A Bicuspid Aortic Valve with Aberrant Coronary Anatomy

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INTRODUCTION

A bicuspid aortic valve (BAV) is the most common congenital heart defect occurring in approximately 1–2% of individuals.[1,2] It can be associated with a number of other cardiac abnormalities including aortic coarctation, ascending aortic aneurysms, and coronary artery anomalies.[3,2] BAV can also be complicated by aortic stenosis and/or aortic regurgitation and infective endocarditis. Patients with bicuspid aortic valve stenosis usually present in their 50s and 60s, one to two decades earlier than older patients with degenerative calcific aortic stenosis, whose disease usually manifests in their 8th and 9th decades.[3] This brief report will review a patient with a BAV, ascending aortic aneurysm, and aberrant coronary anatomy who underwent surgical aortic valve replacement and ascending aortic aneurysm repair. Our discussion will focus on the natural history of BAV, image modalities that aid in its diagnosis, and associated non-valvular anomalies.

CASE PRESENTATION

A 68-year-old female was referred to a cardiologist after presenting to her primary care physician with shortness of breath, dizziness on exertion, and a loud systolic ejection murmur on exam. Transthoracic echocardiogram (TTE) was notable for a severely stenotic BAV and an ascending aortic aneurysm (49 mm). Pre-operative coronary angiography revealed no obstructive coronary disease. However, anomalous coronary anatomy was identified with the left main coronary artery originating from the ostium of the right coronary artery (**Image 1**). Subsequent CT imaging further characterized the course of the aberrant left main coronary artery which traveled anterior to the right ventricular outflow tract (**Image 2**), and confirmed the presence of the ascending aortic aneurysm. A perioperative transesophageal echocardiography (TEE) confirmed BAV stenosis (**Image 3**) and the ascending aortic aneurysm. The BAV was replaced with a 25 mm tissue prosthesis, and a 36mm gelweave aortic graft replacement was inserted from the sino-tubular junction to the mid-ascending aorta. The patient's hospital course was largely unremarkable and she was discharged home on postoperative day 11.

Image 1. Coronary Angiography demonstrating the right coronary artery (RCA) and left main coronary artery (LMCA) originating from a single right coronary ostia.

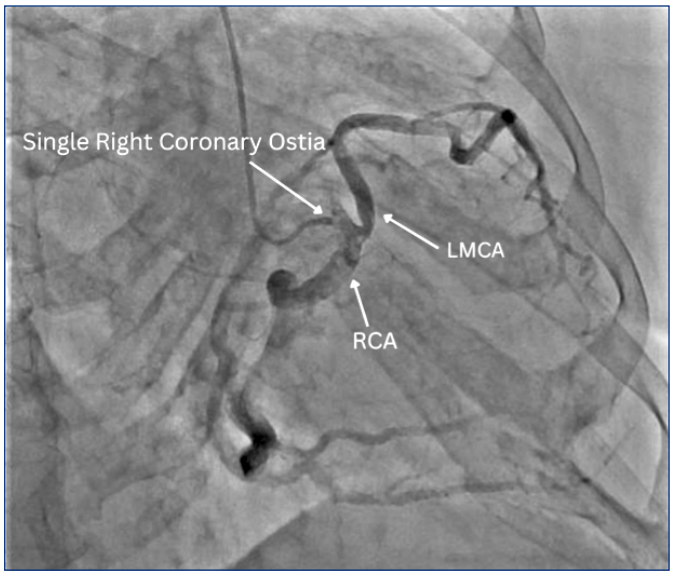


Image 2. CT Angiography demonstrating RCA and LMCA originating from a single right coronary ostia. Additional CT cuts showed the left main coronary artery traveling anterior to the right ventricular outflow tract before bifurcating into the left anterior descending and circumflex coronary arteries.

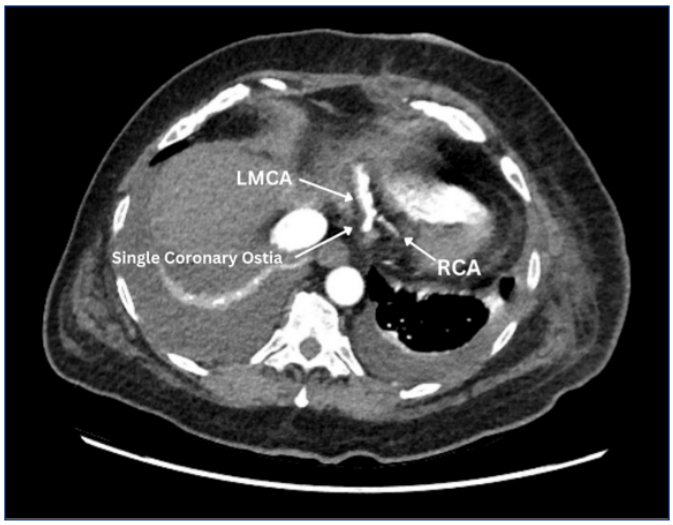


Image 3. Transesophageal Echocardiographic view of the bicuspid aortic valve in short axis (SAX). Here you can see the single coaptation line separating the anterior cusp (A) and posterior cusp (B).

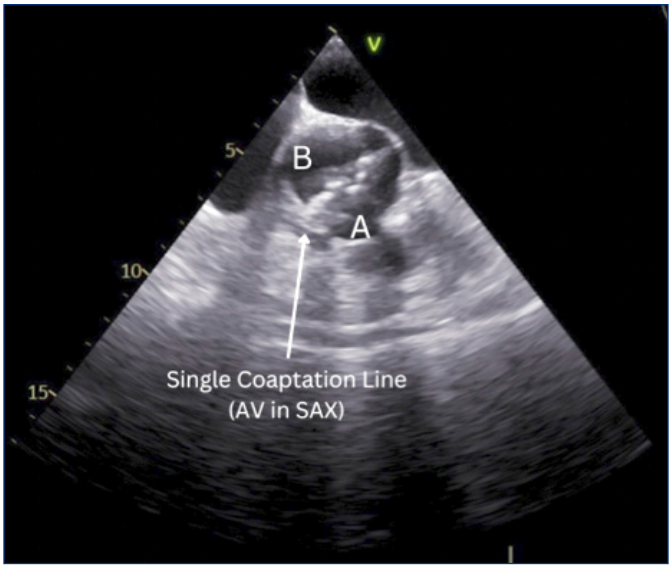
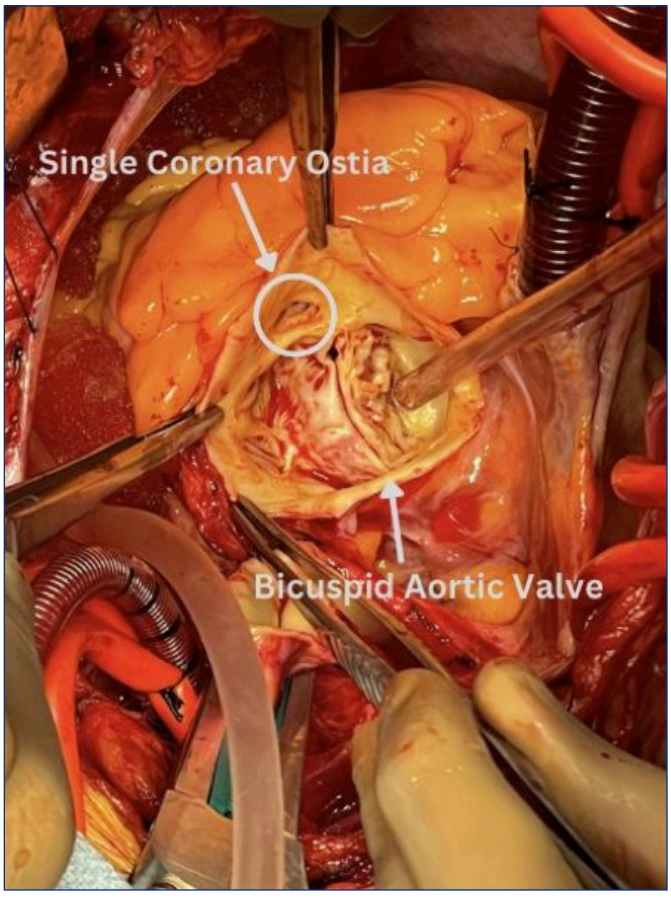


Image 4. Surgical view of bicuspid aortic valve with RCA and LMCA originating from a single right coronary ostia.



DISCUSSION

A congenitally BAV is more prone to degeneration resulting in aortic stenosis (AS) and/or aortic regurgitation.[2,4] As the BAV becomes more stenotic, dyspnea on exertion, left ventricular hypertrophy, pulmonary edema, atrial fibrillation, and syncope become more common.[5]

TTE is the primary diagnostic imaging modality used to diagnose AV abnormalities.[6] The TTE exam must characterize the leaflet morphology and degree of hemodynamic compromise. Severe AS is defined as a valve orifice area less than 1.0 cm^2, a mean gradient greater than 40 mmHg, or jet velocity greater than 4 m/s.[7] The short axis view of a bicuspid AV shows a clear fusion of the two AV leaflets with a single line of coaptation (**Image 3,4**). Once identified, a BAV warrants close surveillance as it can rapidly progress to severe stenosis or regurgitation. The two-year mortality rate for severe AS is roughly 50%.[8]

As seen in this patient, other non-valvular anomalies can be associated with BAV disease. Ascending aortic aneurysms may be present in up to 50% of BAV patients.[9] Aberrant coronary anatomy is uncommon,[4,10] and more likely to originate from the anterior aortic cusp.[2] Most congenital coronary abnormalities are discovered incidentally on coronary angiography or CTA imaging.[11] The identification of aberrant coronary anatomy has important implications for management of BAV patients both during open surgical and percutaneous interventions.

Congenital BAV may have a genetic component. BAV disease is believed to be autosomal dominant with variable penetrance.[9] The ACC/AHA Thoracic Aortic Disease Guidelines from 2010 state that first-degree relatives of patients with BAV or premature onset of thoracic aortic disease should be screened for BAV and associated aortic abnormalities.[12]

In conclusion, BAV may present with a systolic ejection murmur in a younger patient with echocardiographic evidence of a bi-leaflet valve. Once a bicuspid valve has been identified, it is important to monitor the patient for disease progression and associated cardiovascular conditions. If surgical or percutaneous intervention is required, knowledge of the patient's coronary anatomy is key to providing safe periprocedural care.

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