cambridge.org/cty

Images in Congenital Cardiac Disease

Cite this article: Xiao R, Niaz T, and Doan TT (2023) Perforated posterior mitral valve leaflet in mitral annular disjunction and infective endocarditis. *Cardiology in the Young* **33**: 1184–1185. doi: 10.1017/S1047951123002615

Received: 15 March 2023 Revised: 20 May 2023 Accepted: 28 June 2023 First published online: 24 July 2023

Keywords:

Mitral annular disjunction; infective endocarditis; mitral valve perforation; 3-D echocardiography

Corresponding author: T. T. Doan; Email: tam.doan@bcm.edu

Perforated posterior mitral valve leaflet in mitral annular disjunction and infective endocarditis

Ran Xiao¹, Talha Niaz² and Tam T. Doan¹

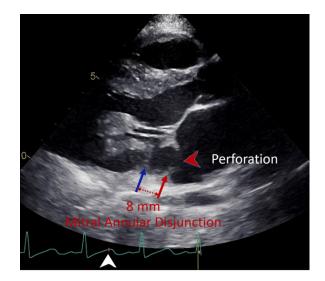
¹Department of Pediatrics, The Lillie Frank Abercrombie Division of Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, TX, USA and ²Division of Pediatric Cardiology, Mayo Clinic, Rochester, MN, USA

Abstract

We herein presented a 17-year-old female with history of mild mitral valve prolapse who was admitted for methicillin-sensitive *Staphylococcus aureus* endocarditis and diagnosed with mitral annular disjunction and perforated posterior mitral valve leaflet on two-dimensional and three-dimensional echocardiography (P_1 - P_2). A perforation in the posterior leaflet was confirmed and repaired during surgical intervention. This is a rare presentation of leaflet perforation in the area of mitral annular disjunction.

Mitral annular disjunction (MAD) is a fibrous separation between the attachment of the mitral valve posterior leaflet hinge and basal left ventricular myocardium.¹ Mitral annular disjunction is often seen with mitral regurgitation in myxomatous mitral valve disease and mitral valve prolapse (upwards of 50.8 and 32.6%, respectively).² It has been reported in patients with ventricular tachyarrhythmia and sudden cardiac arrest.³ Inferolateral mitral annular disjunction can be readily identified on an echocardiographic parasternal long-axis view.²

A 17-year-old female with previously diagnosed mitral valve prolapse with moderate mitral regurgitation who presented with 5 days of fever, petechiae and *Staphylococcus aureus* bacteremia. While being treated for infective endocarditis, she acutely decompensated with respiratory distress and hypotension. Emergent echocardiography showed new concern for perforation of the posterior leaflet and mitral annular disjunction, which was not previously recognised (Fig. 1, Video 1). A three-dimensional echocardiogram demonstrated a 15-mm X 8-mm perforation in P₁ and P₂ (Fig. 2a–b, Video 2). Given haemodynamic instability in the setting of *Staphylococcus aureus* endocarditis and the new finding of mitral valve perforation,⁴ surgical intervention was indicated. The defect measured 12–15 mm in diameter in P₁–P₂ scallop of the mitral valve consistent with findings from the 2-dimensional and 3-dimensional echocardiogram (Fig 2c–d). The damaged posterior leaflet was excised, and the defect was repaired using an autologous pericardial patch. Pathology examination of the excised leaflet



© The Author(s), 2023. Published by Cambridge University Press.



Figure 1. Parasternal long-axis view of mitral annular disjunction and perforation of the posterior leaflet near the disjunction. Mitral annular disjunction is defined as the fibrous separation between the attachment of posterior leaflet (red arrow) and basal left ventricular myocardium (blue arrow) measured at end systole (white arrowhead). A perforation in the posterior leaflet is suspected (red arrowhead).



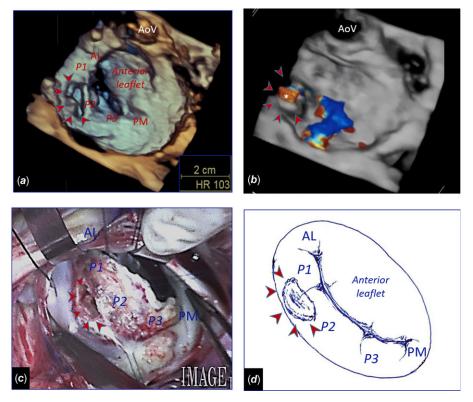


Figure 2. Three-dimensional echocardiographic image and surgical inspection of the perforated posterior mitral valve leaflet. (a, b) Three-dimensional echocardiographic images with and without colour mapping of a large perforation in the P_1-P_2 scallops (arrowheads). (c, d) Surgical view and diagram of the mitral valve from a left atrial view demonstrating a 15 X 8 mm perforation (arrowheads) between P1 and P2 scallop of the posterior leaflet. AL = anterolateral commissure, AoV = aortic valve, P1, P2, P3 = scallops of the posterior mitral valve leaflet, PM = posteromedial commissure.

showed a fibropurulent plaque with abundant fibrin and neutrophils consistent with infective endocarditis. Tissue culture resulted methicillin-sensitive *Staphylococcus aureus*, matching her blood cultures. The patient had an uneventful postoperative recovery, completed the course of antibiotics, and had mild mitral regurgitation at discharge. She has been followed up over the last two years with no evidence of ventricular arrhythmia. However, she unfortunately had recurrent regurgitation at the perforation possibly due to active inflammation at the time of her initial surgery. She is scheduled for repair in the coming months at the time of writing this manuscript.

Mitral annular disjunction is associated with an area of fibrous thin wall exposing to the high-pressured left ventricle.¹ This is a rare case of mitral valve perforation near the area of disjunction. Awareness of mitral annular disjunction allowed for the diagnosis to be recognised, although much remains to be learned about mitral annular disjunction in the paediatric population. Three-dimensional echocardiography provided helpful information to explain for clinical deterioration and surgical planning, which aided in the successful management of this patient.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123002615.

Acknowledgements. The authors appreciate Dr. Michiaki Imamura for the image of the mitral valve from surgical inspection and Ms. Kaitlin Doan for her assistance in editing the videos.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interests. None.

References

- Toh H, Mori S, Izawa Y, et al. Prevalence and extent of mitral annular disjunction in structurally normal hearts: comprehensive 3D analysis using cardiac computed tomography. Eur Heart J Cardiovasc Imaging 2021; 22: 614–622. DOI: 10.1093/ehjci/jeab022.
- Bennett S, Thamman R, Griffiths T, et al. Mitral annular disjunction: a systematic review of the literature. Echocardiography 2019; 36: 1549–1558. DOI: 10.1111/ECHO.14437.
- Dejgaard LA, Skjølsvik ET, Lie Ø.H, et al. The mitral annulus disjunction arrhythmic syndrome. J Am Coll Cardiol 2018; 72: 1600–1609. DOI: 10. 1016/J.JACC.2018.07.070.
- Otto CM, Nishimura RA, Bonow RO, et al. ACC/AHA guideline for the management of patients with valvular heart disease: a report of the American college of cardiology/American heart association joint committee on clinical practice guidelines. J Am Coll Cardiol 2020; 77: e25–e197. DOI: 10.1016/j. jacc.2020.11.018.