

Initial Experience of Atrial Septostomy Treatment for Symptomatic Nonobstructive Hypertrophic Cardiomyopathy: A Case Report

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Abstract

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Abstract

Nonobstructive Hypertrophic Cardiomyopathy with preserved EF is a well-known ejection fraction preserved heart failure. Oral medical therapy is the major treatment for nonobstructive hypertrophic cardiomyopathy. While interatrial shunt devices have preliminarily been shown to be safe and efficacious for the treatment of HFpEF, their utilization in nonobstructive hypertrophic cardiomyopathy patients has not previously been described.

Key words: heart failure, left-to-right shunt, hypertrophic cardiomyopathy

INTRODUCTION

No medications has been proven to improve long-term outcomes in patients with nonobstructive HCM so far^[1], so the treatment is focused on relieving symptoms, and improving exercise capability and quality of life. Dyspnea and chest discomfort are common symptoms in patients with nonobstructive HCM with preserved EF. These can be a result of increased LV filling pressures related to diastolic dysfunction. There has been improvement in outcomes of those with HFpEF managed with an interatrial shunt device by increasing left-to-right shunting as a pressure release^[2]. But for HCM, due to the poor ventricular myocardial relaxation,

the cardiac output greatly depends on the atrial systole for ventricular preloading. So, in the REDUCE-LAP HF clinical trial, the hypertrophy cardiomyopathy was an exclusion criterion^[2]. But the hemodynamic of HCM in different stages is not all the same. If the PCWP was over 25mmHg on exertion, yet not over 15mmHg at rest, at the same time, the cardiac output was accessible and blood pressure did not drop sharply in exercise Swan-Ganz inspection, it is reasonable to make an atrial septostomy to download the preload of left ventricle.

CASE PRESENTATION

A 52-year-old woman with a history of diagnosis of nonobstructive hypertrophic cardiomyopathy and atrial fibrillation for 4 years, presented with New York Heart Association(NYHA) Class II-III dyspnea, lower extremity swelling Occasionally; On presentation, blood pressure was 119/86mmHg, heart rate(HR) 84 beats/min, oxygen saturation 95%, respiratory rate 15/min; Her symptom was notable for severe dyspnea on exertion.

Hospital workup included NT-proBNP 2102 pg/ml, a 12-lead electrocardiogram with atrial fibrillation as well as a chest x-ray that demonstrated pulmonary edema. An echocardiogram revealed a preserved ejection fraction 60%, mild biatrial dilation(54mm for left atrium in the parasternal long-axis), Moderate tricuspid regurgitation, left ventricular hypertrophy with middle ventricular septal thickness of 17 mm, Left ventricular outflow tract flow velocity was normal and E/e' was 11.5.

Prior to further workup, the patient was aggressively dehydrated with intravenous furosemide. He subsequently underwent right cardiac catheterization at rest and during supine bicycle exercise which revealed a resting mean pulmonary artery wedge pressure(PAWP) of 13mmHg, and exercise peak PCWP of 28 mmHg(EP-PAWP); Mean pulmonary artery pressures of 20mm Hg, and a mean right atrial pressure of 5mmHg. At baseline, the 6-min hall walk distance was 325 m; Coronary angiography was normal; the Kansas city cardiomyopathy questionnaire (KCCQ) quality of life score was 52. As dyspnea was not alleviated significantly by diuretics, an interventional course was pursued. Following careful evaluation by the heart failure team, the patient was considered a suitable candidate for the atrial septostomy.

With no evidence of severe right-sided heart failure, the patient underwent atrial septostomy with sequential balloon sizes of 8, and 10mm (Figure 1 A, B). This was accomplished with high-pressure/non-compliant Mustang Balloons (Boston scientific). Per transthoracic echocardiography, the resulting atrial septal defect measured 5~7.3mm in diameter with evidence of left-to-right shunting. After this intervention, he underwent exercise right cardiac catheterization immediately revealed mean PAWP of 14mmHg, EP-PAWP of 19 mmHg and a mean right atrial pressure of 9mmHg. On the second day after operation, the 6-min walk distance was 450 m. The patient noted improvement in dyspnea and was discharged.

At the third month, she continued doing well with NYHA class I symptoms, requiring half dosage of diuretics, the 6-min walk distance was 417m, KCCQ score was 26, NT-proBNP was 2416 pg/ml. A follow-up echocardiogram revealed that a preserved ejection fraction 57%, mild biatrial dilation (55mm for left atrium in the parasternal long-axis), Moderate tricuspid regurgitation, left ventricular hypertrophy with middle ventricular septal thickness of 17 mm, Left ventricular outflow tract flow velocity was normal and E/e' was 9, and no obvious left-to-right shunt was observed.

DISCUSSION

This case report is the first known to employ an atrial septostomy treatment for nonobstructive hypertrophic cardiomyopathy with preserved EF. To avoid the irreversible result, only balloon dilation septostomy was made with gradually increased balloon diameter in case the unacceptable large interatrial shunt and hemodynamical collapse appeared.

In the three-month follow-up, the patient's NYHA grades, exercise capacity, KCCQ improved apparently, and NT-proBNP, left atrium diameter, LVEF kept in the same level. Theoretically, the septostomy by balloon angioplasty could only keep the septum defect patent for several month^[4]. In this case, the defect kept patent at the third month, and give us a chance to study whether the septostomy stay efficacious in

hemodynamic dimension. it was proved only if the artificial atrial septum defect be still patent, the symptom of heart failure is supposed to be relieved significantly.

Atrial septostomy is not a fundamental treatment for HCM pathologically, so we don't expect it could improve the patients' long term prognosis greatly, theoretically we even worried about the increased pulmonary circulation overloading and the right ventricle will lead to heart function deterioration. But it seemed atrial septostomy be well tolerated in this HCM case. In fact, REDUCE LAP-HF II presented us with a neutral result [2] for heart failure with preserved and mildly reduced ejection fraction treated with atrial shunt device. The MACE of atrial septostomy is even significantly improved in the subgroup with a peak exercise PVR of less than 1.74 Wood units (n=382, 63.5% of all patients recruited). That is to say atrial septostomy is a beneficial treatment to HCM conditionally.

CONCLUSION

Treatment of nonobstructive hypertrophic cardiomyopathy with preserved EF could be challenging with medical therapy alone, especially for those diuresis intolerance and resistance patients, and septostomy may provide significant symptomatic benefit in these patients. The long-term prognosis is not sure at present, which need more randomized control trial to provide more evidence.

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CONFLICT OF INTEREST

The authors report no conflict of interest.

AUTHOR CONTRIBUTIONS

YM, Y-HZH and J-ZH: studied the conception and designed the study. T-A: involved in acquisition of data and analysis and interpretation of data (eg, statistical analysis and computational analysis). T-A, YM, Y-HZH and J-ZH: wrote, reviewed, and/or revised the manuscript. YM, Y-HZH and J-ZH: supervised the study.

ETHICAL APPROVAL

The study was approved by the institutional review board.

DATA AVAILABILITY STATEMENT

Data in the current study are available from the corresponding author on reasonable request.

Reference:

- [1] 2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy. Michael A. Burke, Sharlene M. Day, etc, JACC 2020.08.045
1. Atrial shunt device for heart failure with preserved and mildly reduced ejection fraction (REDUCE LAP-HF II): a randomized, multicenter, blinded, sham-controlled trial Sanjiv J Shah, Barry A Borlaug, Eugene S Chung, etc. Lancet 2022.2.25
2. Shah SJ, et al. One-year safety and clinical outcomes of a transcatheter interatrial shunt device for the treatment of heart failure with preserved ejection fraction in the reduce elevated left atrial pressure in patients with heart failure (REDUCE LAP-HF I) trial: a randomized clinical trial. JAMA Cardiol. 2018;3(10):968–77.
3. Chaowu Yan, Guodong Niu, etc. Stable interatrial communication combining balloon septostomy and radiofrequency catheter ablation, journal of the American College of Cardiology, Volume 72, Issue 15, 9 October 2018, Pages:1873-75

Figure 1

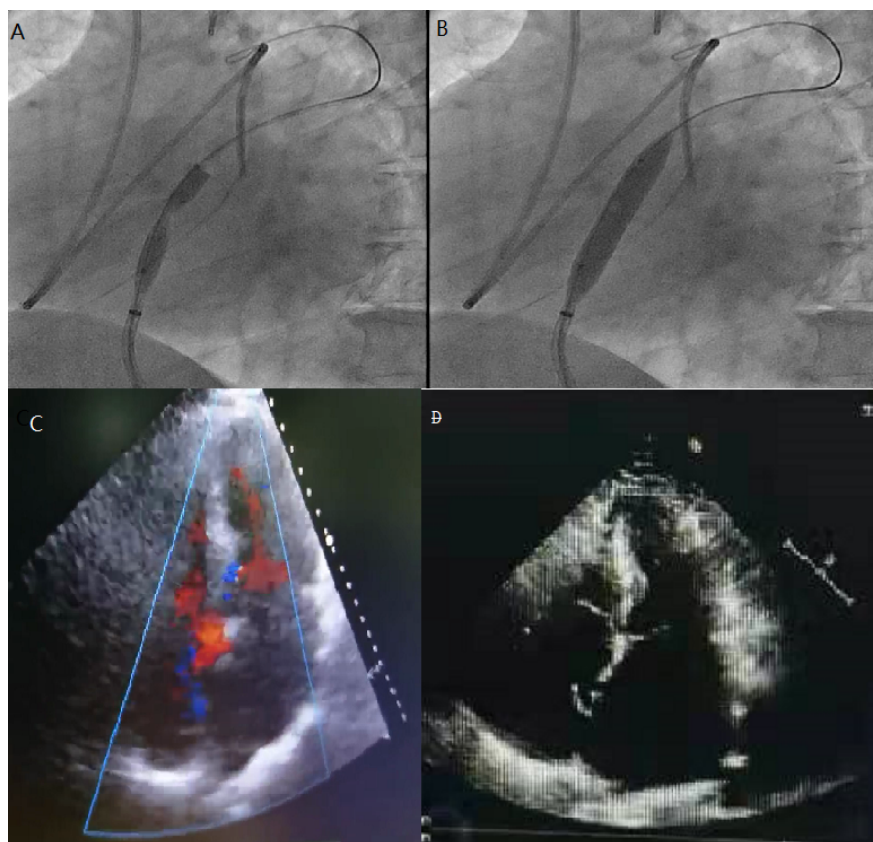


FIGURE 1 A and B, balloon dilation of atrial septum