Aortic root replacement in children: a word of caution about valve-sparing procedures

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Abstract
Objective: Evaluate the results of various surgical procedures used for aortic root replacement in children with aortic root aneurysm. Methods: Between 1986 and 2007, 23 children (less than 16 years of age, mean age: 8.1 ± 5.1 years) underwent elective aortic root replacement for aortic root aneurysm (with associated aortic insufficiency in 10 patients). All had connective tissue defect syndromes. Nine patients underwent composite valve graft repair using a mechanical valve. Fourteen children underwent valve-sparing aortic root replacement (remodeling procedure in 11, reimplantation procedure in 3). Mean follow-up (100% complete) was 7.3 ± 5.5 years (range 6 months—21 years). Results: There was one early death (4.3%)(after valve-sparing remodeling) and no late mortality. Following valve-sparing remodeling operation (10 patients, mean follow-up: 7.8 ± 3.0 years), there were 9 reoperations in 6 patients and only 5 patients retained their native aortic valve. In patients who underwent valve-sparing reimplantation operation (three patients, mean follow-up: 2.1 ± 0.3 years), one underwent reoperation for endocarditis. Fifteen patients had composite valve graft replacement either as a primary operation (nine cases) or at reoperation for valve-sparing failure (six cases); mean follow-up was 8.2 ± 6.2 years; there was no thrombo-embolic or hemorrhagic event and one reoperation for patient-prosthesis mismatch. Conclusions: (1) composite valve graft aortic root replacement provides excellent long-term results in children with aortic root aneurysm due to connective tissue disorder. This remains the first choice procedure in patients with more than minimal aortic insufficiency, with distorted aortic leaflets or needing concomitant mitral valve replacement. (2) Valve-sparing remodeling surgery yields disappointing results and should probably be abandoned in the pediatric population. (3) Valve-sparing reimplantation repair may achieve superior outcome but needs further evaluation.

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1. Introduction

Aortic root aneurysms are rare in children and are typically associated with connective tissue defect syndromes. These aneurysms may lead to rupture, dissection or severe aortic regurgitation. To avoid complications and risky life-saving operations, prophylactic aortic root replacement is indicated. Guidelines for management and timing of surgery have been advocated, according to the size of the aortic root and the severity of the underlying connective tissue disorder [1]. There remains, however, controversy regarding the optimal surgical management.

Composite mechanical valve graft repair (Bentall operation) has long been the standard procedure for complete replacement of the aortic root in patients with connective tissue syndromes. In the pediatric population, the Bentall procedure carries two potential drawbacks: need for long-term anticoagulation and difficulty in implanting an adult-size composite conduit in small children. Several valve-sparing operations have been designed and used extensively in adults. These procedures have also been applied to children but long-term results are lacking, particularly regarding the development of aortic insufficiency and the need for reoperation.

The present study was undertaken to evaluate our experience in children who have undergone aortic root replacement using various surgical procedures.

2. Materials and methods

2.1. Patient population

Twenty-three children (age 10 months to 16 years. mean: 8.1 ± 5.1 years) underwent aortic root replacement between
1986 and 2007. Twelve patients (52%) were less than 5 years of age. There were 18 boys (78%) and 5 girls. All patients had connective tissue disorder: Marfan syndrome in 14 (61%), Loeys–Dietz syndrome in 1, Turner syndrome in 1 and bicuspid aortic valve syndrome in 7 patients. Patients who underwent aortic root replacement late after neonatal repair of heart malformation (transposition of the great arteries, common arterial trunk or pulmonary atresia/VSD) were excluded.

Five patients (22%) had previous cardiovascular operations (coarctation repair, patent arterial duct closure, Ross operation, aortic commissurotomy and pulmonary artery aneurysm repair in one patient each).

All patients underwent elective surgery. The primary indication was either major dilatation of the aortic root or rapid progression of the aortic diameter. At the time of surgery, the mean ascending aorta diameter was 47 mm (range 27–63 mm) and the mean progression of the aortic root diameter during the year preceding surgery was 7.5 mm (range 4–26 mm). Thirteen patients (56%) had no or minimal aortic regurgitation while the other 10 patients had moderate (4 patients) or severe (6 patients) insufficiency.

2.2. Operative techniques

Nine patients (39%) underwent composite graft replacement by a modified Bentall procedure. The standard technique included complete excision of the aneurysm, direct Anastomosis of the mobilized coronary arteries to the Dacron graft and full-thickness anastomosis of the distal graft to the transected distal ascending aorta.

All patients, except one, had moderate to severe aortic insufficiency. The aortic valve was bicuspid in five patients. Three children had Marfan syndrome. In two patients, an enlargement of the aortic annulus by incision and patching of the conal septum (Konno principle) was necessary to accommodate an adult-size composite graft. Associated procedures included replacement of the aortic arch (one patient) and closure of a patent arterial duct (one patient).

Composite graft replacement was the procedure of choice in children with moderate to severe aortic regurgitation. Mean age at operation was 10.2 ± 5.3 years; mean weight was 42 ± 24 kg.

Eleven patients (48%) underwent a valve-sparing remodeling procedure, as first described by Sarsam and Yacoub [2]. The entire aortic root was resected and replaced with three separate tongue extensions of a tubular Dacron graft into which the coronary arteries were reimplanted. Only two patients had more than minimal aortic regurgitation. Eight patients had Marfan syndrome. The aortic valve was bicuspid in three cases. Mitral valve repair was performed concomitantly in two patients (both with Marfan syndrome).

The last three patients (13%) underwent a valve-sparing reimplantation procedure (David I operation) in which the native aortic valve was resuspended in a straight Dacron tube implanted onto the ventriculo-aortic junction [3]. All patients had Marfan syndrome and minimal aortic regurgitation. The aortic valve was bicuspid in one.

For all valve-sparing procedures (remodeling or reimplantation), an external ring was never used to support the ventriculo-aortic junction.

The size distribution of aortic root prostheses is illustrated in Fig. 1.

A valve-sparing procedure was, in most cases, chosen in children with no or minimal aortic regurgitation. Surgery was undertaken at a younger age (in comparison with the composite graft group); mean age was 6.9 ± 5.1 years and mean weight was 29 ± 16 kg. However, the differences in age and weight did not reach statistical significance (p = 0.173 and p = 0.102, respectively).

2.3. Follow-up and data analysis

This study was approved by the Paris V University ethics committee. The need for individual consent was waived.

Data were obtained from clinical records, patient interviews and contacts with primary physician and cardiologist. Transthoracic echocardiographic data were obtained in all survivors within 6 months of study closure. Composite graft replacement patients were managed with sodium warfarin to maintain an international normalized ratio (INR) of 2–3. Valve-sparing procedure patients received long-term aspirin therapy. Beta-blockers were routinely used to minimize hemodynamic stress on the distal aorta. Mean follow-up was 7.3 ± 5.5 years (range 6 months to 21 years). Follow-up data were obtained for all patients.

All data were presented as mean ± SD unless otherwise specified. Crude ratios were given with 70% confidence limits. Actuarial freedom from late events was calculated using the Kaplan–Meier method. The probability of freedom from events was compared using the log-rank test.

3. Results (Table 1)

3.1. Early results

There was one early death (4.3%, 70% confidence limits: 0.6–13.7%). A 1.5-year-old Marfan patient underwent valve-sparing remodeling procedure for aortic root aneurysm (aortic diameter 50 mm) with minimal aortic regurgitation. Because of moderate aortic insufficiency at routine intraoperative transesophageal echocardiography, a Bentall procedure was immediately performed. The following day, severe mitral regurgitation was demonstrated and the child underwent mitral valve replacement. Because of low cardiac output syndrome, he was put on mechanical assist device and died 3 days later of multi-organ failure.
Another patient (12-year-old Marfan patient with moderate aortic insufficiency) underwent valve-sparing remodeling procedure and concomitant mitral valve repair; conversion to composite graft replacement was necessary on the first postoperative day because of severe aortic regurgitation.

Two patients following Bentall procedure with conal septum enlargement (Bentall–Konno procedure) developed complete heart block and underwent pacemaker implantation.

3.2. Late results

3.2.1. Composite graft replacement patients (nine patients)

All patients were alive after a mean follow-up of 8.6 ± 7.7 years (range 6 months to 21 years). No episode of clinically evident thrombo-embolic event, hemorrhagic complication or endocarditis was noted. No reoperation was necessary.

3.2.2. Valve-sparing remodeling procedure patients (10 patients)

The results of this sub-group are summarized in Table 2. All patients were alive after a mean follow-up of 7.8 ± 3.0 years (range 4—12 years).

Only four patients did not undergo reoperation. Six patients underwent reoperation after a mean interval of 26 months (range 1 day—60 months). Two patients underwent a second reoperation 42 and 51 months after the first one. One patient had a third reoperation 2 years after the second one. At last follow-up, only five patients had retained their native aortic valve (no or minimal insufficiency in four, moderate regurgitation in one).

3.2.3. Valve-sparing reimplantation patients (three patients)

One patient developed severe post-endocarditis aortic regurgitation and needed a Bentall procedure 26 months postoperatively. All three patients were alive after a mean follow-up of 2.1 ± 0.3 years (range 22—29 months). One had a composite graft; one had minimal aortic regurgitation and the last one had moderate regurgitation (with associated moderate mitral insufficiency).

3.2.4. Late results in all patients (22 patients)

There was no late mortality. At last follow-up, all patients were in NYHA functional class I, leading a fully active normal life.

In 15 patients (68%), a prosthetic aortic valve had been implanted, either primarily (9 cases) or at reoperation (6 cases). In this sub-group (mean follow-up: 8.2 ± 6.2 years), there was no thrombo-embolic or hemorrhagic event. Seven patients (32%) had retained their native aortic valve which exhibited no or minimal regurgitation in five patients and moderate insufficiency in two cases. Among the four patients with a bicuspid aortic valve who underwent a valve-sparing operation, only one retained his native aortic valve. The actuarial freedom from death or need for aortic root reoperation is depicted in Fig. 2. The actuarial freedom from adverse events was significantly lower in the valve-sparing

### Table 1

Results in children undergoing aortic root replacement.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No. of patients</th>
<th>Mortality</th>
<th>Aortic root reoperations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Composite graft replacement</td>
<td>9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Valve-sparing remodelling</td>
<td>11</td>
<td>1</td>
<td>6/5</td>
</tr>
<tr>
<td>Valve-sparing reimplantation</td>
<td>3</td>
<td>0</td>
<td>1/1</td>
</tr>
<tr>
<td>Total</td>
<td>23</td>
<td>1</td>
<td>7/6</td>
</tr>
</tbody>
</table>

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### Table 2

Summary of 10 operative survivors after valve-sparing remodeling procedure.

<table>
<thead>
<tr>
<th>Patients</th>
<th>Connective tissue disorder</th>
<th>Age (years)</th>
<th>Reop # 1</th>
<th>Reop # 2</th>
<th>Reop # 3</th>
<th>Follow-up (years)</th>
<th>Aortic valve regurgitation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Turner</td>
<td>12</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>9.7</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>Bicuspid aortic valve</td>
<td>12</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>7.1</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>Marfan</td>
<td>3.5</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>Marfan</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>3</td>
<td>++</td>
</tr>
<tr>
<td>5</td>
<td>Marfan</td>
<td>1</td>
<td>MV + TV repair</td>
<td>—</td>
<td>—</td>
<td>8.1</td>
<td>+</td>
</tr>
<tr>
<td>6</td>
<td>Marfan</td>
<td>1.5</td>
<td>Bentall</td>
<td>—</td>
<td>—</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Marfan – BAV</td>
<td>16</td>
<td>AVR</td>
<td>—</td>
<td>—</td>
<td>11.8</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Loeys Dietz – BAV</td>
<td>4</td>
<td>Bentall + arch</td>
<td>—</td>
<td>—</td>
<td>8.2</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Marfan</td>
<td>12</td>
<td>Bentall</td>
<td>Bentall – Konno</td>
<td>—</td>
<td>6.3</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Marfan</td>
<td>4.5</td>
<td>Bentall + arch</td>
<td>MV + TV repair</td>
<td>DTA replacement</td>
<td>12.1</td>
<td></td>
</tr>
</tbody>
</table>

remodelling group (compared to the composite graft group) 
\( p = 0.021 \) at 2 years and \( p = 0.058 \) at 5 years).

The aortic arch had been replaced in three patients and one patient had replacement of the descending thoracic aorta. The mitral valve had been repaired in three patients and replaced in one.

4. Discussion

In contrast with adult patients, aortic root aneurysms are, in children, almost always associated with connective tissue defect syndromes. Marfan syndrome, Loeys–Dietz syndrome and bicuspid aortic valve syndrome account for the majority of cases. Aortic root aneurysms may lead to catastrophic complications (dissection, rupture, acute aortic insufficiency) and, then need urgent and high-risk life-saving operations. Aortic root replacement is therefore indicated before the occurrence of such complications. Although still the matter of some debate, guidelines for the timing of surgery have been established, taking into consideration the importance of the aortic dilatation, the rate of progression of the dilatation and the severity of the underlying connective tissue disorder [1]. In the current series, such guidelines were used. All children, except one, were operated upon either because the aortic root was severely dilated (aortic root z-score >3 according to body size) or because the aneurysm was expanding rapidly (more than 5 mm per year). The last patient, with Turner syndrome, had only moderate aortic dilatation and the rate of progression was only 4 mm in the last year, but hormonal treatment was indicated to initiate puberty and the risk of dissection was thought to be increased [4].

The standard surgical treatment for complete aortic root replacement is represented by the composite valve graft repair or Bentall procedure. Various procedures have been described to replace the aortic root while preserving the native aortic valve (valve-sparing procedures). There is an extensive literature which reports the indications and results of these operations in the adult population [5–12]. On the contrary, pediatric reports are scarce and the optimal surgical management remains controversial [13–15].

4.1. Main findings of the present study

Our experience, although limited, leads to two main conclusions: (1) composite valve graft repair provides excellent long-term results, (2) the results of valve-sparing procedures may be disappointing, particularly regarding the remodeling approach.

The late results of composite valve graft replacement are excellent in the pediatric population [13,14,16]. Our own series includes 15 patients (9 who underwent a Bentall procedure as the first operation and 6 who had the same repair after failure of a valve-sparing operation). There was no late mortality and all patients were leading an unrestricted normal life. There was no valve-related morbidity (endocarditis, thrombo-embolism or hemorrhagic event). Only one patient needed reoperation for patient-prosthesis size mismatch. In most children who need surgery, aortic root dilation is such that the aortic annulus is large enough to accommodate an adult-size prosthesis. When this is not the case, in infancy or early childhood, the aortic annulus can be enlarged by dividing the conal septum (Bentall–Konno procedure); this operation was, however, associated, in our experience, with an increased risk of complete heart block. In infants with a small aortic annulus, aortic root replacement using homografts has been recommended and performed with satisfactory early results. However, homografts degenerate rapidly and reoperation is inevitable and may be risky because of intense calcification of the graft [16]. This approach cannot be recommended anymore.

Even if very rare in children, thrombo-embolic events and endocarditis are potentially lethal and remain a concern after the Bentall operation. Furthermore, long-term anticoagulation may be a major drawback in some patients (girls with potential future pregnancies, patients having lifestyles making anticoagulation hazardous). Procedures in which the native aortic valve is preserved and long-term anticoagulation avoided, represent attractive alternatives.

The first valve-sparing aortic root replacement was described by Sarsam and Yacoub and it is known as valve-sparing remodeling procedure [2]. Satisfactory results have been reported in the adult population, even in patients with Marfan syndrome [6,11]. However, reoperation may be necessary for progressive aortic regurgitation, due to subcommissural triangle stretching and absence of annular stabilization. Aortic root dilatation appears to be earlier and more rapid in children than in adults, leading to a high risk of reoperation [14,17]. This may reflect that patients who require surgery earlier in life have the more severe form of connective tissue defects. In our experience, valve-sparing remodeling procedure was associated with a high rate of failure. Severe aortic regurgitation occurred early in two patients and late in five; in the remaining patients, one had moderate insufficiency at last follow-up. These disappointing results led us to abandon the remodeling surgical approach in children who require aortic root replacement, even in the absence of preoperative aortic regurgitation. The same recommendation has already been made by other groups [14].

The other valve-sparing approach, known as reimplantation technique, provides efficient annular stabilization by implanting a prosthetic conduit directly onto the ventriculo-aortic junction. In the first version (David I procedure), a straight conduit was used and a risk of damage of the aortic leaflets by impingement against the Dacron graft was described. Various technical modifications were reported, all striving to stabilize the ventriculo-aortic junction and recreate the sinuses of Valsalva [10,12,18]. In adults, reimplantation procedures provide superior results in terms of freedom from aortic regurgitation and reoperation, when compared to the remodeling approach [7]. The main concern is the long-term integrity of the aortic leaflets because of increased leaflet stress. Grafts with custom-made [12,18] or prefabricated [10] pseudo-sinuses can be used. Whether the aortic leaflets will last longer when pseudo-sinuses are created remains to be determined. Preliminary results in the pediatric population using the reimplantation technique are encouraging [15]. Our own experience is too limited to draw out useful information. Among three patients, one needed reoperation for endocarditis and one had residual moderate insufficiency.
It has been suggested that, in adults, most bicuspid aortic valves could be preserved during valve-sparing surgery [19]. In our experience, a bicuspid valve was preserved in four cases (remodeling in three, reimplantation in one); there were three failures (one from endocarditis after reimplanta-
tion). In children, in whom at least mid-term valve longevity is desirable, preservation of a bicuspid valve seems justified only when there is no stenosis, no or trivial incompetence and minimal distortion of the leaflets.

Connective tissue disorders may involve the other cardiac valves (particularly the mitral valve) and other distal segments of the aorta. Strict follow-up is mandatory and multiple operations may be necessary in a majority of patients, thus obscuring the long-term prognosis.

4.2. Limitations of the study

The information provided by the present study is obviously limited by the small number of patients with each surgical procedure. This is particularly true for the valve-sparing reimplantation approach. For patients undergoing composite valve graft repair or valve-sparing remodeling surgery, the length and completeness of follow-up make inferences more valid; however, the results must be interpreted with caution since the two groups are not strictly comparable (the patients with valve-sparing surgery were operated upon at a younger age).

5. Conclusions

Children with aortic root aneurysm have connective tissue defects, and most probably, the most severe forms of these disorders. Pathologic tissue must be removed as completely as possible. This goal is achieved using the composite valve graft replacement. The Bentall procedure remains the standard operation and very satisfactory long-term results can be anticipated. It should remain the first choice procedure, particularly when aortic regurgitation is more than minimal, when the aortic leaflets are distorted or when concomitant mitral valve replacement is indicated. In patients with no or minimal aortic insufficiency or in whom long-term anticoagulation is undesirable, valve-sparing procedures are attractive. Remodeling approach should probably be abandoned. Valve-sparing reimplantation with creation of pseudo-sinuses may yield acceptable mid-term results, but further experience is mandatory.

Acknowledgement

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