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Double-outlet Right Ventricle with Nonrelated Ventricular Septal Defect: Surgical Results Using the Multiple Patches Technique

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ABSTRACT

Objective: Introduce a new surgical technique for biventricular correction of double-outlet right ventricle with noncommitted ventricular septal defect.

Methods: From April 1987 to February 1996, 15 patients with double-outlet right ventricle with noncommitted ventricular septal defect were operated on using a new technique for biventricular repair with multiple bovine pericardial patches to create a tunnel between the left ventricle and the aorta. Ages ranged from two months to 13 years (mean age 4.8 years). Thirteen patients had situs solitus and levocardia, one patient had situs inversus and dextrocardia, and one patient had situs solitus and dextrocardia. Construction of the tunnel began at the right atrium. The ventricular septal defect (VSD) was enlarged anteriorly, if restrictive or small, and the first patch was sutured in the infero-posterior edge of the VSD. The second, third and sometimes the fourth patches were sutured in sequence, through the right ventriculotomy, directing the tunnel to the aortic annulus.

Results: Overall mortality was 20%, with two early and one late death. The surviving patients were followed-up for a period ranging from ten months to nine years (mean 33 months), and all were in functional class I (NYHA). Minimal residual ventricular septal defect was observed in one patient, stenosis in two patients and moderate pulmonary insufficiency in one. There was no obstruction of the intraventricular tunnel between the LV and the aorta.

Conclusion: Based on these data, we conclude that this technical modification for the biventricular repair of the double-outlet right ventricle with noncommitted VSD allows for the construction of a tunnel with adequate internal diameter, respecting the spatial changes between the VSD and aorta. In addition, the intraventricular bovine pericardial tunnel takes up less space, thus reducing the incidence of right ventricle outlet obstruction.

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INTRODUCTION

According to Lev et al. [Lev 1972], the double-outlet right ventricle (DORV) is a form of ventriculoarterial connection in which the great arteries arise completely or nearly so from the right ventricle. Mitral-aortic or mitral-pulmonary connection may or may not be present. When more than 50% of each artery arises from the right ventricle, the condition is termed double-outlet right ventricle [Anderson 1983] and is classified, according to Zamora et al. [Zamora 1975], based on the position of the intraventricular communication in relation to the great vessels of the base in subaortic, subpulmonary, doubly related and nonrelated. Uncommonly, there is no intraventricular communication and the blood from the left chambers gets to the right atrium by an interatrial communication, the right ventricle, and the aorta. The DORV is considered a noncommitted ventricular septal defect (VSD) when the VSD is far removed from both semilunar valves and is represented by defects in the inlet septum, an atrioventricular type of septal defect, or a defect at the trabecular portion of the muscular septum.

The objective of this study is to present our experience with a technical modification using multiple patches for the biventricular repair of the DORV with noncommitted VSD.

Table 1. Associated Lesions

	Age	Gender	Associated Lesions
1	4 y	F	AoC + TVS
2	5 y	F	LAo + AJP
3	6 y	F	IVPS + Single atrium + TVS
4	18 mo	F	IVPS
5	6 y	F	IVPS + TVS
6	4 y	M	IVPS + RC with Cx
7	11 y	F	LAo + PA
8	5 y	F	Situs inversus + Dextrocardia + IVPS + SCO
9	6 mo	M	SAS + AoC + ACP
10	1 y	M	SAS + PLSVC + anomalous RV Band
11	2 mo	M	SAS + AoC + ACP
12	5 y	M	IVPS + LAo
13	2 mo	F	SAS + ACP
14	13 y	F	IAC + IVPS + LAo + AJP
15	10y	M	Dextrocardia + Left pulmonary arterial stenosis + TVS

Mo: months; y: years; M: male; F: female; AoC: aortic coarctation; IVPS: infundibular-valvar pulmonary stenosis; SAS: subaortic stenosis; RC: right coronary; Cx: circumflex coronary; ACP: arterial canal persistence; IAC: interatrial communication; PLSVC: persistent left superior vena cava; L-Ao: aorta at the right of the pulmonary trunk; TVS: tricuspid valve straddling; AJP: auricular JP; PA: pulmonary atresia; SCO: single coronary ostium

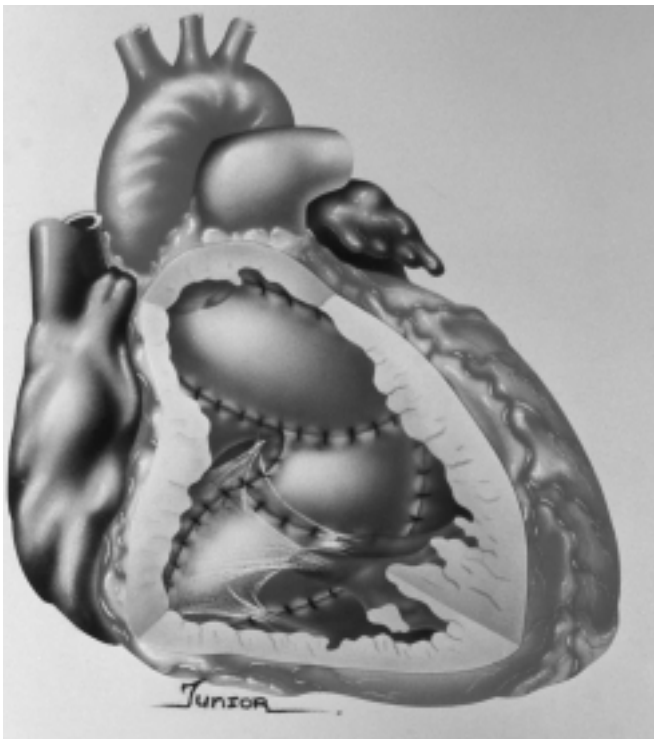


Figure 1. Technical illustration of double-outlet right ventricle with noncommitted ventricular septal defect with multiple bovine pericardial patches. The use of one or more pericardial patches allows better positioning of the tunnel, respecting directional change between the ventricular septal defect and the aorta. In addition, it occupies less space in the right ventricle outlet.

MATERIALS AND METHODS

Fifteen patients with DORV with noncommitted VSD were operated on using this technique from April 1987 to February 1996. Patient ages ranged from two months to 13 years (mean age 4.8 years). Thirteen patients had situs solitus and levocardia, one patient had situs inversus and dextrocardia and one patient had situs solitus and dextrocardia. Two patients had right atrial isomerism and auricular juxtaposition. The associated anomalies observed were (see Table 1 ◎): pulmonary stenosis (7 patients); aortic coarctation (3 patients); subaortic stenosis (4 patients), tricuspid valve straddling (4 patients); arterial canal persistence (3 patients); pulmonary atresia (1 patient); interatrial communication (1 patient); persistent left superior vena cava draining into the coronary sinus (1 patient).

Echocardiogram and cardiac catheterization were used for diagnosis of the patients. More recently, nuclear magnetic resonance, carried out in the last four patients, was used to confirm the diagnosis, and to try to establish the distance between the VSD and the aorta.

Of the 15, five patients underwent the repair surgery. Three of them had modified Blalock-Taussig systemic-pulmonary anastomosis and two had pulmonary trunk binding.

Repair was carried out by median sternotomy. The great vessels, ventricular disposition, and the coronary anatomy were examined. Extracorporeal circulation was established by aortic and vena caval cannulation with hypothermia at 20° C. Cold cardioplegia was performed by the infusion of crystalloids at 4° C in the aorta at 20 ml/kg and maintained at 10 ml/kg every 20 minutes.

Intraventricular communication was identified by longitudinal right atriotomy, pushing away or removing the septal cuspid from the tricuspid valve. The relationship and distance between the VSD and the aorta was established.

If small or restrictive, the VSD was enlarged anteriorly to allow broad blood flow from the left ventricle to the aorta. The first bovine pericardial patch was sutured in the infero-posterior edge of the VSD with separate Prolene 6-0 stitches sutured leftward in small pericardial patches, giving special attention to the atrioventricular node. In three cases of tricuspid valve straddling, the papillary muscles were sectioned for the tunneling and later reimplanted in the patch. Subsequently, the second, third and sometimes fourth patches were sutured by direct longitudinal ventriculotomy directing the VSD to the aortic annulus (see Figure 1 ◎). In the last two cases an endoscope (Pentax EPM 3000 FG 2901) was used after the repair to evaluate the left ventricular outlet (see Figure 2 ◎).

In the presence of associated pulmonary stenosis, infundibular resection was carried out by right atriotomy, which might be completed by ventriculotomy. Valvar pulmonary stenosis was repaired by valvotomy after opening the pulmonary trunk. Enlargement of the right ventricular outlet, when necessary, was done with bovine pericardial patches when pulmonary annulus was adequate for the body surface (5 cases). Otherwise right ventricular outlet enlargement was accomplished using a pericardial monocuspid graft (1 case) or valved corrugated bovine pericardial tube (2 cases).

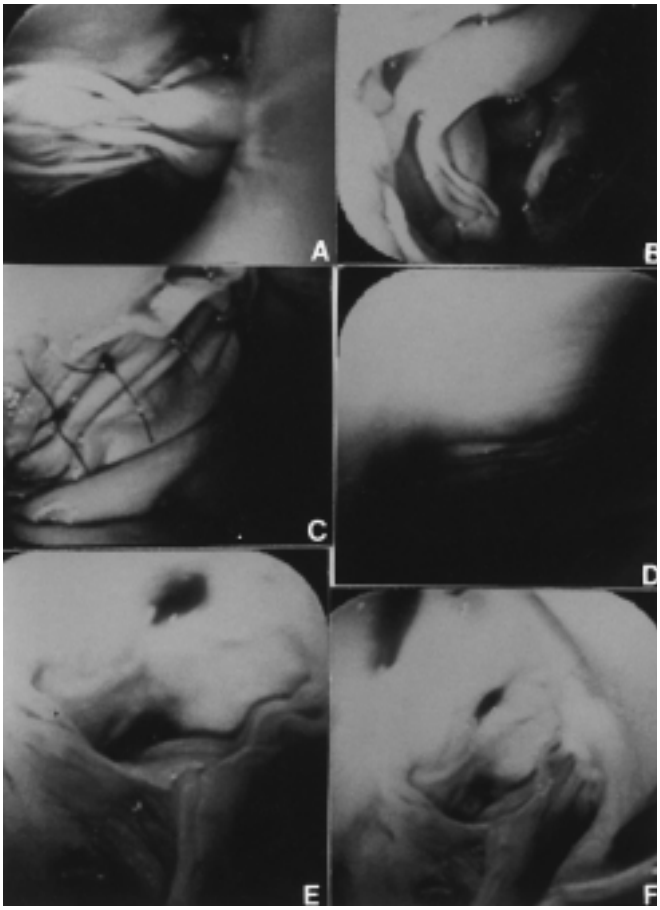


Figure 2. Intraoperative videoscopy showing the final aspect of ventricular septal defect (VSD) tunneling to the aorta in one case of double-outlet right ventricle with noncommitted VSD. The endoscope was introduced through the ascending aorta. The LV-aorta tunnel was photographed at different levels as it was removed: A and B: Beginning of the tunnel. Mitral valve papillary muscles and the VSD edge were seen. C and D: Aspects of bovine pericardial patches are seen in the midway of the tunnel. E and F: Photographs of the annulus, aortic valve and left coronary ostium taken from the end of the tunnel.

In the cases with persistence of the arterial canal or aortic coarctation, we chose simultaneous surgical repair by median sternotomy prior to cardiopulmonary bypass installation.

After the intracardiac repair, extracorporeal circulation was temporarily discontinued and pressure gradients were measured between the left ventricle and the aorta, and between right and left ventricles. There was no systolic gradient between the left ventricle and the aorta and the RV systolic pressure/LV systolic pressure ratio was at all times ≤0.5. Subsequently, cardiopulmonary bypass was restarted and maintained until warming had taken place.

Clinical data and echocardiographic study were used for the postoperative evaluation in addition to cardiac catheterization in five patients.

RESULTS

There were three postoperative deaths (20.0%) — two hospital deaths and one late death. The first one was a six-year old patient who was reoperated at postoperative month five due to residual VSD. The patient had a course of bacterial endocarditis, septicemia and death. The second patient, 11 years old, with an echocardiographic diagnosis of residual VSD, was discharged in good conditions and was asymptomatic, but had sudden death on postoperative day 35, probably due to arrhythmia. The third patient was a two-month old baby who died immediately after the surgery due to low cardiac output and coagulopathy.

Intra-hospital postoperative complications were as follows: supraventricular tachycardia in three patients, which was controlled by amiodarone (2 cases) and electric cardioversion (1 case); right pleural effusion (2 patients); and bronchopneumonia (1 patient).

The 12 remaining patients were followed-up for a period of ten months to nine years (mean 33 months). All of them were in functional class I (NYHA). Echocardiography and cardiac catheterization (5 cases) showed minimal residual VSD in one patient, residual pulmonary stenosis with a gradient of 22 mmHg and 40 mmHg in two patients, pulmonary insufficiency in one patient who underwent enlargement of the right ventricle outlet with a monocuspid graft, and right bundle-branch block in three patients (see Graph 1 ◎). Obstruction of the LV-Aortic tunnel was not observed in any of the patients (see Figure 3 ◎).

DISCUSSION

The prevalence of noncommitted VSD among DORV is 10% to 20%. Surgical repair is aimed at: 1) reestablishing the connection between the left ventricle and the aorta, usually by the creation of a wide tunnel between the VSD and the aorta; 2) establishing adequate communication between the right ventricle and the pulmonary trunk by pulmonary valvotomy, infundibular resection, outlet enlargement using valved or non-valved pericardium, or by overriding the valved tubular graft; and 3) repair of associated anomalies.

The surgical repair of double-outlet right ventricle with noncommitted ventricular septal defect is a challenge for the surgeon because biventricular repair is directing at making the left ventricle a systemic ventricle. The challenge is tunneling the left ventricle to the aorta, due to the distance and the position of the VSD in relation to the aorta, the presence of atrioventricular valve malformations (tricuspid and mitral valves), and the overriding of anatomic structures between the VSD and the aorta. The late results are not satisfactory, with a high incidence of reoperation due to obstruction of the right and left ventricular outlet and increased mortality, ranging from 29% to 66%. [Kirklin 1986, Musmeci 1988, Piccoli 1983, Sondheimer 1977, Stewart 1979].

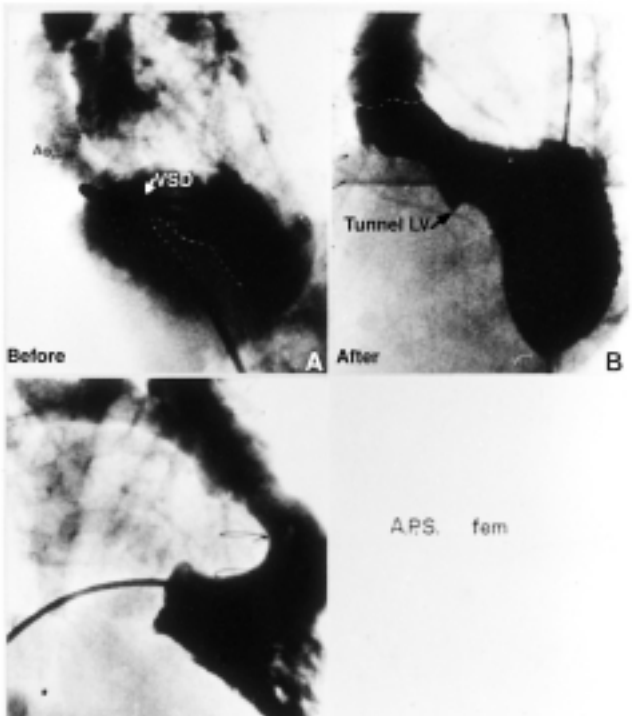
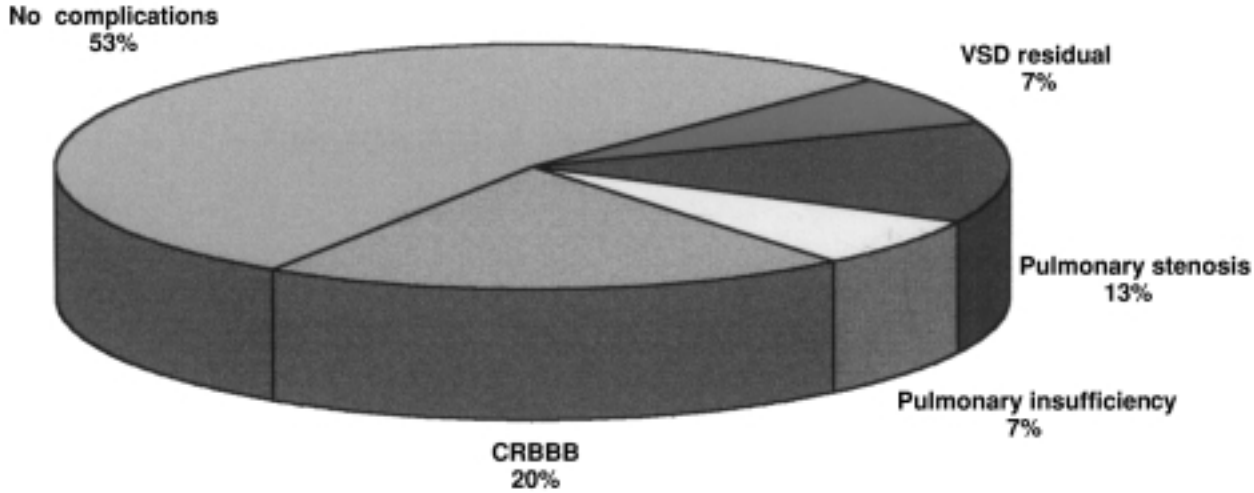


Figure 3. Pre- and postoperative angiography carried out in patient 15: A – Preoperative study showing ventricular septal defect far removed from the aorta. B – Postoperative study with left ventricular injection, left anterior oblique projection, showing extensive tunnel built between the left ventricle and the aorta. C – Postoperative study with left ventricular injection, right anterior oblique projection, showing left ventricle outlet. VSD: ventricular septal defect; VE-left ventricle; Ao – aorta.

The first biventricular repair in DORV with noncommitted VSD was carried out by Kirklin & Castañeda [Kirklin 1977], in 1977 with the tunneling of the left ventricle and the aorta through the VSD using a Dacron graft and repair of the right ventricle outlet using a valved tubular graft.

In patients not eligible for intraventricular approach, the defect can be repaired by the closure of the VSD and the pulmonary trunk with reestablishing the communication between the right ventricle and the aorta with a valved conduit followed by inversion of the atrial circulation (Senning or Mustard's operation) or the arterial circulation (Jatene's operation) [McGoon 1976]. The disadvantage of this repair is that it transforms the right ventricle into a systemic ventricle. In complex cases, with multiple VSDs, pulmonary stenosis, tricuspid or mitral valve straddling and left ventricle hypoplasia, the univentricular correction, the Fontan operation is indicated [Gonzales 1977].

Using the multiple patch repair technique, our overall mortality rate was 20%. All of the surviving patients (12) were in functional class I (NYHA) with residual VSD in one, pulmonary stenosis with pressure gradients of 22 mmHg and 40 mmHg measured by echo-Doppler in two, pulmonary insufficiency in one patient who underwent enlargement of the right ventricle outlet with monocuspid grafting, and right bundle-branch block in three patients. In the three cases with tricuspid straddling who had the papillary muscles sectioned and reimplanted in the bovine pericardial patch there was no valvular insufficiency or stenosis in the postoperative follow-up. None of the patients had obstruction of the tunnel between the left ventricle and the aorta. When these data were analyzed and compared to the literature, it might be concluded that this technique for a step-by-step correction allows for the evaluation of the tunnel's internal diameter after the placement of each pericardial patch, keeping adequate dimensions in the direction changes between the VSD and the aorta. The mortality and complication rates are acceptable, making it possible for patients previously eligible only for univentricular repair to undergo complete biventricular repair with preservation of systemic left ventricle.



Graph 1. Late Postoperative Complications. VSD: ventricular septal defect; CRBBB: complete right bundle-branch block.

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