Valve-sparing root replacement in children with aortic root aneurysm: mid-term results†

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Abstract

OBJECTIVES: We aimed at evaluating the results of aortic valve-sparing root replacement (AVSRR) in children with aortic root aneurysm (ARA) due to genetic disorders in terms of mortality, reoperation and recurrent aortic valve regurgitation (AVR).

METHODS: Thirteen patients (mean age 9.7 ± 6.5 years, 10 months–18 years) underwent AVSRR for ARA between 2002 and 2011. Six of the 13 patients had Marfan syndrome, 3 Loëys–Dietz syndrome (LDS), 2 bicuspid aortic valve syndrome and 2 an unspecified connective tissue disorder. AVR was graded as none/trace, mild and severe in 5, 7 and 1 patient, respectively. The mean pre-operative root diameter was 45 ± 10 mm (mean Z-score 10.3 ± 2.0). Remodelling of the aortic root was performed in 4 patients, reimplantation of the aortic valve in 9 and a concomitant cusp repair in 4. The diameter of the prostheses used for root replacement varied from 22 to 30 mm (mean Z-score = 2.3 ± 3). The follow-up was 100% complete with a mean follow-up time of 3.7 years.

RESULTS: There was no operative mortality. One patient with LDS died 2.5 years after the operation due to spontaneous rupture of the descending aorta. Root re-replacement with mechanical conduit was necessary in 1 patient for severe recurrent AVR 8 days after remodelling of the aortic root. At final follow-up, AVR was graded as none/trace and mild in all patients. Eleven patients presented in New York Heart Association functional Class I and 1 in Class II.

CONCLUSIONS: In paediatric patients with ARA, valve-sparing root replacement can be performed with low operative risk and excellent mid-term valve durability. Hence, prosthetic valve-related morbidity may be avoided. Due to the large diameters of the aortic root and the ascending aorta, the size of the implanted root prostheses will not limit later growth of the native aorta.

Keywords: Aortic aneurysm · Aortic valve · Valve repair · Congenital

INTRODUCTION

The goal of aortic valve-sparing operations, in children as well as adults, is to replace the pathological part of the aortic root complex while preserving the anatomical and functional integrity of the aortic valve leaflets. Nevertheless, the main limitation of aortic valve-sparing procedures is the potential need for reoperation. In contrast to adult patients, for whom reoperations are mainly due to recurrent aortic valve regurgitation (AVR) [1], in growing children the reconstructed aortic valve and the aortic root may become too small for their later body size.

Aortic root or ascending aortic aneurysms in infants and children are almost exclusively associated with rare genetic disorders. Clinical data regarding the surgical therapy of these pathologies are sparse, especially for valve-sparing root procedures.

The principal surgical strategy for infants and children with aortic root aneurysms (ARAs) should not only focus on the replacement of the ascending aorta, but also on a longer-lasting surgical strategy for preserving aortic valve function. Should the valve be replaced or reconstructed? Is reconstruction durable despite the underlying genetic disorder, which may also affect the leaflet tissue?

In the present investigation, we evaluated the results after aortic valve-sparing root replacement (AVSRR) in children with these types of pathologies in terms of mortality, recurrent AVR and reoperation.

PATIENTS AND METHODS

Patient selection and data collection

This study was approved by the Ethics Committee of the Technische Universität München. Two hundred and twenty
patients who were scheduled for aortic valve-sparing procedures between April 2000 and September 2011 at the German Heart Centre Munich were included in our valve-sparing database. All patients who were younger than 18 years of age at time of the operation were included in the present study. Accordingly, root replacement with aortic valve-sparing was performed in 13 infants and children. All paediatric patients without previous operations on the aortic root who were diagnosed with aortic root or ascending aorta aneurysm with or without concomitant AVR were scheduled for aortic valve-sparing operations. The essential requirement for planning the valve-sparing operation was the existence of morphologically intact aortic valve leaflets.

In addition to the 13 patients from this cohort, 3 other patients were scheduled for aortic valve-sparing root replacement. During the operation, it turned out that the valve leaflets were very thin and fenestrated or too thick and fibrotic. As a consequence of the intraoperative findings, Bentall operations with a mechanical conduit had to be performed on all 3 patients. Pre- and postoperative AVR and left ventricular dimensions were assessed by transthoracic and transoesophageal echocardiography. According to Anderson and co-workers, the diameters of the aortic valve annulus (virtual ring formed by joining basal attachments of aortic valvar leaflets), the aortic root (sinus of valsalva diameter), the sino-tubular junction, the aortic arch and the descending thoracic aorta at diaphragm were measured by magnetic resonance imaging (MRI), computed tomography (CT) or echocardiography and expressed as an absolute value and an indexed score for body surface area (Z-score) [2]. The Z-scores were calculated based on the regression equations for the calculation of Z-scores of cardiac structures according to Pettersen et al. [3].

AVR was graded as none/trace, mild, moderate or severe. The graduation was not only based on the length/area of the regurgitant jet, but also on the location and duration of colour-coded backflow in the ascending, descending or abdominal aorta [4, 5]. The ventricular function was graded using 2-dimensional echocardiography and evaluated as normal (ejection fraction ≥55%), moderately impaired (ejection fraction 30–54%) or severely impaired (ejection fraction <30%). All patients were prospectively studied with annual clinical assessment and echocardiography. The functional status was determined according to the New York Heart Association (NYHA) class.

**Study group**

There were 12 boys and 1 girl in our study group, with a mean age of 9.9 ± 6.3 years (range 10 months–18 years). Table 1 shows the demographic data of all patients. Six of the 13 patients had Marfan syndrome (MFS), 3 Loeys–Dietz syndrome (LDS), 2 bicuspid aortic valve (BAV) syndrome and 2 an unspecified connective tissue disorder. In all patients, genetic tests were performed and a genetic disorder was confirmed in the Marfan and Loeys–Dietz patients. The 2 patients with the unspecified connective tissue disorder were not genetically tested until now. Both had clinical signs of MFS, as presented in the revised Ghent criteria, but insufficient to confirm the MFS. They were diagnosed with a pectus excavatum and both had a positive family history. Furthermore, 1 of the patients presented also with myopia and an inguinal hernia. None of the patients received an emergency operation. Two patients had undergone a previous cardiac operation. One patient received a mitral valve and tricuspid valve repair 12 days prior to the VSRR. Another patient had had repair of an aortic coarctation 9 years earlier.

Preoperative echocardiographic data were available in all patients. One patient with LDS also presented with a BAV. The AVR was diagnosed to be none/trace, mild and severe in 5, 7 and 1 patient, respectively. All patients presented in sinus rhythm. The mean preoperative root diameter was 45 ± 10 mm (mean Z-score = 10.3 ± 2.0). Preoperative diameters of the aortic annulus, the aortic root, the sinotubular junction, the aortic arch, the descending thoracic aorta and the left ventricle with the corresponding Z-scores are depicted in Table 2.

**Surgical technique**

The techniques performed for replacement of the dilated aortic root were remodelling in 4 patients and reimplantation in 9. If prolapse of the aortic valve leaflets was present, a concomitant cusp repair was performed. The remodelling of the aortic root technique was performed by replacing the dilated aortic root with a scalloped tubular Dacron graft using a single suture line above the surgical annulus and thus creating artificial sinuses of Valsalva as described by Yacoub and co-workers [6]. In none of the patients was a reinforcement of the aortic annulus performed. Starting in 2004, the reimplantation technique was performed by resection of the aortic sinuses and implantation of the aortic valve annulus inside a straight Dacron or a prefashioned Gelweave Vascutek Valsalva graft (Vascutek) using two proximal suture lines, as described by David and co-workers [7]. The remodelling technique was mainly performed in the earlier patients, and the reimplantation technique was started after our team had made a general switch to the respective technique in all patients. Regarding the diameter of the prosthesis, we follow the recommendation of David and co-workers [8] and De Paulis and et al. [9] measuring the aortic annulus and adding 4–6 mm. In this special group of paediatric patients, the prosthesis size cannot always be chosen using this rule, due to the abnormality of almost all components of the aortic root. Hence, also as described by David, we always pull the three commissures upward and approximate them until the cusps touch each other centrally. The diameter of the circle that includes all three commissures represents the diameter of the vascular graft we choose.

**Follow-up**

Follow-up was conducted annually after the operation by telephone contact with the patient and the referring cardiologist. The clinical assessment and the echocardiography were accomplished either by the referring cardiologist or in our institution. The latest follow-up examinations included electrocardiogram, Doppler echocardiography and physical examination. All data were entered into our database. Follow-up was 100% complete with a mean follow-up time of 3.7 years (range 2 months–9.6 years). The cause of death was determined from hospital records.

**Statistical analysis**

Categorical variables were reported using the number and percentage of observations. Continuous variables were reported as mean ± standard deviation or median with ranges. The outcome
parameters were defined as the time interval from the primary operation to death, aortic valve or aortic root-related reoperation or a recurrent AVR. The probability of freedom from events was estimated according to the Kaplan–Meier method.

**RESULTS**

Operative characteristics and in-hospital complications

The operative data of the 13 patients are depicted in Table 3. The diameter of the prosthetic graft implanted varied from 22 to 30 mm with a mean Z-score of 2.3 ± 3. Four of the 12 patients underwent a concomitant aortic valve repair during the initial operation, by means of subcommissural annuloplasty in 3 patients and free edge plication in 1. In 1 patient, the non-coronary cusp had to be reinforced using an autologous pericardium.

One patient needed a patch enlargement of the left main coronary artery and a subsequent stent implantation. The patient needed temporary support by extracorporeal membrane oxygenation and was weaned successfully on the 10th postoperative day. One patient for whom the aortic valve reimplantation procedure was followed with concomitant mitral and tricuspid valve repair developed complete atrioventricular block and underwent pacemaker implantation.

**Survival**

There were no operative deaths. One patient with LDS died 2.5 years after the primary operation due to rupture of the
The estimated overall 5-year survival probability in the presented cohort is 86 ± 13% (Fig. 1).

Reoperations

During the initial hospitalization, 1 patient with MFS who had undergone a remodelling procedure and extensive aortic cusp repair developed recurrent AVR and underwent reoperation 11 days after the primary operation. A Bentall operation with a mechanical conduit (St. Jude 27 mm, St. Jude, USA) was performed upon the request of patient’s parents. The patient recovered uneventfully after the reoperation and was discharged on the 51st postoperative day. At final follow-up, the patient was alive and NYHA Class I.

Another with MFS exhibited an acute dissection of the residual native ascending aorta, the aortic arch and the descending aorta originating from the distal anastomosis with the aorta 4 years and 7 months after the initial operation. Upon reoperation, the patient underwent a complete replacement of the aortic arch with a 28-mm Hemashield vascular conduit and an antegrade 26-mm Medtronic Valiant stent implantation into the descending aorta. The patient recovered uneventfully after the reoperation and was discharged on the 26th postoperative day. For months prior to the reoperation, the patient had presented for his annual clinical examination in NYHA Class I, showing the normal aortic arch and descending aorta diameters on CT scan.

A third reoperation was necessary in 1 patient with LDS due to dilatation of the native aortic arch 2 years and 10 months after the initial operation. Upon reoperation, the patient underwent a replacement of the residual ascending aorta and the aortic arch with a 24-mm Hemashield vascular conduit. The patient recovered uneventfully and was discharged on the 21st postoperative day.

Table 3: The operative data

<table>
<thead>
<tr>
<th>Patients</th>
<th>Age (month/year)</th>
<th>VSRR operation</th>
<th>AV repair</th>
<th>Concomitant procedure</th>
<th>ACCT (min)</th>
<th>Prostheses Ø (mm)</th>
<th>Z-score</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10 months</td>
<td>David</td>
<td>No</td>
<td>–</td>
<td>124</td>
<td>22</td>
<td>6.7</td>
</tr>
<tr>
<td>2</td>
<td>11 months</td>
<td>David</td>
<td>Yes</td>
<td>PDA ligation</td>
<td>147</td>
<td>24</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>3 years</td>
<td>Yacoub</td>
<td>No</td>
<td>ASD patch</td>
<td>143</td>
<td>24</td>
<td>2.7</td>
</tr>
<tr>
<td>4</td>
<td>4.5 years</td>
<td>Yacoub</td>
<td>No</td>
<td>Fistula closure</td>
<td>57</td>
<td>26</td>
<td>3.9</td>
</tr>
<tr>
<td>5</td>
<td>5 years</td>
<td>David</td>
<td>No</td>
<td>LCA patch</td>
<td>227</td>
<td>26</td>
<td>5.5</td>
</tr>
<tr>
<td>6</td>
<td>11 years</td>
<td>David</td>
<td>No</td>
<td>–</td>
<td>134</td>
<td>30</td>
<td>4</td>
</tr>
<tr>
<td>7</td>
<td>11.4 years</td>
<td>Yacoub</td>
<td>No</td>
<td>–</td>
<td>1128</td>
<td>26</td>
<td>−0.6</td>
</tr>
<tr>
<td>8</td>
<td>11.6</td>
<td>David</td>
<td>Yes</td>
<td>–</td>
<td>142</td>
<td>24</td>
<td>2</td>
</tr>
<tr>
<td>9</td>
<td>13.5</td>
<td>David</td>
<td>No</td>
<td>PDA ligation</td>
<td>110</td>
<td>28</td>
<td>1.7</td>
</tr>
<tr>
<td>10</td>
<td>15.3 years</td>
<td>Yacoub</td>
<td>No</td>
<td>–</td>
<td>105</td>
<td>26</td>
<td>−1.1</td>
</tr>
<tr>
<td>11</td>
<td>16.5 years</td>
<td>David</td>
<td>Yes</td>
<td>–</td>
<td>185</td>
<td>30</td>
<td>−0.5</td>
</tr>
<tr>
<td>12</td>
<td>16.5 years</td>
<td>Yacoub</td>
<td>No</td>
<td>TVP</td>
<td>113</td>
<td>26</td>
<td>0</td>
</tr>
<tr>
<td>13</td>
<td>18 years</td>
<td>David</td>
<td>Yes</td>
<td>–</td>
<td>101</td>
<td>30</td>
<td>2.7</td>
</tr>
</tbody>
</table>


Left ventricular and aortic valve function

Left ventricular function at final follow-up was graded as normal (≥55%) in all patients. AVR was assessed none or less or equal to mild in all patients. None of the patients presented with aortic valve stenosis.

Thromboembolic and neurological events

During the entire follow-up period, there were no thromboembolic or neurological events in the presented cohort.

DISCUSSION

Aneurysms of the aortic root or ascending aorta are uncommon pathologies in the paediatric population and are generally associated with connective tissue disorders, such as MFS, LDS or the BAV syndrome [10, 11]. The clinical manifestation of these
genetic disorders may be highly variable, however, their common feature is the pathology of the aortic complex [10, 11]. Due to an improved understanding of these disorders and the availability of non-invasive screening by echocardiography, the aortic pathologies may be identified early during childhood. For adult patients, the risk of aortic dissection and rupture has been well described on the basis of the diameter of the ascending aorta [12], and consequently, recommendations for treatment have been given in numerous reports [13–15]. However, for children, in whom the risk of rupture is not known, guidelines are not available.

During the last decade, different groups reported good long-term results with the Bentall operation using composite mechanical valved conduits in paediatric patients presenting with aneurysms of the ascending aorta [16, 17]. However, patients with a mechanical prosthesis require life-long anticoagulation with the cumulative risk of thromboembolism, bleeding events and lifestyle impairment. Thus, for adult patients with normal or repairable aortic valves, the Bentall technique has been replaced by valve-sparing root replacement techniques. The rationale for valve-sparing procedures is to preserve the functionality and superior haemodynamics of the native aortic valve, while replacing the dilated aortic root, and thus avoiding the typical complications associated with mechanical or biological prostheses.

As a consequence of the encouraging results of valve-sparing techniques in adults, the indications have been extended to paediatric patients [18, 19] also. A main limitation of aortic valve-sparing procedures is the potential need for reoperation, which may be occasionally warranted in adult patients because of recurrent AVR. However, in paediatric patients, there are additional aspects to consider: What are the indications for this kind of major surgery in infants and children? Should aortic valve-sparing root replacement techniques be performed with a risk similar to that in adult patients and are these techniques successful in the mid- and long-term, considering the children present almost exclusively with connective tissue disorders?

To date, no guidelines are available for infants and children, and therefore, the indication for surgery has to take into consideration the underlying connective tissue disease and the degree of aortic dilatation, the rate of diameter progression per year, the left ventricle function and the severity of AV regurgitation [20, 21]. Patel et al. recently published their excellent series of valve-sparing surgery in 56 children with MFS and LDS. For patients with MFS, they recommend surgery at an aortic root diameter of 5.0 cm or an annular progression of 1.0 cm/year. They do not give specific recommendations for the indication for surgery in children with MFS, arguing that these children usually present late when the threshold diameter of 5.0 cm is already reached. For patients with Loesys-Dietz Type I (severe craniofacial features), they advise surgery at an aortic root Z-score of 3.0 or an aortic root expanding rate of 0.5 cm/year and for children with Loesys-Dietz Type II (mild craniofacial features) at an aortic root Z-score of 4.0 or an aortic root that expands 0.5 cm/year [20].

In children with MFS, we also advocate operation if the 5-cm level is reached or if the progression rate of the aortic root dilatation is >0.5–1 cm/year, depending on the concomitant aortic root growth [13, 17, 22]. Nevertheless, knowing that early mortality in children with MFS may occur due to cardiovascular complications [23] and that the results of cardiac operations in these children compare favourably with those in adults [17], we advocate root replacement before reaching the range of 5 cm if the size of the aortic root exceeds a Z-score of 5 and also accepts the implantation of a vascular prosthesis, which will be sufficient for the children to grow. Children with the Loesys-Dietz syndrome usually present earlier than the Marfan patients. In this special group, the Z-score may reach values far beyond the recommended values of 3–4 as suggested as threshold value for root replacement by Patel and co-workers. Therefore, the indication for surgery may not in all cases be based on the aortic root Z-score alone. We advocate that root replacement should be postponed despite the possible high Z-scores, until the annulus diameter reaches a value of at least 18 mm so that a prosthesis of not less than 22 mm may be used for the reconstruction.

For valve-sparing operations, we require an annulus diameter of at least 18 mm so that a prosthesis of not less than 22 mm may be used for the reconstruction. If smaller prostheses are implanted, the child will face reoperation with valve replacement later in life. In the present cohort, the diameter of the aortic annulus was at a minimum of 18 mm in all children. Based on a Z-score of 5.0 for the annulus as the threshold for surgery, the diameter of the annulus would theoretically have been only 13–17 mm in 3 children (age 6 months–5 years), which we would have considered too small for valve-sparing operations. Hence, the Z-score of the aortic root may have only limited value in small children, and we advise waiting until the aortic annulus diameter has reached at least 18 mm for these patients. Since the risk of spontaneous rupture is reported to be low in children, this recommendation may be justified.

In this small group of 13 paediatric patients presenting with aortic root or ascending aorta aneurysm, with or without AVR, we demonstrated that aortic valve-sparing procedures could be performed at a very low risk with excellent results. Furthermore, the durability of the spared aortic valve in the mid-term is at least similar to the results observed and reported for adult patients with connective tissue diseases with freedom from AVR more than trace having been shown to be between 90 and 100% after 5 years [24]. In the present investigation, the mean Z-score for the aortic annulus was 5.6 ± 3.2 and for the aortic root, 10.3 ± 2.0. Therefore, we believe that valve-sparing root replacement is generally feasible in paediatric patients with connective tissue disorders, because the preoperative diameters are large enough to allow the implantation of an adult-size vascular prosthesis. Consequently, the prostheses implanted in paediatric patients with ARA will usually not be outgrown later in life.

In our initial experience, the remodelling technique was applied to all patients with ARA in order to minimize leaflet stress by recreating the aortic sinuses. However, due to a higher reoperation rate observed in our adult Marfan patients for progressive annular dilatation and recurrent AVR [25], we abandoned the remodelling operation in favour of the reimplantation technique. In all patients, except for one, a prefashioned Valsalva graft was used for the reimplantation technique. Only in one 10-month-old infant, a 22-mm straight Dacron prosthesis was implanted, because of the very small annulus (18 mm) and the very small proximal part of the aortic arch (12 mm). In this special case, the smallest commercially available prefashioned Valsalva graft (24 mm) would have led to a severe graft to aorta mismatch.

None of our patients underwent arch replacement at primary operation, as almost none presented with a dilated aortic arch. Nevertheless, during the follow-up period, reoperations on the aorta were necessary in 2 of the 13 patients, 1 Marfan patient and 1 Loesys–Dietz patient. Both underwent a replacement of the residual native ascending aorta and aortic arch.
In our experience with >70 Marfan patients, a reoperation on the aortic arch after ascending aorta replacement was necessary in 7% of the patients. Of course, by replacing a normal aortic arch during the aortic root replacement operation, one may expose the patient to additional morbidity, which in our opinion, is outweighed by the risk of reoperation. Hence, in Marfan patients, we advocate concomitant aortic arch replacement in patients with a dilated or normal-diameter aortic arch but frail and thin aortic walls.

As described by the group from Baltimore, the Loeys-Dietz patients do develop arch enlargement early. Thus, in order to avoid early reoperation or dissection, a concomitant prophylactic arch replacement during the initial root replacement operation should be taken into consideration even in the presence of normal diameters, depending also on the age and the clinical state of the patient.

Postoperative patient surveillance is mandatory in these patients. In the present cohort, in 2 of the 3 patients with major adverse events, such as death and aortic reoperation, the incidents occurred as a consequence of the structural deterioration of the arterial wall due to the connective tissue disorder. Hence, we advocate echocardiographic and extended CT or MRI examinations in these special patients every 6–12 months after the operation [20].

Study limitations

The main limitation of this study is the small number of patients, which precludes a subanalysis regarding detailed surgical techniques or age groups. Furthermore, the reduced follow-up time also does not allow statements on long-term results.

CONCLUSIONS

In paediatric patients with ARA and connective tissue disorders, AWSRR can be performed at low-operative risk and with excellent mid-term valve performance. Hence, prosthetic valve-related morbidity may be avoided. Due to the large diameters of the aortic root and the ascending aorta, the size of the implanted root prostheses will not limit later growth of the native aorta. However, the aortic annulus should be at least 18 mm for a 22-mm prosthesis to fit. Following this recommendation, reoperation due to a potential outgrowth of the annulus diameter may not be required if the valve function remains stable in the long-term.

Conflict of interest: none declared.

REFERENCES

APPENDIX. CONFERENCE DISCUSSION

Dr R. Prêtre (Zurich, Switzerland): This is a relatively large series of a difficult subset of patients, and the challenge actually, in my opinion, is not so much to perform a valve-preserving operation (we more or less know how to do that), as to decide when to do it.

Even if you are dealing with a paediatric population, I think you have two groups of patients: those kids who have not yet reached full growth and those teenagers who have. What are your criteria for operating on these patients? Because in your manuscript you said you use the same criteria as in adults, but I saw you used z-values of 6 or 7. So, can you elaborate a little bit on the indication criteria in these two groups of patients?

And my second question relates to the very small kids; you had five such kids under 10 years of age. We have described some overgrowth of leaflet tissue and redundancy of leaflet tissue leading to valve dysfunction, stenosis or insufficiency when a valve-preserving operation was done too soon. Have you seen that in your population?

Dr Badiu: Regarding your first question, we did not use the adult criteria for the paediatric population; of course, for the 18 year-old Marfan patient, we used the adult criteria. In children, we are trying to postpone the operation for as long as it is possible. We see these patients for several months or years prior to the operation, and we decide to perform the operation only when we see that there is a progressive dilatation of the aortic root.

In the Loeys-Dietz children, it is a little bit different. We had three Loeys-Dietz patients in the group (10 months, 11 months, and 5 years of age at the time of primary operation) who were seen two or three months prior to the operation. When they presented, they had huge aortic roots of 30 mm or more with z-scores around 12. The heart team (including the cardiologist) decided that it was very risky to postpone the operation in these circumstances.

You are right, there are two different age groups. Maybe one can wait a little bit longer in the 10 or 12 year-old patient with a 40 mm root, but we advocate early operation in very young patients with these huge roots with massive Z-scores.

Dr Prêtre: I will ask the last question again. You did not operate on any kids on an emergency basis for an aortic event? And are you aware of some kids who have died before operation, for instance?

Dr Badiu: No. In our clinic, we have not had any patients dying prior to operation, that is right. But the reason for this may be that we performed surgery early when these excessively dilated roots were diagnosed.

Concerning your second question, in this small group we had five children aged less than 10 years at primary operation. In three out of the five, we had follow-up of at least three years, and until now we have good aortic valve function without recurrent regurgitation. Echocardiography has not shown any outgrowth tissue interfering with valve movement.