Aortic valve replacement in children: are we on the right track? 1

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Abstract

Objective: The choice of the ideal prosthesis for aortic valve replacement (AVR) in children is still controversial. Early degeneration of bioprostheses and the potential risks related to anticoagulation in the child have renewed the interest of many surgeons towards the Ross operation. This study concerns our 22-year experience with AVR in children.

Methods: Forty-six children, aged 4 months to 16 years (mean 11.6 years), had AVR between April 1974 and December 1996. Preoperative diagnosis revealed aortic regurgitation (AR) in 25 cases, aortic stenosis (AS) in ten, combined AS and AR in nine and LVOTO in two patients. Of the 46 patients, 26 had 37 previous procedures. Isolated AVR was performed in 19 cases, 27 children underwent 36 concomitant intracardiac procedures. Mechanical prostheses were implanted in 30 children, bioprostheses in eight, aortic homografts in three. Five patients underwent a Ross procedure.

Results: There was one hospital death in the latter group (2.1%). Six of seven late deaths occurred in patients who underwent complex intracardiac procedures (15.2%). Reoperation rate was 19.5% (n = 10), differentiating 16.6% for mechanical (5/30 patients) and 50% (4/8 patients) for bioprostheses. The mean follow-up period was 8.01 years, ranging from 0.45 to 21.66 years (304.04 patient-years). There was one hemorrhagic event (2.1%) due to anticoagulation, thrombosis of the mechanical valve occurred in two patients (4.2%).

Conclusions: AVR can be performed with acceptable mortality rate and good long-term results in children. We perform the Ross operation only in selected cases. According to our experience, mechanical prostheses show excellent performances in children with a low incidence of complications related to anticoagulation. © 1998 Elsevier Science B.V. All rights reserved

Keywords: Valve replacement; Children; Ross operation

1. Introduction

The choice of the ideal prosthesis for aortic valve replacement (AVR) in children is controversial. The implantation of bioprostheses in young patients has been abandoned because of early degeneration. Cryopreserved homografts also do not offer a definitive solution, due to limited durability. Although the performance of mechanical prostheses is excellent, the potential risks related to anticoagulation have renewed the interest towards the AVR with a pulmonary autograft (Ross operation). We therefore retrospectively reviewed our 22-year experience with AVR in children.

2. Materials and methods

Between April 1974 and December 1996, 46 children, aged 4 months to 16.1 years (mean age 11.6 years), underwent AVR at our institution. There were 33 boys and 13 girls. Body surface area (BSA) ranged between 0.30 and 2.16 m² (mean 1.29 m²). Indications for AVR were: aortic regurgitation (AR) in 25 (54.3%) patients, aortic stenosis (AS) in 10 (21.7%), combined AR and AS in 9 (19.5%) and LVOTO in 2 (4.3%) patients. Of the 46 patients, 27 (58.6%) had a total of 37 previous cardiac procedures.
Reconstructive surgery of the aortic valve had been attempted in 17 of the 46 patients (36.9%). Five additional children had aortic balloon valvuloplasty. The previous operations are summarized in Table 1.

### 3. Operative data

All patients were operated on with standard cardiopulmonary bypass, single or bicaval cannulation and moderate hypothermia. Cold (2°C) crystalloid Bretschneider’s cardioplegic solution (Custodiol®, 30 ml/kg body weight) was infused in the aortic root or, in presence of significant AR, directly into the coronary ostia. Mechanical valves and bioprostheses were implanted in subcoronary position using mattress, interrupted 2/0 or 3/0 Ethibond® sutures, in part reinforced with Teflon pledgets. Of the three implanted aortic homografts, one was implanted as a free graft in subcoronary position, the remaining two as aortic root replacement with implantation of the coronary ostia. The latter technique was performed in all the five children who underwent the Ross operation. Cardiopulmonary bypass and cross-clamping time were significantly longer in the patients undergoing the Ross procedure (mean 146 and 115 min. respectively), not only compared with those receiving AVR alone (mean 75 and 51 min.), but also with patients with AVR plus concomitant intracardiac procedures (mean 120 and 88 min). Mechanical prostheses were implanted in 30 children (65.2%), bioprostheses in 8 (17.3%) and cryopreserved aortic homografts in three patients (6.5%). Since 1994, five patients (10.8%) underwent AVR with the pulmonary autograft (Ross operation). Types and diameters of the prostheses are listed in Table 2.

Isolated AVR was performed in 20 cases (43.4%), 26 children (56.5%) had additional 37 intracardiac procedures. In five patients double aortic and mitral valve replacement was performed, three patients had mitral valve repair and seven additional children underwent concomitant replacement of the ascending aorta.

### 4. Results

#### 4.1. Early mortality

There was one hospital death (2.1%) in the Ross procedure group. He was a 4-month-old infant with critical AS and parachute mitral valve, who had been admitted with signs of severe heart failure. By admission, high CRP value (90 mg/dl) and leukocytosis (16 000/l) were evident but, because of progressive deterioration of his general conditions, aortic balloon valvuloplasty was performed. Unfortunately avulsion of the aortic valve occurred and massive

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**Table 1**

<table>
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<tr>
<th>Type</th>
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<th>No. of patients</th>
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<td>27</td>
</tr>
<tr>
<td>Aortic valvotomy + others</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>Aortic VVPL + others</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>LVOTO resection + others</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Truncus arteriosus repair</td>
<td>2</td>
<td></td>
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<tr>
<td>Tetralogy repair</td>
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<td></td>
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<tr>
<td>MVP</td>
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<td></td>
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<tr>
<td>BWG repair, mitral VVPL</td>
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<td>Arterial switch, VSD</td>
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<td></td>
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<tr>
<td>Others</td>
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<td></td>
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<tr>
<td>Extracardiac</td>
<td>7</td>
<td></td>
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<tr>
<td>Balloon aortic valvotomy</td>
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<tr>
<td>Others</td>
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**Table 2**

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<tr>
<td>CM</td>
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</tr>
<tr>
<td>SJ</td>
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<td>1</td>
</tr>
<tr>
<td>Starr A 6</td>
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<td>ATS</td>
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<tr>
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<tr>
<td>Hancock</td>
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<tr>
<td>CE</td>
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<tr>
<td>Bicor</td>
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</tr>
<tr>
<td>Homografts</td>
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<td>1</td>
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<tr>
<td>Autografts</td>
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<tr>
<td>Total</td>
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</tr>
</tbody>
</table>

VVPL, valvuloplasty; BWG, Bland–White–Garland syndrome; LVOTO, left ventricular outflow tract obstruction.

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SJ, St. Jude Medical; BS, Björk–Shiley; CM, CarboMedics; CE, Carpentier–Edwards.
AR developed, requiring urgent surgical intervention. A Ross operation with concomitant mitral annuloplasty was successfully performed, but the patient died on the fifth postoperative day because of Klebsiellae and Candida sepsis. He was the youngest of the patients included in this study.

4.2. Late mortality

There were seven late deaths (15.2%), with an overall mortality rate of 17.1%. Six of these seven deaths occurred in patients who had undergone complex intracardiac procedures. The causes of late mortality are reported in Table 3. The cumulative survival curve is shown in Fig. 1.

5. Follow-up

The 38 survivors were regularly followed-up at 3-, 6- or 12-month intervals at the outpatient department of the Clinic of Pediatric Cardiology at our hospital (Director: Prof. Dr. J. Hess), yielding a 100% complete follow-up. Follow-up time ranged between 0.67 and 21.66 years (mean follow-up time 8.01 years). Cumulative follow-up was 304.04 patient-years. Kaplan–Meier’s and linearized statistical techniques were used to analyze patient survival and incidence of valve related complications. Valve-related complications were evaluated according to the guidelines reported by Edmunds [1].

6. Anticoagulation

Of the 30 patients who had received mechanical prostheses, 29 were anticoagulated with warfarin sodium, keeping the prothrombin time between 15 and 25% (INR = 2.5–3.5, Innovin®, Baxter). Anticoagulation was started on the first or second postoperative day. Out of this group, 16 patients (55%), or their parents, were trained to perform the test by themselves with a self-testing device (Biotrack 512®, CoaguChek®, Boehringer Mannheim). One patient, who underwent AVR with a 21 mm Björk–Shiley valve at the age of 4.2 years, refused to take anticoagulants. She is now living for more than 21 years with the same valve and has never shown any problem.

7. Valve-related complications

7.1. Haemorrhagic events

There was one anticoagulation-related hemorrhagic event (3.3%) in the group of 30 patients undergoing AVR with a mechanical valve.

This patient had double mitral and aortic valve replacement with mechanical valves at the age of 13 years. Because

![Fig. 1. Cumulative survival after AVR in children. Kaplan–Meier’s analysis.](https://academic.oup.com/ejcts/article-abstract/13/5/565/418144)}
later on a Willebrand–Jürgens syndrome was diagnosed, anticoagulation was changed to acetylsalicylic acid only (Asasantin®, 300 mg daily). It was well tolerated for more than 12 years, then the patient developed acute gastric hemorrhage. After successful endoscopic treatment, anticoagulation was turned to warfarin again without further complications.

7.2. Thromboembolic events

Thrombosis of the mechanical valve occurred in two out of 30 patients (6.6%) after AVR with mechanical prostheses, after 0.6 and 8.7 years, respectively.

The first patient had undergone repair of tetralogy at 5 years of age. Two months later, reoperation because of a residual VSD and AR was necessary. AVR with a 19 mm Björk–Shiley prosthesis was performed. She was admitted 8.7 years later with dyspnea at rest and echocardiography showed thrombosis of the valve. Immediate operation was successful and the valve was replaced with a 21 mm St. Jude prosthesis after patch enlargement of the aortic ring. The postoperative course was uneventful, the patient is still doing well 10 years later.

The second patient had AVR with a 27 mm Björk–Shiley prosthesis at the age of 14 years. Seven months later, he was readmitted with acute abdominal pain and clinical signs of left renal artery embolism. Echocardiography confirmed the suspected valve thrombosis. At reoperation, a wide, plane thrombus, occluding the upper surface of the valve was found. It was removed, the valve reoriented and the operation was terminated without further problems.

The actuarial curve for freedom from thromboembolism is illustrated in Fig. 2. These results compare well with other published data [2,3].

8. Reoperations

The reoperation rate 0.5–14 years after AVR (mean 6.6 years) was 19.5% (9 patients). It accounted for 16.6% (5/30) mechanical and 50% (4/8) bioprostheses. Furthermore, the mean time interval between AVR and replacement of a bioprosthesis was 4.2 years, whereas replacement of a mechanical valve occurred after a mean time interval of 8.6 years. Since 1983, no patient received a bioprosthesis. Causes of reoperation were thrombosis (two patients) and significant pressure gradient (three patients) in the mechanical prostheses group, tissue failure (three patients) and pressure gradient (one patient) in the bioprostheses group.

The actuarial curve for freedom from reoperation after AVR is summarized in Fig. 3.

9. Discussion

The ideal prosthesis for AVR in children has not been found yet and some authors even regard AVR in pediatric patients as a palliative procedure [4]. Actually, this statement can be considered true, as all available prostheses have significant drawbacks.

In those cases where reconstructive surgery of the aortic valve fails, or is impossible, there are basically four types of prostheses that can be chosen for AVR: bioprostheses, mechanical prostheses, homografts and the pulmonary autograft.

9.1. Bioprostheses

The initial enthusiasm for bioprostheses extinguished, as the first mid- and long-term follow-up reports showed a very
high incidence of early degeneration, calcification and structural failure [5,6]. This led to a high incidence of reoperations, which became even more evident in the younger patients. Our results with AVR with bioprostheses in children confirm these findings: between 1974 and 1982, we implanted eight biological prostheses in the aortic position. After a mean time of 4.2 years, 50% of them had to be replaced, mainly because of structural failure of the valve. Since 1983, no more bioprostheses were implanted in children. Although the new types of bioprostheses (e.g. porcine stentless valves) seem to offer better hemodynamic performances and longer durability, long-term results are needed to confirm these findings. We believe that today implantation of bioprostheses in children and young adults should be abandoned, or strictly limited to particular cases.

9.2. Homografts

The first report of Mr Donald Ross about AVR using an aortic homograft 35 years ago [7] awoke huge interest but it still does not seem to offer a definitive solution to the problem of AVR in children. This kind of operation is more difficult and the operative mortality is higher than after AVR with mechanical or bioprostheses [8]. The reoperation rate is lower than for bioprostheses, but may be higher than after AVR with mechanical valves. Furthermore, the availability of such homografts, especially the small sized ones, is limited. Our experience with AVR with homografts in this group is limited to three patients only.

Two of them had Marfan syndrome with AR and aneurysm of the ascending aorta: aortic root replacement was performed. In one of these cases re-AVR with a mechanical valve was performed 5 years later because of significant regurgitation of the homograft valve. The third case was a 3.8-year-old girl with diagnosis of Bland–White–Garland syndrome and significant mitral regurgitation, who had undergone direct reimplantation of the left coronary artery in the aorta and a mitral valve annuloplasty. On the 13th postoperative day, echocardiography showed significant mitral and aortic regurgitation; at reoperation, MVR with a bileaflet mechanical prosthesis and AVR with a 10 mm aortic homograft was performed. This child is still doing well, more than 4 years later. Today we would prefer a Ross operation especially in such a young patient.

9.3. Mechanical prostheses

Mechanical prostheses represent for most surgeons the safest alternative for AVR in children, very good results have been reported with their use [9,10]. On the other hand, the potential risk of thromboembolic complications, the fixed diameter of the prosthesis in the growing organism, and the need for long-life anticoagulation in the very active child, with the risk of hemorrhage, represent important drawbacks of AVR with mechanical valves.

Out of the 46 patients included in this study, 30 (65.2%) had AVR with mechanical prostheses with no hospital mortality. In the long-term follow-up (0.4–21.6 years, mean 6.6 years), late mortality in this group was 10% (3/30). Reoperation rate was 16.6% (5/30): one of the late deaths was related to the reoperation and occurred in a 13-year-old girl who underwent AVR with a Starr A16 prosthesis at age 5.8 years. Re-AVR was performed 7.3 years later because of a significant transvalvular pressure gradient. At reoperation, aortoventriculoplasty according to Konno [11] was performed and a 23 mm Björk–Shiley prosthesis was...
implanted. The postoperative course was uneventful; on the 15th postoperative day, the patient showed signs of acute right ventricular failure, followed by bradycardia and hypotension. Cardiopulmonary resuscitation was necessary, while echocardiography showed an important LV-RV shunt due to dehiscence of the ventricular patch. The patient died before emergency surgery was possible.

The incidence of thromboembolism related to anticoagulation was 6.6% (2 patients). One hemorrhagic event (3.3%) occurred in a patient after double valve replacement.

9.4. Ross operation

In the last years there has been an increasing interest towards AVR with the pulmonary autograft, according to the operative technique created by Donald Ross 30 years ago [12]. Theoretically, the pulmonary autograft is the ideal prosthesis, as no anticoagulation is required, the autograft does not degenerate, it shows excellent hemodynamic performance and seems to grow with the child [13,14]. We begun to perform the Ross procedure in June 1994, and since that time five children underwent AVR with the pulmonary autograft (10.8%). In the same period a total of ten patients under 16 years of age underwent isolated AVR. That means, in the last 3 years 50% of the cases needing isolated AVR have received a pulmonary autograft. Although hospital mortality in this small group of patients is as high as 20% (1/5 patient), this is misleading since, as reported above, the cause of death was neither cardiac nor valve related.

All the survivors show in the short follow-up period an excellent function of the pulmonary autograft and of the right placed homograft as well.

Since our first results with the Ross procedure are very encouraging, we will continue performing this operation. The Ross procedure is technically a very skill demanding operation, and since the first report in 1967, until now only 127 surgeons worldwide have performed a total of 1976 operations (Ross Procedure International Registry Home Page). As for other technical difficult operations, the learning curve has a significant impact on the results in term of morbidity and mortality.

It is a time consuming operation: in our experience the cross clamping time was more then twice as long for a Ross procedure (mean 115 min), compared with simple AVR with mechanical or bioprostheses (mean 51 min.) and it was also significantly longer compared to AVR plus concomitant procedures (mean 88 min). Since 26 of the 46 patients included in this study (56.5%) needed some other complex intracardiac procedures it is clear that, despite optimal myocardial protection, this procedure cannot be indiscriminately performed on every patient requiring AVR.

There have been reports about failure of the pulmonary autograft in patients with endocarditis [14] or juvenile rheumatism [15].

Another point that deserves attention, is the destiny of the conduit implanted for the replacement of the pulmonary valve. Most of the authors prefer a cryopreserved pulmonary homograft for the reconstruction of the RVOT, which seems to have a very long life with low incidence of structural degeneration. Reports with the use of porcine stentless valves for RVOT reconstruction have been published too. Our 3 years follow-up time in this group is too short to come to definitive conclusions, but our 20 years experience with 344 aortic and pulmonary homografts for reconstruction of the right ventricular outflow tract (Homann et al., unpublished data) shows that the mean time interval between the first implantation and replacement of the conduit was 12 years for conduits with a diameter less than 20 mm, and 14 years for homografts with a diameter more than 20 mm. This means, that these patients, whose mean age at the first Ross operation was 11.8 years, facing a life expectancy of around 50 years, will have at least two or three reoperations.

Still, the Ross procedure represents the only and maybe best solution for infants needing AVR and for those cases with isolated aortic valve lesions with or without LVOTO. For other young patients, especially those presenting concomitant heart diseases, our first choice remains the bileaflet mechanical prostheses, since in the long-term follow-up the rate of anticoagulation-related complications has been low. While AVR with aortic homografts is performed in those patients with aortic root aneurysm or with florid endocarditis, AVR with bioprostheses should be strictly limited to very few cases or abandoned.

References


