INTRAVENTRICULAR REPAIR OF DOUBLE-OUTLET RIGHT VENTRICLE WITH NONCOMMITTED VENTRICULAR SEPTAL DEFECT: ADVANTAGES OF MULTIPLE PATCHES

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Objective: The objective of this paper is to report our experience with biventricular repair of double-outlet right ventricle with noncommitted ventricular septal defect by means of multiple patches that simplify and render feasible the intraventricular correction of this complex anomaly. **Methods:** From April 1987 to April 1999, in 18 patients with double-outlet right ventricle and noncommitted ventricular septal defect, a technical modification that used multiple patches of bovine pericardium was used to construct an intraventricular tunnel connecting the left ventricle to the aorta. Ages ranged from 2 months to 13 years (mean age 4.73 ± 3.41 years). **Results:** The early mortality was of 11.1% (2 patients). Surviving patients were followed up for a mean of 2.65 years. Three late deaths (16.6%) occurred: 5 months, 7 months, and 7 months after the operation. All but 1 patient are in New York Heart Association class I. **Conclusion:** The use of multiple patches for biventricular correction of this anomaly simplifies and renders feasible the intraventricular repair in cases in which the 1-patch technique was deemed impossible. (J Thorac Cardiovasc Surg 1999;118:1056-67)

Biventricular repair of double-outlet right ventricle (DORV) with noncommitted ventricular septal defect (VSD) remains a surgical challenge.^{1,2} Several reports have revealed that application of the intraventricular rerouting technique offers less than satisfactory immediate and late results.³⁻⁶ In this anomaly, the VSD is usually located within the right ventricular inlet portion, which makes it difficult to construct an unobstructed tunnel from the left ventricle to the aorta. Therefore Fontan-type procedures have been used as an alternative approach.⁷

The purpose of this report is to present our operative and late results with the biventricular repair of DORV with noncommitted VSD, emphasizing our technical modification of using multiple patches to reroute the left ventricle to the aorta.

Patients and methods

Between April 1987 and April 1999, at the Heart Institute, 18 patients underwent biventricular repair for DORV with noncommitted VSD with the use of multiple patches. Situs solitus was present in 17 patients and situs inversus in 1 patient. Nine patients were boys and 9 girls. Ages at the operation ranged from 2 months to 13 years, with a mean age of 4.73 ± 3.41 years. Seven patients had a preliminary palliative procedure, 4 had a modified Blalock-Taussig shunt, and 3 had pulmonary artery banding. In the same period, the intraventricular rerouting technique was deemed impossible in 4 patients, and they were excluded from subsequent analysis. In 3 of them, the VSD was more related to the pulmonary trunk; therefore a left ventricular-pulmonary arterial tunnel was constructed, and an atrial (2 patients) or arterial switch operation (1 patient) was thereupon performed. In the other patient, a Glenn anastomosis was preferred because of hypoplasia of the left ventricle.

The distance between the tricuspid and pulmonary valves was not considered a restriction to intraventricular rerouting. In the presence of pulmonary stenosis, the pulmonary valve was closed, the left ventricular-aortic tunnel was constructed passing over the pulmonary orifice, and the right ventricular outflow tract was reconstructed with an extracardiac conduit. The other patients, without pulmonary obstruction, generally had right ventricular dilatation. Thus the distance from the tricuspid valve to the pulmonary trunk was larger, facilitating the intraventricular rerouting repair.

Clinical data were obtained by review of clinical records, operative reports, and preoperative 2-dimensional echocardi-

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Fig 1. **A**, Anterosuperior enlargement of the VSD to create a larger anterior and medial passageway. **B**, Bovine pericardial patch secured with interrupted 5-0 or 6-0 mattress sutures, reinforced with pledgets, to the margin of the VSD. **C**, The long axis of the first patch forms an almost 45° angle to an imaginary line drawn through the center of the aortic annulus. The second, more distal patch is secured around the aortic orifice and subaortic conus, with the long axis of the oval patch almost perpendicular to the long axis of the first patch. **D**, Final aspect of the intraventricular tunnel constructed with 3 bovine pericardial patches. The papillary muscles of the tricuspid valve are reinserted at the patch.

Table I. DORV with noncommitted VSD

<i>N (age)</i>	<i>Previous procedure</i>	<i>VSD</i>	<i>Subaortic conus</i>	<i>Pulmonary stenosis</i>	<i>Ao/PA</i>	<i>Coronary</i>
1 (1 y)	None	Inlet septum Restrictive	Present	Present	RSS	Normal
2 (6 y)	Pulmonary banding	Trabecular septum Restrictive	Present	Banding	LAAo	TGA type
3 (6 y)	None	Inlet septum Restrictive	Present	Present	LAAo	TGA type
4 (1 y)	None	Inlet septum Restrictive	Present	Present	RSS	Normal
5 (11 y)	Modified right BT shunt	Inlet septum Restrictive	Absent	Atresia	RAAo	Normal
6 (6 y)	Modified right BT shunt	Inlet septum Restrictive	Present	Present	RSS	Normal
7 (5 y)	None	Trabecular septum Restrictive	Present	Present	LAAo	Single coronary ostium
8 (2 mo)	None	Inlet septum	Present	Absent	RAAo	Normal

<i>Associated anomalies</i>	<i>Surgical repair</i>	<i>Postop findings</i>
None	Two patches RVOT enlargement with transannular patch	Post CPB: LV-Ao gradient = 5 mm Hg Systolic RV/LV pressure ratio = 0.48 Lost to follow-up 1 y postop
Juxtaposition of atrial appendages Persistent LSVC	Three patches Debanding of PA Resection of segment of PA with the band and end-to-end anastomosis	Post CPB: LV-Ao gradient = 4 mm Hg Systolic RV/LV pressure ratio = 0.57 Cath: LV-Ao gradient = 3 mm Hg, RV-PT gradient = 22 mm Hg 8 y of follow-up: FC I (NYHA) without medication Palpitations with exercise Echo: RV-PA gradient = 25 mm Hg; normal EF; LVOT without obstruction 24-h Holter: Isolated ventricular extrasystoles (74 episodes/day)
Single atrium Right isomerism	Three patches Excision of septal papillary muscles and reinsertion at the first patch Atrial septation RVOT reconstruction with 18-mm crimped bovine pericardial conduit	7 y of follow-up: FC I (NYHA) without medication Echo: Normal EF; conduit obstruction with RV-PA gradient = 57 mm Hg; LVOT without obstruction
Pulmonary stenosis	Two patches Infundibular pulmonary resection RVOT reconstruction with monocusp valve	Post CPB: LV-Ao gradient = 10 mm Hg 5 y of follow-up FC I (NYHA) without medication Echo: Normal EF; LVOT without obstruction; severe pulmonary insufficiency
None	Three patches RVOT reconstruction with 18-mm extracardiac conduit BT shunt occlusion	Reoperation for residual VSD on first postop day Echo: RV-PA gradient = 19 mm Hg Electric cardioversion for atrial flutter Right thoracentesis and pericardiocentesis 13 days after definitive repair Sudden death at home 40 days postop
Tricuspid and mitral straddling type A Anomalous RV muscle band Pulmonary stenosis	Two patches RVOT enlargement with transannular patch Resection of anomalous muscle band	Post CPB: LV-Ao gradient = 0 mm Hg Third-degree AV block; definitive pacemaker 1 mo later Bacterial endocarditis Reoperation for residual VSD 2 mo later Reoperation for patch endocarditis with reconstruction of intraventricular tunnel 4 mo after definitive repair Death 5 mo after operation for bacterial endocarditis
Situs inversus Dextrocardia Pulmonary stenosis	Three patches RVOT enlargement with transannular pericardial patch	Post CPB: LV-Ao gradient = 0 mm Hg Systolic RV/LV pressure ratio = 0.3 Echo: RV-PA gradient = 40 mm Hg Sudden death at home 7 mo postop
Aortic coarctation SAS PDA	Two patches Resection of SAS Resection of coarctation and end-to-end anastomosis PDA ligation	Post CPB: LV-Ao gradient = 17 mm Hg Hospital death on first postop day Low cardiac output, coagulopathy, and bleeding

Table I. *Continued*

<i>N (age)</i>	<i>Previous procedure</i>	<i>VSD</i>	<i>Subaortic conus</i>	<i>Pulmonary stenosis</i>	<i>Ao/PA</i>	<i>Coronary</i>
9 (13 y)	None	Trabecular septum	Present	Present	LAAo	TGA type
10 (7 y)	None	Inlet septum Restrictive	Present	Present	AP	TGA type
11 (7 y)	Pulmonary banding	Inlet septum Restrictive	Present	Banding	RSS	Normal
12 (5 y)	None	Inlet septum	Present	Present	RAAo	Normal
13 (2 y)	None	Inlet septum Restrictive	Present	Absent	RSS	Normal
14 (4 y)	Pulmonary banding	Trabecular septum Restrictive	Present	Banding	RSS	Normal
15	Modified right BT shunt	Inlet septum Restrictive	Present	Present	LAAo	TGA type
16 (2 y)	None	Inlet septum Restrictive	Present	Present	RAAo	Normal

<i>Associated anomalies</i>	<i>Surgical repair</i>	<i>Postop findings</i>
None	Three patches RVOT reconstruction with 18-mm extracardiac conduit	Echo: Severe AR, moderate MR, minimal residual VSD, LV-Ao gradient = 25 mm Hg 7th postop day Cath: LV-Ao gradient = 18 mm Hg, RV-PA gradient = 9 mm Hg 3 y 4 mo of follow-up FC I (NYHA) without medication Echo: Mild AI; normal EF; RVOT without obstruction
ASD	Three patches ASD closure Pulmonary infundibular resection	Post CPB: LV-Ao gradient = 4 mm Hg; systolic RV/LV pressure ratio = 0.68 10th postop day Cath: Residual VSD; LV-Ao gradient = 9 mm Hg 2 y 3 mo of follow-up FC I (NYHA) without medication Echo: Minimal residual VSD; normal EF; RVOT without obstruction
Straddling TV	Three patches Detachment of TV and reinsertion on the second patch Bicuspidization of the TV Debanding; resection and end-to-end anastomosis	Post CPB: LV-Ao gradient = 6 mm Hg Postop ventricular arrhythmias Sudden death at home 7 mo postop
None	Three patches Resection of septal cusp of TV Bicuspidization of TV	Post CPB: LV-Ao gradient = 10 mm Hg; systolic RV/LV pressure ratio = 0.52 2 y of follow-up FC I (NYHA) without medication Echo: Minimal residual VSD; normal EF; LVOT without obstruction
Supra-annular mitral ring Parachute mitral valve	Three patches Resection of supra-annular ring Division of single papillary muscle	Post CPB: LV-Ao gradient = 10 mm Hg Systolic RV/LV pressure ratio = 0.48 1 y 6 mo of follow-up FC II (NYHA) with digoxin and furosemide Echo: Moderate TV insufficiency; moderate ventricular dysfunction; moderate pulmonary hypertension
Subaortic stenosis	Three patches Resection of SAS Debanding; Resection and end-to-end anastomosis	Post CPB: LV-Ao gradient = 30 mm Hg 2 y of follow-up FC I (NYHA) without medication Echo: LV-Ao gradient = 0 mm Hg
Persistent LSV Juxtaposition of atrial appendages	Two patches RVOT reconstruction with 16-mm extracardiac conduit BT shunt closure	2 y 3 mo of follow-up FC I (NYHA) without medication Echo: Normal EF; no RVOT or LVOT obstruction
LPA stenosis	Two patches Detachment of TV Enlargement of septal cusp of TV with pericardial patch RVOT enlargement with 14-mm monocusp transannular patch Enlargement of LPA	Post CPB: LV-Ao gradient = 4 mm Hg 2 y 3 mo of follow-up FC I (NYHA) without medication Echo: Normal EF; no RVOT or LVOT obstruction; severe pulmonary regurgitation (monocuspid valve)

Table I. Continued

<i>N</i> (age)	Previous procedure	VSD	Subaortic conus	Pulmonary stenosis	Ao/PA	Coronary
17 (3 y)	Modified BT shunt	Inlet septum Restrictive	Present	Atresia	AP	Normal
18 (1.5 y)	None	Inlet septum Restrictive	Present	Absent	RSS	Normal

DORV, Double-outlet right ventricle; *VSD*, ventricular septal defect; *Ao*, aorta; *PA*, pulmonary artery; *RSS*, right side by side; *RVOT*, right ventricular outflow tract; *CPB*, cardiopulmonary bypass; *LV*, left ventricle; *RV*, right ventricle; *LAAo*, left anterior oblique; *TGA*, transposition of the great arteries; *LSVC*, left superior vena cava; *Cath*, catheterization; *PT*, pulmonary trunk; *FC*, functional class; *NYHA*, New York Heart Association; *Echo*, echocardiography; *EF*, ejection fraction; *LVOT*, left ventricular outflow tract; *BT*, Blalock-Taussig; *RAAo*, right anterior oblique; *AV*, atrioventricular; *SAS*, subaortic stenosis; *PDA*, patent ductus arteriosus; *AR*, aortic regurgitation; *MR*, mitral valve regurgitation; *AI*, aortic insufficiency; *IVR*, intraventricular rerouting; *CHF*, congestive heart failure; *ASD*, atrial septal defect.

graphic, angiographic, and nuclear magnetic resonance imaging studies.

Follow-up data were collected in all patients from their parents and from referring physicians. Electrocardiographic and echocardiographic studies were performed on all hospital survivors. The mean follow-up was 2.65 ± 3.23 years, ranging from 1 month to 12 years, with a median follow-up of 2.0 years. In 5 patients cardiac catheterization was also performed.

Anatomic features. DORV with noncommitted VSD was defined as a subgroup of DORV in which the VSD was distant from both the aortic and pulmonary valves and was located within the inlet septum without perimembranous extension (atrioventricular canal type defect) or within the trabecular intraventricular septum and was not related to the septal band or its division.⁸⁻¹⁰

The VSD was an atrioventricular canal type defect (or inlet septum defect) in 14 of the patients and a muscular defect in 4. The VSD was considered restrictive in 14 patients, and in 4 of them the diameter of the VSD was less than 8 mm. None of the VSDs was more closely committed to the pulmonary trunk or to the aorta (Table I).

The spatial relationship of the aorta with regard to the pulmonary trunk was as follows: right side-by-side aorta in 7 patients, left anterior oblique in 5, right anterior oblique in 4, and anteroposterior in 2.

The pulmonary outflow tract was obstructed in 10 patients. One patient had a juxtaductal left pulmonary artery stenosis. Two patients had pulmonary atresia.

A subaortic conus was present in all but 1 patient of this series and caused moderate obstruction of the left ventricular outflow tract in 2.

Coexistent cardiac anomalies were present in 9 patients:

aortic coarctation and persistent ductus arteriosus in 1, straddling tricuspid valve in 2, straddling mitral valve in 1, juxtaposition of the atrial appendages associated with a persistent left superior vena cava in 2 patients, and atrial septal defect in 1 patient. One patient also had both a supra-annular mitral ring and a parachute mitral valve, and 1 patient had right atrial isomerism.

Surgical technique. In 16 patients, intraventricular rerouting was performed through a combined right atrial and right ventricular approach. In 2 patients with juxtaposition of the atrial appendages and subsequent small right atrium, the intraventricular rerouting procedure was done exclusively through a right ventriculotomy.

All patients underwent continuous moderate hypothermic cardiopulmonary bypass. Myocardial protection was achieved by repeated infusions of crystalloid cardioplegic solution into the aortic root. Direct caval cannulation was performed so as to facilitate the right atriotomy. The intraventricular anatomic structures were examined through the right atrium. The position and size of the VSD and the tricuspid valve, its chordae, and its papillary muscles were carefully evaluated. The distance between the tricuspid and pulmonary valves and between the inlet VSD and the aortic valve were also assessed.

Once the anatomy was clearly identified, the next and very important step was to enlarge the VSD anterosuperiorly, even in nonrestrictive VSDs, to create a larger anterior and medial passageway (Fig 1, A).

The first bovine pericardial patch was tailored in a semilunar shape and secured to the margin of the VSD with interrupted 5-0 or 6-0 mattress sutures reinforced with pledgets (Fig 1, B). The long axis of the first patch forms an almost

<i>Associated anomalies</i>	<i>Surgical repair</i>	<i>Postop findings</i>
Postop LPA stenosis after BT shunt Pulmonary atresia	Three patches for IVR Reconstruction of LPA RV-PA extracardiac conduit BT shunt ligation	Sepsis (sinusitis) CHF—severe mitral valve incompetence Reoperation after 30 days for mitral valvuloplasty Reoperation after 40 days for left diaphragmatic plication 1 y 6 mo of follow-up FC I (NYHA) Echo: Normal EF; no RVOT or LVOT obstruction
PDA	Two patches Ligation of PDA RV-PA extracardiac conduit	Post CPB: Systolic RV/LV pressure ratio = 0.54 Immediate postop cardiac arrest due to endotracheal tube obstruction; partial convulsive crisis; bilateral ischemic temporo-parieto-occipital lesion 1 y of follow-up

45° angle to an imaginary line drawn through the center of the aortic anulus (Fig 1, C).

As the tricuspid valve—mainly the septal cusp—overhung the defect and therefore was interposed between the VSD and the aortic orifice, its papillary muscles were detached from the septum and secured with interrupted stitches for later reinsertion at the tunnel patches. On the other hand, when it was not feasible to reinsert the papillary muscles, their resection and that of a portion of the corresponding septal cusp was performed. After that, bicuspidization of the tricuspid valve was performed by suturing the left margin of the anterior cusp to the remaining septal cusp and also by suturing the left edge of the posterior cusp to the corresponding margin of the septal cusp. Other authors¹¹ have reported a similar technique for repair of mitral insufficiency, detaching the posterior cusp of the tricuspid valve and its chordae tendineae for use as a graft to supply the anterior leaflet of the mitral valve with chordae tendineae. After that, the tricuspid valve was turned into a bicuspid valve through plication of the valve anulus.

The second step was performed through a transverse or oblique right ventriculotomy, depending on the course of the conal right ventricular artery branches. The location of the enlarged and partially tunneled VSD was reassessed, as well as its relationship to the aortic and pulmonary orifices. An obstructive subaortic conus and often a portion of the infundibular septum were partially excised at this time.

The second, more distal patch was trimmed to an appropriate size and oval configuration. It was secured around the aortic orifice and subaortic conus, with the long axis of the oval patch almost perpendicular to the long axis of the first patch (Fig 1, C).

Finally, the third or intermediate patch was sized and

trimmed according to the distance and spatial requirements between the 2 previously placed patches. It generally had a trapezoidal configuration, with the shorter side contouring the septal portion of the tricuspid valve. It was sutured in place with continuous sutures of 5-0 polypropylene and reinforced with interrupted 5-0 polypropylene sutures (Fig 1, D).

The pulmonary stenosis was relieved either by a pulmonary valvotomy and/or infundibular resection, with or without the insertion of a transannular bovine pericardial patch (5 cases), or by a valved extracardiac conduit (6 cases).

Associated anomalies were corrected concomitantly during the same surgical procedure. Straddling of the tricuspid valve was treated with resection of the papillary muscle and the chordae tendineae.

Cardiopulmonary bypass was discontinued as soon as the patient was fully rewarmed and sinus rhythm had returned. The ratio between the peak pressure in the right ventricle and that in the left was measured, as well as the left ventricular–aortic gradient. The mean intraoperative left ventricular–aortic systolic pressure gradient measured after repair was 8.7 mm Hg and the mean peak systolic right ventricular/left ventricular pressure ratio was 0.5.

Results

Early mortality. There were 2 hospital deaths (11.1%). One patient, a 2-month-old boy, had severe aortic coarctation and subaortic stenosis, in addition to DORV with noncommitted VSD. He had severe cardiac failure and required preoperative mechanical ventilation. He died shortly after the operation of low cardiac output and coagulopathy. The autopsy revealed

additional severe pulmonary hemorrhage. The other patient, an 11-year-old girl, required reoperation on the first postoperative day for repair of a residual VSD, had atrial flutter, and required electrical cardioversion in the postoperative period. She died suddenly, 30 days after discharge.

Late deaths. There were 3 late deaths (16.6%). One death occurred in a 6-year-old girl whose postoperative course had been complicated by low cardiac output and a complete permanent atrioventricular block, necessitating pacemaker implantation. She was reoperated on twice: 2 months after repair to close a residual VSD and 4 months after repair because of bacterial endocarditis. At this operation all pericardial patches were replaced. She died of bacterial endocarditis and generalized sepsis during the fifth month after the operation. The other 2 patients were all in New York Heart Association (NYHA) class I without medication and both died suddenly, 7 months after total repair. Retrospectively, during the early postoperative period these patients had occasional episodes of ventricular or atrial tachyarrhythmias.

Postoperative hospital complications. Supraventricular tachyarrhythmias occurred in 3 patients and were successfully managed with electric cardioversion in 1 and with amiodarone in the other 2. One patient had a ventricular arrhythmia and was treated with lidocaine and amiodarone. A right pleural effusion occurred in 1 patient and responded to diuretic therapy. One patient had bronchopneumonia, and another had complete atrioventricular block and required permanent pacemaker implantation.

Follow-up information. All hospital survivors but one, who was lost to follow-up 1 year after repair, were followed up for a mean of 2.65 years, ranging from 1 month to 12 years. Of the 12 patients available for follow-up (Table I), all but 1 were in NYHA class I. A hemodynamically insignificant residual VSD was present in 2 patients. Two patients had residual right ventricular outflow tract obstruction (pressure gradients of 25 and 57 mm Hg). Moderate pulmonary valve insufficiency was found in 2 patients, despite the use of a monocuspid right ventricular outflow patch. No echocardiographic or angiographic evidence of left ventricular outflow tract obstruction was observed (Fig 2). Mild mitral valve incompetence after the operation appeared in 1 patient, and moderate tricuspid valve regurgitation was observed in another.

One 3-year-old boy was reoperated on twice. The first reoperation, 30 days after repair, was for correction of significant mitral valve regurgitation. A mitral cleft was found and repaired successfully. Forty days later, a second reoperation was needed for diaphragmatic plication.

Discussion

Noncommitted VSDs are reported in 10% to 20% of patients with DORV. In these cases, the noncommitted VSD provides the only exit from the left ventricle. The ideal surgical solution should therefore include restoration of a physiologic circulation by a complete 2-ventricle repair. To achieve this, one must (1) establish anatomic continuity between the left ventricle and the aorta through the VSD, creating a large intraventricular tunnel connecting the left ventricle through the defect and the aorta; (2) establish an unobstructed connection between the right ventricle and the pulmonary artery; and (3) repair the associated anomalies.

With a noncommitted VSD, the intraventricular tunnel is technically more difficult to construct because (1) the VSD is remote or distant from the aorta, often located within the right ventricular inlet or muscular septum, and frequently is restrictive; (2) there is interference of anatomic structures, mostly the tricuspid valve and its tension apparatus between the VSD and the aorta; (3) the different spatial orientation between the VSD and the aorta requires that the intraventricular tunnel partially surround the tricuspid valve to reach the aorta at an almost 160° angle; (4) an obstructive subaortic conus is present; (5) the right ventricular outflow tract is interposed along the course of the intraventricular tunnel; and (6) associated anomalies are present.

The early reports of intracardiac repair of DORV were made by Patrick and McGoon¹² and Kawashima and associates.¹³ One horseshoe- or kidney-shaped patch was used to construct an intraventricular tunnel to connect the VSD to the aorta in patients with Taussig-Bing anomaly or with subaortic VSD.

Since the initial report of a successful repair of DORV with noncommitted VSD by Kirklin and Castaneda⁶ in 1977, several other reports have described the use of an intraventricular repair. However, both the surgical mortality and the late survival of these patients proved to be less than satisfactory. Hospital mortality exceeded 30% and the late mortality as reported by Judson and associates⁴ and Kirklin and Castaneda⁶ was 44%.

Since a definitive biventricular repair in infancy was considered an independent risk factor for death by Kirklin and Castaneda,⁶ we decided to postpone definitive repair to an older age. When necessary, we first carry out a palliative procedure.

Other alternative techniques have also been described for repair of DORV with noncommitted VSD, including (1) biventricular repair with the right ventricle used as the systemic ventricle, adding an atrial switch oper-

Fig 2. Cineangiograms in a patient with DORV with noncommitted VSD. **A**, Preoperative study made in the long axial projection showing a remote VSD. **B**, Postoperative study showing an unobstructed LV-to-aorta tunnel. DORV, Double-outlet right ventricle; VSD, ventricular septal defect; LV, left ventricle; Ao, aorta; PRE, preoperative; POST, postoperative.

ation (Senning or Mustard), and tunneling the left ventricular outflow to the pulmonary artery; (2) univentricular heart repair; and (3) interposing an apical valved conduit between the left ventricular apex and the descending thoracic aorta with VSD closure and reconstruction of the right ventricle outflow tract.

Biventricular repair connecting the left ventricle to the aorta has the advantage of establishing normal anatomy and physiology. As Sakata and associates¹⁴ suggested, intraventricular repair is feasible whenever the distance from the tricuspid valve to the pulmonary valve is sufficiently wide or when this distance is at least equal to the aortic valve diameter. In patients without significant pulmonary stenosis and when the intraventricular repair is not feasible, then the arterial switch operation added to a tunnel connecting the left ventricle to the pulmonary artery^{15,16} becomes the alternative of choice.

In our series, the distance between the tricuspid and pulmonary valves was not considered a contraindication to construction of the left ventricular–aortic intraventricular tunnel. In the presence of pulmonary stenosis, the pulmonary valve was closed, the intraventricular tunnel was constructed crossing the pulmonary orifice, and an extracardiac conduit was interposed between the right ventricle and the pulmonary trunk. In the absence

of pulmonary obstruction, the right ventricular cavity was dilated and the distance between the valves was widened, facilitating the construction of the tunnel.

Tunneling the VSD to the pulmonary artery added to an atrial switch procedure is a technique that should be restricted and replaced to exclusive use of the arterial switch. Serious atrial rhythm disturbances, tricuspid regurgitation, systemic and/or pulmonary venous pathway obstruction, and reduced systemic ventricular function are significant concerns after this type of procedure.^{17–22}

Univentricular repair is technically simpler than intraventricular rerouting in patients with complex forms of DORV because it avoids the technical difficulties of creating an intracardiac tunnel in the presence of (1) a noncommitted VSD, (2) anomalies of the atrioventricular valves, and (3) hypoplasia of the right or left ventricle.²³ Delius and colleagues²⁴ reported early and intermediate results of biventricular and univentricular repair of complex heart defects with 2 functional ventricles of adequate size. Although the univentricular repair showed better early and intermediate results, only 5 of the 16 patients of this group and none of the 34 patients subjected to biventricular repair had a noncommitted VSD. Thus no group was available for com-

parison of our results with noncommitted VSD. In our experience, unlike that of other authors,^{23,24} straddling of one of the atrioventricular valves is not an absolute contraindication for intraventricular rerouting. Straddling of the tricuspid valve was satisfactorily solved by resection of the papillary muscles and its chordae tendineae and reimplantation on the second patch after construction of the tunnel; when these muscles were blocking the passageway of the tunnel, they were resected together with part of the septal cusp, with subsequent bicuspidization of the tricuspid valve. The bicuspidization of the tricuspid valve was performed by suturing the anterior cusp to the remaining septal cusp and the posterior cusp to the corresponding margin of the septal cusp. Two patients had straddling of the tricuspid valve and 1 had straddling of the mitral valve. In these patients, the papillary muscles of the tricuspid and mitral valves were detached and reimplanted to the first bovine pericardial patch.

In a recent report, Serraf and associates²³ published the results of the surgical approaches of DORV associated with straddling of atrioventricular valves. Fifteen patients had DORV and 4 had noncommitted VSDs. The presence of ventricular hypoplasia and the existence of abnormal chordal insertion (type C) were the only limiting factors for biventricular repair.

This study includes 18 patients with an echocardiographic, angiographic, or intraoperative diagnosis of DORV with a noncommitted VSD. Intraventricular repair was possible in all. The use of multiple patches significantly simplified construction of the intraventricular tunnel because it permitted the surgeon to better visualize and tailor the shape and size of the tunnel and change the spatial orientation of the tunnel to reroute the VSD to the aorta.

In the majority of cases the VSD proved restrictive. Consequently, it had to be enlarged anterosuperiorly to allow for an unobstructed tunnel. Through a right ventriculotomy, the second patch was sutured around the aortic orifice and subaortic conus, with the long axis of the oval patch oriented perpendicular to a line drawn through the center of the VSD and the center of the aortic orifice. The third and sometimes the fourth patch were trimmed according to the distance and spatial disposition of the other 2 patches. This stepwise construction also permits a better gauging of the internal dimensions of the tunnel throughout its course.

Because the intraventricular baffle connecting the left ventricle to the aorta occupies quite a bit of space within the right ventricle, particularly in the presence of an associated pulmonary stenosis, the right ventricular outflow tract had to be enlarged in 5 patients or

reconstructed by interpositioning of an extracardiac conduit in 6 patients.

The 3 late deaths were sudden and were most likely related to postoperative tachyarrhythmias. Two of the patients who died had evidence of ventricular and atrial arrhythmias in the immediate postoperative period. One patient had complete atrioventricular block develop after repair, requiring pacemaker implantation. This patient died of bacterial endocarditis 5 months after repair. Shen and his colleagues²⁵ reported an 18% incidence of late sudden death in 89 survivors of DORV repair. Cox proportional hazards multivariate analysis disclosed that older age at the time of operation, perioperative or postoperative ventricular tachyarrhythmias, and complete atrioventricular block were significant risk factors for late sudden death.

The new intraventricular tunnel made of bovine pericardium does not provide a rigid septum but a rather floppy one. Hence blood flow may lose part of its kinetic energy and also become turbulent and therefore cause left ventricular dysfunction. One patient in our series had severe low cardiac output in the postoperative period and died. All remaining patients had normal left ventricular function at echocardiography. Probably, some compensatory hyperkinesis of the other left ventricular segments coexists, helping to maintain adequate ventricular function. A similar left ventricular dysfunction occurs after ventricular septation with repair of single ventricle.²⁶⁻²⁸ In these cases, bulging of the prosthetic septum results in bigger left ventricular end-systolic volume, and the paradoxical movement of the septum reduces the left ventricular ejection fraction. Right ventricular contraction is augmented by a shift of the patch toward the right ventricle. Nevertheless, cardiac output seems to be normal at rest and depressed during exercise.²⁹

Left ventricular outflow tract obstruction along the intraventricular tunnel because of residual restrictive VSD is a widely known and described complication after biventricular repair.^{30,31} This complication was responsible for most of the reoperations or late deaths in other reported series. Only 2 of our patients had very mild left ventricular outflow tract obstruction develop. We believe that this low incidence could be explained by the generous enlargement of all VSDs and by the stepwise fashioning of the intraventricular tunnel, creating a very large connection between the VSD and the aorta.

This technical modification, using multiple (2-4) patches for intraventricular rerouting of left ventricular blood to the aorta in cases of DORV with noncommitted VSD, is simple, safe, and applicable to the great majority of patients with this anomaly. With additional

experience and improved selection criteria, it is hoped that both the hospital and late mortality and morbidity can be substantially reduced.

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