Early Results of the Ross Procedure in Simple and Complex Left Heart Disease

Bradley S. Marino, MD, MPP; Gil Wernovsky, MD; Jack Rychik, MD; John R. Bockoven, MD; Rodolfo I. Godinez, MD, PhD; Thomas L. Spray, MD

Background—The Ross procedure has been used increasingly to treat aortic valve disease in children and young adults. Benefits include the lack of anticoagulation after surgery and the potential growth and durability of the autograft. The purpose of this study was to review our institutional experience with the Ross procedure and to compare early outcome in simple aortic valve disease and complex left heart disease.

Methods and Results—Between January 1995 and October 1998, 66 patients (median age, 10.8 years; range, 6 days to 34.8 years) underwent the Ross procedure. The primary indication for surgery was isolated valvular disease in 41 patients: aortic stenosis (AS; n=3), aortic insufficiency (AI; n=11), and AS/AI (n=27). The remaining 25 patients had multiple levels of left ventricular outflow tract obstruction, 12 of whom had at least moderate AI. Additional left heart disease in the complex group included subaortic stenosis (n=20), arch obstruction (n=7), mitral valve disease (n=5), apical aortic conduit stenosis or insufficiency (n=3), and supravalvar AS (n=2). There were 123 prior interventions performed in 51 patients, including aortic valvotomy/valvuloplasty (n=56), coarctation repair (n=21), subaortic stenosis resection/Konno procedure (n=10), ventricular septal defect closure (n=8), apical aortic conduit placement (n=3), aortic valve replacement (n=3), and other (n=22). An isolated Ross procedure was performed in 41 patients, 10 of whom required concurrent aortic annulus enlargement procedure to accommodate the larger pulmonary autograft. In the remaining 25 patients, 49 concurrent procedures were performed, including the Konno procedure (n=17), aortic annulus enlargement (n=2), subaortic membrane resection (n=9), arch augmentation (n=5), mitral valvuloplasty (n=5), ventricular septal defect closure (n=4), apicoaortic conduit division (n=3), and other (n=4). One patient (1.5%) died 3 days after a Ross-Konno procedure, which included arch reconstruction, from presumed arrhythmia. There were no other early deaths. One patient required ECMO (extracorporeal membrane oxygenation) for 3 days after a ventricular tachycardia (VT)–related cardiac arrest. Transient complete heart block was seen in 4 patients; the duration was <5 days. No patient had left ventricular outflow tract obstruction on discharge echocardiography. Neo-AI was graded as none (n=5), trivial-mild (n=57), or moderate (n=3). All 3 patients with moderate neo-AI at discharge had abnormal pulmonary valves before surgery. Perioperative VT was noted in 18 patients (27.2%), 2 of whom were discharged on arrhythmia medication.

Conclusions—The Ross procedure can be performed in isolation or in combination with other complex procedures with low mortality (1.5%) and acceptable short-term results, even in patients with complex left heart disease and multiple prior interventions. Postoperative VT is common. Anatomic abnormalities of the pulmonary valve preclude its use as an autograft. (Circulation. 1999;100[suppl II]:II-162–II-166.)

Key Words: aorta ■ valves ■ heart defects, congenital ■ pediatrics

The Ross procedure, first described in 1967,1 involves replacement of the diseased aortic valve with a pulmonary autograft and placement of a pulmonary or aortic homograft between the right ventricle and the main pulmonary artery. After the Ross procedure, patients do not require anticoagulation, and the autograft has been shown to be durable and to grow in proportion to somatic growth.2,3 Because of these attributes, the pulmonary autograft is an attractive alternative to mechanical, porcine, and homograft valves in the treatment of aortic valve disease in children and young adults. Although mechanical valves provide a satisfactory hemodynamic result, they frequently require replacement during somatic growth and lifelong anticoagulation. Thromboembolism and hemorrhage remain important complications, especially in the pediatric population.4 Porcine bioprostheses, which do not require anticoagulation, deteriorate rapidly in young patients, and have limited durability.5 Although aortic valve homograft placement generally results
in excellent postoperative hemodynamics, does not require anticoagulation, and is associated with a low incidence of thromboembolic phenomena, these homografts also have limited durability in the pediatric population and do not grow with the child.6–8

Several reports have documented the effective use of the Ross procedure for isolated aortic valve disease in children.9–12 Recent reports have extended the pulmonary autograft to children with complex left ventricular outflow tract obstruction13–16 and to neonates and infants. 13,17

The purpose of this study was to review our institutional experience with the Ross procedure and to compare the early outcome in simple aortic valve disease to complex left heart disease.

## Methods

### Study Design

This study is a case series focusing on early surgical results. All patients at the Children's Hospital of Philadelphia who underwent the Ross procedure in isolation or in combination with other interventions between January 1995 and October 1998 were included in this investigation. Charts were reviewed for age at surgery, sex, original anatomic diagnoses, and prior interventions. Prospective data were obtained during the hospital stay. Operative notes were reviewed for additional surgical procedures and the duration of cardiopulmonary bypass, myocardial ischemia, and circulatory arrest. The perioperative course was reviewed for conduction abnormalities and arrhythmias, duration of ventilation, neurological sequelae, and other postoperative issues. Early morbidity was defined as events occurring <30 days from operation. Echocardiography at discharge was reviewed to assess for neo-aortic insufficiency, pulmonic insufficiency, pericardial effusion, and the presence of left or right ventricular outflow tract obstruction.

### Patient Population

There were 66 patients, 45 male and 21 female patients, included in the study. The median age of the group was 10.8 years at the time of surgery (range, 6 days to 34.5 years). The frequency distribution of age at Ross procedure shown in Figure 1 reveals that most patients underwent surgery between 5 and 18 years of age.

At the time of the pulmonary autograft, patients were stratified into simple and complex groups. Patients were defined as having simple aortic valve disease if they had aortic stenosis, insufficiency, or both with no other structural abnormalities present. Patients were defined as having complex left heart disease if they had aortic stenosis, insufficiency, or both and multiple levels of obstruction or additional hemodynamic abnormalities that required surgical intervention.

### Simple Aortic Valve Disease

There were 41 patients identified with simple aortic valve disease. The median age for the group was 14.6 years (range, 1.3 to 26.5 years). Isolated aortic stenosis or insufficiency was the predominant initial diagnosis. For 10 of these patients, the pulmonary autograft was their first intervention. Prior procedures (n=46) were performed in 31 of the 41 patients (Table 1).

### Complex Left Heart Disease

There were 25 patients with complex left heart disease. The median age for this group was 6.5 years (range, 6 days to 34.8 years). For 5 of the patients in this group, the pulmonary autograft was their first intervention. Seventy-seven prior procedures were performed in the other 20 patients with complex left heart disease (Table 2).

### Surgical Indications

Most patients had a combination of aortic stenosis and aortic insufficiency at the time of the pulmonary autograft (Figure 2). At least moderate aortic insufficiency was present in 53 of 66 patients (80%) at the time of surgery. Additional left heart disease in the complex group is described in Table 3.

### Surgical Technique

The Ross procedure, as previously described,18 was performed in 41 patients with isolated aortic valve disease, 10 of whom required a concurrent aortic annulus enlargement procedure to accommodate the size of the larger pulmonary autograft. Twenty-five patients with

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**Table 1. Simple Aortic Valve Disease**

<table>
<thead>
<tr>
<th>Original anatomic diagnoses, n</th>
<th>Aortic stenosis</th>
<th>Aortic insufficiency</th>
<th>Aortic stenosis/aortic insufficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior procedures (46 in 31 patients), n</td>
<td>Surgical valvotomy</td>
<td>Balloon valvotomy</td>
<td>Surgical valvuloplasty</td>
</tr>
<tr>
<td>------------------------------</td>
<td>----------------</td>
<td>---------------------</td>
<td>-------------------------------------</td>
</tr>
<tr>
<td>n=41.</td>
<td>16</td>
<td>13</td>
<td>8</td>
</tr>
</tbody>
</table>

**Table 2. Complex Left Heart Disease**

<table>
<thead>
<tr>
<th>Original anatomic diagnoses, n</th>
<th>Aortic stenosis/coarctation of the aorta</th>
<th>Aortic stenosis</th>
<th>Interrupted aortic arch</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior procedures (77 in 20 patients)</td>
<td>Surgical valvotomy</td>
<td>Balloon valvotomy</td>
<td>Subaortic stenosis resection</td>
<td>Apical aortic conduit placement</td>
</tr>
<tr>
<td>------------------------------</td>
<td>------------------------------------------</td>
<td>----------------</td>
<td>------------------------</td>
<td>-------</td>
</tr>
<tr>
<td>n=25.</td>
<td>10</td>
<td>7</td>
<td>4</td>
<td>4</td>
</tr>
</tbody>
</table>
complex left heart disease underwent the Ross procedure and 49 concurrent procedures (Table 4). In all 3 patients with apical-aortic conduits, a median sternotomy and left thoracotomy were used for takedown of the apical aortic conduit. Valved pulmonary homograft was used in all patients to reconstruct the right ventricular outflow tract.

Statistical Analysis
Summary statistics are expressed as medians and ranges. Statistical differences were assessed by the Wilcoxon rank-sum test.

Results

Mortality
There was 1 death in the series (1.5%), in a 6½-year-old male patient born with aortic stenosis and arch obstruction who had had 2 previous repairs of aortic coarctation and severe residual severe arch obstruction, aortic stenosis, and aortic insufficiency. The child also had left ventricular hypertrophy with a posterior wall thickness of 2 cm and marked coronary dilation (1 cm) at the time of surgery. Despite an apparently successful Ross-Konno procedure with arch reconstruction, the child had a sudden cardiac arrest on postoperative day 2 and could not be resuscitated.

Morbidity
In the simple group, 1 patient had a neo-aortic valve commissuroplasty, 1 had transient complete heart block, and 1 had a transient brachial plexus injury. Three patients were readmitted after discharge for pericardial effusion, 2 of whom required pericardiocentesis.

Perioperative morbidity was more frequent in the complex group. Three patients had reoperation for bleeding; 3 had transient complete heart block, none of whom required permanent pacemaker placement; and 2 had delayed sternal closure. One patient had a hyperkalemic cardiac arrest on postoperative day 2 and was successfully resuscitated with extracorporeal membrane oxygenation; the child sustained a cerebrovascular accident and has residual neurological deficits. One patient with interrupted aortic arch and aortic valvar and subvalvar stenosis who had surgical repair at an outside institution underwent stent placement in the right pulmonary artery because of compression of the right pulmonary artery by the ascending aorta. One patient with complex left heart disease was readmitted for pericardial effusion and had a pericardiocentesis performed.

Conduction abnormalities and arrhythmias for both simple and complex groups are summarized in Table 5. Eighteen patients had ventricular tachycardia in the postoperative period. Of the 18 patients, 16 had nonsustained ventricular tachycardia limited to the first 24 hours. Two patients had nonsustained ventricular tachycardia after postoperative day 1 and were discharged on antiarrhythmic medication. The perioperative conduction and rhythm disturbances have previously been summarized.19

The duration of mechanical ventilation, cardiac intensive care unit length of stay, and total hospital length of stay were significantly less in the simple group (Table 6). Notably, the median period to extubation in the complex group was 17 hours, and the median total hospital length of stay was 6 days.

Perfusion Data
The patients in this study had the entire operation performed, including the right ventricle to pulmonary artery homograft reconstruction, during a single period of aortic cross clamping. The median cross-clamp and cardiopulmonary bypass

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subaortic stenosis</td>
<td>20</td>
</tr>
<tr>
<td>Aortic arch obstruction</td>
<td>7</td>
</tr>
<tr>
<td>Mitral valve disease</td>
<td>5</td>
</tr>
<tr>
<td>Residual ventricular septal defect</td>
<td>4</td>
</tr>
<tr>
<td>AAC stenosis or valvar insufficiency</td>
<td>3</td>
</tr>
<tr>
<td>Supravalvar aortic stenosis</td>
<td>2</td>
</tr>
</tbody>
</table>

AAC indicates apical aortic conduit.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Konno procedure</td>
<td>17</td>
</tr>
<tr>
<td>Aortic annulus enlargement</td>
<td>2</td>
</tr>
<tr>
<td>Subaortic membrane resection</td>
<td>9</td>
</tr>
<tr>
<td>Arch augmentation</td>
<td>5</td>
</tr>
<tr>
<td>Mitral valvuloplasty</td>
<td>5</td>
</tr>
<tr>
<td>Ventricular septal defect closure</td>
<td>4</td>
</tr>
<tr>
<td>AAC division</td>
<td>3</td>
</tr>
<tr>
<td>Other</td>
<td>4</td>
</tr>
</tbody>
</table>

AAC indicates apical aortic conduit.

<table>
<thead>
<tr>
<th>Arrhythmia</th>
<th>Simple (n=41)</th>
<th>Complex (n=25)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVT</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>VT</td>
<td>10</td>
<td>24</td>
</tr>
<tr>
<td>CHB</td>
<td>1</td>
<td>2.5</td>
</tr>
<tr>
<td>Ventricular rhythm</td>
<td>1.25</td>
<td>3</td>
</tr>
<tr>
<td>Coronary artery</td>
<td>1.25</td>
<td>1</td>
</tr>
</tbody>
</table>

SVT indicates supraventricular tachycardia; VT, ventricular tachycardia; and CHB, complete heart block.
times were significantly shorter in the simple compared with the complex group. Circulatory arrest was used during arch reconstruction in 7 patients, with a median circulatory arrest time of 19 minutes (range, 10 to 32 minutes; Table 7). Of the 7 patients who had circulatory arrest, 5 had arch augmentation, whereas 2 neonates required circulatory arrest to permit reconstruction of the aortic arch.

**Discharge Echocardiography**

All patients underwent an echocardiogram before discharge. Greater than trivial-mild neo-aortic insufficiency was found in 3 of 65 patients at discharge. The grade distribution of neo-aortic insufficiency at discharge is shown in Table 8. The 3 patients who had moderate neo-aortic insufficiency at discharge all had structurally abnormal pulmonary valves at the time of the Ross procedure. One patient had supravalvar stenosis with 2 large pulmonic valve leaflets and 1 hypoplastic leaflet; 1 patient had previous pulmonary artery band placement, which resulted in pulmonary valve distortion; and 1 patient was noted to have a gap between 2 of the pulmonary valve commissures. No patient had residual left ventricular outflow tract or arch obstruction at discharge. Greater than trivial-mild neo-pulmonic insufficiency was found in 2 of 65 patients at discharge. The grade distribution of neo-pulmonic insufficiency at discharge is shown in Table 9. In addition, 1 patient had a pressure gradient of 20 mm Hg across the right ventricular outflow tract at discharge.

**Discussion**

This study found that the Ross procedure can be performed with a low mortality risk, 1.5%, in both simple and complex left heart disease. Compared with the simple group, the duration of mechanical ventilation and hospital length of stay was longer in patients with complex left heart disease. Neo-aortic insufficiency greater than trivial-mild was uncommon at discharge.

**TABLE 7. Perfusion Data**

<table>
<thead>
<tr>
<th></th>
<th>Simple (n=41)</th>
<th>Complex (n=25)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Median</td>
<td>Range</td>
<td>Median</td>
</tr>
<tr>
<td>Cross-clamp time, min</td>
<td>74</td>
<td>53–126</td>
<td>89</td>
</tr>
<tr>
<td>CPB, min</td>
<td>101</td>
<td>75–171</td>
<td>114</td>
</tr>
<tr>
<td>Circulatory arrest, min</td>
<td>19</td>
<td>10–32</td>
<td></td>
</tr>
</tbody>
</table>

CPB indicates cardiopulmonary bypass.

*P < 0.001.

**TABLE 8. Discharge Echocardiography: Neo-Aortic Insufficiency**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Simple (n=41)</th>
<th>Complex (n=24)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>2</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Trivial-mild</td>
<td>13</td>
<td>32</td>
<td>12</td>
</tr>
<tr>
<td>Moderate</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>0</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>

The mortality rate in this series (1.5%) is equivalent to that in prior reports. In general, children who are candidates for the Ross procedure have a myocardium “at risk” because of long-standing pressure and or volume overload and the intraoperative ischemia that results from surgical repair. These factors lead to ventricular dysfunction and ectopy (nonsustained ventricular tachycardia and nonsustained supraventricular tachycardia) in the early postoperative period. Reddy et al, who reported extending the Ross procedure to children with complex left heart disease, found isolated cases of transient atrial and ventricular dysrhythmias. Elkins et al reported 1 early death secondary to a fatal arrhythmia. Similarly, in a 20-year follow-up of a pulmonary autograft cohort by Matsuki and colleagues, there was 1 early and 1 late death attributed to arrhythmia. The higher incidence of identified perioperative ventricular tachycardia in this study may result from our methodology, incorporating 24-hour full-disclosure telemetry, but is also more likely to be attributable to the complexity of this cohort of patients. Most patients had multiple prior procedures, and many had concurrent additional surgical procedures performed at the time of autograft placement.

The incidence of complete heart block after the Ross procedure varies from 0% to 6%. In the present series, permanent, complete heart block did not occur, despite the fact that 19 of 25 patients (76%) within the complex group had an annulus-enlarging procedure. Although there was transient heart block in 4 patients within this cohort, all were discharged in normal sinus rhythm. Possible causes of complete heart block in this popula-

**TABLE 9. Discharge Echocardiography: Neo-Pulmonic (Homograft) Insufficiency**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Simple (n=41)</th>
<th>Complex (n=24)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>27</td>
<td>66</td>
<td></td>
</tr>
<tr>
<td>Trivial-mild</td>
<td>13</td>
<td>32</td>
<td>12</td>
</tr>
<tr>
<td>Moderate</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>0</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>
tion include damage to the septal perforating coronary artery during autograft harvest, suture placements during anastomosis of the proximal end of the autograft, resection of subaortic stenosis, and the Konno procedure for aortic annulus and left ventricular outflow tract enlargement.

Reoperation for mediastinal bleeding was required in 4.5% of our study group. All 3 patients who required reoperation had complex left heart disease. Reoperation for mediastinal bleeding was required in 8.5% of a cohort with complex left heart obstruction followed up by Reddy et al., in 4.0% of a cohort followed up by Elkins et al., and in 1.2% of patients in a group followed up by Matsuaki et al. Both Starnes et al. and Reddy et al. have reported results for the Ross procedure in patients with complex left heart disease with durations of mechanical ventilation and hospital length of stay similar to those of the present series. For the complex group within our cohort, the median duration of mechanical ventilation was 17 hours, median length of stay in the intensive care unit was 3 days, and median total hospital length of stay was 6 days.

Similar to other pulmonary autograft and aortic homograft series, discharge echocardiography revealed moderate neo-aortic insufficiency in very few (3 of 65) patients in our series, all of whom had abnormal pulmonary valves before the Ross procedure.

Long-term issues include autograft durability, autograft growth, homograft durability, and the long-term significance of perioperative arrhythmias. It is unclear whether use of the Ross procedure at an earlier age alters the natural history of simple or complex left ventricular outflow tract disease. It is also not known if the risk of late reoperation on the neo-aortic valve is higher if performed in the neonate, infant, or child because of aortic root sinuses dilation and valve distortion over time. Similarly, given the lack of growth of the cryopreserved homograft, late reintervention is likely with interval replacement of the homograft. Long-term changes in left ventricular mechanics await further study.

This study is limited in that it focuses solely on early surgical results. Long-term follow-up is underway to determine whether there is a significant difference in late outcome between simple and complex left heart disease groups and between neonates/infants and children/adolescents.

In conclusion, the Ross procedure can be used for simple aortic valve disease and complex left heart disease with low morbidity and mortality in children and young adults. Most patients had a short period of mechanical ventilation, cardiac intensive care unit length of stay, and total hospital length of stay. Significant neo-aortic insufficiency is uncommon in the short term.

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References
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