

## Subaortic Stenosis in Association with Patent Ductus Arteriosus in a Young Woman from the Andes Region

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### Background

Defects affecting the left ventricular outflow tract (LVOT) are considered major congenital heart defects accounting for 6 out of 10,000 live births.<sup>1</sup> Subaortic stenosis accounts for approximately 1% of all congenital heart defects and 15% to 20% of all LVOT obstruction defects.<sup>2</sup> Subaortic membrane stenosis (SMS) is the most common type of subaortic stenosis, although it is a rare condition scarcely reported in the medical literature.<sup>2,3</sup> Previous studies have reported its association in some cases with other congenital heart defects such as ventricular septal defects and patent ductus arteriosus (PDA).<sup>3,4</sup> Its association with PDA is particularly rare, to the extent that a case series published in 2020 mentioned only 3 previous cases.<sup>3</sup> The objective of this article is to report a case of SMS in association with PDA in a woman from the Andes region.

### Case description

A 24-year-old female patient from the Andean region of Peru, who had previously been diagnosed with severe SMS and PDA at age 13, was evaluated by the cardiology service of our hospital because of dyspnea and pain in left upper limb on exertion. At age of 5, she was diagnosed with heart murmur and remained without further medical assessment until she was 13 years old. At baseline clinical assessment 10 years prior, physical examination revealed faint second heart sound (S2), systolic aortic heart murmur III/VI, apical impulse displaced toward the sixth intercostal space, and palpable liver 1 cm below the right costal margin. Echocardiogram showed SMS, PDA sized 13 mm in length with bidirectional flow in diastole, enlargement of the left ventricle and left atrium, preserved ejection fraction (60%), left ventricular hypertrophy, and severe pulmonary hypertension. At that time, the proposed management comprised subaortic membrane resection, PDA closure, and new atrial septal defect creation. However, the patient lost adherence to the management plan,

and she sought a new medical approach 10 years after the first cardiovascular assessment.

In our hospital, her physical examination findings were dyspnea classified as NYHA II, systolic aortic heart murmur II/VI, along with systolic murmur in the pulmonary, tricuspid, and mitral valves. Electrocardiogram showed normal sinus rhythm, 75 bpm, and left ventricle systolic overload. Echocardiogram findings were consistent with enlargement of the left ventricle and left atrium, ejection fraction 57%, left ventricle diastolic dysfunction type III, preserved right ventricular function (tricuspid annular pulmonary systolic excursion = 25 mm), moderate mitral valve insufficiency, mild mitral valve stenosis, moderate aortic valve insufficiency, severe subaortic stenosis with mean transvalvular gradient 73.6 mmHg, peak aortic velocity 5.34 m/s, moderate tricuspid valve insufficiency, and pulmonary hypertension (pulmonary artery systolic pressure = 76 mmHg) (Figure 1A and 1B). It was not possible to characterize the PDA through the echocardiogram due to an inappropriate acoustic window. Therefore, the patient underwent computed tomography angiography (Figure 2A and 2B). The angiogram showed PDA, Krichenko angiographic classification type A (conical ductus) sized 28 × 29 mm at aortic opening and 18.7 × 23.3 mm at pulmonary opening (Figure 2B). Cardiac catheterization findings showed severe mixed pulmonary hypertension, right ventricle-to-pulmonary artery shunt, ratio of pulmonary blood flow (Qp) to systolic blood flow (Qs) of 3.5, resting left ventricular-aortic systolic gradient ( $\Delta P$ ) of 100 mmHg, and pulmonary vascular resistance (PVR) of 3.79 WU. The patient was transferred to a specialized cardiovascular health center in Lima in order to undergo surgical procedure.

### Discussion

SMS in association with PDA is a particularly rare pathological condition mostly diagnosed during childhood.<sup>3</sup> We have reported the case of a young female patient seeking care for this cardiovascular condition, who was diagnosed when she was 13 years old. Our patient was diagnosed later than other previous cases reported in the literature, in which the latest diagnosis age was 8 years old.<sup>3</sup> On the other hand, there is not a probable association between male or female sex and the presence of these heart defects. We found that Steinherz et al. reported that the majority of their cases of SMS and PDA were female, with a ratio of 5:1 in New York,<sup>4</sup> but Mofa et al. reported the predominance of male patients at a ratio of 4:3 in a city in Saudi Arabia.<sup>3</sup>

Aortic stenosis is frequently associated with other heart defects.<sup>3,5</sup> For instance, Xie et al. reported that 11 out of 213 children who underwent PDA closure also had aortic

### Keywords

Subvalvar Aortic Stenosis; Ductus Arteriosus; Patent; Andean Ecosystem

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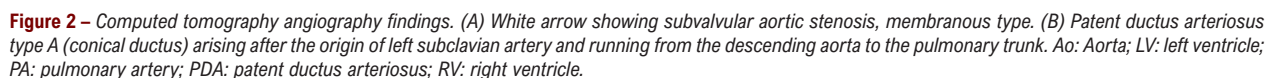
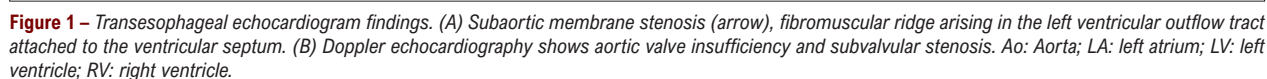
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Our patient presented Qp/Qs 3.5,  $\Delta P$  100 mmHg, and PVR 3.79 WU. These findings were similar to those reported in other studies where Qp/Qs ranged between 3.0 to 4.6, and  $\Delta P$  ranged from 38 mmHg to 139 mmHg.<sup>5,9</sup> Due to the echocardiogram

Finally, the prompt intervention in these heart defects should be addressed as the primary objective, as soon as they are diagnosed, because of highly probable complications, such as pulmonary hypertension and further aortic valve damage leading to aortic insufficiency, as previously described in this case report and other previous studies.<sup>3,9</sup>

In conclusion, SMS and PDA is a very rare association of heart defects rarely reported in the literature that can progress to more complex cardiovascular pathology by adolescence or early adulthood. Therefore, it requires special consideration by cardiologists in order to preserve the functionality of the heart and lungs in patients affected by this pathological entity.

## Case Report

### Author Contributions

Conception and design of the research, analysis and interpretation of the data, writing of the manuscript and critical revision of the manuscript for intellectual content: Sandoval RH, Cachicatari-Beltran A, Baltodano-Arellano R, Levano-Pachas G; acquisition of data: Sandoval RH.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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### Study Association

This study is not associated with any thesis or dissertation work.

### Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

### References

1. Kitchiner D, Jackson M, Malaiya N, Walsh K, Peart I, Arnold R. Incidence and Prognosis of Obstruction of the Left Ventricular Outflow Tract in Liverpool (1960-91): A Study of 313 Patients. *Br Heart J*. 1994;71(6):588-95. doi: 10.1136/hrt.71.6.588.
2. Mulla S, Siddiqui WJ. Subaortic stenosis [Internet]. Bethesda: StatPearls Publishing LLC; 2022 [cited 2023 Feb 20]. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK526085/>.
3. Moafa H, Alnasef M, Diraneyya OM, Alhabshan F. Subaortic Membrane and Patent Ductus Arteriosus in Rare Association-Case Series. *J Saudi Heart Assoc*. 2020; 32(3):410-4. doi: 10.37616/2212-5043.1199.
4. Steiner L, Ehlers KH, Levin AR, Engle MA. Membranous Subaortic Stenosis and Patent Ductus Arteriosus. *Chest*. 1977;72(3):333-8. doi: 10.1378/chest.72.3.333.
5. Xie C, Xia C, Zhao Z, Zhou Y. Hemodynamic Features of Congenital Aortic Valve Stenosis with Patent Ductus Arteriosus in Children. *Pediatr Int*. 2007;49(6):908-10. doi: 10.1111/j.1442-200X.2007.02490.x.
6. Chen BT, Zhang JQ, Dong R. Congenital Aortic Valve Stenosis. In: Wang HL, editor. *Congenital Heart Disease in children*. Beijing: Beijing Publishing Company; 1998. p. 925-938.
7. Champsaur G, Trusler GA, Mustard WT. Congenital Discrete Subvalvar Aortic Stenosis. Surgical Experience and Long-Term Follow-Up in 20 Paediatric Patients. *Br Heart J*. 1973;35(4):443-6. doi: 10.1136/hrt.35.4.443.
8. Reis RL, Peterson LM, Mason DT, Simon AL, Morrow AG. Congenital Fixed Subvalvular Aortic Stenosis. An Anatomical Classification and Correlations with Operative Results. *Circulation*. 1971;43(5 Suppl):I11-8. doi: 10.1161/01.cir.43.5s1.i-11.
9. Newfield EA, Muster AJ, Paul MH, Idriss FS, Riker WL. Discrete Subvalvular Aortic Stenosis in Childhood. Study of 51 Patients. *Am J Cardiol*. 1976;38(1):53-61. doi: 10.1016/0002-9149(76)90062-x.
10. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al. 2020 ESC Guidelines for the Management of Adult Congenital Heart Disease. *Eur Heart J*. 2021;42(6):563-645. doi: 10.1093/eurheartj/ehaa554.



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