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CASE REPORT
Valvular heart disease

Acute severe aortic insufficiency during cardiopulmonary bypass in a bicuspid aortic valve with unrecognized annular displacement and fibrous strands: a case report

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Background Bicuspid aortic valve (BAV) with displacement of the attachment of the conjoined aortic leaflet and fibrous strands is a rare cardiac malformation. We report a case of BAV that presented as acute massive aortic regurgitation during cardiopulmonary bypass for a planned non-valve–related procedure and was successfully treated by emergency aortic valve replacement.

Case summary A 70-year-old man with triple vessel coronary disease and severe left ventricular systolic dysfunction underwent coronary bypass grafting and graft replacement of the ascending aorta. Acute aortic regurgitation occurred during ventricular fibrillation and after declamping of the aortic graft. Intra-operative findings included a fused BAV (right–left cusp fusion), very asymmetrical leaflet (commissure angle of the non-fused leaflet 135°), three aortic sinuses, and conjoined leaflets originating from the myocardium in the inter-ventricular septum. The aortic leaflets were resected and replaced with a prosthetic aortic valve at the attachment site of the conjoined leaflets. Post-operatively, no peri-valvular leaks were observed, and left ventricular function was improved.

Discussion Intra-operative acute massive aortic regurgitation may be caused by a morphologically abnormal aortic leaflet and root complex in patients with a BAV. The dilated aortic root, asymmetrical leaflet, and abnormal aortic leaflet insertion, with thick septal myocardium of the coronary aortic sinus, might have caused unstable leaflet co-aptation, leading to deformation of the aortic leaflets influenced by the change in myocardial tone and intra-operative change in the sinotubular junction. Familiarity with the classification of congenital BAV, and the anatomy of the normal and abnormal aortic root complex, is important.

Keywords Bicuspid aortic valve • Displacement of attachment of aortic valve • Myocardial crescents • Fibrous strand • Acute aortic regurgitation • Case report

ESC curriculum 2.2 Echocardiography • 4.1 Aortic regurgitation • 7.5 Cardiac surgery • 9.7 Adult congenital heart disease • 9.1 Aortic disease

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Learning points

- To describe the anatomy of the aortic valve and aortic root according to the international consensus statement.
- To gain knowledge about the anatomy of the normal and abnormal aortic root complex and about the mechanisms of intra-operative acute aortic regurgitation caused by the abnormal aortic root complex.
- To learn tips for performing aortic valve replacement in patients with an abnormal insertion of aortic conjoined leaflets.

Introduction

Bicuspid aortic valve (BAV) with displacement of the attachment of the conjoined aortic leaflet and fibrous strands is a rare cardiac malformation. Herein, we report a case in which a BAV presented as acute massive aortic regurgitation during cardiopulmonary bypass for a planned non-valve–related procedure and was treated by emergency aortic valve replacement.

Summary figure

Time	Event
September 2018 (age 70 years)	A 70-year-old man with hypertension, dyslipidaemia, and emphysema was transferred to a previous hospital because of dyspnoea on exertion and general fatigue. He was treated for acute heart failure, and trans-thoracic echocardiography (TTE) showed severe left ventricular (LV) dysfunction with regional akinesis. Coronary angiography revealed chronic total occlusion of the right coronary artery (#1 100%) and left anterior descending artery (#7 proximal 100%), and severe stenosis of the left circumflex coronary artery (#13 75%). He was diagnosed with severe triple vessel coronary disease and reduced LV function.
October 2018	The patient was transferred to our hospital for bypass surgery. The chest radiograph showed cardiomegaly (cardiothoracic ratio 60%), and the electrocardiogram showed sinus rhythm with abnormal Q waves in II, III, and aVF. His brain natriuretic peptide and troponin I levels were elevated to 532 and 231 ng/mL. Pre-operative TTE showed moderate LV dilation (LVDd/Ds = 54/46 mm), severe LV systolic dysfunction [LV ejection fraction (LVEF) 38%] with regional akinesis, and a probable tricuspid aortic valve with trivial aortic regurgitation. Pre-operative computed tomography showed dilatation of the ascending aorta measuring 51 mm, mild dilatation of the sinotubular junction of 41 mm, and Valsalva sinus of 47 mm.
18 October 2018	The patient underwent coronary artery bypass grafting and graft replacement of the ascending aorta, sparing the normally functioning aortic valve. Intra-operatively, massive acute severe aortic regurgitation was observed, which made it difficult to maintain the cardiopulmonary bypass flow. The aorta was re-clamped, and we found that the aortic valve was a fused type (right–left fusion) BAV with displacement of the attachment of the conjoined aortic leaflet and fibrous strands. Emergency aortic valve replacement was performed, and the patient was successfully weaned off cardiopulmonary bypass. The postoperative course was uneventful.
November 2022	At 4 years after the operation, the patient had no heart failure symptoms and TTE showed improved LV function (LVEF 60%).

Case presentation

A 70-year-old man with dyspnoea on exertion was diagnosed with triple vessel coronary disease (*Figure 1A and B*) with reduced LV systolic function (LVEF 32%). The patient's past medical history included hypertension, dyslipidaemia, and emphysema. Trans-thoracic echocardiography revealed what appeared to be a tricuspid aortic valve with trivial aortic regurgitation (*Figure 1C–E*). Computed tomography showed dilatation of the ascending aorta of 51 mm, mild dilatation of the sinotubular junction of 41 mm, and a Valsalva sinus of 47 mm. His brain natriuretic peptide and troponin I levels were elevated to 532 and 231 ng/mL. Cardiac CT was considered to be relatively contraindicated because of the presence of renal dysfunction (estimated glomerular filtration rate of <30 mL/m^2) and heart failure due to reduced LV function. The patient was taking anti-hypertensive drugs, beta-blockers, aldosterone receptor antagonists, diuretics, and statins, which consist of candesartan 4 mg, bisoprolol fumarate 1.25 mg, spironolactone 12.5 mg, azosemide 30 mg, tolvaptan 3.75 mg, and atorvastatin 10 mg. The patient was recommended coronary artery bypass grafting due to triple vessel disease with a high SYNTAX score (40.5), reduced LV function, and ascending aortic pathology with an indication for surgery.

We performed coronary artery bypass grafting and graft replacement of the ascending aorta (28 mm J Graft; Japan Lifeline Inc., Tokyo, Japan) with sparing of the normally functioning aortic valve. Acute aortic regurgitation occurred with ventricular fibrillation during cooling for deep hypothermic circulatory arrest prior to the graft replacement. After de-clamping the aortic graft, the left ventricle rapidly became distended and the heart did not resume beating. Trans-oesophageal echocardiography revealed massive aortic regurgitation without dissection of the aortic root. The aorta was re-clamped, and we found that the aortic valve was the fused type (right–left fusion) BAV, very asymmetrical leaflet (commissure angle of the non-fused

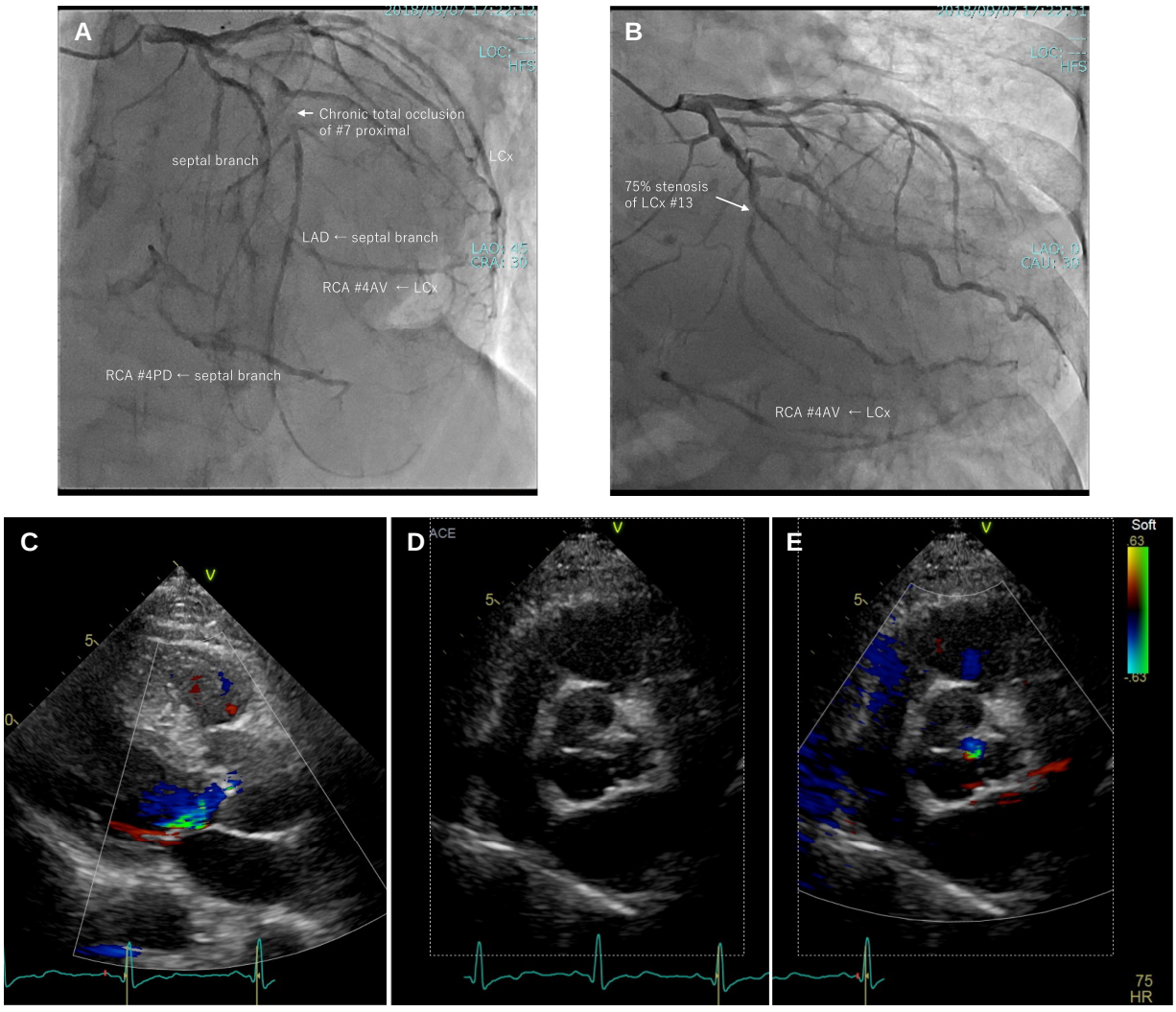


Figure 1 (*A*, *B*) Pre-operative coronary angiography. (*A*) Left anterior oblique 45°, cranial 30°; chronic total occlusion of the proximal left anterior descending artery (#7) and right coronary artery (#1). The right coronary artery #4PD and left anterior descending artery were supplied by collateral circulation from the septal branch, while the right coronary artery #4AV was supplied by collateral circulation from the left circumflex branch. (*B*) Left anterior oblique 0°, caudal 30°; 75% stenosis of the left circumflex artery (#13). (*C*–*D*) Pre-operative trans-thoracic echocardiography. (*C*) Trivial aortic regurgitation in a parasternal longitudinal view. (*D*) A probable trileaflet aortic valve. (*E*) Trivial aortic regurgitation in a short axis view. AV, atrioventricular node artery; LAD, left anterior descending artery; LAO, left anterior oblique; LCx, left circumflex artery; PD, posterior descending artery; RCA right coronary artery.

leaflet 135°), and three aortic sinuses. The conjoined aortic leaflet originated from the myocardium in the coronary aortic sinuses without the normal attachment to the fibrous portion of the aortic annulus (*Figure 2A*). The fibrous strands originated from the raphe of the conjoined leaflet connected to the aortic wall (*Figure 2B*). The aortic leaflet, fibrous strands, and aortic root were not injured by manipulation of the proximal anastomosis of the ascending aorta. We resected the aortic leaflets and found that the attachment of the conjoined leaflet was separate from the anatomical ventriculo-arterial junction without fibrous connection and was displaced downwards into the left ventricle. A prosthetic valve (CEP MAGNA EASE #25 mm; Edwards Lifesciences, Irvine, USA) was implanted at the supra-annular position using non-everting mattress sutures from the site of attachment of the conjoined aortic leaflet. The patient was successfully weaned from cardiopulmonary bypass (*Figure 3*). We reviewed the pre-operative echocardiographic findings and found that the width of the myocardium between the attachment of the leaflets and their supporting coronary sinuses was 17.1 mm, which was consistent with the intra-operative findings (*Figure 4*). One year later, there was no paravalvular leak, and the patient's LV function had recovered well (LVEF 63%).

Discussion

Bicuspid aortic valve with displacement of the attachment of the conjoined aortic leaflet and fibrous strands is a rare cardiac malformation that can cause aortic regurgitation. There has been a report of a case in which a BAV was displaced downwards by fibrous strands and complicated by chronic aortic regurgitation due to poor co-aptation of the aortic valve following aortic dilatation.[1] Moreover, there have been

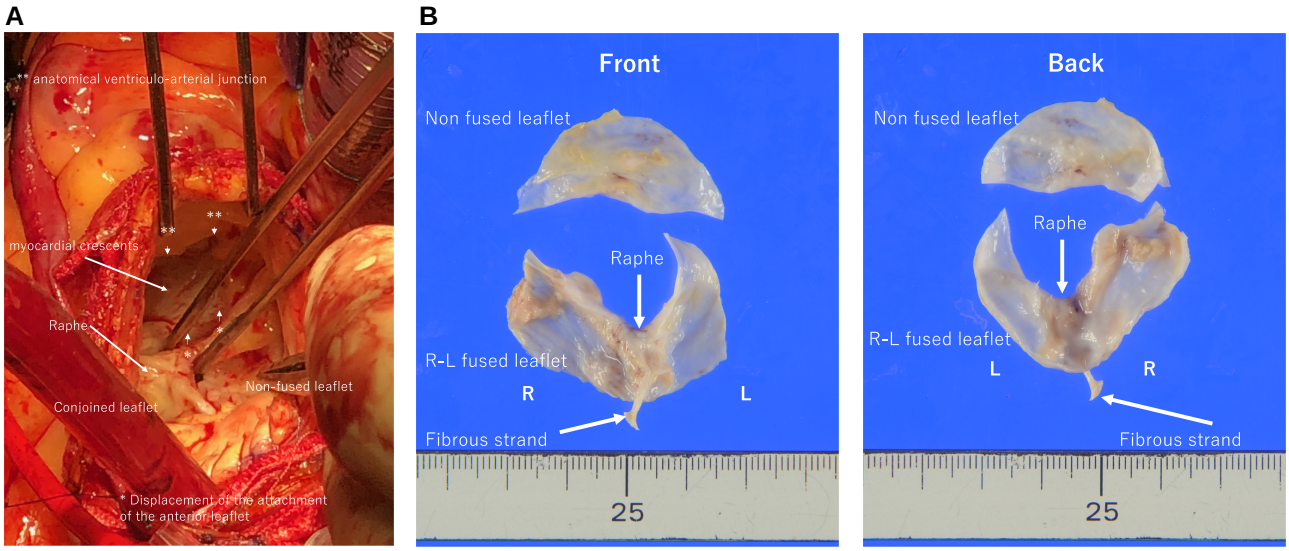


Figure 2 (*A*) Surgical view of the aortic leaflets with fibrous strands and displacement of the attachment of the conjoined leaflet onto the interventricular septum. (*B*) Pathophysiological findings: resected congenital bicuspid aortic leaflets with fibrous strands.

Figure 3 Coronary artery bypass surgery (LITA-LAD, Ao-4PD-4AV with saphenous vein graft) and graft replacement of the ascending aorta. An unscheduled aortic valve replacement was also performed. Ao, aorta; AV atrioventricular node artery, LAD left anterior descending artery, LITA left internal thoracic artery, PD posterior descending artery.

three reports of chordae tendineae strands on a BAV with acute aortic regurgitation due to spontaneous rupture of the chordae.[2–4]

In our case, the acute massive intra-operative aortic regurgitation during cardiopulmonary bypass could have been caused by a morphologically abnormal aortic root complex. Our patient had a fused BAV (right–left cusp fusion), very asymmetrical leaflet (commissure angle of the non-fused leaflet 135°), and three aortic sinuses, as classified according to the European Journal of Cardio-Thoracic Surgery guidelines.[4] This bicuspid valve is classified as Type 1 (L/R) according to the Sievers classification.

Very asymmetrical valves may exhibit retraction of the free edge of the fused cusp at the raphe level,[5] which is best appreciated by direct surgical visualization or pathological inspection and not reliably shown by imaging. This retraction may make the aortic valve more susceptible to restriction by operative procedures.

We found a ventricular myocardium (myocardial crescent) at the base of the conjoined leaflet in the coronary aortic sinuses. The myocardial crescent is the superior aspect of the ventricular septal myocardium supporting the base of the aortic valvular sinuses that gives rise to the coronary arteries in patients with a tricuspid aortic valve.[6] In patients with a fused BAV (right–left cusp fusion), this type of myocardium is found at the base of the conjoined leaflet supported by the aortic coronary sinuses.[5] De Kerchove *et al.*[7] reported that the root base thickness is thicker at the levels of the left/right commissure and the right coronary sinus, and the mean width of the muscular inclusion at the base of the right coronary sinus is 6.2 mm in tricuspid aortic valves. De Kerchove *et al.*[5] also reported that the median muscular width at the base of the right coronary sinus is 5 mm and is similar in three phenotypic groups of BAV.

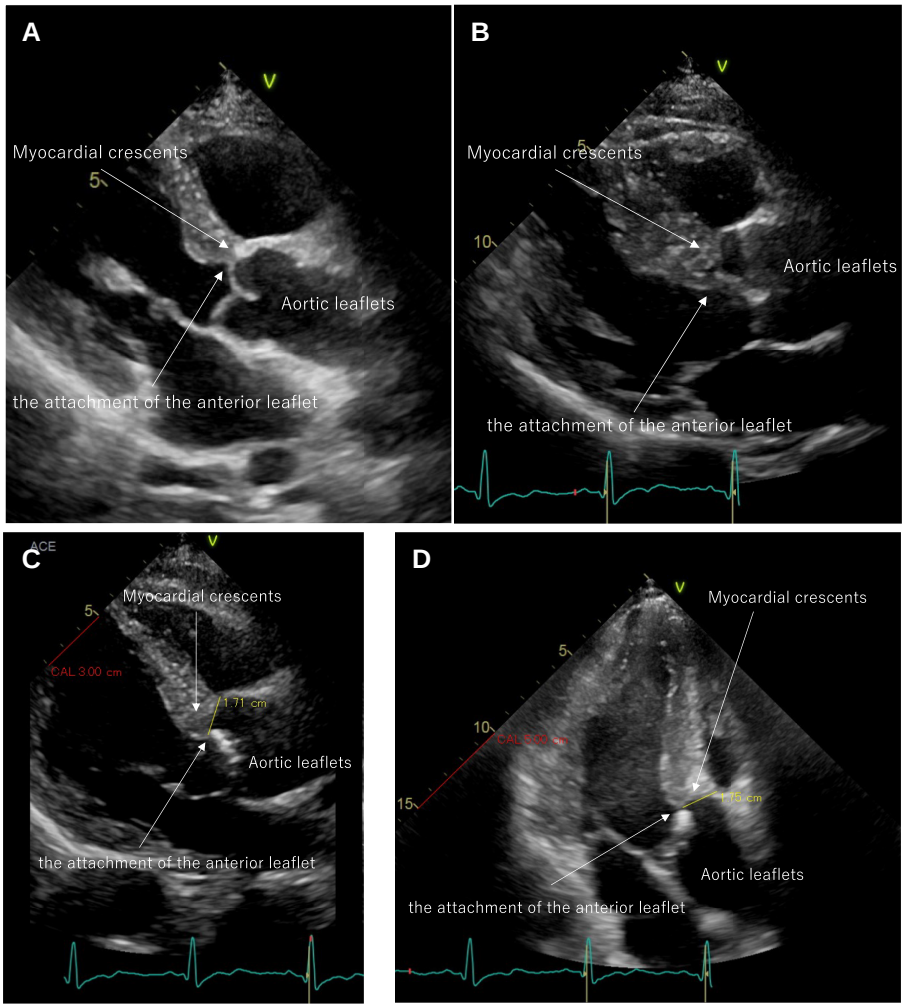


Figure 4 Trans-thoracic echocardiography. (*A*) Parasternal longitudinal view of a normal tricuspid aortic valve. (*B*) The conjoined leaflet originated from the inter-ventricular septum; the attachment of aortic leaflets was separated from the anatomical ventriculo-arterial junction and was displaced downwards into the left ventricle. (*C*) Parasternal longitudinal view showing that the width of the myocardium between the attachment of the leaflets and their supporting coronary sinuses was 17.1 mm. (*D*) Apical three-chamber view showing that the width of the myocardial crescent was 17.5 mm.

In our case, the width of the myocardium between the attachment of the leaflets and their supporting coronary sinuses was 17.1 mm (*Figure 4*), which is almost triple the length reported previously.[5,7] Furthermore, in our patient, there was no fibrous continuity between the attachment of the aortic leaflet and the supporting coronary sinuses, and the attachment of the aortic leaflet was displaced downwards into the left ventricle. We speculate that myocardial tone may easily have affected the co-aptation of the aortic leaflets in our case. The dilated aortic root, very asymmetrical leaflet (commissure angle of the non-fused leaflet 135°), and abnormal insertion of the aortic leaflet with the thick septal myocardium of the coronary aortic sinuses might have caused unstable leaflet co-aptation, leading to deformation of the aortic leaflets influenced by the change in myocardial tone and change in the ST junction during the operation (*Figure 5*). In fact, acute aortic regurgitation also occurred with ventricular fibrillation during cooling for deep hypothermic circulatory arrest prior to the operative intervention. Therefore, this massive aortic regurgitation was not only due to surgical handling but also due to the myocardial tone of the root base of the conjoined leaflet.

The fibrous strands may be embryonic remnants of the cusp formation process at an early stage of aortic valve development, which could leave fibrous tissue between the aortic valve and the aortic wall.[3] Fibrous strands have been reported to cause chronic aortic regurgitation by restricting the aortic valve leaflets following aortic dilatation[8] or to cause acute aortic regurgitation when the strands rupture.[1–3] In the present case, we believe that the fibrous strands had a minimal effect on aortic regurgitation because the strands remained intact during the operative procedure and the diameters of the sinotubular junction and the Valsalva sinus changed slightly after graft replacement (from 41 to 35 mm and from 47 to 44 mm, respectively).

There have been reports of aortic valve replacement in patients with downwards displacement of the aortic annulus. The site at which the prosthesis is sutured is variable, being at the site of the attachment of the leaflet in some cases[8,9] and at the normal site of the anatomical ventriculo-arterial junction in other cases.[10,11] Mo *et al.*[11] reported a case in which they anchored a mechanical prosthesis at a normally sited anatomical ventriculo-arterial junction using interrupted mattress sutures instead of suturing the site of attachment of the leaflets. However, their patient developed a para-valvular leak at 6 months after the operation, and redo surgery was required. In our case, we implanted a prosthesis using non-everting mattress sutures at the site

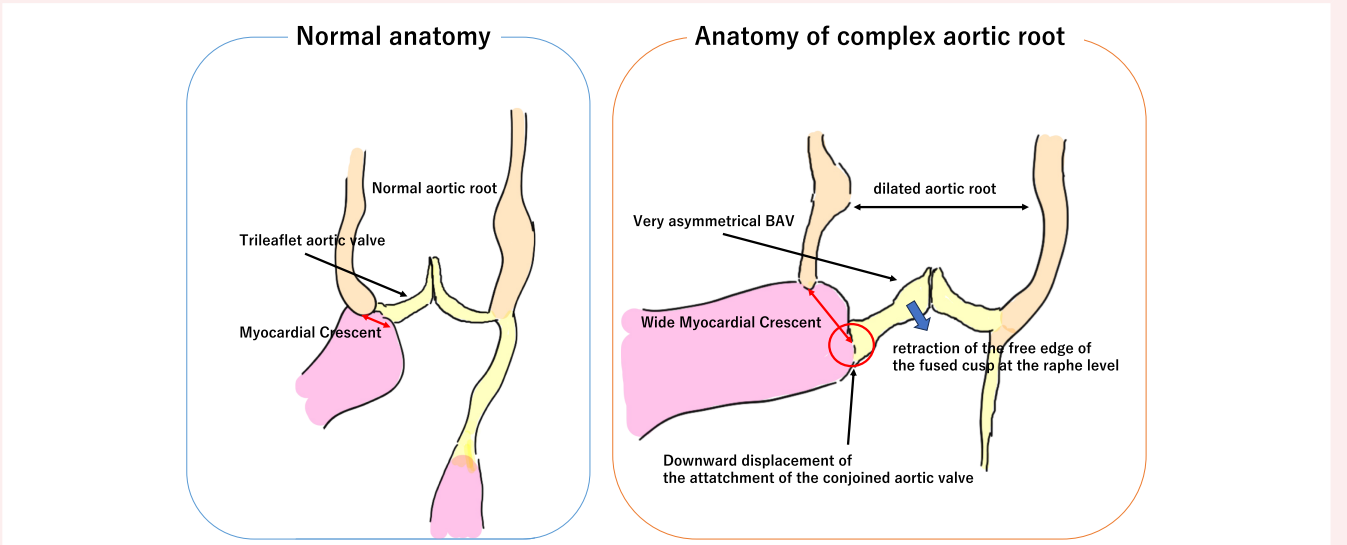


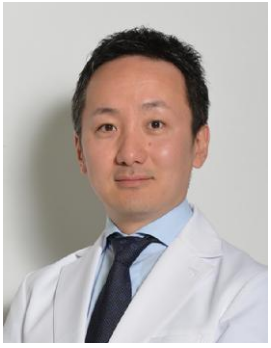
Figure 5 Schematic illustration of the anatomy of normal and complex aortic roots. BAV, bicuspid aortic valve.

of the hinge of the conjoined leaflet. There was no para-valvular leak at the 1-year follow-up visit. However, longer follow-up is needed.

Conclusion

Bicuspid aortic valve with abnormal insertion of the conjoined leaflet and fibrous strands is a rare cardiac abnormality. We experienced a case of acute massive aortic regurgitation intra-operatively that may have been caused by a morphologically abnormal aortic leaflet and root complex.

Lead author biography



Dr Saito graduated from Osaka University School of Medicine in 2003. He worked at Osaka National Hospital and Osaka University, Department of Cardiothoracic Surgery. His specialties include adult congenital surgery, valvular disease, coronary artery disease, and mechanical circulatory support.

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Data availability

No new data were created or analysed in this article.

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