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Primary Intraventricular Cardiac Tumors in Children: Contemporary Diagnostic and Management Options

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SUMMARY. **We reviewed the intraventricular cardiac tumors presenting at our institution between 1985–1991, studying the presentation, modes of investigation, and evidence of hemodynamic compromise. Thirteen patients presented with intraventricular tumors during the study period. Two of the tumors were rhabdomyosarcomas, one was a myxoma, and 10 were rhabdomyomas. All patients were evaluated with two-dimensional and pulsed Doppler echocardiography and B-color imaging was undertaken in three patients. Four patients presented for elective scans to complement investigations for tuberous sclerosis, seven patients had cardiac symptoms, and two patients presented prenatally. Obstruction to intracardiac flow was present in five patients. Two patients had the tumor excised and one had an open biopsy of the tumor. One patient had an transvascular biopsy at cardiac catheter. Early detection of cardiac tumors is increasing, particularly rhabdomyomas. With fetal echocardiography, more patients should come to attention prenatally. B-color may be a useful addition in assessing cardiac tumors, aiding detection and definition of intramural lesions.**

KEY WORDS: **Fetal — Heart — Tumor — Echocardiography — Pediatrics**

Primary cardiac tumors are rare both in infancy and childhood, rhabdomyomas being the most common [4, 11]. Many have been discovered at postmortem. Aided by noninvasive technology, over the last two decades, interest has changed from pathological features to early, noninvasive diagnosis. Our recent experience of primary intraventricular cardiac tumors diagnosed in vivo is reviewed, with special reference to the changing processes leading to diagnosis and altered management implications in some patients.

Methods

The echocardiography database was searched for all patients with the diagnosis of cardiac tumor between January 1985 and May 1991. Those with primary intraventricular cardiac tumors formed the basis for review. Patient case notes and echocardiograms were separately analyzed for site, size, shape, and consequences of the intraventricular tumor (including disturbances to blood flow).

Echocardiograms (ECGs) were performed with an ATL 600 (1985–1987), an ATL UM8 (1988–1989), or an Acuson 128 (1990–1991). Standard subcostal, apical, parasternal, and suprasternal views were performed, with B-color supplementation in the three most recent cases (after May 1991). Pulsed, continuous wave, and color Doppler were used as required. Results were analyzed by both a pediatric echocardiographer and a senior cardiology registrar or fellow. When used, different B-color formats (real-time images displayed with a dynamic compression scale of varying colors) were sequentially superimposed on the two-dimensional image to allow the best differentiation between tumor and normal myocardium.

Obstruction to flow was defined as greater than or equal to 50% of cross-sectional area of the outflow or inflow tract (quantitated as deciles) with associated turbulent flow and/or chamber enlargement or dysfunction. A gradient (mean diastolic at atrioventricular valves and peak instantaneous at semilunar valves) was calculated where possible.

Results

Between January 1985 and May 1991, there were 13 children with primary cardiac tumors located in the

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Table 1. Patient data

Case no.	Age	Presentation	Surgery	Pathology
1	12 years	Right-heart failure	Excision	Myxoma
2[a]	6 years	Syncope	Excision	Rhabdomyo-sarcoma
3	1 week	Arrhythmia	Biopsy	Rhabdomyoma
4	3 years	Tuberous sclerosis	Nil	Rhabdomyoma
5	9 months	Tuberous sclerosis	Nil	Rhabdomyoma
6	fetal	Routine	Nil	Rhabdomyoma
7	2 months	Arrhythmia	Nil	Rhabdomyoma
8	5 years	Tuberculosis	Catheter	Rhabdomyo-sarcoma
9	fetal	Routine	Nil	Rhabdomyoma
10	1 month	Murmur	Nil	Rhabdomyoma
11	2 months	Tuberous sclerosis	Nil	Rhabdomyoma
12	1 month	Left-heart failure	Nil	Rhabdomyoma
13	2 years	Tuberous sclerosis	Nil	Rhabdomyoma

[a] Previous case report [17].

ventricles. Presentations varied considerably depending on the location of the tumor (Table 1). Two patients had heart failure (cases 1 and 12), one had syncope, one had recurrent episodes of supraventricular tachycardia, and four presented for investigation of tuberous sclerosis. Cases 7 and 10 presenting with cardiac abnormalities (arrhythmia and murmur, respectively) were subsequently diagnosed as having tuberous sclerosis. Case 8 presented with tuberculosis and was noted during the admission to have right-heart failure (Figs. 1 and 2). In two cases, prenatal obstetric scanning raised the possibility of a cardiac tumor. This was confirmed by fetal echocardiography and later postnatal echocardiography in our unit.

Tumor appearance on echocardiography varied according to pathology. Well-defined, multiple spherical or ovoid lesions in association with tuberous sclerosis were presumed to be hamartomatous lesions, most probably rhabdomyomas. The two rhabdomyosarcomas were large and irregular. One had evidence of tumor fragmentation, possibly explaining the widespread metastases. The myxoma (case 1) was also irregular and appeared to traverse the tricuspid valve. Table 1 indicates the site of the lesions and the maximal size as seen on the two-dimensional picture.

B-color image processing was used in three patients, all of whom had rhabdomyomas. Fig. 3 compares the images with and without B-color.

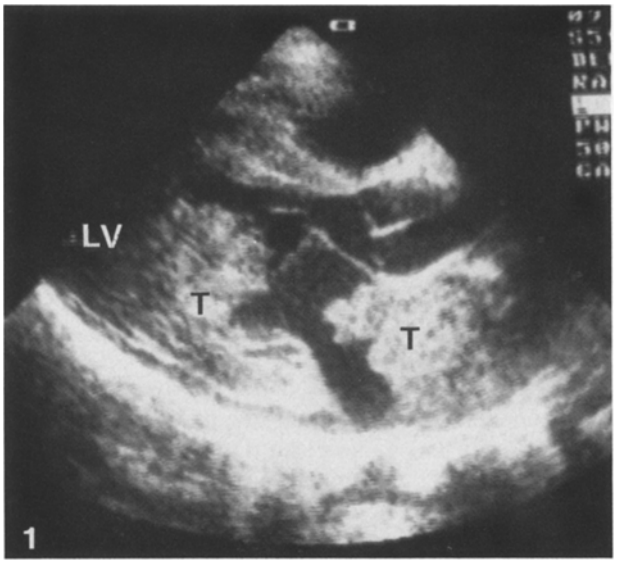

Fig. 1. A parasternal long-axis view showing large irregular tumor (T) occupying most of the left ventricle and left atrium (case 8).

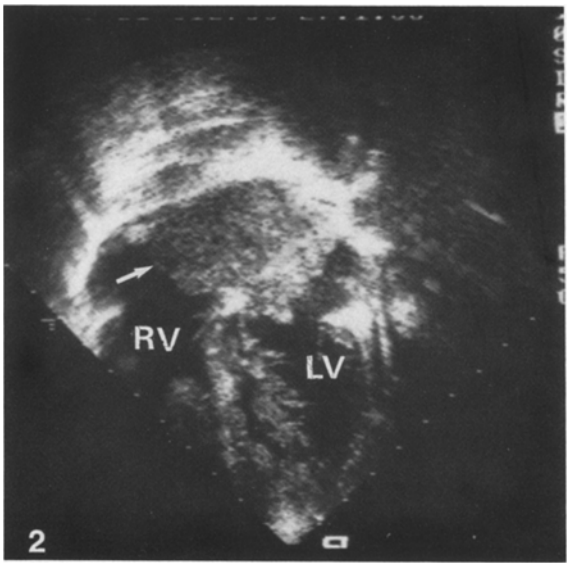

Fig. 2. An apical four-chamber view showing tumor masses in both atria and ventricle (case 8).

Obstruction was present in five patients (Table 2): two involved the left ventricular outflow, one the left ventricular inflow, one the right ventricular outflow, and one the right ventricular inflow with the lesion crossing the tricuspid valve. The severity of effects varied from 50–59% obstruction of the left ventricular outflow tract (case 2) to nearly complete obstruction, with a gradient of 65 mmHg (case 12). The remainder had no demonstrable hemodynamic

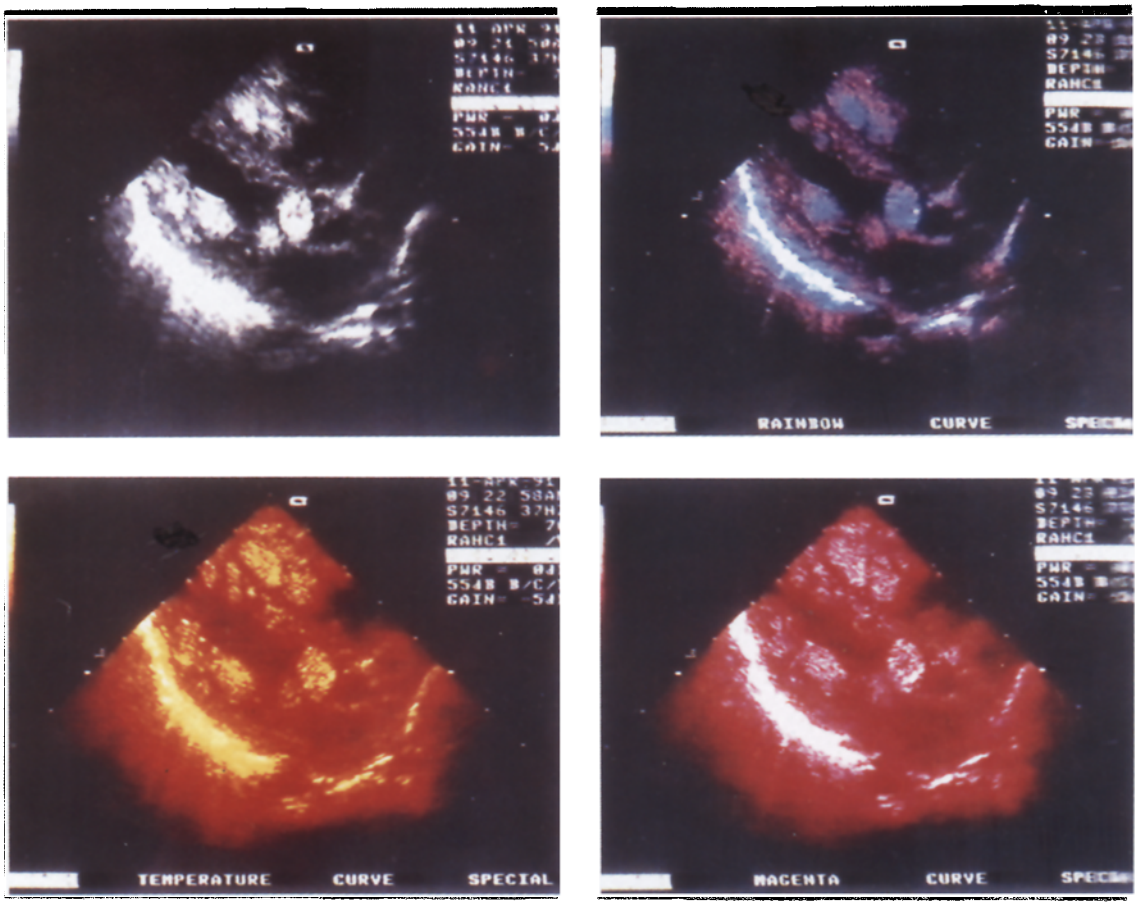

Fig. 3. Parasternal long-axis view. B-mode color accentuates small lesions in interventricular septum. The left ventricular outflow tract is nearly completely occluded by tumor (case 12).

effect, despite some of the lesions occupying up to 50% of the ventricular cavity.

Surgery was performed in three patients. Cases 1 (myxoma) and 2 (rhabdomyosarcoma) were extensive lesions that caused significant hemodynamic compromise. In case 3 (single rhabdomyoma), surgery was performed for pathological diagnosis. Operative findings closely correlated with echocardiographic findings (Table 2). In case 1, there was a multilobar gelatinous tumor occupying approximately two thirds of the right atrial cavity and virtually all of the right ventricular cavity. In case 2, there was a large yellow fleshy tumor occupying most of the left ventricular cavity with origins from the posterior chordal apparatus of the mitral valve. It involved the posteromedial papillary muscle and chordae as suspected on the preoperative echocardiogram. In case 3, there was an almond-shaped sessile firm pale structure 3.0 × 2.0 × 1.5 cm located in the infundibular wall. There was a second similar mass, approximately 1 cm in diameter, palpated posterobasally near the atrioventricular groove.

The two patients with rhabdomyosarcomas died from metastases at 3 years and 2 months, respectively, after diagnosis. Four other patients have had sequential echocardiograms—a reduction in lesion size was seen in two patients (cases 4 and 10). No change was seen at 2 years in case 6. A further study at 3 months in case 12 has shown no difference in the size of the lesion but enlargement of the left ventricular outflow tract (relative reduction in size).

Discussion

The value of echocardiography in diagnosing cardiac tumors has been recognized, particularly over the last decade [2, 3, 10]. With better image resolution and the addition of Doppler, the need for cardiac catheterization—which exposes the patient to the risks of an invasive procedure, tumor hemorrhage, and embolization—has decreased. Echocardiography readily demonstrates the site, size, and hemodynamic consequences of the lesion, often allowing management decisions without invasive in-

Table 2. Echocardiography reports

Case no.	No. lesions	Site[a] (Size[b])	Abnormal function (ventricular and valve)	Obstruction site (% / mmHg)
1	1	RA, RV (75%)	Septal motion TV not seen	RVI /70–79/ 0
2	1	LV (50%)	—	LVO /50–59/ 0
3	2	RVO (1.2 × 2.2 cm)	? TV involved	—
4	2	RV (1.2 × 0.6 cm)	—	—
5	1	RVO (6.0 × 6.0 cm)	—	—
6	1	IVS (2.0 × 1.5 cm)	—	—
7	Multiple	LV, RV (50%)	—	—
8	Multiple	RV, RA LV, LA (50%)	—	LVI /70–79/ 6
9	Multiple	RV, LV (1.3 × 1.6 cm)	—	—
10	Multiple	RV, LV (2.5 × 1.8 cm)	—	RVO /90–99/ 20
11	Multiple	IVS (1.1 × 1.1 cm)	—	—
12	Multiple	LV, RA, RV, IVS (0.8 × 0.5 cm)	—	LVO /90–99/ 65
13	1	LV (0.7 × 1.5 cm)	—	—

FS, fractional shortening; LV, left ventricle; LVO, left ventricular outflow; LVI, left ventricular inflow; LA, left atrium; RV, right ventricle; RVO, right ventricular outflow; RVI, right ventricular inflow; RA, right atrium; TV, tricuspid valve; IVS, intraventricular septum.

[a] Dominant chamber involvement underlined.

[b] Maximal size of lesion on two-dimensional image or area of chamber when irregular.

vestigation. Of our 13 cases, in only one was a cardiac catheterization performed (to obtain tissue diagnosis). Of the three cases that underwent surgery, there was close echocardiographic and surgical anatomical correlation.

Reports in the literature of cases with significant ventricular hemodynamic obstruction diagnosed in vivo are uncommon, though there have been several individual case reports [8, 12, 17]. Of our series of 13 patients, five showed either impaired ventricular filling or outflow obstruction (Table 2). Depending on the site and significance of obstruction, presentations may vary considerably with varying degrees of right- and left-heart failure. In our study, some patients improved with simple medical measures, or had spontaneous reduction in actual or relative size (rhabdomyomas) [5]. Some tumors (more commonly malignant) required surgery [7, 9, 13].

Fetal ultrasound is a valuable tool in diagnosing congenital cardiac disorders in utero [1, 15]. Similarly, cardiac tumors can be detected at an early stage and followed, as with cases 6 and 9 [6, 16, 18]. The wider use and availability of fetal echocardiography should lead to increasing numbers of cases presenting prenatally. In many, this may prove at first to be a diagnosis of tuberous sclerosis.

B-color appeared to augment the identification and definition of the cardiac tumors. The human eye has a lesser capacity to detect varying degrees of gray scale than color scale images. B-color is currently being investigated in the differentiation between normal and abnormal tissue. We have found that B-color assisted in tumor edge definition and small tumor recognition. This was especially the case with intramural lesions surrounded by normal tissue. It may be that large areas of irregular myocardial fiber orientation are accentuated by this

technique or that inherent tissue characteristics are highlighted.

We have found that ventricular obstruction is common among primary cardiac tumors located in the ventricles (especially the malignant ones). It also seems likely that an increasing number of patients will come to attention prenatally because of an increased awareness of prenatal cardiac evaluation. B-color may prove to be a useful addition in detecting and defining the extent of such cardiac tumors.

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