

[ CASE REPORT ]

## Atypical Shone's Complex Diagnosed at 70 Years Old: Presenting with Double-orifice Mitral Valve, Bicuspid Aortic Valve, and Aortic Coarctation

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### Abstract:

Atypical Shone's complex is a rare congenital anomaly involving a left-sided obstructive lesion of two or three cardiovascular levels. A 70-year-old man with dyspnea on exertion was diagnosed with severe aortic stenosis (AS) with a bicuspid valve, complicated by severe aortic coarctation (CoA) and a double-orifice mitral valve. He underwent surgery for AS and CoA in one session. It is important to search for complicated malformations, even in cases of bicuspid aortic valve found in old age.

**Key words:** Shone's complex, forme fruste of Shone's complex, double-orifice mitral valve, bicuspid aortic valve, aortic coarctation

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### Introduction

Shone's complex is a rare congenital anomaly involving a left-sided obstructive lesion of four cardiovascular levels (1). Cases with only two or three levels of stenosis are termed partial forms of the complex or forme frustes. There are few reports describing partial forms, which account for the majority of Shone's complex cases, and cases with double-orifice mitral valve (DOMV), as mitral valve lesions are especially rare (2).

Shone's complex has multiple levels of stenosis, which may lead to underestimation of the severity of each lesion and concealment of symptoms (3). The complex hemodynamics and rarity of the disease make the diagnosis difficult, thereby rendering a delay and difficulty in appropriate treatment (4).

We herein report a patient over 70 years old diagnosed with a partial form of Shone's complex presenting with DOMV.

### Case Report

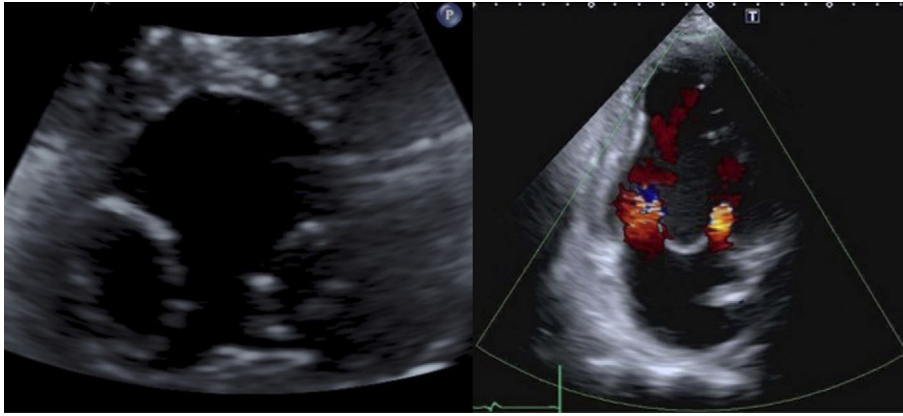
A 70-year-old man with dyspnea on exertion [according to the New York Heart Association (NYHA), Class III] was diagnosed with moderate to severe aortic stenosis and referred to our hospital for surgical treatment. The patient was completely blind due to congenital glaucoma and had severe hearing impairment due to idiopathic sudden deafness. There were no coronary risk factors other than hypertension and no family history of congenital heart disease. The patient was taking irbesartan 100 mg daily, amlodipine 10 mg daily, and carvedilol 2.5 mg twice a day. His blood pressure was 140/70 mmHg, and his heart rate was 60 beats/min.

A physical examination result was significant for a 2/6 systolic murmur at the right upper sternal border. Femoral pulses were weakly palpable. Electrocardiography showed a regular sinus rhythm with first-degree atrioventricular block and left bundle branch block. Laboratory data was notable for an elevated level of brain natriuretic peptide 205.8 pg/mL.

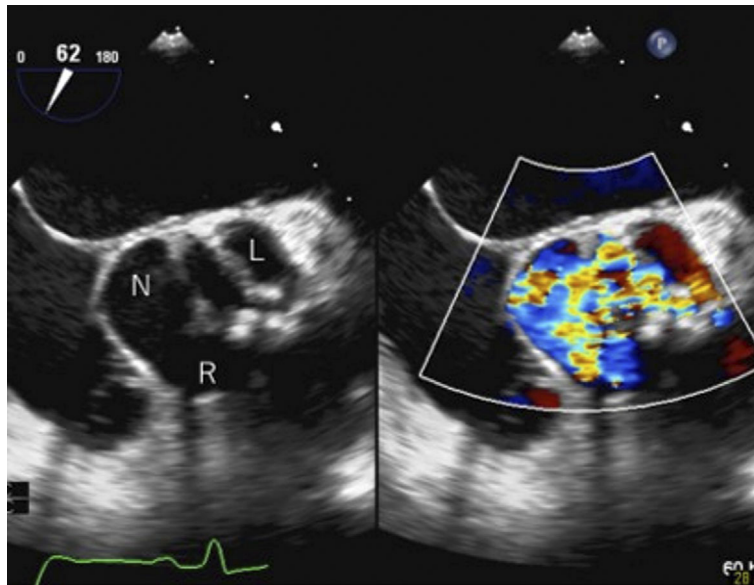
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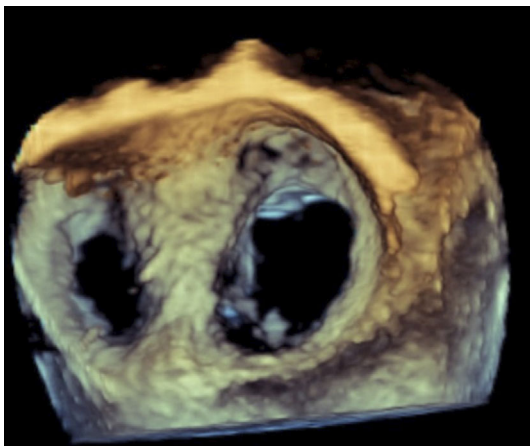
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**Figure 1.** Transesophageal echocardiography shows two separate mitral valve orifices.



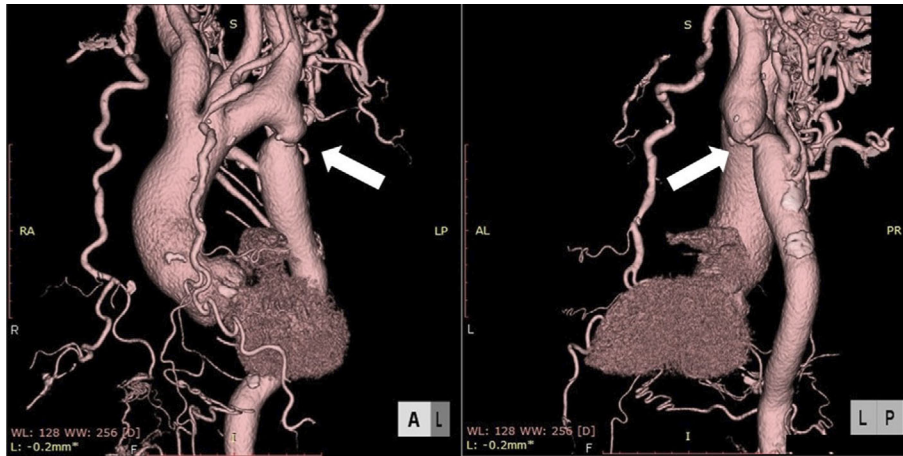
**Figure 2.** Three-dimensional transesophageal echocardiography shows a bicuspid aortic valve (right coronary cusp-noncoronary cusp fusion type).



**Figure 3.** Three-dimensional transesophageal echocardiography shows a complete-bridge-type double-orifice mitral valve with two symmetric orifices.

Transthoracic echocardiography (TTE) revealed a left ventricular ejection fraction of 45% with mild hypertrophy. The

aortic valve (AV) was bicuspid [right coronary cusp (RCC)-noncoronary cusp (NCC) fusion type] with severe aortic stenosis (AS) (a peak AV velocity of 4.0 m/s, a mean AV pressure gradient of 38 mmHg, and an AV area of 0.62 cm<sup>2</sup>) and trivial aortic regurgitation. The images were suggestive of two separate mitral valve orifices (Fig. 1) without stenosis (mean gradient, 1 mmHg) and only trivial mitral regurgitation. The parasternal short-axis view showed two papillary muscles (anterolateral papillary muscle and posteromedial papillary muscle). From this finding, it is clear that parachute mitral valve does not coexist. Three-dimensional transesophageal echocardiography (TEE) showed a bicuspid aortic valve (BAV) (Fig. 2) and a complete-bridge type DOMV with two symmetric orifices (Fig. 3). Computed tomography angiography (CTA) demonstrated severe coarctation of the descending thoracic aorta located 1.9 cm distal to the left subclavian artery and measuring 9.7 mm in diameter at the narrowest point (Fig. 4). Extensive collaterals were present. Coronary angiography showed moderate stenosis in the anterior descending branch of the left coronary artery. The



**Figure 4.** Enhanced computed tomography shows severe coarctation of the descending thoracic aorta. The white arrows indicate the narrowest point.

peak-to-peak pressure gradient across the coarctation was 56 mmHg. His preoperative EuroSCORE II was 6.5%.

The heart team reviewed the case and decided to perform aortic valve replacement and descending aorta replacement in one session. The patient received one-stage repair of AS and aortic coarctation (CoA). First, he underwent bioprosthetic aortic valve replacement via a standard sternotomy with left axillary artery perfusion through an interposition Dacron graft. He was then placed in a right-sided supine position, and descending thoracic aorta replacement was performed through a left thoracotomy, using temporary extra-anatomic axillary-to-femoral bypass. The patient had a good postoperative course and was discharged 15 days after surgery.

## Discussion

There are a few reports on the partial form of Shone's complex presenting with DOMV, BAV, and CoA (4-9). It is well recognized that BAV can coexist with CoA. This case is unique in that the partial Shone's complex with DOMV had 2 levels of severe obstructive lesions (AS + CoA) and was diagnosed at 70 years old.

The complete Shone's complex consisted of parachute mitral valve, supra-annular ring of the left atrium, subaortic stenosis of either the muscular or membranous type, and CoA (1). However, complete Shone's complex is rare, and a majority of Shone's complexes develop as partial forms (10). DOMV is often associated with various other cardiac anomalies, such as atrioventricular septal defect (11), but rarely constitutes Shone's complex. Ma et al. reported that DOMV was found in 2.6% (1/38), severe AS associated with BAV in 31.5% (12/38), and focal CoA in 39.5% (15/38) of the 38 cases of partial forms of Shone's complex included in their review (2).

In Shone's complex, the overlap of multiple obstructive lesions makes it difficult to assess the severity of each obstructive lesion, and mitral stenosis is typically associated

with a poor prognosis among obstructive lesions (12, 13). Isolated severe CoA also has a poor prognosis, causing cerebrovascular and aortic events, and 90% of patients are reported to die before 58 years old (14). However, the partial form of Shone's complex with multiple obstructive lesions, such as severe AS and CoA, without mitral stenosis (as in this case), may not cause clinical events until old age, including subjective symptoms. Even in the elderly, other congenital cardiovascular diseases should not be neglected, especially in patients with severe aortic stenosis associated with bicuspid valve.

**The authors state that they have no Conflict of Interest (COI).**

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