Ventricular Tachycardia with Congenital Ventricular Diverticulum*

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A 24-year-old man presented with symptomatic, recurrent, sustained ventricular tachycardia (VT). He was found to have a basal inferior left ventricular diverticulum. His sustained VT was reproduced by programmed electrical stimulation and was unresponsive to procainamide, tocainide, propafenone, and flecainide. Endocardial mapping followed by resection and cryoablation surgery was performed. The patient had only one recurrence after 18 months, with subsequent control with procainamide for over 14 months.

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Congenital ventricular aneurysms or diverticula are rare, with less than 100 cases in the literature. They may be asymptomatic or may present with systemic embolization, congestive heart failure, valvular regurgitation, and ventricular rupture. Ventricular tachyarrhythmias are an unusual but significant complication, with only six cases reported previously. We present the first case successfully treated with mapping-guided surgery.

CASE REPORT

The patient was a previously healthy 24-year-old Korean-Japanese man. In March 1984, he had a four-hour episode of rapid palpitations with lightheadedness and flushing. It resolved spontaneously. Subsequently, his cardiac examination was notable for a grade 2/6 apical systolic murmur and a parasternal lift. The 12-lead ECG revealed inferior Q waves with T-wave inversion. The chest x-ray film was normal. A treadmill stress test elicited several four-beat to five-beat runs of repetitive ventricular complexes. The patient was started on therapy with disopyramide empirically.

Two months later, the patient again had palpitations with jaw pain and lightheadedness. He was found to have a wide complex tachycardia with left bundle-branch block configuration, superior axis, and heart rate of 210 beats per minute (Fig 1), which was unresponsive to intravenous verapamil. Direct-current (DC) cardioversion restored sinus rhythm. Propanolol was added to the regimen.

Subsequently, the patient had several recurrences of the same tachycardia which again required DC cardioversion. Coronary angiography revealed normal coronary anatomy. A left ventriculogram showed a wide-mouthed, multilobulated 4-cm to 5-cm diverticulum of the posterior left ventricular wall, which had asynchronous contraction with the rest of the left ventricle. Overall left ventricular contractility was moderately depressed. Magnetic resonance imaging also clearly defined the diverticulum (Fig 2).

Electrophysiologic testing was performed while the patient was off antiarrhythmic agents. Sustained VT, identical in morphology to the clinical tachycardia, was inducible with three extrastimuli from the right ventricular apex. After procainamide, the same tachycardia was still inducible, with cycle length slowed from 310 ms to 300 ms. Trials with tocainide and propafenone were likewise unsuccessful, and the patient had increased ventricular ectopy and nonsustained VT with flecainide.

Finally, ablative surgery was chosen. Before surgery, endocardial catheter mapping during VT revealed the earliest endocardial ventricular electrogram to be inferoseptal, near the base of the left ventricle (Fig 3), measuring 40 ms earlier than the onset of the surface QRS complex.

During surgery the diverticulum was found to consist entirely of

FIGURE 1. Twelve-lead ECG, showing patient's VT.

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greyish-white fibrous tissue, arising from the base of the left ventricle along the coronary sinus, bordered by the posterior descending and distal circumflex marginal vessels. Its motion was asynchronous with the rest of the left ventricle. The VT was induced in the beating heart and the earliest site of epicardial breakthrough was at the base of the right ventricle, very close to the septum. A small ventriculotomy was performed through the diverticulum, and endocardial VT mapping was repeated. The earliest endocardial breakthrough was similar to the preoperative catheter map, with the earliest site near the superior rim of the aneurysm, at about 11 o’clock. Extensive resection was performed up to the base of the mitral annulus. Then a cryoprobe was used to freeze the base, concentrating on the focus identified by mapping. The papillary muscles were preserved. A double Teflon patch was placed over the resected area, with a Dacron hood over the patch, reconstructing the left ventricle. After surgery, ventricular programmed stimulation was entirely negative, and the patient was discharged on no antiarrhythmic therapy.

Early after discharge, the patient developed mild heart failure, with a nocturnal cough and fatigue on exertion. An echocardiogram revealed a residual bulge in the inferobasal region of the left ventricle. The overall left ventricular contractility was still moderately depressed. The patient was treated with furosemide, and his symptoms resolved. He had one recurrence, with the identical VT, 18 months after surgery. He became free of symptoms again after initiation of oral procainamide, with follow-up of over 14 months after the episode.

DISCUSSION

Congenital ventricular diverticula are rare. The largest series was from the Texas Heart Institute, reporting ten cases between the years 1965 and 1984 (accounting for 0.076
percent of over 13,000 congenital heart operations.1 The total number in the literature is about 100. These diverticula may occur in the left or right ventricle, with the former more common. Rarely, they may arise from both ventricles.2 Pathologically, ventricular diverticula may be differentiated into muscular and fibrous types.3 The muscular type arises from the apex of the ventricle as an outpouching involving the myocardium, endocardium, and occasionally the pericardium. The point of connection is generally narrow. It is frequently accompanied by midline abnormalities (defects in pericardium, diaphragm, or anterior abdominal wall). There is an association with cyanotic congenital heart disease, and thus, the condition is often diagnosed early in life. The fibrous type arises either in apical or subvalvular positions and predominantly occurs in blacks or Africans. The subvalvular form, which is more common, is frequently accompanied by mitral or aortic insufficiency. Both the apical and subvalvular fibrous forms have been complicated by systemic embolization. Fibrous diverticula are never associated with midline defects or congenital cardiac malformations. The etiology of these diverticula is unclear. They have been postulated to represent congenital epicardial cysts,5 to derive from abnormal attachment of the heart tube to the yolk sac, or to arise from weakness in the ventricular muscle with gradual outpouching from high ventricular pressures.6

The age of discovery has been as early as 38 hours after birth7 and as late as in a 62-year-old man.8 The diverticulum may be asymptomatic, found incidentally with an abnormal ECG or chest x-ray film, or detected during investigation of congenital heart disease or midline abnormalities. They may also present with embolic events,9 congestive heart failure,10 valvular insufficiency,9 or even sudden death. Sudden death may occur by actual rupture of the diverticulum,1 more commonly with the fibrous than the muscular type. Some cases of sudden death probably result from ventricular tachyarrhythmias.

There have only been six cases of congenital diverticula with documented VT in the literature.6-7 Maloy et al.6 reported the findings in a 26-year-old woman with an apical aneurysm and refractory VT. She had a blind aneurysmscotomy and was asymptomatic on five-month follow-up. Fellows et al.9 described three patients, two with aborted sudden deaths and one with nonsustained VT and syncpe. One patient with aborted sudden death was put on disopyramide and died without electrophysiologic evaluation. The patient with syncpe had inducible VT and unsuccessful epicardial cryoablation of the VT focus, which was at the site of a small anterior aneurysm close to the left main coronary artery. This patient was stable on therapy with imipramine. The third patient had inducible VT and was alive on therapy with amiodarone. Our patient had a subvalvular fibrous aneurysm, which was symptomatic with recurrent sustained VT refractory to antiarrhythmic therapy. His VT origin was identified before and during surgery by mapping, which permitted accurate ablation of the focus. Even though the patient had a recurrence 1½ years later, the focus was probably sufficiently modified to allow a prolonged arrhythmia-free duration and eventual suppression by a previously ineffective agent.

The natural history of patients with congenital diverticula is unknown. In view of their high rate of complications and the risk of spontaneous rupture, some authors advocate surgical resection, even if asymptomatic.1 In patients with associated VT, electrophysiologic testing and mapping-guided ablative therapy should be an integral part of the diagnostic and therapeutic regimen.

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Life-Threatening Status Asthmaticus at 12.5 Weeks’ Gestation

Report of a Normal Pregnancy Outcome

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A 26-year-old woman had a life-threatening attack of status asthmaticus at 12.5 weeks of pregnancy. Subsequently, an apparently normal male infant was born at full-term.

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Asthma in pregnancy has been reviewed extensively.11,12 However, few cases of pregnancy outcome after life-threatening status asthmaticus have been described. We report the birth of an apparently normal full-term male infant after status asthmaticus at 12.5 weeks13 of gestation (10.5 weeks postconception).

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