

Hypertrophic Obstructive Cardiomyopathy with Infundibular Stenosis Treated by Alcohol Ablation Therapy

This report describes an uncommon case of hypertrophic obstructive cardiomyopathy (HOCM) accompanying infundibular stenosis of the right ventricle treated by alcohol ablation therapy, in a 28-yr-old male patient presenting with dyspnea on exertion. HOCM with infundibular stenosis was detected by echocardiogram and cardiac catheterization and patient has dynamic obstructions of both ventricular outflow tracts. We performed alcohol ablation therapy to improve clinical symptoms and to relieve dynamic obstructions of both ventricular outflow tracts. This is the first case in which HOCM with infundibular stenosis of the right ventricle was treated by alcohol ablation therapy.

Key Words : *Cardiomyopathy, Hypertrophic; Pulmonary Subvalvular Stenosis; Ablation, Alcoholic*

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INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) involves the ventricular septum and less frequently segments of the right ventricle (1). Outflow obstruction of the left ventricle is the usual hemodynamic finding in HOCM and hypertrophic cardiomyopathy has right ventricular obstruction in 15% of cases (2). However, it is uncommon to find infundibular stenosis of the right ventricle in the presence of left ventricular outflow tract obstruction (2, 3). Management options for patients with HOCM accompanying infundibular stenosis of the right ventricle may include surgical removal of hypertrophied muscle bundles (4-6). Alcohol ablation therapy, known as a non-surgical therapy of HOCM, has been reported in the treatment of hypertrophic cardiomyopathy or infundibular pulmonary stenosis (7-9). However, alcohol ablation therapy in HOCM with infundibular stenosis of the right ventricle has not been reported. We report a case of HOCM with infundibular stenosis of the right ventricle treated by alcohol ablation therapy, in a 28-yr-old male patient presenting with dyspnea on exertion.

CASE REPORT

A 28-yr-old male patient was referred to Inha Medical Center in July, 2000 for an echocardiogram. The patient had a 6-yr history of exertional dyspnea, for which he was treated medically. He had no prior history of medical and surgical diseases. His family members were healthy and had no history of congenital heart disease. On physical examination, the peripheral pulses and blood pressure measurements were normal. A 5/6 harsh, systolic ejection murmur accompanied by a thrill was heard between the apex and the lower left sternal border.

Basic laboratory tests, including CBC, lipid profiles and liver function were not remarkable. There was no significant change in the levels of cardiac enzymes. Urinalysis revealed no abnormalities. The chest roentgenogram showed a normal heart size. Electrocardiogram revealed increased amplitude of the P wave in lead II exceeding 2.5 mV and increased QRS voltages with inverted T waves in the precordial chest leads.

The echocardiogram showed asymmetric hypertrophy of the interventricular septum and systolic anterior motion of the mitral valve (Fig. 1), accompanying the left ventricular outflow tract obstruction and mitral incompetence. The

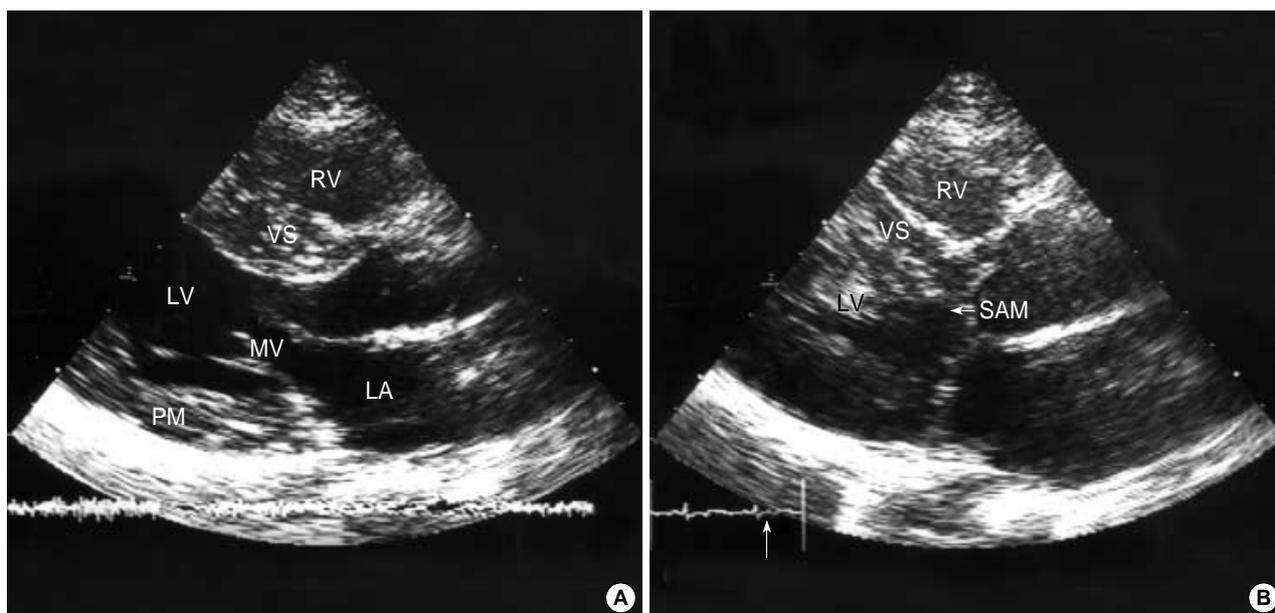


Fig. 1. Asymmetric septal hypertrophy (A) and systolic anterior motion of the anterior mitral leaflet (B) is shown in the parasternal long axis view. RV: right ventricle; VS: ventricular septum; LV: left ventricle; MV: mitral valve; LA: left atrium; PM: papillary muscle; SAM: systolic anterior motion.

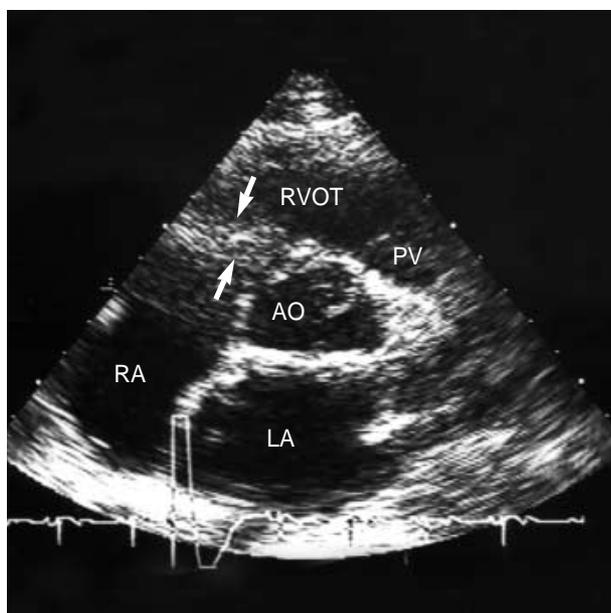


Fig. 2. Right ventricular outflow tract obstruction (RVDT) in the parasternal short axis view. PV: pulmonary valve; AO: aorta; RA: right atrium.

pressure gradient across the left ventricular outflow tract was 100 mmHg. The right ventricular outflow tract obstruction was also observed in the parasternal short axis view (Fig. 2). The ejection fraction was increased to 83%.

Left and right heart catheterization was carried out under local anesthesia. Simultaneous hemodynamic tracings of left ventricular and aortic pressures during pullback of the catheter from the left ventricle showed a large aortic-left ventricular

pressure gradient. Peak-to-peak aortic-left ventricular pressure difference was 100 mmHg at baseline and 200 mmHg after an extrasystolic beat (Fig. 3A). The selective right ventricular angiography revealed infundibular narrowing of the right ventricle due to hypertrophied muscle bundles. The systolic pressure in the right ventricle was elevated up to 80 mmHg and right ventricular diastolic pressure was increased to 10 mmHg (Fig. 4A). An intracardiac shunt was not observed during oxymetry procedure.

Surgical treatment was offered to the patient, but he was reluctant to undergo operation. After he had been offered all other surgical and non-surgical treatment option, he agreed to have an alcohol ablation therapy. The right ventricular angiography displayed that the hypertrophied muscle mass, extending from the infundibulum to the portion of the adjacent interventricular septum, bulged toward the outflow tract during the systolic phase and obstructed the blood flow across the right ventricular outflow tract. According to these findings, we thought that the obstruction across the right ventricular outflow tract was mainly attributed to the infundibular stenosis and the hypertrophied septal mass was partially involved in the obstructions of right ventricular outflow tract. The patient was transferred to the cardiac catheterization laboratory for alcohol ablation therapy. A 5-Fr multipurpose catheter was placed at the apex of the left ventricle through the left femoral artery in order to measure pressure gradients of the left ventricular outflow tract. A catheter of the temporary pacemaker was positioned at the apex of the right ventricle. Through a femoral approach, a 7-Fr Judkins type guiding catheter was introduced into the left coronary ostium. After passing a 0.014" extra-support long guidewire to the

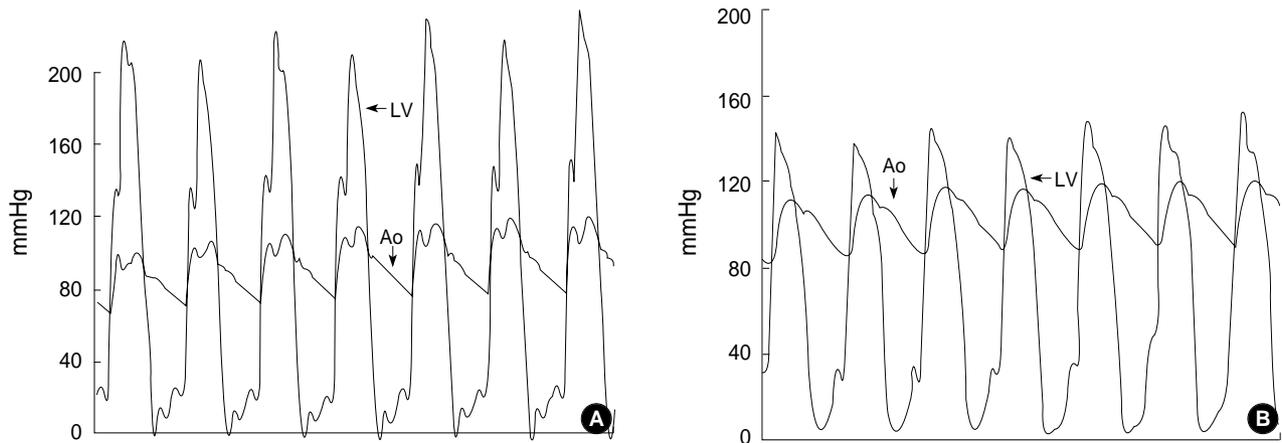


Fig. 3. Simultaneous aortic and left ventricular outflow tract pressure tracings before (A) and after (B) alcohol ablation.

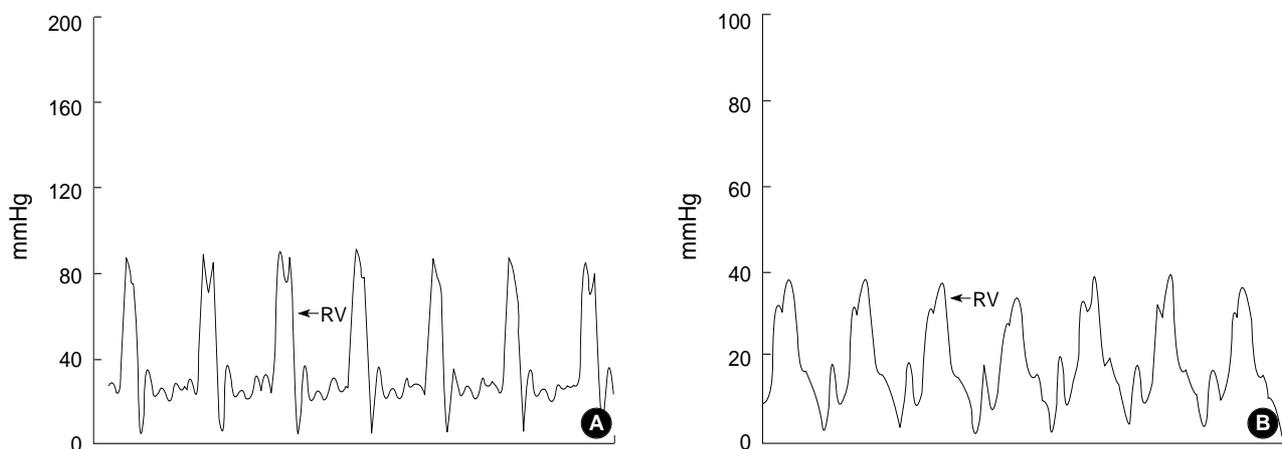


Fig. 4. Right ventricle pressure tracings before (A) and after (B) alcohol ablation.

first septal artery, a 1.5-mm angioplasty balloon was advanced over the guidewire to the first septal artery. Prior to the alcohol ablation, pressure tracing was done by occlusion of septal artery with balloon and the pressure decrease in both LVOT and RVOT were observed. To perform a contrast echocardiogram, we injected 10 mL of agitated saline through the central lumen of the balloon catheter. Under contrast echocardiogram, enhancement was observed up to RVOT of septal area. After fulfilling the contrast echocardiogram, we performed a prolonged balloon inflation to obstruct the ostium of the first septal artery with 4 atmospheres and 5 mL absolute alcohol was administered directly into the first septal artery through the central lumen of the balloon catheter. After 5 min of continuous inflation, the balloon catheter was deflated and a subsequent angiography showed the first septal artery to be occluded at its origin, without evidence of alcohol extravasation. Immediately after the procedure, the pressure gradient between the left ventricle and the aorta was decreased to 20 mmHg at baseline and to 60 mmHg after extrasystole (Fig. 3B). The systolic pressure in the right ventricle was also decreased to 32 mmHg (Fig. 4B). The patient remained hemodynamically

stable and complained of a mild chest pain without any change in the ECG findings. The patient was transferred to the coronary care units and the subsequent hospital course was uneventful and the patient was discharged after 7 days of in-hospital observation.

DISCUSSION

Hypertrophic obstructive cardiomyopathy with infundibular stenosis of the right ventricle is an uncommon congenital anomaly in which outflow tract obstructions of both ventricles may occur due to the hypertrophied muscle bundles. In such cases, outflow tracts of both ventricles may have dynamic obstructions via hypertrophied muscle bundles. Hypertrophied muscle bundles alter the dynamic outflow by causing narrowing of the outflow tracts and the dynamic gradients are produced through the outflow tracts. In the present case, HOCM with infundibular stenosis was detected by echocardiography and cardiac catheterization and the patient had dynamic gradients of both ventricular outflow tracts.

The patient represents a unique example of both ventricular outflow obstructions due to hypertrophied muscle bundles. In patient with obstruction of right ventricular outflow tract, the ventricular septal defect is sometimes accompanied (10, 11). In the present case, there was no evidence of intracardiac shunts by cardiac catheterization.

Reduction of hypertrophied muscle bundles is necessary not only for lowering dynamic gradients of the outflow tracts, but also for improving the clinical symptoms. Treatment for HOCM accompanying infundibular stenosis of the right ventricle may include surgical removal of anomalous muscle bundles. In our patient, due to his refusal to any surgical therapy, we decided to adapt non-surgical therapy under his consent. The hypertrophied septal mass was partially involved in the obstructions of right ventricular outflow tract according to right ventricular angiography and the pressure tracing prior to alcohol ablation revealed that occlusion of septal artery with balloon decreased the pressure in both LVOT and RVOT. So we thought that reduction of the septal mass had led to relief of dynamic obstructions across the left ventricular outflow tract and partially across the right ventricular outflow tract. For HOCM patients, a couple of mechanisms are involved in the relief of the obstruction after alcohol ablation. However, concerning no significant septal thinning after the ablation and also pressure decrease in both LVOT and RVOT, we assume that the decreased septal motion might be the main mechanism in the relief of obstruction. To the best of our knowledge, this is the first case of which HOCM with infundibular stenosis of the right ventricle treated by alcohol ablation therapy. At a 1-yr follow-up, the patient was still asymptomatic without any complications from the treatment.

In conclusion, alcohol ablation therapy is an emerging treatment modality, and it may be used on behalf of surgical therapy in patients with hypertrophic obstructive cardiomyopathy accompanying infundibular stenosis. However, more experiences are needed to establish it as an effective therapeutic approach in this disease.

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