Growth of the pulmonary autograft after the Ross operation in childhood

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Abstract

Objective: Excellent hemodynamic performance has been demonstrated after aortic valve replacement using the autologous pulmonary valve as described by D. Ross. However, in the pediatric population there is concern in regard to growth of the autograft and late dilatation in the systemic circulation. Methods: Since 1991, 30 children (mean age, 11.3 ± 3.1 years) had aortic valve replacement with the pulmonary autograft as a root replacement. All children had yearly clinical and echocardiographic follow-up. Results: There were no perioperative deaths; one child died late in a car accident. At the last follow-up (mean follow-up, 4.3 ± 2.6 years), all patients were in NYHA class I. There was one early reoperation, in which the autograft had to be reconstructed due to a leaflet perforation. There were no major valve related events. All children showed normal somatic growth. The annulus diameter increased significantly from 18 ± 2 at surgery to 20 ± 3.5 mm at the latest follow-up (P < 0.004). The sinus also increased significantly in diameter from 29 ± 4 at surgery to 34 ± 2 mm at the last follow-up (P < 0.001). This increase in autograft size, both for the annulus and the sinus, paralleled the increase in body surface area with no evidence for unproportional dilatation. Hemodynamic measurements demonstrated physiological peak gradients of 6.8 ± 2.9 mmHg and no or trivial aortic insufficiency in 95% of this rapidly growing patient population. Conclusion: These data demonstrate growth of the pulmonary autograft parallel to somatic growth without undue dilatation in the systemic circulation. The hemodynamics are excellent with regard to physiological gradients and no increase in aortic insufficiency.

Keywords: Aortic valve replacement; Pulmonary autograft; Autograft growth; Congenital

1. Introduction

The pulmonary autograft is now preferred by many surgeons as an aortic valve substitute in the pediatric age group and young adults. Short- and mid-term results have been excellent with low rates of valve related deaths and complications [1–4]. The hemodynamic performance at rest and during exercise [5] shows physiological flow velocities and gradients across the autograft in the majority of patients. This leads to rapid remodeling of the left ventricle with normalization of left ventricular dimensions and mass after surgery [6,7]. These observations are seen in both the adult, and importantly, in the pediatric age group as well.

We hypothesized that stable graft function in the adult would indicate that the autograft is simply able to withstand systemic arterial pressures without accelerated degeneration or dilatation. In the pediatric age group, however, somatic growth must be accompanied by growth of the pulmonary autograft to maintain physiological gradients and valve competence over time.

The purpose of this study was to review our results with the Ross operation in the pediatric age group with particular emphasis on growth. We assessed the dimensional changes of the pulmonary autograft, valve gradients and competence in relation to somatic growth in our pediatric Ross population.

2. Methods

Thirty patients younger than 18 years of age were identified in the entire population of 139 patients who received a Ross operation since 1991, and are included in this study. The patient age was 11.3 ± 3.1 years (range, 6–17 years). Nine children were younger than 10 years at the time of operation. There were 21 males and nine females. Twenty
patients had undergone previous aortic valve interventions, 19 surgical valvotomies and six balloon dilatations. Table 1 shows the patient valve pathology and dominant hemodynamic lesions. None of the children had associated congenital cardiac defects at the time of operation.

Aortic valve replacement was performed using standard cardio-pulmonary bypass techniques, with moderate hypothermia and combined antegrade and retrograde blood cardioplegia. The pulmonary autograft was used in all patients as full root with coronary artery reimplantation. Particular attention was given to mismatch of the autograft with both the aortic annulus and the sino-tubular junction, which were surgically addressed if needed. Since our experience did not encompass neonates and small infants until recently, none of the patients presented with a hypoplastic annulus. Annular plication had to be performed in three adolescents with a significantly dilated annulus which was more than 5 mm larger than the autograft. In all other patients, there were no, or only minor, discrepancies between the annulus and the autograft which could be addressed during suturing of the autograft and by using a wider muscle cuff on the pulmonary root. A continuous running suture technique was used in all patients. The autograft was always implanted in an intra-annular position. The dilatation of the sino-tubular junction had to be adjusted through wedge resection of the ascending aorta and appropriate tailoring in two cases. A cryopreserved pulmonary homograft was used for reconstruction of the right ventricular outflow tract and was oversized whenever possible.

All patients were followed on a yearly basis with clinical and echocardiographic follow-up. The echocardiographic assessment included color flow Doppler to assess the severity of autograft and homograft insufficiency, which was graded semiquantitatively according to established criteria and graded as 0–4 +. The mean and peak transvalvular flow velocities were measured with continuous wave Doppler, and mean and peak gradients were calculated. The autograft dimensions were measured as the cross-sectional diameter at the level of the aortic annulus, the maximal diameter of the sinus and at the sino-tubular junction. We also compared the time related longitudinal performance of the autograft with that of the pulmonary homograft used to reconstruct the right ventricular outflow tract.

All data are expressed as means ± SD. The paired Student’s t-test was used for analysis of the differences between measurements taken after surgery and those at the latest follow-up. Statistical significance is assumed with a P value of less than 0.05. Cross-sectional diameters of the annulus and autograft root were plotted against normal values for body surface area [8].

3. Results

There were no perioperative deaths.

A follow-up of at least 1 year was available in 20 children, with a mean of 4.3 ± 2.6 years. These children are the subject of this report. One child died 1 year after surgery in a car accident. All other patients are well and in NYHA class I. There was one early reoperation due to perforation of an autograft leaflet which was reconstructed. No valve related events or autograft endocarditis were observed.

Echocardiography demonstrated excellent autograft function. Ninety-five percent of the patients showed no or trivial aortic insufficiency on color flow Doppler, and 5% have mild (grade I) aortic regurgitation. The peak aortic gradient was 6.7 ± 3.7 mmHg at the last follow-up (Table 2).

Patients showed significant somatic growth. During follow-up, the body weight increased from 42 to 58 kg, and height increased from 147 to 165 cm, with an increase in body surface area from 1.3 ± 0.4 at surgery to 1.6 ± 0.3 m² (P < 0.002) at the latest follow-up. However, valve performance remained stable over time, as was reflected by the echocardiographic data showing no increase in either the number of patients developing new or increasing severity of aortic insufficiency. Similarly, valve gradients showed no increase (Table 2). The annulus diameter increased significantly from 18 ± 2 at surgery to 20 ± 3.5 mm at the latest follow-up (P < 0.004). The sinus also increased significantly in diameter from 29 ± 4 at surgery to 34 ± 2 mm at the last follow-up (P < 0.001). This increase in autograft size, both for the annulus and the sinus, paralleled the increase in body surface area (Figs. 1 and 2). While the annulus diameter was within normal limits for body surface area after surgery and at the latest follow-up, the sinus diameter was significantly larger than normal after surgery when plotted against normal values for aortic sinus of

<table>
<thead>
<tr>
<th>Valve pathology and dominant hemodynamic lesion</th>
<th>n</th>
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<tbody>
<tr>
<td>Bicuspid aortic valve</td>
<td>18</td>
</tr>
<tr>
<td>Monocuspid aortic valve</td>
<td>1</td>
</tr>
<tr>
<td>Post endocarditis</td>
<td>1</td>
</tr>
<tr>
<td>Aortic insufficiency</td>
<td>19</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Combined lesion</td>
<td>8</td>
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</tbody>
</table>

Table 1

Valve pathology and dominant hemodynamic lesion

<table>
<thead>
<tr>
<th>Post surgery</th>
<th>Follow-up</th>
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</thead>
<tbody>
<tr>
<td>BSA (m²)</td>
<td>1.3 ± 0.42</td>
</tr>
<tr>
<td>Peak gradient (mmHg)</td>
<td>7.2 ± 3.3</td>
</tr>
<tr>
<td>Mean gradient (mmHg)</td>
<td>3.7 ± 1.2</td>
</tr>
<tr>
<td>AI</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>5</td>
</tr>
<tr>
<td>Trivial (0–1)</td>
<td>14</td>
</tr>
<tr>
<td>Mild (1–2)</td>
<td>1</td>
</tr>
</tbody>
</table>

a BSA, body surface area; AI, aortic insufficiency.
b N = 20.
Valsalvae for body surface area. However, the increase in size thereafter was again parallel to somatic growth.

In contrast to the autograft, the pulmonary homograft did not show any increase in size. This was reflected by a significant increase in the peak gradient from $9.6 \pm 3.5$ mmHg after surgery to $17.0 \pm 10.8$ mmHg (range, $4-46$ mmHg; $P < 0.01$). Eighty-five percent of patients had no or trivial pulmonary insufficiency and 15% had mild regurgitation.

4. Discussion

Our study confirmed that the Ross operation can be performed with low operative risk and excellent clinical and hemodynamic outcome in the pediatric age group. Most importantly, we were able to document that the autograft increased significantly in size by 17% during follow-up. This increase paralleled the significant somatic growth of the children with a 23% increase in body surface area from surgery to the latest follow-up. With our surgical technique [9] of root replacement, careful adjustment of size discrepancies of the annulus and sino-tubular junction and the pulmonary autograft and strict intra-annular implantation, the annulus and sinus showed a distinctly different behavior. The annulus was within normal limits for body surface area after surgery and showed growth along the normal growth curve when plotted against normal values of annular diameter for body surface area. In contrast, the sinus was significantly larger than normal after surgery. This would indicate early postsurgical dilatation of the pulmonary sinus in the systemic high pressure circulation. However, size increase was then again parallel to normal growth curves without indication for further undue aneurysmal dilatation, suggesting normal growth of the autograft. The early increase in pulmonary autograft sinus size with stabilization during follow-up is also corroborated by the findings of Solymar et al. [10]. Since we had no reoperations during our follow-up period in this series, we were unable to obtain tissue samples as direct evidence of growth. However Schoof et al. clearly demonstrated, in their experimental study in growing pigs, active growth of the pulmonary valve cusps and autograft wall. In fact, pulmonary cusp weight, height and width, as well as wall thickness, increased significantly more than in control pulmonary roots, suggesting normal growth of the autograft. The early increase in pulmonary autograft sinus size with stabilization during follow-up is also corroborated by the findings of Solymar et al. [10]. Since we had no reoperations during our follow-up period in this series, we were unable to obtain tissue samples as direct evidence of growth. However Schoof et al. clearly demonstrated, in their experimental study in growing pigs, active growth of the pulmonary valve cusps and autograft wall. In fact, pulmonary cusp weight, height and width, as well as wall thickness, increased significantly more than in control pulmonary roots, suggesting normal growth of the autograft.

The pulmonary homograft showed a very different behavior to the autograft in our series. During follow-up, there was no increase in the size of these grafts. In contrast to the autograft, transvalvular gradients increased significantly, and 15% of the patients showed a mild to moderate (grade 1.5–2+) degree of pulmonary insufficiency, even though the homograft valve leaflets appeared normal on echocardiography. This indicates a lack of adaptation to the increase in heart size and stroke volume, and possibly, early degeneration.

The excellent durability of the pulmonary autograft is also in sharp contrast with the poor performance of pulmonary homografts in the aortic position with rapid graft failure [12]. While cryopreservation maintains the vitality of the tissue to some extent, which is indicated by the immune response seen after implantation [13], the ability for active adaptation and growth is lost.

Our study may be limited by the fact that there are no neonates and small infants in this series, who, in general, present with a hypoplastic annulus. The finding of seemingly appropriate growth in our patients may not be simply transferred to the infant population. None of the patients in this series were suitable for valve repair, which is preferred over replacement whenever possible. Excellent mid- to long-term results can be obtained with mechanical valve replacement as well [14]. However, this is at the cost of lifetime anticoagulation. No patient younger than 18 years has received a mechanical valve since the introduction of the Ross operation in our institution, and we feel, at this point, supported by the excellent results up to 9 years.
We concluded from our data that there was strong evidence that the vital autograft tissue is able to rapidly adapt to the systemic pressure environment with early dilatation of the sinus. Normal growth occurs thereafter along established growth curves. This facilitates the maintenance of the excellent hemodynamic properties and durability, without signs of degeneration in the rapidly growing pediatric patient. This, in addition to minimal late valve related complications, makes the pulmonary autograft uniquely suited for aortic valve replacement in children.

References


Appendix A. Conference discussion

Dr K. Samir (Marseille, France): What is the difference between growth and dilatation of the pulmonary autograft in the aortic position?

Dr Simon: Well, we obviously don’t have direct evidence by getting tissue. We didn’t have any reoperations in this series where we could have harvested tissue to actually prove that there was true growth. So, we have only indirect evidence. We feel that the fact that the valves remain competent over a very long period does actually reflect growth and not dilatation, because if it was just passive dilatation, at one point, coaptation should decrease. There is actually experimental evidence in, I believe, a pig model which was done some years ago where the authors were actually able to demonstrate that, after the Ross operation, the pulmonary valve exhibited very accelerated increase in tissue mass, both the leaflets and the root.

Dr M. Muxra (Barcelona, Spain): I have noticed that most of the patients that had been operated in this series had a bicuspid aortic valve. You didn’t question that probably the best indication is not just with the bicuspid aortic valve. It’s because of the morphological malformation of the pulmonary artery and even the valve. What is your opinion about that?

Dr Simon: I share your concerns about dilatation in patients with bicuspid valves. Unfortunately, the majority of patients we see who undergo the Ross operation do have bicuspid valves, and at this point, at least, our data indicates that we are on the safe side. We are watching this very carefully, and we are right now analyzing our adult population with regard to late root dilatation, especially in the bicusps.