Long-Term Results after Biventricular Repair for Double-Outlet Right Ventricle

Chih-Yuan Lin, Chien-Sung Tsai, Gou-Jieng Hong, Pi-Chang Lee, Betau Hwang and Zen-Chung Weng

Purpose: The aim of this study was to review our surgical strategy and long-term follow-up results in patients with double-outlet right ventricle who underwent biventricular repair.

Methods: From 1995 to 2003, a total of 15 patients (12 boys, 3 girls; mean age, 3.3 years) underwent biventricular repair for DORV. Among these patients, 8 patients received palliative procedures before definitive repair. According to the location of the VSD, these patients were divided into four groups: Five patients had subaortic VSD, 6 had subpulmonary VSD (Taussig-Bing type), 2 had doubly committed VSD, and 2 had remote VSD. In the subaortic, doubly committed, and non-committed VSD groups, the intraventricular baffle procedure to direct blood from the left ventricle to the aorta combined with right ventricular outflow tract (RVOT) reconstruction with or without extracardiac conduit implantation was performed. In the Taussig-Bing group, 4 patients underwent VSD closure and the Jatene operation, while 2 patients underwent VSD closure and the Mustard operation.

Results: There was one early death, and no late death. One patient in the remote VSD group had complications of complete heart block and underwent pacemaker implantation. Patients were monitored for a mean period of 107.3 ± 31.0 months (range, 66-164 months). Two patients underwent re-operation during the follow-up period. All the survivors were in New York Heart Association (NYHA) class I or II.

Conclusion: We suggest that most of DORV patients with adequate size of both ventricles can be successfully managed using biventricular repair. The initial survival was high, and long-term follow-up results were acceptable.

Key Words: Biventricular repair • Double-outlet right ventricle • Taussig-Bing anomaly • Ventricular septal defect

INTRODUCTION

Double-outlet right ventricle (DORV) is a congenital anomaly in which both the aorta and the pulmonary artery originate from the right ventricle. The only outlet from the left ventricle is a ventricular septal defect (VSD). Although the presence of aortic-mitral discontinuity and bilateral coni are important descriptors, they should not serve as absolute prerequisites for the diagnosis of DORV. From a surgical perspective, it is most useful to adopt the ‘50% rule’ in defining DORV. With this rule, a heart has DORV if > 50% of both arteries arises from the right ventricle. Usually, all of one great artery and 50% or more of the other great artery arise from the right ventricle in DORV.

DORV includes a broad spectrum of anatomical variants and anatomical malformations. Since the only outlet from the left ventricle is a VSD, the ejected stroke volume of the left ventricle passes through the VSD. The relationship between the VSD and the great arteries, the
relative outflow tract obstruction, and the relative systemic-to-pulmonary artery resistance determine the hemodynamic situation. DORV may mimic VSD, Tetralogy of Fallot, and transposition of the great arteries. There is always some degree of arterial desaturation. Based on the work of Lev and colleagues in 1972, 4 DORV is classified into four groups according to the relationship of the VSD to the great arteries: subaortic, subpulmonary, doubly committed, and non-committed (remote).

In the present era, the surgical approach to DORV for biventricular repair is to connect the left ventricle to the systemic circulation and the right ventricle to the pulmonary circulation with the use of an intraventricular baffle, either alone or in association with extracardiac procedures such as an arterial switch operation or the insertion of an extracardiac conduit. Modified Fontan procedures were advocated when biventricular DORV repair was either impractical or extremely complex. 5 With the improving short-term and medium-term outcome for Fontan procedures in recent years, this approach might also be extended to patients who are at increased risk with a conventional biventricular repair. 6,7

This report reviews the anatomic findings, surgical strategies, and results among patients with DORV presenting for biventricular repair at Taipei Veterans General Hospital between 1995 and 2003.

MATERIALS AND METHODS

Patients

From August 1995 to August 2003, 15 children (age range, 20 days to 11 years; mean age, 3.3 years) who received biventricular repair for DORV were enrolled into this study. The medical records were retrospectively reviewed. There were 12 males and 3 females. A diagnosis of DORV was made if both great arteries originated predominantly from the right ventricle on application of the ‘50% rule,’ which requires one great artery to arise completely, and the other more than 50%, from the right ventricle. Additionally, the absence of aortic-mitral continuity was included in the definition of DORV. The diagnosis and anatomic findings were based on a combination of angiography, echocardiography, and surgical inspection. Based on the relationship of the VSD to the great arteries, patients with DORV enrolled in the study were classified into four groups: subaortic, subpulmonary, doubly committed, and non-committed (remote).

Patients’ demographics and clinical characteristics are summarized in Table 1.

Palliative procedures

Eight of 15 patients (53%) had palliative procedures preceding complete DORV repair. The palliative procedures for each group are summarized in Table 2.

Operative data

All patients were operated on with standard cardiopulmonary bypass, bicaval cannulation, and moderate hypothermia with cold potassium cardioplegic arrest. In all patients, the repair was performed through a right ventriculotomy. Three types of definitive repairs were performed: 1) intraventricular tunnel repair with a baffle from the left ventricle to the aorta together with right ventricle outflow tract (RVOT) patch reconstruction, 2) Rastelli operation with an extracardiac valved conduit, and 3) the arterial switch operation with a patch commit-

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<th>Table 1. Patient characteristics</th>
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<td>Age</td>
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<td>Gender (male/female)</td>
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<tr>
<td>Associated anomalies</td>
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<td>Atrial septal defect</td>
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<td>Peripheral pulmonary stenosis</td>
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<td>Patent ductus arteriosus</td>
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<td>Right aortic arch</td>
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<td>Coarctation of aorta</td>
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<td>Single coronary artery</td>
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<th>Table 2. Palliative procedures</th>
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<td>Group</td>
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<td>Subaortic VSD</td>
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<td>Doubly committed VSD</td>
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<td>Subpulmonary VSD</td>
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<td>Non-committed VSD</td>
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BTS: Blalock-Taussig shunt; PAB: pulmonary artery banding; CoA: coarctation of the aorta; VSD: ventricular septal defect; PDA: patent ductus arteriosus.
ting the left ventricle to the pulmonary artery (neoaorta). All procedures were done by one surgeon (Z-C Weng). In the subaortic VSD group (five patients), two patients required conal septum resection to enlarge the orifice of the VSD. Among the five patients, four patients underwent transannular RVOT patching and one patient required implantation of an extracardiac conduit. In the doubly committed group (two patients), one patient required conal septum resection and transannular RVOT patching and the other patient underwent separate RVOT and main pulmonary artery (MPA) patching. In the subpulmonary VSD group (six patients), the arterial switch operation was done in two patients and the Mustard operation was performed in two patients. In the remote VSD group (two patients), both patients required VSD enlargement with conal septum resection. Among these two patients, one required tricuspid valve chordae detachment and reinsertion as well as extracardiac conduit implantation, while the other patient underwent transannular RVOT patching.

Follow-up

All surviving patients received serial physical examinations and periodic echocardiography in the follow-up period. Only when cardiac function deteriorated was cardiac catheterization done to clarify the etiology.

RESULTS

Early and late mortality

There was one early fatality in the subpulmonary VSD group. The patient underwent VSD closure and the Jatene operation at 20 days of age. Acute left ventricular failure due to iatrogenic aortic regurgitation occurred after the arterial switch operation. There was no late mortality.

Complications

There were three operation-related complications. One patient with remote VSD complicated by complete heart block required pacemaker implantation. Chylothorax occurred in one patient, which was successfully managed with conservative treatment. One patient developed sternal wound dehiscence; this patient successfully underwent wound debridement and rewiring of the sternal wound.

Re-operation and follow-up

Follow-up information was available for all 14 surviving patients. Patients were monitored for a mean of 107.3 ± 31.0 months (range, 66-164 months). During the follow-up period, two patients underwent re-operations at a mean time interval of 58 months (range, 36-80 months) after first definitive repair, and no operative mortality occurred. One patient in the subaortic VSD group developed subaortic stenosis and underwent reconstitution of the left ventricular outflow tract 36 months after their initial surgery (Figure 1). The other patient, also in the subaortic VSD group, had conduit stenosis and underwent conduit replacement 80 months after initial biventricular repair (Figure 2).

Of the 14 patients available for long-term follow-up, 10 patients were in New York Heart Association class I, while 4 patients were in class II.

DISCUSSION

In 1964, Kirklin and colleagues reported successful correction of DORV, subaortic VSD, and concordant AV connections in a child. Since then, complete correction through the use of a variety of surgical techniques has been achieved in more complex forms of DORV. Early reports of successful surgical repair of the Taussig-Bing anomaly were reported by Daicoff and Kirklin in 1967.

DORV is characterized by malposition of the great arteries and includes a wide spectrum of anatomic subtypes requiring a variety of surgical approaches for correction. Previous reviews reported hospital mortality figures of up to 25%, especially among patients with non-committed or subpulmonary VSDs. Since 1980, new surgical techniques including the arterial switch operation have significantly improved the outcomes of complex forms of DORV.

Biventricular repair for DORV is usually accomplished within the first 6 to 12 months of life, thus obviating the need for palliative procedures. If it is anticipated that the final repair will utilize an extracardiac valved conduit or a complicated intraventricular tunnel, it is reasonable to delay correction by performing a palliative procedure including pulmonary artery banding or a systemic-to-pulmonary artery shunting. It is also critical to protect the pulmonary vascular bed early with ap-
propriate palliation to allow for patient growth.

In DORV with subaortic and double-committed VSD, intraventricular tunnel repair, connecting the left ventricle to the aorta, is preferred. In patients with refractory congestive heart failure, who are not immediate candidates for complete repair, pulmonary artery banding is required. In the presence of pulmonary stenosis, the techniques of repair are similar to those utilized for repair of tetralogy of Fallot. If the branch pulmonary arteries are severely hypoplastic or an extracardiac conduit is required for the final correction, palliative systemic-to-pulmonary artery shunting is preferred by some prior to performing the definitive repair later in life. During an intraventricular tunnel repair, if the VSD appears to be restrictive (diameter less than that of the aortic valve), it is enlarged by making an antero-superior incision or by resecting a wedge of the interventricular septum in this area. Prominent right ventricular muscle bundles are resected if necessary. Based on our findings and those in the literature, complications after the intraventricular tunnel repair of subaortic DORV with pulmonary stenosis or doubly-committed VSD without pulmonary stenosis are rare. Complete heart block is uncommon, and the functional status of the most survivors is New York Heart Association class I. Complications requiring reoperation include subaortic obstruction (tunnel related and non-tunnel related), pulmonary outflow tract obstruction and residual VSD.

In DORV with subpulmonary VSD (the Taussig-Bing heart) patients, because of the location of the VSD, oxygenated left ventricular blood preferentially streams through the VSD into the pulmonary artery and O2-desaturated right ventricular blood streams into the aorta, as in transposition of the great arteries with VSD. Many approaches to the repair of this complex subset of patients with DORV are employed: 1) patch tunnelling of the VSD to the pulmonary artery combined with an atrial switch procedure (Mustard or Senning), 2) tunnelling of the VSD to the pulmonary artery, aortopulmonary connection (Damus-Kaye-Stansel procedure) and placement of a valved extracardiac conduit from the right ventricle to the distal pulmonary artery, 3) direct tunnelling of the VSD to the aorta, 4) tunnelling of the VSD to the pulmonary artery combined with an arterial switch procedure, and, in selected situations, 5) tunnelling of the VSD to the aorta with translocation of the pulmonary artery (Reparatio l’étage Ventricular-REV procedure of Lecompte). In our series of 6 patients, we performed arterial switch operations in 4 patients; the other two patients underwent the Mustard operation due

Figure 1. The angiographic study shows left ventricular outflow tract obstruction in a patient with DORV and subaortic VSD at 31 months of age after biventricular repair.

Figure 2. The angiographic study demonstrates extracardiac conduit stenosis in a patient with DORV and subaortic VSD at 45 months after biventricular repair. This patient underwent palliative balloon dilatation before re-operation for conduit replacement.
to severe adhesions caused by previous palliation and single coronary arteries. From our experience and that of others, when technically possible, the arterial switch operation is now the procedure of choice for patients with subpulmonary VSD without significant right ventricular outflow obstruction, as well as for other patients in whom baffling from the left ventricle to the pulmonary artery is technically easier than baffling to the aorta. This approach has led to good results with low operative mortality and low necessity for reoperation.

In DORV with non-committed (or remote) VSD, patients often have inlet (atrioventricular canal-type) VSD. The term ‘non-committed’ is used to define the condition in which the VSD is anatomically related to, or is close to, neither great vessel, being separated from both by considerable heart muscle. This subset of patients has poor outcomes, higher risks for operation, and is frequently treated by univentricular repair. Some patients undergo biventricular repair with an intraventricular tunnel, however, it is usually necessary to enlarge the VSD superiorly and anteriorly to make this possible. If the intraventricular tunnel obstructs the right ventricular outflow tract, it is necessary to place a transannular patch or a valved, extracardiac conduit. Since this tunnel procedure presents a high risk of obstruction during growth, it is a serious limitation of this technique; many groups have been faithful to univentricular palliation as a less risky alternative, despite the uncertainties of the long-term results of the Fontan operation. In another group that still prefers biventricular repair, to overcome the limitation of the presence of tricuspid chordae and by the pulmonary artery to tricuspid valve distance, VSD rerouting to the pulmonary artery followed by arterial switch is suggested. In our study, there were two DORV patients with non-committed VSD, both of them underwent VSD enlargement and conal septum resection. Among them, one patient required tricuspid valve chordae detachment and reininsertion to establish an intraventricular tunnel as well as extracardiac conduit implantation. However, this patient had complications of complete heart block and then underwent pacemaker implantation. The complication of complete heart block could be related to VSD enlargement and conal septum resection. Another patient underwent successful DORV repair with an intraventricular baffle to direct blood from the left ventricle to the aorta through the remote VSD and transannular right-ventricular outflow tract patching. Although the Fontan operation is proposed as a solution to the complex forms of this anomaly, it was always our policy to extend the indications of biventricular repair as much as possible.

In summary, DORV includes a wide spectrum of anatomic subtypes requiring a variety of approaches for correction. We found that biventricular repair is feasible in most patients with DORV. The surgical mortality, operation-related complications, and long-term follow-up results were acceptable for DORV patients undergoing biventricular repair.

REFERENCES

13. Aoki M, Forbes JM, Jonas RA, et al. Results of biventricular re-


