Midterm Results of the Modified Ross/Konno Procedure in Neonates and Infants

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Background. **The management of congenital aortic stenosis in neonates and infants continues to be a surgical challenge. We have performed the modified Ross-Konno procedure for patients who have severe aortic insufficiency or significant residual stenosis after balloon aortic dilation. The midterm results of this procedure were evaluated in this subset of patients.**

Methods. **Between 1994 and 2010, a total of 24 patients younger than 1 year of age underwent the modified Ross-Konno procedure. The diagnoses were aortic stenosis with or without subaortic stenosis (n = 16), Shone's complex (n = 7), and interrupted aortic arch with subaortic stenosis (n = 1). The aortic root was replaced with a pulmonary autograft, and the left ventricular outflow tract (LVOT) was enlarged with a right ventricular infundibular free wall muscular extension harvested with the autograft.**

Results. **Age at operation ranged from 1 to 236 days (median 28 days). The median follow-up period was 81 months (range 1–173 months). There was 1 early death and no late mortality. Overall the 1-, 2-, and 5-year survival rate was 95% ± 4.5%. Freedom from aortic stenosis was 94.7% ± 5.1% at 1, 2, and 5 years. Less than mild aortic insufficiency was 93.3% ± 6.4% at 2 years, and 74.7% ± 12.9% at 5 years. In total, 23 reoperations and reinterventions were performed; 14 were allograft conduit replacements. Two patients required aortic valve plasty. None required valve replacement. The reintervention-free rate was 64.6% ± 10.8% at 2 years and 36.9% ± 11.3% at 5 years.**

Conclusions. **Pulmonary autografts demonstrated good durability with low mortality and morbidity. This study shows that the modified Ross-Konno procedure can be a practical choice in selective cases for complex LVOT stenosis in neonates and infants.**

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Congenital aortic stenosis is often complicated by a hypoplastic annulus or multiple levels of left ventricular outflow tract (LVOT) obstruction. The management of these patients in the neonatal or infant period continues to pose significant challenges. Typically, patients who have significant stenosis are managed initially in a palliative fashion through infancy by repeated balloon aortic valvotomy (BAV) or palliative surgical valvotomy [1]. However, BAV can often result in significant aortic insufficiency or residual LVOT obstruction, which can result in left ventricular (LV) dysfunction or hemodynamic instability [2].

Furthermore, surgical options for this patient population are especially limited because of patient size and include (1) prosthetic valve (mechanical or bioprosthetic) replacement with or without annular enlargement, (2) conversion to single-ventricle physiology, (3) cardiac transplantation, and (4) aortic root replacement using allografts or autografts. The problems of aortic valve replacement are numerous, such as the availability of appropriate small-sized prosthetic valves, difficulty in managing anticoagulation, and the need for reoperation for valve replacement at a relatively early period in life because the prosthesis has no growth potential [3]. Conversion to single-ventricle physiology in patients with 2 adequately sized and functioning ventricles may not be optimal because of the need for multiple operations for the Fontan pathway and the lack of real long-term survival data after cavopulmonary palliation. Cardiac transplantation in neonates is limited owing to the availability of appropriate-sized donors, and the long-term outcome of neonatal cardiac transplantation is not satisfactory [4]. One alternative has been the use of an allograft for aortic valve replacement; however major questions exist regarding even short-term and midterm durability of these valves in children [5]. Moreover the lack of growth potential of the homograft valve remains a recurrent problem.

Hence in recent years, the Ross procedure has been advocated with a Konno annular enlargement using a pulmonary autograft [6, 7]. It has been proved that the

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Abbreviations and Acronyms

AS = aortic stenosis
BAV = balloon aortic valvotomy
ECMO = extracorporeal membrane oxygenation
EFE = endocardial fibroelastosis
IAA = interrupted aortic arch
LOS = low-output syndrome
LVOT = left ventricular outflow tract
MVP = mitral valve plasty
MVR = mitral valve replacement
RCA = right coronary artery
RVOT = right ventricular outflow tract
RV-PA = right ventricle to pulmonary artery

pulmonary homograft has some durability as well as growth potential in older children [8, 9]. We have reported the short- and midterm results of 11 children younger than 17 years of age [10]. However the efficacy of this technique in neonates and infants remains to be demonstrated.

To understand the utility of the Ross-Konno procedure in this population, we retrospectively reviewed a group of patients younger than 1 year of age who underwent the Ross-Konno procedure in our institution. Major outcomes studied included survival, freedom from reoperation/reintervention, neoaortic valve function, and homograft function.

Patients and Methods

Between May 1994 and August 2010, 24 children younger than 1 year underwent a modified Ross-Konno procedure. This study was approved by the Institutional Review Board at Stanford University. Medical records including patient charts, operative records, diagnostic reports, and outpatient clinic records were reviewed. Individual patient consent was waived because of the retrospective nature of the study. Preoperative and postoperative aortic insufficiency was qualitatively graded by color Doppler echocardiography using a scale of 0 (none) to 4 (severe). In the same period, 6 patients underwent Ross surgery without the Konno incision. Three patients with hypoplastic left heart syndrome who underwent Ross-Konno procedures because of social issues (families' strong requests) were excluded.

Surgical Technique

Standard neonatal and pediatric techniques of cardiopulmonary bypass, bicaval cannulation, and moderate hypothermia were used. None of the patients required deep hypothermic circulatory arrest. Myocardial protection was achieved by intermittent blood cardioplegia infusion.

A modified complete root replacement technique with annular enlargement and coronary transfer, described previously by our group, was used for the Ross/Konno procedure [11]. After initiation of cardiopulmonary bypass, the pulmonary autograft was harvested along with an attached extension of infundibular free wall muscle. After aortic cross-clamping and cardioplegia infusion, the aorta was transected at the level of the sinotubular junction. The coronary arteries were explanted with large coronary buttons comprising almost the entire wall of the sinuses of Valsalva. The remaining aortic root tissue was excised along with the aortic valve up to the native annulus. The interventricular septum was incised to the left of the right coronary artery in a fashion similar to that used in the Konno procedure. In patients with long-segment subaortic stenosis, the septal incision was extended beyond the obstruction. Additional endocardial fibroelastosis (EFE) resection and ventricular myectomy were performed if deemed necessary at this point. The pulmonary autograft was seated with the infundibular muscle extension fitting into the Konno incision in the interventricular septum. The autograft was then sutured to the native aortic annulus with a continuous 7-0 polypropylene suture starting at the posterior midpoint. This was then continued onto the infundibular muscle extension, which was sutured to the interventricular septal incision and reinforced with nonabsorbable, interrupted, pledgeted mattress sutures. The coronary ostia, with their entire sinus of Valsalva buttons, were then anastomosed to the appropriate sinuses of the autograft with a continuous 7-0 polypropylene suture. The proximal neoaorta was then sutured to the ascending aorta with a continuous polypropylene suture. The right ventricular outflow tract (RVOT) was reconstructed with an allograft valved conduit. The allograft was sutured directly to the right interventricular muscle. Patients were separated from bypass in the usual manner, and transesophageal or epicardial surface echocardiography was performed in all cases.

Statistical analysis was performed using Statview (SAS Institute, Cary, NC). Descriptive statistics are expressed as mean ± 1 standard deviation for continuous variables and as median (range) for categorical variables. From the follow-up data, the Kaplan-Meier method was used to determine survival curves. These survival estimates were then compared with a log-rank test. A p value less than 0.05 was considered significant.

Results

Patient Demographics

The patients ranged in age from 1 to 236 days (median, 28 days), with a median weight of 4.0 kg (range, 2.7 −7.7 kg). Fourteen patients were boys and 10 were girls.

The preoperative hemodynamic indication for the Ross/Konno procedure was critical aortic stenosis with or without subvalvular stenosis in 16 patients, Shone's complex in 7 patients, and interrupted aortic arch with ventricular septal defect in 1 patient. Aortic valve size was 5.0 ± 2.1 mm (z score, −3.0 ± 2.0), and 19 cases were bicuspid, 3 cases were unicuspid, and 2 cases were tricuspid. Pulmonary valve size was 11 ± 2.1 mm. Fourteen patients had greater than mild aortic insufficiency. The mean LVOT pressure gradient was 47 ± 24.8 mm Hg.