Outcomes of patients born with single-ventricle physiology and aortic arch obstruction: The 26-year Melbourne experience

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Background: To review the long-term outcomes of patients born with single-ventricle physiology and aortic arch obstruction.

Methods: Follow-up of 70 consecutive neonates undergoing single-ventricle palliation and arch repair, excluding hypoplastic left heart syndrome, between 1983 and 2008, was reviewed. Dominant arch anomalies were coarctation (n = 48), interrupted arch (n = 10), and hypoplastic arch alone (n = 12). Neonatal Damus procedure with arch repair and shunt became the dominant approach, being performed in 1 (10%) of 10 in 1983 to 1989, 9 (32%) of 28 in 1990 to 1999, and 23 (72%) of 32 in 2000 to 2008.

Results: All patients underwent an initial procedure at a median of 6 days (range, 4-12 days): pulmonary artery banding and arch repair (n = 35); Damus, arch repair, and shunt (n = 33); and other (n = 2). Twenty-six patients died before Fontan completion. Of the 34 survivors of initial banding, 17 (50%) later required a Damus and 4 (12%) required subaortic stenosis relief. Forty patients underwent Fontan completion at a median age of 5 years (range, 4-7 years). After a mean of 5 ± 6 years after Fontan, there was 1 hospital death and 1 Fontan takedown. Overall survival was similar if patients initially underwent a Damus or pulmonary artery banding ($P = .3$). Overall survival at 10 years was 53% (95% confidence interval, 42%-67%).

Conclusions: Patients born with single-ventricle physiology and arch obstruction have a high risk of mortality in the first years of life. Their outcomes seem excellent once they reach Fontan status. It is likely that, in patients with single-ventricle and arch obstruction, strategies to avoid systemic outflow tract obstruction should be implemented in early life, and regular monitoring of blood pressure is warranted. (J Thorac Cardiovasc Surg 2014;148:194-201)

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It is widely accepted that aortic arch obstruction is associated with poor outcomes in patients with single-ventricle physiology because of the potential for myocardial hypertrophy and subaortic stenosis in the first year of life.[1,2] There has been much debate on the best approach for these patients in the neonatal period. All teams agree that the aortic arch obstruction should undergo an early relief.[3] There is, however, contention between those who favor the use of an extensive Norwood-type operation, including an aortic arch repair, a Damus-Kaye-Stansel (DKS) procedure, and a systemic-pulmonary shunt and those who prefer to perform an initial banding of the pulmonary artery (PA) at the time of the arch repair and reserve later completion of a DKS procedure to those developing subaortic obstruction. The presence of coarctation or arch obstruction should strongly suggest the potential for systemic obstruction at the bulboventricular foramen,[4] and previous studies have demonstrated that most patients who undergo initial PA banding will ultimately develop subaortic stenosis early in life, necessitating further intervention.[5-8] Yet, initial banding is still favored by many teams today.[6-8] An arterial switch operation may be performed in a few of these patients, but they have a high propensity to develop subaortic stenosis after this operation, and the adjustment of pulmonary blood flow is unpredictable.[9,10]

We have realized that up to half of the patients with biventricular circulation undergoing aortic arch repair may require reintervention in the decades after the initial intervention.[11-13] We know that the development of diastolic dysfunction consequential to the arch obstruction is a factor leading to the failure of the Fontan circulation, and we wondered whether the patients requiring aortic arch

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This study was supported by the Victorian Government's Operational Infrastructure Support Program. Dr d'Udekem is a Career Development Fellow of The National Heart Foundation of Australia (CR 10M 5339). Dr Iyengar is a cofounded Postgraduate Scholar of the National Health and Medical Research Council/National Heart Foundation of Australia (APP1038802), and holds an Royal Australian College of Surgeons Catherine Marie Enright Kelly Postgraduate Scholarship.

Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication Feb 5, 2013; revisions received June 4, 2013; accepted for publication July 26, 2013; available ahead of print Sept 26, 2013.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2013.07.076

Abbreviations and Acronyms

BCPS = bidirectional cavopulmonary shunt
BT = Blalock-Taussig
CI = confidence interval
DKS = Damus-Kaye-Stansel
DORV = double-outlet right ventricle
HR = hazard ratio
PA = pulmonary artery

surgery who undergo single-ventricle palliation would have poor outcomes.[14] Little is known of the progression of these patients toward the Fontan pathway, and there has been no information about their outcome beyond Fontan completion.

This study reviews our experience of patients born with single-ventricle physiology and neonatal aortic arch surgery during a period of 26 years, during which time we gradually adopted Norwood-type procedures as our primary approach.

METHODS

Study Population

The design of the study was approved by The Royal Children's Hospital Human Research and Ethics Committee, and the need for consent was waived because of the retrospective nature of the study.

The hospital database was screened to identify the patients who underwent single-ventricle palliation and aortic arch repair for coarctation, interrupted aortic arch, or aortic arch hypoplasia at The Royal Children's Hospital between 1983 and 2008. Patients with hypoplastic left heart syndrome were excluded because of the severity of this condition and associated poor early outcomes in the first era of the study. Seventy patients were identified: 46 (66%) were male and 24 (34%) were female.

Morphologic Data

Morphologic diagnoses of the patients were established by 2-dimensional echocardiography and Doppler flow imaging, and confirmed on macroscopic visualization at the initial neonatal palliation. Characteristics of the patients are displayed in Table 1.

Aortic coarctation was defined as a discrete narrowing at the isthmus of the arch between the left subclavian artery proximally and the ductus arteriosus distally. *Interrupted aortic arch* was defined as an anatomic lack of continuity between the proximal and distal segments of the aortic arch. The arch was considered hypoplastic if the echocardiographic report, the cardiologist's report, or the surgical notes labeled it as hypoplastic, or if any portion of the arch had a z score diameter of less than −2.0.

Surgical Management

All patients underwent palliation in the neonatal period at a median age of 6 days (range, 4-12 days), with the goal to ultimately achieve Fontan circulation: 33 (47%) had a DKS procedure; 35 (50%) had a PA band and aortic arch repair, with 4 of them undergoing the arch repair as a previous separate procedure; and 2 (3%) had arterial switch and aortic arch repair.

A strategy of performing a neonatal DKS procedure became the dominant approach during the study period, being performed in 1 (10%) of 10 patients in the initial years of 1983 to 1989, in 9 (32%) of 28 patients in 1990 to 1999, and in 23 (72%) of 32 patients in the recent 2000 to 2008 period (Figure 1). Consequently, PA banding has become less favored in recent times, being performed in 8 (80%) of 10 patients in 1983 to 1989, 18 (64%) of 28 patients in 1990 to 1999, and in only 9 (28%) of 32 patients in 2000 to 2008.

The DKS procedure involved complete transection of both the PA and the aorta, followed by the anastomosis of these 2 vessels such that no more than one third of the smaller of the 2 vessels was sutured to the larger vessel. The descending aorta was anastomosed to the ascending aorta in an end-to-side manner in most of the cases. Additional pericardial patch was added to the arch repair in 21 patients (64%) using homograft pericardium in 14 and autologous pericardium in 7.

Follow-up

The files and echocardiographic reports of the patients were reviewed, and their follow-up was gathered from hospital databases and their referring cardiologists.

Hospital mortality was defined as death that occurred within 30 days after surgery or during hospital stay. *Late mortality* was defined as death that occurred after this period.

Resting hypertension for children and adolescents was defined as a systolic or diastolic blood pressure of greater than the 95th percentile for age and height, and *prehypertension* was defined as between the 90th and 95th percentile or if blood pressure was greater than 120/80 mm Hg.[15] *In adults, resting hypertension* was defined as a systolic blood pressure of greater than 140 mm Hg or a diastolic blood pressure of greater than 90 mm Hg; and *prehypertension* was defined as a systolic blood pressure between 120 and 139 mm Hg or a diastolic blood pressure between 80 and 89 mm Hg.[16]

Aortic arch reobstruction was defined as a peak gradient exceeding 25 mm Hg across the repair site on echocardiogram, or an upper limb to lower limb blood pressure gradient of more than 20 mm Hg.

Statistical Analysis

All data were exported to and analyzed using STATA, version 12.1 (Stata Corporation, College Station, Tex). Data quoted in the text are summarized as either mean ± SD or median (interquartile range). A nonpaired Student *t* test, a Pearson χ^2 test (when appropriate), or a Fisher exact test was used to compare patients who underwent neonatal DKS with patients who underwent PA banding for each of the characteristics listed in Table 1. A univariable analysis of risk factors for hospital mortality after initial surgery was assessed using either a Fisher exact test or logistic regression. Cox regression analysis was used to examine the association of reintervention for systemic outflow tract obstruction and mortality with all collected patient and surgical characteristics. Because of the few outcomes, it was not feasible to perform multivariable risk analyses.

RESULTS

The postoperative courses of the 70 patients in this study are summarized in Figure 2. The mean follow-up from initial neonatal palliation was 7 ± 7 years.

Initial Neonatal Surgery

Comparative characteristics of the 70 patients at their first palliative procedure are given in Table 1. The sources of pulmonary blood flow used for the DKS procedure were as follows: modified Blalock-Taussig (BT) shunt (n = 30) and right ventricle–pulmonary artery shunt (n = 3).

Fifteen patients (21%) had the following 16 concomitant procedures at their initial neonatal surgery: atrial septectomy (n = 9), cor triatriatum repair (n = 2), subaortic stenosis relief (n = 2), tricuspid valve repair (n = 1),

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