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 ventricular cavity (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.
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.

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

### Abbreviations and Acronyms

**AR** = aortic regurgitation

**BAV** = bicuspid aortic valve

### Results

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

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

*Patient is included in the present study (Patient #1). The author described the bicuspid aortic valves as containing “either a fenestrated raphe or a raphal 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

<table>
<thead>
<tr>
<th>Patient #</th>
<th>Age, yrs</th>
<th>Precordial Murmur First Heard</th>
<th>S/S of Heart Failure First Appeared</th>
<th>SH</th>
<th>LVEF, %*</th>
<th>Rupture of Anomalous Cord</th>
<th>AVR (yr)</th>
<th>Type of Valve Implanted</th>
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</thead>
<tbody>
<tr>
<td>1*</td>
<td>27</td>
<td>17</td>
<td>25</td>
<td>0</td>
<td>NA</td>
<td>–</td>
<td>1966</td>
<td>Mechanical</td>
</tr>
<tr>
<td>2</td>
<td>45</td>
<td>NA</td>
<td>+</td>
<td>50</td>
<td>–</td>
<td>–</td>
<td>2013</td>
<td>Mechanical</td>
</tr>
<tr>
<td>3</td>
<td>49</td>
<td>49</td>
<td>49</td>
<td>0</td>
<td>55</td>
<td>+</td>
<td>2012</td>
<td>Bioprosthesis</td>
</tr>
<tr>
<td>4</td>
<td>55</td>
<td>55</td>
<td>55</td>
<td>0</td>
<td>NA</td>
<td>+</td>
<td>2012</td>
<td>Bioprosthesis</td>
</tr>
<tr>
<td>5</td>
<td>71</td>
<td>70</td>
<td>70</td>
<td>+</td>
<td>50</td>
<td>–</td>
<td>2012</td>
<td>Bioprosthesis</td>
</tr>
</tbody>
</table>

*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.
Figure 1  **Patient #3: Pre-Operative Transesophageal Echocardiogram Showing a Congenitally Bicuspid Aortic Valve**

(A) Long-axis view showing the conjoined cusp (arrow) prolapsing toward the left ventricular cavity during diastole. (B) Long-axis view showing the anomalous cord (arrow) prolapsing toward the left ventricular cavity during diastole. (C) Long-axis color Doppler view showing aortic regurgitation. (D) Short-axis view showing the flail cord (arrow). AV = aortic valve; LV = left ventricle.

Figure 2  **Patient #1**

(A) A congenitally bicuspid aortic valve devoid of calcific deposits containing an anomalous cord attached to the conjoined cusp in a 27-year-old man who was found to have a murmur consistent with aortic regurgitation at age 17 years. He remained asymptomatic until age 25 years when exertional dyspnea appeared. On pre-operative cardiac catheterization, the left ventricular pressure was 125/35 mm Hg, the brachial arterial pressure was 125/55 mm Hg, and the cardiac index (Fick method) was 2.2 l/min/m². The patient underwent aortic valve replacement with a mechanical aortic valve prosthesis for chronic aortic regurgitation and died 13 months later. His heart weighed 800 g. (B) Diagram of a short-axis view of the aortic aspect of the bicuspid aortic valve before aortic valve replacement showing the attachment of the anomalous cord from the raphe to the wall of the aorta. (C) Diagram of a long-axis view of the same valve. Reproduced with permission from Roberts [1].
Figure 3  Patient #2

A congenitally bicuspid aortic valve with an anomalous cord extending from the raphe of the conjoined right and left coronary cusps in a 45-year-old man with a history of dyspnea. Pre-operative echocardiogram revealed 4+/4+ aortic regurgitation and a 50% left ventricular ejection fraction. He underwent successful aortic valve replacement with a 27 mm St. Jude Medical Regent mechanical aortic valve (St. Jude Medical Inc., St. Paul, Minnesota). (A) View of valve from ventricular aspect. (B) View from aortic aspect. (C) Lateral view showing the length of the anomalous cord. (D) Similar to C.

Figure 4  Patient #3

(A) The aortic aspect of a congenitally bicuspid aortic valve with an anomalous cord extending from the raphe in a 49-year-old man who heard a loud “pop” while working in his yard with subsequent chest pain and dyspnea. Two days later, he presented to the emergency department with severe dyspnea, and a chest radiograph revealed bilateral pulmonary edema. A precordial murmur was later heard, and echocardiography revealed a “flail” aortic valve cusp causing severe aortic regurgitation. The left ventricular ejection fraction was 55%. Aortotomy disclosed an anomalous cord attached to his bicuspid aortic valve, and it had ruptured. The valve was replaced with a 23-mm Carpentier-Edwards PERIMOUNT pericardial bioprosthesis (Edward Lifesciences Inc., Irvine, California). (B) The torn edge of the anomalous cord. (C) From the side of the same valve.
(76%) had evidence of chronic AR as a result of having an intact anomalous cord, and 8 (24%) patients presented with acute AR due to rupture of the anomalous cord. Waller et al. (14) in 1973 described 2 other patients, not included in Table 2, with similar BAVs: 1 was an 8-year-old boy with coarctation of the aorta, and 1 was an 81-year-old man with no other associated congenital anomalies. There was no mention of function of the BAV in these 2 patients. In 1994, Walley et al. (11) described 8 patients with this anomalous cord attached to a stenotic BAV in addition to the 9 patients with a purely regurgitant BAV included in Table 1. The presence of an anomalous cord extending to the wall of the aorta from the raphe of a BAV thus may be associated with stenosis but far more commonly with pure AR.

In none of the 5 men included in the present study was the diagnosis of this variant of the purely regurgitant BAV made pre-operatively. Most symptomatic patients with this congenital anomaly present with chronic AR and slowly progressing symptoms. Rupture of an anomalous cord should be considered as a possible etiology of pure acute AR when infection, aortic dissection, and trauma have been ruled out.

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REFERENCES


Key Words: aortic regurgitation ▪ bicuspid aortic valve ▪ congenital heart disease.