Biventricular Repair With the Yasui Operation (Norwood/Rastelli) for Systemic Outflow Tract Obstruction With Two Adequate Ventricles

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Background. **The Yasui procedure is employed in neonates with interrupted aortic arch and left ventricular outflow tract obstruction (IAA/LVOTO) or aortic atresia-severe stenosis with ventricular septal defect (AA/VSD) and 2 adequate-sized ventricles. This combines a Norwood arch reconstruction with a Rastelli operation establishing a biventricular repair.**

Methods. **From 2002 to 2011, 21 neonates aged 3 to 55 days (mean 12.2 days, median 7 days) had IAA/LVOTO (n = 13), AA/VSD (n = 7), or AA/IAA with aortopulmonary window (n = 1); ten (48%) had genetic abnormalities (8 with DiGeorge syndrome). Based on clinical characteristics and surgeon preference, 6 had a primary Yasui repair (4 AA/VSD, 2 IAA/LVOTO); 15 were staged with an initial Norwood repair (3 AA/VSD, 12 IAA) followed by Yasui completion in 13 (2 await completion) 4.3 to 26.6 months later (median 6.9 months).**

Results. **Early mortality was zero with no interstage deaths in the staged patients. One patient died 2 months after staged repair. Since biventricular repair, 8 survivors (44%) had reoperation for conduit replacement (n = 6), recurrent LVOTO (n = 1), or a residual VSD (n = 1). No patient requires a pacemaker. There were 3 late deaths after biventricular repair, all in patients with genetic syndromes and IAA/LVOTO. Actuarial survival after initial operation was 100% at 1 year and 75% at 5 years. Actuarial freedom from reoperation or death after biventricular repair was 14% at 5 years.**

Conclusions. **The Yasui operation is effective for patients with IAA/LVOTO and AA/VSD. Primary and staged repair have comparable results. Reoperation after biventricular repair seems inevitable, mostly for conduit replacement. Genetic factors may affect long-term survival.**

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Interrupted aortic arch and severe left ventricular outflow tract obstruction (IAA/LVOTO) or aortic atresia with a ventricular septal defect (AA/VSD) and 2 well-formed ventricles presents surgical challenges. An early success was a staged approach described by Norwood and Stellin in 1981 [1] for a newborn with interrupted aortic arch, VSD, and AA. Initially, a valved conduit from the left ventricle to the descending aorta was placed with an 8-mm descending to ascending aortic conduit and pulmonary artery banding. One month later, the repair was completed with VSD closure and pulmonary artery band removal.

A one-stage primary repair of IAA/LVOTO was first reported in 1987 by Yasui and colleagues [2]. Two patients had an 8-mm ascending aorta to descending aorta interposition graft, a Damus-Stansel-Kaye connection of the proximal pulmonary trunk to the small ascending aorta, closure of the VSD channeling the left ventricular outflow through the VSD to the pulmonary valve, and construction of a right ventricle-to-pulmonary artery conduit (RV-PA) with a 14-m Hancock valve (Medtronic, Minneapolis, MN). The concepts of the Yasui operation (redirection of left ventricular outflow through the VSD to the pulmonary valve, placement of a conduit from the right ventricle to the pulmonary circulation) were also applied to infants with AA/VSD as a primary repair, initially with a conduit from the pulmonary trunk (the new systemic outflow) [3, 4], and then with a Norwood-type arch reconstruction [5–7]. In addition to use as a primary neonatal repair, the Yasui operation has also been employed as a staged repair both for IAA/LVOTO [8–12] and for AA/VSD [6, 12].

With this background in mind, we reviewed our experience with 21 infants from 2002 to 2011 with either IAA/LVOTO or AA/VSD for whom the Yasui operation was applied, either as a primary or as a staged repair.

Patients and Methods

Approval for this retrospective study was obtained from the Emory University School of Medicine Human Investigation Committee and the Institutional Review Board of

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Abbreviations and Acronyms

AA/VSD = aortic atresia or severe aortic valve hypoplasia with two well-formed ventricles and a ventricular septal defect
BT = modified Blalock-Taussig shunt
CoA = coarctation
DSK = Damus-Stansel-Kaye anastomosis
IAA/LVOTO = interrupted aortic arch with a ventricular septal defect and left ventricular outflow tract obstruction
LVOTO = left ventricular outflow tract obstruction
RV-PA = right ventricular to pulmonary artery
VSD = ventricular septal defect

Children's Healthcare of Atlanta. The need for patient or family consent was waived.

Patient Population

From January 2002 to October 2011, 21 neonates presented with either interrupted aortic arch and severe left ventricular outflow tract obstruction (IAA/LVOTO; n = 13), aortic atresia or severe aortic valve hypoplasia (aortic annulus < 3 mm) with a VSD and 2 well-formed ventricles (AA/VSD; n = 7) or interrupted aortic arch with aortic atresia, VSD, and aortopulmonary window (n = 1). All had 2 well-formed ventricles. Table 1 shows the patient characteristics at the time of the initial operation. The age at initial operation was similar between the IAA/LVOTO patients (9.3 ± 1.8 days) and the AA/VSD patients (13.4 ± 18.4 days; $p = 0.41$), as was the weight (3.0 ± 0.45 vs 3.1 ± 0.39 kg; $p = 0.71$).

Thirteen patients had IAA/LVOTO; 11 with type B IAA, 1 with type A (patient 8 with an aortic valve annulus diameter of 4.1 mm (z score = −3.15) and a LVOT diameter of 4.0 mm), and 1 with type C. Eight patients (62%) had an aberrant retroesophageal right subclavian artery (Table 1). Nine of the 13 IAA/LVOTO patients (69%) had a genetic syndrome; 8 with 22q11 microdeletion (DiGeorge syndrome) and 1 with mosaic trisomy 22. The diameter of the aortic annulus measured by echocardiography ranged from 2.1 to 5.1 mm (mean 4.0 mm), with a mean calculated z score of −4.2 (range −2.6 to −8.2). The subaortic diameter ranged from 2.0 to 4.7 mm (mean 3.6 mm). Only 1 of the 13 IAA/LVOTO patients (patient 4) had an aortic valve z score larger than −3.0. During the same time, 44 other neonates had conventional primary biventricular repair of interrupted aortic arch and VSD involving closure of the VSD with or without left ventricular outflow tract resection and repair of the interrupted arch.

Seven patients with a VSD and 2 well-formed ventricles had either severe LVOT obstruction defined as an

Table 1. Patient Characteristics at the Time of Original Operation

Patient Number	Age (Days)	Weight (kg)	Anatomic Group	Operative Strategy	Syndrome	IAA Type	Aberrant Right Subclavian
1	9	3.3	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
2	12	2.4	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
3	4	2.7	IAA/LVOTO	Staged	DiGeorge	Type B	No
4	7	3.5	IAA/LVOTO	Staged	None	Type B	No
5	5	3.5	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
6	8	3.4	IAA/LVOTO	Staged	None	Type B	No
7	3	2.8	IAA/LVOTO	Staged	None	Type B	No
8	17	3.1	IAA/LVOTO	Staged	None	Type A	No
9	7	3.5	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
10	4	2.6	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
11	27	2.5	IAA/LVOTO	Staged	DiGeorge	Type B	Yes
12	8	2.4	IAA/LVOTO	Primary	Mosaic trisomy 22	Type B	Yes
13	10	3.3	IAA/LVOTO	Primary	DiGeorge	Type C	Yes
14	10	3.3	Aortic hypoplasia/VSD	Staged	None	Tight CoA	Yes
15	6	2.3	Aortic hypoplasia/VSD	Staged	None	Tight CoA	Yes
16	7	3.1	Aortic atresia/VSD	Staged	None		Yes
17	7	3.5	Aortic atresia/VSD	Primary	None		No
18	55	3.1	Aortic atresia/VSD	Primary	None	Tight CoA	No
19	5	2.9	Aortic atresia/VSD	Primary	None		No
20	4	3.3	Aortic atresia/VSD	Primary	None		No
21	41	4.1	Aortic atresia/IAA/VSD/ aortopulmonary window	Staged	Cat Eye syndrome	Type B	Yes

CoA = coarctation of the aorta; IAA = interrupted aortic arch; IAA/LVOTO = interrupted aortic arch with left ventricular outflow tract obstruction; VSD = ventricular septal defect.