

CASE REPORT: DOUBLE ORIFICE MITRAL VALVE WITH CLEFT IN ANTERIOR LEAFLET OF DOMINANT VALVE IN AN AFRO-CARIBBEAN

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ABSTRACT

Double orifice mitral valve (DOMV) is a rare congenital mitral valve (MV) disorder. It is associated with other types of congenital heart disease in 45% of patients, but it has been documented to be isolated with or without noncompaction of the left ventricle. The focused features of this case report are the transthoracic echocardiographic images of an isolated case of DOMV, with an additional rare association of a cleft in the anterior leaflet of the dominant MV presenting as mitral regurgitation in a 13-year-old Afro-Caribbean female.

Keywords: Double orifice mitral valve, noncompaction, tensor apparatus, rheumatic heart disease.

INTRODUCTION

In 1876 Greenfield¹ described the first case of double orifice mitral valve (DOMV). There have been >200 cases documented of all types of DOMV^{2,3} via autopsy, transthoracic, and transoesophageal echocardiography (TTE and TOE), with anatomy more clearly defined by three-dimensional (3D) echocardiogram.³⁻⁵ Banerjee et al.⁶ indicates that there is an incidence rate of <0.05% in the population. Isolated cases are a rare occurrence in the DOMV group and the true incidence of this specific type is not known. There are >40% of DOMV cases that are associated with other types of cardiovascular disease, the most common of which are atrioventricular septal and ventricular septal defects, comprising 56% to 17%, in post mortem series. There can be up to 39% with left sided obstructive lesions, such as coarctation of the aorta, interrupted aortic arch, or right-sided obstructive lesion such as pulmonary stenosis. The list of associated conditions include: atrioventricular septal defect, primum atrial septal defect, ventricular septal defect, atrial septal defect, truncus arteriosus, coarctation of the aorta, interruption of the aortic arch, tetralogy of Fallot, corrected

transposition, bicuspid aortic valve, pulmonary stenosis, Ebstein's anomaly of the tricuspid valve, dysplastic tricuspid valve, double orifice tricuspid valve, tricuspid atresia, parachute mitral valve, marfanoid features, left ventricular noncompaction, atrial tachycardia, congenital complete heart block, and cardiomyopathy.¹⁻¹⁴

Presentation may not occur, hence being totally asymptomatic with competent DOMV.⁴ Mitral regurgitation (MR) occurs in the majority of cases (45%) and mitral stenosis (MS) in 11%, in either or both MVs to varying degrees of severity. MR and MS can be masked by concomitant cardiac lesions such as interatrial communications of primum atrial septal defect and secundum atrial septal defect, and also tetralogy of Fallot,⁹ which reduces left ventricular inflow. There are varied abnormalities of the MV annulus, the tensor apparatus and papillary muscles which may be duplicated, with each MV having its own complete subvalvular attachments to the left ventricle (LV).¹⁻⁸

DOMV may occur with or without noncompaction of the LV.⁵ They are then classified into three main groups anatomically by Trowitzsch et al.² as Type 1, having a complete bridge going between the valves

as in the index case, Type 2, an incomplete bridge with connections at the edge of the leaflets of the MVs, and Type 3, a hole in the lateral commissure. The TTE findings, the focused features of the index case, are of an isolated DOMV with severe MR in both MVs, in which the index developing country was mistaken initially for rheumatic heart disease (RHD) affecting the MV, and the patient was placed on a 28-day prophylactic penicillin therapy.

CASE REPORT OF TTE FEATURES OF THE INDEX CASE

The index case is a 13-year-old teenager with clinical signs of MR, being reevaluated with TTE. There was situs solitus, atrioventricular concordance, and ventriculoarterial concordance. **Figure 1** shows normal off-setting atrioventricular valves (AV) ruling out atrioventricular septal defect structural morphology. A large left atrium (LA) with an intact interatrial septum (IAS) deviated to the right, indicating a relative increase in left atrial pressure. There are dilated pulmonary veins (PV), indicative of raised left atrial pressures. Relative large LV with LV end diastolic diameter of 4.2 cm with low fractional shortening of 27% and ejection fraction of 53%. The right ventricle (RV) has a prominent moderator band. In this image the MVs are closed (dominant MV1 and smaller MV2) with their tendon apparatus noted.

Figure 2 shows both the MVs, the dominant MV1, and smaller MV2 open. As in **Figure 1**, it also shows large LA with IAS deviated to the right, dilated PVs, relative large LV, and the RV with prominent moderator band. **Figure 3** shows forward, diastolic flow across dominant MV1 and to the left of this flow, a smaller red colour Doppler flow across MV2. **Figure 4** shows regurgitant flow of dominant MV1 deviated to the left and posteriorly with reversal of flow in dilated left PV. **Figure 5** shows pulsed Doppler regurgitant flow of MV1 of 4 m/s. **Figure 6** shows, in rotated four chamber view, smaller MV2 open. This figure also depicts, as previous figures, large LA with IAS deviated to the right, dilated PV, large LV, and in this view, relative large RV with prominent moderator band. **Figure 7** shows with colour Doppler regurgitant flow in MV2 which was directed posteriorly and to the lateral aspect of the LA. **Figure 8** with posterior angulation in four chamber view, shows colour Doppler guided continuous wave (CW) regurgitant flow MV2 of CW 6.5 m/s. There is reversal of flow in left PVs.

Figure 9 shows dilated LA and aorta (AO). The AO:LA ratio 1:3 where the normal ratio is 1:1.1 confirms a markedly dilated LA. Dilated PV is also noted. Left atrial volumes were not necessary to confirm markedly enlarged LA, in this specific case. **Figure 10** parasternal short axis view, shows two unequal MVs with the medial valve being the larger dominant valve and an anterior lateral inferior smaller valve with abnormal tensor apparatus for both valves. There is marked left atrial enlargement. **Figure 10** shows dominant valve closed, whereas **Figure 2** shows dominant valve open. **Figure 11** shows short axis view of LV with MV1 with open cleft in anterior leaflet (MV1) which is partially open and MV2. **Figure 12** shows short axis view of LV with both MV1 cleft and MV2 fully open.

The dominant MV had a cleft in anterior leaflet with severe MR extending to PVs which were dilated. The smaller MV also had a moderate-to-severe form of MR. The two valves were not seen concomitantly in the same four chamber view plane. Hence, the use of separate four chamber views showed severe MR of the medial dominant MV and moderate-to-severe MR in the smaller anterior lateral MV.

The directions of the regurgitant flow from both MV1 and MV2 were to the left and posterior, with far-field beam widening and also attenuation closer to the back of the LA, and hence were not parallel to the beam and were most likely underestimated. The peak E velocities for MV1 and MV2 did not exceed 1.5 m/s, ruling out a concomitant MS. It was not possible to clearly differentiate E and A waves in the absence of atrial fibrillation. Because of the direction of regurgitant flow and the eccentric orifices of MV1 and MV2, the presence of multiple jets from the MV1, the cleft of MV1, and MV2 makes the calculation of vena contracta, jet area, and proximal isovelocity surface area invalid in the assessment of severity of MR. No A waves with poor left ventricular function implies some diastolic dysfunction.

The parasternal long axis view showed dilated LA with an AO to LA ratio (i.e. AO:LA) greater than 1:3 (normal 1:1.1, **Figure 5**). Hence, direct and indirect assessment of MV regurgitant severity was shown by the regurgitant flow hitting the back of the dilated LA with reversal of flow in dilated PVs and bowing of the intact IAS to the right in the apical four chamber view. The presence of markedly dilated right and left PVs demonstrated markedly increased left atrial pressures. The LV was also enlarged. 3D echocardiography, cardiac

magnetic resonance imaging, and computed tomography are not available in the index country.

DISCUSSION

The embryological development of the MVs occurs with the merging of the endocardial cushion and the myocardial ridge by the 40th day of gestation, connecting the final MV to the tensor apparatus and papillary muscle on the left ventricular wall. DOMV is believed to occur when merging is abnormal. The MV abnormality is believed to be secondary to genetic mutations affecting the regulatory proteins in the myocytes.⁸

A familial case involving a brother and sister has been described, indicating an underlying chromosomal or genetic cause;¹¹ however, the index case had no family history of congenital heart disease or MV disease. 85% of DOMV patients have a dominant orifice and smaller vestigial one as in the index case. Two equal valves occur in 15% of cases, or a third duplication of the MV with each its own tensor apparatus.

The varied structural anatomy of the MV, their competence, tensor apparatus, papillary muscle, and noncompaction determines the final clinical presentation; DOMV patients with competent valves would be asymptomatic and diagnosed as an incidental echocardiographic⁴ or autopsy finding,^{1,2}

comparing the patients who present signs of severe pulmonary hypertension (PH) with Eisenmenger's syndrome to the ones presenting with mitral stenotic symptoms and signs. The presentation can be dilated cardiomyopathy when noncompaction is predominant.¹⁴

All the aforementioned symptoms and signs are well documented.³⁻¹⁴ Hence the management of DOMV is based on the patient's individual anatomy and physiology. This is usually only needed when there is significant MR and MS.^{3,14}

This case confirms that complex structural cardiac anatomy can be clearly defined in expert hands. Surgical intervention with prosthetic MV replacement was recommended to avoid use of anticoagulants until after child bearing because of the documented potential teratogenic effects of these drugs; mechanical valves could be used thereafter.

CONCLUSION

This paper reviewed an index case of DOMV with cleft in anterior leaflet of dominant valve with severe MR. In a developing country it is important to note that all mitral regurgitant signs are not due to RHD. Isolated DOMV is a rare congenital cardiac malformation, which can cause irreversible cardiac disease and PH if not detected early, and requires a high index of suspicion.

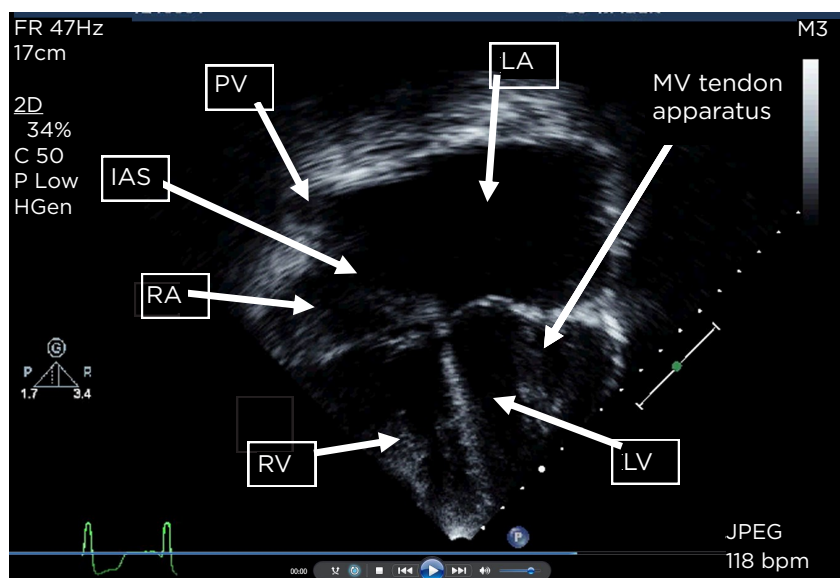


Figure 1: Normal off-setting atrioventricular (AV) valves. Large left atrium (LA) with inter atrial septum (IAS) to the right. Dilated pulmonary veins (PV). Large left ventricle (LV). Large right ventricle (RV) with prominent moderator band. Mitral valves (MV1 & 2) tendon apparatus closed.

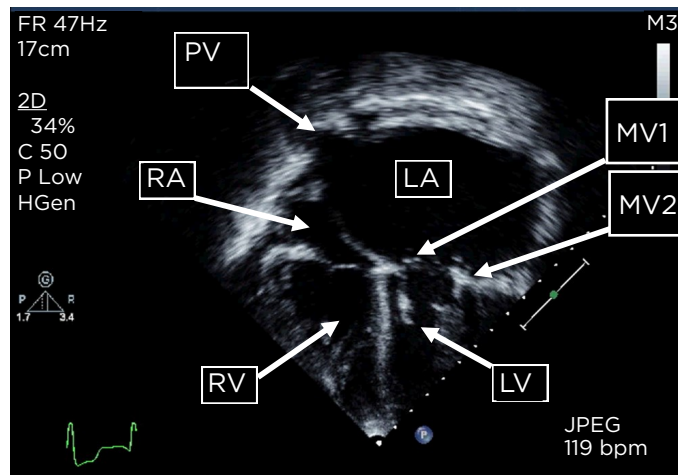


Figure 2: Normal off-setting atrioventricular valves. Large left atrium (LA) with inter atrial septum to the right. Dilated pulmonary veins (PV). Large left ventricle (LV). Large right ventricle (RV) with prominent moderator band. Mitral valves (MV1 & 2 open).

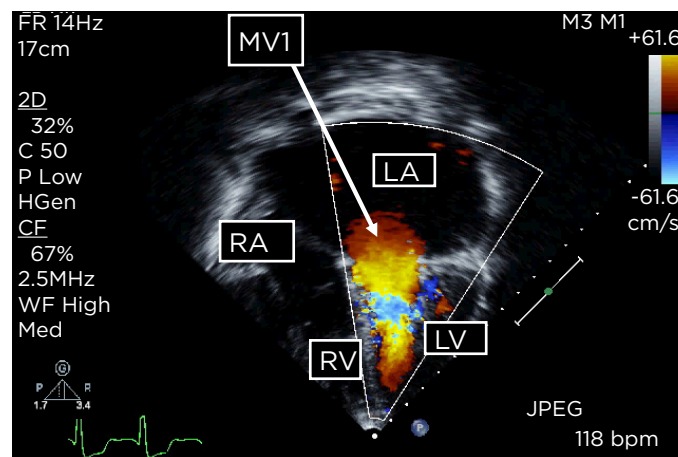


Figure 3: Diastolic flow mitral valve 1 (MV1).
L/RV: left/right ventricle; L/RA: left/right atrium.

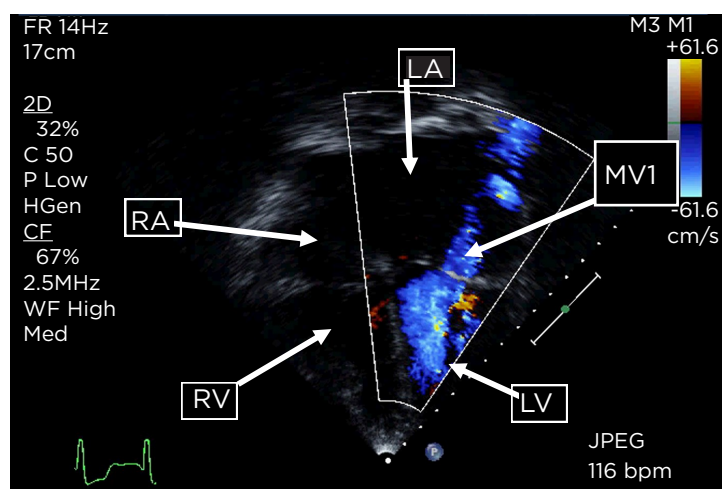


Figure 4: Regurgitant flow mitral valve 1 (MV1) with reversal of flow in left pulmonary vein.
L/RV: left/right ventricle; L/RA: left/right atrium.

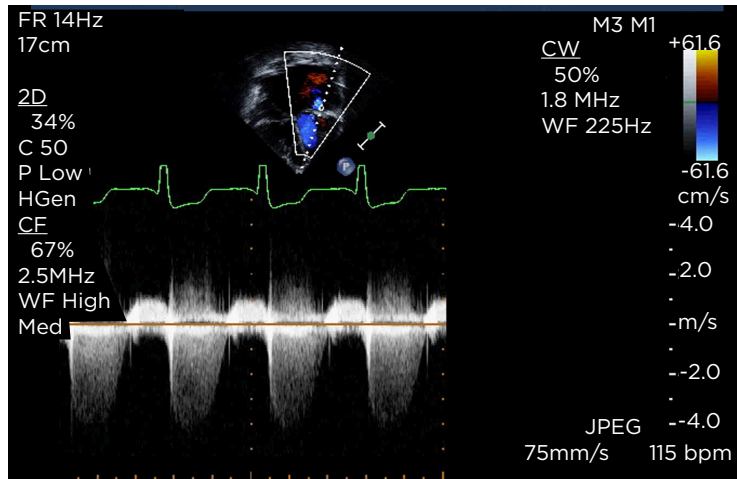


Figure 5: Doppler regurgitant flow mitral valve 1 pulsed wave 4 m/s.

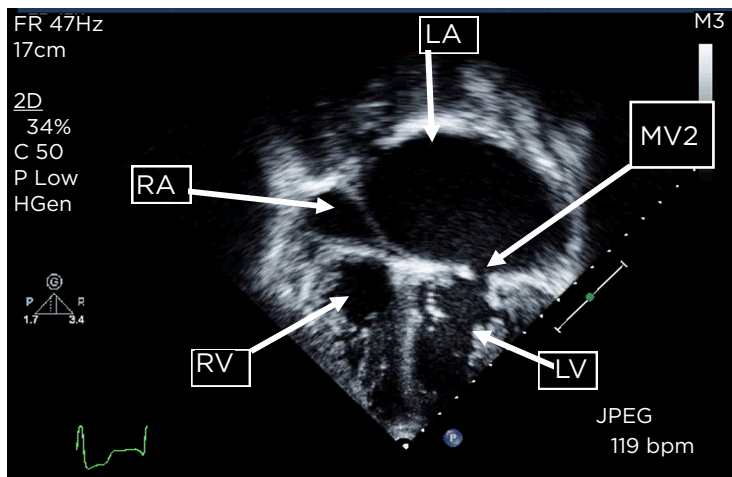


Figure 6: Mitral valves (MV2 open). Large left atrium (LA) with interatrial septum to the right. Dilated pulmonary veins (PV). Large left ventricle (LV). Large right ventricle (RV) with prominent moderator band.

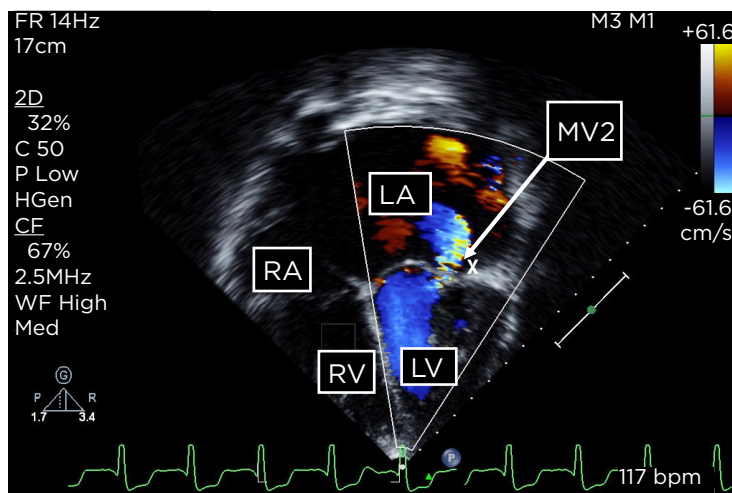


Figure 7: Regurgitant flow in mitral valve 2 (MV2).
L/RV: left/right ventricle; L/RA: left/right atrium.

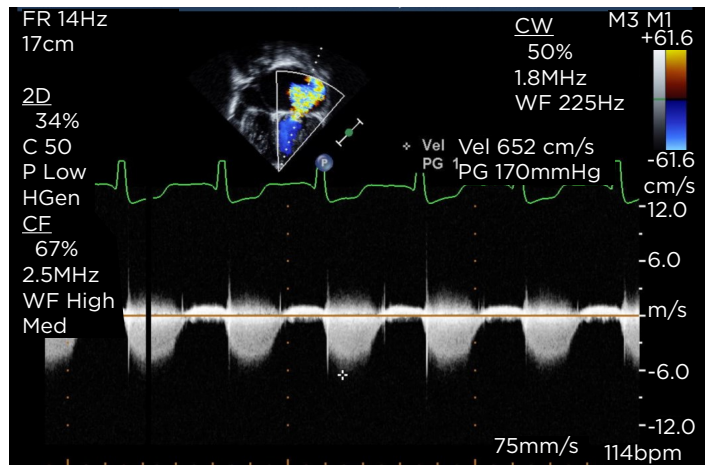


Figure 8: Doppler regurgitant flow mitral valve 2 (MV2) continuous wave 6.5m/s. Reversal of flow in left pulmonary veins.

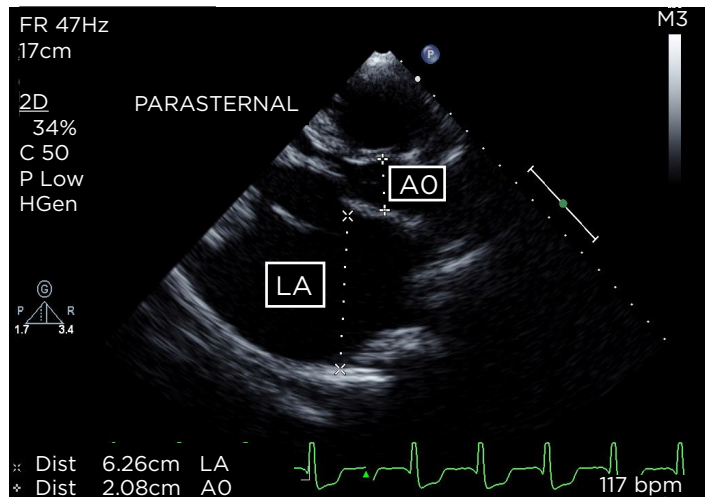


Figure 9: Dilated left atrium (LA), aorta (AO). AO: LA ratio 1:3. Dilated pulmonary veins.

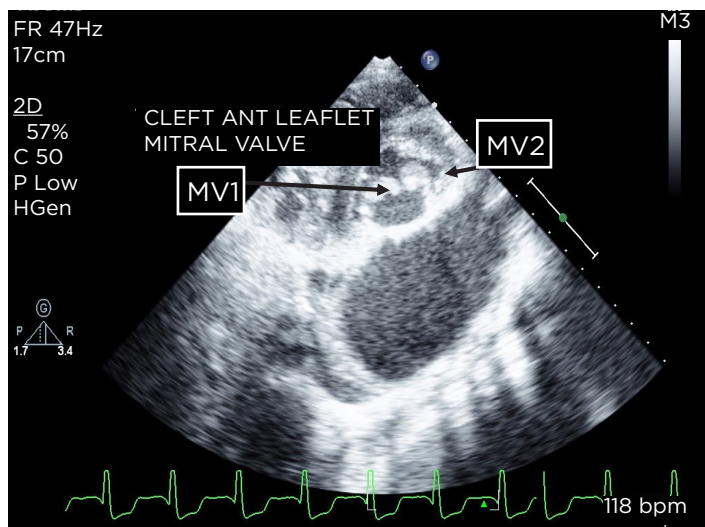


Figure 10: Short axis view of left ventricle with mitral valve 1 (MV1) with cleft closed and mitral valve 2 (MV2).

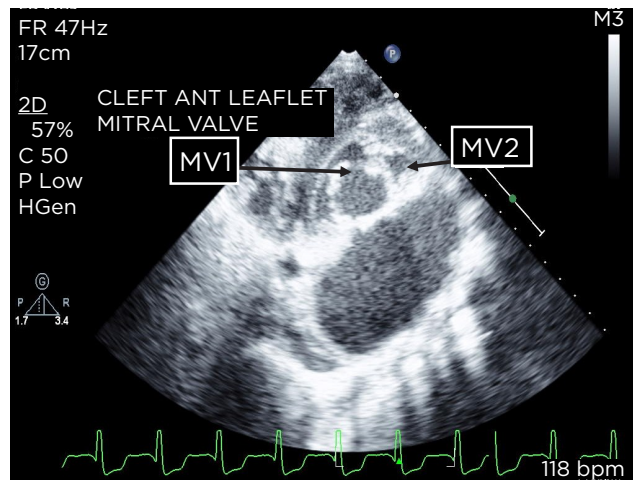


Figure 11: Short axis view of left ventricle with mitral valve 1 (MV1) with open cleft in anterior leaflet partially open and mitral valve 2 (MV2).

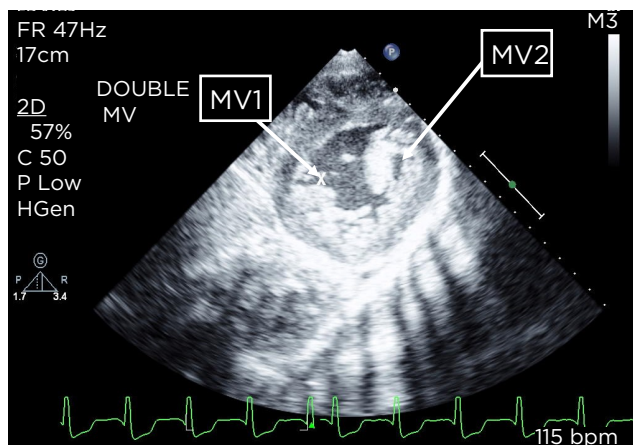


Figure 12: Short axis view of left ventricle with mitral valve I (MV1) cleft fully open and mitral valve 2 (MV2).

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