Effectiveness of Bidirectional Glenn Shunt Placement for Palliation in Complex Congenitally Corrected Transposed Great Arteries

Surgery for complex congenitally corrected transposed great arteries is one of the greatest challenges in cardiovascular surgery. We report our experience with bidirectional Glenn shunt placement as a palliative procedure for complex congenitally corrected transposition.

We retrospectively identified 50 consecutive patients who had been diagnosed with congenitally corrected transposition accompanied by left ventricular outflow tract obstruction and ventricular septal defect and who had then undergone palliative bidirectional Glenn shunt placement at our institution from January 2005 through December 2014. Patients were divided into 3 groups according to subsequent surgeries: Fontan completion (total cavopulmonary connection, 13 patients) (group 1), anatomic repair (hemi-Mustard and Rastelli procedures without Glenn takedown, 11 patients) (group 2), and prolonged palliation (no further surgery, 26 patients) (group 3).

After shunt placement, no patient died or had ventricular dysfunction. Overall, mean oxygen saturation increased significantly from 79.5% ± 13.5% preoperatively to 94.1% ± 7.3% (P < 0.001). The median time from shunt placement to Fontan completion and anatomic repair, respectively, was 2.1 years (range, 1.6–5.2 yr) and 1.1 years (range, 0.6–2.4 yr). Only 2 late deaths occurred, both in group 1. In group 3, time from shunt placement to latest follow-up was 4.5 years (range, 2.3–8 yr). At latest follow-up, mean oxygen saturation was 91.6% ± 10.3%, and no patients had impaired ventricular function.

Bidirectional Glenn shunt placement as an optional palliative procedure for complex congenitally corrected transposition has favorable outcomes. Later, patients can feasibly be treated by Fontan completion or anatomic repair. Use of a bidirectional Glenn shunt for open-ended palliation is also acceptable.

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Clinical Investigation

Kai Ma, MD, PhD
Lei Qi, MD, PhD
Zhongdong Hua, MD
Keming Yang, MD
Hao Zhang, MD, PhD
Kai Ma, MD, PhD

From: Pediatric Cardiac Surgery Center, State Key Laboratory of Cardiovascular Disease, National Center for Cardiovascular Disease, Fuwai Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100037, People’s Republic of China

Address for reprints: Shoujun Li, MD, Department of Cardiac Surgery, Fuwai Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, 167 Beilishi Rd., Xicheng District, Beijing 100037, PRC

E-mail: drlshoujunlsj@yahoo.com

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Patients and Methods

We retrospectively identified 50 consecutive patients who had been diagnosed with CCTGA accompanied by LVOTO and VSD and had then undergone palliative BDG at our institution from January 2005 through December 2014. The patients were divided into 3 groups according to subsequent surgery (Fig. 1): Fontan completion (total cavopulmonary connection [TCPC], 13 patients) (group 1); anatomic repair (hemi-Mustard and Rastelli procedures without Glenn takedown, 11 patients) (group 2); and prolonged palliation (no further surgery, 26 patients) (group 3). Patients who underwent hemi-Mustard and Rastelli procedures concomitantly with BDG were excluded from this cohort. This study was approved by the Ethics Committee at Fuwai Hospital, which waived the need for patient consent for publishing follow-up data on these patients. Complete follow-up information was obtained for all patients.

Indications and Timing of Bidirectional Glenn Shunt Placement

Patients underwent BDG when they presented with cyanosis or tricuspid regurgitation (TR). In groups 1 and 3, all patients had an anatomic contraindication to biventricular repair (for example, unbalanced ventricles, remote VSD, or substantial chordae straddling). In group 2, all patients had cardiac malposition; of those, 7 underwent BDG as a first step toward single ventricular repair at other institutions, but were then selected for 1½ ventricular repair at our center. Another 4 patients with severe dysfunction of other organs underwent first-stage BDG before undergoing hemi-Mustard and Rastelli procedures because cardiopulmonary bypass would have been unsafe at that time.

Surgical Choice after Bidirectional Glenn Shunt Placement

Patients were selected for subsequent surgical treatment according to anatomic or socioeconomic factors. In group 1, all patients had an anatomic contraindication to septation and underwent TCPC as the ending stage of univentricular repair. In group 2, all patients had a potentially septatable anatomy despite the presence of positional anomalies that substantially increased the complexity of the Senning procedure. Therefore, 1½ ventricular repair with a simpler atrial approach (hemi-Mustard) was performed in group 2. In group 3, all patients had an anatomic contraindication to septation, so Fontan completions were planned. However, none underwent the second-stage TCPC, mainly for reasons that included low medical compliance and financial constraints.

Surgical Techniques

Bidirectional Glenn Shunt Placement. Before BDG, major aortopulmonary collateral arteries were occluded and ligated, and previously placed systemic-to-pulmonary shunts were removed. For the BDG anastomosis, the superior vena cava (SVC) was anastomosed to the pulmonary artery (PA) branch in an end-to-side fashion. Concomitant repair of total anomalous pulmonary venous connection (TAPVC) and pulmonary arterioplasty were performed when indicated. Cardiopulmonary bypass was instituted when these associated procedures were performed. Concomitant tricuspid repair was performed only when an associated procedure necessitated aortic cross-clamping. Pulmonary artery pressure was monitored with transducers through the main PA before Glenn anastomosis and through the SVC after Glenn anastomosis. Ligation or banding of the main PA was avoided when pulmonary stenosis or atresia was present. The main PA was banded with a polytetrafluoroethylene strip to achieve a mean PA pressure or central venous pressure $< 16$ mmHg. If the central venous pressure was lower than 16 mmHg, the azygos vein was ligated. In patients younger than 6 months, a modified Blalock-Taussig shunt operation was performed before BDG to relieve cyanosis and promote the growth of branch PAs.

1½ Ventricular Repair. Hemi-Mustard and Rastelli procedures were performed as the second-stage surgery to achieve 1½ ventricular repair. The procedures were done with patients under cardiopulmonary bypass and moderate hypothermia. After aortic cross-clamping and administration of cold crystalloid cardioplegia solution, the hemi-Mustard procedure was performed first. The atrial septum was completely excised, the coronary sinus was unroofed, and a bovine pericardial baffle was constructed to divert blood flow from the inferior vena cava and coronary sinus (including the left SVC if needed) to the tricuspid valve and RV. For the Rastelli procedure, a Dacron intraventricular baffle connecting the LV to the aorta was fashioned through a right ventriculotomy. Right ventricular–PA continuity was then established by constructing a bovine jugular vein conduit. Temporary epicardial pacemaker leads were regularly placed.
Total Cavopulmonary Connection. Either a lateral tunnel or extracardiac conduit technique (with or without cardiopulmonary bypass, depending on the surgeon’s preference) was used to create a cavopulmonary connection in patients with anatomic restrictions precluding septation (for example, unrouteable VSD, severe chordae straddling, or unusual coronary artery pattern and unbalanced ventricle). The decision whether to fenestrate was made intraoperatively according to guidelines that included TCPC pressure >18 mmHg or more than mild atroventricular valvular regurgitation. The technique used for TCPC was similar to that described by Talwar and associates.6

Data Collection and Definitions
Demographic and clinical data were obtained from our local institutional database. Every patient with CCTGA had undergone angiography and catheterization before surgery. Echocardiographic analysis was used to characterize systolic ventricular function and the severity of TR. A single cardiologist reviewed all previous echocardiograms and performed independent measurements. In this study, So2 always referred to pulse oximeter oxygen saturation. Postoperative pleural effusion lasting longer than 10 days was considered prolonged. The degree of valvular regurgitation was graded from 0 to 4 according to the Sellers classification and then expressed numerically as follows: 0, absent or trivial; 1, mild; 2, moderate; and 3, severe.7 Valvular regurgitation was considered significant when documented as moderate or severe, and ventricular dysfunction was defined as an ejection fraction (EF) <0.50. New York Heart Association (NYHA) functional classification was based on subjective symptoms. Thus, we documented the time at which a patient reported related symptoms as the time at which NYHA class III/IV function was achieved.

Statistical Analysis
Results are presented as mean ± SD for continuous variables with normal distribution, as median and range for variables with nonnormal distribution, and as number and percentage for categorical variables. The Student t test, analysis of variance, χ2 test, or Fisher exact test was applied in univariate analysis. The Bonferroni test was used in pairwise comparisons. Multivariate analysis could not be performed because the ratio of events per variable was too small. P <0.05 was considered statistically significant. Data were analyzed with SPSS version 17.0 for Windows (SPSS, an IBM company).

Results
From January 2005 through December 2014, 123 patients with a diagnosis of CCTGA accompanied by LVOTO and VSD underwent surgery at our center. Of those, 50 patients underwent BDG as a palliative procedure. All 50 patients had substantial LVOTO and nonrestrictive VSD; 34 patients (68%) also presented with cardiac positional anomalies. Five patients had pulmonary atresia, and the other 45 had pulmonary stenosis with a trans-LVOTO pressure gradient >70 mmHg. Table 1 documents the incidence of associated anomalies, preoperative TR, and previous modified Blalock-Taussig shunt operation.

The patients in group 3 were significantly older at the time of BDG than those in groups 1 and 2, which by comparison were similar in age. The mean weight at BDG was significantly higher in group 3 than in groups 1 and 2. Mesocardia was more frequent in group 2 than in groups 1 and 3. However, there was no statistically significant difference between the 3 groups in mean preoperative So2, mean PA pressure, mean Nakata index, or mean systemic ventricular EF or in the incidence of TR, previous modified Blalock-Taussig shunt operation, concomitant major aortopulmonary collateral arteries, heterotaxy syndrome, or TAPVC.

Perioperative Outcomes of Bidirectional Glenn Shunt Placement
At the time of BDG, 5 patients underwent TAPVC repairs; 2, tricuspid repairs; and 4, major aortopulmonary collateral artery occlusions. No patients needed pulmonary arterioplasty. The mean cardiopulmonary bypass and aortic cross-clamp times for the patients who underwent TAPVC were 83.6 ± 28.2 and 52.9 ± 17.6 min. No patients died, and none required Glenn take-down after BDG. Three patients had prolonged pleural effusions lasting for 21, 14, and 17 days, respectively. Immediately after BDG, the mean So2 increased significantly, from 79.5% ± 13.5% preoperatively to 94.1% ± 7.3% (P <0.001). Neither the mean PA pressure nor the mean systemic ventricular EF changed significantly. There was no statistically significant difference between groups in postoperative mean So2, mean PA pressure, mean systemic ventricular EF, or prolonged pleural effusion. Tricuspid regurgitation improved, although not significantly (Fig. 2A).

Outcomes of Fontan Completion (Group 1)
The median time from BDG to Fontan completion in group 1 was 2.1 years (range, 1.6–5.2 yr). All 15 patients in this group underwent TCPC (10 lateral tunnel and 3 extracardiac conduit) as the Fontan procedure. Five patients (38.5%) underwent fenestration, and 4 patients (30.8%) underwent concomitant tricuspid repair.

Early after TCPC, no patients died, and none had ventricular dysfunction. The mean So2 increased but not significantly from 85.3% ± 13.4% to 93.6% ± 15.7%. During the 4.3-year follow-up period (range, 1.5–6.2 yr), 2 late deaths (15.4%) occurred. One patient had refractory low cardiac output syndrome 19 months after TCPC and died one month later; the
other had a brainstem embolism and died 5 months after TCPC. Among the 11 survivors, one patient experienced protein-losing enteropathy (9.1%), 2 patients were in NYHA class III/IV (18.2%), and 2 patients had substantial atrioventricular regurgitation (18.2%). However, neither tricuspid reintervention nor transplantation was necessary. Figure 2B shows dynamic changes in TR, although the changes were not significant.

### Outcomes of Anatomic Repair (Group 2)

The median time from BDG to 1½ ventricular repair in group 2 was 1.1 years (range, 0.6–2.4 yr). The median follow-up time was 3.4 years (range, 1.2–4.5 yr). At the time of 1½ ventricular repair, 2 patients underwent tricuspid repairs; 2, mitral repairs; and 1, permanent pacemaker implantation. The mean So2 increased from 83.6% ± 10.9% to 96.5% ± 4.2% (P=0.006). The mean cardiopulmonary bypass and aortic cross-clamp times were 223.8 ± 34.1 and 158.2 ± 28.3 min. In one case, systemic venous obstruction was noted after the patient’s discharge from the operating room, so a repeat hemi-Mustard procedure was performed. The mean mechanical ventilation time and mean intensive care unit stay were 146.3 ± 30.6 h and 8.2 ± 2.7 d. No

### TABLE I. Patient Characteristics and Outcomes of Bidirectional Glenn Shunt Placement

| Variable | Group 1 (Fontan Completion) | Group 2 (Anatomic Completion) | Group 3 (Prolonged Palliation) | Total | P Value |
|----------|-----------------------------|-------------------------------|-------------------------------|-------|---------|
| **Patient Characteristics** | | | | | |
| Age at BDG (yr) | 2.1 ± 1.8 | 1.1 ± 1.3 | 4.5 ± 3.7 | 3.7 ± 2.2 | 0.004* |
| Weight at BDG (kg) | 14.6 ± 4.2 | 8.5 ± 3.1 | 20.3 ± 7.2 | 17.2 ± 14.9 | <0.001* |
| Preoperative So2 (%) | 80.7 ± 7.3 | 82.9 ± 10.6 | 75.3 ± 14.7 | 79.5 ± 13.5 | 0.181* |
| Preoperative mPAP (mmHg) | 14.5 ± 3.5 | 14.8 ± 2.7 | 13.9 ± 3.3 | 14.3 ± 3.1 | 0.686* |
| Nakata index (mm²/m²) | 243.3 ± 73.5 | 252.6 ± 80.3 | 240.8 ± 68.5 | 245.3 ± 72.7 | 0.891* |
| Systemic ventricular EF (%) | 62.3 ± 10.6 | 64.7 ± 8.9 | 58.7 ± 11.5 | 61.8 ± 10.3 | 0.243* |
| Tricuspid regurgitation | — | — | — | — | 0.948** |
| None | 3 (23.1) | 3 (27.3) | 6 (23.1) | 12 (24) | — |
| Mild | 7 (53.8) | 5 (45.5) | 11 (42.3) | 23 (46) | — |
| Moderate | 2 (15.4) | 1 (9.1) | 3 (11.5) | 6 (12) | — |
| Severe | 1 (7.7) | 2 (18.2) | 6 (23.1) | 9 (18) | — |
| Previous modified BT shunt | 1 (7.7) | 0 | 2 (7.7) | 3 (6) | 0.638** |
| MAPCA | 1 (7.7) | 1 (9.1) | 2 (7.7) | 4 (8) | 0.989** |
| Heterotaxy syndrome | 2 (15.4) | 0 | 3 (11.5) | 5 (10) | 0.425** |
| TAPVC | 2 (15.4) | 0 | 3 (11.5) | 5 (10) | 0.425** |
| Levocardia | 5 (38.5) | 0 | 9 (34.6) | 14 (28) | 0.062** |
| Dextrocardia | 5 (38.5) | 1 (9.1) | 10 (38.5) | 16 (32) | 0.183** |
| Mesocardia | 3 (23.1) | 10 (90.9) | 7 (26.9) | 20 (40) | <0.001** |
| Hematocrit (%) | 52.2 ± 9.5 | 50.5 ± 11.3 | 53.8 ± 12.7 | 52.4 ± 10.8 | 0.853* |

### BDG Outcomes

| Variable | Group 1 (Fontan Completion) | Group 2 (Anatomic Completion) | Group 3 (Prolonged Palliation) | Total | P Value |
|----------|-----------------------------|-------------------------------|-------------------------------|-------|---------|
| Postoperative So2 (%) | 93.7 ± 5.3 | 93.6 ± 9.2 | 95.2 ± 7 | 94.1 ± 7.3 | 0.748* |
| Postoperative mPAP (mmHg) | 12.2 ± 3.4 | 13.1 ± 3.7 | 12.6 ± 3.1 | 12.8 ± 3.4 | 0.803* |
| Systemic ventricular EF (%) | 64.7 ± 9.8 | 65.1 ± 10.3 | 62.5 ± 13.1 | 64.3 ± 12.7 | 0.773* |
| Prolonged pleural effusion | 1 (7.7) | 0 | 2 (7.7) | 3 (6) | 0.638** |

BDG = bidirectional Glenn shunt placement; BT = Blalock-Taussig; EF = ejection fraction; MAPCA = major aortopulmonary collateral arteries; mPAP = mean pulmonary artery pressure; So2 = oxygen saturation; TAPVC = total anomalous pulmonary venous connection

* Student t test
** χ² test

Data are presented as mean ± SD or as number and percentage. P <0.05 was considered statistically significant.
death, ventricular dysfunction, or pleural effusion was noted after 1½ ventricular repair. At the latest follow-up, the mean systemic ventricular EF was 0.63 ± 0.09, and no patient was in NYHA class III/IV. The mean So₂ was 95.5% ± 6.9%, and only one patient had an So₂ below 95%. Figure 2C shows dynamic changes in TR, although the changes were not significant.

**Outcomes of Prolonged Palliation (Group 3)**
The median follow-up time in group 3 was 4.5 years (range, 2.3–8 yr). During the follow-up period, no patients died, and none had impaired ventricular function. The mean So₂ at latest follow-up was 91.6% ± 10.3%. One patient (3.8%) was documented in NYHA class III/IV. However, pulmonary arteriovenous fistula (presenting as obvious cyanosis and a sharp decrease in So₂) developed in 3 patients at 4.6, 6.2, and 5.2 years, respectively, after BDG. Figure 2D shows a slight, though not significant, increase in TR at follow-up.

**Overall Mortality Rates and Functional Status**
For the entire cohort, the estimated survival rate after last surgical procedure was 98% at 1 year, 95.7% at 3 years, and 95.6% at 5 years; the estimated proportion (expressed as a percentage) of patients in NYHA class I/II at the respective times was 100%, 96.6%, and 85.2% (Fig. 3).

**Discussion**
From January 2005 through December 2014, 50 patients who had a diagnosis of CCTGA accompanied by LVOTO and VSD underwent BDG as a palliative procedure at our center. Mid- to long-term outcomes were favorable in the entire cohort, including patients who subsequently underwent second-stage TCPC (group 1), second-stage hemi-Mustard and Rastelli procedures (group 2), or prolonged palliation with BDG (group 3). During the same period, 48 patients underwent a double-switch procedure at our center (those patients were not included in this study).

Other investigators have reported excellent in-hospital and midterm survival rates after the double-switch operation. However, patients with atrioventricular discordance usually have an associated cardiac abnormality such as a VSD and morphologic LVOTO. The hemo-
dynamics in such complex cases are quite different from those in cases of simple CCTGA. Malposition of the cardiac apex, which substantially increases the difficulty of surgical repair, is also often seen. In addition to VSD and LVOTO, most of our patients had positional anomalies; many also presented with TAPVC or heterotaxy syndrome. The optimal palliative procedure in this challenging group remains largely unknown.

Bidirectional Glenn shunt placement has become an effective palliative procedure and a standard intermediate step toward TCPC. In our center, we prefer 2-stage effective palliative procedure and a standard intermediate procedure over conventional atrial baffle procedures, which might be complicated by positional abnormalities, is its relatively simple technique. Hence, 1½ ventricular repair is indicated when dextrocardia or mesocardia is present. In our cohort, all 11 patients who underwent the hemi-Mustard and Rastelli procedures had associated cardiac malposition. The excellent survival rate (100%) and excellent functional status (all in NYHA class I/II) in this group were consistent with the report- ed advantages of 1½ ventricular repair.

In our cohort, patients who underwent prolonged palliation (group 3) were older and weighed more than the other patients at the time of BDG. In addition, none of the patients in group 3 achieved Fontan completion, mainly because of low medical compliance or financial constraints (neither factor is rare in developing countries). We hypothesize that late referral in this group contributed the most to the differences in age and weight between the groups. For patients undergoing univentricular palliation, atrioventricular valve regurgitation remains an independent risk factor for unfavorable outcomes. Atrio- ventricular valvuloplasty or replacement remains one of the most challenging fields in pediatric cardiac surgery. Moreover, tricuspid problems often present in patients with CCTGA and can lead to deteriorating ventricular function after BDG and TCPC. However, TR did not increase in any of our 3 groups either early after BDG or at latest follow-up (Fig. 2), which is evidence of BDG’s usefulness as a palliative procedure for CCTGA associated with VSD and LVOTO.

Investigators have reported favorable surgical outcomes for patients with CCTGA through the application of multiple surgical strategies, including univentricular repair, double switch and modifications, and 1½ ventricular repair. In our cohort, with its more complex cardiac anatomy, the overall survival rate (95.6% at 5 y), functional status (85.2% in NYHA class I/II), and low incidence of reintervention are comparable, which further supports BDG as an optional palliative procedure. However, chronic cyanosis, polycythemia, and reduced exercise capacity may develop after BDG, thereby increasing the need for long-term follow-up. Therefore, we recommend palliative BDG and subsequent Fontan completion.

![Graph shows curves for estimated survival and proportion of patients in New York Heart Association (NYHA) functional class I/II, with 98% confidence intervals, after last surgical procedure.](Figure 3)
or 1½ ventricular repair, when feasible, in patients who have CCTGA accompanied by IVOTO and VSD and who have a contraindication to biventricular repair of the cardiac positional anomaly.

Limitations of our study include its single-institutional and retrospective nature. The relatively small sample size may lead to a false-negative result when comparing variables between groups. The groups of patients were not strictly comparable because of their different anatomic factors. Lack of exercise testing data was a substantial limitation and should be investigated further. Moreover, the follow-up duration remains short, and longer-term follow-up is warranted.

In conclusion, BDG can be used to palliate complex CCTGA. It produces favorable short- to midterm outcomes and can be feasibly followed by Fontan completion and anatomic repair. Use of BDG for open-ended palliation is also acceptable but necessitates careful long-term monitoring of patients who are at risk for further hemodynamic compromise.

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