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Case Report

# Pulmonary Valvar Stenosis from the Fetal to the Infantile Period: A Case Report

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## ABSTRACT

***Background:*** Fetal echocardiography is a useful tool for diagnosing fetuses with congenital heart diseases, and it is best to be conducted between 17 and 19 weeks of gestational age. However, fetal echocardiography can be performed at other ages of pregnancy for a variety of reasons. This study describes one fetus with pulmonary valvar stenosis based on the fetal echocardiogram in the uterus.

***Case report:*** This study describes one fetus with pulmonary valve stenosis based on the fetal echocardiogram in the uterus. We referred the family to a hospital with neonatal intensive care unit admission. After birth, we followed her serially and confirmed pulmonary valve stenosis, which increased in severity after two months. Subsequently, we performed a percutaneous balloon valvuloplasty.

***Conclusion:*** Our findings showed that some cardiac defects could vary in severity during pregnancy and post-birth. There was clear evidence that pulmonary valvar stenosis was a lesion developed during the fetus's lifetime to tolerate the lesion. Although pulmonary stenosis progressed in the early months after birth, it was easily treated through balloon angioplasty.

***Keywords:*** Fetal, Infantile, Pulmonary stenosis

## Introduction

The prevalence of congenital heart disease (CHD) is 5 to 8 per 1,000 live births, and fetal echocardiography assists in prenatal diagnosis (1). Pulmonary valve stenosis is one of the most common congenital heart defects, the overall incidence of which is 0.5-1 per 1,000 births. Pulmonary artery stenosis means a mixed defect with a wide variety of morphology and etiology, and various types of genetic syndromes can be associated with CHD (2). It can occur in isolated form or combination with other congenital cardiac malformations (3). There are three types of pulmonary stenosis, namely subvalvular, valvar, and supravalvar forms. Its diagnosis could be made during fetal life by echocardiography. If it is critical pulmonary valve stenosis (PS) in the fetus that warrants intervention, it can be accomplished; however, it is accompanied by a risk of fetal failure. Fetal intervention using balloon valvuloplasty can increase blood flow over a severely obstructed valve in some fetuses, resulting in improved ipsilateral heart growth (4). During fetal life, the degree of stenosis may progress to the end of pulmonary atresia or may be sustained or even resolve (5).

## Case report

A 27-year-old woman was referred to our pediatric cardiology unit for fetal echocardiography by her gynecologist since her sonography results revealed a suspicious gradient. A pediatric cardiologist achieved all cardiac segments (four-

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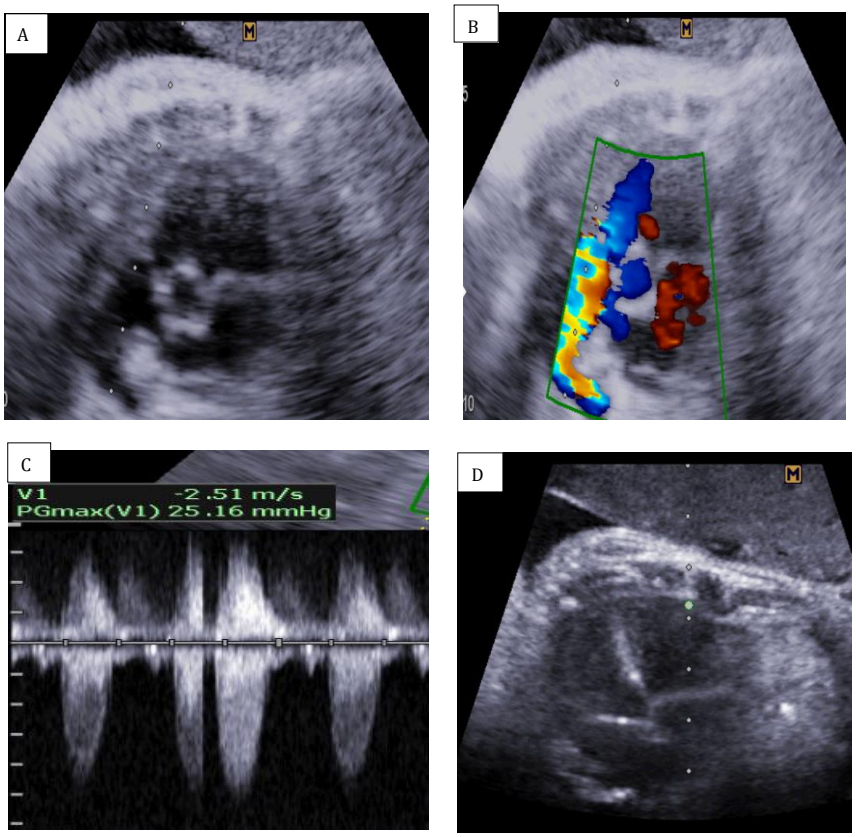


**Figure 1.** Neonatal echocardiogram of pulmonary valve stenosis
A) a 2D short axis demonstrating the stenosis of the pulmonary valve
B) a short axis color echocardiogram displaying turbulence through the pulmonary valve
C) the gradient of the pulse wave through the pulmonary valve (25 mmHg)
D) a 4-chamber showing right ventricle enlargement

chamber view, the long axis of ventricles, short axis, aortic arch planes, and ductus arteriosus) in fetal echocardiography. As shown in Figure 1, there was a mild enlargement of the right atrium (RA) and right ventricle (RV) and a two-dimensional view of PS. When Doppler imaging was used, we observed the turbulent flow through the stenotic pulmonary valve, and the pressure gradient between the two sides of the stenosis was 25 mmHg. Doppler did not show tricuspid regurgitation.

The diagnosis was PS with an intact ventricular septum. A pediatrician referred the family to a hospital with a neonatal intensive care unit (NICU) and first-line care for neonates with CHD, such as prostaglandin E1, cardiac intervention. A 3,300-g baby girl was born at the end of the day. An echocardiogram on the first day of the neonatal period revealed moderate PS with an intact interventricular septum and patent ductus arteriosus (PDA) (Figure 2). She was following up

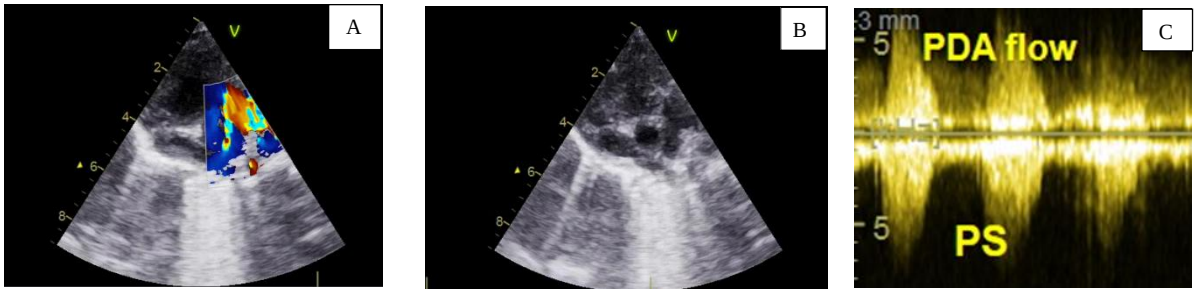


**Figure 2.** Transthoracic echocardiography of 1-day-old neonate with pulmonary valve stenosis
A) a 2D of pulmonary valve stenosis
B) turbulence through pulmonary valve stenosis and patent ductus arteriosus
C) gradient through pulmonary valve stenosis and continuous flow over patent ductus arteriosus

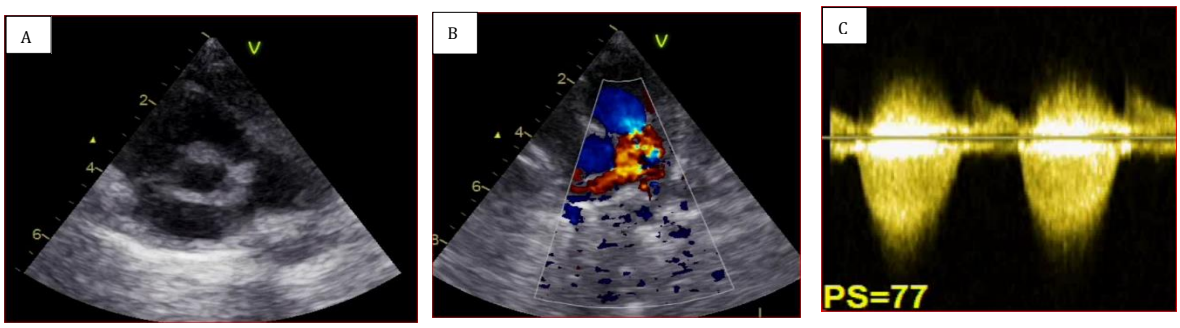


**Figure 3.** Transthoracic echocardiography of 4-month-old neonate with pulmonary valve stenosis
A) a 2D short axis demonstrating stenosis and pulmonary valve dysplasia
B) a turbulent color interrogation through dysplastic pulmonary valve
C) Doppler echocardiogram of turbulence through pulmonary valve indicating severe pulmonary valve stenosis

to two months of age. On physical examination, she had grade 3/6 systolic ejection murmur at the left upper sternal border. Two-dimensional transthoracic echocardiography showed severe PS with an instantaneous peak gradient of 77 mmHg and a closed PDA (Figure 3).

Cardiac catheterization confirmed severe PS with an intact interventricular septum. During angiography, the RV pressure was 90/0-10 mmHg, and the pulmonary artery pressure was 20/10 mmHg, resulting in a peak gradient around the pulmonary valve of 70 mmHg. Percutaneous balloon pulmonary valvuloplasty was performed using an Ever cross 10×20 mm balloon (Figure 4). After that, the RV pressure was 50/0-10 mmHg. It indicated a 60% reduction in the transpulmonary gradient after the intervention. She was discharged home after the intervention, and the transpulmonary gradient was 25 mmHg at the clinical follow-up two weeks and two months later.

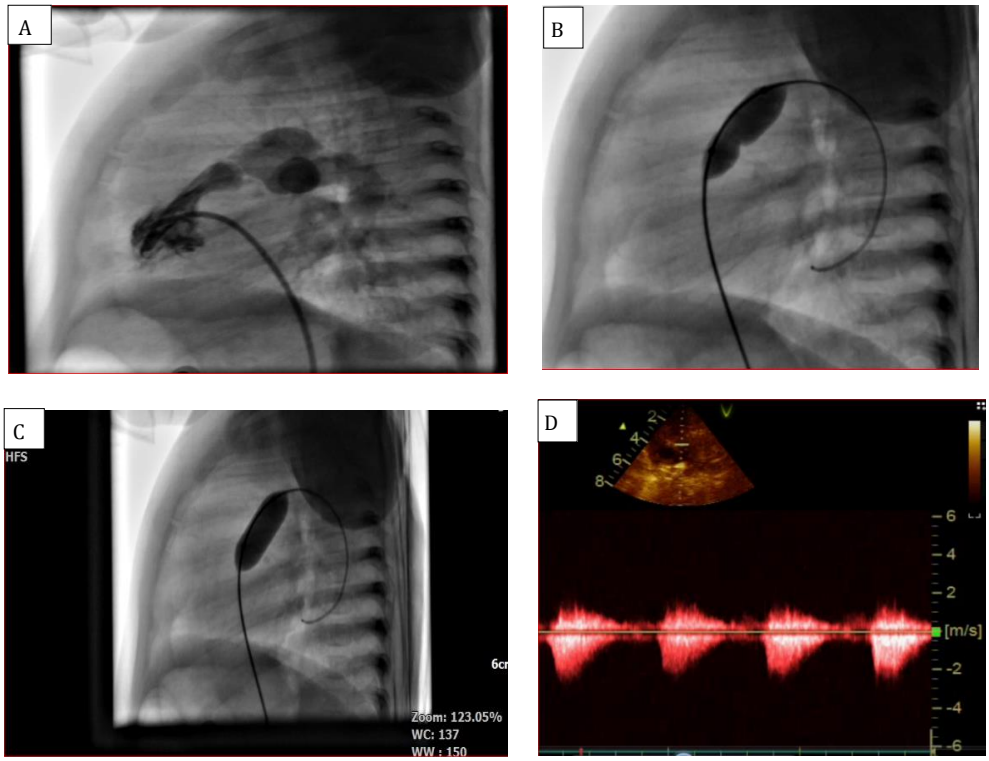


**Figure 4.** Percutaneous balloon pulmonary valvuloplasty of dysplastic valve
A) lateral view of the heart during angiography; right ventricle injections displaying severe valvar pulmonary valve stenosis
B) a lateral view showing the balloon wasting during angioplasty using a 10×20 mm balloon
C) a lateral view after percutaneous balloon pulmonary valvuloplasty showing a successful percutaneous balloon pulmonary valvuloplasty with Ever cross balloon over Extra stiff wire
D) Doppler interrogation through pulmonary valve showing minimal pulmonary valve stenosis

## Discussion

PS can be diagnosed by a pediatric cardiologist in the fetal life. Fetal ventricular outflow obstruction can progress before birth due to the nature of the fetal circulation. It depends mostly on the timing of development, the severity of stenosis, the influence of stenosis on the fetal atrioventricular valve, and myocardial function (6).

In the case of pulmonary stenosis, the lesion can exhibit noticeable variability (7, 8). The RV could be small, normal, or dilated. In addition, the main pulmonary artery can be normal or small. The ductus arteriosus can be normal or narrow. These varying types of manifestations contribute to the influence of different expressions of disease in different fetal ages. In our patient, there was a PS with a tortuous ductus arteriosus that tells us that pulmonary stenosis was developed early in fetal life, resulting in a turbulent flow across the ductus arteriosus which made it tortuous. If PS occurred late in the fetal period, the ductus arteriosus would be straighter (9, 10).

Mild or moderate PS in fetal life is firstly doubted by the finding of a doming pulmonary valve and dilated main pulmonary artery, usually progressing to more severe stenosis due to low annular growth (11). As it is a slow trend, this is tolerated by increasing gradients through the pulmonary valve as can be measured by Doppler. These increasing gradients occur in right ventricular hypertrophy to maintain a satisfactory output several months before birth. However, if the stenosis is severe, the systolic right ventricular function can decrease. Following that, backflow through the ductus arteriosus can be detected by echocardiography, and we would have critical PS, which deserved balloon intervention soon after birth (12).

PS during pregnancy has different severities that can cause severe right ventricular failure due to heart failure. In some cases, the severity of stenosis changes during pregnancy and can be assessed by serial fetal echocardiography. Serial follow-up is performed on these patients. In these cases, it is essential to collaborate with pediatric cardiologists, perinatologists, and neonatologists. Usually, it is safer for the patient to be born in a hospital equipped with NICU. Progression or regression of stenosis is unpredictable and can become worse/better, or unchanged (13).

In our patient, the pattern of valvar pulmonary stenosis remained unchanged during pregnancy and was not associated with right ventricular failure and tricuspid regurgitation. After the neonate's birth, she was admitted to the NICU with a suitable general condition. On examination, mild PS was observed without cyanosis and tricuspid valve insufficiency, and a decision took to follow up the patient. In serial echocardiography, due to increased stenosis severity, interventional angiography (balloon valvuloplasty) was performed, and the condition of the patient was completely normal at the follow-up. Some CHDs, particularly semilunar valve stenosis, could be progressive even after birth. It can be noted that mild stenosis in a young infant can be increased in severity. It is impossible to predict who would have developed more stenosis over time (5, 14).

## Conclusion

Accordingly, this case report provides clear evidence that valvar pulmonary stenosis is a lesion acquired during the lifetime of the fetus, and the fetus can tolerate the lesion. We also had PS progression months after birth that could be easily treated with balloon angioplasty.

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## Conflicts of interest

The authors declare that they have no conflict of interest.

## References

1. Bravo-Valenzuela NJ, Peixoto AB, Araujo Júnior E. Prenatal diagnosis of congenital heart disease: A review of current knowledge. Indian Heart J. 2018;70(1):150-64.
2. Straub L, Huybrechts KF, Bateman BT, Mogun H, Gray KJ, Holmes LB, et al. The Impact of Technology on the Diagnosis of Congenital Malformations. Am J Epidemiol. 2019;188(11):1892-901.
3. Liu L, Wang H, Cui C, Li Y, Liu Y, Wang Y, et al. Prenatal echocardiographic classification and prognostic evaluation strategy in fetal pulmonary atresia with intact ventricular septum. Medicine. 2019;98(42):e17492.
4. Edwards LA, Justino H, Morris SA, Rychik J, Feudtner C, Lantos JD. Controversy About a High-Risk and Innovative Fetal Cardiac Intervention. Pediatrics. 2018;142(3):e20173595.
5. Latson LA. Critical pulmonary stenosis. J Interv Cardiol. 2001;14(3):345-50.
6. Yamamoto Y, Hornberger LK. Progression of outflow tract obstruction in the fetus. Early Hum Dev. 2012;88(5):279-85.
7. Gottschalk I, Strizek B, Menzel T, Herberg U, Breuer J, Brockmeier K, et al. Severe Pulmonary Stenosis or Atresia with Intact Ventricular Septum in the Fetus:

The Natural History. Fetal Diagn Ther. 2020; 47(5):420-428.
8. Masura J, Burch M, Deanfield JE, Sullivan ID. Five-year follow-up after balloon pulmonary valvuloplasty. J Am Coll Cardiol. 1993;21(1):132-6.
9. Elzenga NJ, Gittenberger-de Groot AC. The ductus arteriosus and stenoses of the pulmonary arteries in pulmonary atresia.Int J Cardiol. 1986;11(2):195-208.
10. Arlettaz R, Archer N, Wilkinson AR. Closure of the ductus arteriosus and development of pulmonary branch stenosis in babies of less than 32 weeks gestationArch. Dis. Child. Fetal Neonatal Ed. 2001;85(3):F197-200.
11. Wang JK, Wu MH, Lee WL, Cheng CF, Lue HC. Balloon dilatation for critical pulmonary stenosis. Int J Cardiol. 1999;69(1):27-32.
12. Khalil M, Jux C, Rueblinger L, Behrje J, Esmaeili A, Schranz D, et al. Acute therapy of newborns with critical congenital heart disease. Transl Pediatr. 2019;8(2):114-26.
13. Bronshtein M, Blumenfeld Z, Khoury A, Gover A. Diverse outcome following early prenatal diagnosis of pulmonary stenosis. Ultrasound Obstet Gynecol. 2017;49(2):213-8.
14. Zucker EJ. Cross-sectional imaging of congenital pulmonary artery anomalies. The Int J Card Imaging. 2019;35(8):1535-48.