Surgical palliation in patients with a single ventricle and dextrocardia

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Background: Dextrocardia is found in a significant proportion of patients undergoing a single-ventricle repair. Surgical outcomes in this cohort are unclear.

Methods: The records of 41 consecutive patients with single-ventricle physiology and dextrocardia presenting in a single institution from 1990 to 2008 were reviewed. Of this cohort, 19 patients had heterotaxy syndrome. Twenty-five of the 41 patients had atrioventricular valve regurgitation (AVVR) on presentation (mild, 13; moderate, 9; severe, 3).

Results: One patient died before intervention. Initial surgical palliation was performed in 31 patients. Four patients died postoperatively and 4 interim deaths occurred between initial palliation and bidirectional cavopulmonary shunt (BCPS). Thirty of the surviving 32 patients underwent BCPS, with 2 perioperative deaths. There were 4 additional deaths before Fontan surgery. Twenty-two of the surviving 25 patients underwent a Fontan procedure. There was 1 postoperative mortality. Survival to the age of 15 years was 56% (95% confidence interval [CI], 39%-70%). Patients with moderate or severe regurgitation had higher mortality if they were managed conservatively rather than by surgery (5 of 6 vs 2 of 6; $P = .24$). Patients with bilateral BCPS had better operative outcomes and survival compared with peers with unilateral anastomosis (odds ratio, 27; $P = .005$; 95% CI, 2.7-269). The side of the systemic venous pathway did not seem to influence outcomes.

Conclusions: Surgical outcomes of single-ventricle palliation seem poor in patients with dextrocardia. Aggressive management of congenital AVVR might improve the long-term prognosis. (J Thorac Cardiovasc Surg 2014;148:1475-80)

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Dextrocardia, defined as cardiac positioning with a rightward base-apex axis, is a rare cardiac anomaly with an estimated incidence of 1 in 10,000 to 12,000 births.[1,2] Albeit sparse in the general population, a recent review of 212 patients who underwent a univentricular repair in our institution revealed a high incidence of dextrocardia at 11.3%.[1] The prevalence of univentricular anatomy in patients with dextrocardia has been reported to be 16% to 25%.[2-4] Currently, there are no reports describing the impact of dextrocardia in patients undergoing univentricular repair. After the Fontan procedure, the bulk of the ventricular mass on the right side may cause obstruction to the systemic venous pathway draining the inferior vena cava. It is unclear whether a right-sided atrial tunnel or extracardiac conduit should be avoided in this instance.

In addition, there is a known high incidence of heterotaxia in patients with dextrocardia.[4] The associated variations in both systemic venous and pulmonary drainage pose some operative challenges. The aim of this study was to investigate the outcomes of patients with dextrocardia undergoing single-ventricle palliation with a particular focus on the impact of the side of the routing of the systemic venous pathway.

PATIENTS AND METHODS

The study protocol was approved by our institutional Human Research Ethics Committee. Between 1990 and 2008, 543 consecutive patients with a functionally single ventricle were admitted to the Royal Children's Hospital, Melbourne, Australia. The records of 41 patients (7.6%) diagnosed with dextrocardia were reviewed. Their most current follow-up data were gathered from the hospital database and referring cardiologists.

Dextrocardia status, along with all other visceral and vascular parameters were identified from echocardiography, and further verified against intraoperative reports to ensure accuracy. The anatomic details of the patients are described in Tables 1 and 2.

Fifteen of the 41 patients (37%) had a common atrioventricular (AV) valve. A common AV valve was present more frequently in patients with heterotaxy (13 of 19, 68% vs 2 of 22, 9.1%). Twenty-five patients had AV valve regurgitation at diagnosis. AV valve regurgitation (AVVR) was

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This research project was supported by the Victorian Government's Operational Infrastructure Support Program.

Disclosures: Yves d'Udekem is a Career Development Fellow of The National Heart Foundation of Australia (CR 10M 5339). All other authors have nothing to disclose with regard to commercial support.

Received for publication Nov 21, 2012; revisions received Sept 17, 2013; accepted for publication Oct 22, 2013; available ahead of print Feb 16, 2014.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2013.10.077

Abbreviations and Acronyms
AV = atrioventricular
AVVR = atrioventricular valve regurgitation
BCPS = bidirectional cavopulmonary shunt
CI = confidence interval
HR = hazard ratio

more common in patients with heterotaxy (16 of 19, 84% vs 9 of 22, 41%; $P = .009$). The morphology of the regurgitant valve of these 25 patients was as follows: common AV valve (12), tricuspid (10), both mitral and tricuspid (2), and mitral (1).

Statistical Analysis

All continuous data were expressed as the median (interquartile range). Categorical data were expressed as the frequency (percentage). Proportions were evaluated with χ^2 analysis. Risk factors for mortality were examined by univariate Cox regression and overall survival of the patient population from birth was illustrated via Kaplan-Meier analysis. The small sample size precluded use of a multivariate analysis to separate the effects of different risk factors on mortality. Logistic regression was used to compare the impact of having a bilateral bidirectional cavopulmonary shunt (BCPS) rather than a unilateral one on mortality and/or postoperative complications after BCPS.

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RESULTS

The list of surgical interventions is shown in Figure 1. One patient with associated cerebral lesions died before surgery took place.

There were 4 in-hospital deaths after the initial neonatal surgery related to myocardial dysfunction. An additional 4 patients died in the interim period between initial palliation and subsequent BCPS procedure. Autopsy was performed in all 4 patients and the following causes of death were identified: aspiration pneumonia (1), respiratory failure (1), unknown (2).

Thirty patients underwent BCPS at a median age of 1.4 years (0.5-5.9 years). Fourteen patients had single BCPS, 10 received bilateral BCPS, and the remaining 6 patients had Kawashima procedures. The cavopulmonary shunts were placed in the left, right, and bilaterally in 7, 9, and 14 patients, respectively.

Two patients died postoperatively. Both patients had difficult postoperative courses with low output syndrome requiring extracorporeal membrane oxygenation. Five patients had the following postoperative complications: arrhythmia (4), thromboembolism (1), heart failure (1), and reoperation for take-down of the BCPS to a Blalock-Taussig shunt (1). There were 4 deaths between hospital discharge after BCPS and Fontan surgery or last follow-up, all sudden unexpected deaths.

Fontan Procedure

Of the 25 surviving patients, 22 underwent a Fontan procedure at a median age of 5.4 years (1.7-17.2 years). An extracardiac Fontan operation was performed in 17 patients and a lateral tunnel Fontan operation was done in 5 patients. Fourteen of the Fontan conduits were fenestrated. The extracardiac conduits coursed along the left side of the heart in 11 patients and the right side in 6. Thirteen of the 17 patients who underwent an extracardiac conduit had their conduit on the ipsilateral side of an existing BCPS anastomosis. One patient died after Fontan secondary to low output syndrome. Eleven patients had pleural effusions persisting for more than 2 weeks after surgery. Postoperative recovery course was complicated in 13 patients by arrhythmia (3), bleeding (1), and reoperation for 2 patients for management of postoperative bleeding and mediastinitis.

AVVR

Twenty-five of the 41 patients (61%) had AVVR diagnosed on initial presentation: mild (13), moderate (9), and severe (3). Six of the 12 patients with moderate or severe regurgitation and 2 with mild regurgitation underwent valve repairs. The AV valve repair was performed shortly after the initial palliation surgery (2), during BCPS (4), after BCPS (1), and during Fontan (1). Two patients required further mechanical valve replacement during the same hospital stay. Later reoperation was necessary in 2 additional patients for further progression of the AVVR.

A total of 10 of the 25 patients with AVVR died. Of the remaining 15 patients, progression in severity from mild to moderate was only noted in 1 patient who was managed conservatively. Amongst the 12 patients with moderate or severe regurgitation, 4 of the 6 who were conservatively managed died and 1 of the 6 who underwent surgery died.

Late Follow-up and Survival Analysis

There were no further deaths after Fontan completion. Four patients were lost to follow-up. After a median follow-up of 11.2 years (range, 1.2-19.2 years), 20 patients were still alive. Overall survival to 15 years of age was 56% (95% confidence interval [CI], 39%-70%) (Figure 2). Survivors were in New York Heart Association class I (13), II (4), and III (3). By univariate analysis, transposition of great arteries (hazard ratio [HR], 3.4, $P = .057$; 95% CI, 0.97-11.72) and dominant right ventricle (HR, 2.6; $P = .059$; 95% CI, 0.96-7.08) were associated with poorer postoperative outcomes, but failed to achieve statistical significance (Table 3).

Side of Anastomosis and Outcomes

Analysis of the relationship between the side of anastomosis and postoperative outcomes failed to reveal a significant relationship for both BCPS and Fontan patients. Two of 9 patients (22.2%) with right BCPS anastomoses and 4 out of 7 patients (57.1%) with left BCPS anastomosis experienced either a postoperative complication or death. Four of 6 patients (66.7%) and 7 of 11 patients (63.6%) with