

Images in
Cardiovascular Medicine



Surprising Course of a Pregnant Patient With Mosaic Turner Syndrome

David S. Majdalany , MD¹, Hong S. Lee , MD¹, Timothy Barry , MB, BCh, BAO¹, Katherine Singh , MD², Jeff Chapa , MD³, and Gosta B. Pettersson , MD, PhD⁴

¹Department of Cardiovascular Medicine, Mayo Clinic, Scottsdale, AZ, USA

²Department of Obstetrics, Gynecology and Women's Health Institute, Cleveland Clinic, Cleveland, OH, USA

³Department of Obstetrics and Gynecology, Cleveland Clinic, Cleveland, OH, USA

⁴Department of Thoracic and Cardiovascular Surgery, Cleveland Clinic, Cleveland, OH, USA

 OPEN ACCESS

Received: Mar 14, 2024

Revised: May 7, 2024

Accepted: May 28, 2024

Published online: Jun 3, 2024

Correspondence to

David S. Majdalany, MD

Department of Cardiovascular Medicine,
Mayo Clinic, 13400 E. Shea Blvd, Scottsdale,

A 32-year-old female with mosaic Turner syndrome, coarctation of the aorta status post balloon valvuloplasty in childhood, aortic root dilation (44 mm) and a bicuspid aortic valve presented to the Cardio-Obstetrics clinic at 32 weeks gestation of her second pregnancy. Her first pregnancy was unremarkable although she did not have regular cardiac follow-up and was not taking any cardiac medications. There was no family history of aortopathy or congenital heart disease. Transthoracic echocardiography at 32 weeks gestation revealed an eccentrically dilated aortic root on the parasternal long (45 mm; **Figure 1A**) and short axis (48 mm; **Figure 1B**) and subcostal images revealing normal abdominal aortic Doppler pattern suggesting no residual coarctation (**Figure 1C**). Magnetic resonance imaging revealing the bicuspid aortic valve (**Figure 1D**) and eccentric aortic root dilation measuring 65 mm (**Figure 1E, F**).

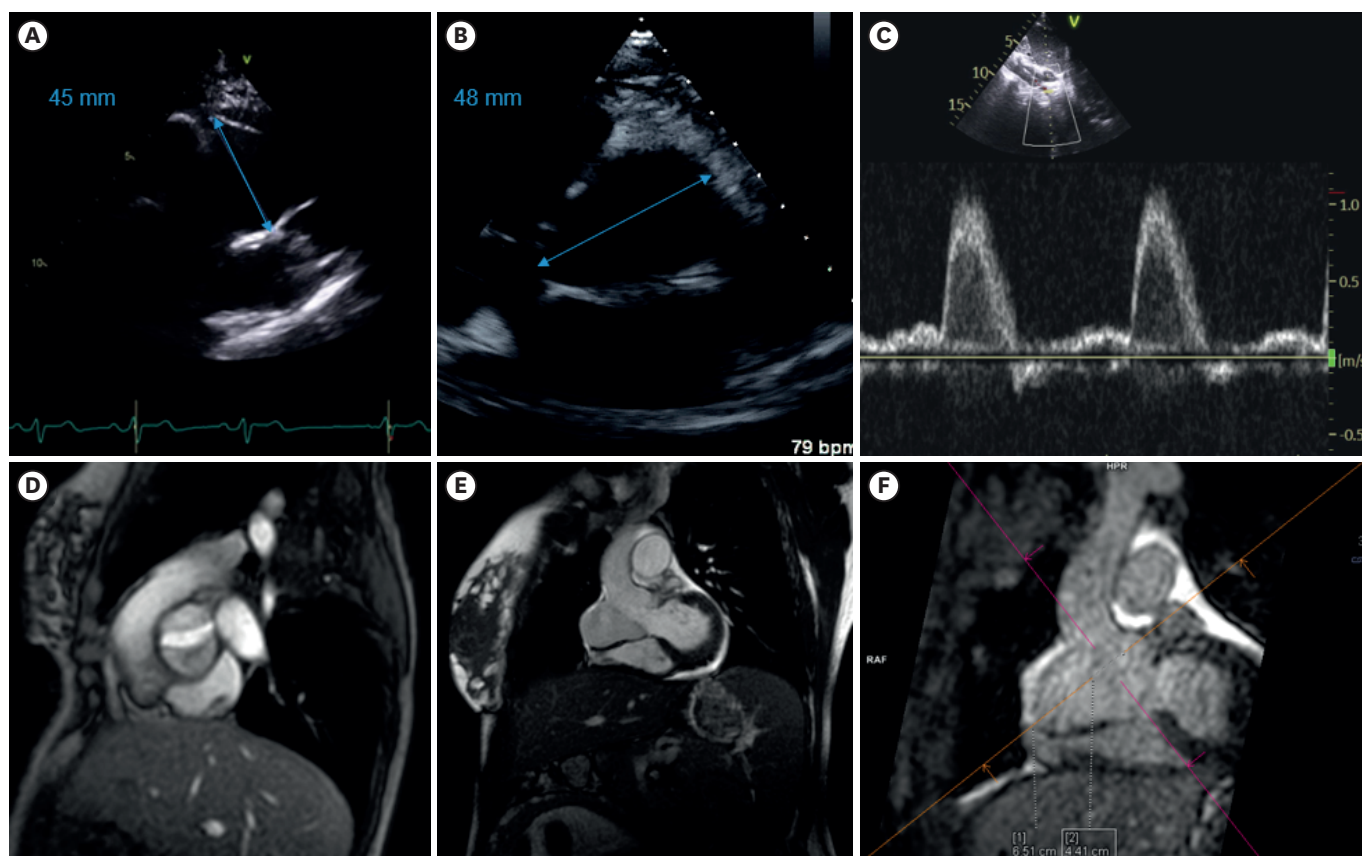


Figure 1. Select cardiac images of a pregnant patient with mosaic Turner syndrome. Echocardiographic parasternal long axis images revealing dilated aortic root at 45 mm (A), parasternal short axis images revealing an eccentrically dilated aortic root at 48 mm (B) and subcostal images revealing normal abdominal aortic Doppler pattern suggesting no residual coarctation (C). Magnetic resonance imaging revealing the bicuspid aortic valve (D) and eccentric aortic root dilation measuring 65 mm (E, F).

AZ 85259, USA.

Email: majdalany.david@mayo.edu


Copyright © 2024. The Korean Society of Cardiology

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<https://creativecommons.org/licenses/by-nc/4.0>) which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original work is properly cited.


ORCID iDs

David S. Majdalany 


<https://orcid.org/0000-0002-5472-6689>

Hong S. Lee 


<https://orcid.org/0000-0002-8847-1039>

Timothy Barry 

<https://orcid.org/0000-0002-0576-2990>

Katherine Singh 

<https://orcid.org/0000-0003-2183-5616>

Jeff Chapa 

<https://orcid.org/0009-0006-0413-6468>

Gosta B. Pettersson 

<https://orcid.org/0000-0003-3298-7279>

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Conflict of Interest

The authors have no financial conflicts of interest.

Data Sharing Statement

The data generated in this study is available from the corresponding author upon reasonable request.

Author Contributions

Supervision: Majdalany DS; Writing - original draft: Majdalany DS, Lee HS, Barry T, Chapa J, Pettersson GB; Writing - review & editing: Majdalany DS, Lee HS, Barry T, Singh K, Chapa J, Pettersson GB.

(48 mm; **Figure 1B**) views. Her ascending aorta was mildly dilated at 41 mm with normal transverse aorta. The bicuspid aortic valve had mild regurgitation with normal Doppler pattern in the abdominal aorta (**Figure 1C**) suggesting no residual coarctation. Given eccentric nature of the aortic root dilation, non-contrast cardiac magnetic resonance imaging was ordered with better visualization of the bicuspid aortic valve (**Figure 1D, Supplementary Video 1**) and surprising discovery of severe eccentric dilation of the aortic root at 65 mm (**Figure 1E and F, Supplementary Video 2**) without a fistula with the right ventricle. The patient was admitted to the hospital, started on beta-blocker therapy, and received antenatal corticosteroids for fetal lung maturity prior to undergoing a cesarean section and delivery of a healthy baby boy. On post-operative day 6, the patient underwent repair of the sinus of Valsalva aneurysm with autologous pericardium and had an uneventful post-operative course.

There is limited data on optimal care of patients with mosaic Turner as they may have heterogeneous presentations during pregnancy.^{1,2} Pre-pregnancy counseling is important as well as regular follow-up in the Cardio-Obstetrics clinic during and after pregnancy to monitor for possible aortic pathology and other cardiovascular abnormalities. Tomographic imaging should be considered to supplement echocardiographic assessment.

Informed consent has been obtained from the patient to publish this work.

SUPPLEMENTARY MATERIALS

Supplementary Video 1

Bicuspid aortic valve.

Supplementary Video 2

Eccentrically dilated aortic root.

REFERENCES

1. Shahrokhi Sabzevar S, Mirzaei F, Tanipour MH, Eslahi A, Hasanzadeh Nazarabadi M. Pregnancy in a patient with mosaic turner syndrome: a case report. *CRCP* 2020;5:58-62. **CROSSREF**
2. Yousif A, Wheatley M, Abuzeid MA. A case report of spontaneous pregnancy in a mosaic turner syndrome patient. *Obstet Gynecol Cases Rev* 2020;7:181. **CROSSREF**