Permanent Pacing in a Patient with Left Ventricular Mid-Cavity Obstruction and Apical Aneurysm

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Summary

Hypertrophic cardiomyopathy with left ventricular (LV) mid-cavity obstruction and LV apical aneurysm is associated with high morbidity and mortality rates. However, consensus is lacking on the treatment modality for LV mid-cavity obstruction and LV apical aneurysm. Here, we report a case of reduced LV mid-cavity pressure gradient and symptoms, treated using permanent pacing. The effect of permanent pacing on pressure gradient and symptoms lasted for 4 years. As pacing is relatively non-invasive compared to surgical therapy, permanent pacing is a good option, especially in the elderly patients with LV mid-cavity obstruction and apical aneurysm.

Key words: Hypertrophic cardiomyopathy, Echocardiography, Pacemaker

Case Report

A 76-year-old woman was admitted to our hospital with heart failure. Although she was diagnosed with HCM 20 years ago, no examination was performed. She had a medication history of carvedilol and a long-acting calcium channel blocker for hypertension. Four months prior to the admission, she had dyspnea upon climbing stairs. The dyspnea and chest pain worsened, and she was transferred to our hospital. The patient’s height, body weight, heart rate, blood pressure, and oxygen saturation (room air) were 153 cm, 47.9 kg, 65 beats per minute, 111/67 mmHg, and 98%, respectively. A late systolic to diastolic heart murmur (grade 2/6) was audible, most prominently at the left sternal border. Bibasilar lung crackles were detected. The abdominal findings were normal. A slight but symmetrical lower leg edema was noted. Laboratory data showed slight renal dysfunction (serum creatinine: 0.91 mg/dL; estimated glomerular filtration rate: 45.9 mL/minute/body surface area), 1226.2 pg/mL brain natriuretic peptide (BNP), and 14.5 g/dL hemoglobin. A chest X-ray demonstrated cardiomegaly; the cardiothoracic ratio was 58% (Figure 1A). Electrocardiography showed sinus rhythm at the rate of 67 beats per minute and LV hypertrophy with ST-T abnormalities that was most notable in leads I, aVL, and V3-V6 (Figure 1B). On transthoracic echocardiography, LV mid-cavity hypertrophy at the level of the papillary muscle and obstruction was revealed with an apical aneurysm, resulting in an hourglass configuration of the LV (Figure 2). The LV basal end-diastolic dimension, end-systolic dimension, and ejection fraction...
Figure 1. A: Chest X-ray at admission. Chest X-ray showed cardiomegaly. B: Electrocardiogram at admission. Electrocardiography showed sinus rhythm at the rate of 67 beats per minute and LV hypertrophy with ST-T abnormalities that was most notable in leads I, aVL, and V3-V6.

Figure 2. Transthoracic echocardiography. LV mid-cavity hypertrophy at the level of the papillary muscle and obstruction was revealed with an apical aneurysm, resulting in an hourglass configuration of the LV of the apical long-axis view (A) and four-chamber view (B). AN indicates aneurysm; LA, left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle.

was 48 mm, 33 mm, and 50%, respectively. Color Doppler and continuous-wave Doppler imaging demonstrated that blood flow from the LV apex to the base was interrupted during systole, but not during diastole, which is known as paradoxical jet flow (Figure 3). The peak flow velocity from the apex to the base was 3.0 m/second, corresponding to the gradient of 36 mmHg. No mural thrombus was detected in the LV apical aneurysm. The ratio of peak early mitral inflow velocity (E) and peak late mitral inflow velocity (A) of the transmitral flow was 0.7. Moderate mitral regurgitation and moderate tricuspid regurgitation were evident. The estimated right ventricular (RV) systolic pressure was 40 mmHg. No significant ventricular arrhythmia was detected on monitor electrocardiography.
Cardiac catheterization revealed mean pulmonary artery wedge pressure of 10 mmHg, mean pulmonary artery pressure of 27/9/17 mmHg, end-diastolic RV pressure of 31/0/10 mmHg, and mean right atrial pressure of 4 mmHg. The cardiac index was 2.63 L/minute/m². The mean LV apical pressure, LV basal pressure, and aortic pressure were 247/15, 93/9, and 99/50/70 mmHg, respectively. Coronary angiography revealed no significant coronary artery stenosis.

The administration of bisoprolol (2.5 mg/day) and cibenzoline succinate (150 mg/day) was ineffective for symptomatic relief and LV mid-cavity obstruction. Thus, we decided to proceed with pacing: RV temporary pacing was performed to estimate the effectiveness of LV mid-cavity obstruction. Before RV apical pacing, the mean LV apical pressure and aortic pressure were 226/13 and 153/65/96 mmHg, respectively, while after RV apical pacing, they were 198/10 mmHg and 143/68/97 mmHg, respectively. RV apical pacing reduced the peak pressure gradient between the LV apex and aorta from 73 mmHg to 55 mmHg. Temporary pacing from the RV apex improved the patient’s symptoms that were interfering with daily activities. Transthoracic echocardiography revealed that the peak flow velocity from the apex to base in early systole and during diastole was significantly reduced to 1.0 m/second by RV apical pacing (Figure 4). Then, a dual-chamber pacemaker with electrodes positioned at the RV apex and right atrial appendage (DDD mode) was implanted (Figure 5). The AV delay was set to 120 ms to maximize RV pacing. The transverse strain recordings of mid-septum and posterior wall myocardial segments were measured using the speckle tracking strain before and after RV apical pacing (Figure 6). Before pacing, the time to peak strain for both segments coincided during systole. After RV apical pacing, however, the motion of both segments was dyssynchronous. Bisoprolol (2.5 mg/day) was continued and warfarin was started to prevent thrombus formation in the LV apical aneurysm. The patient’s functional class improved significantly, and we did not perform stress echocardiography to induce LV mid-cavity pressure gradients. The BNP level was decreased to 696.9 pg/mL, and she was discharged. The symptomatic and hemodynamic improvements sustained for 4 years of follow-up (Figures 7, 8).

**Discussion**

HCM patients with mid-ventricular obstruction and LV apical aneurysm represent a high-risk subgroup within the disease spectrum. The high risk is attributed to the presence of mural thrombosis, progressive heart failure, stroke, and sudden death. Although the mechanism of LV apical aneurysm formation in LV mid-cavity obstruction remains unknown, several hypotheses have been made. LV mid-cavity obstruction leads to the cessation of systolic flow at the mid-ventricular level, resulting in blood trapping within the apical chamber. When LV mid-cavity obstruction is relieved, blood flows from the apex to base in
late systole or early diastole, which is known as paradoxical jet flow. Increased pressure at the apex caused by blood trapping may lead to an apical oxygen supply-demand mismatch. Resulting microvascular ischemia is evident as thickened coronary walls with intraluminal narrowing in the absence of discrete coronary obstruction. Microvascular ischemia is exacerbated by the coronary flow obstruction that occurs during early diastole, leading to coronary flow reversal and decline in flow reserve. Therefore, the formation of a high-pressure apical zone with increased wall stress leads to subendocardial ischemia and infarction. Consequently, this leads to the formation of diffuse myocardial fibrosis and thinning with a resultant apical aneurysm.

In obstructive HCM, the medical management largely aims to alleviate symptoms. The most commonly used drugs are negative inotropic agents, such as beta-blockers. Currently, there is no evidence that medical therapy alleviates mid-ventricular obstruction and prevents the development of LV apical aneurysm. Surgical aneurysmectomy or aneurysm ligation is an alternative to resolve the complications of thromboembolism, ventricular arrhythmias, and cardiac dysfunction associated with LV apical aneurysm formation. Other surgical interventions include thrombectomy, myectomy, and alcohol septal ablation. Kunkala et al. found that patients with preoperative echocardiographic

Figure 4. Continuous-wave Doppler imaging revealed that the peak flow velocity from the apex to base in early systole and during diastole significantly reduced from 3.0 m/second to 1.0 m/second by RV apical pacing.

Figure 5. Chest X-ray and electrocardiogram after dual-chamber pacemaker implantation.
features of mid-ventricular obstruction commonly experience symptomatic relief after myectomy with a significant improvement in the New York Heart Association class.\textsuperscript{7} Although data are currently insufficient to suggest the optimum timing for the surgical approach, surgery should be considered after symptom onset. Despite promising early results, permanent pacing is disregarded as a first-line therapy for drug-resistant HCM with LV outflow tract obstruction because of its low success rate in randomized studies.\textsuperscript{11,12} The LV outflow tract gradient due to systolic anterior motion (SAM) of the mitral valve is highly variable and dynamic.\textsuperscript{13} Velchev et al. reported the effectiveness of permanent pacing in patients with LV mid-cavity obstruction after alcohol septal ablation for LV outflow tract obstruction.\textsuperscript{14} The mechanism of LV mid-cavity obstruction improvement in the absence of subaortic obstruction by SAM may be associated with the dyssynchronous contraction of the septum and posterior wall caused by pacing of the RV apex.\textsuperscript{15} Because pacing is relatively non-invasive compared to surgical therapy, permanent pacing is a good option, especially for the elderly patients with LV mid-cavity obstruction and apical aneurysm.
Disclosure

Conflicts of interest: The authors have no conflicts of interest.

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