

# Severe Aortic Regurgitation Resulting from a Downward Displacement of Anterior Aortic Annulus and Fibrous Strands in the Bicuspid Aortic Valve

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**Aortic regurgitation is occasionally caused by fibrous strands and more rarely by downward displacement of the aortic annulus. The present report describes an 18-year-old man with aortic regurgitation resulting from an anterior-posterior type of bicuspid aortic valve with fibrous strands and downward displacement of the anterior aortic annulus. A pair of fibrous strands at the anterior cusp of the bicuspid valve lifted the free margin of the cusp, and the anterior cusp originated from the intraventricular septum. We considered that the aortic regurgitation was due to poor coaptation of the cusps because of these two conditions. After resection of the cusps and the strands, the aortic valve was replaced at the intra-annular position. (Ann Thorac Cardiovasc Surg 2010; 16: 57–59)**

**Key words:** bicuspid aortic valve, aortic regurgitation, fibrous strand, downward displacement

## Introduction

Fibrous strands comprise one cause of aortic regurgitation, but this is very rare, and downward displacement of the aortic annulus is extremely rare. Here we describe an 18-year-old man who presented with aortic regurgitation caused by an anterior-posterior type of bicuspid aortic valve with fibrous strands and downward displacement of the anterior aortic annulus.

## Case Report

An 18-year-old man (183.8 cm tall; weight 80.2 kg) became fatigued during exercise in early 2006. Electrocardiography

during a routine school physical revealed an abnormality in April 2006. He was further examined at a nearby clinic and referred to our hospital with a diagnosis of aortic regurgitation. A physical examination upon admission revealed blood pressure of 130/68 mmHg and diastolic murmur at the 3rd left sternal border. Laboratory tests were normal, and electrocardiography showed left axis deviation and left ventricular hypertrophy. Computed tomography revealed no calcification at the aortic valve and no dilation of the ascending aorta. Preoperative ultrasonic cardiography revealed an anterior-posterior type bicuspid aortic valve, and a strand suspended the margin of the anterior coronary cusp from the aortic wall. Severe aortic regurgitation was also identified. Preoperative cineangiography of the ascending aorta revealed IV/IV aortic regurgitation.

Under median sternotomy and standard cardiopulmonary bypass with warm blood cardioplegia, the aortic valve was approached through an oblique aortotomy. The bicuspid aortic valve was anterior-posterior, and the two coronary arteries originated from the anterior sinus. A strand suspended the free margin of the anterior cusp from the aortic wall and seemed to lift the margin of the

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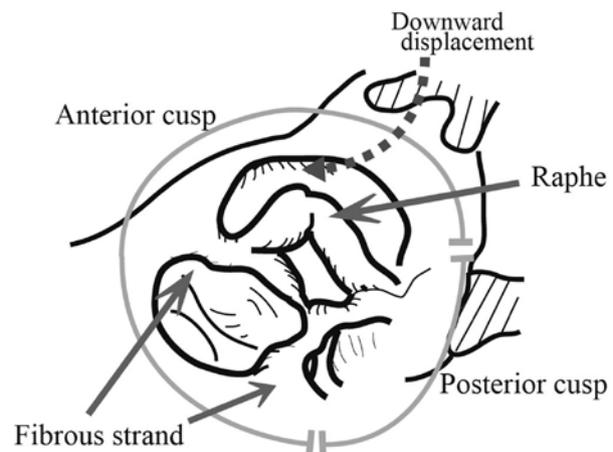
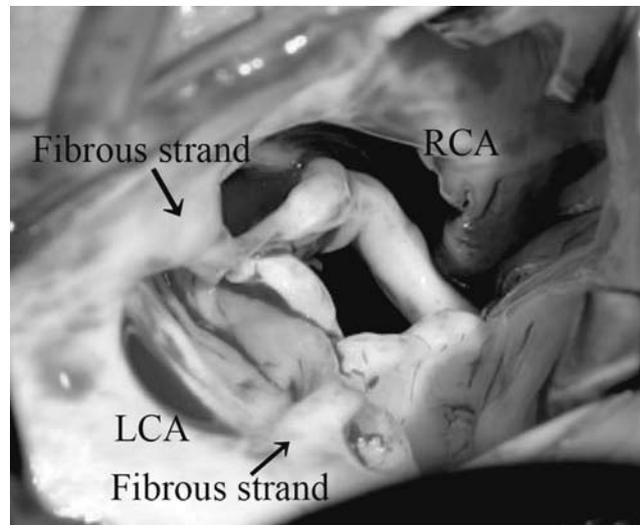
cusps. The other fibrous strand was attached to the posterior cusp at the left side of the commissure (Fig. 1). Infectious processes were absent. The anterior cusp of the aortic valve originated from the intraventricular septum, in other words, downward displacement of the right aortic annulus. Aortic regurgitation was thought to arise because of poor coaptation of the cusps as a result of these two causes. The aortic valve was resected and sutured with an everting mattress at the site of the anterior cusp origin; it was replaced with a mechanical heart valve (ATS 25 mm, ATS Medical, Inc., Lake Forest, CA, USA) at the intra-annular position. The postoperative course of the patient proceeded smoothly without rupture of the mechanical valve.

### Discussion

Congenitally bicuspid aortic valves occur in 0.9% to 2% of the general population.<sup>1)</sup> Haiden and colleagues reported that 542 (29%) of 1,877 surgically excised aortic valves were bicuspid at the Mayo Clinic from 1991 to 1996 and that pure aortic regurgitation occurred in 73 (13%) of them.<sup>2)</sup> The etiology of aortic regurgitation in patients with a bicuspid aortic valve is complex. It can occur in isolation, usually as a result of prolapse of the larger of unequally sized cusps, but also in association with aortic root dilation or infective endocarditis.<sup>1,3)</sup> Pure aortic regurgitation with fibrous strands is considered rare.

Misawa and colleagues described the origin of fibrous strands as embryonic remnants.<sup>4)</sup> From an embryological perspective, the aortic valve and sinuses of Valsalva are formed by distal aortic bulges that extend toward the ventricular cavity. Thus this evolution could leave fibrous tissue between the aortic valve and the aortic wall.<sup>5)</sup> The persistence of such tissue might not normally contribute to aortic valvular function, and pure aortic regurgitation does not develop only as a result of fibrous strands. Two causes of aortic regurgitation have been suggested. Coaptation of the aortic valve can be disturbed by dilation of the ascending aorta to which fibrous strands become attached as a result of growth and aging,<sup>6)</sup> and spontaneous rupture of fibrous strands might be the other cause.<sup>7,8)</sup> However, rupture is considered to be a cause of acute aortic regurgitation. A heart murmur and other symptoms were not recognized in our patient during childhood, so we postulated that a dilation of the ascending aorta because of growth resulted in fibrous strands lifting the margin of the cusp.

The attachment of the anterior cusp was displaced



**Fig. 1.** Intraoperative findings and schema of bicuspid aortic valve.

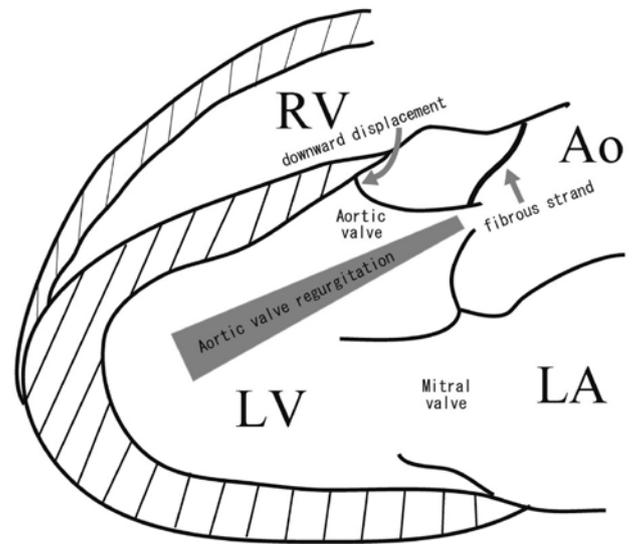
A: Intraoperative findings.

RCA, right coronary artery; LCA, left coronary artery.

B: Schema.

Arrows indicate strands from aortic wall lifting free margin of cusps. The other arrow indicates raphe. The attachment of the anterior cusp was displaced downward onto the interventricular septum (dashed arrow).

downward onto the interventricular septum in our patient (Fig. 2). This condition is extremely rare, since this is only the fourth description of this kind in the literature of congenital aortic annulus displacement. No general consensus exists about the embryological pathogenesis of this condition. Hou and colleagues described combined aortic and mitral regurgitation caused by downward displacement of the left and noncoronary aortic annulus, but they could not rule out the possibility of rheumatic changes.<sup>9)</sup> Matsukawa and colleagues described a semilunar



**Fig. 2.** Schema of downward displacement and the fibrous strand.

Arrows indicate the downward displacement and the fibrous strands. The downward displaced cusp is lifted strongly by the fibrous strands, and the aortic regurgitation is caused by the poor aortic valve coaptation.

RV, right ventricle; LV, left ventricle; Ao, aorta; LA, left atrium.

valve that developed from the bulbar ridges, which also participated in annular formation, and noted that the right coronary cusp could originate from the ventricular myocardium.<sup>10</sup> In this case, the annular origin of the anterior cusp was deep on the interventricular septum, so we emphasize the possibility of a congenital abnormality of the anterior aortic annulus. Such downward displacement was thought to be one cause of the aortic regurgitation in our patient as a result of poor aortic valve coaptation, because the downward displaced cusp was lifted more strongly by the fibrous strands. In summary, we described an 18-year-old man with pure aortic regurgitation of the bicuspid aortic valve. We postulated that the causes of poor aortic valve coaptation were displacement of the origin of the anterior cusp downward to the interventricular septum and lifting of the cusp margin by fibrous strands.

## References

1. Nuran Y, Levent O, Dilek E, Murat Y, Ali Y. Bicuspid aortic valve. *Ann Thorac Cardiovasc Surg* 2002; **8**: 264–7.
2. Haiden YS, William DE, Henry DT, Richard CD. Congenitally bicuspid aortic valves: a surgical pathology study of 542 cases (1991 through 1996) and a literature review of 2715 additional cases. *Mayo Clin Proc* 1999; **74**: 14–26.
3. Ward C. Clinical significance of the bicuspid aortic valve. *Heart* 2000; **83**: 81–5.
4. Misawa Y, Hasegawa T, Oyama H, Sudo H, Hasegawa N, et al. Congenital bicuspid aortic valve with regurgitation—a rare case showing a fibrous band between the conjoined cusp and the ascending aorta—. *Nippon Kyobu Geka Gakkai Zasshi (J Jpn Assn Thorac Surg)* 1993; **41**: 2156–9. (in Jpse.)
5. Nakajima M, Tsuchiya K, Naito Y, Hibino N, Inoue H. Aortic regurgitation caused by rupture of a well-balanced fibrous strand suspending a degenerative tricuspid aortic valve. *J Thorac Cardiovasc Surg* 2002; **124**: 843–4.
6. Yamagishi M, Anzai N, Yamada M. An exceptional form of congenitally bicuspid aortic valve resulting in pure aortic regurgitation. *Jpn Heart J* 1986; **27**: 267–71.
7. Hamada Y, Iijima M, Yoshida I, Tsuda K, Ishikawa S, et al. Bicuspid aortic valve with sudden onset of aortic insufficiency due to rare causes: report of two cases. *Kyobu Geka* 1992; **45**: 519–21. (in Jpse.)
8. Tamura K, Shirai T, Ohshima N. Aortic regurgitation caused by rupture of a fibrous strand: report of a case. *Kyobu Geka* 2006; **59**: 411–3. (in Jpse.)
9. Hou SH, Chu SH, Wang SS, Tsai BW, Chen MF, et al. Downward displacement of aortic annulus leading to aortic and mitral regurgitation. *J Thorac Cardiovasc Surg* 1986; **92**: 308–10.
10. Matsukawa T, Yoshii S, Hashimoto R, Maeda T, Yoshizaki T, et al. A rare case of congenital downward displacement of right aortic annulus resulting in severe aortic regurgitation. *Yamanashi Med J* 1990; **5**: 75–8.