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A contemporary comparison of the effect of shunt type in hypoplastic left heart syndrome on the hemodynamics and outcome at stage 2 reconstruction

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Objective: We compare the hemodynamics and perioperative course of shunt type in hypoplastic left heart syndrome at the time of stage 2 reconstruction and longer-term survival.

Methods: We retrospectively reviewed the echocardiograms, catheterizations, and hospital records of all patients who had a stage 1 reconstruction between January 2002 and May 2005 and performed a cross-sectional analysis of hospital survivors.

Results: One hundred seventy-six patients with hypoplastic left heart syndrome and variants underwent a stage 1 reconstruction with either a right ventricle–pulmonary artery conduit (n = 62) or a modified Blalock–Taussig shunt (n = 114). The median duration of follow-up is 29.1 months (range, 0-57 months). By means of Kaplan–Meier analysis, there is no difference in survival at 3 years (right ventricle–pulmonary artery conduit: 73% [95% confidence limit, 59%–83%] vs modified Blalock–Taussig shunt: 69% [95% confidence limit, 59%–77%]; $P = .6$). One hundred twenty-four patients have undergone stage 2 reconstruction (78 modified Blalock–Taussig shunts and 46 right ventricle–pulmonary artery conduits). At the time of the stage 2 reconstruction, patients with right ventricle–pulmonary artery conduits were younger (153 days [range, 108–340 days]; modified Blalock–Taussig shunt, 176 days [range, 80–318 days]; $P = .03$), had lower systemic oxygen saturation (73% [range, 58%–85%] vs 77% [range, 57%–89%], $P < .01$), and had higher preoperative hemoglobin levels (15.8 g/dL [range, 13–21 g/dL] vs 14.8 g/dL [range, 12–19 g/dL], $P < .01$) compared with those of the modified Blalock–Taussig shunt group. By means of echocardiographic evaluation, there was a higher incidence of qualitative ventricular dysfunction in patients with right ventricle–pulmonary artery conduits (14/46 [31%] vs 9/73 [12%], $P = .02$). However, no difference was observed in common atrial pressure or the arteriovenous oxygen difference.

Conclusion: Interim analyses suggest no advantage of one shunt type over another. This report raises concern of late ventricular dysfunction and outcome in patients with a right ventricle–pulmonary artery conduit.

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In the absence of transplantation, hypoplastic left heart syndrome (HLHS) and its variants require staged surgical reconstruction to palliate the anatomic abnormalities. Stage 1 reconstruction (S1R) includes aortic arch reconstruction, anastomosis of the proximal main pulmonary artery with the ascending aorta, ligation of the distal main pulmonary artery, atrial septectomy, and provision of a source of pulmonary blood flow. Recently, an alternative strategy to S1R has become popularized: replacing the modified Blalock–Taussig shunt (mBTS) with a right ventricle–pulmonary artery (RV-PA) conduit as the source of pulmonary blood flow.[1] The theory behind this modification is that the use of the RV-PA conduit will

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Abbreviations and Acronyms

AVVR = atrioventricular valve regurgitation
DHCA = deep hypothermic circulatory arrest
HLHS = hypoplastic left heart syndrome
mBTS = modified Blalock–Taussig shunt
RV-PA = right ventricle–pulmonary artery
S1R = stage 1 reconstruction
S2R = stage 2 reconstruction
S3R = stage 3 reconstruction

result in less diastolic runoff into the pulmonary vasculature and therefore improved coronary and myocardial perfusion. Several case series using the mBTS as a historical control have documented improved S1R early outcome with the use of the RV-PA conduit.[2-6] However, other case series using both contemporary and historical controls have not found significant differences in early survival with the use of the RV-PA conduit.[7-9] Most recently, contemporary comparison of the immediate hemodynamics after S1R with both shunt types in a randomized study did not demonstrate benefit of one shunt type versus the other.[10]

Using contemporary controls, we recently demonstrated no difference in surgical mortality, time to extubation, or length of hospital stay in 149 infants undergoing S1R for HLHS or variants palliated with either an mBTS or an RV-PA conduit.[8] We undertook the current study to compare the midterm hemodynamics and outcomes for the 2 shunt strategies at stage 2 reconstruction (S2R). The current study objectives include defining the difference between the 2 shunt groups with regard to (1) preoperative hemodynamic parameters, as defined by cardiac catheterization and echocardiography; (2) angiographic pulmonary artery size and architecture at the S2R; (3) age and degree of illness (as defined by anticongestive medication use, use of home oxygen, and tube feedings) at the time of S2R; and (4) stage 2 morbidity and mortality.

Materials and Methods

This study is a cross-sectional case series including all patients who underwent an S1R for HLHS or variants at The Children's Hospital of Philadelphia between January 1, 2002, and May 1, 2005 (n = 176), with attention paid to S1R hospital survivors (n = 138). This is a consent-waived study approved by The Children's Hospital of Philadelphia Institutional Review Board. Sources of information included the hospital medical records, the cardiac center and cardiac intensive care databases, records and reports provided by referring cardiologists, and pertinent information obtained from the parents on telephone interview.

We reviewed the initial, pre-S2R and pre–stage 3 reconstruction (S3R) echocardiographic reports of the study patients. Qualitative measurements of systolic ventricular function, atrioventricular valve regurgitation (AVVR), and neoaortic valve regurgitation were recorded.

TABLE 1. Single-ventricle anatomy among patients with stage 1 reconstruction shunt surgical technique

Cardiac anatomy	BTS (n = 114)	RV-PA (n = 62)	*P* value
Usual hypoplastic left heart syndrome	75 (66%)	54 (87%)	.004
Malaligned atrioventricular canal defect	12 (10.5%)	4 (6.5%)	.53
Double-outlet right ventricle with mitral or aortic stenosis	9 (8%)	3 (4.9%)	.65
Single left ventricle	6 (5%)	1 (1.6%)	.16
Other*	12 (10.5%)	0	.01

Data are counts and percentages within groups. Comparison is with the χ^2 test. *BTS*, Blalock–Taussig shunt; *RV-PA*, right ventricle–pulmonary artery conduit. *Nine cases of double-inlet left ventricle with transposition of the great arteries and aortic stenosis or interrupted aortic arch, 1 interrupted aortic arch with aortic stenosis and ventricular septal defect, 1 tricuspid stenosis with transposition of the great arteries, and 1 L-transposition of the great arteries with aortic stenosis and single left ventricle.

Oximetry, hemodynamic, and angiographic data were obtained at cardiac catheterization before S2R after achievement of conscious sedation or general anesthesia. Measured data included venous and arterial saturations, common atrial pressure, pulmonary artery pressure or pulmonary venous wedge pressure, and aortic pressure. Calculated data included arteriovenous oxygen difference and coronary perfusion pressure. Angiographic measurements were obtained from the anteroposterior or anterior oblique projections offline by a single observer to assess branch pulmonary artery anatomy and growth. The Nakata index was calculated as previously described.[11] The narrowest portion of the proximal branch pulmonary artery within 1 cm of the shunt insertion was measured by using standard offline techniques and compared with the vessel diameter just before the take-of the lobar branches to assess pulmonary artery stenosis. The ratio of these 2 measurements was used as a measure of severity of pulmonary artery stenosis and was graded as follows: moderate-to-severe stenosis, ratio less than 0.65; mild stenosis, ratio 0.66 to 0.85; and no stenosis, ratio greater than 0.85. The appearance of the branch pulmonary arteries was also recorded to ascertain the degree of pulmonary artery distortion qualitatively. The appearance was graded as follows: normal, diffusely narrowed, or discretely narrowed at the shunt or conduit insertion.

The operative technique for S1R has been previously described.[8] There was a surgical preference for the use of the RV-PA conduit in aortic atresia and the mBTS for lesions with a single left ventricle (Table 1). S2R consisted of either a bidirectional Glenn or hemi-Fontan procedure. Patients with bilateral superior vena cavae underwent either the bilateral bidirectional Glenn procedure or the right hemi-Fontan and left bidirectional Glenn procedure. The selection of a bidirectional Glenn or hemi-Fontan procedure was at the discretion of the surgeon. Patients with heterotaxy syndrome with interrupted inferior vena cavae underwent the Kawashima procedure. Additional procedures, such as tricuspid valve annuloplasty or valvuloplasty, were documented. Age and size at S2R and comparative operative data, including additional cardiac