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Mitral valve dysplasia syndrome with severely restricted atrial septum: Insights from two cases of fetal atrial septal stenting

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Introduction

- Mitral valve dysplasia (MVD) is characterized by a dysplastic mitral valve, moderate to severe mitral regurgitation (MR), and left atrial (LA) and ventricular (LV) dilation
- MVD is often associated with aortic stenosis and restrictive/intact atrial septum (R/IAS) leading to elevated LA pressure, lung abnormalities, and poor prognosis with 50-76% fetal and neonatal mortality
- Objectives:
 - Review two cases of mitral valve dysplasia (MVD) that underwent fetal atrial septal stenting (FASI),
 - Demonstrate the role of FASI in improving pulmonary hemodynamics and postnatal stability

Clinical Presentation

- Two fetuses were referred for evaluation and potential intervention with mitral valve dysplasia with restrictive/intact atrial septum
- Fetal echocardiography demonstrated:
 - MVD with moderate to severe MR (Patient 1), severe MR (Patient 2)
 - Forward to reverse pulmonary vein velocity time interval ratio (F/R VTI) <3, predicting severe hypoxemia at birth
 - Severe aortic valve stenosis
 - Severe LA and LV dilation with endocardial fibroelastosis
 - Severely depressed LV systolic function
- Fetal MRI (fMRI) revealed pulmonary lymphangiectasia in both cases

Cases

- Case 1:** Patient 1 had an IAS with a small pericardial effusion
- Successful FASI at 29 weeks with a 2.5x12mm Rebel stent
 - F/R VTI ratio improved to > 3
 - Follow-up fMRI at 35 weeks showed resolution of the effusion and significant improvement in pulmonary lymphangiectasia
 - Infant was delivered at 38 weeks, underwent balloon aortic valvuloplasty, and achieved a biventricular repair

- Case 2:** Patient 2 had a severely RAS
- FASI performed at 29 weeks with a 2.5x12mm Rebel stent
 - Stent thrombosis was noted on post-operative day 1 despite adequate flow noted in the operating room.
 - Despite this, follow up evaluation showed reduced MR, increased antegrade flow across the mitral and aortic valves, and improved pulmonary lymphangiectasia on follow-up fMRI
 - Infant delivered at 39 weeks and underwent emergent atrial septal stent placement on DOL 0 followed by a Norwood at DOL 6
 - Doing well in the inter-stage period

Conclusion

- MVD is a rare left-sided heart disease with high mortality
- In these two MVD cases, pulmonary lymphangiectasia improved after FASI by decompressing the left atrium
- Despite stent thrombosis in the second case, MR and antegrade aortic flow improved possibly due to changes in left atrial configuration
- FASI may prevent hydrops and support a more stable postnatal transition by improving pulmonary hemodynamics

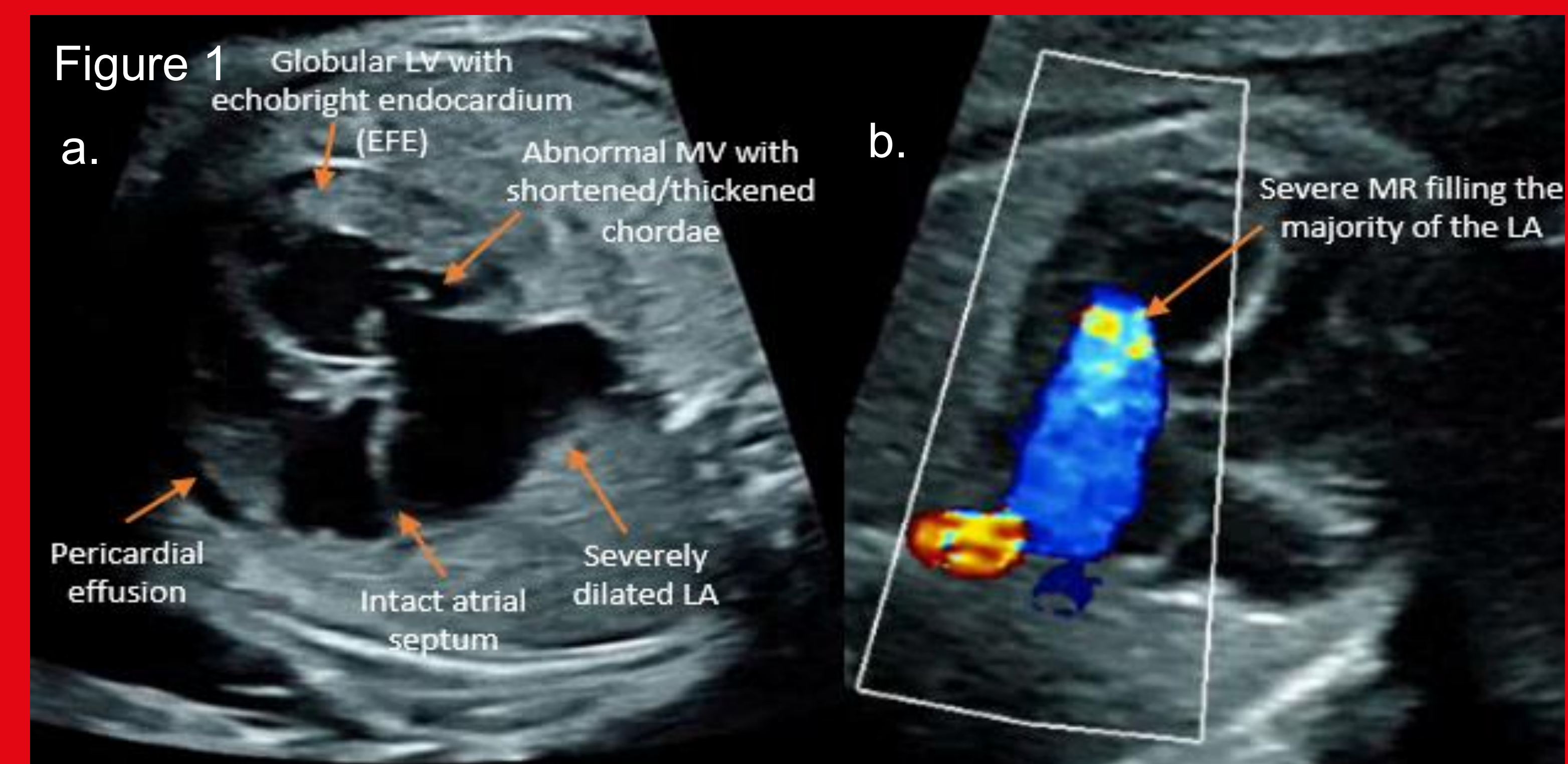


Fig 1a-b. Baseline 2D image demonstrating an abnormal mitral valve with severe LA dilation, globular LV with EFE, small pericardial effusion and color image demonstrating severe MR originating at the leaflet coaptation point in Patient 1

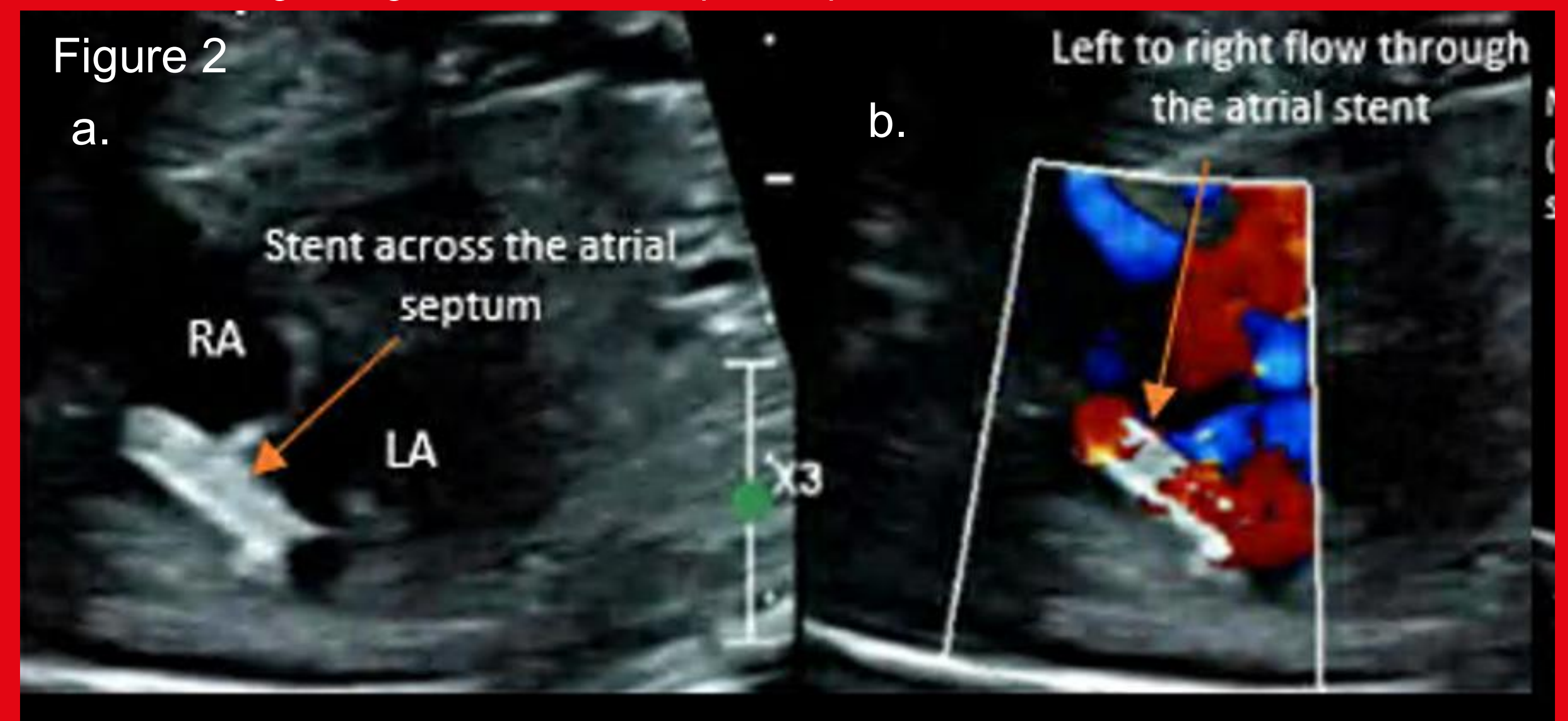


Fig 2a-b: Atrial stent visualized across the atrial septum with left to right shunting in Patient 1.

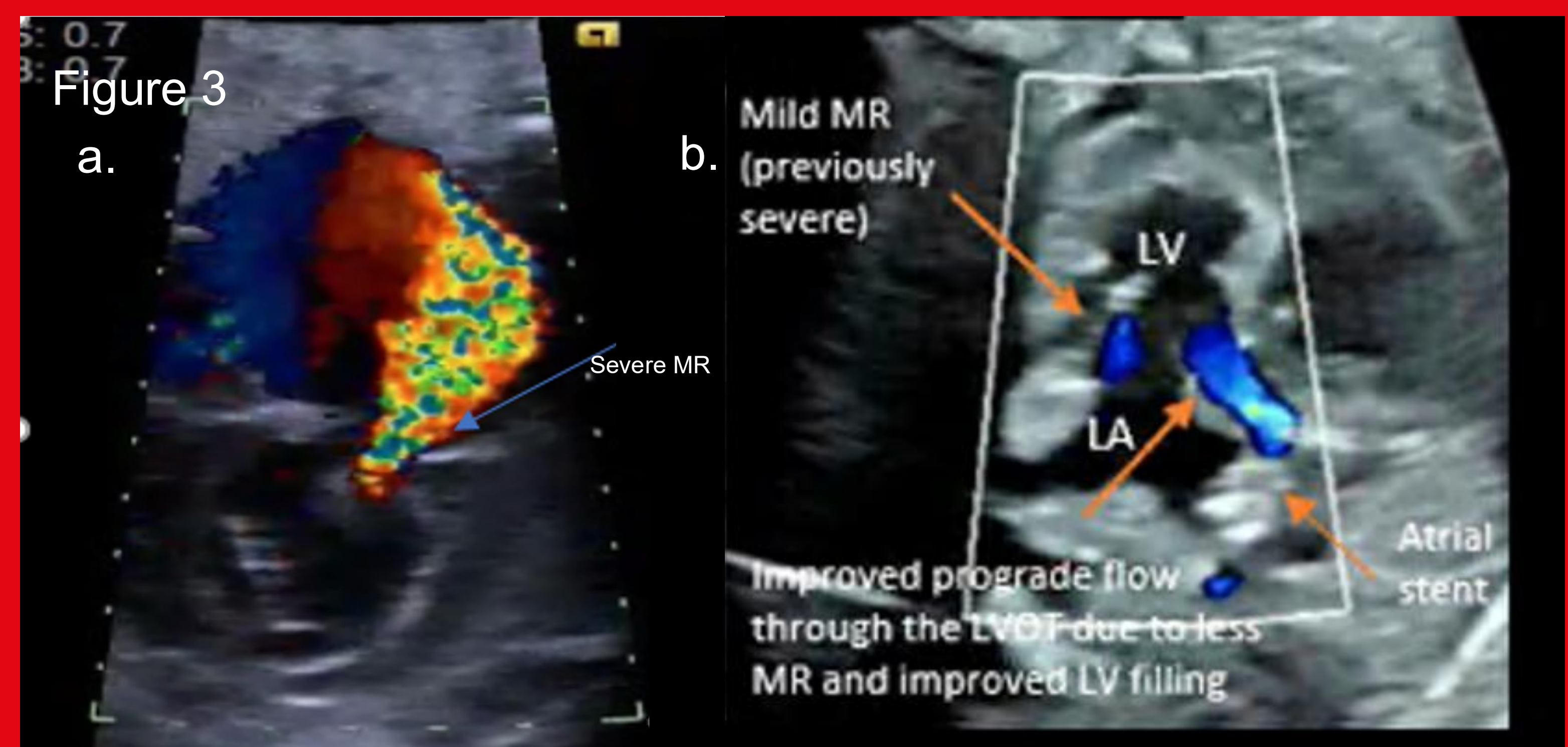


Fig 3a. Baseline color image demonstrating severe MR through an abnormal mitral valve and a dilated globular LV in Patient 2. 3b. Color image demonstrates improvement in mitral regurgitation and prograde flow across the LVOT post stent placement