Total right ventricular exclusion procedure: An operation for isolated congestive right ventricular failure

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Objective: To prevent possible deleterious effects of right ventricular volume overload on cardiorespiratory function, we developed a total right ventricular exclusion procedure for the treatment of end-stage isolated congestive right ventricular failure.

Methods: Since 1996, this procedure has been performed in 5 patients in New York Heart Association functional class IV: 2 adults with arrhythmogenic right ventricular dysplasia and 3 children with Ebstein anomaly. The entire right ventricular free wall was resected along the atrioventricular groove and then parallel to the interventricular septum, sparing the pulmonary valve and a skeletonized right coronary artery. The orifice of the tricuspid valve was closed with either a polytetrafluoroethylene patch or with its leaflets. The defect of the right ventricular free wall was covered with a polytetrafluoroethylene patch in the 2 patients with arrhythmogenic right ventricular dysplasia and directly closed with the remnant of the free wall in the 3 children with Ebstein anomaly. After resection of a redundant right atrial wall, coronary sinus blood flow was rerouted into the left atrium through an atrial septal defect. A total cavopulmonary connection was constructed in 4 patients and a bidirectional superior cavopulmonary anastomosis in 1 infant. The heart was controlled with a DDD pacemaker in 3 patients.

Results: The patients were extubated at a mean of 14 hours postoperatively (range, 1–38 hours). There were no early or late deaths. At follow-up, ranging from 8 to 57 months, the mean cardiothoracic ratio had decreased from 74% $\pm$ 7% before the operation to 52% $\pm$ 6% ($P < .01$). All patients are in functional class I. Neither of the patients with arrhythmogenic right ventricular dysplasia have had attacks of ventricular tachycardia nor are they using antiarrhythmic medication.

Conclusions: The total right ventricular exclusion procedure provides effective decompression of the lung, as well as the left ventricle, and may result in more effective volume loading of a surgically created single ventricle with increased systemic output. We believe that this new surgical option offers rescue treatment for isolated end-stage right ventricular failure in critically ill patients.

Isolated right ventricular dysfunction caused by myocardial diseases or long-lasting volume overload gradually results in right ventricular and right atrial dilatation, leading to fatal cardiorespiratory failure. Arrhythmogenic right ventricular dysplasia is a specific myocardial disease of unknown cause characterized by fatty and fibrofatty replacement of the right ventricular myocardium associated with life-threatening ventricular tachycardia.^{1,2} The tachycardia is often refractory to medical treatment. Nonpharmacologic thera-

TABLE 1. Preoperative status and echocardiographic assessment in the 5 patients undergoing the total right ventricular exclusion procedure

	Patient no.				
	1	2	3	4	5
History of CPR	+	+	+	+	+
CTR	66	70	74	84	77
Mechanical ventilation	+	—	+	—	+
Inotropic support	+	—	+	+	+
Tricuspid regurgitation	Severe	Severe	Severe	Severe	Severe
Pulmonary regurgitation	None	Mild	Moderate	Severe	Moderate
Mitral regurgitation	Moderate	None	None	Moderate	None
Leftward shift of IAS	+	+	+	+	+
Paradoxical motion of IVS	+	+	+	+	+

CPR, Cardiopulmonary resuscitation; CTR, cardiothoracic ratio; IAS, interatrial septum; IVS, interventricular septum.

pies, such as catheter ablation,³ implantation of an automatic implantable cardioverter-defibrillator,⁴ and surgical intervention to interrupt the re-entrant circuit,^{5,6} have been used. Because any area of the right ventricular myocardium is potentially arrhythmogenic, surgical isolation of the right ventricular free wall has been an effective treatment of multifocal or polymorphic ventricular tachycardia.⁷⁻⁹ However, only cardiac transplantation has been life saving for patients with severe right ventricular dysfunction caused by the frequent occurrence of ventricular tachycardia.^{10,11}

We developed a total right ventricular exclusion procedure composed of resection of the entire right ventricular free wall and subsequent total cavopulmonary connection. This procedure was first performed in 1996 as rescue surgery for a moribund patient with arrhythmogenic right ventricular dysplasia. Indications for this procedure have extended to pediatric patients with an extremely enlarged nonfunctioning right ventricle associated with Ebstein anomaly of the tricuspid valve. In this article we describe the surgical techniques and early- to medium-term results of the total right ventricular exclusion procedure in 5 patients.

Patients and Methods

Between June 1996 and July 2000, the total right ventricular exclusion procedure was performed in 2 adults with arrhythmogenic right ventricular dysplasia and 3 children with Ebstein anomaly. Preoperative clinical data on the 5 patients are summarized in Table 1.

Case Presentations

Patient 1. A 27-year-old man first experienced syncope and ventricular tachycardia in December 1995 and was admitted to a local hospital, where he was given a diagnosis of arrhythmogenic right ventricular dysplasia on the basis of electrophysiologic and histologic studies. Because of failed catheter ablation, he was transferred to Okayama University Hospital for possible antiarrhythmic therapy on June 12, 1996. The electrocardiogram documented 5 distinct morphologies of ventricular tachycardia (rate, 110–120 beats/min) mixed with atrial tachycardia. Cardiac cath-

eterization demonstrated a mean pulmonary artery pressure of 26 mm Hg and a pulmonary capillary wedge pressure of 23 mm Hg. Endocardial mapping confirmed the earliest recorded activity for each morphology to be located in the right ventricular free wall. Over the next 2 weeks, cardiorespiratory failure developed as a result of recurrent ventricular tachycardia, and the patient subsequently required mechanical ventilation and intra-aortic balloon pumping. On June 24, however, systolic blood pressure dropped to less than 90 mm Hg, with right atrial pressure of 23 mm Hg, and the cardiac index progressively decreased to $1.3 \text{ L} \cdot \text{min}^{-1} \cdot \text{m}^{-2}$; thus extracorporeal membrane oxygenation was established by means of femoral cannulation. Despite volume unloading on the right ventricle with extracorporeal membrane oxygenation, ventricular tachycardia and occasional ventricular fibrillation became refractory to cardioversion. On June 26, 1996, after 8 hours of sustained ventricular tachycardia, he was transferred to the operating room with maximal life-support systems and underwent the total right ventricular exclusion procedure.

Patient 2. A 44-year-old woman who underwent closure of an atrial septal defect at 8 years of age and tricuspid valve repair at 36 years of age first experienced ventricular tachycardia in 1991. After the initial episode, she was hospitalized 5 times for recurrent ventricular tachycardia and required cardioversion at least 6 times. At the time of referral in September 1997, she was cachectic as a result of protein-losing enteropathy and liver dysfunction. Cardiac catheterization showed a mean pulmonary artery pressure of 14 mm Hg. Echocardiography demonstrated an extremely dilated right ventricle with markedly diminished contractility, massive tricuspid regurgitation, and normal left ventricular function. Electrophysiologic study revealed a focus of ventricular tachycardia in the right ventricular free wall. Results of histologic examination led to the suspicion of arrhythmogenic right ventricular dysplasia. All possible surgical treatments, including redo tricuspid valve repair or replacement, implantation of a cardioverter-defibrillator, right ventricular isolation, and exclusion procedures, were proposed to the patient and her family. In consideration of ventricular tachycardia arising from the right ventricular myocardium, poor right ventricular function, and severe tricuspid regurgitation, we elected to perform the total right ventricular exclusion procedure, which was carried out on January 21, 1998.

Figure 1. The right ventricular free wall was resected along the atrioventricular groove and then parallel to the interventricular septum, sparing the pulmonary valve and a skeletonized right coronary artery. The dotted lines show incisions for resection of the right ventricular free wall and the redundant right atrial wall.

Patient 3. A 14-month-old girl who had a factor VII coagulation defect was admitted to Okayama University Hospital on September 9, 1998, for surgical treatment of Ebstein anomaly. On admission, she had deep cyanosis, with an arterial oxygen tension of 30 mm Hg. Echocardiography revealed an atrial septal defect with a diameter of 7 mm, Ebstein anomaly with severe tricuspid regurgitation, and moderate pulmonary regurgitation. A large atrialized right ventricle resulting from a 33-mm downward displacement of a dysplastic septal leaflet had a paper-thin noncontracting wall, whereas the small functional right ventricle had a systolic wall thickness of 2.4 mm. She had a cerebral hemorrhage on September 11 and cardiac arrest on September 23. Because of recurrent cardiopulmonary collapse, she underwent an emergency operation on September 29, 1998.

Patient 4. A 5-year-old boy who was born with Ebstein anomaly and critical pulmonary stenosis underwent pulmonary valvotomy at 7 days of age and tricuspid valve repair and right ventricular outflow reconstruction with a monocusp-valved onlay patch at 1 year of age. Eight months later, he required a third operation, including tricuspid valve replacement and pacemaker implantation. At 5 years of age, he had severe congestive right ventricular failure, with the highest brain natriuretic peptide value being 1819 pg/mL, which was treated with continuous infusion of dobutamine and phosphodiesterase inhibitor (olprinone). Echocardiography showed a dilated and poorly contracting right ventricle, a huge right atrium, severe tricuspid regurgitation with back flow reaching the hepatic vein, and severe pulmonary regurgitation. After 3 months of bed rest, he underwent a fourth operation on March 15, 2000.

Patient 5. A 5-month-old boy, weighing 5.5 kg, had dyspnea and severe cyanosis on July 13, 2000. On admission to the hospital, systemic acidosis was present with a pH of 6.9. Mechanical ventilation and inotropic support were required to stabilize his cardiorespiratory condition. Echocardiography showed Ebstein anomaly with severe tricuspid regurgitation, a restrictive but patent foramen ovale, and moderate pulmonary regurgitation. The septal leaflet was greatly displaced into the right ventricular outflow tract at a distance of 3.8 mm from the pulmonary valve. Because the greater part of the right ventricular free wall was thin and atrialized, and the anterior leaflet was also displaced downward from the atrioventricular junction, we performed a univentricular palliation that incorporated total resection of the right ventricular free wall and bidirectional superior cavopulmonary anastomosis on July 17, 2000.

Operative Procedure

Arrhythmogenic right ventricular dysplasia. Through a mid-line sternotomy, hypothermic cardiopulmonary bypass (34°C) was commenced with bicaval and aortic cannulation. The right atrium was opened to drain coronary sinus flow into the bypass circuit. Keeping the heart beating, the entire right ventricular free wall, including the tricuspid valve apparatus, was excised as follows. A longitudinal right ventriculotomy was made in the anterior right ventricle parallel to the atrioventricular groove and then around the tricuspid annulus, sparing the skeletonized right coronary artery and its posterior interventricular branches (Figure 1). This incision was extended across the right ventricular outflow tract (leaving the pulmonary valve intact), down the anterior surface parallel to the interventricular septum, around the right ventricular apex onto the posterior wall, and to the level of the tricuspid annulus just to the right of the crux of the heart. Special care was taken to avoid injury to the left anterior descending coronary artery. Any hemorrhage from branches of the right coronary artery was effectively controlled with electrocautery. During the ventriculotomy, the sustained ventricular tachycardia abruptly terminated in patient 1. After complete removal of the right ventricular free wall from the heart, the papillary muscles of the tricuspid valve and the septoparietal trabeculations were resected from the interventricular septum and the right ventricular apex.

The tricuspid valve orifice was closed with a 1-mm-thick polytetrafluoroethylene (PTFE) patch secured with continuous 4–0 polypropylene sutures. The membranous interventricular septum was left above the patch to avoid heart block (Figure 2). Several cryolesions were placed along the ventriculotomy for latent foci of ventricular tachycardia. The right atrial free wall was almost totally resected to prevent potential supraventricular tachyarrhythmia,¹ leaving a small triangular flap of the wall attached to the atrioventricular groove. A 24-mm-diameter PTFE tube for a cavopulmonary connection was anastomosed to the inferior vena caval orifice. At this stage, the aorta was crossclamped, and cold blood cardioplegic solution was administered in patient 2, whereas the whole procedure was completed with the heart beating in patient 1. An atrial septal defect was created, and the coronary sinus was roofed with the right atrial flap to reroute blood flow into the left atrium (Figure 3). The superior vena cava was transected and anastomosed to the upper surface of the right pulmonary artery. The aorta was declamped, and rewarming commenced. An

Figure 2. The tricuspid valve orifice was closed with a 1-mm-thick PTFE patch secured with continuous 4-0 polypropylene sutures. The membranous interventricular septum was left above the patch to avoid heart block. **ASD**, Atrial septal defect; **CS**, coronary sinus; **MS**, membranous septum.

extracardiac total cavopulmonary connection was completed by anastomosing the distal end of the PTFE tube to the lower surface of the right pulmonary artery. A small communication between systemic and pulmonary venous returns was created with a 6-mm PTFE graft to preserve the ventricular preload while limiting an eventual rise in central venous pressure. Finally, the defect of the right ventricular free wall was covered with a small 1-mm-thick PTFE patch secured with continuous 4-0 polypropylene sutures (Figure 4). Unipolar epicardial pacing leads were placed on the left atrium and the left ventricle so that the heart could be controlled by a DDD pacemaker.

Ebstein anomaly. Three children underwent hypothermic cardiopulmonary bypass with bicaval and aortic cannulation. In patient 3, with an interatrial venous communication, the aorta was cross-clamped, and crystalloid cardioplegia was infused. Heart beat was maintained in patients 4 and 5, with an intact atrial septum. After complete unloading of the right ventricle, the thin redundant right ventricular free wall was resected by incisions made 1 to 2 cm away from its attachment to the interventricular septum in these 3 patients. The pulmonary valve was closed in patients 3 and 4 and repaired in patient 5. The tricuspid valve was partially excised, and its orifice was closed with a PTFE patch in patients 3 and 4, as described for patients with arrhythmogenic right ventricular dysplasia, and a 4-mm atrioventricular communication was created in the PTFE patch to drain blood through the Thebesian vein from a remnant right ventricle into the atrium. In patient 5 the tricuspid orifice was closed by suturing the anterior leaflet to the tricuspid

Figure 3. An atrial septal defect was created, and the coronary sinus was roofed with the right atrial flap to reroute blood flow into the left atrium. The *dotted line* indicates the suture line of the atrial flap to the interatrial septum.

anulus or to the displaced septal leaflet, thereby placing the membranous interventricular septum on the right atrial side. The defect of the right ventricle was directly closed with the remnant of the free wall by using a continuous polypropylene suture (Figure 5). Patients 4 and 5 underwent cardioplegic arrest at this point. The interatrial septum was resected, the redundant right atrial free wall was removed, and the atriotomy was closed. After aortic declamping, a total cavopulmonary connection was constructed with an inferior vena caval channel by using the right atrial wall¹² in patient 3 and a 20-mm-diameter PTFE tube in patient 4. Bidirectional superior cavopulmonary anastomosis was performed in patient 5.

Results

Operative results are presented in Table 2. The patients were extubated between the 1st and 38th postoperative hour (mean, 14 hours), had a slow but uneventful recovery, and were discharged from the 25th to 76th postoperative day. There were no in-hospital or late deaths. At follow-up, ranging from 8 to 57 months (mean, 24 months), the mean cardiothoracic ratio decreased from the $74\% \pm 7\%$ preoperative value to $52\% \pm 6\%$ ($P < .01$, Mann-Whitney test; Figure 6). Most recent echocardiography revealed no paradoxical motion of the interventricular septum and a left ventricular ejection fraction of greater than 60% in all patients.

Neither patient with arrhythmogenic right ventricular dysplasia has had an attack of ventricular tachycardia nor is receiving antiarrhythmic medication. At follow-up catheter-

Figure 4. In patients with arrhythmogenic right ventricular dysplasia, a fenestrated extracardiac total cavopulmonary connection was constructed. The defect of the right ventricular free wall was covered with a small 1-mm-thick PTFE patch.

ization, central venous pressures were less than 15 mm Hg. The extracardiac cavopulmonary pathway and the skeletonized right coronary artery are demonstrated in Figure 7. Left ventricular function in patient 4 improved sequentially over the year after total right ventricular exclusion; ejection fraction evaluated with echocardiography was 38% at 1 month after the operation, 55% at 6 months, and 62% at 1 year. All 5 patients are currently in New York Heart Association functional class I; patient 1 is employed in a full-time job, patient 2 is a housewife, patient 3 is doing well without neurologic deficits, patient 4 is fully active and goes to kindergarten, and patient 5 is awaiting the Fontan procedure.

Discussion

Surgical treatment of arrhythmogenic right ventricular dysplasia has focused on the management of associated life-threatening ventricular tachycardia. In 1983, Guiraudon and associates⁷ introduced an operation for complete isolation of multiple origins of ventricular tachycardia by disconnecting the entire right ventricular free wall. This procedure theoretically confines any arrhythmic activity arising from the diseased myocardium in the isolated right ventricular wall. Despite encouraging results, Cox and colleagues⁸ and Nimkhedkar and coworkers⁹ warned against the routine use

Figure 5. In children with Ebstein anomaly, the defect of the right ventricle was directly closed with the remnant of the free wall.

of the isolation procedure because of the high incidence of right ventricular failure. Our 2 patients with arrhythmogenic right ventricular dysplasia had intractable ventricular tachycardia, a markedly dilated right ventricle, and severe tricuspid regurgitation. The resultant congestive right ventricular failure and the presence of tricuspid regurgitation precluded the indication for the isolation procedure. The only life-saving option for these patients appeared to be cardiac transplantation. We devised the operation as rescue surgery for a moribund patient on extracorporeal membrane oxygenation (patient 1). By resecting the diseased myocardium, creating a single left ventricle, and constructing a cavopulmonary connection, the total right ventricular exclusion procedure provided a definitive cure of end-stage right ventricular failure and refractory ventricular tachycardia associated with arrhythmogenic right ventricular dysplasia. This new surgical option should be indicated when the left ventricle, including the interventricular septum, is proved nonarrhythmogenic¹³ and functionally normal.

Ebstein anomaly is a spectrum of disease characterized by varying degrees of downward displacement of the septal and posterior leaflets of the tricuspid valve into the right ventricular cavity and marked dilatation of the anulus, resulting in tricuspid valve incompetence.^{14,15} The right ventricular myocardium is generally abnormal, with a decrease in the number of myocardial fibers.¹⁶ The results of ana-

Figure 6. Chest roentgenograms of patient 1 with arrhythmogenic right ventricular dysplasia (top) and patient 3 with Ebstein anomaly (bottom) before the operation (left) and at the last follow-up (right).

TABLE 2. Operative results of the 5 patients undergoing the total right ventricular exclusion procedure

	Patient no.				
	1	2	3	4	5
CPB time (min)	198	234	144	190	102
Aortic clamp time (min)	0	21	82	16	27
Intubation time (h)	19	1	38	2	11
Postoperative hospital stay (d)	76	72	25	37	34
Heart rhythm	Pacing	Pacing	Sinus	Sinus	Sinus
Complications	None	Pleural effusion	None	Pleural effusion	None

CPB, Cardiopulmonary bypass.

tomic correction of this anomaly have improved in the last decades since tricuspid valve repair combined with ventriculoplasty by plication of the atrialized right ventricle^{17,18} has been performed in preference to valve replacement. Recent reports^{19,20} demonstrated beneficial effects of a bi-directional cavopulmonary connection in association with intracardiac repair on operative outcome and postoperative tricuspid valve function in patients with poor ventricular function.

Management of severe forms of Ebstein anomaly has remained a surgical challenge, particularly in symptomatic

children presenting with congestive right heart failure and deep cyanosis. In 1991, Starnes and coworkers²¹ described an innovative univentricular palliation by creating a functional tricuspid atresia for critically ill neonates with this anomaly. Although the surgical concept of the right ventricular exclusion procedure described in this report is the same as that of Starnes' operation, our technique differs in some important ways. First, a large part of the right ventricular free wall is resected. Resultant volume reduction of the right ventricle promises normal interventricular motion, as evidenced in our patients. Second, the coronary sinus is

Figure 7. Angiograms 2 years after the total right ventricular exclusion procedure in patient 2 with arrhythmogenic right ventricular dysplasia, showing an inferior vena cava–pulmonary artery pathway with a patent fenestration (left) and skeletonized right coronary artery and intact posterior interventricular branches (right).

included in the atrium, whereas with the Starnes' technique, it is left on the ventricular side, thus requiring a functioning pulmonary valve. Finally, the tricuspid valve orifice is closed, leaving the membranous interventricular septum above the patch to reduce the risk of heart block. No definite statement can be made as yet concerning an indication for univentricular repair in this anomaly. Because repair of the tricuspid valve is the key to successful biventricular or one-and-a-half ventricular repair of Ebstein anomaly, particularly in the presence of pulmonary regurgitation, determination of operative procedure should be based on the exact assessment of right ventricular function and the surgeon's skill in repairing the tricuspid valve.

Hemodynamic improvement in our patients is attributable to radical removal of deleterious effects of right ventricular congestion and associated tricuspid regurgitation on systemic venous return and left ventricular function. Although we did not exactly evaluate preoperative left ventricular function, echocardiography revealed a distortion of the left ventricular geometry caused by paradoxical motion of the interventricular septum in all patients. Previous clinical studies^{22,23} in patients with isolated right ventricular

volume overload indicated that late diastolic leftward displacement of the interventricular septum may impair distensibility and contractility of the left ventricle. In the setting of tricuspid regurgitation, compression of the left atrium by a distended right atrium and leftward bulging of the interatrial septum during right ventricular systole may decrease left atrial stroke volume and further reduce left ventricular filling.²⁴ By means of complete decompression on the left heart, the total right ventricular exclusion procedure normalizes interventricular septal motion, increases left atrial volume, and subsequently achieves effective volume loading of the left ventricle.

Other beneficial effects of the right ventricular exclusion procedure are to relieve lung compression by the enlarged heart, as well as to make more space for the lung to expand. Despite long-lasting lung compression, all but one patient could be weaned from the ventilator within 24 hours after the operation. Adequate lung expansion may improve respiratory function and play an important role in successful univentricular circulation by reduction in pulmonary vascular resistance.

In conclusion, the total right ventricular exclusion pro-

cedure is hemodynamically feasible if left ventricular function is well preserved. This procedure provides a definitive cure of ventricular tachycardia associated with arrhythmogenic right ventricular dysplasia, when the left ventricle is proved nonarrhythmogenic. Removal of a dilated right ventricle and downsizing the right atrium leads to effective decompression of the lung, as well as the left ventricle, and may result in more effective volume loading of a surgically created single ventricle. We believe this new surgical option offers rescue treatment for isolated congestive right ventricular failure in critically ill patients.

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