SURGERY FOR CONGENITAL HEART DISEASE

PARTIAL BIVENTRICULAR REPAIR FOR COMPLEX CONGENITAL HEART DEFECTS: AN INTERMEDIATE OPTION FOR COMPLICATED ANATOMY OR FUNCTIONALLY BORDERLINE RIGHT COMPLEX HEART

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Background: Partial biventricular repair consists of bidirectional cavopulmonary anastomosis in the presence of separated systemic and pulmonary circulations, with antegrade flow of inferior caval return through an intact or reconstructed pulmonary outflow tract. This versatile procedure may be used as a definitive repair for patients with a functional right heart complex incapable of supporting an entire cardiac output or in patients with complicated anatomy. **Methods:** From July 1992 to April 1997, 23 patients (median age 5.2 years) underwent partial biventricular repair. In 15 of these cases the entire repair, including bidirectional cavopulmonary anastomosis, intracardiac repair, and right ventricular outflow reconstruction, was performed as a planned procedure at our institution. The other eight patients had previously been placed on a Fontan track and had undergone bidirectional cavopulmonary anastomosis; their circulations were converted to a partial biventricular circulation. **Results:** There were no early deaths. Complete atrioventricular block developed in two patients with straddling tricuspid valve. At a median follow-up of 17 months, there were no late deaths and three patients had undergone reintervention. Partitioning of the pulmonary arteries to create a classic Glenn anastomosis with antegrade flow to the left lung was performed in one case; another patient underwent an atrial septectomy, and the third patient required revision of tricuspid valve repair. All patients are in New York Heart Association functional class I. **Conclusions:** Partial biventricular repair is a versatile strategy that can be used to manage a variety of forms of complex congenital heart disease. Cases for which the repair is useful include those in which complete biventricular repair is unlikely to be achieved because of limited size or function of the right side of the heart and those in which a patient with a ventricle capable of supporting inferior vena caval return was previously placed on a Fontan track. (J Thorac Cardiovasc Surg 1998;116:21-7)

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As experience accumulates with single-ventricle palliation for complex congenital heart disease, it has become clear that there is a spectrum of palliative options that may be appropriate in different cases. The choice between univentricular and biventricular repair remains unclear for a range of borderline lesions.¹⁻³ Moreover, there is controversy surrounding such issues as whether to supplement a bidirectional Glenn anastomosis with additional pulmonary blood flow⁴⁻⁷ and whether to fenestrate a Fontan procedure.^{8,9} Other innovative approaches

Table I. Demographic and diagnostic features of group II patients

Patient	Age (yrs)	Diagnosis	Procedures performed at the time of conversion to partial biventricular repair
16	2.4	TGA, VSD, ASD, straddling tricuspid valve, mild right ventricular hypoplasia	Arterial switch, ASD and VSD closure, tricuspid valve repair, resection of main pulmonary artery aneurysm
17	2.1	TGA, pulmonary atresia, VSD, ASD	Rastelli procedure, ASD and VSD closure, tricuspid annuloplasty
18	3.4	LVOT obstruction (valvular, subvalvular, supraaortic), pulmonary stenosis, VSD	LVOT myectomy, supraaortic aortic augmentation, pulmonary valvuloplasty, pulmonary artery augmentation VSD closure, take-down DKS
19	7.6	Double-outlet left ventricle, pulmonary stenosis, VSD, ASD	Pulmonary root translocation to right ventricle, VSD and ASD closure
20	10.6	Pulmonary atresia, VSD, hypoplastic branch pulmonary arteries	Right ventricle-pulmonary artery allograft conduit, RVOT myectomy, VSD and ASD closure, branch pulmonary artery augmentation
21	7.8	Corrected TGA, pulmonary atresia, VSD	Double-switch procedure (Rastelli and modified Mustard procedures), pulmonary artery augmentation
22	2.1	TGA, pulmonary stenosis, VSD	Rastelli procedure, VSD enlargement, ASD closure, main pulmonary artery ligation
23	7.4	Corrected TGA, pulmonary atresia, multiple VSDs	Double-switch procedure (Rastelli and modified Mustard procedures)

TGA, Transposition of the great arteries; VSD, ventricular septal defect; ASD, atrial septal defect; LVOT, left ventricular outflow tract; DKS, Damus-Kaye-Stansel procedure; RVOT, right ventricular outflow tract.

have also been suggested.¹⁰⁻¹⁴ An intermediate option that may be suitable in particular cases is partial biventricular repair, in which intracardiac repair is supplemented with a bidirectional cavopulmonary anastomosis.¹⁵⁻²⁰ This approach allows the maintenance of equal and separate pulmonary and systemic circulations while at the same time incorporating the pulmonary ventricle into the circulation with a reduced volume load. Since July 1992, partial biventricular repair has been performed in 23 patients who either had a functionally borderline right side of the heart or had previously been placed on a Fontan track primarily because of complicated anatomy.

Patients and methods

Patients. Between July 1992 and April 1997, 23 patients underwent partial biventricular repair. The left and right sides of the heart were completely separated by closing all septal defects, maintaining or reconstructing the pulmonary outflow tract, and routing superior vena caval flow directly to the lungs through a bidirectional cavopulmonary anastomosis. The median age was 5.2 years (7 months to 32 years). This is a significantly older population than our overall cohort of patients undergoing bidirectional cavopulmonary anastomosis during the same period (median 7 months; $p < 0.001$). Patients were categorized in two groups. Group I patients ($n = 15$) underwent planned partial biventricular repair with bidirectional cavopulmonary shunt and intracardiac repair at

our institution. Group II patients ($n = 8$) had previously undergone bidirectional cavopulmonary anastomosis in preparation for an eventual Fontan operation, in most cases because of a combination of complicated anatomy and mild to moderate ventricular hypoplasia, and underwent conversion to partial biventricular repair at our institution. Ages were similar between the two groups, but group I patients had undergone significantly fewer previous procedures than had group II patients (not including previous bidirectional Glenn anastomosis, mean $\pm$ standard deviation 0.9 ± 1.0 vs 2.3 ± 1.3 , $p = 0.01$). Diagnoses of group I patients were Ebstein's anomaly ($n = 6$), pulmonary atresia with intact ventricular septum ($n = 4$), pulmonary atresia or stenosis with hypoplasia of the right side of the heart and ventricular septal defect ($n = 4$), and other hypoplastic right ventricle ($n = 1$). Other associated defects in these patients were straddling tricuspid valve ($n = 1$), partially anomalous pulmonary venous drainage to a left superior vena cava ($n = 1$), and coronary sinus septal defect ($n = 1$). Nine patients had significant tricuspid valvular regurgitation: two mild cases, six moderate cases, and one severe case. Among the nine patients with hypoplastic right ventricle and tricuspid valve, tricuspid Z values ranged from -1 to -5 (median -2.5). Significant ventricle-coronary arterial connections were not present in any of the patients. There was a range of diagnoses among group II patients (Table I), all of whom had undergone bidirectional Glenn anastomosis between 20 months and 4 years before conversion to partial biventricular repair.

Surgical procedures. All patients in group I underwent bidirectional cavopulmonary anastomosis by standard techniques.²¹ In three cases the bidirectional Glenn anas-

tomosis was added after intracardiac repair was completed and right atrial pressure was found to be elevated (either >12 mm Hg or twice left atrial pressure). Complete intracardiac septation and repair was performed in all cases, with procedures including atrial septal defect closure ($n = 10$), tricuspid valvuloplasty ($n = 8$), right ventricular outflow tract reconstruction ($n = 6$, including three cases with allograft conduits), ventricular septal defect closure ($n = 5$), and repair of partially anomalous pulmonary venous drainage ($n = 1$) or coronary sinus septal defect ($n = 1$). All but two of the patients with Ebstein's anomaly underwent some form of tricuspid valve repair, ranging from closure of fenestrations in the anterior leaflet to placement of a simple horizontal annuloplasty stitch to reconstruction of the septal leaflet with pericardium. Cardiopulmonary bypass was used in all cases, with total perfusion times ranging from 60 to 312 minutes (median 92 minutes).

Patients in group II had previously undergone bidirectional cavopulmonary anastomosis. Procedures performed at the time of conversion to partial biventricular repair are listed in Table I. Cardiopulmonary bypass was used in all cases and ranged in duration from 85 to 209 minutes (median 136 minutes).

Results

There were no early deaths. The only early reoperation was a partitioning of the bidirectional Glenn anastomosis to create a classic Glenn anastomosis to the right lung and antegrade flow from the right ventricle to the left lung in a group I patient with hypoplasia of the right side of the heart, pulmonary atresia, and ventricular septal defect who developed superior vena cava syndrome. Among the nine patients with preoperative tricuspid regurgitation, the regurgitation was reduced to none ($n = 3$), trace ($n = 5$), or mild ($n = 1$). No patient required prolonged chest tube drainage for pleural or pericardial effusions. On the day of extubation, pressures in the superior vena cava ranged from 8 to 18 mm Hg (median 12 mm Hg), and pressures in the right atrium ranged from 2 to 11 mm Hg (median 5 mm Hg). Patients were discharged after a median hospital stay of 8 days (range 3 to 25 days).

Cross-sectional follow-up was obtained at a median of 19 months (range 2 to 58 months) and was complete in all cases. There were no late deaths, and no patients required take-down of the Glenn anastomosis. Two group I patients underwent late reoperation. A patient with pulmonary atresia and intact ventricular septum had a small atrial septectomy performed 7 months after partial biventricular repair to decompress the right atrium and ventricle. This patient had a preoperative tricuspid valve Z value of -4 (Fig. 1). A patient with Ebstein's anomaly and severe tricuspid regurgitation required re-

Fig. 1. Preoperative apical four-chamber echocardiogram from a group I patient with pulmonary atresia with intact ventricular septum and a tricuspid valve Z value of -4 who had a small atrial septectomy performed 7 months after partial biventricular repair as a result of failure of the right side of the heart to grow. LA, Left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

sion of the tricuspid valve repair 6 months later for recurrent moderate regurgitation. At follow-up echocardiography, tricuspid valve Z values in patients with hypoplastic right ventricle and tricuspid valve did not differ significantly from before the operation, and tricuspid regurgitation had not returned or progressed in any of the other patients with preoperative regurgitation. No patient had undergone follow-up cardiac catheterization. Arterial oxygen saturation ranged from 93% to 99%, with a median of 96%.

Discussion

Rationale for partial biventricular repair. Despite modifications that have led to improved outcomes,^{22, 23} the Fontan operation remains a suboptimal form of definitive palliation for many patients with complex congenital heart disease. Bidirectional cavopulmonary anastomosis, which has proved a useful intermediate step in staged palliation for patients with functional single ventricle,²⁴⁻²⁶ is probably not an adequate alternative for definitive palliation in most cases. Although a bidirectional Glenn anastomosis with additional pulmonary blood flow has been proposed as a potentially acceptable definitive palliation for patients at high risk for a Fontan operation.⁷ There is no strong evidence that additional pulmonary blood flow in a parallel circulation can sufficiently mitigate the pulmonary vascular complications that often occur in patients with a

superior cavopulmonary anastomosis. For patients with a left or right heart complex that is functional but unable to support the entire cardiac output, options intermediate between total cavopulmonary connection (modified Fontan procedure) and complete biventricular repair may be preferable for definitive palliation. Several such intermediate procedures have been described,¹²⁻¹⁴ including complete intracardiac repair with supplementary bidirectional Glenn anastomosis, or "partial biventricular repair."¹⁵⁻²⁰

The principle of partial biventricular repair is to create a modified in-series circulation, with no left-right or right-left shunting, the systemic ventricle pumping a single cardiac output, and the pulmonary circulation receiving full cardiac output through contributions of both the pulmonary ventricle and the superior cavopulmonary connection. This approach allows the maintenance of equal pulmonary and systemic flows, and the bidirectional cavopulmonary shunt simultaneously reduces the volume load on the right ventricle. This allows a hypoplastic right heart complex to adequately handle the reduced preload. It also enables aggressive tricuspid valve repair in patients with Ebstein's anomaly without the risk of iatrogenic tricuspid stenosis.

Although bidirectional cavopulmonary anastomosis is widely used for patients with a variety of forms of complex congenital heart disease, there are several concerns with this procedure as a form of long-term palliation. These include progressive desaturation, as the proportion of flow to the upper body (and thus through the superior vena cava) decreases in relation to patient size, and pulmonary vascular changes, such as pulmonary arteriovenous fistulas,²⁷ aortopulmonary collateral arteries,²⁸ systemic venous collaterals,²⁹ and potentially poor pulmonary artery growth.³⁰ However, these potential complications might be reduced or eliminated by allowing antegrade flow through the main pulmonary artery, which provides pulsatility, increased absolute volume and flow rate, and hepatic venous blood to both lungs.

Indications for partial biventricular repair. Possible indications for partial biventricular repair can be classified in a number of different ways. Van Arsdel and colleagues¹⁸ divided patients into the following four groups: small physiologic right ventricle (group A), chronic right ventricular dysfunction (group B), facilitation of biventricular repair without hypoplasia or functional impairment of the pulmonary ventricle (group C), and acute right

ventricular dysfunction (group D).¹⁸ Patients in our series fell into two basic groups: those with borderline function of the right side of the heart (group I) and those in whom previous bidirectional Glenn anastomosis had been performed, usually on the basis of complicated anatomy, but for whom complete or partial biventricular repair was possible with additional complex procedures (group II). Other published series have generally included patients with a single lesion or surgical indication.^{15-17, 19, 20}

In our cohort of patients with borderline function of the right side of the heart, partial biventricular repair was indicated on the basis of a small right ventricle, functional compromise, or both. This category thus includes both groups A and B of the series of Van Arsdel and colleagues.¹⁸ The former category may include patients with pulmonary atresia and intact ventricular septum or other lesions with a hypoplastic right ventricle. The latter group may include such lesions as Ebstein's anomaly and isolated tricuspid stenosis without right ventricular hypoplasia. It is difficult to posit definitive size or functional guidelines for partial biventricular repair on the basis of our relatively limited experience and the available literature, especially in light of the diversity of lesions repaired. In the 1993 Congenital Heart Surgeons' Society multiinstitutional study² on neonatal management of pulmonary atresia with intact ventricular septum, the only risk factor militating against biventricular repair was small tricuspid valve Z value (continuous variable). The predicted prevalence of biventricular repair at 5 years was less than 30% for a Z value of -3 and less than 50% for a Z value of -2. According to this study, almost 50% of patients with a tricuspid valve Z value of -2 would be predicted to have a mixed (incompletely separated) circulation at the age of 5 years. Patients with a tricuspid valve diameter in this range may be the most suitable candidates for partial biventricular repair, as has been the case in our experience. We were able to perform partial biventricular repair in patients with tricuspid valve Z values as low as -5, but the need to create an atrial septectomy (in effect failure of the partial biventricular repair) in a patient with a tricuspid Z value of -4 suggests that such severe hypoplasia of the right side of the heart may not be consistently amenable to this intermediate form of definitive palliation.

For moderate and severe forms of Ebstein's anomaly, partial biventricular repair adds another dimension to the range of surgical approaches.

Decreasing right ventricular preload by performing a bidirectional Glenn anastomosis may lead to improved right ventricular and tricuspid valvular function, with or without tricuspid valvuloplasty. In most of our cases of Ebstein's anomaly, some form of tricuspid valvuloplasty or annuloplasty was performed, but two patients had improvement in tricuspid regurgitation from mild or moderate to trivial without any valve intervention, presumably as a result of improvement in coaptation caused by decreased right ventricular volume and tricuspid annular dilatation. For all patients with borderline function of the right side of the heart, the decision to proceed with partial biventricular repair need not be made before the operation. For patients who may be able to tolerate complete biventricular repair, this is attempted and the patient is then separated from bypass. If right atrial pressure is too high (more than 12 mm Hg or twice left atrial pressure) or tricuspid valvular function is inadequate according to transesophageal echocardiography, a bidirectional Glenn anastomosis is added. The bidirectional Glenn anastomosis can be performed without cardiopulmonary bypass, adding little risk to the operation.

Group II patients, who had previously undergone bidirectional Glenn anastomosis in preparation for an ultimate Fontan operation, had a range of anomalies. Among them were two patients with corrected transposition of the great arteries and pulmonary atresia, one with double-outlet left ventricle, and one with transposition of the great arteries and a straddling tricuspid valve, all of which were revised to partial biventricular repair with the use of fairly new surgical approaches. In the patients with corrected transposition of the great arteries, we performed a double-switch procedure, with a Rastelli procedure and a modified Mustard procedure, in which the atrial baffle extended around the orifice of the inferior vena cava but not of the superior cava. This approach, also referred to by the Toronto group for two of their group C patients,¹⁸ simplifies the Mustard component of a double-switch procedure, reduces bypass time, decreases atrial suture line, and may lead to a lower incidence of venoatrial obstruction than is the case with a standard full Mustard baffle. In our patient with double-outlet left ventricle, we transposed the pulmonary root into the right ventricle. Although this would probably have been adequate for complete biventricular repair, the bidirectional Glenn anastomosis was left in place. In the patient with transposition of the great arteries, hypoplastic right ventricle, and a straddling tricuspid

Fig. 2. Preoperative apical four-chamber echocardiogram from a group II patient with transposition of the great arteries, right ventricular hypoplasia, and straddling of the tricuspid valve (*arrow*). This patient underwent arterial switch and reimplantation of the straddling chordae on the right ventricular side of the septum 22 months after bidirectional Glenn anastomosis. Abbreviations are as in Fig. 1.

valve, an arterial switch was performed and the valvular anomaly was corrected by transecting the straddling chordae and reattaching them in the right ventricle, as we previously described.²⁰ The bidirectional Glenn anastomosis was also left in place in this patient because of the small size of the right ventricle (Fig. 2).

Conclusion. Partial biventricular repair, in which complete intracardiac repair and pulmonary outflow tract reconstruction (if necessary) are supplemented by a bidirectional superior cavopulmonary anastomosis, is a valuable concept in the management of a variety of forms of complex congenital heart disease. The widest and most reliable application of partial biventricular repair is likely to be for patients with hypoplasia or functional compromise of the right heart complex, or both.¹⁵⁻²⁰ There are other applications in which it may have an important role, however, such as facilitating biventricular repair for lesions such as corrected transposition of the great arteries or left superior vena cava to left atrium, or conversion to partial biventricular repair among patients with complex lesions who have previously undergone bidirectional Glenn anastomosis in single-ventricle staging. Limited long-term follow-up reported by other investigators suggests that this approach confers lasting benefits as a form of definitive palliation for pulmonary atresia with intact ventricular septum.¹⁷ Of particular interest in com-

paring this strategy with a modified Fontan procedure are exercise tolerance, rhythm status, and the effects on cerebral function of pulsatile and mildly elevated pressure in the superior vena cava. Long-term follow-up data will be necessary before the merits of this approach can be assessed with respect to other forms of intermediate palliation.¹¹⁻¹⁴ We are following the courses of these patients closely and plan to perform full functional evaluation, including the previously mentioned tests, in the future.

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Discussion

Dr. Hillel Laks (*Los Angeles, Calif.*). This is a fascinating and interesting area in cardiac surgery, the distinction between biventricular, univentricular, and what is described as "partial biventricular" or "one and a half ventricle" repair. Attempted biventricular repair for patients with hypoplastic right ventricle is associated with a high mortality rate, and another group showed that unless the right ventricle was within the 90% confidence limits of normal size, the mortality rate was extraordinarily high. In 1989 we described what we called the "partial biventricular" repair, which included leaving an adjustable atrial septal defect, snare controlled, that would allow as much as one third of the systemic venous return to cross from the right atrium to the left atrium and allow a smaller right ventricle to carry as much as two thirds of the venous return. In that same publication, we included a description of the bidirectional Glenn shunt for those ventricles that were between one third and one half normal size, which would correlate with Z values of tricuspid valve size down to -5 or -6. This allowed one to do these partial biventricular repairs with acceptable results.

In your series, which included six cases of Ebstein's anomaly, in which right ventricular size is generally normal, could some of these patients have undergone a biventricular repair with the adjustable atrial septal defect without the need for the Glenn shunt?

Dr. Reddy. I think that it is a matter of the surgeon's preference whether an adjustable atrial septal defect or a cavopulmonary shunt is used. I am pretty certain that these patients could have had adjustable atrial septal defect. The reason that we generally have gone on to do bidirectional cavopulmonary anastomoses in these patients is that with an adjustable atrial septal defect there is still some mixing, whereas there is none with the cavopulmonary shunt. Also, we were able to aggressively repair the tricuspid valve and try to eliminate all the tricuspid regurgitation by doing an aggressive annuloplasty, to the point at which a stenosis would have been created if one cardiac output is pumped through the tricuspid valve.

Dr. Laks. The advantage of the adjustable atrial septal defect is that as the right ventricle grows, one can go back and close it without, of course, needing bypass. My other question is related to the reconstruction of the right ventricle. If one does a bidirectional Glenn shunt, the

pulmonary artery pressure will usually rise to a mean of 12 to 15 or 16 mm Hg, and in the absence of a confident pulmonary valve the right ventricular end-diastolic pressure may rise to similar levels. If the atrial septal defect is closed, the right atrial pressure will then rise to that same level again, which would negate some of the advantages of partial biventricular repair. We have therefore incorporated as part of this operation the insertion of a pulmonary valve to lower the right ventricular end-diastolic pressure. Have you have had a similar experience? What have your postoperative right atrial pressures been, and have you begun to insert valves as part of the initial repair?

Dr. Reddy. We have not inserted any valves as a part of the repair as yet, but I completely agree with you that pulmonary insufficiency is a major concern in the cases in which you have to do a right ventricular outflow tract procedure. We have generally tried to do a careful valvotomy if necessary and have tried to place a small transannular patch and limit the pulmonary insufficiency by not resecting too much of the right ventricle. However, some of these patients may need pulmonary valve insertion during follow-up.

Dr. Laks. Although there are advantages to the bidirectional Glenn shunt, there is also something of a down side. If there indeed is differentiation between the superior vena caval and inferior vena caval pressures, venous collaterals may develop to divert blood from the higher pressure superior vena cava down to the lower pressure inferior vena cava, as we have seen in long-term follow-up of some of these patients. This has yielded a semicircular circulation, in which blood is being diverted down and recirculating. We also had one case, and when we reviewed the literature we found another, in which the highly pulsatile flow from the entire right ventricular cardiac output going to the pulmonary arteries resulted in an aneurysm formation in the superior vena cava. In our case we had to go back and reduce this and put in a pulmonary valve. Have you seen this complication? In view of that concern and the fact that some of these patients have severely pulsatile neck veins when there is a large pulse pressure, I think that one should emphasize trying to go to a biventricular repair whenever possible and to avoid the Glenn anastomosis, which can be done with the adjustable atrial septal defect.

Dr. Reddy. We have not seen that complication in our experience, but I certainly agree that it would be important to follow up these patients to observe for development of any of these complications. I think that with long-term follow-up, we will be able to clearly define the groups that would benefit from a cavopulmonary shunt and those that would benefit from adjustable atrial septal defect.