock, I’ll get home and have something to eat. And usually by eleven, between eleven and twelve, I go to bed.”

— Dr. Michael E. DeBakey

DeBakey ME. Personal interview on American Academy of Achievement website. Available at: http://www.achievement.org/autodoc/page/coo0int-7

This tribute is reprinted with permission from Heart Views.
Beginning in this issue, we will initiate a Poet’s Corner edited by Dr. Michael W. Lieberman, former chair of the Department of Pathology at The Methodist Hospital and former director of The Methodist Hospital Research Institute. Our goal is to provide for your enjoyment a poem in each issue of the journal from an award-winning, modern poet selected by Dr. Lieberman. We welcome your comments on the Poet’s Corner and, as always, on articles appearing in the journal. MDCVJ does not accept unsolicited poems for publication in the Poet’s Corner.

Returning to the Luxapalila

The river is the color of earth, fed by runoff
from pastures and fields of cotton and corn
and forestland heavy with humus.
Kneeling by the water near a shale outcropping,
I shatter my face and settle my outspread hand,
palm up, until it fades from sight
like something drowned in history’s dark pages —
now you see it, now you don’t.
Watching the hand disappear, I see
the face of a girl eased down by the pastor,
her paleness and blond hair darkened,
held below that brown rush
until she broke the surface again,
arms flung wide in the flaring sun,
face shining like an angel’s,
white marble with thin blue veins
trailing from her temples to blend
with water whispering off her hair,
dress sheer and tight on her tiny breasts,
Thank you, Jesus, he cried to the water and the woods.
Here as a boy I curled at the end
of a cable swing, flung out, released,
hung there, wingless creature floating on air
until gravity snatched me and I dropped
breathless to the river, the flash of green bank,
the sun a yellow something spinning on blue,
then my feet entering the water, my body
going down through that wet tunnel,
the color of weak whiskey across my eyes,
a darker stronger bourbon, then nothing,
slipping into the earth itself, and deeper,
until my feet touched the bottom,
the spongy primordial end of the world.
A thrust and I rose through the tunnel,
eyes uplifted toward the brightness,
hands and arms battering like wings
to burst breathless to green and blue,
the steady round face of the sun,
my vision bleared by water,
the taste of Earth upon my tongue.
My feet uncertain against the muddy slope,
I clamber back to the level of brush and briar
on the bluff, watch the brown ribbon below
weaving around a grassy bar, and see —
is it a simple slant of light
breaking from behind me? —
the girl’s marble-white face rising free,
hair streaming, cupped by my hand,
her arms stretched out to the mounting sun.

Paul Ruffin, author of 2 novels, 3 collections of short stories, 4 books of essays, and 7 collections of poetry, is the 2009 Texas State Poet Laureate and Texas State University System Regents’ Professor and Distinguished Professor of English at Sam Houston State University, where he edits The Texas Review and directs Texas Review Press. Printed with the author’s permission.
May 12, 2010

Re: Volume 6, Number 1, pages 42–44

Dear Dr. Winters:

It is refreshing to read the tribute to Dr. Michael E. DeBakey by Philip A. Salem. It is of great importance to keep Dr. DeBakey’s numerous contributions in the fields of medicine and cardiothoracic-cardiovascular surgery alive, and to portray his human nature as a true humanitarian, and a life devoted selflessly to loving his father, mother and brother, and sisters, as well as to friends — in a way beyond service to science — and to his patients, that to this day, no other physician has demonstrated the same qualities.

Dr. Salem did not know fully his qualities as a physician, the integrity, and the dynamic nature of the demands for perfection he imposed on his peers and residents working on the surgical-medical service at The Methodist Hospital, as I did during my residency training.

Drs. Dennis, Don Chapman and Bob Hettig, all of whom mentored me as I worked up the patients to clear them for surgery, be it patients with aneurysms, congenital heart defects, coronary artery disease, or peripheral occlusive arterial diseases. No less than 80 inpatients were under the care of Dr. DeBakey at any one day at the Methodist.

As a treat, Dr. Don Chapman, after morning rounds and discharging his patients home, would invite me for breakfast at the Toddle House right across from The Methodist Hospital on Fannin Street, keeping our contact with operators at the hospital.

Another humanitarian aspect of Dr. DeBakey occurred to my wife and me, after she delivered our first born son. No beds were available in the obstetrics unit, yet Dr. DeBakey brought his mother’s bed to a special space in the hospital where my wife could rest and nurse our baby. I could not thank the great Dr. DeBakey enough. With all humility, he smiled and continued on his rounds.

This is just a brief narrative of the work and deeds of a great man and great surgeon.

Sincerely,

Mounir E. Nassar, M.D.
The Methodist DeBakey Heart & Vascular Center continues the groundbreaking work begun by famed heart care pioneer, Dr. Michael E. DeBakey and his associates, who developed many of today’s life-saving techniques, tools and procedures at The Methodist Hospital. Located in Houston, Texas, the Methodist DeBakey Heart & Vascular Center combines research, prevention, diagnostic care, surgery and rehabilitation services in a coordinated multidisciplinary program with one focus: delivering compassionate, effective care and treatment to patients suffering from heart disease.