Total Disconnection of the Right Ventricular Free Wall: Surgical Treatment of Right Ventricular Tachycardia Associated with Right Ventricular Dysplasia

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SUMMARY Arrhythmogenic right ventricular dysplasia is a myopathy that affects the right ventricular free wall (RVFW) and gives rise to recurrent reentrant ventricular tachycardia (VT). Because the entire right ventricle is potentially arrhythmogenic, ablating a single site of VT may not eliminate the arrhythmia. We developed an operation to confine any arrhythmic activity arising from the right ventricle to that chamber: total disconnection of the RVFW from the left ventricle.

We performed RVFW disconnection in two patients with refractory VT associated with arrhythmogenic right ventricular dysplasia. At least two sites of origin of morphologically distinct VT were identified in the RVFW in each patient. RVFW disconnection was carried out under normothermic cardiopulmonary bypass. An encircling incision was made along the attachment of the RVFW to the aortoventricular unit and the tricuspid annulus; the right coronary artery and its RVFW branches were left intact. Electrical activity of the two chambers became dissociated, and VT arising from the RVFW was confined to that chamber. Postoperatively, there was no clinical evidence of hemodynamic impairment (follow-up 4 months and 3 months).

Left ventricular function was unchanged and right ventricular flow was maintained by atrial contraction and motion of the septum toward the RVFW during left ventricular systole. One patient had incessant right ventricular tachycardia confined to the RVFW for 3 weeks. We conclude that RVFW disconnection is feasible and applicable to patients with refractory VT originating in the diffusely diseased RVFW.

ARRHYTHMOGENIC right ventricular dysplasia (ARVD) is a clinical entity characterized by predominantly right-sided cardiomyopathy associated with ventricular tachycardia (VT). Since 1973, the surgical treatment of refractory VT associated with ARVD has consisted of simple ventriculotomy at the site of earliest epicardial activation during VT in an attempt to interrupt the presumed reentrant circuit responsible for tachycardia. This surgical procedure was not uniformly successful; some patients subsequently developed VT at a right ventricular site distant from the original one. Since any area of the right ventricular myocardi- um is potentially arrhythmogenic in this disease, we thought that surgical treatment must be directed at "isolating" arrhythmic activity within the entire right ventricle. This could be achieved by dissecting the right ventricular free wall (RVFW) from its left ventricular attachments. When subsequently resutured, the fibrous scar would confine right ventricular VT to that chamber.

We describe two patients who successfully underwent RVFW disconnection for medically refractory VT associated with ARVD.

Case Reports

Patient 1

Patient 1 was a 35-year-old man who first experienced VT in January 1980. His father, paternal uncle and a brother died suddenly at ages 36, 42 and 33 years, respectively. Tachycardia was associated with chest pain and lightheadedness. Over the next 18 months, he was hospitalized 10 times with sustained VT and required cardioversion three times. During this time, he was treated with propranolol, disopyramide, procainamide, quinidine and amiodarone, alone and in combination. At the time of referral in September 1981, he had been taking amiodarone, 400–600 mg/day, for 5 months and was in virtually incessant VT. During interludes of sinus rhythm, the ECG showed left-axis deviation and Q waves in the anterosepal leads.

The physical examination was unremarkable. The chest radiograph showed a cardiothoracic ratio of 0.53. A two-dimensional echocardiogram revealed enlargement of the right ventricle, most marked in the outflow region. Radionuclide wall motion studies showed that the right ventricular ejection fraction was 0.36 and the left ventricular ejection fraction was 0.40. At cardiac catheterization, the patient was in VT. He had a mean right atrial pressure of 8 mm Hg, a right ventricular pressure of 27/9 mm Hg, a mean pulmonary artery pressure of 15 mm Hg and a left ventricular-end diastolic pressure of 12 mm Hg. The right ventricle was enlarged and contracted poorly, and there was aneurysmal dilatation in the outflow area. The left ventricle was slightly hypokinetic at the apex. Electrophysiologic study revealed two distinct mor-
phologies of spontaneous VT. Endocardial mapping showed earliest recorded activity for each morphology to be located in the anterior portion of the RVFW. Tachycardia was incessant and could only be terminated momentarily by bursts of ventricular pacing.

At operation, the heart was exposed through a median sternotomy. The right ventricle was enlarged with purplish discoloration and fatty streaks. There were aneurysmal, dyskinetic bulges over the infundibulum and over the basal portion of the inferior wall. The left ventricle appeared normal. Epicardial mapping was performed under normothermia in the absence of cardiopulmonary bypass. During sinus rhythm, low-amplitude, fractionated electrograms were recorded diffusely over the RVFW. The epicardial activation sequence of the RVFW in sinus rhythm was abnormal, showing slowing of conduction over anterior right ventricle. One morphology of VT showed earliest epicardial activation in the right ventricular outflow region and the second over the anterolateral wall of the right ventricle.

RVFW disconnection was then carried out under normothermic cardiopulmonary bypass (rectal temperature greater than 36.5°C) (figs. 1 and 2). The heart was kept beating and left and right ventricular electrograms were continuously monitored. A full-thickness ventriculotomy was performed along the superior, anterior and inferior attachment of the RVFW to the aortoventricular unit. All the trabeculae were cut. The tricuspid valve apparatus was carefully inspected: the inferior papillary muscle inserted into the septal area of the aortoventricular unit and the anterior papillary muscle inserted into the RVFW. A Y-shaped moderator band bridged the insertion of the anterior and inferior papillary muscle. The anterior papillary muscle and the common limb of the Y-shaped band were cut. The attachment of the posterolateral wall situated between the pulmonary artery and the tricuspid ostia was cut through an endocardial approach. The wall at that site was very thick, and total disconnection was ensured by a deep dissection of the right aortic sinus, sparing the right coronary artery. The attachment of the RVFW along the tricuspid annulus was then cut to complete the disconnection. Alternatively, the RVFW can be left attached to the tricuspid annulus and this final step can be achieved by dissection or cryoablation of all “interventricular” fibers where the tricuspid annulus joins the membranous septum.

The RVFW and the left ventricle contracted independently only after the section was completed by a dissection of the atroventricular sulcus to sever all connecting fibres. The appearance and contractility of the RVFW did not change after RVFW disconnection. Various unstable rhythms confined to the RVFW were observed, but no comprehensive mapping could be done. The anterior papillary muscle was reimplanted at its previously marked insertion over the RVFW by supporting Teflon pledgets. The RVFW was resutured and the right ventricular chamber was closed with a 4-0

**Figure 1.** Method of right ventricular free wall disconnection. (A) The curved arrows delineate the incision along the attachment of the right ventricular free wall. (B) The right ventricular free wall disconnection is completed. The anterior papillary muscle is disinserted.
monofilament running suture. Cardiopulmonary bypass was stopped uneventfully without the need for inotropic support. The surface ECG showed normal sinus rhythm. VT was recorded over the right ventricle, but was dissociated from normal sinus rhythm activating the left ventricle.

The postoperative course was uneventful. No attacks of VT were observed over a follow-up period of 4 months. The postoperative ECG showed sinus rhythm. The QRS morphology was slightly altered in comparison to the preoperative record, showing increased prominence of QS waves in the inferior leads. Intrinsic electrical activity of the RVFW could not be recorded and the RVFW could not be paced. Postoperatively, the echocardiogram demonstrated right ventricular enlargement with tricuspid leaflet prolapse. Prominent paradoxical septal motion, previously absent, was recorded. A radionuclide study demonstrated a right ventricular ejection fraction of 0.24 and left ventricular ejection fraction of 0.46. The right ventricular ejection fraction rose to 0.45 during exercise, and the left ventricular ejection fraction remained at 0.49. At right-heart catheterization, the right ventricle was enlarged and akinetic. The mean right atrial pressure was 10 mm Hg (previously 8 mm Hg). The right ventricular pressure was 32/20 mm Hg with a very prominent “A” wave (20 mm Hg). The mean pulmonary artery pressure was 16 mm Hg. During these studies, no active contraction of the RVFW was ever seen.

Patient 2

Patient 2 was a 32-year-old man who was well until October 1980, when he had a syncopal spell and was found to be in VT. His father and paternal uncle died suddenly at ages 26 and 39 years, respectively. Three paternal cousins died suddenly before the age of 30 years and another cousin had recurrent VT. After the initial episode, the patient was hospitalized at least six times...

**Figure 2.** Operative views of the heart in patient 1. (A) The right ventricle is dilated and has the typical appearance of right ventricular dysplasia. (B) The encircling ventriculotomy is completed. The Y-shaped band is seen, and its free wall insertion is about to be cut off.
times for recurrent VT and required cardioversion three times. Quinidine, propranolol, procainamide and amiodarone, alone and in combination, were ineffective. The patient had been taking amiodarone, 400–800 mg/day, for 9 months before admission. At the time of referral in September 1981, his medications included amiodarone, 600 mg/day, and procainamide, 4 g/day. Between episodes of tachycardia, the patient was well and had no chest pain, edema or orthopnea.

Physical examination revealed no evidence of cardiac disease. The ECG showed poor R-wave progression during normal sinus rhythm (fig. 3) and left bundle branch block morphology during VT. The chest radiograph showed borderline cardiomegaly (cardiothoracic ratio 0.52). The two-dimensional echocardiogram showed diffuse enlargement and hypokinesia of the right ventricle. The radionuclide angiogram showed a large right ventricle with an ejection fraction of 0.24 at rest and 0.40 during exercise. The left ventricular ejection fraction was 0.56 at rest and 0.54 during exercise.

At cardiac catheterization, the mean right atrial pressure was 2 mm Hg, the right ventricular pressure was 24/2 mm Hg, the pulmonary artery pressure was 22/5 mm Hg and the left ventricular end-diastolic pressure was 5 mm Hg (fig. 4). The right ventricle was diffusely enlarged and hypokinetic, and very slow washout and marked fissuring of contrast were evident. The left ventricle and coronary arteries were normal.

At electrophysiologic study, endocardial mapping of the right and left ventricle did not demonstrate delayed or fractionated electrograms. Five distinct morphologies of VT could be induced by the ventricular extrastimulus technique (fig. 5). The clinically observed VT was most frequently induced and endocardial mapping showed the site of earliest recorded electrical activity to be the infundibulum of the right ventricle. The other “nonclinical” tachycardias could not be mapped extensively. However, in all instances, right ventricular data points were considerably earlier than left ventricular data points.

At surgery, the right ventricle was diffusely enlarged and hypokinetic, with bulges over the infundibulum, the apex and the basal part of the inferior wall. The right ventricular epicardium had a purplish discol-

**Figure 3.** (A) Preoperative and (B) postoperative (4-week) ECGs of patient 2.
The patient's postoperative course was unremarkable at 3-month follow-up. Left pleural effusion occurred and was treated transiently with furosemide. The QRS morphology of the surface ECG was not significantly different from the preoperative tracing (fig. 3). For 3 weeks postoperatively, a pseudo-atrial-flutter pattern was observed on the ECG over the precordial leads and was demonstrated to be right ventricular tachycardia (cycle length 235 msec) dissociated from sinus rhythm activating the remainder of the heart (fig. 8). Two-dimensional echocardiography demonstrated diffuse enlargement and akinesia of the RVFW. The left ventricle contracted well, and there was prominent paradoxical motion of the septum toward the right ventricle (fig. 9). At cardiac catheterization, the right ventricular pressure was 27/6 mm Hg, the "a" wave was 20 mm Hg and the pulmonary artery pressure was 28/14 mm Hg (fig. 4). Radionuclide studies demonstrated a right ventricular ejection fraction of 0.22 at rest and 0.20 during exercise. The left ventricular...
Discussion

Our patients had predominantly right-sided cardiomyopathy associated with VT refractory to medical management. They had the typical clinical and radiographic features of ARVD.7

We believed that a new procedure aimed at confining arrhythmic activity in the right ventricle — RVFW disconnection — offered the only possibility for definitive treatment of recurrent VT. The multiple sites of origin of tachycardia associated with diffusely abnormal right ventricular myocardium precluded localized surgical ablation. The profound disability of our patients and the striking family history of sudden death underscored the need for definitive therapy. RVFW disconnection excludes the entire RVFW but leaves intact the septum, which belongs anatomically and functionally to the left ventricle. Our previous experience suggests that the arrhythmogenic areas are located over the RVFW.1-3,7 Consequently, this new procedure is indicated when the left ventricle, including the septum, is proved "nonarrhythmogenic."

Hemodynamic Feasibility of Right Ventricular Disconnection

The right ventricular chamber consists of the attachment of the RVFW to the aortoventricular unit. The septal wall delineated by the attachments of the RVFW may be considered a part of the aortoventricular unit.4 The RVFW and the aortoventricular unit differ in volume, thickness and curvature. The contraction of each is determined by separate branching of the His bundle. Different functional anatomy suggests a different role for each. The role of the RVFW has long been questioned. In 1943, Starr et al.5 demonstrated that the central venous pressure did not increase significantly in the anesthetized dog after severe undermining damage of the RVFW by thermal injury. In 1949, Rodbard and Wagner9 performed the first partial right ventricular bypass in the dog. In 1958, Robicsek and colleagues10 successfully bypassed the right ventricle in the dog with good long-term results. In 1968, Glenn et al.11 performed the first successful partial right ventricular bypass in man. The first total right ventricular bypass was performed by Fontan et al.12 in 1971. His patients had increased central venous pressure (15 mm Hg) postoperatively.

Ebstein’s anomaly constitutes another model of chronic RVFW dysfunction. The review of 65 cases of Ebstein’s anomaly by Bialostozky et al.13 strongly suggests that Ebstein’s patients without associated lesions and without arrhythmia have a normal life expectancy. Right ventricular failure was uncommon in this series, occurring only in association with severe dysrhythmia or high pulmonary vascular resistance.

The results of right ventricular disconnection in our

![Figure 7](image_url) Right ventricular free wall epicardial map during clinical ventricular tachycardia in patient 2. Earliest activity is recorded over the posterolateral wall of the infundibulum. Abbreviations are as in figure 6.

![Figure 8](image_url) Surface ECG in patient 2 recorded 2 weeks postoperatively. Right ventricular tachycardia is evident. The flutter-like activity in leads V1 and V2 corresponds to right ventricular tachycardia. RVFW eg -- bipolar right ventricular free wall electrogram.
patients verified the hemodynamic feasibility of this procedure. Left ventricular function was not affected and both patients had good exercise tolerance postoperatively. Neither patient had sinus tachycardia at rest. Possible compensatory mechanisms maintaining right ventricular function included an increase in right ventricular preload with a prominent atrial contraction and systolic (“paradoxical”) movement of the septum toward the RVFW postoperatively, associated with peak systolic pressure in the right ventricle. This latter finding suggested that the aortoventricular unit plays a definite role in right ventricular function.15 There are other important considerations. First, the integrity of the tricuspid and pulmonary valves was maintained. Second, pulmonary vascular resistance and left ventricular filling pressures were normal. Finally, right ventricular contractility was already grossly impaired in both patients preoperatively, possibly allowing for some degree of chronic compensation.

The Concept of Exclusion

The concept of exclusion was described in 1975 as a rationale for the surgical management of infarct scar responsible for ventricular arrhythmia. A deep endocardial ventriculotomy encircled the visible margin of the infarction or border zone.15 This ventriculotomy was aimed at containing abnormal arrhythmic electrical activity within the confines of the ventriculotomy scar and thereby preventing ventricular tachycardia. Evidence supporting the rationale for this procedure was provided by Ungerleider et al.,16 who showed impaired conduction from within the confines of the encircling endocardial ventriculotomy in experimental animals. Williams et al.17 applied the principle of exclusion at the atrial level to exclude the left atrial wall and Gallagher et al.18 applied the concept of exclusion at the right ventricular level to exclude an arrhythmogenic area over the acute margin of the right ventricle.

The concept of exclusion is readily applied to the surgical treatment of arrhythmogenic right ventricular dysplasia, a diffuse disease of the RVFW with multiple potential areas of arrhythmogenesis. Local ventriculotomy directed at specific arrhythmogenic areas has met with recurrence of VT of different morphology originating at right ventricular sites not dealt with at the initial operation.3 The confinement of electrical activity within the right ventricle by right ventricular disconnection provides a definitive cure of VT arising anywhere within the RVFW. The hemodynamic consequences of right ventricular tachycardia are absent if the remainder of the heart is dominated by sinus rhythm.

Atypical Features of Arrhythmogenic Right Ventricular Dysplasia

Both of our patients described a striking history of sudden death at an early age in male family members. We could not obtain autopsy records, but these people might have died from ventricular arrhythmia associated with ARVD. This possibility suggests that ARVD may be genetically transmitted and that family members of affected patients should be screened for this disorder.

Typically, the ECGs of patients with ARVD show nonspecific repolarization changes in the right precordial leads or notching at the beginning of the ST segment (“postexcitation”) in the same leads.7 The ECGs of our patients during normal sinus rhythm showed Q waves in the anterosetal and inferior leads preoperatively, which suggests the diagnosis of coronary artery disease. Interestingly, the QRS morphology did not change appreciably after RVFW disconnection.

We conclude that RVFW disconnection is hemody-
namically feasible if left-heart pressures are normal and if tricuspid valve integrity is maintained. RVFW disconnection offers definitive treatment of recurrent VT associated with diffuse disease of the RVFW.

References


Severe Pulmonary Hypertension Associated with a Small Ventricular Septal Defect

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SUMMARY A case of progressive pulmonary hypertension in a child with a small ventricular septal defect is presented. Natural history studies have indicated that children with small ventricular septal defects can be followed conservatively. This case represents a contradiction to that rule and suggests that further study must be directed toward defining the etiology of pulmonary hypertension in patients with congenital heart disease.

SMALL to medium-sized ventricular septal defects (VSDs) are usually associated with minimal risk of development of pulmonary vascular disease, particularly in early years.1-8 Most authorities have advocated nonsurgical, conservative management of children with pulmonary arterial systolic pressures less than 40% of systemic pressure, and pulmonary-to-systemic flow ratios (Qp:Qs) less than 2:1.1-8 However, in this report, we describe a child with progressive pulmonary vascular disease in association with a small VSD that underwent spontaneous closure.

Case Report

JF was born after an uncomplicated pregnancy and delivery in rural Louisiana. His mother was an 18-year-old, gravida 1, para 1, abortus 0, healthy white female. At 3 days of age, the child was discharged from the hospital with a normal physical examination. At 4 weeks of age, he was seen by a pediatrician, who noted a cardiac murmur. He was referred to Charity Hospital of New Orleans, where tachypnea with a respiratory rate of 65 beats/min was noted. The pulse rate on admission was 170 beats/min; systolic blood pressure 80 mm Hg; and height and weight at the tenth percentile. The examination revealed a mildly distressed, acyanotic infant with a hyperactive precordium. The S1 was normal and S2 was split with a loud P2. A grade III/IV holosystolic murmur was noted at the lower left sternal border. A mid-diastolic rumble was heard at the apex. The lungs were clear, and the liver edge was palpable 3 cm below the right costal margin. The child had mild peripheral edema and all pulses were easily palpable. A chest roentgenogram revealed moderate cardiomegaly and increased pulmonary vascularity. The ECG revealed biventricular hypertrophy and left atrial enlargement. The hematocrit was 40%. Therapy for congestive heart failure was initiated with digoxin and furosemide.

The infant responded well and underwent cardiac catheterization at 2 months of age. The study revealed right ventricular and pulmonary artery pressures of 60/5 and 60/20 mm Hg (mean 31 mm Hg), respectively. The left ventricular pressure was 85/5 mm Hg. The Qp:Qs ratio was 3.7:1. Angiographically, the patient had a large, subaortic VSD.

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