

## Pregnancy in a Rare and Acquired Mitral Stenosis Different from Rheumatic Mitral Involvement: Mitral Ring Mismatch

### Hamilelikte Romatizmal Mitral Tutulumdan Farklı ve Nadir Edinilmiş Bir Mitral Darlık: Mitral Ring Uyumsuzluğu

#### ABSTRACT

There is an increasing incidence of diagnosing both congenital and acquired heart valve diseases in women during pregnancy. Factors such as elevated rates of pregnancy, older maternal age, and coexisting conditions like diabetes, hypertension, and obesity contribute significantly to the symptomatic manifestation and detection of valvular heart diseases in women planning for pregnancy. Pregnancy induces cardiovascular changes across the maternal physiology, posing a heightened risk of morbidity and mortality, especially in women afflicted with valvular heart diseases. In this article, we present a case of a 25-year-old patient who underwent mitral ring annuloplasty surgery at the age of 9. Her echocardiography revealed a mismatch of the mitral ring during a follow-up examination at the age of 20. Despite neglecting further follow-ups, the patient became pregnant and presented to us at the 6<sup>th</sup> week of pregnancy. She underwent evaluations at the 6<sup>th</sup>, 10<sup>th</sup>, and 22<sup>nd</sup> weeks of pregnancy, as well as before delivery. Following the resumption of follow-up appointments, intervention planning was initiated during the postpartum period. This case exemplifies successful management, resulting in a healthy delivery after navigating through the pregnancy.

**Keywords:** Heart valve diseases, mitral ring annuloplasty, mitral stenosis, pregnancy

#### ÖZET

Hamilelik sırasında kadınlarda hem doğuştan hem de edinsel kalp kapak hastalıklarının tanısı artmaktadır. Yüksek gebelik oranları, ilerleyen anne yaşı ve diyabet, hipertansiyon ve obezite gibi eşzamanlı faktörler, gebelik planlayan kadınlarda kapak hastalıklarının semptomatik belirti gösterme ve tespitinde önemli ölçüde rol oynamaktadır. Hamilelik, anne fizyolojisinde kardiyovasküler değişikliklere neden olur ve özellikle kapak hastalığı olan kadınlarda morbidite ve mortalite riskini artırır. Bu makalede, 25 yaşında bir hastanın vaka sunumu yapılmıştır; hastamız 9 yaşında mitral yüzük anuloplasti cerrahisine tabi tutulmuştur. Ekokardiyografisi, takip muayenesinde 20 yaşında mitral ring uyumsuzluğu ortaya çıkarmıştır. Takiplerini ihmal ettiği sırada hasta gebe kalmış ve gebeliğinin 6. haftasında bize başvurmuştur. Hastamız, gebeliğinin 6., 10. ve 22. haftalarında ve doğum öncesinde değerlendirmelere tabi tutulmuştur. Takip muayenelerine yeniden başladıktan sonra, müdahale planlaması doğum sonrası dönem için planlanmıştır. Bu vaka, gebelik sürecinde başarıyla yönetimin bir örneğini sergilemektedir ve hamilelik sürecinde sorunsuz bir doğumla sonuçlanmıştır.

**Anahtar Kelimeler:** Kalp kapak hastalıkları, mitral ring anuloplasti, mitral stenoz, gebelik

Cardiovascular diseases are significant contributors to maternal morbidity and mortality, mirroring trends observed in the general population. Among these, valvular heart diseases constitute a prevalent subset.<sup>1</sup> Approximately 1% to 3% of pregnancies encounter complications from cardiovascular disease, contributing to 10% to 15% of maternal fatalities.<sup>2</sup> Although heart valve conditions typically progress favorably during pregnancy, inadequate monitoring and intervention can lead to poor outcomes.<sup>3</sup> Heart disease during pregnancy frequently correlates with maternal mortality rates. Hence, expectant mothers grappling with heart valve disease warrant collaborative evaluation by both cardiology and obstetrics specialists to navigate care decisions. Pregnancy induces considerable physiological transformations within the cardiovascular system, serving as a natural stress test that reveals latent cardiac conditions.<sup>4</sup> Particularly, pre-pregnancy assessments hold

#### CASE REPORT OLGU SUNUMU

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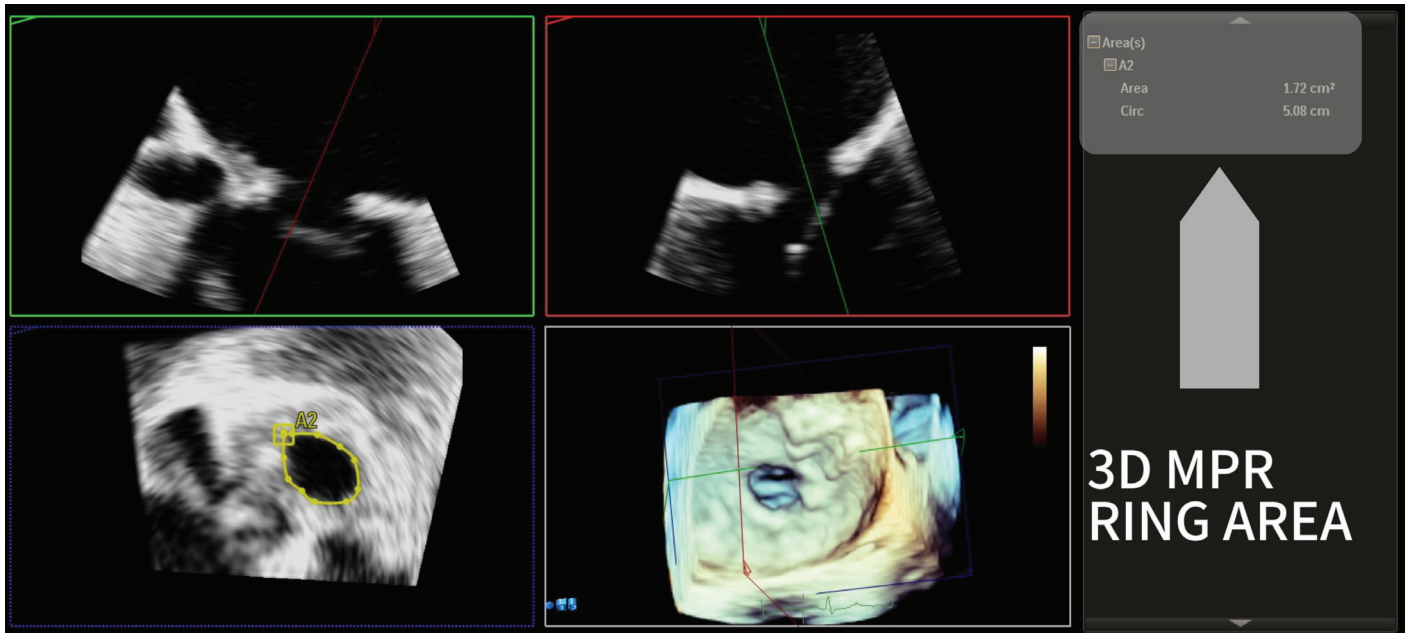
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**Figure 1. Three-dimensional mitral valve area measured on transesophageal echocardiography (TEE) at the 6<sup>th</sup> week of pregnancy. (3D MPR Ring Area, Three-Dimensional Multiplanar Reconstruction Ring Area).**

paramount importance during gestation, necessitating evaluation and treatment of pre-existing valve diseases. Significantly, women with mechanical valves or a history of repair surgery encounter increased risks of morbidity and mortality, impacting both maternal health and fetal outcomes.<sup>5</sup> Cardiac output surges by 30% to 50% in early pregnancy, peaking between the second and third trimesters. This surge is driven by augmented stroke volume initially, followed by a gradual uptick in heart rate.<sup>4</sup> During pregnancy, having left-sided obstructive lesions like mitral stenosis and aortic stenosis may result in limited cardiac reserve to withstand these hemodynamic shifts.<sup>6</sup> Hence, meticulous serial monitoring is indispensable to detect clinical deterioration during pregnancy.<sup>7</sup> Therefore, pregnant women grappling with heart valve disease necessitate close surveillance before, during, and after delivery, as increased preload during childbirth may precipitate hemodynamic destabilization postpartum. In this case, we will present a perspective on managing a pregnant woman with left-sided obstructive valve disease to provide insight into the management of the pregnancy process.

### Case Report

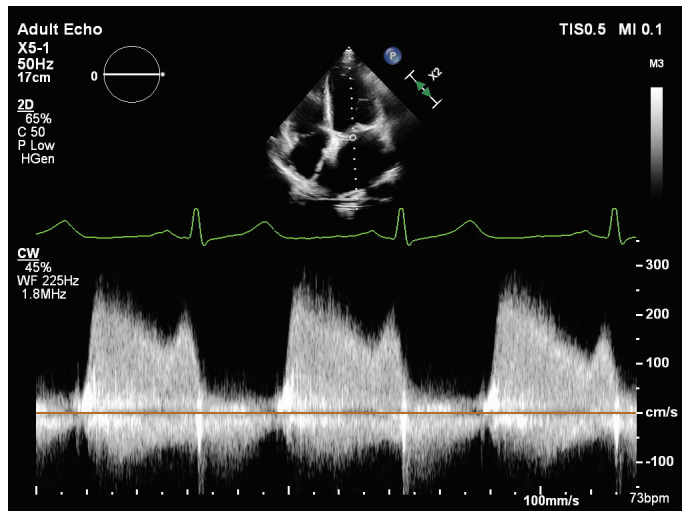
Our case study involves a 25-year-old patient who underwent mitral ring annuloplasty at the age of 9. In the patient's echocardiographic

assessment in 2019, findings were consistent with mitral ring mismatch, showing a mean gradient of 9 mmHg, a Pressure Half-Time (PHT) of 187 ms, an ejection fraction (EF) of 60%, a Systolic Pulmonary Artery Pressure (SPAP) of 31 mmHg, and a tricuspid velocity of 275 cm/s. Due to the patient's asymptomatic status, follow-up was recommended. However, subsequent to this assessment, the patient did not undergo regular follow-up. A year ago (when the patient was 24 years old), she presented to our center with severe exercise intolerance and exertional dyspnea. She also had iron deficiency anemia. Our pregnant patient had a hemoglobin (HGB) value of 8.9 g/dL and a hematocrit (HCT) of 26.7%. Her heart rate, in sinus rhythm on an electrocardiogram (ECG), was 104 beats per minute (bpm). At the same time, she was six weeks pregnant. Additionally, she expressed a strong desire to continue her pregnancy. In the subsequent echocardiographic evaluations, both transthoracic and transesophageal (Video 1), we detected an ejection fraction of 60%, SPAP of 30 mmHg, and tricuspid velocity of 274 cm/s, alongside a gradient of 24/11 mmHg and a PHT of 149 ms, with restricted posterior leaflet motion due to surgery and normal anterior leaflet movement. The patient's mitral ring area was measured as 1.72 cm<sup>2</sup> via 3-D multiplanar reconstruction (MPR) method (Figures 1 and 2). Furthermore, moderate mitral and tricuspid regurgitation were detected (Video 2) identified during the admission.

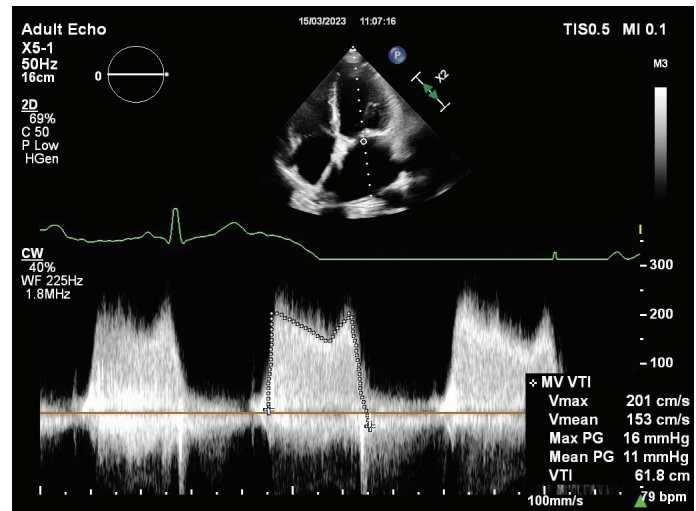
After treating iron deficiency anemia, her HGB rose to 11.9 g/dL and HCT to 35.7%. Her heart rate, in sinus rhythm on ECG, was 76 bpm; her exercise capacity increased, and her dyspnea resolved entirely. In the echocardiography performed at the 10<sup>th</sup> week, the mitral valve gradient was measured as 20/12 mmHg, with a PHT of 165 ms. These values showed improvement compared to the 6<sup>th</sup> week. Also, at the 22<sup>nd</sup> week, the gradient was 16/11 mmHg, with a PHT of 132 ms (Figure 3). The ejection fraction remained at 60%, SPAP increased to 35 mmHg, and

### ABBREVIATIONS

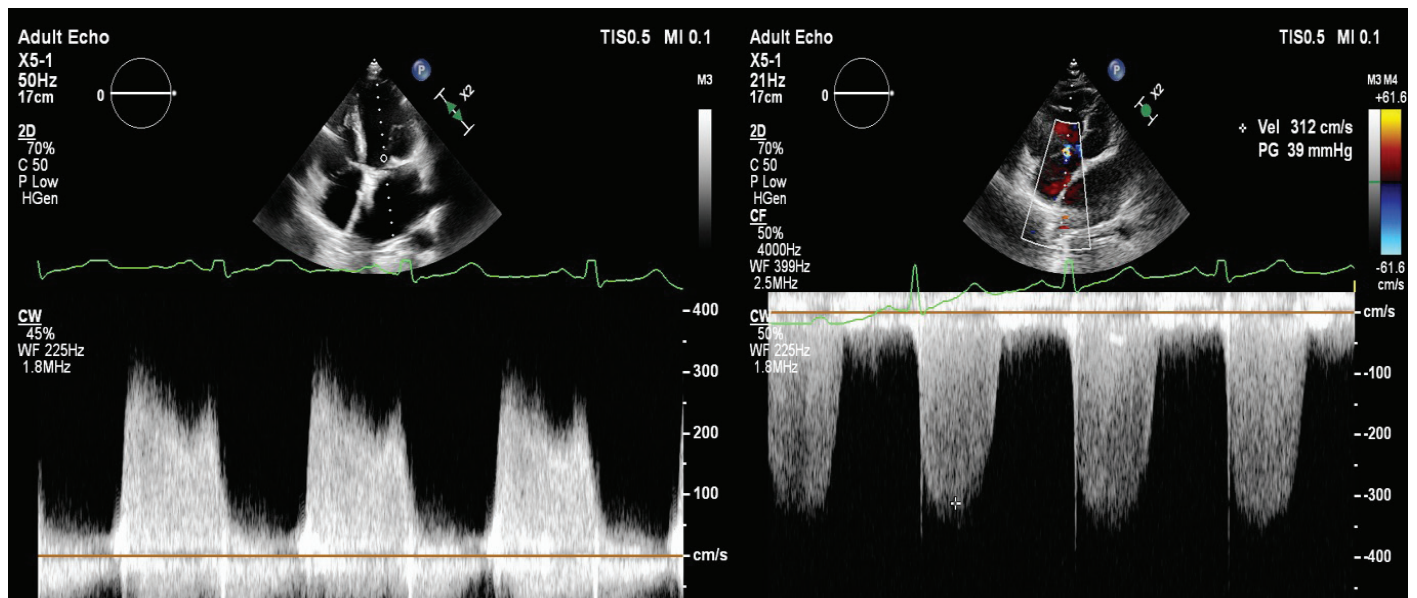
ECG	Electrocardiogram
EF	Ejection fraction
HCT	Hematocrit
HGB	Hemoglobin
MPR	Multiplanar reconstruction
MS	Mitral stenosis
NYHA	New York Heart Association
PHT	Pressure Half-Time
SPAP	Systolic Pulmonary Artery Pressure



**Figure 2.** Mitral valve gradient obtained via Continuous Wave Doppler on transthoracic echocardiography (TTE) at the 6<sup>th</sup> week of pregnancy.



**Figure 3.** Mitral valve gradient obtained via Continuous Wave Doppler over the mitral valve at the 10<sup>th</sup> week of pregnancy.



**Figure 4.** Mitral valve gradient obtained via Continuous Wave Doppler over the mitral valve, along with systolic pulmonary artery pressure (sPAP) values obtained via Continuous Wave Doppler over tricuspid regurgitation in the pre-delivery final assessment. (sPAP, Systolic Pulmonary Artery Pressure).

tricuspid velocity was 268 cm/s. Moderate mitral and tricuspid regurgitation were consistent with previous findings. Up to the last examination before delivery, the patient's symptoms were under control. The mitral valve gradient and movement were 26/14 mmHg, EF was 60%, SPAP was 39 mmHg, and tricuspid velocity was 312 cm/s (Figure 4), similar to the pre-pregnancy period (Video 3). Mitral and tricuspid regurgitations remained at a moderate level (Video 4). The SPAP did not increase during pregnancy compared to the beginning. She did not require diuretic therapy. Afterwards, our patient completed her pregnancy with a successful live birth by cesarean section due to obstetrics reasons. We recommended elective surgical intervention for the mitral valve. She is currently undergoing

further evaluations with her family as part of the decision-making process.

## Discussion

The aim of this case report is to emphasize the management of left-sided valve stenosis, which may pose challenges during pregnancy, through regular monitoring and appropriate treatment until a healthy delivery is achieved. While pregnancy is not recommended in cases of left-sided valve stenosis, unintended pregnancies do occur in clinical practice, necessitating effective management. The growing body of literature on this topic facilitates improved patient care.



**Table 1. High-Risk Cardiovascular Diseases in Pregnancy**

Patients with mitral stenosis and valve area <1.5 cm <sup>2</sup> (especially <1.0 cm <sup>2</sup> )
All symptomatic patients with severe aortic stenosis (AS)
Patients with no symptoms but impaired left ventricular ejection fraction (LVEF <50%) and severe AS
Patients with severe AS who have abnormal exercise test results, even without symptoms
Women with Marfan syndrome and an aortic diameter >45 mm (due to the high risk of aortic dissection)*
Patients with bicuspid aortic valves and an aortic diameter >50 mm (>27 mm <sup>2</sup> per body surface area [BSA]) (although the risk of dissection has not been proven)
Patients with Turner syndrome and vascular Ehlers-Danlos syndrome with an aortic diameter >25 mm/m <sup>2</sup> BSA

\*While an aortic diameter <40 mm is rarely associated with aortic dissection, there is no completely safe diameter. For aortic diameters between 40 and 45 mm, previous aortic enlargement and family history are important for counseling on pregnancy without aortic repair.

Educating all women of reproductive age with heart valve disease about the potential implications of pregnancy on their cardiac health is of paramount importance. Providing detailed information about potential symptoms during pregnancy is crucial, underscoring the significance of vigilant monitoring. Symptoms such as fatigue, shortness of breath, and palpitations, which are common during pregnancy, may mimic signs of cardiovascular diseases, posing challenges in diagnosis. Hemodynamic alterations during pregnancy may lead to the manifestation of symptoms in women with previously asymptomatic valvular heart disease, potentially exacerbating the condition's progression and symptoms. In instances where pregnancy becomes intolerable due to symptomatic aggravation, pre-pregnancy intervention may be warranted. These considerations are outlined in Table 1.

During pregnancy, various physiological changes occur, potentially leading to reduced intrauterine oxygenation, which in turn can result in complications such as intrauterine growth restriction, preterm birth, and fetal demise. These alterations not only pose risks to fetal health but also carry the potential for maternal morbidity or mortality. Of particular importance is the functional capacity reserve of the expectant mother. The period surrounding delivery is marked by increased cardiac demands due to factors like uterine contractions, anxiety, and autotransfusion from the uterine to central circulation. These dynamics elevate the risk of pulmonary hypertension, pulmonary edema, and heart failure. Consequently, the peripartum phase represents the highest risk period for maternal mortality. To illustrate, during pregnancy, pathological conditions such as anemia, which physiologically increase preload, can exacerbate symptoms significantly if left untreated. Therefore, conditions that increase cardiac load, such as anemia in our case, will lead to symptomatic improvement if treated. This will contribute to better tolerating the pregnancy. Despite these challenges, our patient exhibited tolerance to these physiological changes.

Information regarding the management of patients with a mitral ring during pregnancy is currently limited in the literature.<sup>8</sup> There is no class recommendation according to the modified World Health Organization classification about ring mismatch in the latest pregnancy guidelines. Considering the mismatch diagnosis established prior to pregnancy, a similar diagnostic approach to that employed for mitral stenosis was deemed appropriate. In our case, although intervention was not immediately necessary, decisions regarding intervention could be considered if medical management

proves insufficient during pregnancy. The second trimester is recognized as the most opportune time for invasive interventions during pregnancy.<sup>9</sup> Induction, delivery, and postpartum monitoring require the expertise of experienced cardiologists, obstetricians, and anesthesiologists. The mode of delivery should be tailored to each patient's circumstances; for example, vaginal delivery might be suitable for patients with mild mitral stenosis (MS) and no pulmonary hypertension in New York Heart Association (NYHA) class I/II, while cesarean section might be warranted for those with moderate or severe mitral stenosis, NYHA class III/IV, or pulmonary hypertension despite medical therapy.<sup>10</sup> Consequently, thorough cardiology and obstetric follow-up from preconception through postpartum stages are crucial.

## Conclusion

In summary, despite advancements in cardiac and obstetric monitoring and treatment modalities, heart valve disorders persist as a risk factor for both maternal and fetal health across the spectrum from preconception to postpartum periods. Vigilant monitoring assumes heightened significance, especially in cases akin to our patient's, characterized by clinical features resembling mitral stenosis due to a mismatched mitral ring causing left-sided obstruction. As illustrated in our case, the close surveillance and management of such patients should be integral not only during pre-pregnancy planning and throughout pregnancy but also in the postpartum period. A collaborative approach involving cardiologists and obstetricians is paramount in making informed decisions pertaining to pregnancy.

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**Video 1.** Movements of the mitral valve at the 6<sup>th</sup> week of pregnancy in our patient, visualized using transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE). (LA, Left Atrium; RV, Right Ventricle; LV, Left Ventricle; Asc. Aorta, Ascending Aorta).

**Video 2.** Mitral and tricuspid valve regurgitation observed in transthoracic echocardiography and transesophageal echocardiography imaging at the 6<sup>th</sup> week of pregnancy, with systolic pulmonary artery pressure (sPAP) calculated at 30 mmHg. (LA, Left Atrium; LV, Left Ventricle; Asc. Aorta, Ascending Aorta; sPAP, Systolic Pulmonary Artery Pressure).

**Video 3.** Evaluation of mitral valve motion in the parasternal long-axis view on TTE at the pre-delivery assessment. (LA, Left Atrium; RV, Right Ventricle; LV, Left Ventricle; Asc. Aorta, Ascending Aorta).

**Video 4.** Mitral and tricuspid valve regurgitation observed in transthoracic echocardiography at the pre-delivery assessment. (LA, Left Atrium; RA, Right Atrium; RV, Right Ventricle; LV, Left Ventricle; Asc. Aorta, Ascending Aorta).

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