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## *Brief Report*

# Atypical left ventricular outflow tract aneurysm diagnosed by three-dimensional echocardiography

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## Report

A 14-YEAR-OLD WHITE FEMALE WAS BORN AT 37 weeks of gestation with a balanced Rastelli Type A atrioventricular septal defect. There was mild sub-aortic narrowing. There was a persistent left superior caval vein to a dilated coronary sinus. The inferior caval vein was not identified, and the abdominal aorta was to the left of the spine. A large azygous vein was seen draining to the right superior caval vein. The pulmonary venous connections were normal.

At 11 weeks of age, she underwent two-patch repair of atrioventricular septal defect with reconstruction of the mitral valve and tricuspid valves. At 13 weeks, repeat mitral valve repair was performed. At 31 months, an additional repeat mitral valvuloplasty was performed. At 34 months, mitral valve replacement with a No. 21 Saint Jude prosthesis was carried out. A dual-chamber epicardial pacemaker was implanted for post-surgical heart block. Culture of the native mitral valve was found to be positive for bacillus infection and she was treated with clindamycin. At 9 years of age, she underwent mitral valve replacement with a No. 25 Saint Jude prosthesis, and septal myectomy through the aortic valve.

The patient has been clinically stable and doing well for 5 years. Serial echocardiograms were performed and demonstrated stable cardiac findings. Intact atrial and ventricular patches, normally functioning No. 25 Saint Jude prosthesis in the mitral position, no residual or recurrent sub-aortic obstruction, mild aortic valve regurgitation, interrupted inferior caval vein with azygous continuation to the superior caval vein, mild left ventricular enlargement with slightly abnormal/dyssynchronous systolic contractility.

At 13 years of age, a routine complete echocardiogram was performed including three-dimensional imaging.[1] Using an IE33 system (Philips Medical Systems, Andover, Massachusetts, United States of America), two- and three-dimensional imaging was accomplished. The S5-sector array probe was used for the two-dimensional imaging and the X3-1 matrix array probe for the three-dimensional imaging.

In this study, an echolucent area lateral to the aortic valve and contiguous with the left ventricular outflow tract could be seen, and measured approximately 17 millimetre by 22 millimetre. This area appeared to expand and contract with ventricular systole and diastole. Clip 1: two-dimensional parasternal short axis.

In some video clips, this echolucent area seemed to be in communication with the left anterior descending coronary artery, and was first thought to be a coronary artery aneurysm. This area could not be conclusively seen from other views. Clip 2: two-dimensional parasternal short axis sliding medially towards the aortic root.

Using the matrix array three-dimensional transducer, several full-volume data sets were obtained from the parasternal window, taking care to use settings that yield the largest area of view and with the highest frame rate. The data sets were obtained with held respirations over four cardiac cycles.

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These data sets were then transferred to an off-line workstation and analysed using Philips Medical Systems QLAB Advanced Ultrasound Quantification Software.

In addition to an arbitrary cropping plane, three cropping planes for the X, Y, and Z axes allow one to view any area within the pyramidal data set at any angle.

Additional post-processing controls allow changes in gain, compress, smoothing, and brightness, which improve shading, shadowing, and contrast to better delineate depth in the three-dimensional image. Cropping and post-processing was done to best show the area of translucency lateral to the aortic valve in the left ventricular outflow tract. Dynamic three-dimensional imaging clearly showed the presence of an aneurysm immediately inferior and lateral to the aortic annulus in the left ventricular outflow tract.

Clip 3: the left ventricular outflow tract and aortic valve viewed from the left ventricular apical perspective. The aneurysm is seen at the 3 o'clock position and expands and contracts with ventricular systole and diastole.

Clip 4: from the apical perspective, but with the apex angled to the right to show the lateral wall of the left ventricular outflow tract. The three-dimensional image shows the aneurysm to be located just anterior to the left anterior descending coronary artery and posterior to the pulmonary annulus. There is echo dropout at the far end of the aneurysm due to the limits of the three-dimensional volume data set. With ventricular systole, the aneurysm appears to move the left anterior descending coronary artery or slightly compress it. The aneurysm also moves towards the pulmonary annulus with systole.

Clip 5: the apex is tilted down to show the prosthetic mitral valve inflow.

Clip 6: this image is looking back towards the aortic annulus (surgeon's view) from within the ascending aorta. The left main coronary artery is seen arising from the aortic root at the 7 o'clock position, with the left anterior descending coronary artery branching superiorly and coursing posterior to the aneurysm.

Clip 7: seen from a high left atrial perspective the left anterior descending coronary artery is seen posterior to the aneurysm and coursing down the anterior interventricular septum. A cardiac catheterisation was carried out to further investigate the left ventricular outflow tract aneurysm (Video clips 8 and 9).

A follow-up chest computed tomography scan was done as cardiac magnetic resonance imaging was contraindicated due to permanent pacing system, and yielded no additional information.

## Discussion

This is a case report illustrating a unique and novel aneurysm development in a patient with atrioventricular septal defect post mitral valve replacement and sub-aortic resection. The aneurysm is located in the transition zone between the lateral fibrous ring of the aortic annulus and the myocardium of the left ventricular outflow tract, anterior to the left anterior descending coronary artery and posterior to the pulmonary trunk not involving the mitral-aortic intervalvular fibrosa. This area is remote from previous surgical sites. To our knowledge, an aneurysm of the left ventricular outflow tract in this area has never been previously described. We can only speculate as to the cause:

- congenital weakness of this area;
- endocarditis;
- atypical presentation of mitral-aortic intervalvular fibrosa aneurysm.[2,3]

The clinical significance of this lesion is unclear but concerning given its intimate location with the left anterior descending coronary artery. Compression of the coronary artery could lead to ischaemia or infarct.[4–7] The aneurysm's intimate location with the pulmonary trunk may also cause compression of the pulmonary artery, although this is less likely because of the strength of the pulmonary artery wall.[8]

It is well known that left ventricular pseudoaneurysms are prone to rupture, but the area of this aneurysm is likely a more fibrous area.[9] There have been suggestions that patients with thick fibrous or calcified walls might be at a lower risk for rupture.[10] Thrombus formation is also a concern. Although the patient does not manifest signs and symptoms of active endocarditis, this is a potential future morbidity. The present plan for this patient remains unclear. If signs of vascular compression develop, it would prompt intervention.

## Conclusions

An aneurysm of the fibro-muscular lateral wall of the left ventricular outflow tract is a very rare occurrence and has not been described previously. Thus, it can be concluded that three-dimensional echocardiography is an excellent adjunct to help delineate the anatomic features of this and similar lesions.

## Supplementary material

The supplementary material referred to in this article is available online at http://www.journals.cambridge.org/CTY

## References


1. Lopez L, Colan S, Frommelt P, et al. Recommendations for quantification methods during the performance of a pediatric echocardiogram: a report from the Pediatric Measurements Writing Group of the American Society of Echocardiography Pediatric and Congenital Heart Disease Council. J Am Soc Echocardiogr 2010; 23: 465–495.
2. Sudhakar S, Sewani A, Agrawal M, Uretsky B. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): a comprehensive review. J Am Soc Echocardiogr 2010; 23: 1009–1018.
3. Selcuk S, Selcuk M, Tufekcioglu O, et al. The use of three-dimensional echocardiography in the visualization of pseudoaneurysm of the mitral-aortic intervalvular fibrosa. Turk kardiyol Dern Ars Arch Turk Soc Cardiol 2009; 37: 328–331.
4. Bier A, Lamphere J, Daily P. Coronary artery compression caused by a large pseudoaneurysm of the mitral-aortic intervalvular fibrosa. J Am Soc Echocardiogr 1995; 8: 753–756.
5. Parashara D, Jacobs L, Kotler M, et al. Angina caused by systolic compression of the left coronary artery as a result of pseudoaneurysm of the mitral-aortic intervalvular fibrosa. Am Heart J 1995; 129: 417–421.
6. Antonellis J, Kostopoulos K, Routoulas T, et al. Aneurysm of the mitral-aortic intervalvular fibrosa as a rare cause of angina pectoris: angiographic demonstration. Cathet Cardiovasc Diagn 1997; 42: 423–426.
7. Almeida J, Pinho P, Torres J, et al. Pseudoaneurysm of the mitral-aortic fibrosa: myocardial ischemia secondary to left coronary compression. J Am Soc Echocardiogr 2002; 15: 96–98.
8. Cowley M, Wells Jr. J, Hodd Jr. W, Kirklin J. Pulmonary arterial compression syndrome caused by false aneurysm of left ventricular outflow. Am J Cardiol 1976; 38: 959–963.
9. Vlodaver Z, Coe J, Edwards J. True and false left ventricular aneurysms. Propensity for the later to rupture. Circulation 1975; 5: 567–572.
10. Kassim T, Lowery R, Nasur A, et al. Pseudoaneurysm of mitral-aortic intervalvular fibrosa: two case reports and review of literature. Eur J Echocardiogr 2010; 11: E7.


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