Secondary Arrhythmogenic Right Ventricular Cardiomyopathy Decades After Operative Repair of Tetralogy of Fallot

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We describe a 47-year-old man who underwent heart transplantation (HT) for severe right-sided heart failure and periodic episodes of ventricular tachycardia (VT) 43 years after operative repair of tetralogy of Fallot (T of F). The right-ventricular outflow tract, the site where a patch had been placed 4 decades earlier, was aneurysmal. Such development decades after operative repair of T of F of both aneurysm and episodes of VT is probably more common than previously realized. **(Am J Cardiol 2014;114:806−809)**

In July 2011 we reported a 41-year-old man who underwent heart transplantation (HT) because of severe right-sided heart failure and periodic ventricular tachycardia (VT) after operative repair of tetralogy of Fallot (T of F) at age 6.[1] The present report was prompted by studying a similar patient, aged 47, who underwent HT also because of severe right-sided heart failure associated with periodic episodes of VT after operative repair of T of F at age 4. Both patients developed huge right ventricular outflow aneurysms.

Case Description

A 47-year-old male manual laborer had repair of T of F at 4 years of age. Thereafter, he was asymptomatic until age 33 years, when he had his first symptomatic episode of VT and an intracardiac defibrillator was inserted. Evidence of right-sided failure appeared about the same time and it gradually progressed thereafter. An electrocardiogram before an intracardiac defibrillator was inserted is shown in Figure 1. A computed tomographic image of the heart just before HT is shown in Figure 2. Three months after HT (January 2014) the patient was asymptomatic. Photographs of the explanted heart are shown in Figures 3 and 4.

Discussion

Described herein is a patient who had HT at age 47 for severe right-sided heart failure and episodic VT occurring several decades after repair of T of F at age 4. The patient described is virtually identical to a patient reported 2 years earlier with a similar scenario (Table 1). The earlier report also described a huge right ventricular outflow tract aneurysm involving the widening patch utilized when eliminating the subvalvular right ventricular outflow tract obstruction. To study 2 similar patients in a 2-year period suggests that aneurysmal formation in the right ventricular outflow tract several decades after repair of T of F may not be an uncommon occurrence but indeed potentially a common late occurrence. The development of episodes of VT with development of the right ventricular outflow aneurysm of course might be described as secondary or acquired arrhythmogenic right ventricular cardiomyopathy/dysplasia.

Shown in Table 2 are selected reports of patients having late follow up after repair of T of F in childhood.[2−9] Of the 1964 patients included none appeared to have a follow up as long as 40 years as did our present patient and our previously described patient (Table 1). Nevertheless, certainly many of the reported patients having pulmonic valve replacement with or without partial excision of the right ventricular outflow tract did have subvalvular aneurysms and some had "arrhythmias" (type not specified).

The other unusual feature here is the use of HT to eliminate both the right-sided severe heart failure and the periodic episodes of ventricular tachycardia. As shown in Table 2, however, HT is infrequently employed for management of late complications after repair of T of F. Only 4 of the previously mentioned 2638 patients (Table 2) underwent HT late after previous "repair" of T of F, and 3 of them previously had homograft replacement of the pulmonic valve.

Disclosures

The authors have no conflicts of interest to disclose.

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See page 806 for disclosure information.

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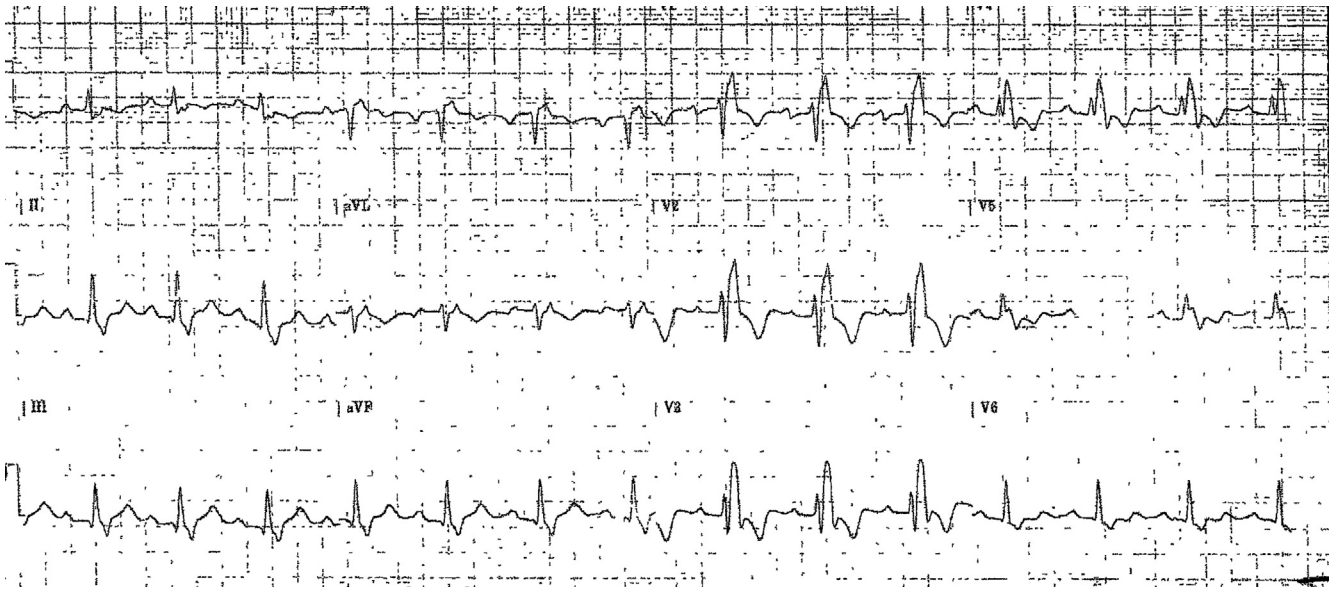

Figure 1. Electrocardiogram in the patient described before an intracardiac defibrillator was inserted. The electrocardiogram discloses a complete left bundle branch block pattern. Shows a complete right bundle branch block pattern. The rhythm is sinus.

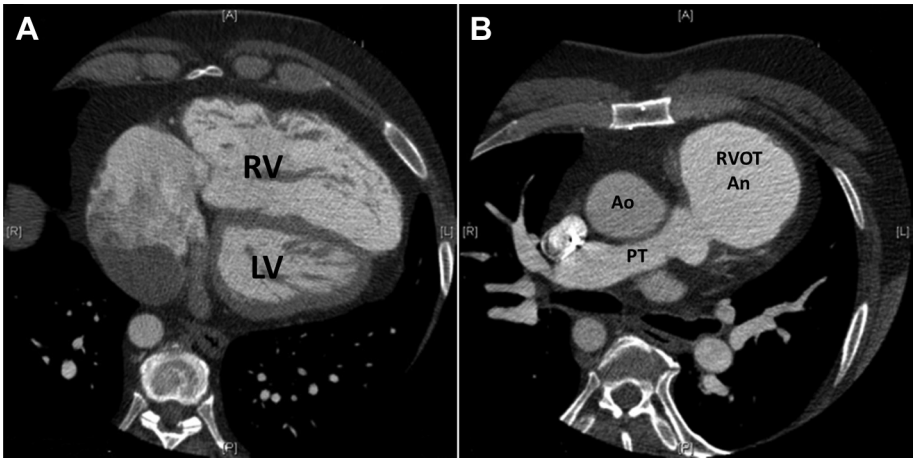

Figure 2. Computed tomographic imaging of the heart in the patient described before cardiac transplantation. *(A)* View showing the right ventricular cavity (inflow portion) and the much smaller left ventricular cavity (LV). The dilated right atrium is also seen as well as the right-sided aortic arch (AO). *(B)* A view showing the marked dilatation of the outflow portion of the right ventricle (RV) just beneath the pulmonic valve (PV). PT = pulmonic trunk; RVOT An = right ventricular outflow tract aneurysm.

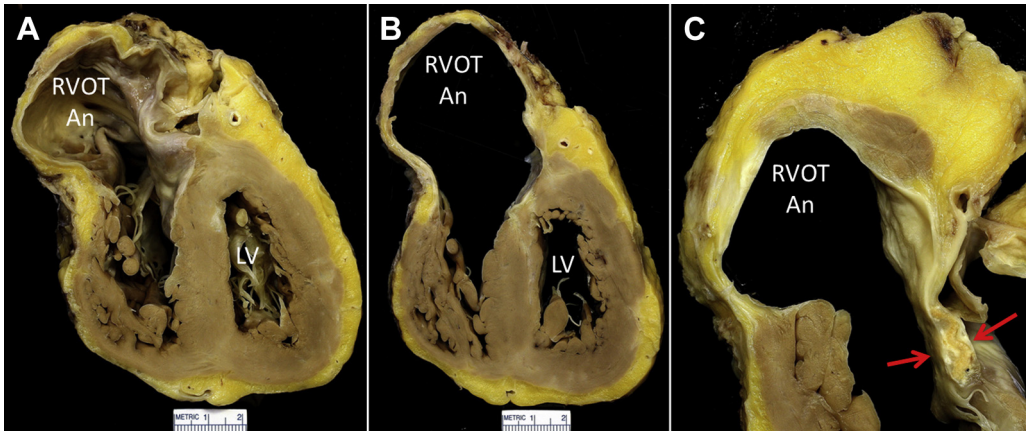

Figure 3. Partial four chamber cuts of the heart in the patient described. *(A)* View of the very dilated right ventricular outflow tract (RVOT), partially destroyed pulmonic valve (PV), left anterior descending coronary artery residing in the excessive subepicardial adipose tissue, right and left ventricular cavities exposing both tricuspid and mitral valves. *(B)* Another view showing the much dilated right ventricular outflow tract and the non-dilated right ventricular inflow tract. The scarring in the cephalad portion of the ventricular septum is the area where the previous ventricular septal defect was closed, approximately 40 years earlier. *(C)* Another view of the right ventricular outflow tract with calcium in the area where a patch *(arrows)* was used to close the ventricular septal defect (CA++). An = aneurysm; LV = left ventricle.