Midterm Follow-Up After Biventricular Repair of the Hypoplastic Left Heart Complex

Jan Erik Freund, BS, Martijn H. T. den Dekker, MD, A. Christian Blank, MD, Felix Haas, MD, and Matthias W. Freund, MD

Departments of Pediatric Cardiology and Pediatric Cardiac Surgery, Wilhelmina Children's Hospital, University Medical Centre, Utrecht, Netherlands

Background. In neonates with hypoplastic left heart complex (HLHC), biventricular repair is considered superior to univentricular repair. The Z-scores of the mitral and the aortic valve annulus are primary factors for the choice of repair. Predictive cutoff values for the feasibility and optimal outcome of biventricular repair are unknown. This study assesses the growth of left side heart structures and the midterm outcome after biventricular repair with an interatrial fenestration in our HLHC population.

Methods. Retrospective study of 19 HLHC patients who underwent biventricular repair in a single tertiary referral center between 2004 and 2013. The cardiac dimensions (mitral and aortic valve annulus, left ventricle inlet length, left ventricular internal diastolic dimension) were measured before and at 6, 12, 24, and 48 months after biventricular repair.

Results. The follow-up ranged from 2 to 98 months. There was no early mortality, and the midterm survival rate was 95%. One patient died of a noncardiac- and nonintervention-related cause. Seven patients (37%) required a total of 8 reinterventions because of recurring or residual obstructive lesions. After biventricular repair, the left cardiac structures grew significantly.

Conclusions. Neonatal biventricular repair is successful and safe in HLHC patients, even with preoperative mitral and aortic valve annulus Z-scores of −4.5 and −5.5, respectively. Residual or restenosis required reintervention in 37% of our HLHC population, but was not significantly correlated with the magnitude of the preoperative Z-scores. Within the first 6 months of follow-up, the Z-scores almost normalized.

(Ann Thorac Surg 2015;■:■–■)

The hypoplastic left heart complex (HLHC) describes an entity of severe coarctation of the aorta and borderline forms of the hypoplastic left heart syndrome that is favorable for biventricular repair [1]. Hypoplastic left heart complex is defined by severe underdevelopment of the left heart–aorta complex, an aortic valve (AoV) or mitral valve (MV) annulus Z-score of less than −2, with absence of significant valvar or subvalvar aortic and mitral stenosis, but with a coarctation or aortic arch hypoplasia, and no significant ventricular septum defect [1–3]. For the Z-score calculation of the MV and AoV annulus, the method of Peterson and associates [4, 5] should be applied. Functionally, HLHC shows continuous antegrade flow in the aortic arch and ductal dependency with bidirectional ductal flow [6–8]. The etiology of HLHC is unclear. Obstruction of left ventricular inflow due to a restrictive foramen ovale or a persistent left superior vena cava may favor the developing of HLHC [9–14].

Neonatal biventricular repair is considered superior to the univentricular approach in HLHC [1, 6, 8, 13]. The biventricular approach consists of an aortic arch reconstruction with an patch enlargement of the aorta up to the sinus of Valsalva, with optionally leaving a small interatrial fenestration. The Z-score of the MV and the AoV annulus are the primary factors in the decision making for the surgical approach [5, 15, 16]. However, a cutoff value to indicate the feasibility of biventricular repair with optimal outcome has not been determined. Moreover, it is unclear how the preoperative cardiac dimensions influence the outcome. The cardiac performance, growth of the left ventricle, and the MV and the AoV annulus have been observed during short-term and midterm follow-up, but the results are divergent [2, 17–19].

In our cohort, the Z-scores of the MV and AoV annulus, the left ventricle (LV) length, and the left ventricular internal diastolic dimension at end diastole (LVIDd) were used to assess the growth of left side heart structures. Survival and the need of reinterventions were analyzed to evaluate the short-term and midterm outcomes of biventricular repair and to examine possible correlations with the preoperative cardiac dimensions.

Patients and Methods

Study Design and Study Population

At the Wilhelmina Children's Hospital (University Medical Center Utrecht, Netherlands), between 2004 and 2013,

Accepted for publication Feb 10, 2015.

Address correspondence to Dr Freund, Department of Pediatric Cardiology, Wilhelmina Children's Hospital, University Medical Center, KG 01.319.0/PO Box 85090, 3508 AB, Utrecht, Netherlands; e-mail: freund.matthias@klinikum-oldenburg.de.

0003-4975/$36.00
http://dx.doi.org/10.1016/j.athoracsur.2015.02.030

Table 1. *Patient Characteristics*

		Z-Scores Before Surgery											
Pt. No.	Diagnosis	MV	AoV	LV Length	LVIDd	Age at Surg (days)	BSA at Surg	Type of Surgery	RT After Surg (days)	ICU After Surg (days)	Total Follow-Up (months)	Reintervention[a]	Outcome[b]
1	HLHC	−3.74	−4.13	−2.31	−2.57	10	0.21	AAR, ASD 4 mm, UCS, ACP, OC	19	99	97.5	Supravalvular membrane/ subaortic fibrous tissue (22 months) & myomectomy (78 months)	II
2	HLHC	−4.51	−1.30	−3.38	1.40	8	0.20	AAR, ASD 3 mm, ACP, DHCA, OC	5	7	83.5	Balloon angioplasty re-CoA (7 months)	I
3	HLHC	−1.89	−2.76	−5.42	−1.96	7	0.21	AAR, ASD 3.5 mm, ACP, DHCA, OC	3	4	90.5	Patch angioplasty AAO (10 months)	I
4	HLHC, small pm VSD	−3.23	−2.55	−2.84	−5.11	5	0.18	AAR, ASD 4 mm, VSD closure, ACP, OC	12	14	79.5	None	I
5	HLHC, small pm VSD	−2.73	0.14	−0.78	−1.85	8	0.22	AAR, ASD 2 mm, VSD patch, ACP, CC	2	8	45.0	None	I
6	HLHC	−2.54	−0.71	−3.92	−1.80	17	0.21	AAR, ASD 3 mm, ACP, CC	3	5	49.0	None	I
7	HLHC	−3.33	−5.58	−2.26	−3.44	11	0.14	AAR, ASD 2 mm, ACP, DHCA, OC	4	6	52.0	Balloon angioplasty re-CoA (4 months)	I
8	HLHC	−3.08	−4.43	−2.78	−3.36	9	0.20	AAR, ASD 2 mm, DHCA, OC	4	6	50.0	None	I
9	HLHC, small muscular VSD	−2.25	−3.37	−2.38	−5.82	14	0.20	AAR, ASD 2.5 mm, VSD closure, DHCA, OC	3	5	49.0	None	I
10	HLHC	−3.65	−4.66	−2.16	−5.67	8	0.18	AAR, ASD 2 mm, ACP, DHCA, CC	4	6	43.0	Subaortic fibrous tissue (18 months)	I
11	HLHC	−4.12	−2.67	−3.49	−4.59	9	0.19	ARR, ASD 2 mm, ACP, OC	4	13	32.5	Balloon angioplasty re-CoA (8 months)	I
12	HLHC	−3.64	−2.12	−3.45	−3.74	14	0.21	AAR, ASD 2 mm, ACP, CC	3	4	34.0	None	I
13	HLHC	−2.33	−3.24	0.15	−3.66	14	0.20	AAR, ASD 2 mm, ACP, OC	7	19	23.0	None	I
14	HLHC	−4.24	−3.14	−3.57	−4.70	10	0.18	AAR, ASD 2 mm, UCS, DHCA, OC	3	4	26.5	None	I
15	HLHC, small pm VSD	−2.65	−2.51	−1.42	−2.10	5	0.21	AAR, ASD 2 mm, ACP, OC	13	17	4.0	None	I
16	HLHC, 2 small muscular VSD	−1.79	−2.59	−2.79	−3.41	13	0.20	AAR, ASD 2 mm, VSD closures, ACP, OC	6	8	5.0	None	I

(*Continued*)