JACC: CASE REPORTS VOL. 4, NO. 12, 2022

CASE REPORT

INTERMEDIATE

CLINICAL CASE

A 16-Year Natural History of a Congenital Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrosa

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ABSTRACT

Pseudoaneurysm of the mitral aortic intervalvular fibrosa is a rare condition most often reported as sequela of endocarditis and surgical trauma, with congenital cases being very uncommon. This case describes a congenital pseudoaneurysm of the mitral aortic intervalvular fibrosa and stable 16-year natural history. (**Level of Difficulty: Intermediate.**)
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HISTORY OF PRESENTATION

A 16-year-old otherwise healthy male patient who was lost to follow-up since 3 years of age presented to reestablish care for a mitral valve (MV) disorder. He was found to have an "aneurysm of the MV" and bicuspid aortic valve (AOV) without stenosis or insufficiency at 2 months of age after being referred for a nonspecific intracardiac finding on a fetal echocardiogram. He remains asymptomatic and participates in competitive track and golf with a negative review of systems. His physical exam revealed bradycardia with a rate of 43 beats/min, otherwise normal vital signs, and a nonspecific 2/6 midsystolic ejection murmur heard equally at the right and left upper sternal borders without radiation.

His initial evaluation consisted of a transthoracic echocardiogram followed by a transesophageal echocardiogram (**Figures 1A, 2A, and 3A,** Video 1). He was started on aspirin after those evaluations but was discontinued due to a rash. He was followed until 3 years of age on no medications before being lost to follow-up.

LEARNING OBJECTIVES

- To develop a differential diagnosis for aneurysmal lesions of the MV.
- To understand the how the natural history of PMAIF differs based on etiology.

PAST MEDICAL HISTORY

He was born full term without complications and has no history of hospitalizations, surgeries, serious illness, or chronic medical problems except the previously mentioned cardiac history.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included a congenital cystic defect of the MV, an aortic pseudoaneurysm entering the left atrium, and a pseudoaneurysm of the mitral-aortic intervalvular fibrosa (PMAIF).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received February 15, 2022; revised manuscript received April 25, 2022, accepted May 3, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.05.002

INVESTIGATIONS

Transthoracic echocardiography showed a sac originating in the mitral-aortic intervalvular fibrosa (**Figures 1B, 2B, and 3B**). There is a 1.3-cm initially spherical-appearing aneurysm seated on the fibrosa of the anterior leaflet of the MV that communicates with to-fro flow with the left ventricular outflow tract through a 0.4-cm orifice (**Figures 1B and 2B**,Videos 2 and 3). Tissue from the pseudoaneurysm extends along the left atrial reflection of the aortic root toward and along the atrial septum (**Figure 3B,** Video 4). The right nonfused bicuspid AOV continued to be free of insufficiency, prolapse, or stenosis. The MV has trace regurgitation, but has no prolapse or stenosis. The remainder of the cardiac anatomy is normal, as are systolic and diastolic function. There is no suggestion of thrombus formation or endocarditis.

When compared with both the initial and most recent prior echocardiogram performed at 2 months of age and 3 years of age, the pseudoaneurysm is grossly unchanged in appearance (**Figures 1 to 3**). There has been an increase in absolute size that appears consistent with somatic growth. The proportions of the pseudoaneurysm relative to other cardiac structures appears unchanged.

A 12-lead electrocardiogram demonstrated a prolonged PR interval of 236 ms in the setting of a marked sinus bradycardia (35 beats/min). A 48-hour Holter monitor was obtained because demonstrated sinus bradycardia with an average rate of 44 beats/min, maximum of 135 beats/min, and minimum of 22 beats/min while sleeping with rare premature atrial contractions (<0.1%). There was neither a second- or third-degree block nor other arrhythmias. He did not have an electrocardiogram during his initial assessment.

MANAGEMENT

The stable appearance of the aneurysm for over a decade, normal ventricular and valve function, and the lack of symptoms allowed for observation of this lesion. We recommended against heavy weightlifting, but the patient was otherwise allowed to continue participating in sports and takes no medications.

We had concerns that any surgical solution could potentially damage the atrioventricular node, placing him at risk for advanced heart block in addition to other risks of surgery, most specifically AOV and MV dysfunction. Aspirin was considered as an antithrombotic prophylaxis, but because of his history of a possible reaction, no evidence of thrombus formation for over a decade while off therapy, and his active lifestyle, we elected against aspirin or other antiplatelet agent. Spontaneous bacterial endocarditis prophylaxis was recommended.

DISCUSSION

PMAIF is most commonly seen as a complication of endocarditis and surgical trauma in patients with a bicuspid AOV.[1,2] Only rare congenital cases have been reported.[3-5] Cases of acquired PMAIF usually undergo operative repair because the lesion causes valve

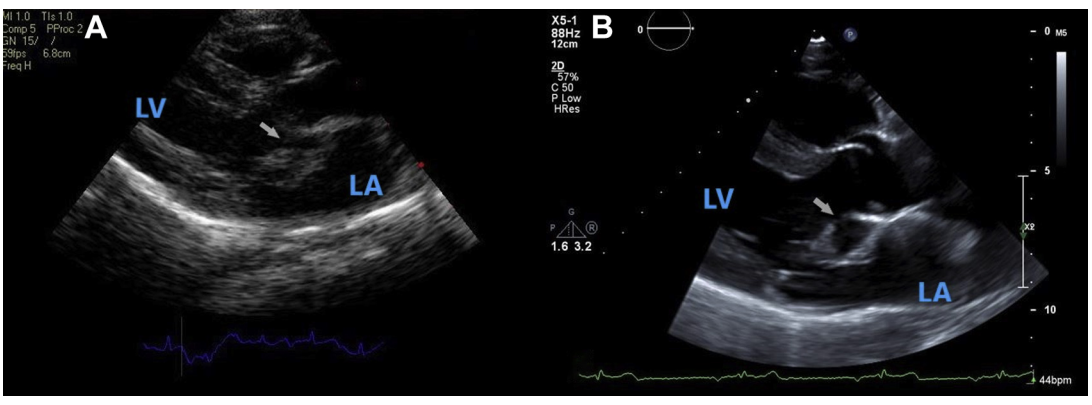
ABBREVIATIONS AND ACRONYMS

AOV = aortic valve

PMAIF = pseudoaneurysm of the mitral-aortic intervalvular fibrosa

MV = mitral valve

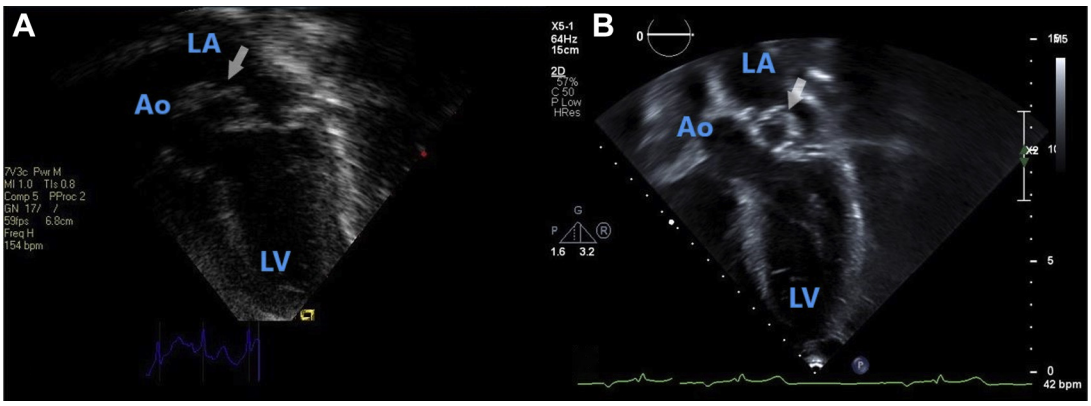
FIGURE 1 Comparison of Long Axis



Comparison of parasternal long-axis view demonstrating the aortic valve and mitral valve with the pseudoaneurysm **(arrow)** seen within the left atrium (LA) posterior to the anterior leaflet of the mitral valve. **(A)** Two months of age. **(B)** Sixteen years of age. LV = left ventricle.

JACC: CASE REPORTS, VOL. 4, NO. 12, 2022
JUNE 15, 2022:706–709

FIGURE 2 Comparison of Apical 4-Chamber



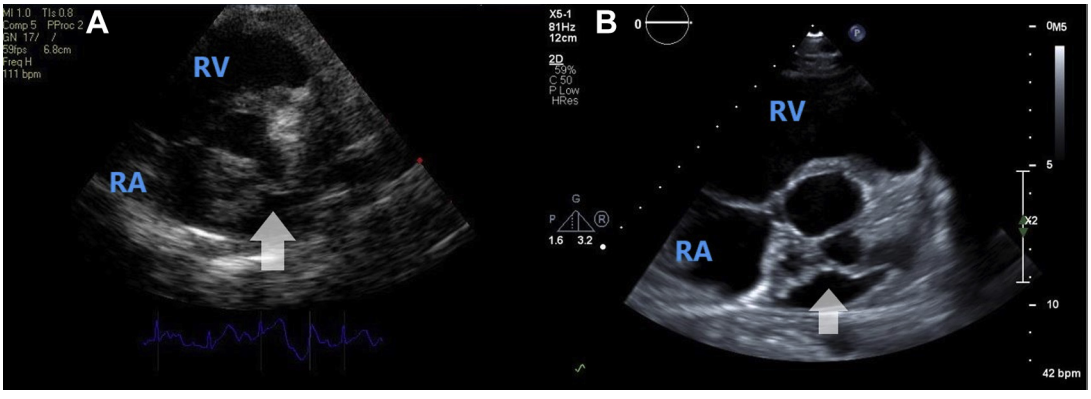
Comparison of an apical 4-chamber view demonstrating the pseudoaneurysm **(arrow)** seen within the LA. **(A)** Two months of age. **(B)** Sixteen years of age. Ao = aorta; other abbreviations as in Figure 1.

dysfunction and is at risk of expansion, rupture, and thrombus formation.[1]

In contrast, congenital cases may not have the same risk as PMIAF from acquired causes. Del Pasqua et al[3] reported a stable 5-year natural history of a congenital PMAIF without any significant change in appearance. Similarly, Harrington et al[4] reported a stable 1-year natural history of a prenatally diagnosed PMIAF.

Caution should be used when describing the case as congenital or acquired, which may imply a different natural history. Lizano Santamaria et al[6] reported a case of a PMIAF in a 4-year-old related to culture-negative AOV endocarditis, and He et al[7] reported a case in a 5-year-old that developed several years after a ventricular septal defect repair. In both cases, the term *congenital* was applied despite the most likely cause being endocarditis and cardiac surgery, respectively. In both cases, there were sequelae that supported surgical intervention. We believe the term *congenital* should only be applied to those without a history of intracardiac surgery, infective endocarditis, or other process that could mechanically disrupt the fibrosa.

FIGURE 3 Comparison of Short Axis



Comparison of parasternal short-axis view showing the spherical pseudoaneurysm **(arrow)** toward the left and irregular tissue adjacent that extends along the underside of the atrial reflection of the aortic root. **(A)** Two months of age. **(B)** Sixteen years of age. RA = right atrium; RV = right ventricle.

Our case is the longest reported natural history of PMAIF, with a similar stable course described by Harrington et al[4] and Del Pasqua et al.[3] It is reasonable to conclude that the natural history of congenital PMAIF can be stable and does not necessitate immediate surgery in the absence of symptoms or other concerning findings.

We would consider a surgical option if there was valve dysfunction secondary to the aneurysm, thrombus formation, increasing size, pulmonary venous or mitral inflow obstruction, or concerns about compression of any part of the left coronary system. Given the limited information on the natural history, offering surgery in the asymptomatic patient could be considered to reduce the hypothetical risk of endocarditis, thrombus, and rupture. In this case, the risk for surgical heart block is not insignificant.

We suspect that the first-degree heart block in our case is from aneurysmal damage to the atrioventricular node. If our patient developed advanced heart block, surgery would be considered. We propose the sinus bradycardia is from athletic conditioning and high vagal tone unrelated to the PMAIF.

FOLLOW-UP

Serial echocardiograms, electrocardiograms, and Holter monitors are planned every 2 years.

CONCLUSIONS

Congenital PMAIF is a rare finding that in the absence of symptoms can be conservatively managed without surgery. We believe the term *congenital* should be applied to those patients without a history of intracardiac procedure or suspicion of endocarditis. Aspirin can be considered for thrombus prophylaxis, but the benefit of this therapy is unclear.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS bicuspid aortic valve, congenital heart disease, echocardiography, mitral valve

APPENDIX For supplemental videos, please see the online version of this paper.