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Surgical Treatment of a 4-Year-Old Child with Hypertrophic Obstructive Cardiomyopathy: Case Report

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ABSTRACT

The incidence of pediatric hypertrophic obstructive cardiomyopathy is low. The lesions usually involve the left ventricle or ventricular septum, leading to either left or right ventricular outflow tract stenosis. However, combined left and right ventricular outflow tract stenosis is rare, and the surgical treatment is limited, especially in children. Surgery to release the obstruction was performed successfully in a 4-year-old child with right and left ventricular outflow tract obstruction together with a hypertrophic cardiomyopathy. The result was excellent.

INTRODUCTION

The incidence of pediatric hypertrophic obstructive cardiomyopathy (HOCM) is uncommon, compared to a relatively higher incidence of HOCM in adults [Kazui 2012; Prinz 2011]. The lesion usually involves the left ventricle or ventricular septum, leading to either left ventricular outflow tract (LVOFT) or right ventricular outflow tract (RVOFT) stenosis. However, combined left and right ventricular outflow tract stenosis is rare, and the surgical treatment is limited, especially in children. A 4-year-old female with both left and right outflow tract obstruction underwent a successful surgical procedure, as described here.

CASE REPORT

A 4-year-old girl was admitted to our hospital due to a cardiac murmur present for 2 years. Her growth and development were below normal, and her exercise intolerance was also below normal. A grade 3/6 harsh systolic murmur was heard at the 3rd to 4th left sternal border.

The chest radiograph indicated a cardiothoracic ratio of 0.52, with lung markings decreased (Figures 1 and 2). Echocardiography revealed that the left ventricular end diastolic diameter was 36 mm, and the right ventricular end diastolic diameter was 33 mm. The echocardiogram also revealed asymmetric septal hypertrophy (the maximum thickness of the septum was 18 mm). The anterior mitral valve leaflet demonstrated systolic anterior motion. The maximum flow

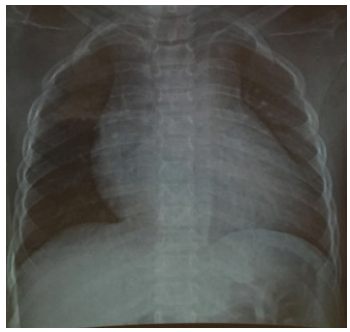

Figure 1. Preoperative chest X-ray.

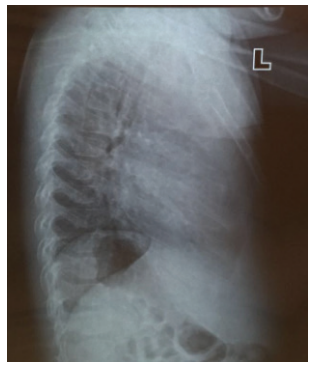

Figure 2. Preoperative chest X-ray.

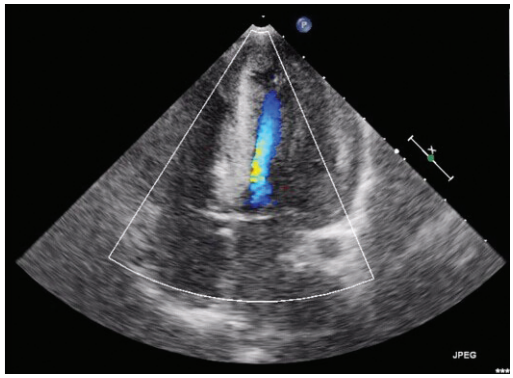

Figure 3. Preoperative ventricular septum.

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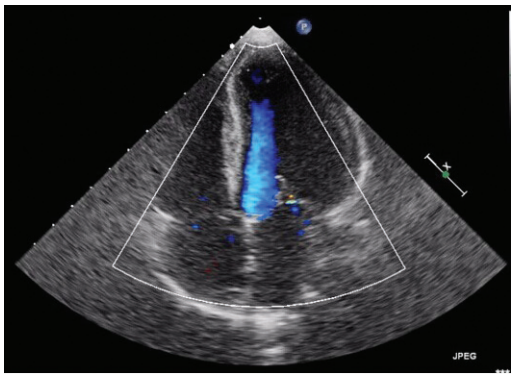

Figure 4. Post-operative ventricular septum.

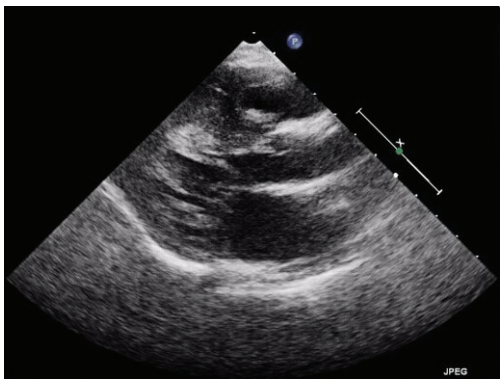

Figure 5. Preoperative LV and LVOFT.

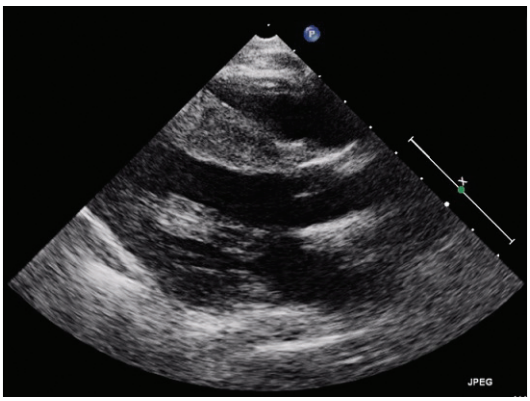

Figure 6. Postoperative LV and LVOFT.

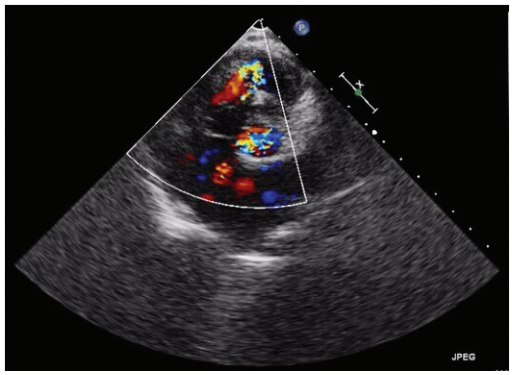

Figure 7. Preoperative RVOFT.

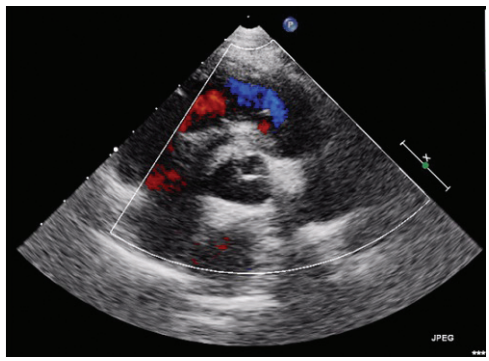

Figure 8. Postoperative RVOFT.

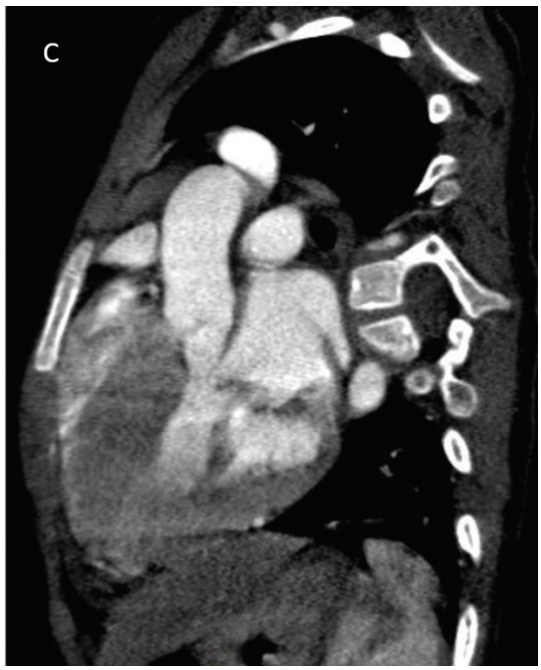

Figure 9. Preoperative CT (left ventricular outflow).

velocity of the left ventricular outflow tract was 3.5 m/s, with a gradient of 50 mm Hg. There was visible muscular stenosis of the RVOFT, and the narrowest diameter was 5 mm. The blood flow velocity of RVOFT was 4 m/s, and the gradient was 64 mm Hg (Figures 3, 5, 7).

A CT angiogram revealed that the diameter of the left ventricle was 35 mm, and the narrowest part of the LVOFT was 6 mm. The ventricular septum was hypertrophied, including a portion of the poster medial wall and papillary muscle. The base of the anterior interventricular septum was 21 mm, and it bulged toward the right ventricle, leading to RVOFT stenosis; the narrowest diameter was 4 mm. (Figures 9 and 11).

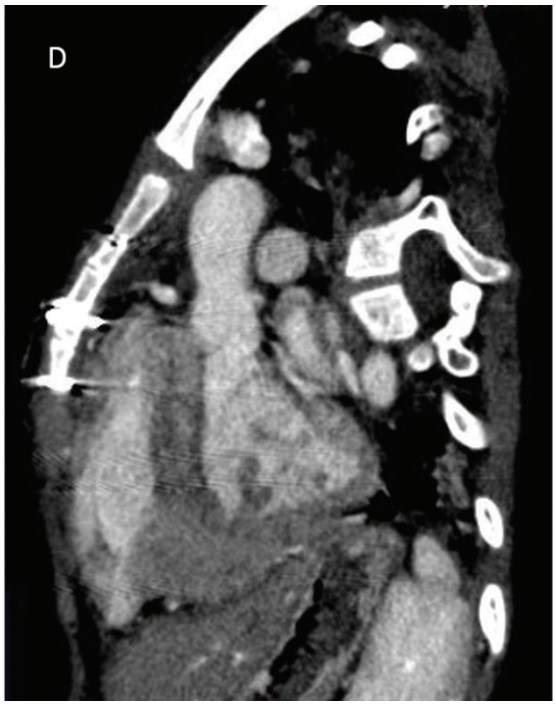

Figure 10. Postoperative CT (left ventricular outflow).

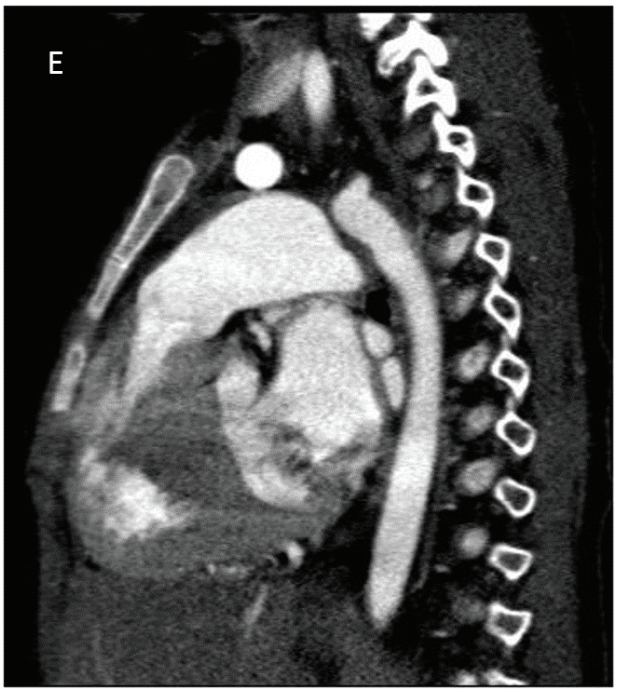

Figure 11. Preoperative CT (right ventricular outflow).

Cardiac catheterization revealed that the gradient in the RVOFT was 60 mm Hg, and the gradient in the LVOFT was 60 mm Hg (Figures 13 and 14; Table 1). The child was diagnosed with a hypertrophic obstructive cardiomyopathy, moderate mitral regurgitation, and mild aortic regurgitation.

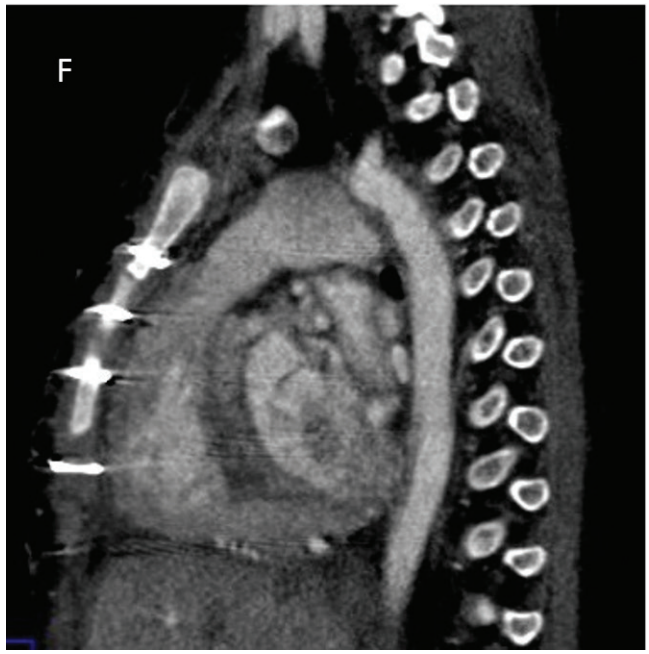

Figure 12. Postoperative CT (right ventricular outflow).

Table 1. Pressure analysis

Catheterization	Pressure (mm Hg)
Right ventricular	87/4/34
Main pulmonary artery	21/12/16
Left ventricular	142/-3/62
Ascending aorta	81/35/51

SURGICAL PROCEDURE

The surgery was performed under general anesthesia, cardiopulmonary bypass, and moderate hypothermia. Before establishment of cardiopulmonary bypass, the pressure and gradient were measured by a needle placed in the heart and connected to the monitor (Table 2). It was noted that the ventricular septum encroached into both the LVOFT and RVOFT, producing bilateral ventricular outflow tract stenosis. The hypertrophied myocardium (4.0 × 1.5 × 1.0 cm in the LVOFT) was removed through an aortic transverse incision, which was then closed with continuous 5/0 Prolene. Abnormal myocardium in the right outflow tract was excised through right atriotomy and trans-PV to release the stenosis. The thickness of reserved ventricular septum was based on normal children's data, assessed by preoperative echocardiography and CT. After the aortic clamp was released, spontaneous sinus rhythm resumed and the right atrial and pulmonary artery incisions were closed. The pressure of the pulmonary artery-right ventricle and the aorta-left ventricle were measured before closing the chest (Table 2).

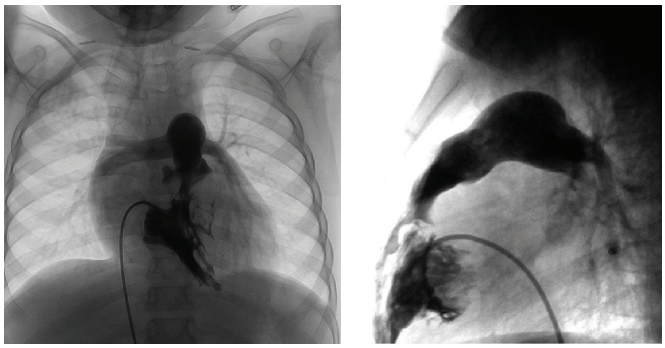

Figure 13. Preoperative catheterization: the chambers of heart, LVOFT, and RVOFT.

Table 2. Pressure measurement

	Pulmonary artery-right ventricle gradient (mm Hg)	Aorta-left ventricle gradient (mm Hg)
Preoperative	83	32
Postoperative	25	5

Table 3. Pressure appraisal

Echocardiography	Preoperative	Postoperative
Left ventricular end diastolic diameter, mm	36	33
Right ventricular end diastolic diameter, mm	33	24
Maximum forward velocity of RVOFT, m/s	4	2
Gradient of LVOFT, mm Hg	50	0
Gradient of RVOFT, mm Hg	64	16

LVOFT indicates left ventricular outflow tract; RVOFT, right ventricular outflow tract.

POSTOPERATIVE CARE

On the first postoperative day, the child developed low cardiac output syndrome and ventricular fibrillation, requiring inotropic therapy. The sternum had to be reopened under emergency conditions. The heart returned to normal sinus rhythm after electrical cardioversion. Following three hours of instability, the blood pressure gradually rose. Eight days later, the chest incision was closed. Then ventilation was discontinued and the patient was extubated. The child was discharged from the hospital on postoperative day 14, and was doing well at one-year follow-up.

The histopathological diagnosis was hypertrophic obstructive cardiomyopathy and myocellular disarray (Figures 15 and 16). Echocardiography performed prior to discharge showed that both the left ventricular end diastolic diameter and the right ventricular end diastolic diameter had significantly decreased, compared to the preoperative status. The

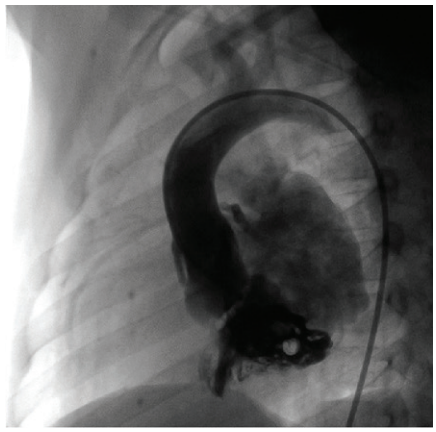

Figure 14. Preoperative catheterization: moderate mitral regurgitation.

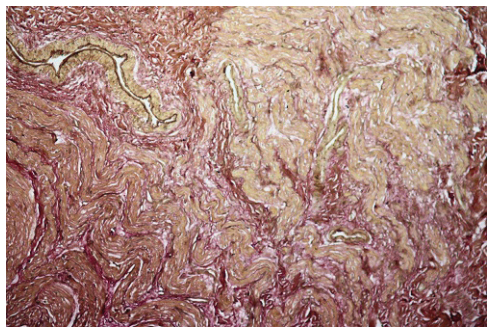

Figure 15. Myocellular disarray (×20).

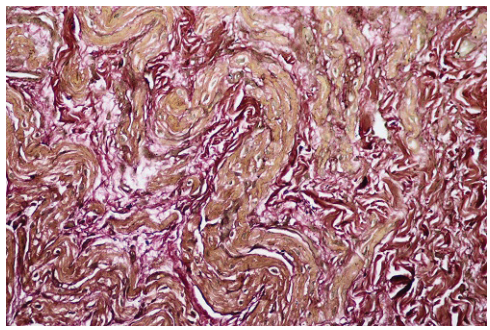

Figure 16. Myocellular disarray (×40).

thickest portion of the ventricular septum was 12 mm. The LVOFT was unobstructed, and the maximum forward velocity of RVOFT decreased, compared to the preoperative status (Figures 4, 6, 8) The gradient of the RVOFT decreased as well (Table 3). A postoperative CT imaged revealed that the diameter of the left ventricle was 33 mm, compared to 35 mm preoperatively (Figures 10 and 12).

DISCUSSION

The incidence of childhood hypertrophic obstructive cardiomyopathy is 0.47/100000 [Lipshultz 2003]. Thus, a

4-year-old child with hypertrophic obstructive cardiomyopathy involving both the left and right ventricular outflow tract is extremely rare.

A septal myectomy should be considered in patients who have chest pain and other symptoms that cannot be completely relieved with pharmacologic treatment, or if the left ventricular outflow tract pressure is ≥50 mm Hg [Maron 2003]. However, in children with mild symptoms (e.g., a resting left ventricular outflow tract pressure gradient >50 mm Hg), a septal myectomy is the preferred treatment [Minakata 2004]. The maximum gradient of both the LVOFT and the RVOFT were 60 mm Hg in this patient.

Myocellular disarray and cardiac muscle cell disorganization are responsible for cardiac dysfunction [Tardiff 1998; Morimoto 2003]. Pathological examination documented the presence of myocellular disarray; this condition, or surgical injury, might result in cardiac dysfunction in the early postoperative period. Postoperatively, the left ventricular end diastolic index and right ventricular end diastolic index significantly decreased; these findings may be due to gradual recovery of cardiac function to a normal level, as well as normalization of the left ventricular end diastolic index and the right ventricular end diastolic index.

The key feature of this operation is the resection of abnormal myocardium (hypertrophy of the ventricular septum as well as the left and right ventricular outflow tracts) as completely as possible. The procedure corrects myocardial lesions via incisions in the aorta and pulmonary artery. However, after a myocardial myomectomy, the myocardium may be damaged, which results in an increase in cardiac diastolic volume that may lead to the low cardiac output syndrome. Therefore, delayed sternal closure after the procedure may be crucial for patient recovery. It is also important to avoid damage to myocardial conduction tissue. A LVOFT myomectomy is usually performed through the aortic valve with extreme care being taken to maintain leaflet integrity and valve function; the same precaution is necessary for a RVOFT through the right atrial and pulmonary arterial incisions.

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