CASE REPORT

Hypertrophic cardiomyopathy with apical aneurysm: a case of catheter and surgical therapy of sustained monomorphic ventricular tachycardia

Massimo Mantica, Paolo Della Bella, Vincenzo Arena

Istituto di Cardiologia dell' Universita' degli Studi, Centro Ricerche Cardiovascolari del Consiglio Nazionale delle Ricerche, Centro Cardiologico Fondazione Monzino, IRCCS, Milan, Italy
M Mantica
P Della Bella
V Arena

Correspondence to:
Dr P Della Bella, Centro Cardiologico Fondazione Monzino, Via Parea 4-20138, Milan, Italy.

Accepted for publication 17 February 1997

Abstract

The case is presented of a patient with hypertrophic cardiomyopathy, midventricular obstruction, apical aneurysm, and very frequent episodes of sustained monomorphic ventricular tachycardia (VT) unresponsive to common antiarrhythmic drugs. Left ventricular catheter mapping during sinus rhythm suggested the presence of an extensively scarred apical region; early fractionated ECGs were recorded at the neck of the aneurysm during monomorphic VT, suggesting a possible role of this region as VT substrate. Radiofrequency delivery at these sites stopped the VT and it was no longer inducible; however, it spontaneously recurred the following day. An apical aneurysmectomy, guided by the results of catheter mapping, was performed and was successful in preventing arrhythmic recurrences during 12 months' follow up.

(*Heart* 1997;77:481–483)

Keywords: hypertrophic cardiomyopathy; ventricular tachycardia; catheter ablation; aneurysmectomy

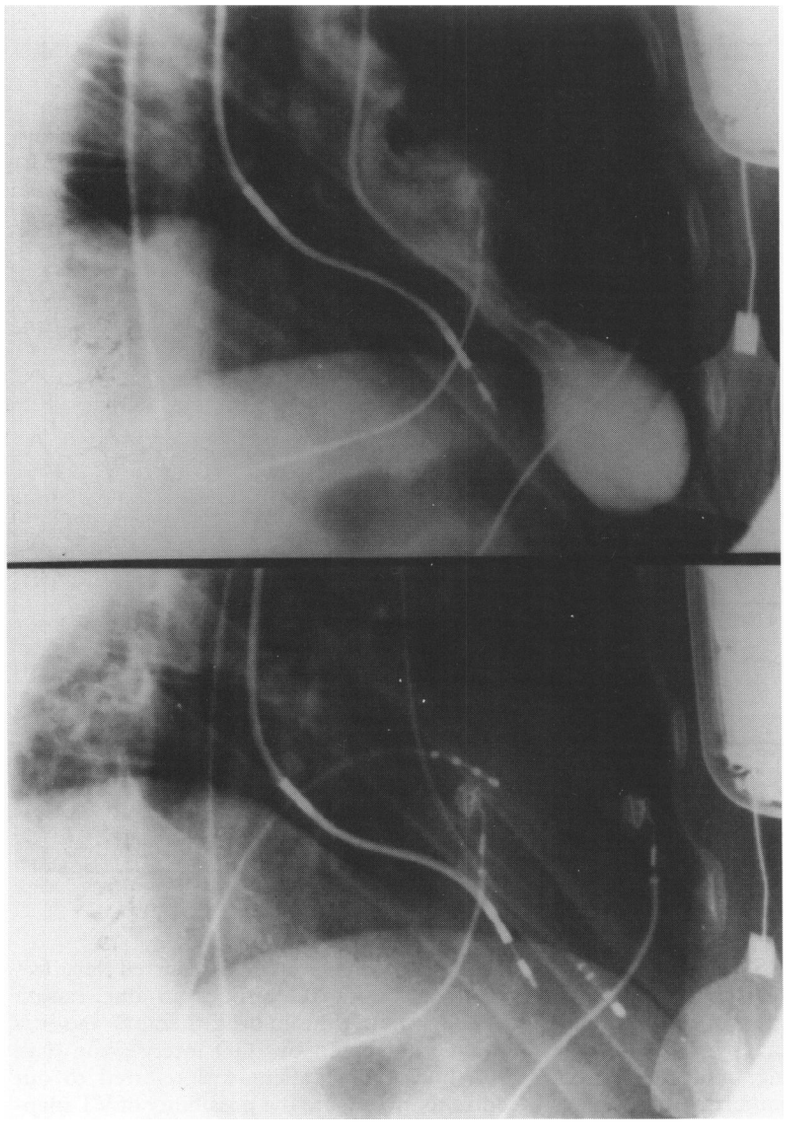

Figure 1 (Top) The left ventriculogram in right anterior oblique projection showing the apical aneurysm. The implantable cardioverter-defibrillator electrode in the right ventricular apex is also evident. (Bottom) The ablation catheter has been advanced through the narrow channel to the neck of the apical left ventricular aneurysm.

The mechanisms of sudden death and syncope in hypertrophic cardiomyopathy (HCM) are not fully known but may be related to lethal ventricular arrhythmias, myocardial ischaemia or to the severity of left ventricular outflow tract obstruction. The most common ventricular arrhythmias observed with programmed electrical stimulation in these patients include polymorphic ventricular tachycardia (VT) and fibrillation (VF), while monomorphic VT is less frequent.[1] Clinically stable VT in these patients is rare: a relation with the presence of midventricular obstruction and apical aneurysm has been proposed.[2]

Case report

A 34 year old male with familial HCM had been followed for about two years because of palpitations and angina, with evidence of clinical episodes of sustained monomorphic VT. Echocardiogram demonstrated asymmetric left ventricular hypertrophy with a midventricular obstruction (20 mm Hg gradient in basal condition), and an apical aneurysm confirmed by left ventriculography (fig 1); coronary angiography was normal. Sustained monomorphic VT was inducible at electrophysiological study (EPS) with characteristics similar to the clinical VT; good haemodynamic tolerability, left bundle branch block morphology,

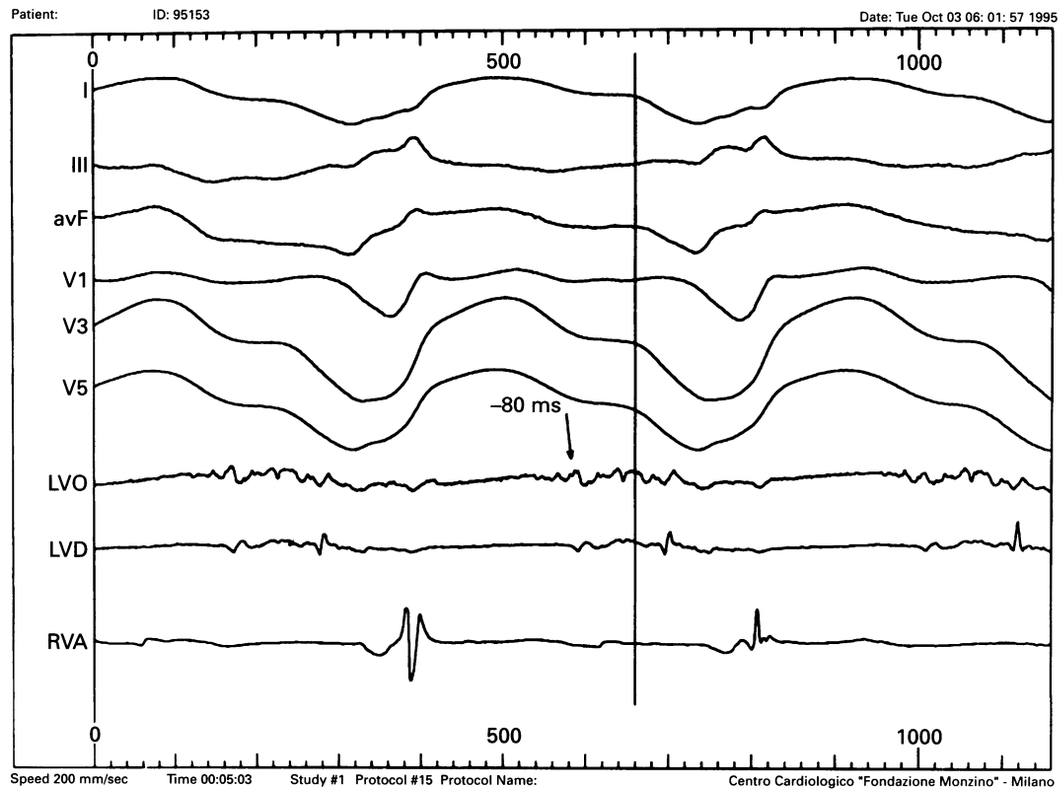

Figure 2 Low amplitude fractionated ECGs recorded during VT at the ablation site, near the neck of the aneurysm, from the tip of the ablation catheter (LV distal). The local prematurity versus ECG leads is about −80 ms.

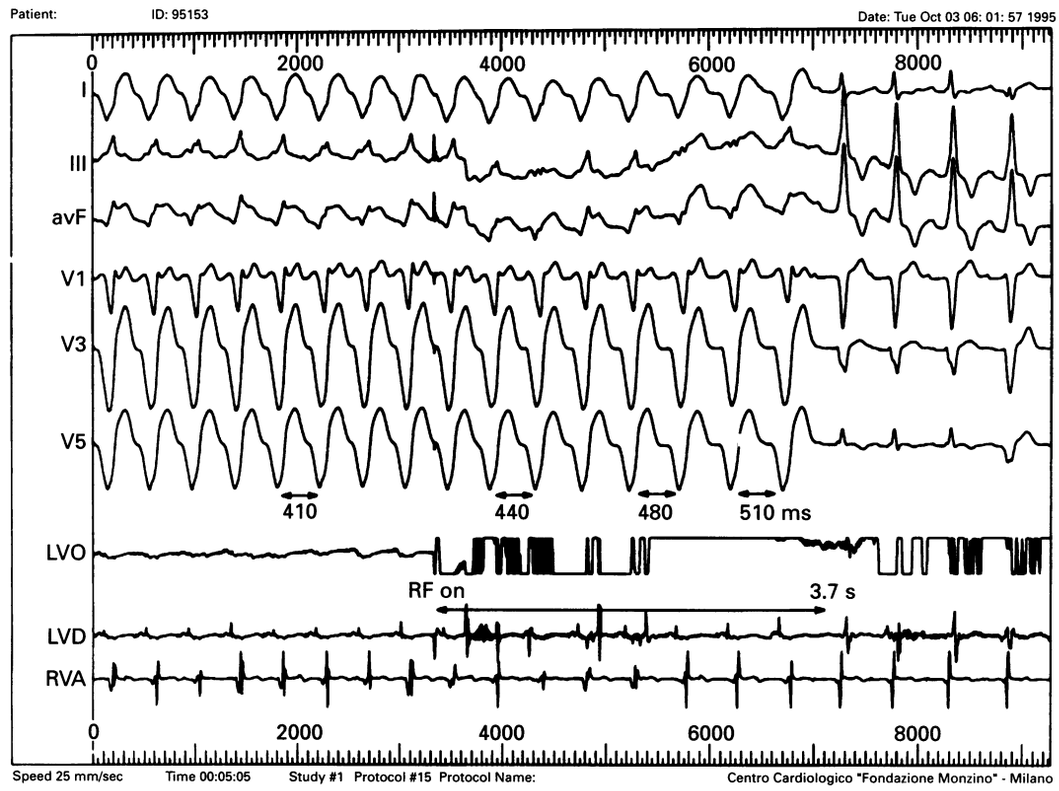

Figure 3 VT stops after 3·7 seconds and sinus rhythm is restored; the VT cycle length increases during radiofrequency catheter ablation from 410 to 510 ms.

inferior axis, negative QRS complexes in the precordial leads, and a cycle length of 410 ms. The arrhythmia frequently degenerated into VF after the common manoeuvres to interrupt it with ventricular pacing, and it was not responsive to common antiarrhythmic drugs (amiodarone or sotalol, also in association with class 1 antiarrhythmic drugs). The patient, therefore, was given an implantable cardioverter-defibrillator (Jewel PCD 7219C, Medtronic). However, VT recurred very frequently (about 70 episodes in one month despite antiarrhythmic drug therapy) and was always interrupted by ICD intervention with DC shock. The patient was referred to our centre to investigate the possibility of VT mapping and radiofrequency catheter ablation.

Left ventricular endocardial mapping was performed during sinus rhythm, advancing a quadripolar 4 mm tip steerable catheter

through the narrow mid-ventricular channel into the aneurysm (fig 1). The catheter mapping of the apical region revealed no electrical activity, thus suggesting complete endocardial scarring of the chamber. Low amplitude fractionated ECGs were recorded at the proximal part of the aneurysm and throughout the channel connecting to the left ventricle. A sustained VT, identical to the clinical one, was induced; early diastolic activity could be recorded in the proximal part of the aneurysm (−50 ms) and in the posterior-middle part of the channel with more prematurity (−80 ms) (fig 2).

Radiofrequency energy, with temperature set at 55°C (Cardiorhythm Atakr, Medtronic), was delivered for 50–60 seconds at sites showing the highest degree of prematurity. The VT stopped without premature events 3·7 seconds after the beginning of the second radiofrequency delivery (fig 3). No ventricular arrhythmias could be induced after this termination. The same VT, however, recurred spontaneously the following day. Thus, surgical resection of the ventricular aneurysm, guided by the results of catheter endocardial mapping, was undertaken.

Under normothermic cardiopulmonary bypass, left ventricular apical aneurysmectomy was associated with septal myotomy and freezing of the channel. At EPS, performed one week after surgery, A-V conduction was normal and no ventricular arrhythmias could be induced. The patient was discharged on β blockers (metoprolol 50 mg twice a day). Twelve months later the patient was free from arrhythmic recurrence.

Discussion

PREVALENCE OF MONOMORPHIC VT IN HCM

Sustained monomorphic VT in patients with HCM is an uncommon finding. Fananapazir *et al* reported inducibility of sustained VT in 82 of 230 patients (36%) selected for EPS because of out of hospital cardiac arrest, syncope, palpitations, VT, or strong familiar history of sudden cardiac death; however, in 66% of the patients the induced VT was polymorphic.[1] No data about the incidence of apical aneurysm in their patients were reported. Clinical sustained monomorphic VT in patients with HCM is uncommon but may be underestimated because of early degeneration into VF that has been well documented in these patients during EPS.[3] Alfonso *et al* found only two patients with clinical monomorphic sustained VT among 51 consecutive patients with HCM.[2] In both patients, they described echocardiographic and ventriculographic evidence of apical aneurysm, with angiographically normal coronary arteries. They also reported the presence of ST segment elevation in precordial electrocardiographic leads, a pattern that was evident in our patient and previously proposed as a marker of left ventricular aneurysm in HCM.[4]

APICAL ANEURYSM IN HCM

Midventricular obstruction is an uncommon variant of left ventricular obstructive HCM.[5,6] It may lead to apical aneurysm development, creating two distinct (basal and apical) left ventricular chambers. Doppler echocardiographic evidence of a paradoxical systolic flow directed towards the apex has been reported.[5] High apical diastolic pressure and wall stress may lead to cellular necrosis, possibly through an ischaemic mechanism. Scintigraphic evidence of fixed and transient inferoapical perfusion defects, after dipyridamole infusion, has been described in these patients.[5]

ELECTROPHYSIOLOGICAL SUBSTRATE OF VT IN HCM

Marked disarray and disorganisation of myofibrils and myofilaments, with variations in diameter and length of myocardial cells and interstitial fibrosis, have been proposed to alter the range of both conduction velocity and refractory periods of myocardial fibres in patients with HCM.[7] Thus, the dispersion of conduction within the ventricles may form one component of the substrate for reentry and VF. In patients with midventricular obstruction, the presence of an apical aneurysm may lead to a different arrhythmic substrate, related to local ischaemic phenomena. In our patient the early fractionated activity recorded during VT mapping at the neck of the aneurysm suggested the presence of an extensively scarred region, a possible substrate for sustained monomorphic VT. This region was the target of radiofrequency catheter ablation and, in case of failure, of a surgical approach, with good success in preventing arrhythmic recurrences.

1 Fananapazir L, Chang AC, Epstein SE, McAreavey D. Prognostic determinants in hypertrophic cardiomyopathy. Prospective evaluation of a therapeutic strategy based on clinical, Holter, hemodynamic, and electrophysiological findings. *Circulation* 1992;**86**:730–40.

2 Alfonso F, Frenneaux MP, McKenna WJ. Clinical sustained uniform ventricular tachycardia in hypertrophic cardiomyopathy: association with left ventricular apical aneurysm. *Br Heart J* 1989;**61**:178–81.

3 Gilligan DM, Missouris CG, Boyd MJ, Oakley CM. Sudden death due to ventricular tachycardia during amiodarone therapy in familial hypertrophic cardiomyopathy. *Am J Cardiol* 1991;**68**:971–3.

4 Gordon EP, Henderson MA, Rakowski H, Wigle ED. Midventricular obstruction with apical infarction and aneurysm formation [abstract]. *Circulation* 1984;**70 (suppl II)**:145.

5 Barbier P, Bartorelli A. Doppler evidence of abnormal intracavitary systolic and diastolic flow in hypertrophic cardiomyopathy with midventricular obstruction. *Am Heart J* 1993;**126**:483–7.

6 Webb JG, Sasson Z, Rakowsky H, Liu P, Wigle ED. Apical hypertrophic cardiomyopathy: clinical follow-up and diagnostic correlates. *J Am Coll Cardiol* 1990;**15**:83–90.

7 Saumarez RC, Camm AJ, Panagos A, Gills JS, Stewart JT, de Belder MA, *et al*. Ventricular fibrillation in hypertrophic cardiomyopathy is associated with increased fractionation of paced right ventricular electrograms. *Circulation* 1992;**86**:467–74.