Gac Méd Caracas 2023;131(4):996-1001
DOI: 10.47307/GMC.2023.131.4.22

Managing Challenges in a Symptomatic Patient with Moderate Pulmonary Stenosis: A Case Report

Manejo de los Desafíos en un Paciente Sintomático con Estenosis Pulmonar Moderada: Reporte de un Caso

Andi Alief Utama Armyn[1a], Yulius Patimang[2a], Anugrah Mahadewa Putra[3a], Muzakkir Amir[4a], Idar Mappangara[5a], Andriany Qanitha[6,a,b*]

SUMMARY

Introduction: *Pulmonary stenosis is a common form of obstruction in the right ventricular outflow tract, characterized by the narrowing of the pulmonary valve, which results in obstructed blood flow.* **Case Presentation:** *A 17-year-old female presented with recurring shortness of breath, chest pain, and palpitations, especially during physical activity. On examination, a grade III/VI continuous murmur was heard at the upper left sternal border. Echocardiography confirmed moderate pulmonary valve stenosis, a left-to-right patent ductus arteriosus (PDA), and mild to moderate pulmonary regurgitation. Treatment involved intravenous normal saline, ceftriaxone premedication at 2 grams/24 hours intravenously, and oral propranolol at 10 mg every 8 hours. Subsequently, a successful balloon pulmonary valvuloplasty (BPV) was performed, and the patient's symptoms improved.* **Conclusion:** *Timely intervention with BPV is the primary therapeutic approach, emphasizing the importance of early treatment, irrespective of symptom onset.*

Keywords*: Balloon pulmonary valvuloplasty, congenital heart disease, case report, pulmonary stenosis, symptomatic moderate pulmonary stenosis.*

RESUMEN

Introducción: *La estenosis pulmonar es una forma común de obstrucción en la vía de salida del ventrículo derecho, caracterizada por el estrechamiento de la válvula pulmonar, lo que resulta en un flujo sanguíneo obstruido.* **Presentación del caso:** *Una joven de 17 años se presentó con episodios recurrentes de*

DOI: https://doi.org/10.47307/GMC.2023.131.4.22

ORCID: 0000-0002-4914-3542[4]
ORCID: 0000-0003-1543-1519[5]
ORCID: 0000-0003-2420-0560[6]

[a]Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Hasanuddin, Makassar 90245, Indonesia.
[b]Department of Physiology, Faculty of Medicine, Universitas Hasanuddin, Makassar 90245, Indonesia.

Recibido: 8 de octubre 2023
Aceptado: 13 de noviembre 2023

[1]MD, FIHA. E-mail: andi.alief@gmail.com
[2]MD, FIHA. E-mail : yulio_patimang@yahoo.co.id
[3]MD. E-mail: anugrahmahadewa@gmail.com
[4]MD, PhD, FIHA. E-mail: dr.muzakkir@gmail.com
[5]MD, PhD, FIHA, FINASIM. E-mail : idar.mks@yahoo.com

*Corresponding author: Andriany Qanitha, MD, MSc, PhD
Faculty of Medicine, University of Hasanuddin Jl. Perintis Kemerdekaan Km. 10, Makassar 90245, South Sulawesi, Indonesia
E-mail: myaqanitha@gmail.com
Tel.: +6281282816699

falta de aire, dolor en el pecho y palpitaciones, especialmente durante la actividad física. En la exploración física, se escuchó un soplo continuo de grado III/VI en el borde esternal izquierdo superior. La ecocardiografía confirmó una estenosis moderada de la válvula pulmonar, un conducto arterioso permeable de izquierda a derecha (PDA) y una regurgitación pulmonar leve a moderada. El tratamiento incluyó suero fisiológico intravenoso, premedicación con ceftriaxona a 2 gramos/24 horas intravenosamente y propranolol oral a 10 mg cada 8 horas. Posteriormente, se realizó con éxito una valvuloplastia pulmonar con balón (BPV), y los síntomas de la paciente mejoraron. **Conclusión:** *La intervención oportuna con BPV es el enfoque terapéutico primario, enfatizando la importancia del tratamiento temprano, independientemente del inicio de los síntomas.*

Palabras clave: *Valvuloplastia pulmonar con balón, enfermedad cardíaca congénita, informe de caso, estenosis pulmonar, estenosis pulmonar moderada sintomática.*

INTRODUCTION

Right ventricular outflow tract (RVOT) obstruction is an anatomic blockage in the right ventricular output and is typically caused by pulmonary valve stenosis, a condition characterized by the narrowing of the pulmonary valve, leading to restricted blood flow. This ailment is most frequently congenital, generally benign, and can affect both pediatric and adult patients (1,2).

While pulmonary stenosis may occur independently in 8 %-10 % of congenital heart diseases, it is frequently observed in conjunction with other congenital anomalies. Congenital pulmonary stenosis is commonly associated with genetic syndromes like Noonan syndrome, Alagille syndrome, Williams syndrome, and congenital rubella. Additionally, it can manifest as an acquired condition, resulting from factors such as rheumatic heart disease, carcinoid syndrome, infective endocarditis, or trauma. Pulmonary valve diseases often coincide with pulmonary regurgitation, attributed to valve irregularities or prior interventions (2).

Therapeutic strategies are determined based on the hemodynamic severity of the obstruction. In cases of severe pulmonary stenosis, intervention is warranted. Furthermore, it is essential to consider intervention in instances of non-severe stenosis accompanied by symptoms such as congestive heart failure, right-to-left interatrial cyanosis, and exercise intolerance. Balloon pulmonary valvuloplasty (BPV) stands as the primary intervention for pulmonary stenosis and should be promptly administered upon diagnosis, regardless of the presence of symptoms (3-5). In this present case, we report our approach to managing symptomatic moderate pulmonary stenosis in a young female patient.

Case Illustration

A 17-year-old female patient has presented with a chief complaint of experiencing shortness of breath, especially during physical activities. Additionally, the patient reports a history of chest pain and palpitations over the past two years, with intermittent symptoms. The patient has a background of recurrent cough, fever, chest pain, and palpitations. Furthermore, the patient has been diagnosed with a congenital heart disease since the year 2020 and is currently undergoing propranolol treatment. There is no evidence of cyanosis, and there is no family history of congenital heart disease.

The patient is the oldest among four siblings, born after a full-term pregnancy, and cried immediately upon birth. She had a birth weight of over 2500 grams. During the pregnancy, the mother attended regular hospital check-ups, followed a vitamin regimen, and consistently took iron supplements.

Upon physical examination, the patient presents with a weight of 44 kg and a height of 163 cm. She is alert, and fully oriented, with vital signs within the normal range, including a blood pressure of 115/66 mmHg, a heart rate of 86 beats per minute, a respiratory rate of 24 breaths per minute, a temperature of 36.8°C, and oxygen saturation consistently ranging between 97 %-99 % in all extremities while breathing room air. There were no observable signs of jaundice or anemia during the head examination. The chest x-ray displayed normal findings (Figure 1).

Echocardiography findings indicated the presence of moderate valvular pulmonary

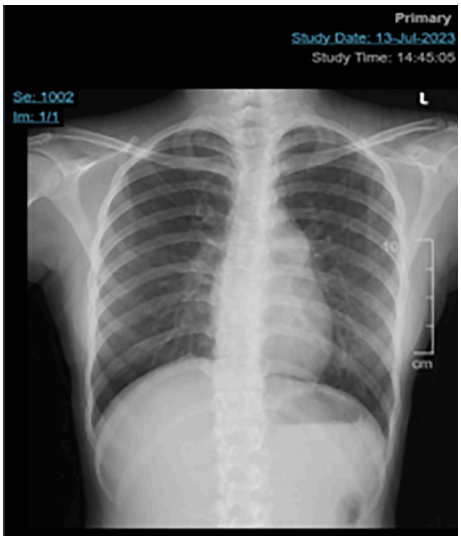

Figure 1. Chest X-ray examination of this patient.

stenosis (maximum pressure gradient (maxPG) of 46 mmHg, a left-to-right shunt through a patent ductus arteriosus (PDA) (sized 2.1 mm, with systolic PG of 12 mmHg and diastolic PG of 2 mmHg), and mild to moderate pulmonary regurgitation (PR PHT 151 ms, width >1/3 RVOT) (Figure 2).

During auscultation, vesicular breath sounds were observed, with no signs of rhonchi, wheezing, or retractions. Cardiac examination revealed a single S1 and a continuous murmur graded as III/VI at the upper left sternal border. Abdominal examination showed no dullness upon percussion and no organomegaly. Examination of the extremities did not reveal any peripheral edema, cyanosis, or clubbing of the fingers. Table 1 provides the laboratory findings of the patient conducted on July 13, 2023.

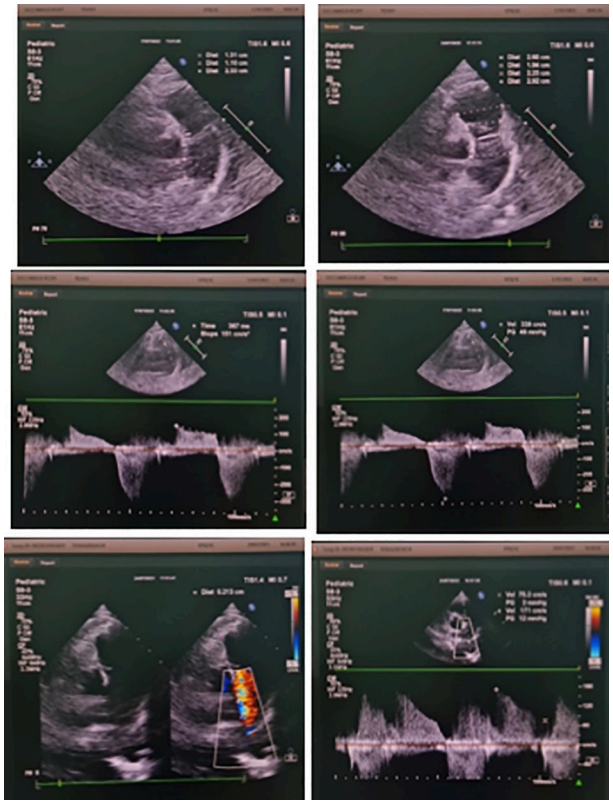

Figure 2. Echocardiography findings of the patient.

Table 1

Laboratory findings in this patient

Laboratory	Results	Normal range
Routine Haematology		
WBC	6.1	4.00 – 10.0
RBC	4.92	4.00 – 6.00
HGB	14.1	12.0 – 16.0
HCT	42	37.0 – 48.0
PLT	380	150 – 400
NEUT	49.6	52.0 – 75.0
LYMPH	41.4	20.0 – 40.0
Coagulation		
INR	1.05	-
PT	11.3	10-14
APTT	31.3	22.0 – 30.0
Blood Chemistry		
Urea	19	10 – 50
Creatinine	0.88	M(<1.3);F(<1.1)
ALT	22	< 38
AST	20	< 41
Electrolyte		
Sodium	140	136 – 145
Potassium	4.0	3.5 – 5.1
Chloride	107	97 – 111
Hepatitis Markers		
Hbs Ag (Elisa)	Non-Reactive	Non-Reactive
Anti-HCV (Elisa)	Non-Reactive	Non-Reactive

Previously, the patient underwent an RHC procedure on April 12, 2023. The RHC findings indicated the presence of moderate subvalvar pulmonary stenosis and a left-to-right shunt through the PDA characterized by low flow and low resistance. A BPV procedure with an estimated balloon size of 25 to 28 mm was scheduled. During hospitalization, the patient received intravenous administration of normal saline (NaCl 0.9 %) at a rate of 500 cc over 24 hours, intravenous ceftriaxone at a dosage of 2 grams every 24 hours as premedication, and oral propranolol at a dosage of 10 mg every 8 hours. The patient then underwent the BPV procedure on July 26, 2023. Following the RHC, which revealed the presence of pulmonary stenosis (pulmonary artery annulus 20.75 mm) and a small PDA, the BPV procedure was successfully performed using a nucleus balloon sized 25 x 40 mm. Oxygen saturation improved from 76 % to 91 % at the pulmonary artery (Figure 3).

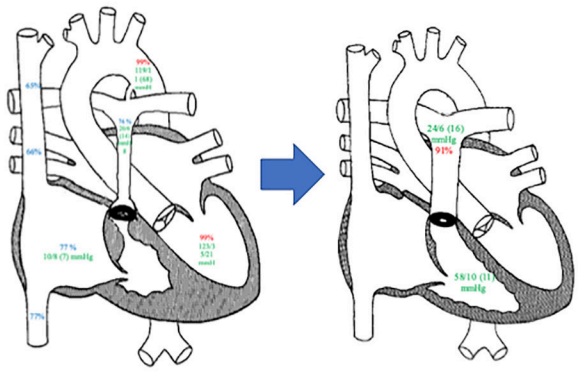

Figure 3. Comparison of oxygen saturation at pulmonary artery before and after BPV.

Following the percutaneous BPV (Figure 4), the patient has reported a significant improvement in her symptoms, and the shortness of breath has disappeared. A physical examination conducted before discharge showed normal vital signs, with oxygen saturation consistently ranging from 98 % to 99 % while breathing room air.

DISCUSSION

Pulmonary stenosis represents a relatively prevalent congenital heart disease, occurring in approximately 8 % to 10 % of the population and frequently co-occurring with other congenital anomalies. In the current case, the patient's primary complaints include dyspnea on exertion, chest discomfort, and intermittent palpitations. These symptoms are consistent with the documented clinical presentations associated with pulmonary stenosis. These manifestations typically tend to emerge and, in many cases, progress over time in patients with moderate to severe stenosis. In conditions marked by severe stenosis, the incapacity of the right ventricle to enhance cardiac output can lead to chest pain during physical activity, instances of fainting, and, although uncommon, sudden cardiac death (2).

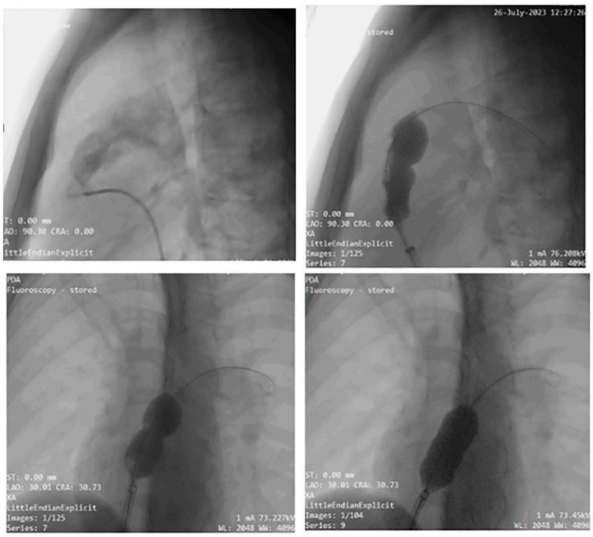

Figure 4. Percutaneous balloon pulmonary valvuloplasty was performed in patients with symptomatic moderate PS.

During the physical examination, the patient showed a singular S1 heart sound, a regular cardiac rhythm, and a continuous grade III/VI murmur at the upper left sternal border. As indicated in the existing medical literature, pulmonary stenosis

commonly presents with a systolic ejection murmur, primarily audible at the left cardiac base, with accentuation during inspiration. With increasing severity of pulmonary stenosis, the ejection click progressively approaches S1, eventually becoming inaudible in instances of severe pulmonary stenosis. In cases of pronounced pulmonary stenosis, jugular venous pressure (JVP) demonstrates conspicuous waves, and manual palpation may uncover an enlarged right ventricle; however, this was not observed in our present case (6).

Echocardiography serves as the primary diagnostic method for evaluating pulmonary stenosis (7). In this examination, the patient was diagnosed with patent ductus arteriosus featuring a left-to-right shunt, moderate valvular pulmonary stenosis, and mild to moderate pulmonary regurgitation. Consequently, the patient is scheduled for a balloon pulmonary valvuloplasty procedure. In cases of moderate pulmonary stenosis, it is advisable to undergo regular follow-up assessments, ideally on an annual basis. If there is evidence of right ventricular (RV) enlargement or the emergence of symptoms indicative of RV dysfunction, intervention becomes necessary (5). In the present case, the patient displays symptoms associated with pulmonary stenosis and an RV pressure exceeding 50 mmHg, necessitating intervention (8).

According to the American Cardiology College/ American Heart Association (ACC/ AHA) guidelines, our patient meets the criteria for undergoing balloon pulmonary valvuloplasty. This procedure is recommended for patients who show symptoms and have been diagnosed with either moderate or severe pulmonary stenosis, as well as those with valvar pulmonary stenosis. In our patient, who has been diagnosed with symptomatic moderate valvar pulmonary stenosis, the indication for balloon pulmonary valvuloplasty is fulfilled.

CONCLUSIONS

Balloon pulmonary valvuloplasty is the first-line treatment for pulmonary stenosis and should be performed promptly upon diagnosis. Intervention is not only considered in patients with severe pulmonary stenosis but also in non-severe cases with symptoms.

DECLARATIONS

Ethics approval and consent to participate.

Not applicable

Consent for publication

The authors certify that the patient and family have obtained all appropriate patient consent forms. The patient's parents understand that names and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Availability of data and material

Not applicable

Competing interests

The authors declare no competing interests.

Funding

The authors have not declared a specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Authors' contributions

AAU, YP, and AMD made the initial conception and idea. AAU and AMD prepared the initial manuscript. AQ revised and prepared the submitted manuscript. YP, AAU, IM, and MZ reviewed and advised for critical revisions. All contributing authors approved the final draft.

Acknowledgments

The authors gratefully acknowledge the patient and family members for their cooperation and willingness to provide written informed consent.

REFERENCES

1. Heaton J, Kyriakopoulos C. Pulmonic stenosis. StatPearls. 2023. Available from URL: https://www.ncbi.nlm.nih.gov/books/NBK560750/

2. Ruckdeschel E, Kim YY. Pulmonary valve stenosis in the adult patient: Pathophysiology, diagnosis and management. Heart. 2019;105(5):414-422.
3. Marchini F, Meossi S, Passarini G, Campo G, Pavasini R. Pulmonary valve stenosis: From diagnosis to current management techniques and future prospects. Vasc Health Risk Manag. 2023;19: 379-390.
4. Muneuchi J, Watanabe M, Sugitani Y, Kawaguchi N, Matsuoka R, Ando Y, et al. Early palliative balloon pulmonary valvuloplasty in neonates and young infants with tetralogy of fallot. Heart Vessels. 2020;35(2):252-258.
5. Mitchell B, Mhlongo M. The diagnosis and management of congenital pulmonary valve stenosis. SA Heart. 2018;15(1):36-45.
6. Mann DL, Zipes DP, Libby P, Bonow RO, Braunwald E. Braunwald's heart disease. A textbook of cardiovascular medicine, 10th edition. Philadelphia: Elsevier Inc; 2015.
7. Mitchell C, Rahko PS, Blauwet LA, Canaday B, Finstuen JA, Foster MC, et al. Guidelines for performing a comprehensive transthoracic echocardiographic examination in adults: recommendations from the American Society of Echocardiography. J Am Soc Echocardiogr. 2019;32(1):1-64.
8. Cuypers JA, Witsenburg M, van der Linde D, Roos-Hesselink JW. Pulmonary stenosis: Update on diagnosis and therapeutic Options. Heart. 2013;99(5):339-347.