

Two Cases of Ventricular Tachycardia with Congenital Left Ventricular Malformation in an Adult

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Two patients without any risk factors for coronary artery disease presented with symptomatic, recurrent, nonsustained ventricular tachycardia. They were found to have a postero-lateral left ventricular aneurysm and diverticulum. Coronary angiography revealed normal coronary arteries. The 12-lead electrocardiogram showed sinus rhythm with frequent premature ventricular contractions. Their nonsustained ventricular tachycardias were reproduced by programmed electrical stimulation and was unresponsive to procainamide, mexiletine, and disopyramide. Aneurysmal resection and cryoablative surgery were performed. The pathological examination of the aneurysmal wall revealed focal defect of muscle fibers in case 1. On the other hand, the wall of case 2 was formed by all three cardiac layers. After surgery, ventricular programmed stimulation was negative, and premature ventricular contraction had disappeared. (*Ann Thorac Cardiovasc Surg* 2004; 10: 42–6)

Key words: congenital left ventricular malformation, ventricular tachycardia, ablating surgery

Introduction

True congenital malformation such as aneurysm and diverticulum of the left ventricle especially in an adult, is very rare. They may be asymptomatic or may present with systemic embolization, congestive heart failure, valvular regurgitation, and ventricular rupture.^{1,2)} Ventricular tachyarrhythmias are an unusual but significant complication.²⁻⁴⁾ We present two cases successfully treated with cryoablating surgery for recurrent nonsustained ventricular tachycardia (NSVT).

Case Report

Case 1

The patient was a previously healthy 60-year-old woman.

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She presented to the hospital for an episode of lightheadedness, flushing and syncope. It resolved spontaneously. On physical examination, no abnormal findings were obtained. The 12-lead electrocardiogram (ECG) revealed sinus rhythm with frequent premature ventricular contractions (PVCs). The long axial images of echocardiography revealed an aneurysm extending from the posterior left ventricular (LV) wall. This pouch showed paradoxical contraction, with a relatively thin wall. Thrombus was not recognized in the left atrium or the ventricle including an aneurysm. Coronary angiography revealed normal coronary anatomy (Fig. 1A, B). A left ventriculogram showed a wide-mouthed, multilobulated, 3 cm to 4 cm sized, aneurysm of the posterior wall, which had asynchronous contraction with LV (Fig. 1C, D). Electrophysiological testing was performed while the patient was off antiarrhythmic agents. NSVT, identical in morphology to the clinical tachycardia, was induced with stimulation from the endocardium of the right ventricular apex (Fig. 2). The patient was started on therapy with mexiletine. Three months later, the patient again had lightheadedness with syncope due to ventricular tachycardia (VT).

Case 2

The patient was a previously healthy 57-year-old man.

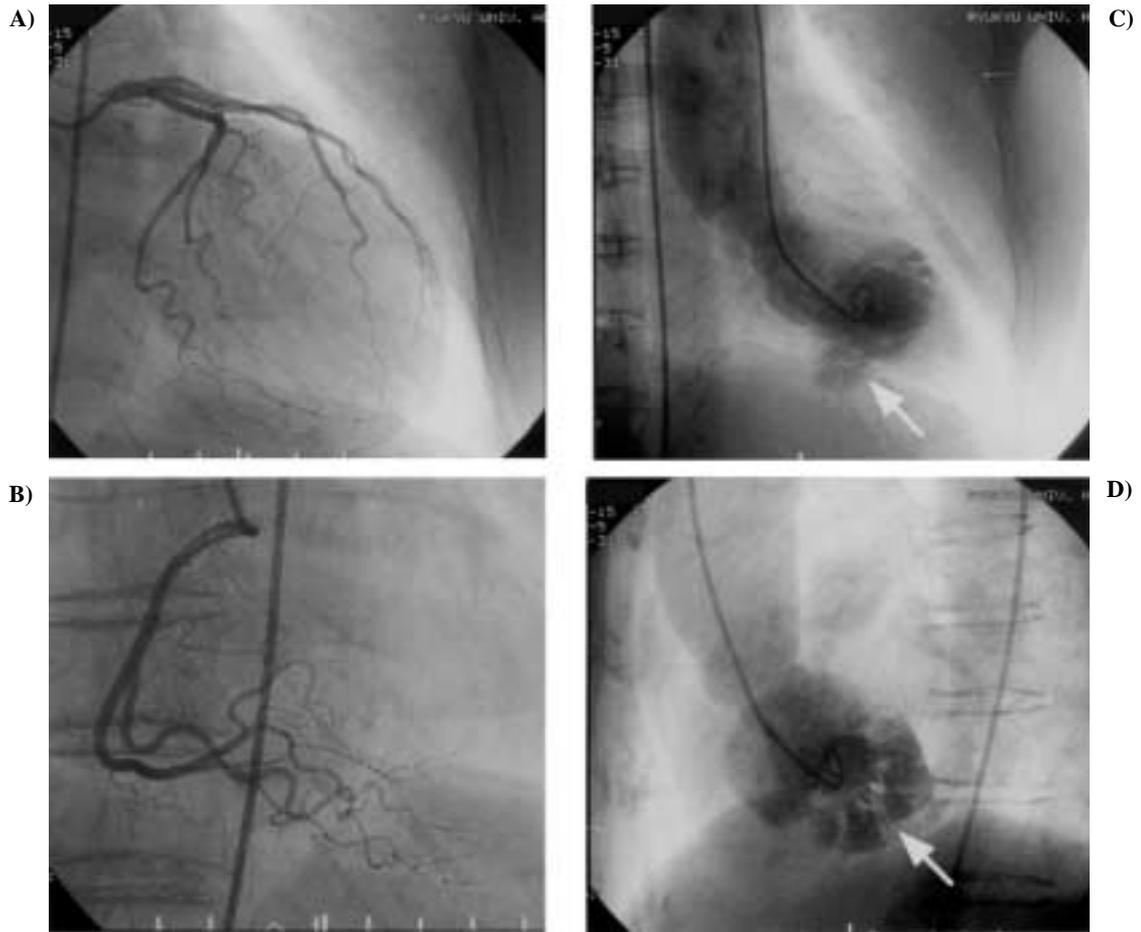


Fig. 1. Coronary angiography revealed normal coronary anatomy (A, B).

A left ventriculogram showed a wide-mouthed, multilobulated aneurysm (arrow) of the posterior wall, which had asynchronous contraction with LV in case 1. C: Anterior-posterior (A-P) view, D: lateral view of the left ventriculogram.

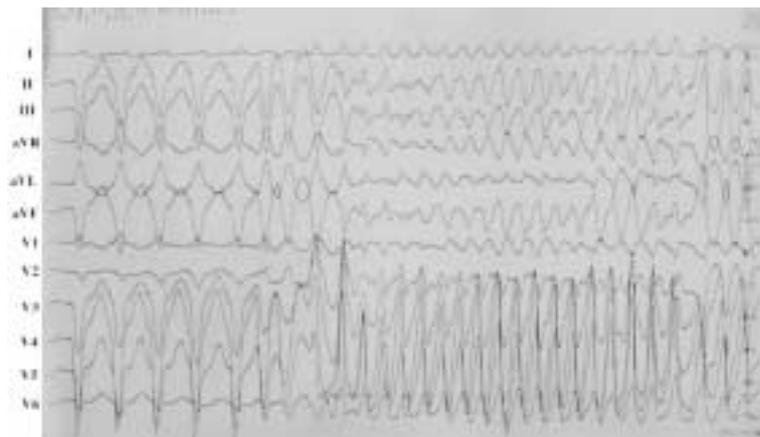


Fig. 2. Result of electrophysiological test of case 1. Nonsustained ventricular tachycardia was induced with intrastimuli from the right ventricular apex.

He presented to the hospital for general fatigue. On physical examination, no abnormal findings were obtained. Laboratory examination results were within normal limits. The 12-lead ECG revealed sinus rhythm with frequent premature PVCs as in case 1. The long axial images of

echocardiography revealed diverticulum in posterior LV wall. Coronary angiography revealed normal coronary anatomy. A left ventriculogram showed a small-mouthed, multilobulated diverticulum of the posterior wall (Fig. 3). He was found to have wide complex tachycardia with a

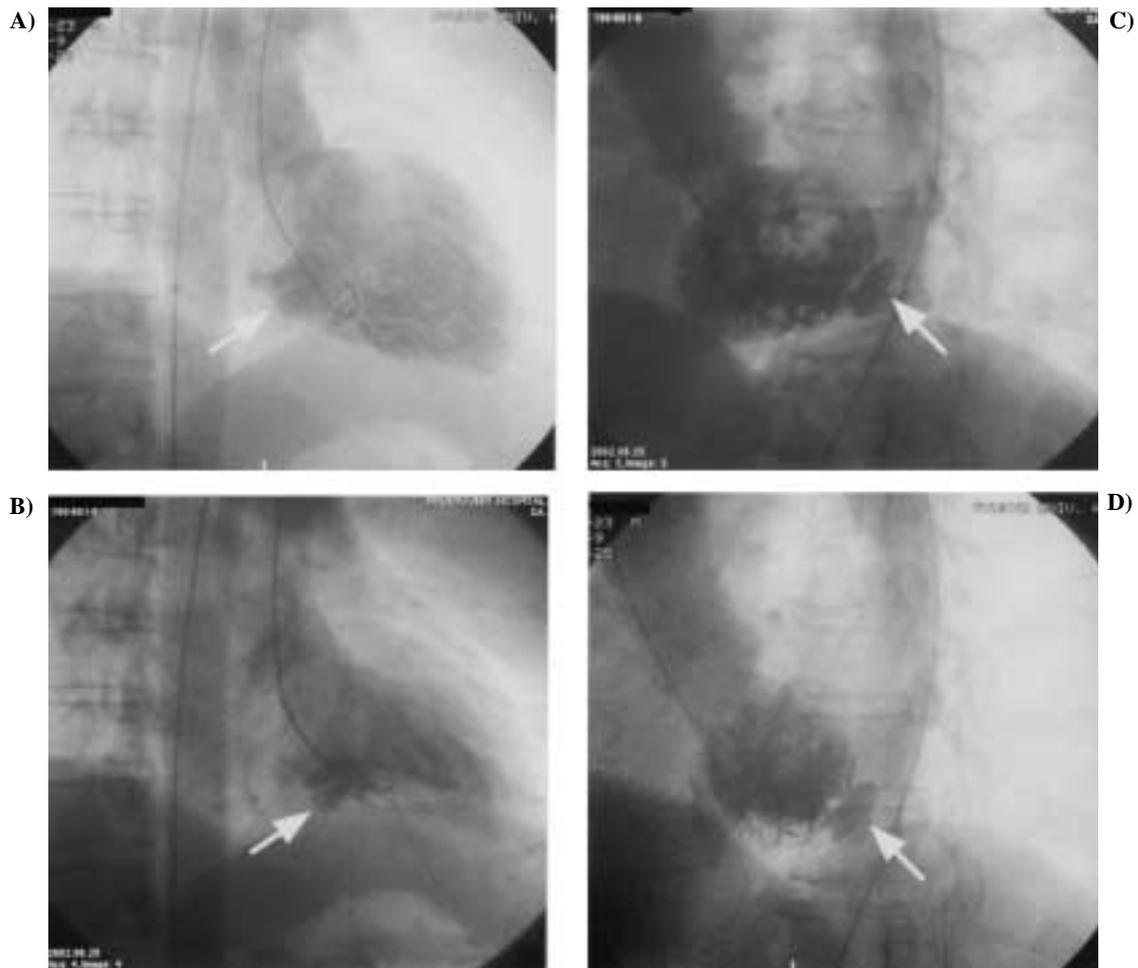


Fig. 3. A left ventriculogram showed a small-mouthed, multilobulated diverticulum (arrow) of the posterior wall in case 2. A: A-P view, diastolic phase, B: A-P view, systolic phase, C: lateral view, diastolic phase, D: lateral view, systolic phase of the left ventriculogram.

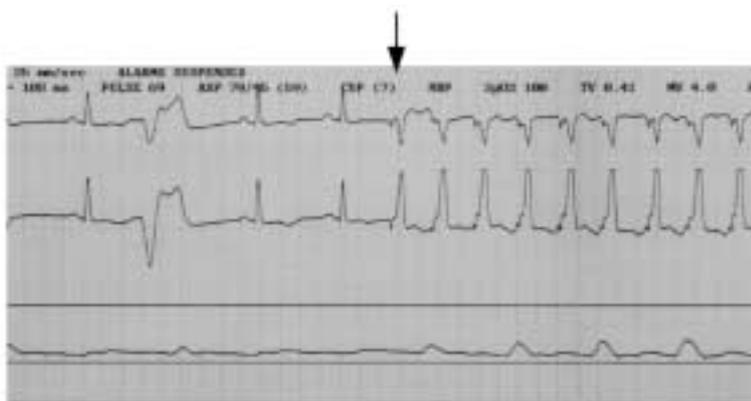


Fig. 4. Result of electrophysiological test of case 1. Wide complex tachycardia with left bundle-branch block configuration was induced with extrastimuli from the right ventricular apex.

left bundle-branch block configuration, and his heart rate was 200 beats per minute. In addition, a 24-hr Holter ECG demonstrated several two-beat to three-beat runs of repetitive ventricular complexes.

Ablating surgery was chosen in these two patients. During surgery, it was identified that NSVT had been induced

with extrastimuli from the right ventricular apex (Fig. 4). The aneurysm and diverticulum were found to consist entirely of grayish-white fibrous tissue, arising from the postero-lateral wall of the left ventricle bordered by the posterior descending and distal circumflex marginal vessels. Its motion was paradoxical with the rest of the left

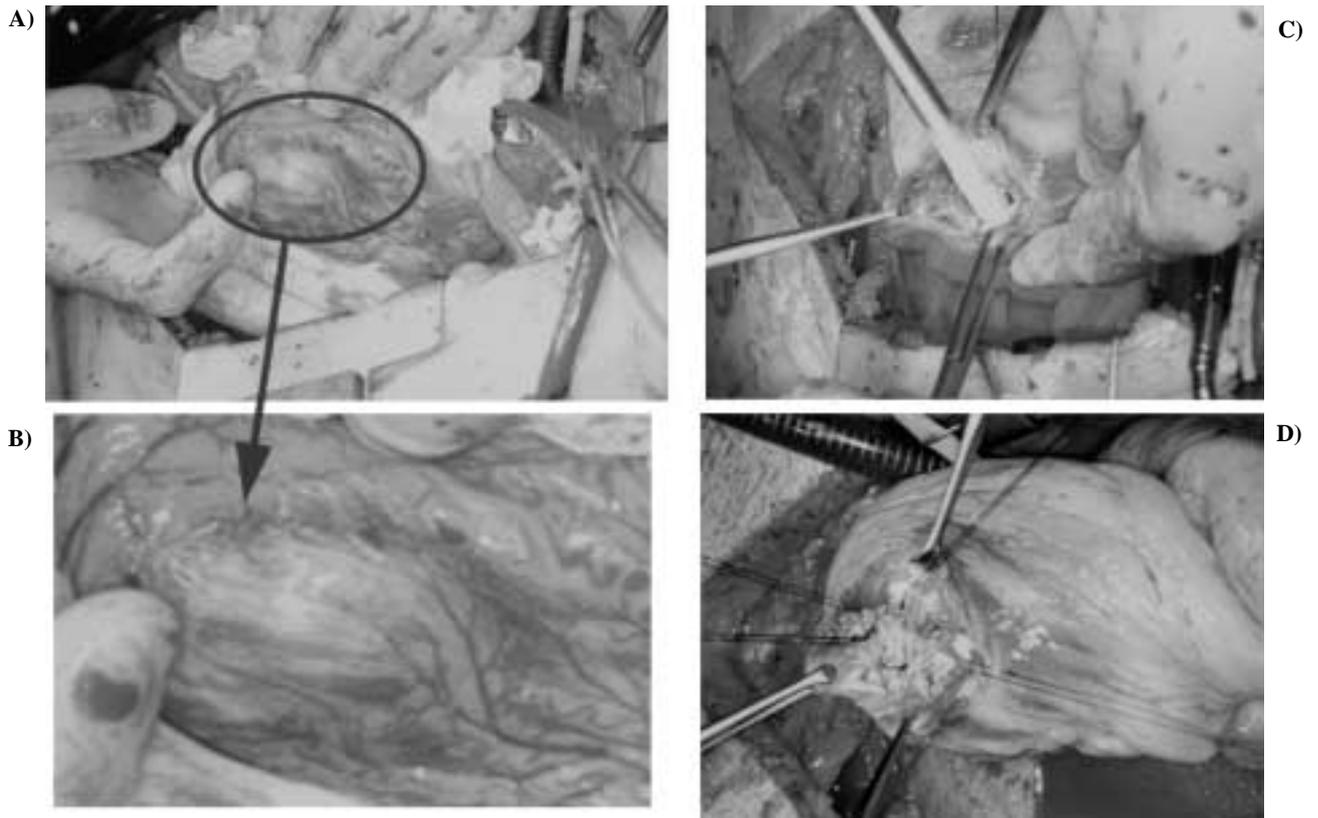


Fig. 5. Operative findings. The aneurysm was found to consist entirely of grayish-white fibrous tissue, arising from the posterior wall of left ventricle bordered by the posterior descending and distal circumflex marginal vessels (A, B). Cryoprobe was used to freeze the edge of resection concentrating on the focus of ventricular tachyarrhythmias (C). Endoaneurysmorrhaphy was performed as described by Dor using endoventricular circular sutures and a Dacron patch graft to reconstruct the left ventricle (D).

ventricle in case 1. Extensive resection of aneurysm and diverticulum was performed. Thrombus was not presented. The fibrous endocardial scar and the border zone between the totally fibrous tissue and the muscular tissue was unclear. Then a cryoprobe was used to freeze the resected margin concerning on the focus of ventricular tachyarrhythmias. Endoaneurysmorrhaphy was performed using endoventricular circular sutures with 2-0 prolene monofilament and Dacron patch graft to reconstruct the left ventricle. Double layers of continuous running suture were placed with reinforcing felt strips (Fig. 5). In case 1, pathological examination of the aneurysm wall revealed focal defect of muscle fibers. Various degrees of vacuolization of muscle fibers is present, and cross striations may be lost in some areas. On the other hand, the wall of the case 2 was formed by all three layers. After surgery, ventricular-programmed stimulation was negative in both cases, and PVCs disappeared on 24-hr Holter ECG. The patients were discharged on no antiarrhythmic therapy.

Discussion

Treisman et al. classified the defect as an aneurysm when its root of connection to the left ventricle was wide, and diverticulum if the connection was narrow.⁵⁾ The wall of true congenital diverticulum is formed by all three layers and contracts normally, whereas an aneurysm is generally a fibrous saccular region with paradoxical contraction.⁶⁾ Therefore, we diagnosed that case 1 as an aneurysm, and case 2 as diverticulum. Contraction of LV wall is different in case 1 (aneurysm) and case 2 (diverticulum), aneurysms expand, whereas diverticulum contract during ventricular systole. The etiology of these aneurysms and diverticula is unclear. It has been postulated to 1) represent congenital epicardial cysts, 2) derive from abnormal attachment of the heart tube to the yolk sac, 3) arise from weakness in the ventricular muscle with gradual outpouching from high ventricular pressures.⁴⁾

Ventricular tachyarrhythmias associated with the congenital LV aneurysm and diverticulum in an adult are an

unusual but significant complication, with only a few cases reported previously.²⁻⁴⁾ Maloy et al. reported the findings in 26-year-old woman with an apical aneurysm and refractory VT.¹⁾ She had a blind aneurysmectomy and was asymptomatic after five-months follow-up. Fellows et al. described three patients, two with sudden deaths and one with NSVT and syncope.²⁾ Their patient with syncope had inducible VT and unsuccessful epicardial cryoablation of the VT focus. However, this patient was stable on therapy with imipramine. Shen et al. reported a successfully treated case with mapping-guided surgery.⁴⁾ Even though the patient's VT had a recurrence one year and six months later, VT was suppressed by a previously ineffective agents.

The natural history of LV aneurysm and diverticulum is unknown. Therefore, Treisman et al. concluded that patients with LV aneurysm and diverticulum should be treated medically unless some other associated cardiac anomalies needed surgical correction or medical treatment cannot control the symptoms.⁵⁾ However, some cases of sudden death probably result from ventricular tachyarrhythmias.⁴⁾ Moreover, sudden death may occur by actual rupture of the aneurysm and diverticulum because of the thinness of its wall.¹⁻⁵⁾ In view of their high rate of complications and the risk of spontaneous rupture, some authors advocate surgical resection, even if asymptomatic.⁷⁾ Recently, radiofrequency catheter ablation has been used for treatment of VT.⁸⁻¹⁰⁾ Some investigator reported that in patients with apparently normal hearts, VT might only involve the subepicardial myocardium.⁸⁻¹⁰⁾ They concluded that VT, which has a subepicardial arrhythmogenic substrate, might be amenable to epicardial ablation. In patient with associated VT, electrophysiological testing and ablative therapy should be an integral part of the diagnostic and therapeutic regimen.

Our patients had a postero-lateral left ventricular aneurysm and diverticulum, which was symptomatic with recurrent NSVT and syncope refractory to antiarrhythmic therapy. We performed surgical resection and cryoablative surgery without mapping. To avoid recurrence of ventricular arrhythmia, we ablated the aneurysmal and diverticular base sufficiently. On the other hand, it has been reported that surgical cryoablation of the focus of the tachycardia which was reconfirmed by epicardial and endocardial mapping during operation.^{11,12)} It has been recognized that intraoperative VT mapping was effective and useful for many reports, we reflected that

epicardial and endocardial mapping should be performed during operation. Fortunately, our patients VT disappeared after surgery. Although long-term follow-up is necessary, the patients had been discharged on no antiarrhythmic therapy and have been free of symptoms with follow-up over two years and one year, respectively.

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