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Echocardiography of Intracardiac Filling Defects in Infants and Children

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SUMMARY. **Intracardiac masses are rare in infants and children. Early detection is essential to their successful management. We present seven patients in whom echocardiography established the diagnosis and was crucial in the management. Three of the masses were primary cardiac tumors and four were thrombi. *Patient 1*: an infant with a calcified left ventricular fibroma. *Patient 2*: a neonate who presented with cyanosis due to obstruction of the right ventricular inflow tract by a fibroblastic tumor. *Patient 3*: an infant with a right atrial myxoma presenting as sepsis. *Patient 4*: a child who had a pulmonary embolus after a pulmonary valvotomy and was found to have a right ventricular thrombus. *Patient 5*: a child with a right atrial thrombus following a Fontan procedure for univentricular atrioventricular connection. *Patient 6*: a child with a left ventricular thrombus due to a dilated cardiomyopathy in association with epidermolysis bullosa. *Patient 7*: An infant with bilateral lobar emphysema, an aorticopulmonary window with left ventricular fibroelastosis, who developed a left ventricular thrombus.**

KEY WORDS: **Echocardiography — Intracardiac filling defect**

Intracardiac filling defects are rare in infants and children. With advances in diagnostic [8, 11, 14, 24] and surgical techniques, the outlook for these patients has been markedly improved [2, 3, 17, 23, 27]. Awareness of the possibility of this diagnosis in an individual is paramount in making the diagnosis. We report seven cases in whom the diagnosis was made by echocardiography.

Case Reports

Case 1

A ten-month-old infant girl presented with a one-month history of persistent cough. She was mildly tachypneic and on auscultation of the heart there was a grade-2/6 pansystolic murmur at the cardiac apex and at the left sternal edge a pericardial friction rub.

The chest x-ray showed a large mass in the left hemithorax with extensive calcification. The electrocardiogram showed sinus rhythm with deep Q waves in leads V_5 and V_6; 24-h ECG monitoring revealed no arrhythmias. Echocardiography (Fig. 1) showed the presence of a moderate pericardial effusion and a mass was seen, involving the free wall of the left ventricle with extension superiorly onto the right ventricular outflow tract and pulmonary artery. There appeared to be some involvement of the posterior papillary muscle of the mitral valve and on Doppler interrogation mitral regurgitation was detected.

At left thoracotomy, the echocardiographic findings were confirmed. Radical excision was not feasible. Multiple biopsies were taken.

Postoperatively, a 24-h ECG recording showed frequent runs of ventricular tachycardia for which the infant was started on propranolol. She is thriving and asymptomatic six months after the surgery and a repeat 24-h ECG recording shows partial suppression of the arrhythmia.

Histology. Microscopically the tumor consisted of dense hypocellular, poorly vascularized fibrous connective tissue containing heavy deposits of calcium and elastic fibers. The tumor cells had an ultrastructural appearance of fibroblasts. These findings are consistent with a fibroelastic hamartoma (fibroma).

Case 2

An infant girl, weighing 2740 g, presented on the first day of life with central cyanosis. On examination, there was moderate cyanosis with no signs of heart failure and no murmurs were heard.

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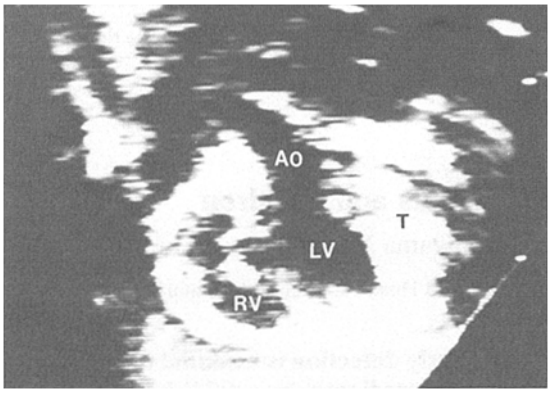

Fig. 1. Subcostal coronal section showing a large echo-dense mass arising from the left ventricle. Echocardiogram recorded on an Advanced Technology Laboratories Mark 600 mechanical sector scanner. *Ao*, aorta; *LV*, left ventricle; *RV*, right ventricle; and *T*, tumor.

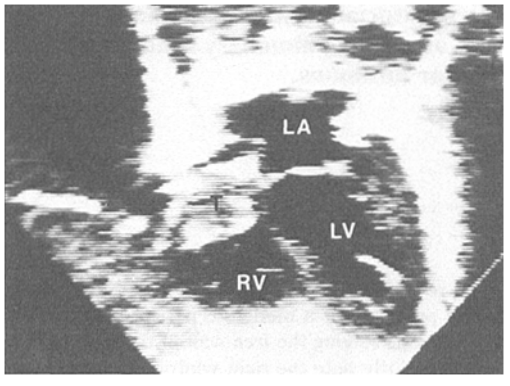

Fig. 2. Subcostal coronal section showing a filling defect in the right atrium. Echocardiogram recorded as in Fig. 1. *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle; and *T*, tumor.

The chest x-ray revealed a normally sized heart with oligaemic lung fields. The ECG was normal with sinus rhythm.

Echocardiography (Fig. 2) demonstrated an echo-dense mass in the right atrium. It was obstructing right ventricular inflow and the right ventricle was small. Doppler interrogation of the pulmonary artery demonstrated continuous flow from the ductus arteriosus and indicated that there was very little flow through the pulmonary valve. There was left-to-right shunting at the atrial level.

Cardiac catheterization confirmed the presence of a filling defect in the right atrium. The right ventricle was small and right ventricular systolic pressure was 40 mmHg.

A right Blalock–Taussig shunt was performed. She then thrived and at the age of six months underwent elective exploratory surgery on cardiopulmonary bypass.

A hard mass was found in the right atrium and this was obstructing the tricuspid valve. The mass was partially excised, relieving the obstruction but leaving some tissue in proximity to the AV node. The Blalock–Taussig shunt was left intact.

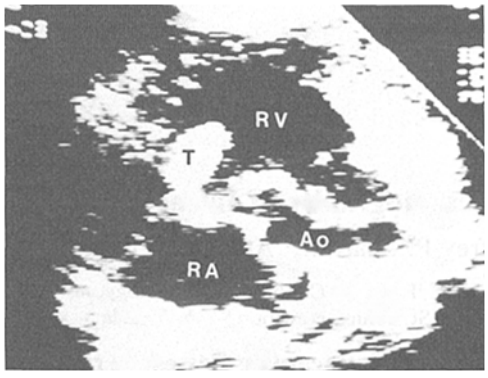

Fig. 3. Parasternal short-axis section in diastole showing the filling defect prolapsing into the right ventricle. Echocardiogram recorded as in Fig. 1. *Ao*, aorta; *RA*, right atrium; *RV*, right ventricle; and *T*, tumor.

Six months after the surgery, echocardiography showed that the right ventricle had increased in size and Doppler interrogation indicated normal flow through the pulmonary valve with minimal diastolic flow through the Blalock–Taussig shunt.

Histology. The tissue consisted of a hypercellular, highly vascular tumor composed of spindle-shaped, mildly pleomorphic cells arranged in whorls surrounding blood vessels. Less cellular areas with dense collagen were present with collections of foamy macrophages. A diagnosis of a fibroblastic tumor which could not otherwise be subclassified was made.

Case 3

A three-month-old infant boy was admitted with a two-day history of fever, feeding difficulty, irritability, and lethargy. The CSF had 38 × 10 WBC/liter, 66% being polymorphs, a protein of 0.71 g/liter, and a sugar of 3.5 mmol/liter. The Gram stain was negative and subsequent culture was sterile. A blood culture did however grow *Staphylococcus aureus*. Examination of the cardiovascular system was normal except for a loud high-pitched first heart sound. He was treated with intravenous antibiotics for two weeks with clinical improvement. However, the white count increased and a thrombocytosis developed. A gallium scan was performed and was negative. An echocardiogram revealed a mass in the right atrium attached to the septal leaflet of the tricuspid valve and this mass was seen to pass into the right ventricle in diastole (Fig. 3). This finding was confirmed at surgery and the mass was completely removed.

Histology. The mass consisted of foci of myxoid tissue with stellate cells present compatible with the diagnosis of a myxoma. A large proportion of the tissue was composed of fibrin with some organization. The antibiotics were continued for six weeks and no recurrence has been seen after six months.

Case 4

An eight-year-old boy underwent an uneventful pulmonary valvotomy for pulmonary valve stenosis. On the seventh postopera-

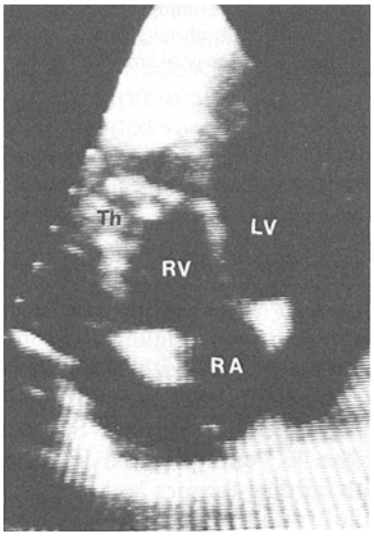

Fig. 4. Apical four-chamber section showing a filling defect in the right ventricle. Echocardiogram recorded as in Fig. 1. *LV*, left ventricle; *R*, right atrium; *RV*, right ventricle; and *Th*, thrombus.

tive day, he had severe abdominal pain and was peripherally cyanosed with a tachycardia, tachypnea, and hypotension. He then suffered a cardiac arrest. Subsequent x-rays of the abdomen and chest were unremarkable. An echocardiogram (Fig. 4) revealed the presence of a small pericardial effusion and a filling defect in the right ventricle which was thought to be a thrombus. A lung scan revealed the presence of multiple filling defects consistent with the diagnosis of pulmonary emboli.

Two more hypotensive episodes occurred despite heparin therapy and the lung scan showed further filling defects. In view of the recurrences, the heparin was stopped and streptokinase started; 48 h later the lung scan showed a marked improvement and the echocardiogram showed no evidence of a thrombus in the right ventricle. Two weeks later, he was off all therapy and cardiac catheterization was performed. Pulmonary angiography showed a decrease in perfusion to the left lower lobe, but the pulmonary artery pressure was normal and no filling defects were seen in the right ventricle. He was reviewed three months later and was asymptomatic with no evidence of pulmonary hypertension. Unfortunately a few weeks later he died in a house fire.

Case 5

A ten-year-old boy at the age of seven years underwent a modified Fontan procedure for a univentricular heart with two AV valves. Initially he was well, but subsequently became more edematous and was found to have a protein-losing enteropathy. An echocardiogram (Fig. 5) done 27 months after the Fontan procedure showed a large filling defect in the right atrium near the junction with the superior vena cava. This was thought to be due to a thrombus. There was no evidence that this was causing pulmonary outflow obstruction and a lung scan was normal. He was started on aspirin and persantin, but three months later the

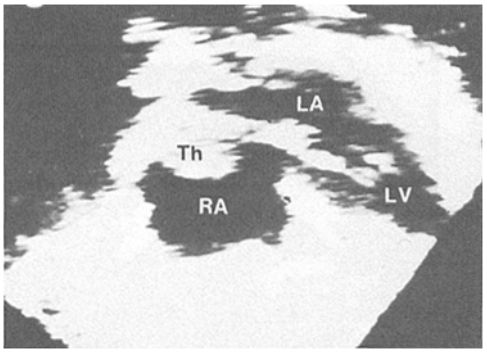

Fig. 5. Subcostal coronal section showing a filling defect in the right atrium. Echocardiogram recorded as in Fig. 1. *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; and *Th*, thrombus.

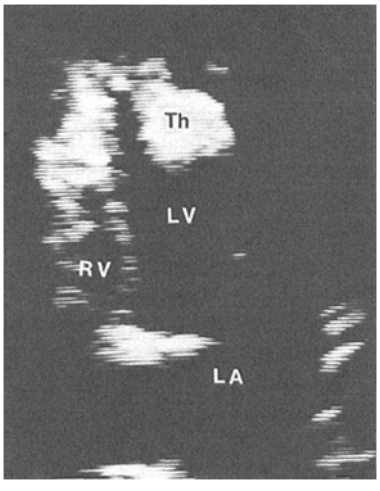

Fig. 6. Apical four-chamber section showing a filling defect at the left ventricular apex. Echocardiogram recorded as in Fig. 1. *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle; and *Th*, thrombus.

echocardiogram showed the development of a second smaller thrombus near the junction of the right atrium and the inferior vena cava. He was then started on warfarin and the aspirin was stopped. The echo subsequently showed resolution of the second thrombus. The initial thrombus became smaller. He currently remains well with some resolution of the protein-losing enteropathy and a reduced diuretic requirement.

Case 6

A seven-year-old boy had epidermolysis bullosa of the dystrophic type causing extensive skin lesions with scarring. At the age of five years, echocardiography showed that the left ventricle was dilated and had reduced contractility consistent with a dilated cardiomyopathy.

Two years later he was admitted with a sudden onset of left hemiparesis and a seizure. Echocardiography (Fig. 6) again

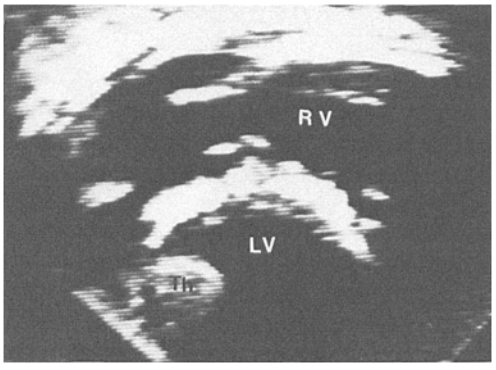

Fig. 7. Subcostal coronal section showing a filling defect in the left ventricle. Echocardiogram recorded as in Fig. 1. *LV*, left ventricle; *RV*, right ventricle; and *Th*, thrombus.

showed that the left ventricle was dilated and at the apex there was a large mobile filling defect. The platelet count was elevated at 857 × 10/liter and the fibrinogen was elevated at 7.0 g/liter. The PT, PTT, and levels of factors 5, 8, 11, and 12 were normal. It was felt that there was imminent risk of further embolization and so the thrombus was surgically removed. The postoperative recovery was uneventful apart from a further seizure.

Despite treatment with heparin and then warfarin, echocardiography demonstrated the development of further intracardiac thrombus. The warfarin was discontinued and he was treated with aspirin and persantin. No further thrombi were noted and no emboli occurred so he was discharged. He died suddenly a few weeks later. At autopsy, there were multiple infarcts in the brain and extensive thrombus and fibrosis in the left ventricle. Histology of the myocardium was nonspecific with no evidence of myocarditis.

Case 7

An infant boy died at the age of four months of bilateral congenital lobar emphysema, an aorticopulmonary window, and left ventricular endocardial fibroelastosis.

He was in severe respiratory distress at birth due to emphysema of the right, middle, and left upper lobes of the lung.

Echocardiography at that time showed considerable distortion of cardiac structures and great vessels. The heart was rotated to the left and superiorly with the right ventricle lying superior to the left ventricle. Ventricular function was normal. There was some narrowing of the left ventricular outflow tract, but Doppler interrogation suggested that there was no gradient.

Resection of the emphysematous lobes resulted in some improvement in ventilatory status, but he remained ventilator dependent. At two weeks, echocardiography showed very poor function of both ventricles.

Because of chalasia and aspiration, intravenous alimentation was used, initially by peripheral vein but, after four weeks, a central venous line was inserted.

Echocardiography (Fig. 7) at six weeks showed that both ventricles were continuing to function very poorly and a large filling defect was seen in the left ventricle. In view of the infant's severe respiratory status, no thrombolytic therapy was given and over the following month the echocardiogram showed gradual spontaneous resolution of the filling defect. However, he then suffered a cardiac arrest and, although initially resuscitated, he died a few days later. At autopsy, a small residual thrombus was found in the left ventricle.

Discussion

Cardiac tumors are rare in infants. In 1965, fifteen European cardiac centers began a collaborative study of tumors in infancy and childhood and collected 22 cases of which only nine were listed as being less than one year old [25]. Boston Children's hospital had three infants with symptomatic tumors out of 11,000 autopsies performed between 1914 and 1966 [15]. The Hospital for Sick Children in Toronto in the 46 years prior to 1972 had seen 11 children with cardiac tumors and five of these were in infancy [26]. The three cases reported here were all seen in a 12-month period. Prior to this, only one other case, a left ventricular fibroma, was found in the hospital autopsy records since the institution opened in 1966. We have no evidence to suggest that this occurrence was anything other than coincidental.

Cardiac tumors may present with features of obstructed blood flow, which may result in cyanosis as in case 2. More commonly, however, this obstruction, rather than being in the right atrium, is of the right [13, 25] or left ventricular outflow tracts [25]. Arrhythmias are frequent and may be the presenting features even with small tumors [16]. In case 1, the arrhythmia was only detected by 24-h ECG monitoring and it illustrates the value of this investigation in such patients. Alternatively the tumor may be revealed as a mass on chest x-ray as in case 1.

In case 1, the histologic diagnosis is that of a fibroma. However, some of the features are unusual. The observation of extensive calcification in the mass is more common with teratoma [25], but in a less conspicuous form has been reported infrequently in cardiac fibroma [1, 2, 25]. Pericardial effusion may occur [19], but is unusual for a "benign" fibroma, and its presence usually implies a pericardial origin to the tumor or malignant change [6].

Cases 2 and 3 are very unusual in that right atrial tumors in infancy are rare and we could find only seven such cases in the English medical literature. Only four of these were neonates. Of the latter, one was a calcified myxoma [5], two rhabdomyomata [14, 22], and one a teratoma [25]. Ilbawi et al. [9] reported a hemangioendothelioma in a six-week-old infant, Sanyal et al. [21] a myxoma in a seven month old, and Rutkowski et al. [19] an atrial fibroma in a ten week old. In addition, the association of bacteremia with a myxoma, as in case 3, is in

itself an unusual event [10, 12, 18]. The vegetations of infective endocarditis are not infrequently seen as filling defects on echocardiography, but usually in the context of structural heart abnormalities, and we have not included such cases in this report.

The occurrence of thrombus in the heart of a child is a rare event and its presence, if undetected, is life-threatening. It is a well-recognized complication of central venous catheterization [4], but presumably the thrombus in the left ventricle in case 7 was unrelated to this. The likely cause was that of the poor left ventricular function in the presence of fibroelastosis. Since the widespread use of two-dimensional echocardiography, left ventricular thrombus is an increasingly recognized complication of dilated cardiomyopathy [11] and myocarditis [14] in children, and case 6 is a further example. This case is, however, unusual in that we are unaware of previous reports of the combination of dilated cardiomyopathy and epidermolysis bullosa. The role of the thrombocytosis and hyperfibrinogenemia in the formation of the thrombus in this case is also undetermined.

The presence of a right atrial thrombus in a patient after a Fontan procedure is of great concern and would have been unsuspected if it were not for the routine use of echocardiography in these patients. In this particular case (case 5), it did not seem to be producing any hemodynamic disturbance and there has been no evidence to suggest embolization. Two-dimensional echocardiography has been demonstrated to be a very useful tool in the detection of right-sided thrombus in adults with pulmonary emboli [7, 20]. In children, pulmonary embolization is a very unusual event and, when it does occur, is most frequently on the basis of intracardiac thrombus. In case 4, the echocardiographic demonstration of a thrombus in the right ventricle was a crucial factor in making the diagnosis and the subsequent successful therapy.

The essential prerequisite for making the diagnosis of an intracardiac filling defect is awareness of the possibility. These cases illustrate that the diagnosis can be made by echocardiography. Tissue characterization is not possible from the echo alone. However, in all cases except case 3, by interpreting the echo in the clinical context, a confident distinction between tumor and thrombus was made. In case 3, there was some doubt about the pathology, but myxoma was suspected on the basis of the characteristic sessile mobility of the filling defect. Myocardial extension of the filling defect, as in case 1, is very much in favor of tumor and, if multiple defects are found, rhabdomyoma is the likely diagnosis.

It is essential that routine screening be done, not only for the more well-recognized indications such as unusual or intractable arrhythmias, syncope, or cyanosis, but for cases where embolization is at all a possible explanation for the clinical presentation, whenever undisclosed sepsis may be present, when central venous lines are used, and routinely in the follow-up of patients with cardiomyopathy or myocarditis.

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