The Heart Surgery Forum #2004-1142
8 (2), 2005 [Epub March 2005]
doi:10.1532/HSF98.20041142

Online address: www.hsforum.com/vol8/issue2/2004-1142.html

Noncompaction of Ventricular Myocardium in a Patient with Congenitally Corrected Transposition of the Great Arteries Treated Surgically: Case Report

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ABSTRACT

Noncompaction of the ventricular myocardium is a rare disorder that represents numerous prominent trabeculations and intratrabecular recesses in the ventricles. It is believed to represent not only an arrest in endomyocardial morphogenesis but also an unclassified cardiomyopathy. The pathology has been almost invariably associated with other congenital cardiac malformations. A female patient with noncompaction of the myocardium of both ventricles and congenitally corrected transposition of the great arteries (cTGA), situs inversus totalis, and atrial and ventricular septal defects is described. When she was 7 days old a permanent pacemaker was implanted because of complete heart block. Prazosin (Minipress), an α-receptor blocker, was administered, and the cardiac ejection fraction showed a striking increase from 20% to 42%. Despite careful and regular follow-up evaluations, the general condition of the patient slowly worsened. Five months after surgery she died of hepatorenal failure and low cardiac output. This case report is thought to be the first description of congenital complete heart block, cTGA, and situs inversus totalis with noncompaction of the myocardium of both ventricles.

INTRODUCTION

During the early phase of embryonic development, ventricular myocardium consists of a trabecular network. The intratrabecular spaces communicate with the ventricular cavity. As the heart develops, the myocardium becomes compacted, and the meshlike pattern disappears. Noncompaction of ventricular myocardium (NVM) is accepted as representing an arrest in endomyocardial morphogenesis. Today NVM is known as an unclassified cardiomyopathy characterized by excessively prominent trabeculations and deep intratrabecular recesses in the left ventricle, at times affecting the right ventricle and interventricular septum as well [Richardson 1996].

Noncompacted myocardium was previously described as persistent intramyocardial sinusoids or spongy myocardium. These conditions, however, are generally associated with congenital obstructive lesions of the left or right ventricular outflow tract, such as pulmonary atresia with intact ventricular septum and aortic atresia [Dusek 1975]. During ontogenesis, regression of the embryonic sinusoids is impaired by ventricular pressure overload that results in deep recesses that communicate with both ventricular cavities and the coronary artery system in these patients [Grant 1926, Chenard 1965, Williams 1951, Feldt 1969, Lenox 1972, Dusek 1975]. Noncompaction of ventricular myocardium can be seen as an isolated malformation [Chin 1990, Ichida 1999, Oechslin 2000] or as associated with other congenital cardiac malformations.

We report a case of myocardial noncompaction involving both ventricles. The originality of the report is that the patient had congenital complete heart block in addition to morphologic anomalies consistent with congenitally corrected transposition of the great arteries (cTGA), situs inversus totalis, atrial septal defect (ASD), and ventricular septal defect (VSD) corrected surgically.

CASE REPORT

The patient was first seen when she was 7 days old. The electrocardiogram (ECG) revealed third-degree heart block with a heart rate of 70 beats/min. The chest x-ray showed situs inversus totalis and an enlarged heart with a cardiothoracic ratio of 0.65. Two-dimensional echocardiography (2D-ECHO) showed situs inversus totalis, cTGA associated with perimembranous VSD, and fossa ovalis type small ASD. A permanent pacing wire was implanted via the subxiphoid approach, and the pulse generator was left in the right retroperitoneal space. One month later, a pulmonary banding operation was performed because signs of a progressive rise in pulmonary pressure were found at 2D-ECHO.

When the patient was 2 months old, 2D-ECHO showed minimal left atrioventricular (AV) valve insufficiency, a 30-mm Hg gradient at the pulmonary artery because of banding, and a VSD 4 mm in diameter.

Received October 19, 2004; received in revised form January 1, 2005; accepted January 7, 2005.

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Table 1. First Catheterization Data

Chamber	Pressure, mm Hg	O_2 Saturation, %
Main pulmonary artery	27	95
Morphologic left ventricular apex	68	63
Morphologic right ventricle	92	96
Aorta	90	-
Right atrium	2	56
Left atrium	-	96
Superior vena cava	-	50

One year later, 2D-ECHO revealed an enlarged left atrium and morphologic right ventricle, which give rise to the aorta. The systolic function of the morphologic right ventricle was depressed with fractional shortening of 18%. There was prominent trabeculation of both ventricular apexes. The data recorded in the catheterization laboratory were as follows: pulmonary artery (PA) pressure, 27/7 mm Hg (mean, 19 mm Hg); morphologic left ventricular apex pressure, 68 mm Hg; morphologic right ventricular pressure, 92 mm Hg; and aortic pressure, 90/45 mm Hg (mean, 64 mm Hg) (Table 1). The patient had second-degree left and minimal right AV valve insufficiency and a 41-mm Hg gradient at the level of pulmonary banding. VSD was found to be closed owing to pulmonary banding. The patient was evaluated at 6-month intervals. When she was 4 years old, the patient was admitted because of exertional dyspnea and fatigue. She had cardiomegaly with a cardiothoracic ratio of 0.68. The ECG showed nothing abnormal because of the presence of the pacemaker. 2D-ECHO revealed an enlarged left atrium and morphologic right ventricle. The systolic function of the morphologic right ventricle was depressed with fractional shortening of 12% and an ejection fraction (EF) of 40%. The left AV valve insufficiency had increased to third/fourth degree, and the pulmonary banding gradient was 50 mm Hg. Prominent trabeculations were clearly seen in both ventricular apexes (Figure 1). In the apical 4-chamber view marked trabeculations and recesses were seen easily in the apex of the morphologic right ventricle. A Doppler echocardiogram showed typical forward and reversed flow between these trabeculations (Figure 2). Results of catheterization were as follows: PA pressure, 48/24 mm Hg (mean, 34 mm Hg); morphologic left ventricular apex pressure, 100 mm Hg; morphologic right ventricular pressure, 90 mm Hg; and aortic pressure, 90 mm Hg (Table 2). Third-degree left AV valve insufficiency and minimal aortic valve insufficiency were observed. The patient underwent open heart surgery. Primary pulmonary arterioplasty was performed after the pulmonary band was removed. Left AV valve replacement was performed with the use of a no. 25 St. Jude mechanical valve. Initially the patient tolerated the operation well. However, her general condition became worse after the second postoperative day. The liver enlarged, and the chest x-ray revealed pulmonary edema. The patient was treated with digoxin, diuretics (furosemide), and an angiotensin-converting enzyme (ACE) inhibitor (Captopril), but her condition did not improve. On the fifth day, inotropic support (intravenous dopamine and dobutamine) was started. 2-D ECHO revealed morphologic right ventricular dysfunction with an EF of 20%, myocardial noncompaction without residual pulmonary gradient, and mechanical valve dysfunction. With optimal doses of dopamine and dobutamine, arterial tension decreased gradually. Inotropic agents were stopped, and prazosin, an α-blocker, was added to the therapy (0.05 mg/kg body weight) to increase pulsatile force, peak velocity, and circulating norepinephrine levels and to reduce afterload. The dose of prazosin was increased slowly from 0.5 mg/kg to 0.25 mg/kg, and the patient's general condition improved. EF increased from 20% to 42%, a level that cor-

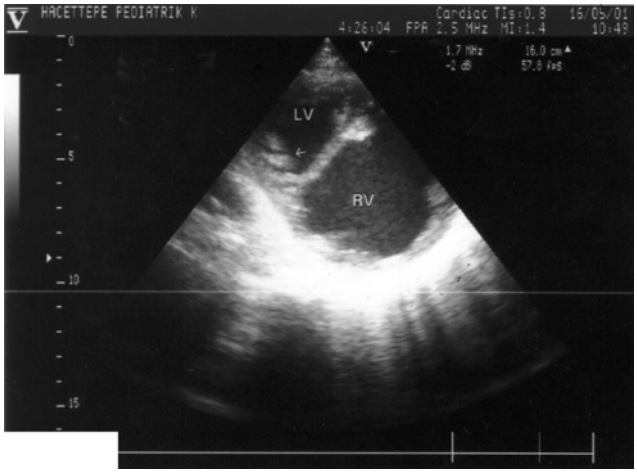

Figure 1. Two-dimensional echocardiogram shows prominent trabeculations and intratrabecular recesses in the apex of the morphologic left ventricle (LV) in the parasternal short-axis view. RV indicates right ventricle.

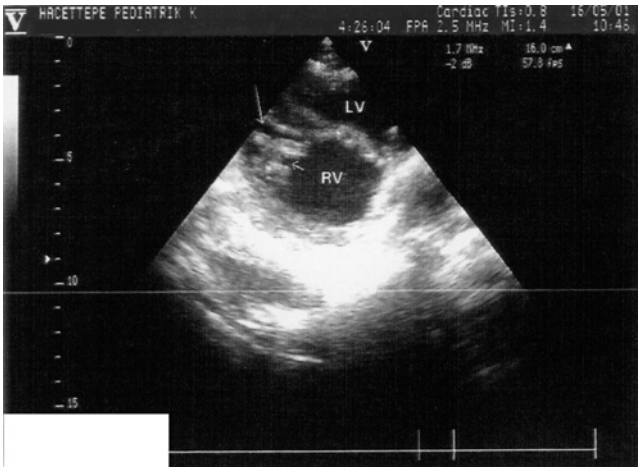

Figure 2. Apical 4-chamber view shows marked trabeculations and recesses in the apex of the morphologic right ventricle (RV). Doppler echocardiogram shows typical forward and reversed flow between these trabeculations. LV indicates left ventricle.

Table 2. Second Catheterization Data

Chamber	Pressure, mm Hg	O2 Saturation, %
Main pulmonary artery	48	53
Morphologic left ventricular apex	100	51
Morphologic right ventricular apex	90	98
Aorta	90	97
Right atrium	-	50
Left atrium	-	98
Superior vena cava	-	50

responded to a 70% to 110% increase in ventricular contraction. The patient was discharged from the hospital on the 12th postoperative day undergoing treatment with digoxin, captopril, warfarin sodium, and furosemide. She was readmitted to the hospital 1 year postoperatively with signs of hepatitis. 2D-ECHO showed an EF of 38%. The laboratory results revealed acute cytomegalovirus (CMV) hepatitis. The result for CMV immunoglobulin M was positive. The levels of hepatic enzymes dramatically increased in a progressive manner, and acute tubular necrosis developed, necessitating peritoneal dialysis. The patient died of septicemia and multiorgan failure despite supportive therapy. Permission for postmortem examination was not granted by the parents.

DISCUSSION

The trabecular structure of the ventricular myocardium fuses and begins to regress in the fifth to eighth weeks of fetal life [Richardson 1996, Dusek 1975]. This process is isochronal with development of the coronary circulation. NVM is caused by a block during this period in which there is continuity of the embryonic myocardial sinusoidal structure after birth and even in adult life. Noncompaction primarily affects the left ventricle, but the right ventricle can be affected as well [Jenni 1986, Chin 1990, Ritter 1997, Ichida 1999, Oechslin 2000]. However, the right ventricle cannot be affected alone. This disease has been reported to occur in any age group (from 1 week to 71 years) [Ichida 1999, Oechslin 2000]. Asymptomatic cases have normal systolic function and increased left ventricular end-diastolic pressure similar to that of restrictive cardiomyopathy. Except for symptomatic cases in children reported by Chin et al [1990], most cases have been initially asymptomatic. When patients initially manifest heart failure, arrhythmia, or embolic events, the clinical course is rapidly progressive. Ventricular and supraventricular tachyarrhythmias and conduction disturbances can be seen in patients with NVM [Jenni 1986, Chin 1990, Ichida 1999, Oechslin 2000]. The complete AV block in our patient was congenital.

The diagnosis of NVM may be delayed because of similarities between NVM and other cardiomyopathies. Prominent multiple myocardial trabeculations and intratrabecular spaces within this trabecular structure, which is continuous with the ventricular cavity, are typical echocardiographic findings of NVM. Most commonly, the apical and midventricular segments of both inferior and lateral walls are affected [Ichida 1999, Oechslin 2000]. Oechslin and colleagues [2000] stated that determination of x-to-y ratio is helpful, but end-diastolic differentiation between noncompacted and compacted myocardium is difficult. Therefore they proposed identifying the segment with a maximal wall-thickness ratio between the noncompacted and the compacted layers. A ratio of noncompaction/compaction >2 is accepted as diagnostic [Oechslin 2000]. Ozkutlu et al [2002] described 12 patients with ventricular noncompaction identified at echocardiographic examinations. Five patients had isolated abnormality, and 3 patients had subnormal left ventricular systolic function. The authors reported that noncompaction of the left ventricle was associated with a complex congenital heart defect in 3 patients. Our patient's findings were suitable for these criteria. Computed tomography, magnetic resonance imaging, ventriculography, and endomyocardial biopsy also can be used as diagnostic tools.

Familial occurrence of NVM in the pediatric population has been described. In all reported cases the patients were boys. This finding strongly suggests X-linked recessive inheritance of this disorder [Chin 1990, Bleyl 1997, Ichida 1999, Oechslin 2000]. Because of the risk of familial recurrence, first-degree relatives of our patient underwent echocardiographic assessment, and there were no positive findings.

Treatment of myocardial noncompaction is not different from that of other cardiomyopathies. Medical treatment is preferred at the beginning of congestive heart failure. Diuretics, ACE inhibitors, and digitalis frequently are preferred for ventricular failure due to ventricular noncompaction.

Using ^{123}I-metaiodobenzylguanidine (a radiolabeled noradrenaline analogue taken up by the sympathetic neuronal terminals) before carvedilol, a mixed α- and β-blocker with antioxidant properties, Toyono et al [2001] found sympathetic nerve dysfunction in noncompacted areas in a patient with isolated left ventricular noncompaction. They also found preserved myocardial viability with the use of scintigraphy but compromised oxidative fatty acid metabolism in the areas of noncompaction. The investigators found a beneficial effect of carvedilol on global and regional left ventricular function, left ventricular mass, and scintigraphic findings in a patient with severe myocardial failure. A similar effect was seen in our patient, who was treated with an α-blocker. Our findings showed a striking decrease in end-systolic and end-diastolic pressures of the noncompacted ventricle and an increase in EF from 20% to 42%. This positive effect may be a slow decrease in ventricular afterload because of the decrease in arterial wall tension after initial treatment with the α-blocking agent. Scintigraphic study was not performed. We performed only bedside transthoracic echocardiography for assessment of myocardial performance status because the echocardiographic results showed clear and valuable data on both ventricular and diastolic function. Sympathetic dysfunction in viable myocardium has been recognized as a substrate for ventricular tachycardia [Oechslin 2000], which often is associated with isolated left ventricular noncompaction [Mitrani 1993]. Prazosin may improve left ventricular function because of a reversed abnormal adrenergic release in

noncompacted ventricle, and this effect may lead to an improved prognosis.

Pulmonary banding should not be performed in patients with cTGA, even if the patient is a newborn. Total correction should be performed in 1 session. In this case, the reason for the adverse outcome of progressive AV valve insufficiency might have been the pulmonary banding. Total correction of left AV valve insufficiency should be done when the defect is moderate. If in this case the valve could have been repaired instead of replaced (the patient's left AV valve was irreparable), we might not have faced the morphologic right ventricle failure. In addition, to decrease afterload and reverse abnormal adrenergic release, we suggest that the α-blocking agent be given to this particular group when the patients have severe cardiac failure. To our knowledge, a case such as this one, in which the patient had myocardial noncompaction of both ventricles combined with cTGA and was successfully treated by administration of prazosin (Minipress), has not been reported previously.

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