Stage II palliation of hypoplastic left heart syndrome without cardiopulmonary bypass

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Objectives: Bidirectional cavopulmonary anastomosis has been performed without cardiopulmonary bypass for some single-ventricle heart defects. Limited data are available for the outcomes of off-pump bidirectional cavopulmonary anastomosis in infants with hypoplastic left heart syndrome. The purpose of this study is to determine the early outcomes for stage II palliation of hypoplastic left heart syndrome without cardiopulmonary bypass.

Methods: This is a retrospective review of infants having surgical palliation of hypoplastic left heart syndrome from April 2003 to March 2010 at a single institution.

Results: Seventy-five infants had a modified Norwood procedure, 65 with a right ventricle–pulmonary artery conduit, 10 with an aortopulmonary shunt, 2 with atrioventricular valve repair, and 3 with extracorporeal life support. Sixty-eight patients had hypoplastic left heart syndrome or one of its variants, and 7 had other single-ventricle lesions. There were 2 stage I deaths. Stage I survival was 97% (95% confidence interval, 88%–99%). Another 5 infants succumbed in the interstage period. Of the 68 stage I and interstage survivors, 61 had bidirectional cavopulmonary anastomoses, 20 without cardiopulmonary bypass. Median age was 6 months (range, 4–13 months), and median weight was 6.1 kg (range, 5.2–9.0 kg). There were no conversions to cardiopulmonary bypass when off-pump bidirectional cavopulmonary anastomosis was attempted. There were no hospital deaths. Median ventilation duration was 10 hours (range, 6–18 hours), and length of stay was 5 days (range, 4–9 days). Follow-up was available on all infants at a median duration of 17 months (range, 3–43 months), with no unplanned reinterventions.

Conclusions: Bidirectional cavopulmonary anastomosis without the use of cardiopulmonary bypass can be performed safely and with low mortality for selected infants with hypoplastic left heart syndrome. Midterm to long-term outcomes remain to be determined. (J Thorac Cardiovasc Surg 2011;141:400-6)

Outcomes for staged palliation of hypoplastic left heart syndrome (HLHS) and its variants have improved significantly over the last few decades. The second stage is a critical juncture at which significant benefit is offered. Stage II palliation attenuates pulmonary overcirculation, reduces the risk of sudden cardiac death, and provides a more stable circulation for infants having undergone the Norwood procedure.[1-3] A number of surgical approaches and strategies have been developed for second-stage palliation, including the bidirectional cavopulmonary anastomosis (BCPA) or hemi-Fontan operation. Cardiopulmonary bypass (CPB) is routinely used with each and can include cardioplegic cardiac arrest, deep hypothermic circulatory arrest, or both.

The off-pump BCPA was first reported by Lamberti and colleagues in 1990[4] and has been applied to single-ventricle lesions with antegrade pulmonary blood flow through a native or banded main pulmonary artery (PA) or a systemic–PA shunt contralateral to the side of the superior vena cava (SVC).[5,6] Data regarding stage II palliation of HLHS without the use of CPB are limited.

Originally described by Norwood and recently popularized by Sano and others, the right ventricle (RV)–PA conduit as a modification of the Norwood procedure has achieved increased use.[7-12] The advantages of the conduit include higher diastolic blood pressure, potentially improved coronary perfusion with lower volume loading, and lower myocardial oxygen demand. The number of postoperative interventions required to balance the pulmonary and systemic circulations might also be reduced when compared with the use of a systemic–PA shunt.[7] A recent report of more than 500 infants undergoing the Norwood procedure who were randomly assigned to the modified Blalock–Taussig shunt (n = 275) or the RV–PA conduit (n = 274) at 15 North American centers showed that survival 12 months after randomization was higher with the RV–PA

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Abbreviations and Acronyms

AV	= atrioventricular
BCPA	= bidirectional cavopulmonary anastomosis
BSID-II	= Bayley Scales of Infant Development-II
CPB	= cardiopulmonary bypass
HLHS	= hypoplastic left heart syndrome
ICU	= intensive care unit
NIRS	= near-infrared spectroscopy
PA	= pulmonary artery
RV	= right ventricle
SVC	= superior vena cava

conduit than with the modified Blalock–Taussig shunt.[13] Another potential advantage of the RV–PA conduit is that it can be used for antegrade blood flow during performance of the off-pump BCPA, thus maintaining adequate oxygenation and gas exchange, whereas the superior cavopulmonary anastomosis is performed with the assistance of a passive venous shunt.

Here we report our experience with BCPA without CPB in selected infants with HLHS who had an RV–PA conduit as a modification of the Norwood procedure.

MATERIALS AND METHODS

Patient Population

The pediatric cardiac surgery database at the University of California, San Francisco, was queried for all neonates having a modified Norwood procedure for HLHS or one of its variants or other forms of single-ventricle lesions with obstruction to systemic blood flow from April 2003 to march 2010. This patient cohort was then tracked for stage I and interstage survival, progression to stage II palliation, and subsequent survival. Infants palliated with an RV–PA conduit modification of the Norwood procedure were candidates for BCPA without CPB if there was no need for concomitant atrioventricular (AV) valve repair for moderate-to-severe insufficiency or left PA reconstruction for any degree of stenosis.

Perioperative and outcome data were also collected for a control group of patients having BCPA with CPB. Infants who had associated procedures, such as AV valve repair, repair of aortic arch stenosis, repair of RV aneurysm, atrial septectomy, and repair of pulmonary veins, were not included in this selected group.

Echocardiographic Analysis and Cardiac Catheterization

Preoperative echocardiographic analysis was performed in all patients, with particular focus on qualitative assessment of ventricular function; AV valve function; aortic arch, PA, and pulmonary venous anatomy; and interatrial septal patency. Ventricular function was subjectively graded as good, fair, or poor, whereas AV valve regurgitation was graded as none–trace, mild, moderate, or severe. Infants who had moderate or greater AV valve regurgitation or PA stenoses were not candidates for BCPA without CPB.

All infants had preoperative cardiac catheterization to assess candidacy for second-stage palliation, evaluate hemodynamics, and determine any anatomic abnormalities. Data acquisition included PA, atrial, ventricular, and aortic/arch pressures. Systemic, PA, superior caval, left atrial, and pulmonary venous hemoglobin–oxygen saturations were also measured.

Operative Technique

After repeat sternotomy, dissection, and mobilization of all mediastinal and cardiac structures, a full dose of systemic heparin was given to achieve an activated clotting time of greater than 400 seconds. The right PA was extensively mobilized up to its hilar branches (Figure 1, *A*). A test occlusion of the right PA was then performed, and if hemoglobin–oxygen saturations remained greater than or equal to 70%, then the off-pump BCPA was performed. The SVC–innominate vein junction and right atrial appendage were cannulated with 12F Pacifico cannulas and were joined with a ¼-inch connector for insertion of a passive venous shunt (Figure 1, *B*). A snare was tightened above the azygous vein, which was doubly ligated. The SVC–right atrial junction was clamped and divided, and the cardiac end was oversewn with polypropylene sutures. After proximal and distal control of the right PA was achieved, a longitudinal arteriotomy was made. The end of the SVC was sewn to the PA with running fine polypropylene sutures (Figure 1, *C*). The anastomosis was deaired, the clamps and snares were removed, and the patient was decannulated. The RV–PA conduit was then dissected, mobilized, and quadruply clipped and divided (Figure 1, *D*). Protamine was administered, hemostasis was ensured, and bilateral pleural and single mediastinal drains were inserted. Chest drains were removed postoperatively when the total volume of drainage was less than 1 mL $\cdot$ kg^{-1} $\cdot$ d^{-1}. Continuous monitoring of arterial blood pressure, pulse oximetry, hemoglobin-oxygen saturation, and near-infrared spectroscopy (NIRS) was performed perioperatively.

Assessment of Outcomes

Outcomes measured included demographic information, morphologic data, intraoperative NIRS data, duration of ventilatory support, intensive care unit (ICU) length of stay, chest tube drainage, hospital length of stay, death, stroke, bleeding, infection, and postoperative hemoglobin–oxygen saturation at various time points (1 hour postoperatively, 6 hours postoperatively, 24 hours postoperatively, and before discharge) were collected.

Cerebral Monitoring and Neurologic Testing

NIRS was performed in all patients by using standard techniques, as previously reported.[14,15] The Bayley Scales of Infant Development-II (BSID-II) was performed at 1 year. The mental development index and psychomotor development index scores are reported as means ± standard deviations. Comparisons of outcomes were performed by means of non-parametric testing with the Mann–Whitney test, assuming that normality or equality of variance is not met by either group.

Statistical Analysis

All data are expressed as frequencies, means ± standard deviations or medians with appropriate ranges. Comparative statistical analysis between groups was performed by using *t* tests, Fisher exact tests, or Mann–Whitney tests, where appropriate. The study was reviewed and approved by the Committee on Human Research of the University of California, San Francisco, and need for patient consent was waived because of its retrospective nature.

RESULTS

Patients' Characteristics

Figure 2 shows a flow diagram of the cohort of patients having undergone a Norwood procedure and the subsequent study population palliated with BCPA without CPB.

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