Outcomes After Operations for Bicuspid Aortic Valve Disease in the Pediatric Population

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Background. Outcomes after operations for bicuspid aortic valve disease in pediatric patients were determined.

Methods. Between 1977 and 2011, 146 consecutive patients underwent surgical repair of bicuspid aortic valve. Median age at operation was 207 days (range, 5 days to 16 years). Indication for surgery was stenosis in 113, insufficiency in 25, and both in 8. Valve debridement was done in 76 patients, and complex repairs in 70, including 61 who required addition of pericardial patches, consisting of the creation of a neocommissure in 55, cusp extension in 33, and a perforation repair in 6. The valve was made tricuspid in 38 patients (29 cusp extensions).

Results. Twenty-year survival was 88% (95% confidence interval, 73% to 95%). After a mean follow-up of 8 ± 7 years, 35 patients needed a reintervention. Freedom from reintervention at 18 years was 43% (95% confidence interval, 73% to 95%). After a mean follow-up of 8 ± 7 years, 35 patients needed a reintervention. Freedom from reintervention at 18 years was 43% (95% confidence interval, 73% to 95%). At the latest follow-up, an additional 13 patients without reoperation had moderate or severe stenosis, and 17 had moderate regurgitation. Seventy-eight patients had an event-free long-term outcome (no reintervention, stenosis, or regurgitation). The only independent predictive factors of an event-free outcome were not having addition of patch material at repair (hazard ratio, 12; p = 0.05) and shorter bypass time (HR, 1.01; p = 0.023). The 10-year freedom from any significant event was 60% (95% confidence interval, 46% to 71%) for those without use of patch material, whereas nearly all those with a patch repair had an adverse event at that time.

Conclusions. Outcomes after surgical repair of bicuspid aortic valves in the pediatric population are excellent, especially if the repair can be performed without the addition of patches. Primary repair should be offered because long-lasting results can be achieved if the disease can be relieved by simple procedures.


Up to 2% of the population bears a bicuspid valve, and longitudinal studies performed in adults estimate that a quarter of them require an operation over a span of 10 to 20 years [1]. The development of techniques of aortic valve repair for the treatment of aortic valve regurgitation in adults has focused a lot of attention on bicuspid aortic valve disease because it seemed ideally suited for repair [2–4]. Bicuspid aortic valve disease is often associated with anomalies of the media, responsible for early dilatation of the aortic root and subsequent dilatation of the ascending aorta [5]. This has prompted the American Heart Association/American College of Cardiology to recommend a lower threshold to replace the ascending aorta of patients undergoing operations when their valve is bicuspid [6].

Very little information has been published on the outcomes of bicuspid aortic valves in the pediatric population. The disease appears to have different characteristics than in its adult counterpart, with a predominance of stenosis at an early age [7]. Balloon valvuloplasty has become the most widespread treatment approach for the management of pediatric aortic valve stenosis. Recent studies have reported favorable outcomes after pediatric aortic valve repair, but the indications of this approach are still disputed [7–11]. Interestingly, the same American
Heart Association/American College of Cardiology guidelines recommend balloon valvuloplasty as the primary treatment of aortic valve stenosis in the pediatric age group and reject this practice in adults [6].

During the course of the last 15 years, our team has favored primary aortic valve repair of all aortic valve lesions. We reviewed the results of this approach to determine the long-term outcomes of aortic valve repair in pediatric bicuspid aortic valve disease.

Patients and Methods

The local Human Research Ethics Committee approved the study design, and the need for consent was waived because of the retrospective nature of the study. The files of all patients who underwent aortic valve repair in the Royal Children’s Hospital, Melbourne, were reviewed. Patients with single-ventricle palliation or rheumatic heart disease or living overseas were excluded. Between 1977 and 2011, 146 consecutive patients (39 females, 107 males) with a bicuspid aortic valve underwent aortic valve operations in our institution.

Operations

The median age at time of intervention was 207 days (interquartile range, 5 days to 16 years). Of these, 39 were neonates (<30 days), 42 were infants (interquartile range, 31 to 354 days), 58 were aged between 1 and 18 years, and 7 were older than 18. The primary indication for operation was aortic stenosis in 113, aortic insufficiency in 25, and mixed aortic stenosis and insufficiency in 8. The aortic valve was exclusively stenotic in neonates and infants, and the distribution of indication in the remaining patients is given in Figure 1.

Thirty-three patients had a prior cardiac intervention. Among those, 21 had a previous intervention on the aortic valve: 2 underwent previous commissurotomy at another institution and 19 had a balloon valvuloplasty. Additional previous cardiac procedures were coarctation repair in 9, ventricular septal defect closure in 1, relief of left ventricular outflow tract obstruction in 2, aortic arch repair in 1, and mitral valve repair in 1. The following associated cardiac anomalies were present in 39 patients: coarctation in 17, mitral regurgitation in 7, mitral stenosis in 4, ventricular septal defect in 8, atrial septal defect in 4, subaortic stenosis in 6, cardiomyopathy in 2, and 1 patient each with endocardial fibroelastosis, bilateral superior vena cava, aortic arch hypoplasia, partial anomalous pulmonary venous drainage, tricuspid incompetence, and pulmonary artery stenosis.

The mean bypass time was 92 ± 51 minutes. An additional 30 procedures were performed in 25 patients concomitantly with the valve operation: coarctation repair in 11, ventricular septal defect closure in 7, relief of left ventricular outflow tract obstruction in 3, atrial septal defect closure in 3, and 1 patient each with mitral valve repair; partial anomalous pulmonary valve drainage repair, relief of supravalvular stenosis, ventriculoplasty, post-switch repositioning of a coronary artery, and pulmonary valve repair.

Hospital mortality was defined as death before hospital discharge or within 30 days of the operation. Late mortality was defined as death after discharge or more than 30 days from the first valve operation. Early reinterventions were defined as any operation on a previous valve repair or replacement before hospital discharge and were considered separate to late reoperations occurring after hospital discharge. Reporting of valve-related outcomes, such as valve thrombosis and bleeding events, was based on published guidelines [12].

Follow-up information was gathered from the hospital database or collected from the referring cardiologists. Because gradients through the aortic valve were not consistently reported, the severity of valve stenosis and regurgitation was extracted from cardiologists reports and were separated in categories of less than moderate and at least moderate.

Statistical Analysis

Statistical analysis was performed using Stata 10 software (StataCorp LP, College Station, TX). Continuous data are expressed as mean ± standard deviation or as median (interquartile range). All tests of patient characteristics were two-tailed. Kaplan-Meier survival analysis was used to estimate freedom from the time-related outcomes of late death, reintervention, valve replacement, restenosis and reintervention combined, and regurgitation before a first reintervention. Risk factors...
for time-related outcomes were tested from the date of the first valve operation using Cox regression analysis. For development of late regurgitation, a patient was considered to be no longer at risk once a surgical reintervention was required. All preoperative and perioperative variables were tested for their association with early and late outcomes. Univariate analysis identified variables most likely to be associated with the given event, which were then entered in a stepwise fashion into multivariable analysis.

Results

During the course of our experience, the techniques applied to perform the repairs varied considerably. Initially, the repairs were limited to blade commissurotomies of fused commissures [13]. Progressively, more extensive resection of fibrotic tissues and nodular dysplasia was performed. The first re-creation of a neo-commissure with patches of glutaraldehyde autologous pericardium was performed in 1996, and that operation became routine thereafter. Starting in 1999, we added cusp extension techniques to our armamentarium of techniques [14]. It was only in 2000 that adult techniques were applied to regurgitant aortic valves (Fig 2). [15]

Simple repairs consisting of commissurotomy, with or without extensive resection of nodular dysplasia and thinning of leaflets, were performed in 76 patients. The remaining 70 patients underwent a more complex repair. From these, 61 underwent repair using pericardial patch(es): a neocommissure was created in 55, and a perforation was repaired in 6. A cusp extension was performed in 33 patients, and in 29 of these, all 3 leaflets were extended. An additional 9 patients underwent tricuspidization without cusp extension. Of the 9 patients who underwent repair without use of patch material, a triangular resection was performed in 6, plication of the free edge in 2, and subcommissural annuloplasty in 5.

Survival

There were 2 early deaths (1%). One death occurred suddenly after hospital discharge in a 14-year-old diabetic patient. Multisystem organ failure led to the death of a 3-month-old patient on the 19th postoperative day. An early reintervention was necessary in 3 patients during the same hospital stay. One patient sustained a

Fig 2. Examples of techniques of repair. (A) Tricuspidization and cusp extension. 1. Bicuspid valve. 2. Incision of the raphe. 3. Cusp extension with patches of pericardium of the tricuspidized valve. (B) Commisurotomy and creation of a neocommissure with a pericardial patch with resection of the raphe and insertion of a triangular patch. 1. Bicuspid valve with thickened raphe and commissural fusion. 2. Incision of the commissure, resection of the dysplastic fibrosis and creation of a neo-commissure with a pericardial patch. 3. Resection of the thickened raphe and insertion of a triangular pericardial patch. (Reproduced from Heart, Congenital heart disease: aortic valve surgery in children, Yves d’Udekem, Vol 97, Pages 1182–89, Copyright 2011, with permission from BMJ Publishing Group Ltd. [15])
cardiac arrest on the second postoperative day. He was urgently supported with extracorporeal membrane oxygenation and ultimately received a heart transplant 2 months later. One neonate who underwent a valvotomy underwent a subsequent balloon dilatation on postoperative day 9. The last patient was a neonate who required a repeat surgical valvotomy 15 days after his initial procedure.

The mean peak gradient through the aortic valve decreased from 67 ± 38 mm Hg before the operation to 27 ± 17 mm Hg immediately after the operation.

The median follow-up in hospital survivors was 4.5 years (interquartile range, 1 to 24 years). Only 2 patients were lost to follow-up. There were 7 late deaths (5%), which occurred between 79 days and 22 years postoperatively. The causes of death were sudden unexplained death in 1 patient, and arrhythmia, a noncardiac cause, and unknown in 2 patients each. Survival was 95% (95% confidence interval [CI], 89% to 97%) at 10 years and 88% (95% CI, 73% to 95%) at 20 years. Univariate analysis identified having another cardiac anomaly as the only independent predictor of death (hazard ratio [HR], 5.2; 95% CI, 1.3 to 21; p = 0.02). No subsequent neurologic or thromboembolic events were reported during follow-up. A reintervention for endocarditis was necessary in 3 patients.

Freedom From Reintervention
Thirty-five patients had the following first late reintervention after a median of 7.4 years (interquartile range, 1.4 to 13 years): Ross procedure in 17, redo aortic valve repair in 9 (6 consisting of tricuspidization and cusp extension), redo commissurotomy in 3, balloon valvuloplasty in 3, and mechanical valve replacement, aortic homograft valve replacement, and Bentall procedure in 1 patient each. The indications for reoperation were aortic stenosis in 14, insufficiency in 11, endocarditis in 1, and mixed stenosis and insufficiency in 7. Ten patients underwent a second reintervention, including a Ross procedure in 9 and an aortic valve repair in 1. The indications for second reintervention were endocarditis in 2, aortic stenosis in 4, and insufficiency in 4.

Freedom from a first reintervention at 5, 10, and 18 years, respectively, was 87% (95% CI, 78% to 92%), 64% (95% CI, 51% to 75%), and 43% (95% CI, 28% to 56%; Fig 3). Multivariable analysis identified longer bypass time (HR, 1.0117; 95% CI, 1.004 to 1.019; p = 0.002) and being a neonate at time of the operation (HR, 2.5; 95% CI, 1.22 to 5.1; p = 0.012) as predictor of reintervention.

Freedom From Valve Replacement
Ultimately, 29 patients underwent a valve replacement, consisting of a Ross procedure in 26, replacement with a mechanical valve in 2, and an aortic homograft in 1.

By univariate analysis, the predictors of valve replacement were creation of a neocommissure (HR, 3.17; 95% CI, 1.3 to 7.7; p = 0.011), use of patch material (HR, 2.97; 95% CI, 1.3 to 7.03; p = 0.013), having a more complex repair (HR, 2.56; 95% CI, 1.1 to 5.92; p = 0.028), and longer bypass time (HR, 1.01; 95% CI, 1.007 to 7.01; p < 0.001). Longer bypass time was the only predictor identified by multivariable analysis (HR, 1.02; 95% CI, 1.01 to 1.02; p = 0.001). Freedom from valve replacement was 67% (95 CI, 64% to 77%) at 10 years and 59% (95% CI, 45% to 71%) at 18 years (Fig 4). The median time to valve replacement was 7 years (interquartile range, 3 to 10 years).

Freedom From Restenosis or Reintervention
At the latest follow-up, 13 of the 108 patients who did not undergo a reintervention were considered by their cardiologists to have at least moderate stenosis. Their mean peak gradient was 61 ± 20 mm Hg. Freedom from both more than moderate stenosis and reintervention was 54% (95% CI, 42% to 65%) at 10 years and 36% (95% CI, 24% to 48%) at 18 years. At the latest follow-up, the gradient of those who did not undergo a reintervention and were considered to have less than moderate stenosis was 28 ± 13 mm Hg. Multivariable analysis identified a higher peak gradient before the operation (HR, 1.012; 95% CI,
1.003 to 1.021; \( p = 0.01 \) and longer bypass time (HR, 1.007; 95% CI, 1.001 to 1.013; \( p = 0.02 \)) as being predictive of restenosis or reintervention.

**Freedom From Regurgitation Before a First Reintervention**

At the latest follow-up or just before the first reintervention, 41 patients had moderate or greater regurgitation. Of these 41 patients, regurgitation was moderate in 23, moderate to severe in 8, and severe in 10. Of the 108 patients who had not needed a reintervention, 17 were left with moderate regurgitation at the last follow-up. By univariate analysis, having a complex repair (HR, 4; 95% CI, 1.9 to 8.9; \( p < 0.001 \)) was predictive of late valve regurgitation. Freedom from more than moderate regurgitation was 64% (95% CI, 52% to 74%) at 10 years and 44% (95% CI, 31% to 57%) at 18 years. Freedom from more than moderate regurgitation at 10 years was 78% (95% CI, 64% to 87%) for those who had a simple repair compared with 40% (95% CI, 21% to 59%) for those whose repair was more complex.

**Event-Free Long-Term Outcome**

At the latest follow-up, 78 patients had an event free long-term outcome, defined as requiring no reintervention and having less than moderate stenosis and moderate regurgitation. Specific outcomes in patients who had previous balloon dilatation and those who underwent the creation of neocommissure, who had addition of a pericardial patch, and those who had a tricuspidization with cusp extension are given in Table 1. Multivariable analysis isolated having repair using patch material (HR, 12; 95% CI, 1.0012 to 136; \( p = 0.05 \)) and longer bypass time (HR, 1.01; 95% CI, 1.001 to 1.02; \( p = 0.023 \)) as significant predictive factors for not having an event free long-term outcome. Freedom from any significant event was 48% (95% CI, 37% to 58%) at 10 years and 30% (95% CI, 19% to 41%) at 18 years (Fig 5). The 10-year freedom from any significant event was 60% (95% CI, 46% to 71%) for those without use of patch material, whereas nearly all patients with a patch repair had experienced an adverse event at that time (Fig 6).

**Comment**

This review of our experience enlightens the pathologic features of bicuspid aortic valve disease in the pediatric age group. It confirms the predominance of stenosis in younger patients and shows that it is only as patients are getting closer to adulthood that their symptoms become predominantly regurgitant. Our experience is that in the pediatric age group, even predominantly regurgitant valves carry stenotic components with some degree of commissural fusion and, often, restriction of the motion of the leaflet bearing the raphe.

The first aortic balloon valvuloplasty was reported in 1983, and the subsequent years saw a formidable expansion of the use of this procedure, so that today only a few centers still favor surgical intervention as the primary line of treatment of aortic valve stenosis in pediatric patients.
The aortic valve operation in neonates and younger children has made enormous progress since the time when it was limited to blade commissurotomies or even transapical balloon dilatation [16]. Today, because of the emergence of refined techniques, historical comparisons of balloon valvuloplasty and surgical intervention have become obsolete, but the belief that the operation and interventional catheterization yields equivalent results persists.

Here we report the long-term outcomes of our experience with surgical intervention for aortic valve repair spanning more than 3 decades. Because the operations described were performed during this historical period, the current report is limited in its ability to predict contemporary outcomes after valve repair because it reflects a “trial-and-error” experience rather than the analysis of a consistent approach. In addition, it is impossible to discern retrospectively whether there were subtle selection biases in the attribution of patients to surgical intervention in the period when balloon valvuloplasty and operative repair were both offered.

Despite these limitations, the remarkable finding of our study is that just over half of our patients undergoing bicuspid aortic valve repair were still with their native valve 20 years after repair, a result that we believe cannot be matched by balloon valvuloplasty [17]. Not surprisingly, being a neonate and requiring a longer procedure, a likely surrogate for the complexity of the procedure, were predictive of the need for reintervention. It has become clear to us over time, and it is well demonstrated in the present study, that the addition of patch material to the repair increases the risk of reoperation or of valve failure, or both.

What came as a surprise was that 60% of our patients undergoing a bicuspid valve repair without patch material were free of any adverse events for 10 years. These were the patients who had undergone a debridement of their valves and were left with only native tissues. We have the conviction that a large proportion of these patients may remain free of any reoperation for even longer periods because the valves resulting from these procedures are very similar to the native ones.

The longest of the follow-up data presented describe outcomes after procedures that were less sophisticated than the concurrent ones, and we hope to observe further improvement in these late outcomes. Although this assumption remains to be proven, these initial results are encouraging. It has become clear that aortic valve disease in the pediatric population will attract several interventions over the course of a lifetime [18]. We believe that the relative benefits of balloon valvuloplasty and surgical repair should be weighed in terms of the total number of procedures over the course of a life time and the possibility of avoiding valve replacement for the longest time rather than in terms of freedom of reoperation after an initial intervention. We believe that surgical repair is more likely to generate a subgroup of patients who will have only one procedure for many decades than balloon valvuloplasty, because the latter is essentially a destructive process that will result in reinterventions in most patients, who will ultimately not be spared valve replacement.

To this day, it has, unfortunately, been impossible to identify preoperatively the patients whose repair will be limited to an extensive debridement of the valve who will have a superior outcome. Until preoperative imaging is able to accurately predict the type of repair necessary, it may therefore be preferable to offer primarily an operation to patients with bicuspid valve disease because it gives them a better chance to be in this group who may hope for a long-lasting event-free outcome.

Some may consider the Ross procedure to be a viable option for the most complex of these patients. We have tended to try to postpone the Ross procedure to the adult age, because it has now become clear that a quarter of them will require the replacement of their autografts within 2 decades [19, 20]. We hope that in adulthood, procedures aiming at reinforcing the support of the autograft root may be more easily performed [21, 22].

Fig 5. Freedom from any significant event.

Fig 6. Freedom from any significant event (no patch used vs use of patch).
The acquisition of the past experience and the present analysis enabled us to draw some technical recommendations. Extensive resection of all nodular fibrosis and thinning of the leaflets is the rule. That may include the resection of the ridge of a raphe and coring out of all fibrosis in the interleaflet triangle lying under a fused commissure. We now try to limit the use of pericardial patches in all possible circumstances, except in neonates to re-create a neocommissure. Even though our results show a slight trend toward better long-term outcomes with limited use of pericardial patches, longer follow-up will be necessary to ascertain the benefits of this strategy.

We have always used glutaraldehyde-preserved autologous pericardium because this material was demonstrated to be superior to untreated pericardium [8]. Devitalization by the glutaraldehyde prevents growth of the patch, which becomes stenotic as the child grows. Even in larger children, patches degenerate by fibrotic thickening and, ultimately, calcification, thus limiting their longevity to a maximum of 15 years [7, 8].

Whenever a complete involution of the commissure has occurred and has been replaced by a dome like-structure, it seems to us essential to support the area that is severed to avoid valve regurgitation. Tricuspidization of the bicuspid valve and 3 leaflets extension with pericardial patches was our favored approach for many years, but we have progressively moved away from this technique and now restrict the additions of patches and cusp extension techniques to patients where we doubt that any other techniques will achieve a durable repair.

Most bicuspid valves in children tend to have at least some degree of commissural fusion. The amount of fusion varies from an aspect close to a monocuspid valve, which will necessitate extensive reconstruction, to a more discrete fusion where limited debridement may restore a normally functioning bicuspid valve. The commissural area not only needs to be opened but also, we believe, thinning of the area is necessary. Very often, commissural fusion is accompanied by the loss of its support extending toward the sinotubular junction. Therefore, when this commissure is opened, again a support of this opened area has to be created using a pericardial patch to avoid this flailed portion of the valve to cause regurgitation.

The restoration of the mobility of these valves necessitates the resection of any thickened raphe, and we found that its resection, followed by its direct closure, does not always leave enough tissue to maintain valve competence. In recent times, we have sometimes replaced the raphe by a triangular patch of glutaraldehyde-treated autologous pericardium. The base of the interleaflet triangle lying under commissural fusion is not enlarged and does not need restriction once the fibrosis is resected at this level. The other commissure may require a subcommissural annuloplasty to improve the length of the coaptation of these valves, but our experience has been that, similarly to adults, these annuloplasties tend to fail, and it is possible that more rigid type of partial annuloplasties may one day benefit these patients.

Most patients with bicuspid valves have some degree of dilatation of the aortic root or the ascending aorta, or both. The enlargement of the root and ascending aorta has been demonstrated to be modest as long as the patients do not have severe aortic valve regurgitation [23]. Because the risk of aortic dissection and rupture seem to be low in the pediatric age group and because we believe that these patients would require an operation at a later stage, we have tended to avoid replacement of the aortic root and the ascending aorta in these patients [24–26].

The benefits of preventing fatal complications have to be balanced against the inconvenience of losing the elastic properties of the aorta at such a young age, which some believe could contribute to the development of hypertension. In some instances, it seems that the dilatation of the sinotubular junction may contribute to the failure of some of our repairs, and we wondered if we should not, in rare instances, apply techniques of remodelling of the ascending aorta that have met some success in adults [25].

In conclusion, outcomes after surgical repair of bicuspid aortic valves in the pediatric population are excellent, especially if the repair can be performed without the addition of patches. Children and adolescents with diseased bicuspid aortic valves should be offered primary repair because they can have long-lasting results if their disease can be relieved by simple procedures.

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My questions are the following: When do you resort to perform tricuspidization of bicuspid valves? What is the youngest age that you would be able to perform the leaflet extension technique without worrying about developing supraaortic obstruction in the small-sized aorta? And finally, what grades of residual lesions do you accept at the end of repair? Obviously, overcorrecting regurgitation with leaflet extension could result in stenosis, and there is a fine balance line between stenosis and regurgitation, so it would be helpful if you can tell us more about your decision making when you are faced with residual lesions at the end of the repair.

DR d’UDEKEM: The indications for 3-cusp extension. Well, in the past, we had a blanket rule to try to repair all patients. Nowadays, we still try to do a primary valve repair and that would be a 3-cusp extension technique if it is the only thing we can do in the early age, because we believe that it gives a benefit for the patients to wait 5 to 10 years to get them to a larger aortic root. Then, we give them to our colleague, Dr Skillington, who will do a perfect Ross at the time. I think we can do a better Ross if they reach adulthood with their native aortic root.

But if at a later age, 13 or 14 years old, we find that we have to do a cusp extension technique, we know that it is going to be failing, likely in early adulthood, at an age when it is not the right time for them to undergo another operation. So we tend more and more to do more Ross at adolescence or at the end of adolescence.
The earlier age for the 3-cusp extension technique. I have to say that I have been burned by doing a 3-cusp extension in a 3-month-old, and I had to reoperate 4 months later. So I believe that you should try to avoid the 3-cusp extension technique at less than 1 year.

And what was the last of your questions?

DR ALSOUFI: Correcting residual lesions.

DR d’UDEKEM: My natural answer would be we do not accept residual lesions. The immediate result is usually good. We easily accept mild regurgitation if it is central. We tend to avoid an eccentric jet. The only exception is when you avoid a 3-cusp extensions technique. You know that when you do a 3-cusp extension you may have a central jet, but no more than that because the material facing each other is the same. But if you do a little patch in front of a native tissue, especially if you have to thin it out, even though you may have enough coaptation, you may have some leak in the commissure, and then that gives you an image of eccentric lesions. That is something that you would accept; they are not important regurgitations. We would not accept moderate regurgitation and moderate stenosis.

DR CHRISTOPHER CALDARONE (Toronto, Ontario, Canada): My compliments on your technique and your very nice presentation. That was very well presented. Thank you.

Your data go a long way to describe your technique and philosophy in Melbourne, but it may be a bit of a stretch to say that you have demonstrated some superiority over balloon valvuloplasty. Of course, you are in a safe room to make that assertion, but I will play the devil’s advocate here. To go intervention-free 60% at 10 years, is that all that different than balloon aortic valvuloplasty in the population you described, typically having a first intervention at about 1 year of age or less?

DR d’UDEKEM: I am happy to make this statement in a room full of cardiologists, and hopefully that time will come. We will not have today, I believe, randomized trials. It is going to be very difficult to have randomized trials comparing balloon valvuloplasty with aortic valve repair. So we will have to progress with this type of experience.

What happened is that if you look at the experience of balloon valvuloplasty, and the best results are coming from Boston, by 15 years, the freedom from reintervention is 27%. And it is very likely when you look at the curves that maybe you are going to have 10%, 20% who will not need reintervention, but likely because it is a destructive process and it is going to evolve. The thing here is that when you are doing a reintervention in these patients, it is going to be almost always a valve replacement. So my point is that when you do a balloon valvuloplasty, you will always end up with a valve replacement.

I believe that today we should stop comparing balloon valvuloplasty and aortic valve repair just in terms of freedom from first intervention. Aortic valve disease, and especially bicuspid aortic valve disease, in children is likely a lifelong disease that will necessitate several interventions. I think you should favor the procedure that allows the patients to be without a mechanical valve for the longest possible time. And I believe that if you manage to have a native valve, as shown in the movie, to have a repair with only native valve tissues, then this valve may last for 15, 20 and maybe more years, and I think that is impossible to get with a balloon valvuloplasty, because it is a destructive process.

DR CALDARONE: I agree with your philosophy, and I agree with the point that this is a lifelong disease, and the only way we are really going to sort out the supremacy of one intervention over another is to look at the lifelong sequence of procedures required to manage these patients. I also agree, the only variable we want to minimize is the amount of time someone has to spend during his or her life with a mechanical valve. I would say the jury is still out. Nice presentation. Thank you.

DR COSMIN DOBRESCU (Los Angeles, CA): Thank you very much for an interesting presentation. I have two questions: First, a technique question; then, an analysis question. The technical question is: With the cases you presented in which you augmented the valve with autologous tissue, I understood that pericardium was used. Was that treated or untreated pericardium? And, if it was treated, then can you describe the technique?

The second question pertains to the series you presented where primary aortic leaflet repair was required. You demonstrated that there were two sort of distinct modes of failure, one of stenosis and one of regurgitation. Did you notice that there was some pattern of the mode of failure depending on which technique that was used? And was one technique more related to a stenotic failure mode and perhaps some other related, for example, to an insufficiency failure mode? Thank you very much.

DR d’UDEKEM: All the patches were treated with glutaraldehyde, and what we changed over the time is that we tend to expose the tissue to less time in the bath; from 6 minutes we went down to 2 minutes of treatment.

And then for the analysis question, we have analyzed specifically the mode of failure—stenosis, regurgitation—and the only predictive factor that came up was, again, longer bypass time, and earlier age at surgery. That was all. We couldn’t find out any techniques that would promote more one or the other mode of failure.