Available online at www.rajournals.in

RA JOURNAL OF APPLIED RESEARCH
ISSN: 2394-6709
DOI:10.47191/rajar/v7i10.11
Volume: 07 Issue: 10 October-2021

International Open Access

Impact Factor- 7.036

Page no.- 2588-2592

Aortic Bicuspidy: Clinical Profile and Surgery

A. Seghrouchni[1,2], N. Atmani[1,2], Y. Moutakiallah[1], Y. El Bekkali[1], M. Ait Houssa[1]
[1]Cardiovascular surgery department, Mohammed V military training hospital, Rabat-Morocco
[2]Faculty of Medicine and Pharmacy of Fes, University of Sidi Mohamed Ben Abdellah Fes.

ARTICLE INFO

Published Online:
25 October 2021

Corresponding Author:
A. Seghrouchni
Department of Cardiovascular Surgery, Hôpital Militaired' Instruction Mohamed V Rabat

ABSTRACT

Objective:
To study the anatomic-clinical profile of aortic bicuspidy and the outcome of surgery.

Patients and Methods:
During an 18-year period, 448 patients had aortic valve replacement. Of these, 24 (5.3%) had aortic bicuspidy (AB). The diagnosis of AB was made by echocardiography or during surgery. All patients underwent surgery under extracorporeal circulation.

Results:
The mean age was 45.2 ± 11.8 years, 14 patients (58.3%) had aortic stenosis and 10 cases (41.7%) had aortic insufficiency, 4 of whom had infective endocarditis. All patients had aortic valve replacement. The operative mortality rate was zero. The mean times of the cardiopulmonary bypass (CPB) and aortic clamping were 99.2 ± 35.4 min and 65.8 ± 24.9 min, respectively.

Conclusion:
Aortic bicuspidy progresses rapidly and becomes symptomatic in young adults. Despite excellent surgical results, early detection is desirable before complications occur.

KEYWORDS: Aortic Bicuspidy, Infective Endocarditis, Aortic Valve Replacement

INTRODUCTION

Aortic bicuspidy is the most common congenital cardiovascular anomaly, affecting 0.5% to 2% of the population [1]. In 10-30% of cases, bicuspidism runs in families and is transmitted in an autosomal dominant fashion. It is accompanied by premature valve dysfunction and abnormalities of the ascending aorta. It is the leading cause of aortic valve replacement in the United States [2]. The risk of aortic dissection is increased 9-fold, and when infective endocarditis occurs in this setting, it has a more guarded prognosis.

We report a series of 24 cases of aortic bicuspidy operated in the cardiac surgery department of the Mohammed V military training hospital.

PATIENTS AND METHODS

From January 2002 to December 2019 (period = 18 years), 448 patients had aortic valve replacement, of whom 24 had aortic bicuspidy (5.3%). All patients in whom aortic bicuspidy was diagnosed preoperatively or discovered intraoperatively were included.

Doppler echocardiography was performed in all patients. The parameters studied were: left ventricular dimensions, left ventricular systolic function, valves and their function, and the ascending aorta.

Transesophageal echocardiography was performed in patients with dilated ascending aorta and in those with infective endocarditis.

Similarly, CT angiography was performed as additional imaging in selected patients.

All patients were operated under general anesthesia and extracorporeal circulation.

Anticoagulation was done with heparin at a dose of 300 ui / kg to have a TCA > 400 s. Bypass surgery was performed with a membrane oxygenator with moderate hypothermia (temperature) (32° - 34°). Heparin was neutralized by protamine dose by dose. Myocardial protection was provided by anterograde cold blood cardioplegia.

Antibiotic prophylaxis used a broad-spectrum antibiotic and reduction of intraoperative bleeding required the use of antifibrinolytics.

Clinical information, echocardiographic data, and operative findings were collected from the patients' medical records. In-hospital mortality was defined as any death occurring within 30 days after surgery. Difficult post-operative

outcomes were defined as any complication responsible for a long stay in the intensive care unit.

RESULTS

24 patients were included in this study, including 4 women (16.7%). The mean age of the patients was 45.2±14.8 years. The clinical characteristics of the patients are summarized in Table 1. Fourteen patients (58.3%) had NYHA class III-IV dyspnea and 3 had stress angina. Half of the patients were smokers; few preoperative comorbidities were noted (01 case of renal insufficiency).

Echocardiographic data are summarized in Table 2: left ventricular diameters were 62.6±12.8 mm for the telediastolic diameter and 45.4±12.4 mm for the telesystolic diameter, respectively. Mean EF was 51 ± 16.3 %, mean systolic pulmonary artery pressure was 55.8 ± 18 mmHg.

14 patients (58.3%) had dominant aortic stenosis and 10 cases (41.7%) had isolated significant aortic insufficiency. Infective endocarditis was found in 6 patients. The surgical indication was urgent in 1 patient (4.2%). 23 patients underwent aortic valve replacement by mechanical prosthesis and one patient had a Bentall procedure. The operative data are summarized in Table 3: the mean times for CPB and aortic clamping were 99.2 ± 35.4 minutes and 65.8 ± 24.9 minutes, respectively. The median mechanical ventilation time was 15 hours (7-20 hours) and the median ICU stay was 72 hours (47-120 hours).

Three patients were ventilated ≥ 48h. Two patients were reoperated for bleeding and no patient died.

During follow-up, one patient died of unknown cause, six patients were lost to follow-up, and the medium-term survival rate (time=36 months) was 70%.

DISCUSSION

Our study showed that the prevalence of aortic bicuspidy in patients who underwent aortic valve replacement was 5.3% which is slightly higher than the prevalence in the general population. The majority of cases were reported sporadically, however, familial forms represent a fairly high prevalence 9% suggesting an autosomal dominant origin [3]. The prevalence is 7-15% according to autopsy studies of deaths after aortic dissection or rupture [4, 5].

Aortic bicuspidy is more frequent in boys than in girls with a sex ratio of 3:1 [6]. Tutar [7] reported a prevalence of 0.71 % in boys and 0.19 % in girls. Evalidas Girdauskas [8] in a series of 530 cases of aortic bicuspidy, 77% of the patients were male.

The bicuspid aortic valve may remain unrecognized for many years; however, the majority of patients will develop complications over time. The risk of acute aortic dissection is increased 9 to 10 times in cases of bicuspidy [9, 10]. When infective endocarditis occurs it is more severe.

Tribouillay [11] studied a population of 310 consecutive patients with aortic bicuspidy complicated by infective endocarditis and found that there is an increase in aortic perivalvular abscesses [+50%] and endocarditis is more likely to be dislocating. Six patients in our series were complicated by infective endocarditis and 3 had aortic ring abscess.

Thus, it may progress to aortic insufficiency and/or aortic stenosis as in patients with a tricuspid valve. Various autopsy studies have shown that pure aortic stenosis is often the predominant lesion (75% of cases); pure aortic insufficiency (13%) and aortic disease (10%) of cases [12].

Hammond [13] found that aortic bicuspidy was responsible for aortic stenosis in 2/3 of patients. Aortic stenosis has a rapid progression (27 mmHg / 10 years) and patients with aortic bicuspidy require surgery 5 years earlier than those with a tricuspid aortic valve [14, 15]. In addition, there are a number of factors known to be associated with rapid progression of the trans-aortic gradient: smoking, hypertension and hyperlipidemia.

The incidence of aortic insufficiency in aortic bicuspidy varies from 1.5 to 3%.It is isolated or associated with ascending aortic aneurysm, aortic coarctation and infective endocarditis [16]. One patient in our series had aortic bicuspidy associated with coarctation of the aorta, aneurysm of the ascending aorta and complicated by infective endocarditis.

The exact mechanism of aortic bicuspidy is unknown. Two theories attempt to explain this condition: the hemodynamic theory states that the decrease in flow through the aortic orifice during embryonic life is not sufficient to allow separation and differentiation of the aortic cusps, but this theory is not well supported. Conversely, a genetic etiology is increasingly documented in the literature. [17, 18 ,19, 20, 21].

Aortic bicuspidy is increasingly described as a pathology of the entire ascending aorta including the aortic annulus, valve, sinus of Valsalva coronary ostia, sino-tubular junction and supra-coronary segment [22, 23].

Dilation of the ascending thoracic aorta is observed in approximately 50 % of patients with aortic bicuspidy, including patients with normal aortic valve function. The annual progression of ascending aortic dilation is approximately 0.5 to 0.9 mm [24, 25].

Dilation is much more frequent in aortic insufficiency than in aortic stenosis [26, 27].

The etiopathogeny of ascending aortic aneurysm in aortic bicuspidy remains a current debate but around 2 theories: one in favor of a hemodynamic cause in which the aneurysm would be a consequence of abnormalities of aortic flow through an abnormal valve [28, 29]. Hemodynamic turbulence in case of valve stenosis and volumetric overload in case of aortic insufficiency have been raised as causes of dilation [30, 31], the other hypothesis supports a genetic cause [32, 33]. Abbatt [34] was the first to raise an association between bicuspidy and aneurysm of the ascending aorta. Recently this association is increasingly documented.

Transthoracic (TTE) and transesophageal (TEE) echocardiography represent the essential examination for the diagnosis of aortic bicuspidy and the detection of dilation of the ascending aorta. Angioscanner provides mainly morphological arguments for the diagnosis of bicuspidy, but it is mainly MRI angiography that seems to be more efficient.

The prevention of infective endocarditis, which used to be mandatory and then optional in this condition according to the French recommendations of 2002, is no longer part of the North American recommendations of the ACC/AHA, because aortic bicuspidy is no longer listed as a condition at high risk of infective endocarditis [35]. In our series 6 patients (25%) had been revealed by infective endocarditis, this should solicit the clinician's common sense regarding prophylaxis against infective endocarditis far from the recommendations.

The surgical indications for aortic stenosis or insufficiency are identical for patients with a bicuspid or tricuspid valve.

CONCLUSION

Aortic bicuspidy is a congenital anomaly of the entire aortic root that requires screening and therapeutic management before serious complications arise.

REFERENCES

1. ROBERTS WC. The congenitally bicuspid aortic valve. A study of 85 autopsy cases. Am J Cardiol, 1970; 26: 72-83.
2. Roudaut R. Bicuspidie aortique et dilatation de l'aorte ascendante chez l'adulte jeune : conduite à tenir. Rendez-vous de la Cardiologie, Hôpital de la Pitié-Salpêtrière. Paris, 11 mars 2011.
3. Huntington K, Hunter MD, Alasdair GW, Chan MD. A prospective study to assess the frequency of familial clustering of congenital bicuspid aortic valve. J Am CollCardiol 1997;30:1809–12.
4. Roberts CS,Roberts WC. Dissection of the aorta associated with congenital malformation of the aortic valve. J Am CollCardiol 1991;17:712-6.
5. Spittell PC, Spittell JA, Joyce JW, Tajik AJ, Edwards WD, Schaff HV, et al. Clinical features and differential diagnosis of aortic dissection: experience with 236 cases (1980 through 1990). Mayo ClinProc 1993;68:642-51.
6. Fedak PWM, Verma S, David TE, Leask RL, Weisel RD, Butany J. Clinical and pathophysiological implicationsof a bicuspid aortic valve. Circulation 2002;106:900–4.
7. TUTAR E, EKICI F, ATALAY S et al. The prevalence of bicuspid aortic valve in newborns by echocardiographic screening. Am Heart Lung Circ, 2006; 15: 297-9.
8. Evaldas Girdauskas, Kushtrim Dishaa, Michael A. Borger and Thomas Kuntze. Risk of proximal aortic dissection in patients with bicuspid aortic valve: how to address this controversy? Interactive cardiovascular and thoracic surgery (2013) 1-5
9. Edwards WD, Leaf DS, Edwards JE. Dissecting aortic aneurysm associated with congenital bicuspid aortic valve. Circulation 1978;57:1022-5.
10. Larson EW, Edwards WD. Risk factors for aortic dissection: a necropsy study of 161 cases. Am J Cardiol 1984;53:849-55.
11. Tribouilloy C, Rusinaru D, Sorel C et coll. Clinical characteristics and outcome of infective endocarditis in adults with bicuspid aortic valves: a multicentre observational study. Heart. 2010; 96(21):1723-9.
12. Hammond GL, Letsou GV. Aortic valve disease and hypertrophic cardiomyopathies. In: Baue AE ed.; Glenn's Thoracic and Cardiovascular Surgery. Connecticut: Appleton & Lange, 6th ed. 1996; pp 1981–2003.
13. Mautner GC, Mautner SL, Cannon RD, et al. Clinical factors useful in predicting aortic valve structure in patients >40 years of age with isolated valvular aortic stenosis. Am J Cardiol1993; 72: 194–8.
14. BEPPU S, SUZUKI S, MATSUDA H et al. Rapidity of progression of aortic stenosis in patients with congenital bicuspid aortic valves. Am J Cardiol, 1993; 71: 322-7.
15. Lamas CC, Eykyn SJ. Bicuspid aortic valve–A silent danger: analysis of 50 cases of infective endocarditis. Clin Infect Dis 2000; 30: 336–41.
16. Nada Abdulkareem, Jeremy Smelt and Marjan Jahangiri. Bicuspid aortic valve aortopathy: genetics, pathophysiology and medical therapy. Interactive cardiovascular and thoracic surgery 17 (2013) 554-559.l
17. B. Fernández, A. C. Durán, T. Fernández-Gallego et al., "Bicuspid aortic valves with different spatial orientation of the leaflets are distinct etiological entities," Journal of the American College of Cardiology, vol. 54, no. 24, pp. 2312–2318, 2009.
18. T. C. Lee, Y. D. Zhao, D. W. Courtman, and D. J. Stewart, "Abnormal aortic valve development in mice lacking endothelial nitric oxide synthase," Circulation, vol. 101, no. 20, pp. 2345–2348, 2000.
19. Huntington K, Hunter AG, Chan KL. A prospective study to assess the frequency of familial clustering of congenital bicuspid aortic valve. J Am CollCardiol. 1997;30:1809 –1812.
20. Clementi M, Notari L, Borghi A, Tenconi R. Familial congenital bicuspid aortic valve: a disorder of uncertain inheritance. Am J Med Genet. 1996; 62:336 –338.
21. J. F. M. Bechtel, F. Noack, F. Sayk, A. W. Erasmi, C. Bartels, and H. H. Sievers, "Histopathological

grading of ascending aortic aneurysm: comparison of patients with bicuspid versus tricuspid aortic valve," Journal of Heart Valve Disease, vol. 12, no. 1, pp. 54–61, 2003.
22. M. Cecconi, M. Manfrin, A. Moraca et al., "Aortic dimensions in patients with bicuspid aortic valve without significant valve dysfunction," American Journal of Cardiology, vol. 95, no. 2, pp. 292–294, 2005.
23. BROGER MA, DAVID TE. Management of the valve and ascending aorta in adults with bicuspid aortic valve disease.SeminThoracCardiovascSurg, 2005; 17: 143-7.
24. Nistri S, Sorbo MD, Marin M et al. Aortic root dilation in young men with normally functioning bicuspid aortic valves. Heart, 1999 : 82 : 19-22.
25. SABET HY. EDWARDS WD. TAZELAAR HD et al. Congenitally bicuspid aortic valves: a surgical pathology study of 542 cases . Mayo Clinproc, 1999;74: 14-26.
26. Yasuda H, Nakatani S, Stugaard M, Tsujita-Kuroda Y, Bando K, Kobayashi J, et al. Failure to prevent progressive dilation of ascending aorta by aortic valve replacement in patients with bicuspid aortic valve: comparison with tricuspid aortic valve. Circulation. 2003;108(Suppl 1):II291-4.
27. Robicsek F. The post stenotic dilatation. Acta Med Acad Sci 1954; (Suppl. IV): 1-4.
28. Jarchow B H, Kincaid O W. Poststenotic dilatation of the ascending aorta: Its occurrence and significance as a roentgenologic sign of aortic stenosis. Proc Staff Meet Mayo Clin. 1961;36:23–33.
29. M. G. Keane, S. E. Wiegers, T. Plappert, A. Pochettino, J. E. Bavaria, and S. M. G. S. John, "Bicuspid aortic valves are associated with aortic dilatation out of proportion to coexistent valvular lesions," Circulation, vol. 102, no. 19, pp. III35–III39, 2000.
30. G. Thanassoulis, J. W. L. Yip, K. Filion et al., "Retrospective study to identify predictors of the presence and rapid progression of aortic dilatation in patients with bicuspid aortic valves," Nature Clinical Practice Cardiovascular Medicine, vol. 5, no. 12, pp. 821–828, 2008.
31. Borger M A, Preston M, Ivanov J J. et al. Should the ascending aorta be replaced more frequently in patients with bicuspid aortic valve disease? J. J Thorac Cardiovasc Surg. 2004;128:677–683.
32. Lindsay J, Jr..Coarctation of the aortic, bicuspid valve and abnormal ascending aortic wall.Am J Cardiol. 1988;61:182–184.
33. M. E. Abbott, "Coarctation of the aorta of the adult type. II. A statistical study and historical retrospect of 200 recorded cases with autopsy, of stenosis or obliteration of the descending arch in subjects above the age of two years," American Heart Journal, vol. 3, no. 5, pp. 574–618, 1928.
34. WILSON W, TAUBERT KA , GEWITZ M et al . Prevention on infective Endocarditis. Guidelines from the American Heart Association. Circulaion, 2007; 115: 1-19.

Table 1: Demographic Data

Variable	Mean ± SD	%
Age (ans)	45,21 ± 11,85	
Sex ratio M/F		83,3 / 16,7
Hypertension		8,3
Smoking		50
Diabetes mellitus		4,2
Hyperlipidemia		8,3
NYHA I – II		41,6
III – IV		58,4
BMI kg/m²	25,2391 ± 3,9854	
CTR	0,5683 ± 0,08597	
RI		4,2
SBP (mmHg)	114,96 ± 15,176	
DBP (mmHg)	64,04 ± 13,836	
Euroscore	2,73 ± 2,857	
Log Euroscore	2,9795 ± 2,93416	

M/F: man/female; **SD**: standard deviation; **BMI** : Body mass index ; **CTR** : rapport cardiothoracic ratio ; **IR** : renal insufficiency ; **SBP** : systolic blood pressure ; **DBP** : diastolic blood pressure.

Table 2: Trans thoracic ultrasound data (24 cases)

Variable		Mean ± SD	%
LA	(mm)	42,67 ± 6,651	
DTS LV ESD	(mm)	45,48 ± 12,464	
DTD LV EDD	(mm)	62,62 ± 12,816	
SF	(%)	23,58 ± 7,596	
EF	(%)	51,08 ± 16,386	
PASP	(mmHg)	55,80 ± 18,054	
AS	(cm^2)	0,6889 ± 0,16159	
Gradient moyen	(mmHg)	55,13 ± 20,049	
AI grade I			29,2
AI grade II			20,8
AI grade III			8,3
AI grade IV			25
MI grade I			43,5
MI grade II			26,1
TI grade I			34,8
TI grade II			17,4

LA: left atrium ; **LV**: left ventricle ; **ESD**: end-systolic diameter ; **EDD**: end-diastolic diameter ; **SF**: shortening fraction ; **EF**: ejection fraction; **PASP**: pulmonary artery systolic pressure; **AS**: aortic surface; **AI**: aortic insufficiency; **MI**: mitral insufficiency; **TI**: tricuspid insufficiency.

Table 3: Operating data

Variable	Moyenne ± ET	%
No elective surgery (%)		4,2
DH (%)		15
UF (%)		10
AVR (%)		95,8333
Bentall (%)		4,1666
Use of inotropic drug (%)		16,7
LOCS (%)		12,5
CPB time (min)	99,25 ± 35,465	
Aortic cross clamping (min)	65,83 ± 24,923	
Mechanical ventilation time (h)	10,96 ± 12,103	
ICU stay (h)	37,67 ± 18,925	
Hospital stay (j)	12,35 ± 6.365	
Reexploration for bleeding (%)		8,3
RI with dialysis (%)		4,2
MV ≥ 48 h (%)		12,5
Hospital mortality (%)		0

DH : deep hemodilution; **UF** : ultra filtration ; **AVR** : aortic valve replacement; **LOCS** : low output cardiac syndrome ; **CPB** : cardiopulmonary bypass ; **ICU** : intensive care unit ; **RI** : renal insufficiency; **MV** : mechanical ventilation