

# Impressive echocardiographic images of a rare pathology: Aneurysm of the mitral valve – Report of two cases and review of the literature

Mustafa A. Al-Refai<sup>a</sup>, Farouk M. Oueida<sup>b</sup>, Raphael C. Lui<sup>b,\*</sup>, Shukri M. Al-Saif<sup>a</sup>, Hamed M. Al-Omran<sup>c</sup>

<sup>a</sup>The Department of Cardiology, Saud Al-Babtain Cardiac Center, Dammam, Eastern Province; <sup>b</sup>The Department of Cardiac Surgery, Saud Al-Babtain Cardiac Center, Dammam, Eastern Province; <sup>c</sup>The Department of Cardiac Anesthesia, Saud Al-Babtain Cardiac Center, Dammam, Eastern Province

<sup>a-c</sup> Saudi Arabia

Aneurysm of the mitral valve (AMV) is rarely reported. The etiology of this unusual pathology is commonly attributed to aortic valve endocarditis (AVE) with aortic regurgitation (AR) or connective tissue disease. We present two recent cases of AMV with good correlation between pre-operative trans-esophageal echocardiography (TEE), intra-operative real-time 3-dimensional echocardiography (RT-3D-Echo) and surgical findings. The importance of diligent surveillance by TEE in patients with AVE for occurrence of AMV is emphasized. The literature on this topic is briefly reviewed.

© 2012 King Saud University. Production and hosting by Elsevier B.V. All rights reserved.

**Keywords:** Mitral valve, Aneurysm, Aortic valve endocarditis, Trans-esophageal echocardiography, Real-time 3-dimensional echocardiography

## Introduction

Aneurysm of the mitral valve (AMV) is rarely reported in medical literature since it was first described more than four decades ago. However, with the increasing usage of TEE during the last decade, it is being detected much more frequently as evidenced by the increasing number of published case reports [1–4]. In 1991, Delgado et al. [5] proposed the following as mechanisms for the formation of AMV: (1) Direct local spread of infection from aortic valve (AV) along the mitral-aortic intervalvular fibrosa to the mitral valve (MV) leaflets with weakening of the leaflet

structure by the inflammatory and reparative process; and (2) The regurgitant jet from the AR repeatedly strikes the mitral valve anterior leaflet (MVAL), causing structural damage and subsequent aneurysm formation which, under the left ventricular systolic pressure, bulges into the left atrial cavity like a wind sock during systole and collapses during diastole. This hypothesis might explain the fact that all of the reported cases of AMV prior to 2005 were associated with the MVAL. Since 2005, however, five out of the eleven case reports on this topic were related to the mitral valve posterior leaflet (MVPL) [6], thus raising the possibility that factors other than AVE, such as

Received 30 July 2012; revised 15 September 2012; accepted 12 November 2012.

Available online 24 November 2012

\* Corresponding author. Tel.: +966 508998187.  
E-mail address: [Raphael\\_lui07@yahoo.com](mailto:Raphael_lui07@yahoo.com) (R.C. Lui).



1016–7315 © 2012 King Saud University.  
Production and hosting by Elsevier B.V. All rights reserved.

Peer review under responsibility of King Saud University.  
URL: [www.ksu.edu.sa](http://www.ksu.edu.sa)  
<http://dx.doi.org/10.1016/j.jsha.2012.11.002>



P.O. Box 2925 Riyadh – 11461KSA  
Tel: +966 1 2520088 ext 40151  
Fax: +966 1 2520718  
Email: [sha@sha.org.sa](mailto:sha@sha.org.sa)  
URL: [www.sha.org.sa](http://www.sha.org.sa)



Production and hosting by Elsevier

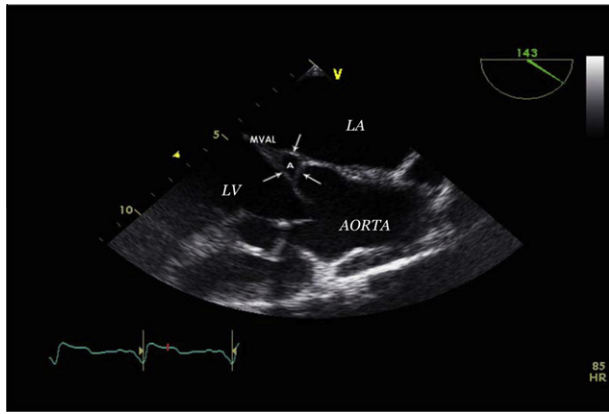


Figure 1a. Patient A, 04/2009 (Ten months before surgery) pre-op TEE showing abnormal "Echo free" space (A) along the mitral-aortic intervalvular fibrosa. MVAL= mitral valve anterior leaflet, LA=Left atrium, LV=Left ventricle.

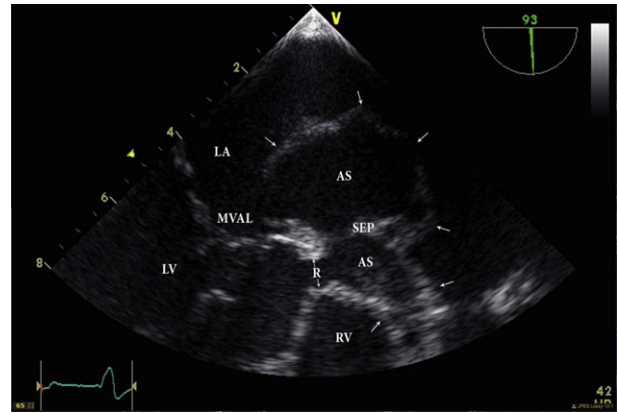


Figure 1b. Patient A: 02/2010 pre-op TEE (long-axis view, Systole) showing a bulging aneurysmal sac (AS, surrounded by arrows) arising from the mitral valve anterior leaflet (MVAL), and was divided by an incomplete septum (SEP) into two communicating chambers. R=rupture of the base of MVAL, LA=Left atrium, LV=Left ventricle, RV=Right ventricle.

connective tissue or myxomatous valvular disease might play a role in the formation of AMV. We describe two recent cases of AMV with good correlation between pre-operative TEE, intra-operative RT-3D-Echo images with surgical findings.

**Patient A:** In April 2009, a 50 year-old Saudi male presented with symptoms of congestive heart failure (CHF). TEE showed severe mitral regurgitation (MR), trace AR, moderate tricuspid regurgitation (TR) and pulmonary hypertension. Left ventricular ejection fraction was reduced to 35%. The MV appeared myxomatous with excessive leaflet motion. In addition, there was an abnormal echo-free space (7 × 10 mm) (Fig. 1a) along the mitral-aortic intervalvular fibrosa. Surgery was recommended but the patient refused. Past medical history was significant for a 2003 TEE at our hospital showing mild MR and LVEF of 70%. Annual follow-up with trans-thoracic echocardiography (TTE) was recommended at that time but apparently not followed.

In February 2010, 10 months later, the patient was re-admitted for worsening of symptoms of CHF, and he finally agreed to surgery. Pre-operative coronary arteriography, carotid Doppler were unremarkable. No history of AVE could be elicited. Repeat TEE confirmed severe MR, trace AR and moderate TR. In addition, a thin-walled aneurysmal sac (AS) 20 × 25 mm arising from the MVAL (Fig. 1b) with blood flow from the left ventricle (LV) to the sac during systole (Fig. 1c). Extension of the AS into the left atrial appendage during systole was also visualized (Fig. 1d).

Under mild hypothermic cardiopulmonary bypass (CPB) and antegrade cold blood cardioplegic arrest, the left atrium (LA) was explored through the inter-atrial groove. An AS measuring 2 cm at

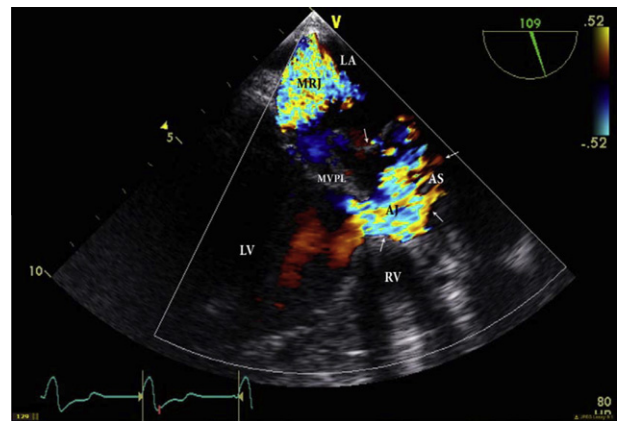


Figure 1c. Patient A, 02/2010 pre-op TEE (long-axis view) showing turbulent jet (AJ) from LV to the aneurysmal sac (AS, surrounded by white arrows) during systole, and turbulent regurgitant jet (MRJ) from left ventricle (LV) to left atrium (LA), MVAL = Mitral valve anterior leaflet.

the oval-shaped base, and 2.5 cm tall was identified arising from the A1 scallop of the MVAL. No vegetation or any evidence of prior endocarditis was noted. The MV was myxomatous in appearance with intact but redundant sub-valvular apparatus. The AS was opened at the roof and no thrombus was found inside. The AS was excised at the base and the defect at the MVAL was primarily closed. MV repair with ring annuloplasty was attempted, but the degree of MR off CPB at physiologic systemic blood pressure remained moderate by TEE. Mitral valve replacement with a mechanical prosthesis was performed. Off CPB TEE showed trace AR as before, and TR was improved. The patient was discharged 20 days after

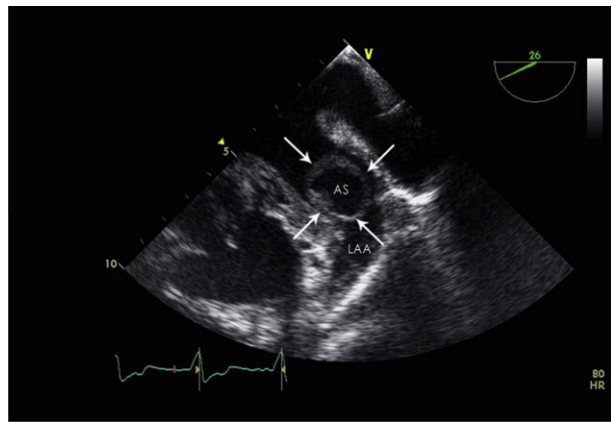


Figure 1d. Patient A, 02/2010 pre-op TEE showing the aneurismal sac (AS) (white arrows) extending into the left atrial appendix (LAA).

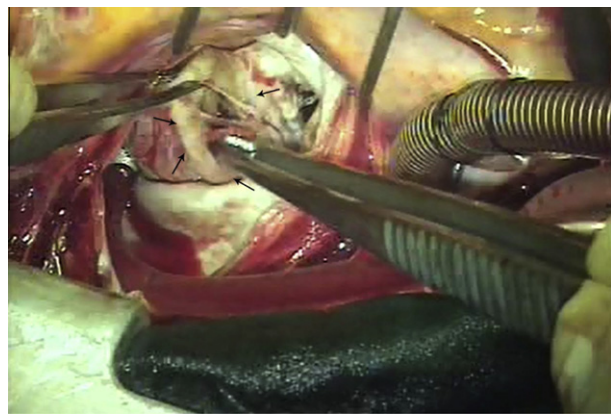


Figure 1e. Patient A, 02/2010 intra-operative photo showing opened aneurismal sac (black arrows) arising from the anterior leaflet of the mitral valve.

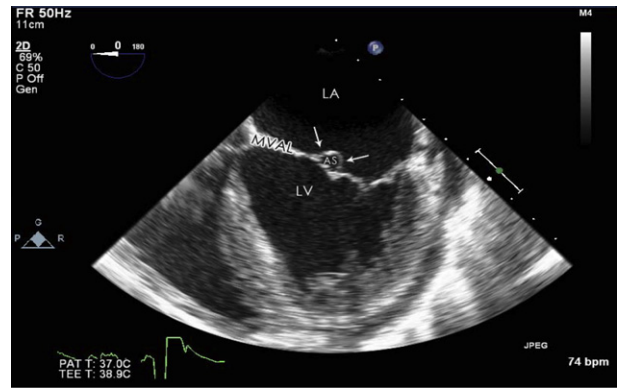


Figure 2a. Patient B, pre-op TEE (long-axis view) showing early development of an aneurismal sac (AS, white arrows) from the mitral valve anterior leaflet (MVAL), LA=Left atrium, LV=Left ventricle.

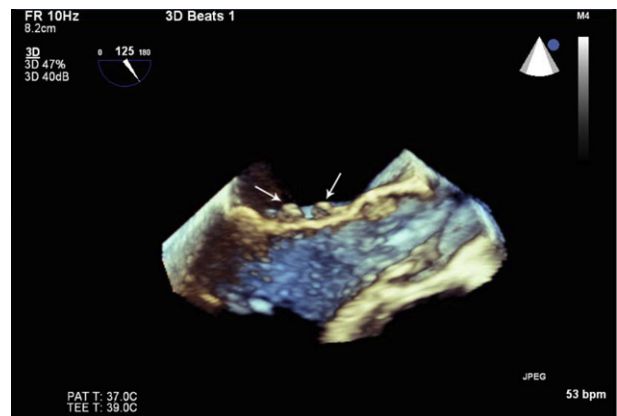


Figure 2b. Patient B, intra-operative RE-3D-Echo showing two small "aneurismal sacs" (white arrows) arising from the mitral valve anterior leaflet.

surgery in good condition but never returned for follow-up.

Patient B: A 25 year-old Saudi male was transferred to us from another facility in April, 2012 with one month history of fever, malaise, poor appetite, fatigue and weight loss of 3 kg. Physical examination revealed systolic and diastolic heart murmur. His WBC count and ESR were moderately elevated. He is afebrile and initial blood cultures twice were negative, most likely due to the two-week course of several broad spectrum antimicrobial agents the patient received prior to transfer. Admission TEE showed severe AR with a mobile vegetation arising from the non-coronary cusp, moderate MR, and mild TR. In addition, a small AS arising from the MVAL was seen (Fig. 2a). A clinical diagnosis of AVE with AMV formation was made. Two weeks after admission, the patient was afebrile on antibiotics but maintained in severe pulmonary congestion due to the severe

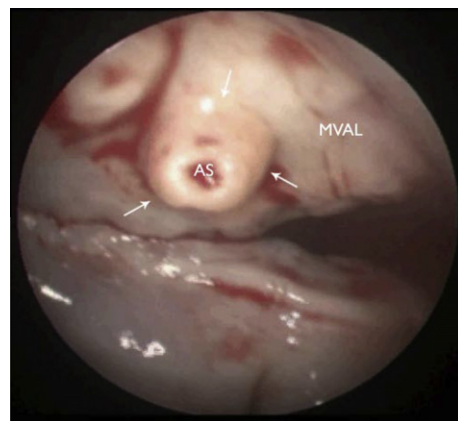


Figure 2c. Patient B, intra-operative thoracoscopic photo showing early development of an aneurismal sac (AS) from the mitral valve anterior leaflet (MVAL). The other smaller aneurismal sac seen by the RT-3D-Echo was not evident by this view.

AR. The decision was made to proceed with surgery. Under general anesthesia, repeat TEE with

RT-3D-Echo confirmed the previous findings and the presence of not one, but two small (5 mm) aneurysmal sacs arising from the MVAL (Fig. 2b). Intra-operatively, the non-coronary cusp of the AV was found to be almost completely torn, and two small aneurysmal sacs were identified protruding from the A1 scallop of the MVAL (Fig. 2c). Aortic and mitral valve replacements with mechanical prosthesis and preservation of the mitral posterior leaflet were performed. Subsequent histological examination of the resected aortic valve showed, in addition to the inflammatory changes, thickened tunica media containing elastic and collagen tissue fibers with some accumulation of mucopolysaccharidases but no overt large pools of mucin. Focal disruption of elastic fibers and a few small foci of alcian-blue positive mucoid material were seen, suggesting focal mild degenerative changes. The patient was discharged one month later in good condition. Follow-ups at one week and one month with TTE did not reveal any problems.

## Discussion

The term AMV first appeared in the English literature in 1963 in an article by Edynak et al. [7]. However, 2 years prior to that, an article (in Italian) by De Annibalis, entitled "Aortic cuspid aneurysm of the mitral valve" could be found. During the following four decades, about 26 sporadic case reports appeared in English medical journals. Due to the much wider application of TEE during the last decade, reports on this rare pathology have increased markedly.

In 1986, Vandenbossche et al. first documented the formation of AMV by cross-sectional echocardiography [8]. One year later, 2D-Echo was shown to be a more sensitive diagnostic tool. In 1995 and 2006, TEE and cardiac magnetic imaging (C-MRI) respectively were introduced into the diagnostic armamentarium with even higher sensitivity. More recently, the complimentary role of RT-3D-Echo in detecting AMV has been described [2,4].

AMV has been linked to a wide range of congenital and adult cardiac structural defects such as congenital left ventricular outflow tract obstruction and partial atrio-ventricular septal defect [9]; hypertrophic obstructive cardiomyopathy; bicuspid and quadricuspid AV; and rheumatic mitral valve disease. Genetic disease such as Libman-Sachs [10] and Marfan's syndrome with aneurysm of the sinus of Valsalva have also been implicated to the formation of AMV in patients without AVE.

Patient A displayed TEE evidence of myxomatous MV disease which was confirmed by intra-

operative inspection of the MV. Unfortunately, we did not submit the resected MV for pathological examination. Interestingly, the first abnormal echocardiographic finding (Echo-free space at the mitral-aortic intervalvular fibrosa) of this patient preceded the formation of a 2.5 cm AS by 10 months. This case clearly lends support to the theory that connective tissue disorder alone, without endocarditis, is a causative factor of AMV.

Patient B did not have any previous echocardiographic images to determine whether his two small aneurysmal sacs were pre-existing or was a result of his AVE with AR. He did have pathological evidence of focal mild degenerative disease of the AV (but not the MV which was not submitted for pathological examination). The etiology for this patient's two aneurysmal sacs might very well be a combination of both. Interestingly, the pre-operative TEE in this patient showed only one AS, and yet the intra-operative RT-3D-Echo identified two aneurysmal sacs which was confirmed intra-operatively, providing support to the claim that 3D does have a higher sensitivity over 2D echocardiography.

While isolated reports of conservative observation of AMV for up to three years without complication could be found, the overwhelming majority of reported cases of AMV were treated by surgical excision of the infected valvular tissues and replacement of the valves. More recently, however, several cases of successful resections of the AMVs only and repair of the MVs were reported [11].

## Conclusion

AMV was thought to be a rare pathology in the past. The advent of more sensitive diagnostic tools such as TEE, cardiac MRI, and RT-3D-Echo suggested that its prevalence might actually be higher among patients with AVE or connective tissue disease. Clinicians caring for this group of patients should be more aware of the recent advances in diagnostic tools available for this not-so-rare pathology. Surgical resection of all the infected or myxomatous valve tissues and replacement with prosthetic valves remain the current choice of treatment.

## Acknowledgment

The authors wish to thank Mr. Abdou Tannous for his expert assistance in the preparation of the graphics in this manuscript.

## References

- [1] Hotchi J, Hoshiga M, Okabe T. Impressive echocardiographic images of a mitral valve aneurysm. *Circulation* 2011 April;123(14):400-2.
- [2] Michelena HI, Suri RM, Enriquez-Sarano M, et al. Ruptured mycotic aneurysm of the mitral valve on real-time 3-dimensional trans-thoracic echocardiography. *J Am Coll Cardiol* 2010 July 6;56(2):154.
- [3] Piazza N, Marra S, Webb J, et al. Two cases of aneurysm of the anterior mitral valve leaflet associated with transcatheter aortic valve endocarditis, a mere coincidence? *J Thorac Cardiovasc Surg* 2010 Sept.;140(3):36-8.
- [4] Hong SN, Perk G, Skolnick A. Evaluation of a posterior mitral valve leaflet aneurysm by real-time 3-dimensional trans-esophageal echocardiography. *Echocardiography* 2009 Oct;26(9):1089-91.
- [5] Delgado R, Martin Duran R, Vazquez de Prada JA, et al. The importance of the echocardiographic diagnosis of a mycotic aneurysm of the mitral septal leaflet in infective endocarditis of the aortic valve: the surgical implication. *Rev Esp Cardiol* 1991 Dec;44(10):672-6.
- [6] Lee CH, Tsai M. An aneurysm of posterior mitral valve mimicking a huge vegetation. *Int J Cardiol* 2006;107(2):277-8.
- [7] Edynak GM, Rawson AJ. Ruptured aneurysm of the mitral valve in a Marfan-like syndrome. *Am J Cardiol* 1963:674-7.
- [8] Vandenbossche JL, Hartenberg D, Leclerc JL, et al. Mitral valve aneurysm formation documented by cross-sectional echocardiography. *Eur Heart J* 1986;7(2):171-5.
- [9] Kathir K, Dunn RF. Congenital obstructive mitral valve aneurysm. *Intern Med J* 2003;33(11):541-2.
- [10] Takayama T, Teramura M, Sakai H, et al. Perforated mitral valve aneurysm associated with Libman Sacks endocarditis. *Intern Med* 2008;47(18):1605-8.
- [11] Hori D, Tanaka M, Yamaguchi A. Extracardiac aneurysm of the interleaflet triangle above the aorto-mitral curtain due to infective endocarditis of the bicuspid aortic valve. *Gen Thorac Cardiovasc Surg* 2008;56(8):424-6.