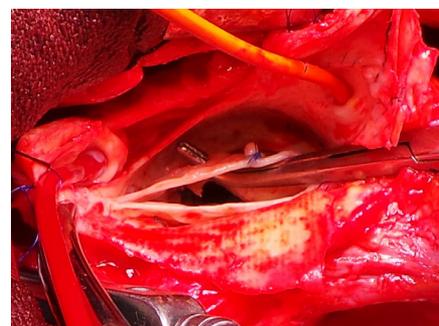


Almost All Incompetent BAV Should Be Repaired



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The short answer to this topic is “No, most incompetent bicuspid aortic valve should not be repaired.” Aortic valve repair certainly plays an important role in the management of incompetent bicuspid aortic valve of young patients, and the selection criteria are like those for incompetent tricuspid aortic valve. The tissue that makes the cusps should be normal or have minor abnormalities easily corrected by surgery such as plication of the free margin. In addition, the anatomic orientation of the conjoint and the normal cusps should be approximately at 180°. If the root is dilated, reimplantation of the aortic valve is likely the most reproducible approach, and if the root is not dilated but the annulus is, a subannular annuloplasty and correction of the cusp prolapse is adequate. Most patients with incompetent bicuspid aortic valve have malformed, fibrotic and calcified cusps and replacement with either pulmonary autograft or a mechanical aortic valve in young patients and a bioprosthetic aortic valve are likely to provide better long-term outcomes than aortic valve repair.



A perforated pericardial patch used to augment the conjoint cusp of an incompetent bicuspid aortic valve.

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Central Message

Most incompetent bicuspid aortic valves should be replaced; repair is reserved for young patients with cusps with normal tissue and satisfactory valve morphology.

INTRODUCTION

Aortic valve (AV) repair is among those heart valve operations performed before the advent of cardiopulmonary bypass.¹ Open AV repair was also performed early on after open-heart surgery became feasible but largely abandoned in favor of AV replacement. Congenital heart surgeons continued to perform AV repair in children with ventricular septal defect and aortic insufficiency (AI).^{2,3} George Trusler from the Hospital for Sick Children of Toronto developed a technique to correct cusp prolapse by plication of the free margin next to one of its commissures in patients with AI and subaortic ventricular septal defect which became known as “Trusler stitch.”³

The introduction of intraoperative transesophageal echocardiography transformed heart valve surgery because it has allowed surgeons to determine valve function in the operating room. It also opened the door to the development of newer operative approaches to correct valve dysfunction. Cosgrove et al⁴ from

the Cleveland Clinic were the first ones to publish on a relatively large series of patients with AI due to bicuspid AV (BAV) disease who had AV repair by triangular resection of the central portion of the prolapsing cusp (and less commonly by plication) and subcommissural plication. The triangular resection of cusps had been described by Alain Carpentier in his famous “Cardiac valve surgery—the ‘French correction’.”⁵ The subcommissural plication had been described by Cabrol et al from Paris.⁶ The Cleveland Clinic surgeons presented the results on 94 patients operated exclusively by Delos Cosgrove at the 12th Annual Meeting of the European Association for Cardiothoracic Surgery in 1998.⁷ After a median follow-up of 5.5 years, 13% of patients had had reoperation, mostly because of dehiscence of the suture line in the repaired cusp.⁷ Interestingly, discussants of that paper mentioned the importance of addressing the dilated aortic annulus, frequently present in patients with BAV and AI.⁷

Several other centers have also reported their experience with AV repair in patients with AI due to BAV with failure rates of 20–30% at 5 years and 40–50% at 10 years.^{8,9} In spite of these suboptimal results, some surgeons believe that it is a good alternative to bioprosthetic AV replacement in young patients.^{8,9}

Pure AI is uncommon in patients with BAV.^{10,11} Aortic stenosis and mixed lesions are more common.^{10,11} AI in patients with BAV is often associated with dilatation of the aortic annulus. This is probably related to the genotype of BAV whereby

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the connective tissue of the aortoventricular junction becomes abnormal by the time surgery is indicated.¹² Gebrine El Khoury from Brussels has greatly contributed to our knowledge of AV repair in both bicuspid and tricuspid AVs.^{13–15} El Khoury's group was one of the first to identify the importance to addressing the dilated aortic annulus at the time of AV repair in BAV.¹⁵ They compared 2 groups of patients with BAV who had cusp prolapse with or without subcommissural plication or reimplantation of the AV and found significantly better results in the latter.¹⁵ Other investigators also found excellent mid-term results with reimplantation of BAV for AI with dilated aortic root.¹⁶

Dilatation of the aortic annulus is not always associated with dilatation of the aortic sinuses or ascending aorta to justify an extensive operation such as reimplantation of the AV. An aortic annuloplasty along the fibrous components of the aortoventricular junction or complete ring on the outside of the left ventricular outflow tract is likely adequate to correct annular dilatation.¹⁷ Thus, an aortic annuloplasty and repair of the prolapsing conjoint cusp is often all that would be required to re-establish AV competence. The long-term durability of such procedure remains unknown.

Hans Joachim Schäefers from Homburg/Saar probably has performed more AV repair in patients with BAV than any other surgeon in the world.¹⁸ Schäefers et al have published extensively on AV repair in BAV and have identified factors associated with failure.^{18–20} A review of their publications indicated that they started repairing incompetent BAV in Homburg/Saar in 1995 and the techniques evolved over the years and more recently converged to the same conclusions as El-Khoury's group, that is, stabilization of the aortic annulus is an important component of AV in BAV.²⁰ I believe that the most important lessons from Schäefers' experience with repair of incompetent BAV are the significance of coaptation height and coaptation length and the geometric orientation of the cusps.¹⁹ The best results in BAV repair are obtained in patients with Sievers type 0 and cusps oriented at 180° followed by Sievers type I with cusps oriented at various degrees but close to 180°.

SURGICAL MANAGEMENT OF PATIENTS WITH INCOMPETENT BAV

Most patients with BAV do very well until adult life and may never need AV surgery.^{10,11} Surgery may become necessary because of AV dysfunction or dilatation of the ascending aorta and arch. BAV-related aortopathy was the subject of a recently published consensus.²¹ The issue in this review is when should AV repair be performed in the setting of incompetent BAV.

Criteria to repair or replace an incompetent BAV are like those for incompetent tricuspid AV. I believe that the most important factor is the quality of the tissue that makes the aortic cusps. If the cusps appear normal or have only mild abnormalities such as myxomatous changes, cusp prolapse or small fenestrations, AV repair should be considered. Other factors such as the orientation of the cusps, their heights, patient's age, and the associated aortopathy should also be taken into

account. BAV with 2 thin and pliable cusps at approximately 180° with or without a raphe in 1 of the cusps are ideal for repair. Approximately one-third of all BAV repaired by us had this phenotype and were associated with dilated aortic root and had reimplantation of the AV.²² No patient in that subgroup has developed more than mild AI after a mean follow that exceeds 10 years and extends up to 25 years.²² Patients with Sievers' type 1 BAV and dilated aortic root are also candidates for repair as long as the cusps are thin and pliable and the orientation of the conjoint cusp and normal cusp is such that with reimplantation of the AV, one can force it to become close to 180°. That is, the sutures for the reimplantation if the subannular area are distributed in the Dacron graft in such way as to reduce the intercommissural distance of the conjoint cusp. There is a physical limitation on this approach because the most common type of BAV results from fusion of the right and left cusps which are largely attached to the muscular interventricular septum and cannot be reduced in length. I believe that BAV with cusps oriented down to 160° can be converted to close to 180° during reimplantation of the AV.

Not all incompetent BAV have dilated aortic root to warrant reimplantation of the AV. In this circumstance, I believe that an aortic annuloplasty with a band or ring can reduce the diameter of the aortic annulus and simple cusp repair can restore AV function.

Younger the patient the harder I have tried to repair incompetent BAV. I have used all sorts of material to augment the aortic cusp (fresh and glutaraldehyde fixed autologous pericardium, bovine and equine glutaraldehyde fixed pericardium and Cormatrix membrane [CorMatrix Cardiovascular Inc., Sunnyvale, CA]) or to replace a malformed, fibrotic or calcified segment of the cups, but my experience has been uniformly disappointing. Small patches of any material work well, particularly if they are not in commissural areas or areas where the patch is bent repetitively during the cardiac cycle. Large patches usually fail and most them do so within the first decade. The central picture of this essay illustrates a case of a young man who had the conjoint cusp augmented with a glutaraldehyde fixed autologous pericardium and developed severe AI only 3 years later due to degeneration of the patch.

Early on in our experience with AV replacement with pulmonary autograft, we identified incompetent BAV as the main cause of failure in young patients.²³ Once we learned that we began to repair incompetent BAV in young patients even if the conjoint cusp was grossly abnormal and required reconstruction with a patch material but a decade later we are back at the crossroad because these repaired valves are leaking again. Prompted by a recent study from Australia on the Ross procedure in patients with incompetent BAV that showed a freedom of moderate or severe AI of 85% at 15–20 years,²⁴ we are now reconsidering this operation in young adults with less than ideal cusps anatomy for AV repair. Obviously, this is not an issue if the patient is willing to accept a mechanical AV, something that I have always advised my young patients to have if a satisfactory AV repair or Ross procedure cannot be performed.

Finally, it is hard to justify AV repair in patients older than 60 years of age with incompetent BAV because the durability of bioprosthetic AVs, particularly the Hancock II bioprosthesis is excellent and the freedom from structural valve failure is greater than 80% at 20 years of follow-up.²⁵

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