The Ross Procedure in Pediatric Patients (Midterm Result)

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Abstract

Objective: This study reviews our experience with the Ross procedure in pediatric patients at the Rajvithi Hospital.

Background: Aortic valve disease in children is a challenging problem in terms of surgical options. Although the Ross procedure provides freedom from anticoagulation and the growth potential of the pulmonary autograft, it is also known to have a future risk of autograft insufficiency and reoperation for the homograft on the right ventricular outflow tract.

Methods: From January 1998 to January 2007, 30 pediatric patients underwent the Ross procedure for aortic valve disease at the Rajvithi Hospital. Their ages ranged from 3 to 15 years (mean 8.3 ± 3.43 years; median 8 years). The operative technique was total aortic root replacement with pulmonary autograft. Reducing aortic annulus was made when the mismatch of the autograft and aortic root occurred. Pulmonary or aortic homografts were used for right ventricular outflow tract reconstruction.

Results: There was 1 hospital mortality (3.3%) in a patient with acute severe aortic regurgitation and active rheumatic carditis. There was no late mortality and the morbidity was limited to 1 reoperation for bleeding and 1 permanent pacemaker implantation. Echocardiograms performed at discharge demonstrated that no patient had more than a trace to 1+ aortic regurgitation and none had evidence of aortic stenosis. During the follow-up period, ranging from 43-103 months (mean 80.7 ± 19.1 months), the majority of patients had no or trivial to mild neoaoortic valve regurgitation up to 4 years of follow-up. Moderate to severe neoaoortic valve regurgitation was not seen in any patients up to 1 year. It was seen in 1 patient after 2 years, 2 patients at 3 years and 4 patients at 4 years of follow-up. Two patients with severe neoaoortic insufficiency underwent reoperations at 84 and 72 months after initial surgery to treat neoaoortic central regurgitation. Neoaoortic annular fixation and reduction of dilated sinus of valsava was done in 1 patient and aortic valve replacement in the other. None of the patients required intervention or reoperation for the homograft on the right ventricular outflow tract.

Conclusions: The Ross procedure is a safe and effective option for aortic valve replacement in pediatric patients. It also has a real potential to grow with the children and offers good midterm results. The risk factors for late autograft dysfunction are a preoperative diagnosis of aortic regurgitation and absence of aortic annular fixation in the initial Ross procedure. The incidence of autograft dysfunction and reoperation are relatively low (13.3% and 6.6%). Surgical correction can be performed in patients with central autograft insufficiency. Aortic valve replacement is inevitable in patients with severe autograft dilatation and valve degeneration.

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of our patients. This technique is very useful in pediatric patients with a small aortic annulus. The usual technique for right ventricular outflow tract reconstruction with a cryopreserved allograft has been widely accepted and performed in most centers around the world.

Methods
From January 1998 to January 2007, 30 children underwent the Ross procedure at the Rajvithi Hospital. The following data were collected retrospectively including operative findings, postoperative courses, length of hospitalization, mortality and morbidity. Echocardiograms were performed immediately before hospital discharge and then at a 6-12 month interval. Follow-up was carried out initially at 2 weeks and then at 3 month-intervals after discharge for the first year, then 6 month-intervals for the second year and then once a year. Follow-up was completed by a cardiac surgeon and pediatric cardiologist.

Surgical Techniques
Via a median sternotomy, cardiopulmonary bypass was established, using one aortic and two caval canulas in all patients. Moderate hypothermia with core cooling to 25 degree Celsius was established. The left ventricle was decompressed by the left atrial vent inserted via the right upper pulmonary vein. Myocardial protection consisted of topical hypothermia and hyperkalemic blood cardioplegia administered antegradely or retrogradely. Repeated doses of cardioplegia were given at 30-minute intervals throughout the operation.

After cross-clamping the aorta, the aortic valve was exposed and inspected through a transverse incision in the ascending aorta. If the aortic valve was severely deformed and not repairable, the pulmonary valve was then inspected through a transverse incision just above its bifurcation to see whether it was intact and suitable to be a neo-aortic valve. Measurement of the annulus size of both valves was done at this time. The pulmonary valve annulus was usually approximately the same size or slightly larger than the aortic valve annulus. If it was found to be more than 2 or 3 mm. smaller than the aortic valve annulus, an aortic root tailoring technique was considered (5).

The pulmonary artery was then transected at its bifurcation and harvested. The harvesting technique began with the incision in the anterior surface of the right ventricular outflow tract about 5 mm. below the pulmonary valve annulus and followed by scalloping and beveling the right ventricular myocardium around the pulmonary valve. Care was taken to avoid injury to the left main coronary artery behind the posterior commissure of the pulmonary valve. The area adjacent to the lateral commissure of the pulmonary valve was also dissected meticulously since the left anterior descending coronary artery and its first septal branch lied in close proximity. The pulmonary valve bed was then inspected and cauterized carefully to obtain homeostasis. The harvested autograft was then placed in cold normal saline while waiting for implantation.

Harvesting the pulmonary autograft in the patient with doubly-committed ventricular septal defect was quite a challenge when there was no subpulmonary conus muscle between the aortic and pulmonary valve. Care is taken not to injure the pulmonary autograft in the area of the kissing commissures of the two valves. Many times when the kissing commissures could not be separated surgically, we would excise part of the aortic wall along with the autograft.

In the patients with perimembranous (subaortic) ventricular septal defect, the defect was closed as usual with a Dacron patch. In patients with doubly-committed ventricular septal defect, the defect was closed by using a Gortex patch to recreate the complete aortic annulus to appropriate size.

In all patients, the pulmonary autograft was implanted as a total root replacement as mentioned above (4). The aorta was then transected a few millimeters above the sinutubular junction and the diseased aortic valve was excised. The coronary ostia were removed with buttons of aortic tissue. In patients with aortic regurgitation and discrepancy between the aortic and pulmonary valve annulus (aortic/pulmonary diameter ratio more than 1.0), the aortic annuloplasty was done to make the aortic and pulmonary autograft equal in order to prevent central regurgitation of the neoaortic valve (5). The aortic annuloplasty techniques we used depended on the size of the autograft. If the size of the autograft was close to adult size (more than 20 mm in diameter), the aortic annular reduction and fixation were done by using 2-0 polypropylene running in the aortic annulus and tied over the 20-mm Hegar dilator as described by Elkins (5).
the autograft was smaller than the usual adult size (less than 20 mm in diameter), aortic annular reduction only was performed by placation of the annulus in the NCC/LCC commissure area by using interrupted 2-0 or 3-0 polypropylene with pledget to reduce the size of the aortic annulus to equal the autograft size.

The autograft was then sutured to the aortic root with 4.0 polypropylene suture in continuous fashion with a strip of pericardium incorporated into the suture line. Care was taken to avoid injury to the autograft leaflets while suturing. The coronary artery buttons were then reimplanted onto the facing sinuses of the autograft with 6.0 polypropylene and the distal aortic anastomosis was then completed. The right ventricular outflow tract was reconstructed with an appropriate-sized cryopreserved aortic or pulmonary homograft beginning with the distal anastomosis. The proximal anastomosis of the homograft to the right ventricle was completed after removal of the aortic cross-clamp. Meticulous stitches on the conal septum were needed to avoid injury to the conducting bundles, left anterior descending coronary artery and its first septal branch lying in close proximity.

**Results**

Data were available from all 30 patients. All patients were operated on by a single surgeon (No.1). There was 1 hospital mortality (3.3%) in a patient with acute severe aortic regurgitation and active rheumatic carditis. There was no late mortality and morbidities were limited to 1 reoperation for bleeding and 1 permanent pacemaker implantation.

The median age of the children was 8 years (range, 3 to 15 years) and mean age was 8.3 ± 3.43 years. There were 17 males and 13 females. All patients underwent preoperative transthoracic echocardiographic evaluation. The diagnosis of preoperative aortic valve pathology is summarized in Figure 1 and other associated cardiac defects in Table 1. All patients were in NYHA class II or above preoperatively. The majority (25/30) of patients had aortic valve regurgitation from prolapse of one or more cusps of the aortic valve caused by either doubly committed ventricular septal defect (VSD) or subaortic VSD. Congenital bicuspid native aortic valve was seen in 3 patients and rheumatic valvulitis was seen in 2. Associated surgical procedures were done in all of the patients (Figure 2). The major associated procedure was closure of the VSD with patch in 25 patients.

Aortic/pulmonary diameter ratio was less than 1.0 in 10, between 1.01-1.20 in 12 and 1.21-1.40 in 8 (Figure 3).
Aortic annuloplasty (annular reduction ± fixation) was performed in 2/3 of the patients (20/30) when the Aortic/pulmonary diameter ratio was more than 1.0. The aortic annular fixation was only performed in 2 adolescent patients.

In 25 patients with a preoperative diagnosis of moderate to severe and severe aortic regurgitation (AR) in association with VSD, the intraoperative valve pathology is shown in Figure 4. The number of deformed valve leaflets seen intraoperatively is demonstrated in Figure 5.

Follow-up ranged from 43 to 103 months (mean 80.7 ± 19.1 months). There was no mortality during follow-up. Morbidities included 1 reoperation for bleeding and 1 permanent pacemaker implantation. Mean ICU stay was 19.3 ± 4.6 hours (range, 12-30 hours) and mean hospital stay was 9.6 ± 2.4 days (range, 7 to 14 days). One patient was lost to the follow-up after 48 months. She was the oldest patient in this study and was 15 years of age at the time of surgery. She underwent the Ross procedure, mitral valve repair and closure of PDA. This patient was doing well in NYHA class I during her last visit. Figure 6 shows the status of the neoaortic valve during follow-up. The majority of patients had no or trivial to mild neoaortic valve regurgitation up to 4 years of follow-up. Moderate to severe neoaortic valve regurgitation was not seen in any patients up to 1 year. It was seen in 1 patient after 2 years of follow-up, 2 patients at 3 years and 4 patients at 4 years of follow-up. All of these 4 patients had a preoperative diagnosis of VSD/AR.

In the group of 4 patients with moderate to severe aortic insufficiency, aortic annular reduction was done in 1 and none had aortic annular fixation. Two patients in this group underwent reoperations at 84 and 72 months after initial surgery. We were able to repair the neoaortic valve by performing annuloplasty and reduction of dilated sinus of valsava in 1 patient. Echocardiograms at discharge showed mild neoaortic regurgitation up to 1 year after reoperation. We had to perform aortic valve replacement with St.Jude aortic prostheses in the other patient due to severe deformity of the autograft leaflet. Both patients were in NYHA class I up to 1 year follow-up. None of the

### Table 1

This table shows preoperative associated cardiac defects in this group of patients.

<table>
<thead>
<tr>
<th>Associated anomalies</th>
<th>No. of patients</th>
</tr>
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<tbody>
<tr>
<td>VSD</td>
<td>9</td>
</tr>
<tr>
<td>Doubly committed</td>
<td>16</td>
</tr>
<tr>
<td>Subaortic</td>
<td>1</td>
</tr>
<tr>
<td>Tunnel type</td>
<td>1</td>
</tr>
<tr>
<td>Discrete type</td>
<td>3</td>
</tr>
<tr>
<td>MR</td>
<td>1</td>
</tr>
<tr>
<td>PDA</td>
<td>1</td>
</tr>
<tr>
<td>Rupture sinus of valsava</td>
<td>2</td>
</tr>
<tr>
<td>Abnormal coronary artery pattern</td>
<td></td>
</tr>
</tbody>
</table>

VSD = ventricular septal defect; MR = mitral regurgitation; PDA = patent ductus arteriosus

**Figure 3.** This chart shows the aortic/pulmonary diameter ratio measured intraoperatively.

Aortic annuloplasty (annular reduction ± fixation) was performed in 2/3 of the patients (20/30) when the Aortic/pulmonary diameter ratio was more than 1.0. The aortic annular fixation was only performed in 2 adolescent patients.

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Figure 4. This figure shows the aortic valve pathology which includes 2 valve leaflets on the left and 3 leaflets on the right. Deformity and distortion of leaflets which precludes repair is demonstrated.

Figure 5. This chart shows the number of deformed valve leaflets found intraoperatively in the group of patients with VSD/AR (RCC = right coronary cusp, NCC = non coronary cusp).

Figure 6. This chart shows the status of the neoaortic valve during follow-up. Four patients had moderate to severe regurgitation at 4-year follow-ups.

patients required intervention or reoperation for the homograft on the right ventricular outflow tract.

Discussion

Management of aortic valve disease in children is a challenge. Beside longevity and durability of the surgical options, the growth potential and quality of life of the children are also main concerns. We would choose to repair the diseased aortic valve whenever it is possible to repair and the repair should yield acceptable results. In the group of patients with severe deformity in more than 1 valve leaflet (Figure 4), replacement of the aortic valve is a better option.

Aortic valve replacement in pediatric patients is associated with a number of problems which significantly limits all kinds of available aortic valve prostheses currently in use from being the most suitable aortic valve replacement for these children. The mechanical valve prostheses have several disadvantages including size mismatch, the need for lifelong anticoagulation and absence of growth potential. Although the bioprostheses do not need long-term anticoagulation, it is known to be calcified much earlier in
children due to high calcium metabolism (6), and as a result, it needs early reoperation. Allograft prostheses (homograft) also provide freedom from anticoagulation and a solution to the small aortic annulus when inserted as a homograft root replacement. Early calcification of a homograft is also a main concern. Performing a repeated aortic root replacement for a calcified homograft is one of the most complicated reoperations in the future for the child. Although there was some evidence of a viable fibroblast in a cryopreserved homograft, it has never been proven to have the ability to grow.

The Ross procedure has become the operation of choice for pediatric patients with severe aortic valve disease who require aortic valve replacement during the past 10 years (7-10). It overcomes nearly all of those limitations mentioned above with a low operative mortality in a majority of the present series. The pulmonary autograft provides an autologous, viable tissue valve in the aortic position that also has the potential to grow, which could not be found in any other valve prostheses available these days (3). In addition, it provides freedom from thromboembolic complication associated with long-term anticoagulation. The durability of the autograft has also been documented in long-term studies conducted by many investigators (2, 11).

One of the main concerns for the Ross patients is neoaortic insufficiency or autograft dysfunction. In our group of patient, this problem occurred in 4 patients starting after the second year of follow-up as described above. Echocardiograms of these 4 patients showed the same abnormalities of neoaortic annular and sinus dilatation causing moderate to severe central regurgitation. The first patient was reoperated 84 months after the Ross procedure and found to have severe deformity of one of the autograft leaflets and a marked dilatation of neoaortic annulus and sinus. We decided to replace the autograft valve with a 23-HP St.Jude Medical aortic prostheses, due to the inability to achieve a good repair. The second reoperated patient was at 72 months after the Ross procedure. Neoaortic annular and sinus dilatation was found. The autograft leaflets were still nice and thin. We were able to repair the autograft by performing an annular reduction/fixation using 2-0 polypropylene running in the neoaortic annulus and sinus reduction using interrupted 3-0 polypropylene with pledget. The 3 cusps of the autograft were then able to come together and resume normal function. Intraoperative transesophageal echocardiogram revealed a mild regurgitation after the procedure.

The early reports of the Ross procedure showed a significant number of early autograft failure (1, 12). With the improvement of surgical techniques and better understanding of the surgical anatomy of the aortic and pulmonary root, operative mortality and early autograft failure have decreased significantly. The current preferred technique including ours is to insert an autograft as a root replacement (13). This technique maintains an anatomic unit and normal coaptation of the pulmonary valve.

Late autograft dysfunction has been described by several investigators with a varied incidence (14-16). All of our 4 patients with autograft dysfunction were in the VSD/AR preoperative category. None of them had a neoaortic annular fixation procedure done at the initial operation, because of small aortic annulus and need for further growth. We identified these 2 factors as our risk factors for late autograft dysfunction. Elkins and colleagues showed that autograft dysfunction was worse in patients with a primary lesion of aortic insufficiency than in those with aortic stenosis (8). Their data correlated well with ours in this aspect. They also demonstrated a lower incidence of autograft dysfunction with the root replacement technique and recommended aortic annulus reduction and fixation at the time of the Ross procedure in patients with significant discrepancy between the aortic and pulmonary valve annulus diameter or patients with annuloaortic ectasia (14). We employed this annular reduction technique in 20 patients with an aortic/pulmonary diameter ratio more than 1.0. Combined annular fixation technique was used in only 2 patients because of the need to preserve growth potential of the autograft in the rest of the group. Although initial aortic annular fixation may help prevent annulus dilatation, the progressive dilatation of the sinus portion of the autograft may not be prevented by this technique. Carrel and colleagues proposed an external reinforcement technique of the whole autograft by using a prosthetic Dacron graft with artificial aortic root configuration in order to prevent this late complication (17). They reported a good early result, but long-term follow-up is needed to see.
In terms of an operative procedure for autograft reoperation, repair of the autograft was always attempted at first. In the first reoperated patient, we were able to repair the autograft when central autograft insufficiency from annular and sinus dilatation was the main pathology. Annular fixation and reduction of the dilated sinus restored the autograft function, leaving only mild aortic regurgitation at discharge and one-year follow-up. In the second reoperated patient, there was a severe annular dilatation and degeneration of the autograft leaflet. We decided to replace the autograft valve with aortic prostheses since we felt that the repair procedure would not yield a satisfactory result. The homograft used to reconstruct the right ventricular outflow tract in the Ross procedure has proved to last longer in many series when compared to those used in the Rastelli operation (2, 8). In our group of patients, none of them had a significant gradient (more than 25 mmHg) on the homograft up to 103 months of follow-up.

Conclusions

The Ross procedure is a safe and effective for aortic valve replacement in pediatric patients. The growth potential of an autograft and freedom from anticoagulation has made the Ross procedure the best surgical option for children in this situation. The risk for autograft dysfunction which needs reoperation remains low. Preoperative diagnosis of aortic regurgitation and absence of aortic annular fixation in the initial Ross procedure are risks factors. Surgical repair can be performed in patients with central autograft insufficiency. Aortic valve replacement can not be avoided in patients with severe autograft dilatation and valve degeneration.

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References

การดำเนินการผ่าตัด Ross Procedure ในผู้ป่วยเด็ก (ติดตามผลระยะกลาง)

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บทคัดย่อ

วัตถุประสงค์: เพื่อรายงานผลการศึกษาการดำเนินการผ่าตัด Ross Procedure ของผู้ป่วยเด็กที่โรงพยาบาลราชวิถี การปรับปรุงการดำเนินการผ่าตัดฟื้นคืนชีพของ pulmonary autograft การตัดสินใจในการผ่าตัดซ้ำกับกับ homograft ที่ต้องหัวใจในอนาคต

วิธีการศึกษา: ตั้งแต่เดือนมกราคม 2541 ถึงเดือนมกราคม 2550 ผู้ป่วยจำนวน 30 รายได้รับการผ่าตัด Ross Procedure ที่โรงพยาบาลราชวิถีโดยมีอายุระหว่าง 3-15 ปี (mean 8.3 ± 3.43 ปี; median 8 ปี) โดยใช้วิธีการผ่าตัดแบบเปลี่ยน aortic root ทั้งอันดับ pulmonic autograft การลดขนาด aortic annulus จะกระทำเมื่อขนาดของ autograft และ aortic root ไม่เท่ากัน และใช้ pulmonary หรือ aortic homograft ให้เหมาะสมในหัวใจข้างขวา

ผลการศึกษา: มีผู้ป่วยเสียชีวิตในโรงพยาบาล 1 ราย (3.3%) โดยเป็นผู้ป่วยที่มีลิ้นหัวใจเอออร์ติกรั่วในузนาระหว่างสาเหตุ active rheumatic carditis ไม่มีผู้ป่วยที่มีโรคหลอดจากอากาศมาก่อนผ่าตัด การผ่าตัดครั้งแรก 1 ราย และมีผู้ป่วย 1 รายที่ต้องการผ่าตัดซ้ำในระยะหลัง การตรวจ echocardiogram ได้ผลการตัดสินใจการทำผ่าตัดซ้ำทั้งอันดับ pulmonic autograft การลดขนาด aortic annulus จะกระทำเมื่อขนาดของ autograft และ aortic root ไม่เท่ากัน และใช้ pulmonary หรือ aortic homograft ให้เหมาะสมในหัวใจข้างขวา

สรุป: การดำเนินการผ่าตัด Ross Procedure เป็นการดำเนินการผ่าตัดที่ปลอดภัยและเป็นทางเลือกที่ดีในการรักษาโรคหัวใจเอออร์ติกในกลุ่มผู้ป่วยเด็ก โดย pulmonary autograft สามารถใช้ได้ไปพร้อม ๆ กัน แต่การตัดสินใจการดำเนินการผ่าตัด futher ได้ผ่านการตัดสินใจที่จะดำเนินการผ่าตัด Ross Procedure ในระยะหลังการผ่าตัดเพื่อแก้ไขปัญหา pulmonary autograft ที่มีการรั่วของ aortic annular fixation ในกรณีที่มี pulmonic autograft และการดำเนินการผ่าตัดซ้ำที่มี pulmonic autograft และการดำเนินการผ่าตัดซ้ำที่มี homograft ที่ต้องหัวใจในอนาคต สามารถทำได้ด้วยการใช้แบบ central autograft insufficiency และการตัดสินใจที่มี severe autograft dilatation และการตัดสินใจที่จะเปลี่ยน homograft ที่ต้องหัวใจในอนาคต