CASE REPORT

Pulmonary balloon valvuloplasty in a pregnant woman with severe pulmonary stenosis

Ciddi pulmoner stenozu olan gebede pulmoner balon valvüloplasti uygulaması

Yusuf Ziya Şener, M.D., Levent Şahiner, M.D., Metin Okşul, M.D., Ergün Barış Kaya, M.D., Kudret Aytemir, M.D.

Department of Cardiology, Hacettepe University Faculty of Medicine, Ankara, Turkey

Summary– Pulmonary valvular stenosis is a rare valvular disease; it accounts for 7% to 12% of all patients with congenital heart disease. Patients with mild or moderate pulmonary stenosis (PS) are usually asymptomatic and the stenosis is often detected incidentally with echocardiography performed for another reason. Severe PS typically presents with symptoms related to increased right ventricular pressure and right heart failure. Valvular heart diseases are associated with increased morbidity and mortality in pregnancy due to cardiovascular alterations that occur during the gestational period, such as increased extracellular volume, a faster heart rate, and decreased venous return due to compression of the vena cava inferior by the enlarged uterus. While mild or moderate PS can be well tolerated in pregnancy, severe PS can lead to maternal and fetal perinatal complications. Presently described is the case of a pregnant patient with severe PS who successfully underwent balloon valvuloplasty in the third trimester.

Özet– Pulmoner stenoz nadir bir valvüler hastalık olup tüm doğuştan kalp hastalıklarının %7–%12'sini oluşturur. Hafif ve orta derecede pulmoner stenoz genellikle semptomsuz olup hastalar sıklıkla başka nedenle yapılan ekokardiyografik değerlendirme sırasında tesadüfen tanı alırlar. Ciddi pulmoner stenoz ise artmış sağ ventrikül basıncı ve sağ ventrikül yetersizliği ilişkili semptomlar ile kendini gösterir. Gebelik sürecinde kardiyovasküler sistemde kalp hızında ve ekstraselüler sıvıda artış ile birlikte büyüyen uterusun inferiyor vena kavaya basısı sonucu venöz geri dönüşte azalma gibi fizyolojik değişiklikler meydana gelir ve bu nedenle kapak hastalıkları gebelerde artmış kardiyovasküler morbidite ve mortalite ile ilişkilidir. Hafif ve orta derecede pulmoner stenoz gebelikte iyi tolere edilirken ciddi pulmoner stenoz maternal ve fetal perinatal komplikasyonlarda artış ile ilişkilidir. Bu yazıda üçüncü trimesterde başarılı pulmoner balon valvüloplasti uygulanan ciddi pulmoner stenozlu bir gebe hasta sunduk.

Pulmonary valvular stenosis is a rare congenital heart disease and is usually diagnosed in adulthood incidentally because of a silent clinical course. Pulmonary stenosis (PS) is well tolerated in pregnancy even when severe, and is associated with typically minor fetomaternal complications. Percutaneous valvular balloon valvuloplasty, which was first performed by Dr. Jean S. Kan in 1982 is the principal treatment option for pulmonary valvular stenosis. Intervention should be considered in pregnant women with severe PS or moderate PS with accompanying symptoms.[1] This report describes the case of a pregnant woman with severe PS who underwent pulmonary balloon valvuloplasty in the third trimester and had a healthy birth without peripartum complications.

Abbreviations:

PS	*Pulmonary stenosis*
RV	*Right ventricle*

CASE REPORT

A 23-year-old pregnant woman was referred to the outpatient clinic for a routine cardiac evaluation before delivery. Her medical history revealed no known disease. This was her first pregnancy and she was in the 34th week of gestation. She described moderate exertional

Received: September 29, 2018 *Accepted:* December 17, 2018

Correspondence: Dr. Yusuf Ziya Şener. Hacettepe Üniversitesi Tıp Fakültesi, Kardiyoloji Anabilim Dalı, Ankara, Turkey.
Tel: +90 312 - 305 28 15 e-mail: yzsener@yahoo.com.tr

dyspnea and her functional class was graded New York Heart Association class II. Her vital signs and physical examination were unremarkable, other than mild pretibial edema, which may occur during pregnancy, and a systolic ejection murmur at the pulmonary valve location. A complete blood count and kidney and liver function test results were all in the normal range. An electrocardiogram demonstrated a sinus rhythm with right axis deviation and was consistent with right ventricular (RV) hypertrophy (Fig. 1). Echocardiography was performed and severe PS with normal systolic left ventricular function was established. RV hypertrophy (RV free wall thickness: 1.6 cm), RV dilatation (RV diastolic diameter at mid region: 4.3 cm), a D-shaped left ventricle and post-stenotic dilatation of the pulmonary artery (pulmonary artery diameter: 4.5 cm) were detected (Video 1*). The pulmonary valvular peak gradient was 122 mmHg, with a mean gradient of 68 mmHg (Fig. 2a). The patient was informed about the possible complications of severe PS that could occur during delivery despite being asymptomatic. Pulmonary balloon valvuloplasty was suggested and she consented. Right heart catheterization was performed with lead protection for the fetus in order to estimate basal pulmonary artery and RV pressure (pulmonary artery pressure: 30/10 mmHg; RV pressure: 120/12 mmHg). After catheterization, a super stiff wire was used to pass the stenotic pulmonary valve and the valve was dilated with a 22x50-mm pulmonary balloon (VACS dilatation balloon catheter; Osypka Medical GmbH, Berlin, Germany) (Video 2/3*). The pulmonary artery and RV pressure were reassessed. The gradient at the pulmonary valve was 50 mmHg and the RV pressure had decreased to 80/12 mmHg. A post interventional echocardiography was performed and a decrease in the pulmonary valvular gradient was confirmed (peak: 48 mmHg; mean: 26 mmHg) (Fig. 2b). The left ventricular eccentricity index improved as a result of decreased right ventricular pressure (Video 4*). The patient subsequently gave birth by cesarean section without any perinatal complications and was discharged per normal.

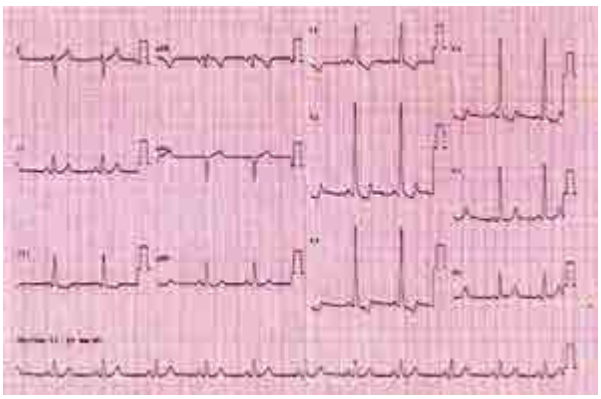

Figure 1. Electrocardiogram results consistent with right axis deviation and right ventricular hypertrophy with a normal sinus rhythm.

DISCUSSION

Pulmonary valvular stenosis is a rare type of congenital heart disease that accounts for 7% to 12% of all cases.[2] Morphologically, there are 6 types of PS: dome-shaped (the most common), tricuspid, bicuspid, hypoplastic annulus, unicommissural, and dysplastic.[3] Mild and moderate PS is usually asymptomatic and often detected incidentally by echocardiography. Patients with mild or moderate PS have a similar

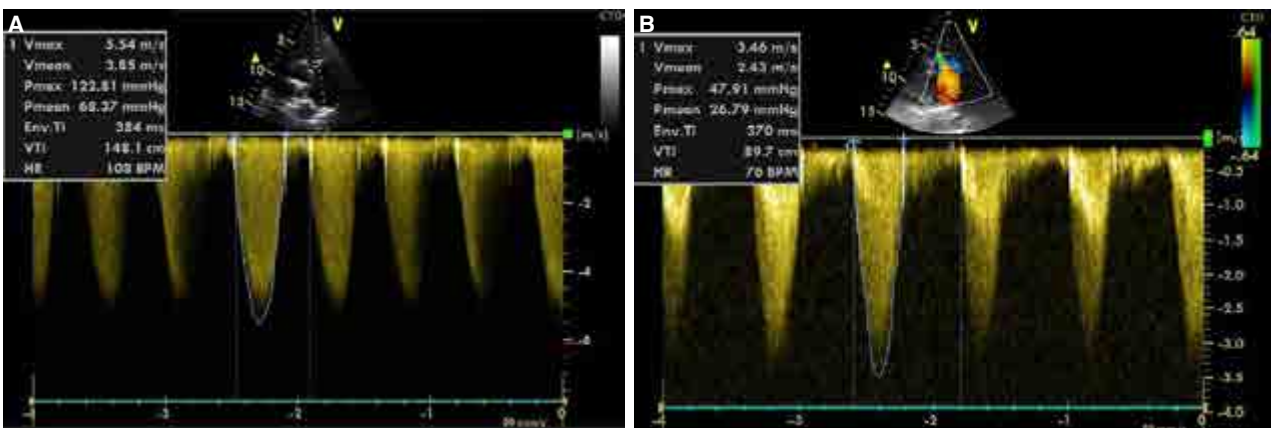

Figure 2. (A) Baseline peak pulmonary valvular gradient of 122 mmHg and mean of 68 mmHg; **(B)** Pulmonary valvular gradient significantly decreased after balloon valvuloplasty.

survival length compared with healthy counterparts. Severe PS causes RV hypertrophy and is related to poor long-term outcomes unless treated with valvotomy or balloon valvuloplasty.[4] The severity of PS is determined by the peak gradient across the pulmonic valve as estimated using echocardiography. Mild PS is defined as a peak gradient of <36 mmHg, moderate PS is a peak gradient of 36–64 mmHg, and severe PS is described as peak gradient of >64 mmHg. Intervention is recommended in patients with severe PS, regardless of the patient's symptom status, while intervention should be offered in patients with mild or moderate disease when the patient is symptomatic or RV function deterioration has developed. Balloon valvuloplasty is the preferred first-line treatment option in patients with valvular or peripheric PS, unless dysplastic pulmonary valvular morphology is present. Surgical valvotomy should be considered in patients not suited to balloon valvuloplasty.[5]

Cardiovascular alterations, such as an increase in plasma volume, cardiac output, and heart rate may occur in pregnancy, and these changes can lead to morbidity in patients with background cardiovascular diseases. Congenital heart diseases are the most common cardiovascular disorders presenting in pregnancy in Western countries, while rheumatic valvular disease is the most common in developing countries. PS is usually detected incidentally during pregnancy due to a typically silent clinical course. Although severe PS is generally well tolerated in pregnancy, it may result in fetomaternal peripartum complications.[6] Yet Hameed et al.[7] reported that fetal outcomes, including the Apgar score and birth weight, were similar in new mothers with mild and severe PS to those of the controls. It has also been reported that vaginal delivery is a safe option in patients with severe PS. Nonetheless, percutaneous balloon valvuloplasty can reduce peripartum complications and the American Heart Association/American College of Cardiology practice guidelines recommend balloon valvuloplasty in pregnant women with a peak pulmonary gradient of more than 40 mmHg.[2]

In our case, a decrease in pulmonary gradient was achieved with successful percutaneous pulmonary balloon valvuloplasty. Pregnant women with severe PS and those who are symptomatic with moderate PS should be treated in order to reduce fetomaternal complications and improve the likelihood of an optimal pregnancy outcome.

**Supplementary video file associated with this article can be found in the online version of the journal.*

Peer-review: Externally peer-reviewed.

Conflict-of-interest: None.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Authorship contributions: Concept: K.A.; Design: L.Ş.; Supervision: L.Ş.; Materials: E.B.K.; Data collection: M.O.; Literature search: Y.Z.Ş; Writing: Y.Z.Ç.

REFERENCES

1. Presbitero P, Prever SB, Brusca A. Interventional cardiology in pregnancy. Eur Heart J 1996;17:182–8.[CrossRef]
2. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;52:e143–e263. [CrossRef]
3. Gikonyo BM, Lucas RV, Edwards JE. Anatomic features of congenital pulmonary valvar stenosis. Pediatr Cardiol 1987;8:109–16. [CrossRef]
4. Hayes CJ, Gersony WM, Driscoll DJ, Keane JF, Kidd L, O'Fallon WM, et al. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvar stenosis. Circulation 1993;87(2 Suppl):I28–37.
5. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al; Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for Practice Guidelines (CPG).ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31:2915–57. [CrossRef]
6. Oylumlu M, Aykent K, Soydinc HE, Oylumlu M, Ertas F, Ozer HO, et al. Pulmonary Balloon Valvuloplasty during Pregnancy. Case Rep Cardiol 2012;2012:353168. [CrossRef]
7. Hameed AB, Goodwin TM, Elkayam U. Effect of pulmonary stenosis on pregnancy outcomes-a case-control study. Am Heart J 2007;154:852–4. [CrossRef]

Keywords: Balloon valvuloplasty; pregnancy; pulmonary stenosis.

Anahtar sözcükler: Balon valvüloplasti; gebelik; pulmoner stenoz.