

Anomalous Cord From the Raphe of a Congenitally Bicuspid Aortic Valve to the Aortic Wall Producing Either Acute or Chronic Aortic Regurgitation

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Objectives	This report calls attention to an unappreciated cause of both acute and chronic aortic regurgitation (AR).
Background	Although stenosis develops in most patients with a congenitally bicuspid aortic valve (BAV), in others with this anomaly, pure AR (no element of stenosis) develops, some in the absence of infection or other clear etiology.
Methods	We describe 5 men who underwent aortic valve replacement for pure AR associated with a BAV containing an anomalous cord attaching the raphe of the conjoined cusp near its free margin to the wall of the ascending aorta cephalad to the sinotubular junction.
Results	Three of these 5 patients had a history of progressive dyspnea, and the anomalous cord, which was intact at operation, appeared to cause chronic AR by preventing proper coaptation of the 2 aortic valve cusps. The other 2 patients heard a “pop” during physical exertion and immediately became dyspneic, and at operation, the anomalous cord was found to have ruptured. Prolapse of the conjoined aortic valve cusp toward the left ventricular cavity resulted in severe acute AR.
Conclusions	This variant of the purely regurgitant BAV may cause either chronic AR (when the anomalous cord does not rupture) or acute severe AR (when the cord ruptures). (J Am Coll Cardiol 2014;63:153–7) © 2014 by the American College of Cardiology Foundation

The congenitally bicuspid aortic valve (BAV) occurs in an estimated 1% of the population, such that in the United States, an estimated 3 million individuals have this malformation (1). Although in some individuals the BAV functions normally for an entire lifetime, stenosis develops in most, superimposed infection (infective endocarditis)

develops in some, and pure aortic regurgitation (AR) unassociated with infective endocarditis or its consequences develops in some (2). A subgroup of those with pure AR unassociated with infection have an anomalous cord extending from the raphe of 1 of the 2 cusps to the wall of the aorta, and the cord serves to keep the raphe cusp from prolapsing toward the left ventricle (Table 1). During a 50-year period, we examined 5 operatively excised purely regurgitant BAVs with an anomalous cord from the raphe to the wall of the aorta. A brief description of these 5 cases is the purpose of this report.

Methods

During a nearly 50-year period, we examined 5 operatively excised aortic valves that were congenitally bicuspid and had a cord attached from the margin of the raphe cusp to the wall of the aorta. The clinical records in all 5 patients were subsequently examined, and all 5 valves were photographed.

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Abbreviations and Acronyms

AR = aortic regurgitation
BAV = bicuspid aortic valve

Results

Pertinent findings in the 5 patients are summarized in Table 2; all patients were men. Three (Patients #1, #2, and #5) had chronic AR and 2 had acute AR. The latter 2 were asymptomatic until 1 or 2 days before aortic valve replacement: both heard a popping noise in his chest, 1 patient while working in his yard and 1 while working on his car beneath the hood. Both became suddenly dyspneic, and the dyspnea progressed rather rapidly, prompting pulmonary edema and hospitalization. A median sternotomy was emergently performed in each for severe AR. The cord extending from the raphe to the wall of the aorta had ruptured, causing the raphe cusp to prolapse toward the left ventricular cavity (Fig. 1). Each of the other 3 patients had chronic AR, and in none of them had the anomalous cord ruptured.

Photographs of the operatively excised BAVs are shown in Figures 2 to 5.

Discussion

This report describes 5 men with a BAV and pure AR with an anomalous cord extending from the raphe of the conjoined cusp to the wall of the ascending aorta cephalad to the sinotubular junction. In the 3 patients who presented with chronic AR, the cord appeared to prevent complete coaptation of the 2 aortic valve cusps by pinning the raphe of the conjoined cusp to the aortic wall. In the 2 patients who presented with acute AR, the cord had ruptured, resulting in the conjoined cusp prolapsing toward the left ventricular cavity and the acute onset of symptoms.

This variant of the BAV has been previously described (Table 1), and its association with AR has been well documented (1,3-13). Of the 33 previously published cases, 25

Table 1 Previous Publications of Patients Having an Anomalous Cord Extending From the Raphe of a Congenitally BAV to the Wall of the Aorta Causing Either Acute or Chronic Pure Aortic Regurgitation

Year of Publication	First Author (Ref. #)	No. of Patients With a BAV Containing an Anomalous Cord	No. With Cord Rupture	Mean Age, yrs (Range)	No. of Men	Severity of AR (No. of Patients)			
						1+	2+	3+	4+
1970	Roberts (1)	1*	0	27	1	0	0	0	1
1971	Carter et al. (3)	1	1	59	1	NA	NA	NA	NA
1977	Becker and Düren (4)	1	1	45	1	0	0	0	1
1984	Olson et al. (5)	11†	1	NA	NA	NA	NA	NA	NA
1986	Yamagishi et al. (6)	1	0	32	1	0	0	0	1
1990	Waller et al. (7)	2	0	53	2	0	0	0	2
1990	Arikawa et al. (8)	2	NA	NA	NA	NA	NA	NA	NA
1992	Hamada et al. (9)	2	1	53	NA	NA	NA	1	NA
1993	Misawa et al. (10)	1	NA	NA	NA	NA	NA	NA	NA
1994	Walley et al. (11)	9	2	61 (46-73)	7	1	1	1	6
2000	Akiyama et al. (12)	1	1	57	1	0	0	1	0
2011	Journigan and Clements (13)	1	1	48	1	0	0	0	1
Total		33	8 (24%)		15/17 (88%)				12/16 (75%)

*Patient is included in the present study (Patient #1). †The author described the bicuspid aortic valves as containing "either a fenestrated raphe or a raphe cord."
BAV = bicuspid aortic valve; NA = not available.

Table 2 Clinical Data for 6 Men Having AVR for a Purely Regurgitant (4+/4+) Congenitally Bicuspid Aortic Valve With an Anomalous Cord Extending From the Raphe of One Cusp to the Wall of the Aorta

Patient #	Age, yrs	Age, yrs		SH	LVEF, %*	Rupture of Anomalous Cord	AVR (yr)	Type of Valve Implanted
		Precordial Murmur First Heard	S/S of Heart Failure First Appeared					
1†‡	27	17	25	0	NA	-	1966	Mechanical
2	45	NA	NA	+	50	-	2013	Mechanical
3	49	49	49	0	55	+§	2012	Bioprosthesis
4	55	55	55	0	NA	+§	2012	Bioprosthesis
5	71	70	70	+	50	-	2012	Bioprosthesis

*LVEF was measured by echocardiography in Patients #2 and #3 and by cardiac catheterization in patient 5. †Patient was previously reported by Roberts (1) in 1970. ‡Patient's aorta coursed over his right main bronchus (right aortic arch). §Producing acute aortic regurgitation.
AVR = aortic valve replacement; LVEF = left ventricular ejection fraction; NA = not available; SH = systemic hypertension; S/S = signs and symptoms; + = positive or present; - = negative or absent.

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