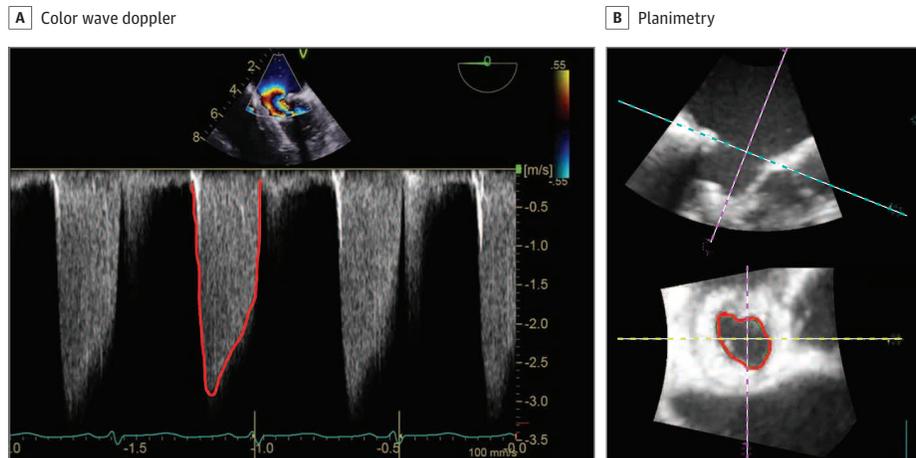


## JAMA Cardiology Clinical Challenge

## A Pregnant Woman With Shortness of Breath

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**Figure.** A, Color wave doppler demonstrating a mean mitral valve gradient of 21 mm Hg at a heart rate of 100 beats per minute. B, Three-dimensional mitral valve area (1.22 cm<sup>2</sup>) as measured by planimetry.

**A 40-year-old woman** presented with a history of mitral valve (MV) endocarditis leading to MV repair (without annuloplasty) 15 years ago. Four years ago while pregnant, she had recurrent endocarditis complicated by severe symptomatic mitral regurgitation, but she was successfully brought to term and had a cesarean delivery. Post partum, her symptoms resolved, but she had persistent marked mitral regurgitation related to a severely deformed posterior leaflet. She wished to have another child, and after extensive counseling she elected to undergo a complex redo MV repair (annuloplasty) 2 years ago. She developed perioperative atrial fibrillation but declined anticoagulation and has not had further arrhythmias. Serial transthoracic echocardiograms have demonstrated normal biventricular function and pulmonary pressures. Her postoperative MV gradient was 10 mm Hg (heart rate, 90 beats per minute [bpm]) and has been stably elevated since surgery.

At presentation, the patient was 19 weeks pregnant at presentation. She had mild exertional dyspnea, a right ventricular (RV) heave, and elevated jugular venous pulsation. Transthoracic echocardiogram demonstrated normal biventricular function, mild RV dilation, RV systolic pressure of 84 mm Hg, normal left atrial size, and mean MV gradient of 18 mm Hg at a heart rate of 97 bpm. Transesophageal echocardiogram revealed mean MV gradient of 21 mm Hg at a heart rate of 100 bpm (Figure). She strongly desired this pregnancy and declined termination.

## WHAT WOULD YOU DO NEXT?

- A. Perform balloon mitral valvuloplasty
- B. Perform transcatheter MV-in-ring procedure
- C. Trial medical management with metoprolol and enoxaparin
- D. Perform surgical MV replacement

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## Diagnosis

**Severe mitral stenosis (MS) after complex MV repair worsened by hemodynamic changes of pregnancy**

## What to Do Next

- C. Trial medical management with metoprolol and enoxaparin

## Discussion

Severe MS in pregnancy is associated with increased risk of maternal heart failure, maternal arrhythmia, and preterm birth.<sup>1</sup> Addi-

tionally, pulmonary arterial hypertension in pregnancy is associated with 15% to 50% maternal mortality.<sup>2</sup> These lesions are classified by the American Heart Association as World Health Organization pregnancy risk category IV (high risk of maternal morbidity and mortality).<sup>3</sup> Patients with severe MS who are contemplating pregnancy should be extensively counseled on these risks and offered transcatheter or surgical management prior to conception when possible; individuals who are already pregnant should additionally be counseled about all reproductive choice options, including termination.

This patient with moderate to severe MS after prior MV repairs presented with a severely elevated MV gradient, pulmonary hypertension, and mild heart failure symptoms in the setting of pregnancy. During pregnancy, physiologic increases in heart rate and stroke volume can lead to increased gradients across an already narrowed MV, as seen in this patient. Echocardiography also revealed a significant increase in pulmonary pressures. Given the importance of distinguishing between pulmonary arterial hypertension (a high-risk lesion in pregnancy) and pulmonary venous hypertension, a right-sided heart catheterization was considered. However, there was no significant RV dysfunction and no characteristic echocardiographic features of pulmonary arterial hypertension (eg, RV hypertrophy; reduced RV systolic function as indicated by reduced tissue Doppler S' velocities and reduced tricuspid annular plan systolic excursion; notching of the RV outflow tract pulse wave Doppler envelope).<sup>4</sup> Furthermore, the left atrium was larger than the right atrium, and the interatrial septum bowed left to right. Thus, the clinical suspicion favored pulmonary venous hypertension, and an invasive hemodynamic study was deferred.

Care of pregnant patients with valvular disease necessitates a multidisciplinary approach at an experienced center, with a care team ideally comprising cardiology, maternal-fetal medicine, obstetric and cardiac anesthesiology, interventional cardiology, and cardiac surgery. Medical management is the preferred initial approach. Selective  $\beta$ -blockers are used to reduce the heart rate and left atrial pressure.<sup>5</sup> This patient also received anticoagulation, which is recommended in pregnant patients with MS and atrial arrhythmias or high-risk features such as left atrial enlargement or history of hypercoagulability or embolic event<sup>6</sup> because of her history of atrial fibrillation.

Interventional and surgical approaches are reserved for pregnant patients who decompensate despite aggressive med-

ical therapy. Percutaneous balloon mitral valvuloplasty is safe and effective in pregnant patients with native MS<sup>7,8</sup> but has only been described in case reports after MV repair, to our knowledge.<sup>9</sup> Transcatheter MV-in-ring procedures can improve hemodynamic and functional status but has not been studied in pregnancy. MV replacement should be performed only in select cases if alternative strategies are not feasible given the high rates of fetal mortality associated with cardiopulmonary bypass.<sup>10</sup> Surgery, if necessary, should ideally be performed after delivery.

### Patient Outcome

The patient was treated initially with metoprolol and enoxaparin with close monitoring and serial echocardiography. At 26 weeks' gestation, her RV systolic pressure was 94 mm Hg and her MV gradient was 23 mm Hg at a heart rate of 76 bpm. Her symptoms were controlled until 34 weeks' gestation when she presented with worsening dyspnea, pulmonary edema, and signs of fetal distress. Although vaginal delivery is preferred in patients with MS in whom pain-induced catecholamine surges can be tempered by neuraxial analgesia, this patient ultimately underwent cesarean delivery under general anesthesia because of progressive fetal distress and maternal inability to receive neuraxial analgesia, given recent enoxaparin use. She delivered a healthy male newborn and was subsequently admitted to the cardiac intensive care unit for close hemodynamic monitoring. She was discharged on postoperative day 4, at which time her RV systolic pressure was 61 mm Hg and her MV gradient was 9 mm Hg at a heart rate of 64 bpm. Her heart failure symptoms resolved by postpartum day 14. She continues to follow up in the cardiology clinic for close monitoring of her symptoms and echocardiographic surveillance.

### ARTICLE INFORMATION

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