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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Bicuspid Aortic Valve and Ascending Aortic Aneurysm in a Twin Pregnancy

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ABSTRACT

Bicuspid aortic valve with ascending aortic aneurysm is a common condition encountered in pregnancy. There are limited data on how to manage these patients. To our knowledge, we report the only case of a bicuspid aortic valve and aortic aneurysm with twin gestations. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:96–100) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 34-year-old G1P0 Caucasian woman with a bicuspid aortic valve (BAV) and a dilated ascending aorta of 4.9 cm presented to a women's heart health clinic at 12 weeks of a twin gestation for evaluation and management of her high-risk pregnancy.

Two months prior, at an outside facility, a positive human chorionic gonadotropin urine pregnancy test led to an ultrasound, which revealed an ovarian mass concerning for cyst versus malignancy. She was too early in the pregnancy for the ultrasound to detect a fetal heartbeat. She then underwent a right ovarian cystectomy for the 9.2-cm right ovarian cyst with pathology demonstrating a conventional type borderline serous tumor. She was also in the process of infertility workup because she had been trying to become pregnant for 12 months prior. It was then recommended that she undergo an oophorectomy; however, when her human chorionic gonadotropin remained positive, she discovered she was pregnant and decided to defer until after pregnancy.

During the workup for her ovarian cystectomy, she underwent an echocardiogram, which revealed a BAV and an ascending aortic aneurysm of 4.9 cm (**Figure 1**). She reported she had a known BAV since the age of 7 years with periodic echocardiograms and clinical monitoring until approximately age 18, at which time she was reportedly told no intervention was required and the BAV could be monitored in the future. She was active all throughout her childhood and remained so at the time of the initial consultation. From a cardiac standpoint, she was asymptomatic and denied chest pain, back pain, palpitations, exertional dyspnea, orthopnea, postural nocturnal dyspnea, dizziness, and syncope (New York Heart Association functional class I).

LEARNING OBJECTIVES

- Monitoring of a bicuspid aortic valve with a dilated aorta during a twin pregnancy in a nonsyndromic patient is discussed.
- Anesthetic and delivery management in a bicuspid aortic valve and a dilated aorta is examined.

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Informed consent was obtained for this case.

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PAST MEDICAL HISTORY

She also had documented elevated blood pressure readings at clinic visits without a diagnosis of hypertension (but was diagnosed with chronic hypertension), obesity (BMI 35 kg/m^2), and gestational diabetes in her first trimester. She had no family history of valvular heart disease or aortopathies. Her examination demonstrated no syndromic features.

INVESTIGATIONS

She was offered elective termination versus carrying a high-risk pregnancy and pre-term delivery. If she elected to carry, the timing and mode of delivery were discussed, as was anesthesia management. The risks and benefits based on limited data extrapolation were discussed with her, and it was elected by the patient to continue.

MANAGEMENT

She was counseled on the risk of pre-term delivery, fetal demise, cardiovascular death, and morbidity related to an aortic dissection. A multidisciplinary maternal-fetal medicine, anesthesia, and cardiac team deemed her pregnancy high risk for cardiovascular morbidity and/or mortality. She and her family were counseled on these risks, expressed understanding, and elected to proceed with the pregnancy. To avoid radiation in pregnancy, magnetic resonance imaging of the aorta (**Figure 2**) was performed to accurately assess aortic dimensions. She met with the cardiothoracic surgery team as a precautionary measure should earlier intervention be required. She was started on a beta-blocker at the initial consultation, given exercise restrictions of avoiding heavy lifting, and monitored with transthoracic echocardiograms every 4 weeks until delivery (**Table 1**). Throughout her pregnancy, she remained clinically asymptomatic, normotensive, and New York Heart Association functional class I.

DISCUSSION

To our knowledge, this is the first published case report of a successful pregnancy in a patient with BAV and a dilated aorta with twin gestation. BAV is the most common congenital cardiac abnormality with coexistent dilatation of the ascending aorta (1,2). In addition to the complications of aortic stenosis or regurgitation, there are potential risks of aortic enlargement or dissection in pregnancy. Prior reports suggest that these risks may be slightly higher in the presence of aortic regurgitation. The risk of aortic dissection is thought to be low at 3.1% per 10,000 patient-years; however, this represents an 8-fold increase over the risk to the general population (3,4).

ABBREVIATIONS AND ACRONYMS

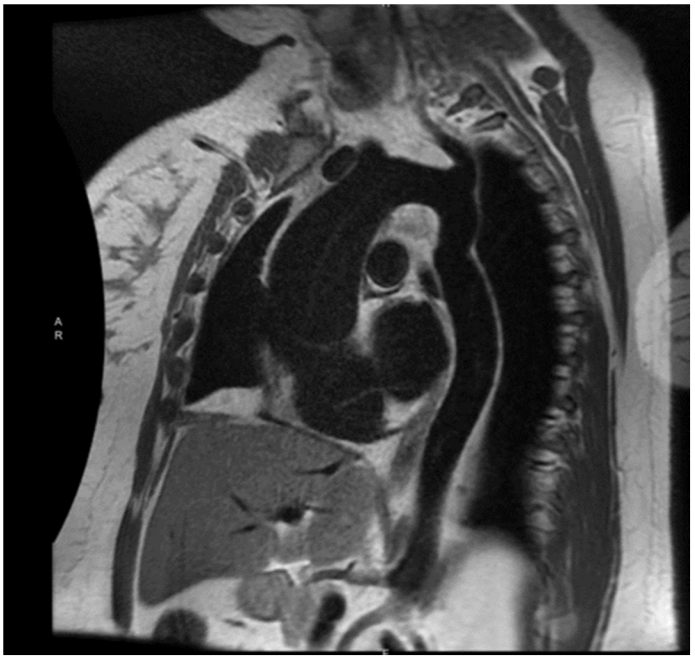
BAV = bicuspid aortic valve

BAV is considered to have a strong genetic component with an inheritance pattern that may be autosomal dominant with variable penetrance or sporadic. It can be isolated or part of other heritable congenital syndromes and aortopathies (1,5). The 2018 American College of Cardiology/American Heart Association guidelines for the management of adults with congenital heart disease and the American Association of Thoracic Surgery consensus guidelines recommend fetal ultrasound screening when the mother has BAV (1,5,6).

Aortic aneurysms are diagnosed when the diameter is >2 SDs above the mean or referenced aortic segment or using *z*-scores. However, current guidelines still use absolute values for intervention recommendations (7). Lastly, much of the data on aneurysms in pregnancy come from extrapolation from Marfan syndrome patients (2,8).

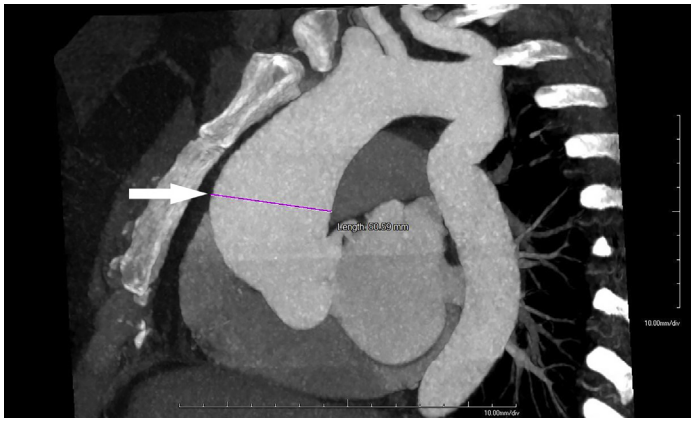
In pregnancy, the increases in heart rate, stroke volume, and blood pressure, along with hormonal shifts, result in increased stress and changes in the

FIGURE 1 MRI of Thoracic Aorta



Magnetic resonance imaging: the oblique sagittal view of the thoracic aorta showing an ascending aortic aneurysm.

FIGURE 2 CTA Thoracic Aorta



Computed tomography angiography: the oblique sagittal maximum intensity projection of the thoracic aorta showing the ascending aortic aneurysm (postpartum).

TABLE 1 Aortic Measurements

Gestational Week	Imaging Modality	Proximal Ascending Aorta	Mean AV Gradient
4	TTE	4.9 cm	14 mm Hg
12	TTE	4.9 cm	12 mm Hg
16	TTE	4.9 cm	17 mm Hg
18	MRI	4.9 × 4.8 cm	NA
22	TTE	5.0 cm	20 mm Hg
26	TTE	5.0 cm	22 mm Hg
30	TTE	4.9 cm	21 mm Hg
6 weeks postpartum	TTE	4.9 cm	17 mm Hg
10 months postpartum	CT	5.1 cm	NA

CT = computed tomography; MRI = magnetic resonance imaging; NA = not applicable; TTE = transthoracic echocardiography.

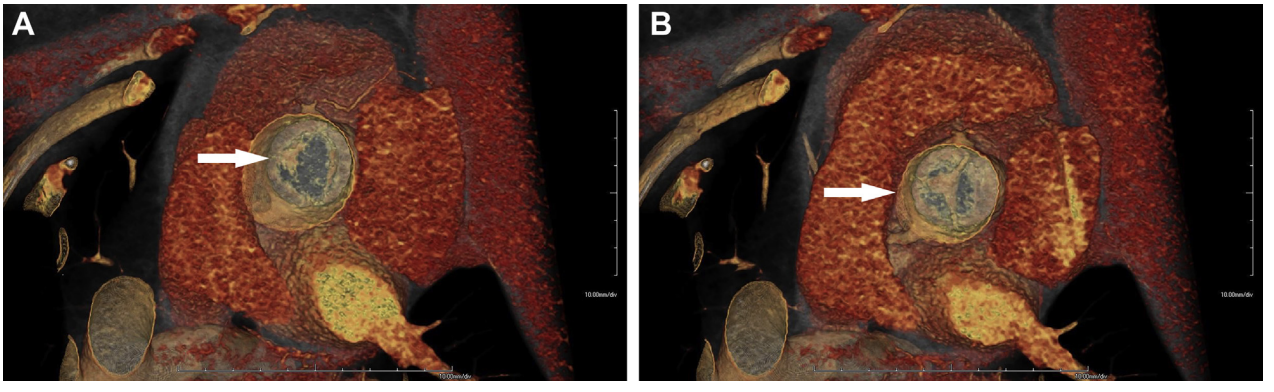
aortic media (1,3,9,10). The increased risk of dissection is greatest in the third trimester or postpartum. Recent studies using monthly to every 6 weeks serial echocardiographic monitoring during pregnancy indicate a 0% maternal mortality.

The American Association of Thoracic Surgery guidelines recommend that in the case of an aortic root diameter >4.5 cm, pregnancy should be discouraged (5). The 2018 European Society of Cardiology guidelines for the management of cardiovascular diseases during pregnancy recommend the avoidance of pregnancy when the aorta diameter is >5.0 cm (6). Because of the increased risk of dissection in the case of BAV, some recommend a lower threshold for prophylactic repair before planned pregnancy of 4.0 cm, especially in the context of Marfan syndrome or other collagen vascular disorders. The European Society of Cardiology guidelines recommend considering surgery in this case with an aortic diameter >5.0 cm (6).

The current clinical standard of management would suggest serial monitoring based on aortic dimension and growth during pregnancy. The consideration of >5.0 mm dilation through the pregnancy should trigger a personalized plan for intervention with elective termination, delivery once fetal lung maturity is established, or surgical repair early in the postpartum period (6).

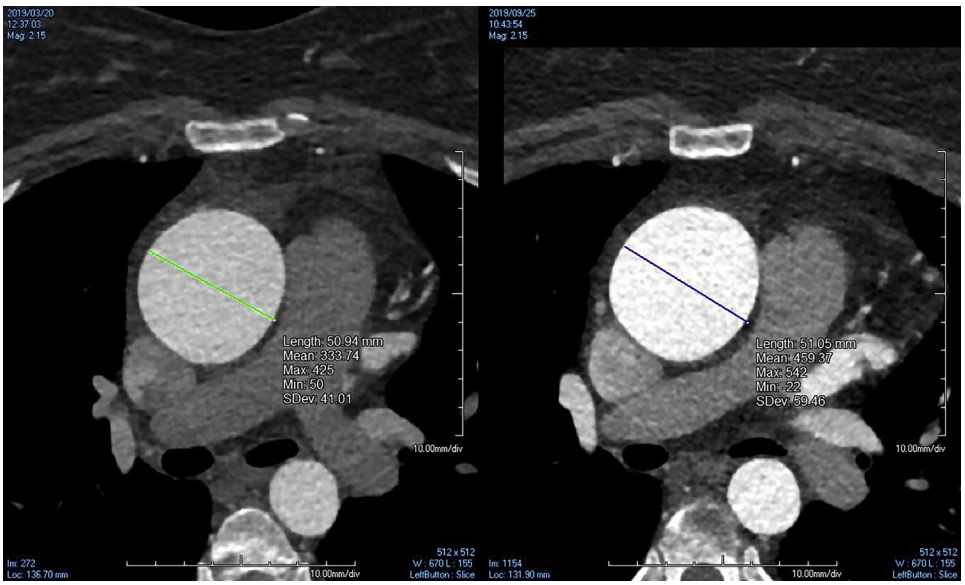
For patients with isolated BAV and a dilated aorta ≥5.0 cm, consideration for prophylactic surgery should be pursued. Prolonged pushing is not advised

FIGURE 3 CT Volume-Rendered Images of Aortic Valve



(A) Volume-rendered images of the aortic valve viewed from above in systole shows an incomplete valve opening due to fusion of the left and right aortic cusps by a raphe (Sievers type I, R-L). **(B)** Volume-rendered images of the aortic valve viewed from above in diastole.

FIGURE 4 CTA Post-Partum



Computed tomographic images **(left)** 2 months' and **(right)** 10 months' postpartum showing the size of the ascending aortic aneurysm at 51 mm.

in those with an aorta >5.0 cm or growth >5.0 mm in pregnancy (1,6). Anesthetic management based on European guidelines suggests if aortic diameters are 4.0 to 4.5 cm, vaginal delivery with regional anesthesia and invasive arterial pressure monitoring is needed. A cesarean section should be considered in aortas larger than this (3,4,6).

Obtaining an accurate and consistent aortic measurement can be challenging whether based on echocardiography or magnetic resonance imaging because techniques for measuring differ slightly. Medical management includes beta-blockers in pregnancy rather than angiotensin-converting enzyme inhibitors or angiotensin receptor blockers, which are contraindicated in pregnancy. They should be used to prevent hypertension with systolic pressure maintained <140 mm Hg and diastolic pressure <90 mm Hg. Some studies advocate a decrease in heart rate by 20 beats/min. Some early reports indicated atenolol could cause fetal growth restriction; therefore, it should be avoided (3,4).

Delivery planning should be done in a multidisciplinary and personalized fashion to include a high-risk obstetrician, anesthesiologist, and cardiac team. It should integrate parity status, gestation status, patient age, family history, and concomitant collagen vascular disorders and syndromes, in addition to the aortic dimensions, aortic valve function, and aortic growth rate.

FOLLOW-UP

She underwent elective cesarean section at 34 weeks' gestation and delivered 2 healthy baby girls. Her aorta continued to dilate postpartum (**Figures 3** and **4**); therefore, a planned elective repair was scheduled after 11 months.

CONCLUSIONS

Pregnancy in a patient with BAV with a dilated aorta has traditionally been associated with high risk; however, when not associated with syndromic aortopathies and with careful monitoring and multidisciplinary management, it can be carried out successfully.

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KEY WORDS aorta, bicuspid aortic valve, pregnancy