Seven children with primary cardiac tumors are discussed. The diagnosis was achieved by cardiac catheterization in all seven patients. In four patients, surgical excision was undertaken with one long-term good result. One of the other three surgical patients died of acute heart failure at the discontinuation of cardiopulmonary bypass. One of the other two surgical patients died suddenly postoperatively, presumably from arrhythmia. The other, affected by fibrosarcoma, also died suddenly while undergoing chemotherapy. The histologic findings on the tumor masses were available in six patients. There were three rhabdomyomas, one fibroma and one hamartomatous mass. In one patient with tuberous sclerosis, a presumptive diagnosis of rhabdomyoma was made clinically on the basis of multiple masses infiltrating the left ventricle.

Two-dimensional echocardiography is the best non-invasive tool to determine the extent and location of the tumor and suitability for surgery. When surgery is indicated, cardiac catheterization for pressure data is also required. Surgery is recommended in symptomatic solitary lesions that most likely are fibromas and have an overall good surgical outcome.

Tumors of the heart are rare in infancy and childhood. Nevertheless, they are reported with surprising frequency. Interest in them, once limited to their pathologic features, is now directed to their early diagnosis and planned surgery because more successful surgical excisions of such tumors have been reported (1–6).

In our experience, surgery has an important role only in carefully selected cases. Recent advances in diagnostic techniques allow for accurate preoperative determination of the extent and location of the lesion and estimation of the probability of successful surgical excision. In this report, we review our cases of cardiac tumors in infants and children and discuss surgical implications.

**Study Patients**

Seven children with primary heart tumors presenting between the age of 1 day and 5 years were studied at the University of Alabama in Birmingham from January 1966 to December 1981 and their findings represent the subject of this report. Patients older than 15 years were not considered. A summary of the clinical presentation, pathologic features and surgical results is shown in Tables 1 and 2.

**Case Reports**

**Case 1.** A 3,100 g black male infant, the fourth child of a 28 year old mother, presented at birth (38 weeks' gestation) with severe respiratory distress and low cardiac output. On examination there was a grade 3/6 systolic murmur over the precordium and gross hepatomegaly. Chest X-ray film showed cardiomegaly and the electrocardiogram revealed nonspecific ST segment and T wave changes. Shortly afterward, he had a cardiac arrest and was resuscitated. Cardiac catheterization demonstrated gross distortion of the right ventricular cavity which was filled by a large mass (Fig. 1). The left ventricle was of normal configuration but displaced upward and posteriorly by the extensive lesion. Cardiac output was insufficient to sustain life and the infant died the next day with acute cardiac and renal failure.

Autopsy revealed extensive multiple rhabdomyomas occupying most of the right ventricular free wall and the interventricular septum. The brain was diffusely involved by the sclerotic plaques of tuberous sclerosis and showed evidence of obstructive hydrocephalus.
Cardiac catheterization revealed a large mass filling the cavity of the left ventricle (Fig. 2). At 5 days of age, the infant underwent median sternotomy for assessment of the suspected cardiac tumor. Tumor masses were found to involve a large area of the posterolateral aspect of the left ventricle as demonstrated by angiography; they were also found in the anterior wall of the right ventricle and throughout the right atrial wall. A nodule of tumor at the site of the sinoatrial node probably accounted for the initial sign of fetal bradycardia.

Biopsy of the tumor showed the features of rhabdomyoma and no further surgery was attempted. At follow-up 6 weeks later, there was no hemodynamic impairment but the infant died suddenly at 3 months of age; autopsy was not performed.

Case 3. A full term, 3,350 g black male infant had severe respiratory distress shortly after delivery. On examination, there were no cardiac murmurs but evidence of severe congestive heart failure was present. Chest X-ray film showed massive cardiomegaly. M-mode and two-dimensional echocardiography demonstrated a large interventricular septal mass with multiple separate masses protruding into the right ventricular cavity (Fig. 3 and 4).

Cardiac catheterization confirmed the presence of a septal mass distorting the right ventricular cavity and displacing the left ventricle posterolaterally. Pressure measurements did not show significant outflow tract obstruction of the left or right ventricle. Surgical intervention was not advised and the child died from acute heart failure 24 hours later.

Photographs of the autopsy specimen are shown in Figure 5. Histologic examination showed the tumor to be a rhabdomyoma. Diffuse sclerotic plaques of tuberous sclerosis were found on brain examination.

Case 4. A previously healthy, 14 month old white boy was studied after a chest X-ray film taken during an upper respiratory tract infection showed cardiomegaly with flecks of myocardial calcification. Auscultation of the precordium revealed a grade 3/6 ejection systolic murmur and the electrocardiogram showed T wave inversion in leads I, aVF and V6.

Cardiac catheterization showed a large mass distorting the cavity of the left ventricle but no significant outflow tract obstruction. Two-dimensional echocardiography showed a huge solid mass in the interventricular septum, extending to the anterolateral aspect of the free wall of the left ventricle. There was no clinical evidence of tuberous sclerosis.

A median sternotomy was performed and the echocardiographic findings confirmed. The left ventricular cavity was stretched over a large, firm globular septal mass the size of a hen’s egg. Cardiopulmonary bypass was undertaken; the tumor was approached through the apex of the heart, the septum opened and the anterior and posterior descending coronary vessels avoided. The tumor itself was well circumscribed, white, whorled and similar in appearance to a uterine fibroid. The mass was carefully dissected.
out of the septum, with care taken to remain close to pathologic tissue. In this way, all macroscopic tumor was removed without damage to the conducting tissue and the septum was then reconstructed. At discontinuation of bypass, the heart functioned well and postoperative recovery was uneventful. Histologic study showed the tumor to be a fibroma.

The patient remains clinically well 3 months later. A repeat two-dimensional echocardiogram showed the huge mass in the ventricular septum to have disappeared even though septal thickness remained above normal limits.

**Case 5.** A previously white, healthy 2 year old boy presented with lethargy and ankle swelling. An electrocardiogram showed low voltage complexes with ST segment elevation in leads II, III, aVF and V6 and chest X-ray film demonstrated cardiomegaly. Pericardial effusion was diagnosed and needle pericardiocentesis produced blood-stained serous fluid. The symptoms soon recurred, however, and cardiac catheterization demonstrated elevated atrial pressures consistent with pericardial tamponade; no lesion was found on angiography. A pericardial window was created surgically and 350 ml of heavily blood-stained fluid was drained. Analysis of this fluid for malignant cells proved negative.

* A median sternotomy was performed and a large fleshy tumor mass was found in the epicardial aspect of the posteroinferior left ventricular wall. Cardiopulmonary bypass was instituted and all macroscopic tumor excised with reconstruction of the ventricular wall. The child made a satisfactory recovery, and when histologic examination showed the tumor to be a fibrosarcoma, chemotherapy with vincristine, cyclophosphamide and actinomycin-D was started. He died suddenly 3 months later; permission for autopsy was not obtained.

**Case 6.** An asymptomatic 2 year old white girl had been diagnosed as having cerebral tuberous sclerosis at the age of 7 months. Auscultation of the heart revealed a grade 3/6 pansystolic apical murmur with radiation to the axilla. Chest X-ray film showed cardiomegaly and the electrocardiogram showed incomplete left bundle branch block. Angiography demonstrated a mass in the posterior left ventricular wall with apical dyskinesia but no left ventricular outflow tract obstruction. Two-dimensional echocardiography showed diffuse infiltration of a large part of the left ventricular free wall and radioisotope cardiac scintigraphy confirmed the presence of a diffuse intramural mass involving the apical, posterior and lateral aspects of the left ventricle.

In view of the extent of the tumor, absence of clinically significant hemodynamic impairment and the presence of cerebral tuberous sclerosis requiring treatment with adrenocorticotropic hormone (ACTH) and anticonvulsive drugs, surgical intervention was considered inappropriate. The tumor is almost certainly a rhabdomyoma and the patient remains in hemodynamically stable condition.

**Case 7.** This 5 year old white girl had been treated since infancy with digoxin and propranolol for paroxysmal tachycardia. At the time of presentation, the electrocardiogram...
Table 1. Seven Cases of Primary Cardiac Tumors in Infants and Children: Clinical Presentation

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Presentation</th>
<th>Heart Murmurs (grade)</th>
<th>Electrocardiogram</th>
<th>2 D Echo</th>
<th>Catheterization</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 day</td>
<td>Severe respiratory distress, low cardiac output, cardiac arrest</td>
<td>Systolic (3/6)</td>
<td>Nonspecific ST-T changes</td>
<td>NA</td>
<td>RV cavity distorted by mass, LV cavity normal but displaced</td>
</tr>
<tr>
<td>2</td>
<td>1 day</td>
<td>Fetal bradycardia, cesarean section</td>
<td>None</td>
<td>Nonspecific ST-T changes</td>
<td>NA</td>
<td>Intracavitary LV mass</td>
</tr>
<tr>
<td>3</td>
<td>1 day</td>
<td>Severe congestive heart failure</td>
<td>None</td>
<td>Nonspecific ST-T changes</td>
<td>Large interventricular septal mass, multiple separate masses in RV</td>
<td>RV cavity distorted by mass, LV cavity normal but displaced</td>
</tr>
<tr>
<td>4</td>
<td>14 mo</td>
<td>Unrelated respiratory infection, cardiomegaly by chest X-ray</td>
<td>Systolic (3/6)</td>
<td>T wave inversion in leads I, aVF and V₆</td>
<td>Huge solid interventricular septal mass</td>
<td>Mass distorting LV outflow tract without pressure gradient</td>
</tr>
<tr>
<td>5</td>
<td>2 yr</td>
<td>Lethargy and fluid retention</td>
<td>None</td>
<td>Low voltage with ST elevation in leads II, III, aVF and V₆</td>
<td>NA</td>
<td>Reduced contractility of RV and LV, pericardial effusion</td>
</tr>
<tr>
<td>6</td>
<td>2 yr</td>
<td>Cerebral tuberous sclerosis diagnosed at age 7/12, cardiac murmur present</td>
<td>Systolic (3/6)</td>
<td>Incomplete left bundle branch block</td>
<td>Diffuse infiltration of a large portion of LV free wall</td>
<td>Mass in posterior LV wall, apical dyskinesia without pressure gradient</td>
</tr>
<tr>
<td>7</td>
<td>5 yr</td>
<td>History of paroxysmal atrial tachycardia</td>
<td>Systolic (3/6)</td>
<td>LV hypertrophy, ST-T changes</td>
<td>NA</td>
<td>Mitral valve incompetence, intramural LV mass</td>
</tr>
</tbody>
</table>

*Each patient had cardiomegaly by chest X-ray film. LV = left ventricular; NA = not available; RV = right ventricular; 2 D Echo = two-dimensional echocardiography.

showed left ventricular hypertrophy with ST segment depression in the left precordial leads. Chest X-ray films showed cardiomegaly with an abnormally prominent left heart border. Left ventricular angiography demonstrated mitral regurgitation and a distorted free wall of the left ventricle. The preoperative diagnosis was that of an intramural mass, most likely a tumor.

The heart was explored through a median sternotomy and a round, hard mass (5 cm in diameter) was found in the posteroinferior aspect of the left ventricular wall extending to the atrioventricular groove. This appeared to be well circumscribed but not encapsulated and biopsy examination by frozen section demonstrated benign hamartomatous tissue. Cardiopulmonary bypass was begun and the tumor was completely excised with reconstruction of the ventricular wall. At discontinuation of bypass, the child died from acute left heart failure.

Discussion

Previous authors (1,2,4–11) have discussed in detail the pathologic features of cardiac tumors in infancy. Though rare, they occur with sufficient frequency to be considered in any infant or child who presents an unexplained heart murmur, arrhythmia or congestive heart failure, especially if accompanied by fever, anemia or nonspecific electrocardiographic changes. A knowledge of the natural history of cardiac tumors is of great benefit in planning a logical approach to their treatment.

Benign tumors. The most common primary cardiac tumors in children are rhabdomyomas and fibromas; both are benign lesions (1,2). Rhabdomyoma, which is probably a hamartomatous malformation rather than a true neoplasm, is the most frequent lesion encountered and occurs most often in infants with tuberous sclerosis, a familial condition characterized by epilepsy, adenoma sebaceum and mental retardation. Many other hamartomas are found in the brain, kidneys and pancreas. The myocardial involvement in this condition is usually in the form of multiple tumor nodules and often results in fetal or perinatal death (9). Their effects result from mechanical obstruction or sudden death from arrhythmia (12). Because they are embedded in the ventricular walls, their true extent is seldom demonstrated angiographically. Fibroma, myxoma, hemangioma, lipoma and cardiac teratoma are seen sporadically (1,6,13). Fibromas constitute approximately 5% of primary cardiac tumors, and 85% are found in infants and children (6). These are usually single, encapsulated and easily removed surgically.

Malignant tumors. Primary malignant tumors of the heart are extremely rare in children (1). They are usually sarcomas, and early death occurs often because of rapid tumor growth, systemic metastatic spread and lack of favorable response to any treatment. There are many more metastatic than primary tumors of the heart and the clinical picture is usually dominated by the underlying disease.
Lymphosarcoma is the most common. Only three cases of metastatic Wilms’ tumor have been reported at Boston Children’s Hospital in 53 years (8). Malignant tumors often present with a blood-stained pericardial effusion and may sometimes be diagnosed by pericardiocentesis and exfoliative cytology. In any event, a search for a primary lesion elsewhere is indicated before surgical exploration is considered.

**Planning surgical treatment.** Until recently, evaluation of a child with a suspected cardiac tumor relied principally on cardiac catheterization, followed by surgical exploration even in the absence of hemodynamic impairment. Angiography, at best, demonstrates only intracavitary involvement with accuracy, and surgical exploration often proves useless if unexpected extensive involvement of the ventricular walls precludes beneficial excision (Case 2). Because most tumors are benign and slow growing, and malignant tumors are, from previous experience, always fatal, there is rarely an indication for urgent operation unless severe hemodynamic impairment exists. Before a child is submitted to a major surgical procedure, it is important that the aims and potential benefits be clearly defined.

Recent advances in two-dimensional echocardiography now provide considerably better visualization of intramyocardial tumors so that accurate assessment of tumor lo-

**Table 2. Pathologic Findings and Surgical Results**

<table>
<thead>
<tr>
<th>Case</th>
<th>Pathology</th>
<th>Operation</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Extensive multiple rhabdomyomas of RV free wall and ventricular septum by autopsy. Obstructive hydrocephalus from tuberous sclerosis</td>
<td>None</td>
<td>Death from cardiorenal failure at age 2 days</td>
</tr>
<tr>
<td>2</td>
<td>Multiple rhabdomyomas of RA, RV and LV at surgery. Nodule at site of sinoatrial node</td>
<td>Exploratory thoracotomy and biopsy</td>
<td>Death at age 3 months</td>
</tr>
<tr>
<td>3</td>
<td>Rhabdomyoma of RV Tuberous sclerosis at autopsy</td>
<td>None</td>
<td>Death at age 2 days from heart failure</td>
</tr>
<tr>
<td>4</td>
<td>5 cm fibroma</td>
<td>Excision of septal tumor, reconstruction of septum Intracardiac pericardiocentesis of serosanguineous fluid with negative malignant cytology; pericardial window with removal of 350 ml fluid; excision of large tumor from posteroinferior LV wall</td>
<td>Well at 3 months postoperative</td>
</tr>
<tr>
<td>5</td>
<td>LV fibrosarcoma</td>
<td>Initial recovery from operation; chemotherapy with vincristine, hydrophosphamide and actinomycin-D; sudden death at 3 months postoperative</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Suspected rhabdomyoma</td>
<td>None</td>
<td>Clinically stable from suspected rhabdomyoma</td>
</tr>
<tr>
<td>7</td>
<td>5 cm hamartomatous lesion</td>
<td>Excision of 5 cm hamartomatous mass in posteroinferior aspect of left ventricle extending to AV groove</td>
<td>Death from acute heart failure</td>
</tr>
</tbody>
</table>

AV = atroventricular; LV = left ventricle; RA = right atrium; RV = right ventricle.
cation and extent can be made by noninvasive means. Angiography and pressure measurements at cardiac catheterization and electrocardiographic assessment of conduction system involvement will then complete the evaluation. Also, after precise location of one or more lesions, it may be possible at the time of cardiac catheterization to obtain a tissue diagnosis by endomyocardial biopsy, though we have no experience with this.

Combined assessment by these methods should lessen the need for exploratory thoracotomy in many cases because the radiologic and echocardiographic features of the tumor are often sufficient to reveal its histologic nature. Hence, the likelihood is that multiple myocardial nodules in infants are rhabdomyomas; the presence of tuberous sclerosis confirms this diagnosis (10,14). Rhabdomyoma is the most common primary tumor of the heart in infants and children (1,7,9,15,16). A large, well circumscribed lesion in the ventricular walls or septum is likely to be a fibroma (12) or intracardiac teratoma (5). Treatment planning is greatly benefited by well considered judgment of the nature of the tumor with an accurate assessment of its size, location, hemodynamic significance and extension into nearby structures.

Surgical indications and treatment. The aim of surgery should be to completely resect the lesion because residual tumors may later result in sudden death by arrhythmia (4,12). Large tumors can be resected successfully if they are solitary, well circumscribed and benign, as in our patient with a fibroma (Case 4), a similar patient of Mustafa et al. (3) and the patient with pedunculated rhabdomyoma of Arciniegas et al. (2). In symptomatic infants with hemodynamically important lesions and congestive heart failure, palliative resection to relieve inflow or outflow tract obstruction is justified because long-lasting symptomatic relief may be achieved by partial resection. Although this does not protect from arrhythmic death, further definitive treatment may be available at a later stage. For instance, Jamieson et al. (17) recently reported cardiac transplantation for a long-standing unresectable ventricular fibroma in a 20 year old woman.

It is now our policy to study all patients initially with two-dimensional echocardiography and then, if indicated, with cardiac catheterization and angiography. In symptomatic infants, surgery is recommended to correct hemodynamic impairment if the extent of tumor involvement allows this treatment. If echocardiography shows multiple lesions in the absence of symptoms, surgery is not recommended. A solitary lesion amenable to surgical resection should be removed, whether symptomatic or not.
because the overall results for fibroma resection are good (10,11).

Surgical results. Because the majority of surgical reports are of "successful" resection in isolated cases, it is difficult to draw comparisons from other series. Arciniegas et al. (2) recently described four infants with rhabdomyoma who underwent surgical resection. Excision was complete in one solitary pedunculated tumor and partial in the other three. One child died at termination of cardiopulmonary bypass and one is mentally retarded. Two are alive and well, the longest follow-up period being 19 months. The survivors have impressive hemodynamic improvement though only one was symptomatic before operation.

Our study includes three neonates and one 2 year old child with multiple rhabdomyomas. Two patients were symptomatic and three of the four had tuberous sclerosis. There was no autopsy and little opportunity for tuberous sclerosis to manifest itself in one of the neonates (Case 2). Surgical exploration was carried out in one early case, but did not contribute to the child's well being because widespread myocardial nodules were found in addition to the intracavitary tumor; therefore, useful resection was impossible. Death occurred suddenly 3 months later, presumably from arrhythmia because there was no hemodynamic impairment. More detailed preoperative investigation with methods available today would have argued against surgical intervention in this child.

Conclusions. Progress in the safety of cardiac surgery in infants and young children now provides gratifying surgical results after complete resection in a few cases of cardiac tumors. We recommend careful preoperative assessment including plain chest X-ray examination, electrocardiography and two-dimensional echocardiography. Evidence for tuberous sclerosis, hamartomas or a primary neoplasm elsewhere should be sought and pericardial fluid examined for malignant cells when appropriate. If signs and symptoms are such that surgical intervention may prove beneficial, cardiac catheterization and angiography would complete the preoperative evaluation. Endomyocardial biopsy should be considered for the future but may prove to be difficult in small infants. With the benefit of this information, a carefully planned surgical procedure increases the likelihood of a satisfactory outcome.

References