It has been well established that dual-chamber pacing reduces the pressure gradients in the left ventricular outflow tract (LVOT) and improves the symptoms of hypertrophic obstructive cardiomyopathy. However, the effect of pacing therapy on a hypertrophic mid-ventricular obstruction (MVO) with an apical aneurysm has rarely been described and we report such a case that was dramatically improved by dual-chamber pacing.

This report is of special interest from 2 points of view. We demonstrate, on the basis of this case, that the pacing therapy was effective in not only reducing intraventricular pressure gradient, but also in preventing sustained ventricular tachycardia (VT). Secondly, we are the first to attempt to evaluate the intraventricular flow dynamics during pacing in such a case.

Case Report

A 63-year-old woman suffered chest discomfort with a syncope attack and was admitted to hospital. Her past history and familial history were unremarkable. The 12-lead electrocardiogram (ECG) on admission showed sustained monomorphic VT (170 beats/min) with a right bundle branch block pattern in V1 and a QS pattern in leads I, II, III, aVF and V2–6, and an axis of –140°. These findings were similar to those seen with VT originating from the septoapical area of the left ventricle (Fig 1). After spontaneous recovery to sinus rhythm 40 s later, the ECG showed negative T waves in leads I, II, III, aVF and V1–6 with slight ST elevation in III, aVF and V4 (Fig 1). Holter monitoring showed 2 episodes of sustained monomorphic VT (30–40 s), 1,220 episodes of nonsustained monomorphic VT and 355 episodes of couplet ventricular extrasystoles per day. Although a β-blocking agent is the drug of choice, it did not completely suppress the VT. The 2-dimensional echocardiographic examination revealed left mid-ventricular hypertrophy at the papillary muscle level and a discrete high apical pressure. The left ventriculogram confirmed a mid-ventricular obstruction with an apical aneurysm. Invasive assessment of intraventricular pressure showed a peak-to-peak gradient greater than 100 mmHg. Treatment with antiarrhythmic agents could not prevent the VT, but dual-chamber pacing reduced the intraventricular pressure gradient and suppressed the VT completely. Continuous wave Doppler showed that the early systolic ejection flow from the apex had disappeared, that there was isovolumetric relaxation flow toward the apex and that there was attenuation of the diastolic paradoxical jet flow toward the basal chamber. Such findings by continuous wave Doppler can be useful in pacing therapy for evaluating changes in the severity of mid-ventricular obstruction. 

Key Words: Apical aneurysm; Continuous wave Doppler; Dual-chamber pacing; Mid-ventricular obstruction; Ventricular tachycardia
There was no evidence of typical systolic anterior motion of the mitral valve or asymmetric septal hypertrophy. The color Doppler image showed turbulent diastolic flow from the sequestered apical chamber toward the main left ventricular cavity. The continuous wave Doppler signal across the region of mid-ventricular narrowing, recorded from the LV apex under the guidance of color Doppler, revealed a very unusual flow pattern (Fig 2). In early systole, there was a short-lived ejection flow from the apical chamber that was abruptly halted by the mid-ventricular obstruction. Thereafter, the apex-to-base flow resumed in early diastole and continued for most of diastole, corresponding with a diastolic paradoxical jet flow (Fig 2A) that implied a significant diastolic intraventricular gradient with high apical pressure. The maximal diastolic flow velocity was 3.1 m/s. The apical chamber filled in late diastole and also during isovolumetric ventricular contraction, but it was difficult to calculated an accurate peak pressure gradient from the modified Bernoulli equation because of attenuation of the Doppler signal during systole. Cardiac catheterization and an electrophysiological study was performed. Coronary angiography did not show any significant stenotic lesions. The left ventriculogram showed a hyperkinetic contraction pattern with systolic obstruction at the mid-ventricle (Fig 3). The sequestered apical cavity persisted throughout the cardiac cycle, indicating apical aneurysm. No communication between the apical and basal chambers was observed at mid systole, but a narrow muscular tunnel between the cavities was evident at end-diastole.

An asynergic or aneurysmal apical chamber in the absence of fixed coronary artery disease has been previously described in a patient with MVO, and it has been suggested that the development of apical aneurysm may be
Patients with MVO are candidates for cardiac surgery, but recently, pacing therapy for pressure gradient reduction in patients with MVO has been reported as is the case for patients with hypertrophic LVOT obstruction.

Previous reports have suggested that the paradoxical motion of the interventricular septum may play a role in the relief of LVOT obstruction and another study has indicated that pacing reduces septal wall motion in patients with LVOT obstruction. These effects of pacing may also relieve MVO, so we decided to use pacing therapy in the present case. Dual-chamber pacing markedly and immediately reduced the intraventricular pressure gradient (Fig 4C), so pacing therapy would be a therapeutic option for pressure gradient reduction in drug-refractory patients with MVO.

Patients with the MVO variant of hypertrophic cardiomyopathy are at a higher risk of developing serious ventricular arrhythmias. A prolonged increase in pressure within the apical chamber may lead to cellular necrosis and produce different arrhythmic substrates, possibly through an ischemic mechanism. We cannot conclude that reentry was the origin of the sustained monomorphic VT in the present case, because it was not inducible by programmed ventricular stimulation. Automaticity of the surviving myocardium might have cause the arrhythmia in this case, because an experimental study showed that automaticity was promoted by an increased afterload and preload.

Regarding therapy, Mantica et al reported that catheter and surgical therapy of sustained monomorphic VT was successful in a case of MVO with apical aneurysm, but there are no other studies so far of pacing therapy in such cases. The present case is the first in which dual-chamber pacing successfully prevented sustained VT through resolution of the high apical pressure.

In a patient with LVOT obstruction, continuous wave Doppler echocardiography can accurately and instantaneously estimate the outflow tract pressure gradient, which is useful for determining the efficacy of therapy. However, in patients with MVO, it is difficult to accurately calculate the peak pressure gradient from the modified Bernoulli equation because the Doppler flow signal from the apical chamber is aborted during late systole. The maximal flow velocity measured by the Doppler signal results in a significant underestimation of the peak pressure gradient. So far, little is known of the change in the Doppler flow pattern in a patient with MVO with apical aneurysm after pacemaker implantation. Takeuchi et al reported that dual-chamber pacing markedly affected the peak velocity and profile of isovolumetric relaxation flow in a case of MVO without apical asynergy. In the present case, when dual-chamber pacing reduced the mid-ventricular pressure gradient, a continuous wave Doppler signal showed disappearance of the early systolic ejection flow from the apex, the appearance of isovolumetric relaxation flow toward the apex and attenuation of the diastolic paradoxical jet flow toward the basal chamber.

The exact mechanism of these flow dynamics is unknown, but we speculate that the paradoxical motion or reduced motion of the interventricular septum by pacing might have cause the arrhythmia in this case, because an experimental study showed that automaticity was promoted by an increased afterload and preload.

Effect of Pacing on Mid-Ventricular Obstruction

Fig 4. Invasive assessment of the mid-ventricular pressure gradient. (A) Pullback pressure recording from the LV apex to the basal LV chamber during sino rhythm. The peak-to-peak pressure gradient was 175 mmHg. (B) Simultaneous pressure recording of the LV apex and the aorta (Ao) during sino rhythm. The peak-to-peak pressure gradient was 122 mmHg. (C) Simultaneous pressure recording of the LV apex and Ao during dual-chamber pacing. The pressure gradient decreased to 12 mmHg. V5 is precordial electrocardiographic lead.

Fig 5. Left ventriculogram with selective injection of contrast medium to the apical aneurysm. (A) The apical aneurysm during sino rhythm at end diastole and (B) during pacing at end diastole. (C) The apical aneurysm during sino rhythm at mid systole and (D) during pacing at mid systole. Arrows indicate the tunnel between the basal LV chamber and the apical aneurysm. Note that the tunnel at mid systole expanded after pacing induction, but the asynergy of the apical aneurysm was not affected.
would be higher than in the apical aneurysm at the isovolumeric contraction time, causing the delayed 2nd peak flow toward the apex. At the time of isovolumetric relaxation, the apical chamber with the aneurysm would have faster decrease in pressure than the basal chamber after pacing induction, because the decreased pressure overload in the apical chamber would induce asynchronous early segmental relaxation, as reported previously.25 Secondly, the apical induction, because the decreased pressure overload in the apical aneurysm would have faster toward the apex. At the time of isovolumetric relaxation, metric contraction time, causing the delayed 2nd peak flow would be higher than in the apical aneurysm at the isovolumetric pressure recording in the basal and apical LV chambers will assist in further analysis of such hemodynamics, but we did not perform such measurements because we were afraid of severe aortic regurgitation if we inserted 2 catheters through the aortic valve.

We conclude that the dual-chamber pacing can be effective for relief of MVO with an apical aneurysm and can prevent the development of sustained VT. In addition, the continuous wave Doppler flow pattern may be useful for evaluating the effect of pacing on the hemodynamics.

References