Outcomes of 1½- or 2-ventricle conversion for patients initially treated with single-ventricle palliation

Takaya Hoashi, MD, Edward L. Bove, MD, Eric J. Devaney, MD, Jennifer C. Hirsch, MD, and Richard G. Ohye, MD

Objective: As outcomes for the Fontan procedure have improved, it has become more difficult to select between a single-ventricle repair or biventricular repair for patients with complex anatomy and 2 ventricles. However, late complications after the Fontan procedure remain a concern. Our strategy, which has favored an aggressive preferential approach for biventricular repair in these patients, has also been applied to patients initially treated on a single-ventricle track elsewhere.

Methods: Nine patients (4 male patients) who had previously undergone the Fontan procedure (n = 3) or bidirectional cavopulmonary shunting (n = 6) with intent for a later Fontan procedure were referred to our center for complex 1½- or 2-ventricle repair over the last 10 years. Indications for conversion in these patients were protein-losing enteropathy (n = 2), pulmonary arteriovenous malformation (n = 1), and preference for biventricular anatomy (n = 6). The conversion mainly consisted of takedown of the Fontan procedure or bidirectional cavopulmonary shunt connection, reconstruction of 1 or both of venae cavae, creation of an intraventricular pathway for left ventricular output, and placement of a right ventricle–pulmonary artery conduit (Rastelli-type operation).

Results: Five patients underwent 1½-ventricle repair, and 4 had complete biventricular repair. Median cardiopulmonary bypass and aortic crossclamp times were 202 minutes (range, 169–352 minutes) and 129 minutes (range, 100–168 minutes), respectively. There were 2 early deaths and 1 late death. At a median follow-up of 27 months (range, 3.3–99.8 months), all survivors are in New York Heart Association class I.

Conclusions: Patients initially treated with intent to perform single-ventricle palliation can be converted to 1½- or 2-ventricle physiology with acceptable outcomes. (J Thorac Cardiovasc Surg 2011;141:419-24)

CHD

The Fontan procedure has undergone continual refinement over the past 2 decades. Technical modifications have resulted in excellent early and midterm results.[1-3] These outcomes have expanded the use of the Fontan procedures not only for anatomically, functionally, or both single-ventricle (SV) patients but also for those thought to have untreatable complex intracardiac anatomy with 2 relatively adequately sized ventricles.

Late complications of the Fontan circulation remain management challenges, such as protein-losing enteropathy (PLE), pulmonary arteriovenous malformations (PAVMs), plastic bronchitis, arrhythmia, thromboembolism, and ventricular failure.[4-7] In addition, the life expectancy and exercise capacity of patients undergoing a Fontan procedure are also of concern.[8-11]

Despite these concerns over the long-term outcomes of the Fontan procedure, certain types of complex anatomy might be too risky to establish biventricular circulation, even in the presence of 2 adequate ventricles.[12-15] In particular, the challenge of ventricular septation and intraventricular rerouting might cause some surgeons to hesitate to pursue a biventricular approach because of the technical difficulty, the risk of surgical heart block, left ventricular outflow tract obstruction (LVOTO) and iatrogenic atrioventricular valve regurgitation, and the need for a right ventricle (RV)–pulmonary artery (PA) conduit. These disadvantages of complex biventricular repair (BVR) must be balanced against the late complications of the Fontan procedure.

In an attempt to avoid late Fontan complications, an alternative procedure, 1½-ventricle repair, was developed[16] for patients with an insufficiently sized pulmonary ventricle. The potential benefit of this operation is the achievement of a physiological separation of the pulmonary and systemic circulations with the maintenance of pulsatile blood flow in the PAs by diverting the superior vena caval blood to return directly into the PAs through a bidirectional cavopulmonary shunt (BCPS). This technique can result in normal peripheral oxygen saturation while reducing the volume load on the pulmonary ventricle.[17]

In general, our strategy has favored an aggressive preferential approach to BVR. This approach has also been applied to patients initially treated on an SV track elsewhere. This report reviews our surgical experience with these challenging patients.

From the Division of Pediatric Cardiovascular Surgery, Department of Surgery, University of Michigan Medical School, Ann Arbor, Mich.

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Address for reprints: Takaya Hoashi, MD, 5144 Cardiovascular Center/SPC 5864, 1500 East Medical Center Dr, Ann Arbor, MI 48109-5864 (E-mail: thoashi@surg1.med.osaka-u.ac.jp).

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Abbreviations and Acronyms

BCPS = bidirectional cavopulmonary shunt
BVR = biventricular repair
CAVSD = complete atrioventricular septal defect
DORV = double-outlet right ventricle
ECC = extracardiac conduit
LV = left ventricle
LVOTO = left ventricular outflow tract obstruction
PA = pulmonary artery
PAVM = pulmonary arteriovenous malformation
PLE = protein-losing enteropathy
PS = pulmonary stenosis
RV = right ventricle
SV = single ventricle
TGA = transposition of the great arteries
VSD = ventricular septal defect

MATERIALS AND METHODS

Patients

From 2000 to 2009, a total of 9 patients (4 male patients) who had previously undergone a Fontan procedure or BCPS with intent for a later Fontan procedure were referred to C.S. Mott Children's Hospital, University of Michigan Health Systems, for further surgical treatment (Table 1). This study was approved by the University of Michigan Medical School Institutional Review Board for Human Subject Research.

The primary diagnosis was l-transposition of the great arteries (TGA) with ventricular septal defect (VSD) and pulmonary atresia (n = 3), double-outlet right ventricle (DORV) with pulmonary stenosis (PS; n = 3), d-TGA with VSD and PS or pulmonary atresia (n = 2), and LVOTO with VSD (n = 1).

These patients underwent a variety of previous interventions, which are summarized in Table 1. The patients had been deemed Fontan candidates because of the existence of an inadequately sized ventricle (n = 4), unsuitable atrial anatomy for atrial switch (n = 3), unsuitable VSD for creating intraventricular rerouting (n = 2), and limited anterior chest space for insertion of an RV–PA conduit (n = 1). All of the patients had undergone 2 or more interventions. The last operation before presentation at our institution was a BCPS with intent for a later Fontan procedure in 6 patients and a Fontan procedure in 3 patients. Eight of the 9 patients had preoperative cardiac catheterization data (Table 2).

Surgical Techniques

The operation was performed with standard bicaval cardiopulmonary bypass. The conversion involved the reconstruction of the systemic venous system by means of takedown of the Fontan or BCPS connections and reconstruction of 1 or both of the venae cavae. Left and right ventricular egress was provided by the creation of an intraventricular pathway from the VSD to the aorta to provide left ventricular (LV) outflow and the placement of an RV–PA conduit (Rastelli-type operation). The VSD was enlarged anterosuperiorly in 4 patients at the discretion of the operating surgeon. The atrial septal defect was completely closed in 6 patients, whereas 3 patients with a hypoplastic RV underwent partial atrial septal defect closure with a 2.8-mm fenestrated polytetrafluoroethylene patch. Delayed sternal closure was applied in 5 patients.

Specific techniques were applied according to individual anatomy. Three patients with a relatively hypoplastic RV underwent RV enlargement by means of division or resection of RV trabecular myocardium.[18] For the 2 patients with bilateral BCPSs, the left superior venae cavae were anastomosed end-to-side to the right superior venae cavae at the $1^1/_2$-ventricle repair in an effort to minimize caval hypertension theoretically exacerbated by means of pulsatile blood flow from an RV–PA conduit directly up the superior vena cava.

Patient 8 (DORV, pulmonary atresia, and complete atrioventricular septal defect [CAVSD]) required a unique reconstruction. She had previously undergone an extracardiac conduit (ECC) Fontan procedure with bilateral BCPS. For the systemic venous reconstruction, both PAs were transected distal to the BCPS anastomoses, leaving the central portion of the PA to serve as the neoinnominate vein. A short segment of the distal ECC was then used as the superior vena cava to the right atrium. The inferior vena cava was reattached to the right atrium with a short segment of the proximal ECC. The DORV and CAVSD were repaired with closure of the VSD to the aorta. A bifurcated pulmonary allograft was used to create an RV to both branch PAs continuity. The proximal end of allograft was anastomosed to RV anterior wall. One of the distal end was anastomosed to the left PA directly, then a 12-mm ringed expanded polytetrafluoroethylene graft was interposed between the right PA and the other distal end of the allograft.

RESULTS

Indications for conversion in these patients were PLE (n = 2), PAVMs (n = 1), and preference for biventricular anatomy (n = 6, Table 1). Patients 7 and 9, who had a Fontan circulation, had persistent PLE. Patient 8, who had undergone an ECC Fontan procedure after the Kawashima procedure, also had extensive left-sided PAVMs and severe cyanosis.

Six patients underwent $1^1/_2$-ventricle repair (one of which was the staging procedure for the later complete BVR), and 3 had a primary complete BVR (Table 3). In addition to the Rastelli-type repairs, the concomitant procedures were atrial switch for l-TGA in 3 patients, inferior vena caval reconstruction for a takedown of Fontan connection in 2 patients, superior vena caval reconstruction for complete BVR in 3 patients, and VSD enlargement in 4 patients. The median cardiopulmonary bypass and aortic crossclamp times were 202 minutes (range, 169–352 minutes) and 129 minutes (range, 100 –168 minutes), respectively.

There were 2 early deaths (Table 3). Patient 5 (l-TGA, pulmonary atresia, and VSD) with moderate-to-severe mitral regurgitation underwent $1^1/_2$-ventricle repair consisting of a Senning procedure combined with Rastelli-type operation and mitral valvuloplasty. Her chest was able to be closed 4 days postoperatively. However, she had junctional ectopic tachycardia with profound hypotension 2 days later and was unable to be resuscitated. Patient 6 (d-TGA, PS, and single coronary artery) had diminished function of both ventricles before the operation. He underwent biventricular conversion consisting of BCPS division, VSD enlargement, and a Rastelli-type operation. At weaning from CPB, his ventricular function was severely decreased, and he required extracorporeal life support. An echocardiogram at that time showed mild-to-moderate global LV dysfunction without segmental wall motion abnormality and was otherwise unremarkable.

TABLE 1. Summary of preoperative patients' characteristics

Patient no.	Sex	Diagnosis (factors believed to favor SV palliation)	Previous interventions (age)	Indication for conversion
1	F	l-TGA, pulmonary atresia, VSD (left juxtaposed atrial appendages)	BTS, PA augmentation (6 d) Left PA unifocalization, central shunt (6 mo) BCPS (13 mo)	Preference for biventricular anatomy
2	M	DORV, PS, VSD, hypoplastic arch (believed to have too limited space for conduit insertion)	BTS, arch repair, modified DKS (12 d) Balloon aortoplasty (9 mo) BCPS (15 mo)	Preference for biventricular anatomy
3	F	DORV, PS, VSD, dextrocardia (hypoplastic LV)	BTS BCPS (9 mo)	Preference for biventricular anatomy
4	F	l-TGA, pulmonary atresia, VSD (situs inversus, levocardia)	BTS BCPS (6 mo)	Preference for biventricular anatomy
5	F	l-TGA, pulmonary atresia, VSD (dextrocardia)	BTS (8 d) BCPS (15 mo)	Preference for biventricular anatomy
6	M	d-TGA, PS	BCPS (15 mo)	Preference for biventricular anatomy (restrictive VSD)
7	M	d-TGA, pulmonary atresia (remote VSD, hypoplastic RV)	Right BTS (1 d) Left BTS (7 mo) Bilateral BCPS, PA augmentation (18 mo) ECC (6 y)	Protein-losing enteropathy
8	F	DORV, PS, CAVSD (hypoplastic RV, left isomerism)	Kawashima, main PA ligation (6 mo) Hepatic vein to PA connection (ECC) (3.5 y)	Severe pulmonary arteriovenous malformations
9	M	LVOTO, VSD (hypoplastic RV)	BTS, modified DKS (age?) Fontan procedure (age?)	Protein-losing enteropathy

SV, Single ventricle; *TGA*, transposition of the great arteries; *VSD*, ventricular septal defect; *BTS*, Blalock–Taussig shunt; *PA*, pulmonary artery; *BCPS*, bidirectional cavopulmonary shunt; *DORV*, double-outlet of right ventricle; *PS*, pulmonary stenosis; *DKS*, Damus–Kaye–Stansel anastomosis; *LV*, left ventricle; *RV*, right ventricle; *ECC*, extracardiac conduit; *CAVSD*, complete atrioventricular septal defect; *LVOTO*, left ventricular outflow tract obstruction.

Because the patient could not be weaned from support, cardiac catheterization was performed 2 days later to assess the coronary anatomy, which detected a tight narrowing in the proximal left anterior descending coronary artery. This stenosis was repaired; however, ventricular function did not recover.

Late Outcomes in Successfully Repaired Patients

Median follow-up was 27 months (range, 3.3–99.8 months). There was 1 late death 1.5 years after surgical intervention caused by sepsis associated with unresolved PLE. All survivors have normal peripheral oxygen saturations. Quantitatively, RV function has remained normal in all late survivors, and although LV function has deteriorated in 1 patient, all patients remain in New York Heart Association functional class I. Further interventions have been required in 3 patients: device closure of residual septal defects in 1 patient, Senning pathway revision and RV–PA conduit replacement in 1 patient, and RV–PA conduit replacement, pacemaker implantation for sick sinus

TABLE 2. Preoperative catheter examination

Patient no.	BSA (m^2)	LVESP (mm Hg)	LVEDP (mm Hg)	RVESP (mm Hg)	RVEDP (mm Hg)	Qp/Qs	PVR ($u \cdot m^2$)	CI ($L/min \cdot m^2$)
1	0.49	98	8	98	8	0.56	2.0	4.1
2	0.59	86	10			0.57	1.0	5
3	0.70	130	8	124	11	0.50	1.6	3.4
4	0.47	95	12			0.67		
5	Did not undergo preoperative catheterization							
6	0.52	86	2	90	4	0.86	1.3	8.9
7	1.00	95	10	90	6	1.00		5.1
8	0.70	82	10			1.00		
9	0.86	115	14	94	10	0.80	3.2	3.1

BSA, Body surface area; *LVESP*, left ventricular end-systolic pressure; *LVEDP*, left ventricular end-diastolic pressure; *RVESP*, right ventricular end-systolic pressure; *RVEDP*, right ventricular end-diastolic pressure; *PVR*, pulmonary vascular resistance; *CI*, cardiac index.

TABLE 3. Summary of patients undergoing $1^1/_2$- or 2-ventricle conversion

Patient no.	Age (y)	Weight (kg)	Conversion procedure	Repair	CPB time (min)	AXC time (min)	Outcome (cause of death)
1	2.4	11	Rastelli, Mustard	1.5	169	100	Alive
2	3.9	14	Rastelli	1.5	195	131	Alive
3	7.8	17	Rastelli, VSD enlargement, mVSD closure	1.5	200	110	Alive
4	2	12	Rastelli, Senning, rt BCPS division, VSD enlargement	1.5 (→2)	202	129	Alive
5	5.7	23	Rastelli, Senning, mitral valve repair	1.5	253	142	Early death
6	2.3	11	Rastelli, BCPS division, VSD enlargement	2	202	109	Early death
7	9.0	31	Rastelli, takedown Fontan, RV sinus myectomy	1.5	352	168	Alive, PLE improved
8	5.3	17	CAVSD repair, Rastelli, takedown Fontan, RV sinus myectomy	2	302	145	Alive, PAVM resolved
9	7.4	26	Rastelli, takedown Fontan, VSD enlargement, RV sinus myectomy	2	196	128	Late death, PLE not improved

CPB, Cardiopulmonary bypass; *AXC*, aortic crossclamp; *VSD*, ventricular septal defect; *mVSD*, muscular VSD; *BCPS*, bidirectional cavopulmonary shunt; *RV*, right ventricular; PLE, protein-losing enteropathy; *CAVSD*, complete atrioventricular septal defect; *PAVM*, pulmonary arteriovenous malformation.

syndrome, and coil embolization of collateral vessels in 1 patient. Four patients have had follow-up cardiac catheterization (Table 4).

DISCUSSION

Our center has pursued a policy of BVR for the majority of patients presenting at birth with 2 reasonably sized functional ventricles, even in the presence of complex intraventricular anatomy. It is our belief that BVR will result in a better long-term functional outcome for these patients. Ohuchi and colleagues[19] compared the long-term cardiopulmonary responses to exercise among 14 patients undergoing the Fontan procedure and 13 patients undergoing BVR for double-inlet left ventricle. They found that the BVR group showed superior exercise capacity and suggested that this finding would relate to a better long-term prognosis.

We have also applied this strategy to patients initially palliated with intent to perform SV palliation at other institutions. Results of the conversion of patients undergoing SV palliation to $1^1/_2$- or 2-ventricle physiology are good. However, as with all complex operations in challenging patients, careful consideration of the risks and benefits must be weighed. The candidates for biventricular conversion can be divided into 2 categories. The first group is patients requiring BVR for significant complications of their Fontan physiology or who are partway through their staged palliation and deemed to be poor candidates for a Fontan procedure. The second group is comprised of those that are doing well with their Fontan procedure but are undergoing conversion for the long-term benefits of normal biventricular physiology.

All 3 of the patients with a completed Fontan circulation were in the former group, with PLE (n = 2) or PAVMs with severe cyanosis (n = 1). For these patients, the PAVMs resolved after biventricular conversion, and the PLE was improved in 1 of 2 patients after $1^1/_2$-ventricle conversion. Other options for late Fontan complications have been described, such as revision of an atriopulmonary Fontan procedure to an ECC.[20,21] However, although these revisions are often successful in resolving issues such as arrhythmia, they

TABLE 4. Postoperative cardiac catheter examination of patients undergoing $1^1/_2$- or 2-ventricle conversion

Patient no.	BSA (m^2)	Interval postoperative (mo)	LVESP (mm Hg)	LVEDP (mm Hg)	mPAP (mm Hg)	RVESP (mm Hg)	RVEDP (mm Hg)	CI (L/min · m^2)
1	0.60	7	115	8	14	50	9	5.1
2	No data (has not undergone a postoperative catheterization)							
3	No data (has not undergone a postoperative catheterization)							
4		6	95	8	32	50	15	
5	No data (early death)							
6	No data (early death)							
7	No data (has not undergone a postoperative catheterization)							
8	0.80	16	88	12	14	49	8	4.3
9	1.03	4	87	8	15	35	9	3.7

BSA, Body surface area; *LVESP*, left ventricular end-systolic pressure; *LVEDP*, left ventricular end-diastolic pressure; *mPAP*, mean pulmonary artery pressure; *RVESP*, right ventricular end-systolic pressure; *RVEDP*, right ventricular end-diastolic pressure; *CI*, cardiac index.

do not change the underlying SV physiology and might not address complications such as PLE, which are thought to be related to the Fontan circulation. In this study 2 patients survived, whereas 1 ultimately succumbed to unresolved PLE. Unfortunately, because even heart transplantation does not always halt the progression of PLE, attempts at BVR in this population with severe life-threatening complications of Fontan palliation would seem indicated.

The latter group, those patients on an SV pathway with satisfactory hemodynamics presenting for conversion to BVR, present a more difficult decision-making process. Of the 6 patients in this category, there were 2 early deaths. One must weigh the potential long-term complications of the Fontan procedure against the operative risk of BVR, the need for reoperation, and the benefits of $1\frac{1}{2}$- or 2-ventricle physiology.

Determining whether there were preoperative characteristics that differentiated survivors from nonsurvivors would be important. However, objective morphological criteria, such as ventricular volumes determined by means of cardiac catheterization, were not recorded, and these subjects predated our routine use of cardiac magnetic resonance imaging. Other objective data, such as routine catheterization hemodynamics, were not different between groups, and the sample size limits the ability to detect subtle differences. However, with regard to the 2 early deaths, these patients differed from the remaining 7 patients in that both had a preoperative functional problem in the left heart; patient 5 had severe mitral valve regurgitation, and patient 6 had diminished LV function. Although not a statistically significant finding, it might be that patients with preexisting left heart issues would be better served by cardiac transplantation or continuing expectant management in the case of the asymptomatic patient.

Another concern are the long-term outcomes of the 4 patients with $1\frac{1}{2}$-ventricle physiology. Whereas $1\frac{1}{2}$-ventricle repair has a more physiological circulation than the Fontan procedure and might avoid late complications associated with Fontan circulation, Numata and associates[22] demonstrated the long-term exercise capacity in patients with pulmonary atresia with intact ventricular septum, which showed that $1\frac{1}{2}$-ventricle physiology showed late exercise intolerance, as seen in patients undergoing the classic Fontan procedure, and was inferior to the biventricular physiology. Also, pulsatile flow in the superior vena cava can also lead to complications.[17] Kim and coworkers[23] reviewed 114 patients who underwent $1\frac{1}{2}$-ventricle repair. Serious superior vena cava–related complications occurred in 3 (2.6%) patients, and mild complications occurred in 5 (4.4%) patients.

A potential role for $1\frac{1}{2}$-ventricle repair is as an interstage procedure before complete BVR. Fontan physiology results in a lower ventricular reserve capacity, or potential compliance under increased loading conditions, than normal biventricular physiology primarily because of reduced systemic ventricular preload.[24] The sudden increase in ventricular preload associated with primary complete biventricular conversion might excessively stress the systemic ventricular, and thus this staging approach might lead to improved results.

Several reasons were cited for not performing an initial BVR in the patients in this report. Three patients had been judged not to be candidates for BVR because of hypoplasia of the RV. However, these patients were able to undergo RV sinus myectomy associated with the conversion operation, and their RV function has been preserved. We and other authors consider that BVR, with or without a BCPS to reduce RV volume load, is possible even if the RV is hypoplastic but there is an adequate sized right-sided atrioventricular valve.[13]

The other indications for SV palliation in this patient group were l-TGA, CAVSD with DORV, DORV with remote VSD, and heterotaxy syndrome with complex ventricular and atrial anatomy. All these anomalies have in common the need for complex intraventricular rerouting of the LV output through the VSD to the semiluminar valve. The complex intracardiac tunnels, which often require VSD enlargement, place the patient at risk for surgical heart block or LVOTO. In this series no patient had heart block, and no long-term survivor has required a reoperation for LVOTO. Thus with enlargement of the VSD in an anteriosuperior direction and careful construction of the intraventricular tunnel, these defects can be repaired without the need for SV palliation.

Whether the patients in this series would have been better served by an initial BVR, thus avoiding the need for complex SV palliation takedown in addition to the challenges of the BVR, is unknown. All of the patients in this series would have undergone BVR at our institution, either primarily (patients 2, 3, and 6–9) or after an interim systemic–PA shunt (double-switch candidates, patients 1, 4, and 5). The choice between a complex BVR and a relatively straightforward SV repair is a difficult one. However, the results of a policy of BVR for patients such as patient 9 with LVOTO and VSD have been published from our center with a 5-year actuarial survival of 78%, which exceeds most published studies for 5-year Fontan survival.[25]

In summary, carefully selected patients initially treated with intent to perform an SV palliation can in some instances be converted to $1\frac{1}{2}$- or 2-ventricle physiology with acceptable outcomes. Although it can be still difficult to select between a complex, potentially high early mortality primary BVR and an SV palliation with the potential for higher late morbidity, a patient with adequately sized atrioventricular valves and adequate ventricular size and function would be a good candidate for initial BVR. Although it is not possible to know what the outcome of a plan for BVR from the outset would have been, these data would suggest that the majority of these patients could have undergone BVR rather than initial SV palliation, thus avoiding the need for complex takedown of previous operations in addition to the challenging reconstruction.

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