Results of palliation with an initial pulmonary artery band in patients with single ventricle associated with unrestricted pulmonary blood flow

Bahaaldin Alsoufi, MD,[a] Cedric Manlhiot, BSc,[b] Alexandra Ehrlich,[c] Matthew Oster, MD,[c] Brian Kogon, MD,[a] William T. Mahle, MD,[c] Kevin Maher, MD,[c] Brian W. McCrindle, MD, MPH,[b] and Kirk Kanter, MD[a]

Objectives: Pulmonary artery banding is the initial palliative surgery in patients with single ventricle cardiac anomalies presenting with unrestricted pulmonary blood flow. Reported mortality in those receiving pulmonary artery banding is high, and its application in patients with single ventricle anomalies and arch obstruction is controversial. We report current-era results after pulmonary artery banding in patients with single ventricle anomalies, including those with arch obstruction.

Methods: Between 2002 and 2012, 73 patients with single ventricle anomalies and unrestricted pulmonary blood flow underwent pulmonary artery banding, including 29 (40%) who received simultaneous arch repair. Competing risk analysis modeled events after pulmonary artery banding (death/transplantation, transition to the Glenn procedure) and subsequently after the Glenn procedure (death/transplantation, transition to the Fontan procedure) and examined risk factors affecting outcomes.

Results: Hospital mortality was 3 of 73 patients (4%). Before the Glenn procedure, 16 patients (22%) underwent 18 reoperations, including shunt (n = 7), Damus–Kaye–Stansel connection plus shunt (n = 5), pulmonary artery banding adjustment (n = 5), and transplantation (n = 1). On competing risk analysis, 2 years after pulmonary artery banding, 8% of patients have died or received transplantation, 88% have undergone the Glenn procedure, and 7% were alive without the Glenn procedure. Five years after the Glenn procedure, 9% have died, 71% have undergone the Fontan procedure, and 20% were alive awaiting the Fontan procedure. Overall survival 5 years after pulmonary artery banding was 86%. On multivariable analysis, risk factors for mortality were associated genetic/extracardiac anomalies (hazard ratio, 3.7; 95% confidence interval, 1.1-12.2; $P = .03$) and high-risk morphology (heterotaxy, unbalanced atrioventricular septal defect, and mitral atresia) (hazard ratio, 4.1; 95% confidence interval, 1.1-15.6; $P = .04$).

Conclusions: Pulmonary artery banding is an acceptable initial palliative strategy of selected patients with single ventricle cardiac anomalies and unrestricted pulmonary blood flow, including those with concomitant arch obstruction. Short- and long-term outcomes are generally good, although results diverge with the worst outcomes noted in patients with heterotaxy, unbalanced atrioventricular septal defect, or associated extracardiac anomalies. (J Thorac Cardiovasc Surg 2015;149:213-20)

See related commentary on pages 220-1.

Multistage palliation is the mainstay in the management of infants born with single ventricle (SV) cardiac anomalies. The principles of first-stage palliation of patients with SV cardiac anomalies are consistent and aim to achieve unobstructed systemic cardiac output, unobstructed and controlled source of pulmonary blood flow, and unobstructed pulmonary venous return to the heart.[1,2] The choice of initial palliative surgery depends on the specific anatomy and the degree of systemic/pulmonary outflow obstruction.

In patients with SV anomalies and unrestricted pulmonary blood flow (UPBF), initial palliation typically has been pulmonary artery banding (PAB).[1-6] Nonetheless, several centers reported disappointing short- and long-term outcomes of patients with SV anomalies typically palliated with PAB, and some authors recommended the use of an aortopulmonary shunt plus division of the main pulmonary artery as an

From the Division of Cardiothoracic Surgery,[a] Children's Healthcare of Atlanta, Emory University School of Medicine, Atlanta, Ga; Labatt Family Heart Center,[b] Hospital for Sick Children, University of Toronto, Toronto, Ontario, Canada; and Sibley Heart Center,[c] Children's Healthcare of Atlanta, Emory University School of Medicine, Atlanta, Ga.

Disclosures: Kevin Maher reports equity ownership in Sensiotec. All other authors have nothing to disclose with regard to commercial support.

Read at the 40th Annual Meeting of The Western Thoracic Surgical Association, Dana Point, California, June 25-28, 2014.

Received for publication June 24, 2014; revisions received July 30, 2014; accepted for publication Aug 1, 2014; available ahead of print Sept 1, 2014.

Address for reprints: Bahaaldin Alsoufi, MD, Division of Cardiothoracic Surgery, Emory University School of Medicine, Children's Healthcare of Atlanta, 1405 Clifton Rd, NE, Atlanta, GA 30322 (E-mail: balsoufi@hotmail.com).

0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2014.08.007

Abbreviations and Acronyms

BTS = Blalock–Taussig shunt
CI = confidence interval
DKS = Damus–Kaye–Stansel
ECMO = extracorporeal membrane oxygenation
HR = hazard ratio
IQR = interquartile range
PAB = pulmonary artery banding
SV = single ventricle
SVOTO = systemic ventricular outflow tract obstruction
TAPVC = total anomalous pulmonary venous connection
UPBF = unrestricted pulmonary blood flow

alternative first-stage palliation strategy that would provide a more reliable source of pulmonary blood flow and more predictable pulmonary vascular protection.[4-7]

Infants with concomitant arch obstruction are especially challenging because they are at an increased risk for development of systemic ventricular outflow tract obstruction (SVOTO).[1-4,7-10] The ventricular hypertrophy as a result of PAB can cause subsequent enlargement of a subaortic conus or reduction in the size of the bulboventricular foramen, both leading to the development of subaortic obstruction.[1-4,7-10] SVOTO can create unfavorable hemodynamic conditions that can have detrimental effects on the future of patients with SV physiology, with failure to progress through palliative stages or late malfunction after final palliation.[1-4,7-10] Therefore, the initial management strategy of patients with SV anomalies with UPBF and arch obstruction remains controversial, with many groups favoring an initial Norwood-type palliation rather than PAB and concomitant arch repair.[1-3,7,9-12]

Given the potential disadvantages of all the different alternative treatment strategies, we aim to report our recent institutional experience with the palliation of patients with SV physiology who have UPBF with initial PAB, including those with simultaneous arch obstruction requiring repair, and to examine the risk factors that affect early and late survival in this challenging group of patients.

PATIENTS AND METHODS

Inclusion Criteria

Between 2002 and 2012, 73 infants with various SV anomalies associated with UPBF underwent PAB at Children's Healthcare of Atlanta, Emory University. PAB was the initial palliation in all 73 patients; none of them had a prior cardiac surgery. Patients who underwent bilateral branch PAB for hypoplastic left heart syndrome and associated anomalies were not included. Patients were identified using our institutional surgical database. Demographic, anatomic, clinical, operative, and hospital details were abstracted from the medical records for analysis. Approval of this study was obtained from the Children's Healthcare of Atlanta Institutional Review Board, and requirement for individual consent was waived for this observational study.

Follow-up

Time-related outcomes were determined from recent office visits present on the electronic chart of Children's Healthcare of Atlanta system or from direct correspondence with other pediatric cardiologists outside of the system. Mean follow-up duration after PAB was 6.0 ± 3.6 years and was 93% complete.

Statistical Analysis

Data are presented as means with standard deviation, medians with interquartile ranges (IQRs), or frequencies as appropriate. Time-dependent outcomes (death/transplantation and survival to the Glenn procedure) after PAB and time-dependent outcomes (death/transplantation and survival to the Fontan procedure) after the Glenn procedure were parametrically modeled. Parametric probability estimates for time-dependent outcomes uses models based on multiple, overlapping phases of risk (available for use with the SAS system at http://www.clevelandclinic.org/heartcenter/hazard). The HAZARD procedure uses maximum likelihood estimates to resolve risk distribution of time to event in up to 3 phases of risk (early, constant, and late). Competing risk analysis was performed to model the probability over time of each of 2 mutually exclusive end points after PAB: death/transplantation and survival to the Glenn; the remaining patients begin alive without the Glenn. After the Glenn procedure, mutually exclusive end points were death/transplantation and survival to the Fontan procedure, the remaining patients being alive awaiting the Fontan. Variables potentially influencing the likelihood of outcomes in the competing risk models were sought from demographic, anatomic, and surgical variables. Multivariable models were created using forward entry of variables given the limited sample size available for analysis. Effects of covariates on the probability of outcomes in competing risk models are given as hazard ratio (HR) with 95% confidence interval (CI). Clinical relevance of covariates of interest on the likelihood of selected outcomes was established by solving the regression equations for multiple "typical" test patients. All statistical analyses were performed using SAS version 9.3 (SAS Institute Inc, Cary, NC).

RESULTS

Patients' Characteristics

During the study period, 73 infants with SV anomalies associated with UPBF underwent PAB. There were 39 male infants (53%). The median age at the time of PAB was 14 days (IQR, 6-43), and the median weight was 3.4 kg (IQR, 3.0-3.9), with 9 patients (12%) weighing 2.5 kg or less. Eight children (11%) were born prematurely at 36 weeks or less gestation, and 9 children (12%) had associated genetic or major extracardiac anomalies. The specific morphology for the SV anomalies included tricuspid atresia (n = 14, 19%), double outlet right ventricle (n = 14, 19%), heterotaxy syndrome (n = 12, 16%), double inlet left ventricle (n = 11, 15%), unbalanced atrioventricular septal defect (n = 8, 11%), mitral atresia (n = 8, 11%), and other (n = 6, 8%). The predominant ventricle was of left morphology in 33 patients (45%) and right morphology in 32 patients (44%), and 8 patients (11%) had 2 equally developed ventricles.