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BICUSPID AORTIC VALVE AND ASSOCIATED AORTOPATHY: SURGICAL CONSIDERATIONS

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Introduction

The human aortic valve is normally a tricuspid structure but may occur as bicuspid or quadricuspid structures (Figure 1). Bicuspid aortic valve (BAV) is one of the most common congenital heart anomalies and the most common congenital anomaly in adults, occurring in 0.5-2% of the population based on echocardiography and autopsy series.[1, 2] It is an inherited defect that appears to occur in an autosomal dominant pattern. BAV is the cause of aortic stenosis in 70-85% of pediatric cases[3] and about 50% of adult cases.[4] Indications for aortic valve replacement (AVR) in BAVs is the same as rheumatic or senile calcific degeneration aortic valves. BAVs are different, however, in that the tissue pathology is not limited to the valves' leaflets but extends from the left ventricular outflow tract to the ascending thoracic aorta. BAVs are associated with several congenital anomalies, infective endocarditis, and acute thoracic aortic emergencies such as aneurysm and dissection. These associations lead to surgical considerations different from other aortic valve pathologies.

Associated Congenital Conditions

BAV is usually an isolated anomaly but may be associated with additional congenital abnormalities in 20-40% of patients. Coarctations of the aorta, ventricular septal defect, and patent ductus arteriosus have known associations with BAV. Those BAVs associated with coarctation of the aorta have an increased risk of ascending thoracic aortic aneurysm and aortic dissection.[5] Ventricular septal defect (VSD) is the most common congenital defect in children. When found in adults, ventricular septal defects are associated with BAV in up the 30% of cases.[6] Determination of BAV prior to VSD repair or VSD prior to BAV replacement is of obvious importance to the surgeon. BAV can also be associated with more severe genetic congenital heart problems such as Turner syndrome,[7] Williams syndrome,[8] and Shone's syndrome.[9] These are generally concerns for the pediatric cardiac surgeon and are not often encountered in an adult practice.

Valvular Complications with BAV

The majority of patients with BAV are asymptomatic and unaware of their abnormality until adulthood, when symptoms, physical findings, or cardiac echocardiography for other reasons lead to the diagnosis. Although undetected for prolonged periods, the majority of patients with BAV will require surgery for their valve abnormality at some point in their life.[10]

Echocardiographic series have demonstrated that valve sclerosis begins in the patient's second decade and that calcification increases after the fourth decade.[11] The natural history is similar to but occurs earlier and progresses faster than senile calcific degeneration of the aortic valve. Valvular complications of BAV include aortic stenosis, aortic regurgitation, and infective endocarditis. The vast majority of cases (at least 75%) presenting to the cardiac surgeon for operative correction have aortic stenosis,[12] with a lesser number presenting with regurgitation or mixed lesions. The indications for surgical intervention are the same as

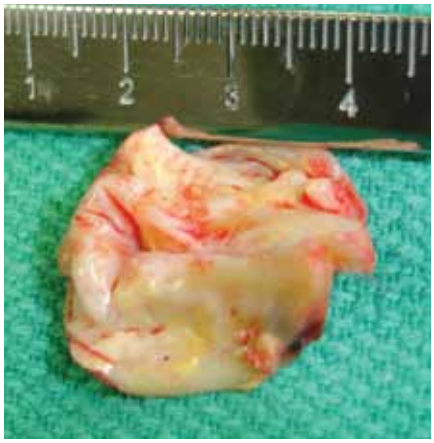

Figure 1A. Bicuspid aortic valve with calcific stenosis.

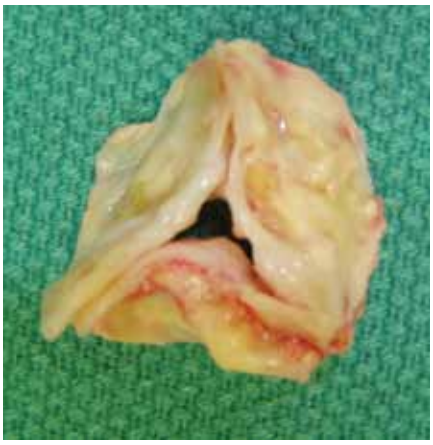

Figure 1B. Tricuspid aortic valve with senile calcific stenosis.

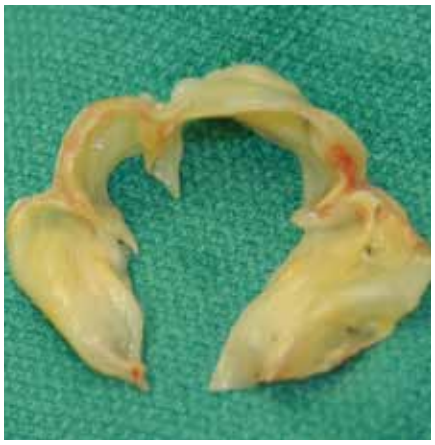

Figure 1C. Quadricuspid aortic valve with severe regurgitation.

other aortic valve pathologies and can be found in the ACC/AHA guidelines.[27] Case series have led to estimates of infective endocarditis in BAV ranging from 10-30%,[13] but the actual population risk is likely closer to 3%.[10] Infective endocarditis is the cause for most cases of severe aortic regurgitation in BAV due to perforation of the leaflets.[13] Indications for surgical intervention are no different than other valve pathologies with endocarditis.

Aortic Complications with BAV

BAV is associated with an aortopathy leading to wall weakness. Whether this affects all patients with BAV or only a subset is not known, but all thoracic aortas are considered at risk. The challenge to the cardiac surgeon is deciding what to do with a slightly to moderately enlarged ascending aorta in the patient with BAV. Cystic medial necrosis, elastic fragmentation, and changes in smooth muscle orientation are significantly more common in aortas with BAV than with tricuspid aortic valves.[14] These changes are similar to, but not as severe as, those seen in Marfan syndrome and lead to increased dilatation and aneurysm formation in the ascending thoracic aorta as well as an increased risk of aortic dissection that is five to nine times greater than in the normal population.[15] Aortic dilatation can be seen by echocardiographic exam in up to 70% of patients with BAV, even in the absence of hemodynamic valve disturbance.[16] Repair of ascending thoracic aortic aneurysm in patients without BAV is commonly recommended when the ascending aorta is 5.5 cm in diameter, since there appears to be an upward inflection point for rupture at 6 cm. In patients with BAV, ascending aortas greater than 4.5 to 4.9 cm in diameter appear to have an increased risk of rupture, leading to the recommendation that they be repaired during AVR if larger than 4.5 cm.[17] This recommendation is made to decrease the risk of both rupture and dissection. The persistence of aneurysm and dissection risk after successful aortic valve replacement underscores the belief that aneurysm formation and dissection are due to intrinsic aortic wall abnormality and not hemodynamic flow perturbations across abnormal valves.[18]

Surgical Procedures with BAV

Repair

Stenotic aortic valves are replaced and regurgitant aortic valves may be replaced or repaired. Early reports of repair for BAV stated good results,[19] but recent follow up shows only an 87% freedom from reoperation at five years.[20] A 10-year follow-up paper shows a competent valve in only 57% of patients.[21] Most cases of BAV repair involve reconstruction of an enlarged annulus or sinotubular junction only; When leaflet reconstruction is needed, the results are more disappointing. We would recommend aortic valve repair in BAV only in carefully selected patients.

Replacement

The indications for AVR and valve selection criteria when using a mechanical or stent mounted tissue valve are the same in BAV as they are in other causes of aortic stenosis, as the annular tissue has good tensile strength. Aortic valve replacement is complicated if the surgeon wishes to use a stentless valve such as a stentless xenograft valve, allograft, or pulmonary autograft due to the risk of aortic dilation and subsequent future distortion of the valve and aortic regurgitation. For a stentless xenograft or allograft, we would recommend annular and sinotubular stabilization with fabric collars

to prevent future dilatation. The pulmonary autograft or Ross procedure presents a special consideration since the arterial wall abnormality present in BAV can also be present in the pulmonary artery being used as the new aortic valve, leading to a high failure rate.[22, 23] We believe that the Ross procedure should be used with caution and should include consideration of graft stabilization with fabric collars when used in patients with BAV.

The ascending aorta

The ascending aorta is repaired in an asymptomatic patient to prevent rupture or dissection. The patient without BAV shows a predilection for rupture or dissection above a diameter of 6 cm.[24] For the patient with BAV, the risk of rupture or dissection increases between diameters of 4.5 to 4.9 cm.[25] This has led to the ACC/AHA Class I guidelines for ascending aortic replacement in patients with BAV at a diameter greater than 5 cm if AVR is not needed and greater than 4.5 cm if AVR is necessary. When appropriate, surgical strategies for the ascending aorta include reduction aortoplasty, replacement from the sinotubular junction to the distal ascending aorta, and full root replacement with coronary reimplantation. Although not technically difficult, reduction aortoplasty has a high rate of future failure due to redilatation, and we do not use this approach.[26] Replacement beyond the sinotubular junction is adequate for most patients and adds little risk to the operative procedure. Full root replacement is necessary when there is evidence of aortic root involvement such as extreme annular dilatation or cephalad displacement of the coronary ostia. Full root replacement can, however, double the risk of the procedure in some series.[25]

Conclusion

Bicuspid aortic valve disease is a common entity that usually necessitates surgery at some point in the patient's life. Surgical decision making is complicated by the coexisting aortopathy of the ascending aorta that may require surgical replacement. Even when replacement of the ascending aorta is unnecessary, it remains at risk after successful AVR and requires life-long surveillance.

References

1. Roberts WC. The congenitally bicuspid aortic valve. A study of 85 autopsy cases. Am J Cardiol. 1970 Jul;26(1): 72-83.
2. Steinberger J, Moller JH, Berry JM, Sinaiko AR. Echocardiographic diagnosis of heart disease in apparently healthy adolescents. Pediatrics. 2000 Apr;105(4 Pt 1):815-8.
3. Mack G, Silberbach M, Aortic and pulmonary stenosis. Pediatr Rev. 2000 Mar;21(3):79-85.
4. Ward C. Clinical significance of the bicuspid aortic valve. Heart. 2000 Jan;83(1):81-5.
5. Oliver JM, Gallego P, Gonzalez A, Aroca A, Bret M, Mesa JM. Risk factors for aortic complications in adults with coarctation of the aorta. J Am Coll Cardiol. 2004 Oct 19;44(8):1641-7.
6. Neumayer U, Stone S, Somerville J. Small ventricular septal defects in the adult. Eur Heart J. 1998 Oct;9(10):1573-82.
7. Roos-Hesselink JW, Schölzel BE, Heijdra RJ, Spitaels SE, Meijboom FJ, Boersma E, Bogers AJ, Simoons ML. Aortic valve and aortic arch pathology after coarctation repair. Heart. 2003 Sep;89(9):1074-7.
8. Sugayama SM, Moisés RL, Wagënfur J, Ikari NM, Abe KT, Leone C, da Silva CA, Lopes Ferrari Chauffaille Mde L, Kim CA. Williams-Beuren syndrome: cardiovascular abnormalities in 20 patients diagnosed with fluorescent in situ hybridization. Arq Bras Cardiol. 2003 Nov;81(5):462-73.
9. Bolling SF, Iannettoni, MD. Dick M 2nd, Rosenthal A, Bove EL. Shone's anomaly: operative results and late outcome. Ann Thorac Surg. 1990 Jun;49(6):887-93.
10. Lewin MB, Otto CM. The bicuspid aortic valve: adverse outcomes from infancy to old age. Circulation. 2005 Feb;111(7):832-4.
11. Mills P, Leech G, Davies M, Leathan A. The natural history of a non-stenotic bicuspid aortic valve. Br Heart J. 1978 Sep;40(9):951-7.
12. Braverman AC, Güven H, Beardslee, MA, Makan M, Kates AM, Moon MR. The bicuspid aortic valve. Curr Probl Cardiol. 2005 Sept;30(9):470-522.
13. Ward C. Clinical significance of the bicuspid aortic valve. Heart. 2000 Jan;83(1):81-5.
14. de Sa M, Moshkovitz Y, Butany J, David TE. Histologic abnormalities of the ascending aorta and pulmonary trunk in patients with bicuspid aortic valve disease: clinical relevance to the Ross Procedure. J Thorac Cardiovasc Surg. 1999 Oct;118(4):588-94.
15. Nistri S, Sorbo M, Marin M, Palisi M, Scognamiglio R, Thiene G. Aortic root dilatation in young men with normally functioning bicuspid aortic valves. Heart. 1999 Jul;82(1):19-22.

16. Cecconi M, Manfrin M, Moraca A, Zanoli R, Colonna PL, Bettuzzi MG, Moretti S, Gabrielli D, Perna GP. Aortic dimensions in patients with bicuspid aortic valve without significant valve dysfunction. Am J Cardiol. 2005 Jan;95(2):292-4.
17. Ortiz JT, Shin DD, Rajamannan NM. Approach to the patient with bicuspid aortic valve and ascending aorta aneurysm. Curr Treat Options Cardiovasc Med. 2006 Dec;8(6):461-7.
18. Russo CF, Mazzetti S, Garatti A, Ribera E, Milazzo A, Bruschi G, Lanfranconi M, Colombo T, Vitali E. Aortic complications after bicuspid aortic valve replacement: long-term results. Ann Thorac Surg. 2002 Nov;74(5):S1773-6.
19. Cosgrove DM, Rosenkranz ER, Hendren WG, Bartlett JC, Stewart WJ. Valvuloplasty for aortic insufficiency. J Thorac Cardiovasc Surg. 1991 Oct;102(4):571-6; discussion 576-7.
20. Casselman FP, Gillinov AM, Akhrass R, Kasirajan V, Blackstone EH, Cosgrove DM. Intermediate-term durability of bicuspid aortic valve repair for prolapsing leaflet. Eur J Cardiothorac Surg. 1999 Mar;15(3):302-8.
21. Rao V, Van Arsdell GS, David TE, Azakie A, Williams WG. Aortic valve repair for the adult congenital heart disease: A 22-year experience. Circulation. 2000 Nov 7;102(19 Suppl 3):III40-3.
22. Kouchoukos NT, Masetti P, Nickerson NJ, Castner CF, Shannon WD, Dávila-Román VG. The Ross procedure: long-term clinical and echocardiographic follow-up. Ann Thorac Surg. 2004 Sep;78(3):773-781; discussion 781.
23. Luciana GB, Casali G, Favaro A, Prioli MA, Barozzi L, Santini F, Mazzucco A. Fate of the aortic root late after Ross operation. Circulation. 2003 Sep 9;108 Suppl 1:II61-7.
24. Elefteriades JA. Natural history of thoracic aortic aneurysms: indications for surgery, and surgical versus nonsurgical risks. Ann Thorac Surg. 2002 Nov;74(5):S187780; discussion S1892-8.
25. Borger MA, Preston M, Ivanov J, Fedak PW, Davierwala P, Armstrong S, David TE. Should the ascending aorta be replaced more frequently in patients with bicuspid aortic valve disease? J Thorac Cardiovasc Surg. 2004 Nov;128(5):677-83.
26. Bauer M, Pasic M, Schaffarzyk R, Siniawski H, Knollmann F, Meyer R, Hetzer R. Reduction aortoplasty for dilatation of the ascending aorta in patients with bicuspid aortic valve. Ann Thorac Surg. 2002 Mar;73(3):720-3; discussion 724.
27. American College of Cardiology; American Heart Association Task Force on Practice Guidelines (Writing Committee to revise the 1998 guidelines for the management of patients with valvular heart disease); Society of Cardiovascular Anesthesiologists, Bonow RO, Carabello BA, Chatterjee K, de Leon AC Jr, Faxon DP, Freed MD, Gaasch WH, Lytle BW, Nishimura RA, O'Gara PT, O'Rourke RA, Otto CM, Shah PM, Shanewise JS, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Fuster V, Halperin JL, Hiratzka LF, Hunt SA, Lytle BW, Nishimura R, Page RL, Riegel B. ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing Committee to Revise the 1998 guidelines for the management of patients with valvular heart disease) developed in collaboration with the Society of Cardiovascular Anesthesiologists endorsed by the Society for Cardiovascular Angiography and Interventions and the Society of Thoracic Surgeons. J Am Coll Cardiol. 2006 Aug 1;48(3):e1-148.