

Double-Orifice Mitral Valve with Severe Regurgitation Associated with Bicuspid Aortic Valve: A Case Report

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Introduction

Double-orifice mitral valve (DOMV) was first described in 1876 by the British pathologist William Smith Greenfield. It is a very rare congenital or acquired cardiac anomaly with limited epidemiologic data and no clear sex predilection.¹

Congenital DOMV rarely occurs in isolation and is commonly associated with other cardiac anomalies, such as aortic coarctation, patent ductus arteriosus, atrioventricular septal defects, and bicuspid aortic valve (BAV), among others. Functionally, it may behave normally and remain asymptomatic; however, symptoms may arise from mitral valve dysfunction, such as stenosis or regurgitation.²

Acquired cases are usually due to calcification of the mitral annulus associated with aging, which, when extensive or irregular, can form a “bridge” of calcified tissue within the annulus, dividing the mitral orifice into two separate passages and creating the appearance of a double orifice. It may also occur after iatrogenic surgical or percutaneous procedures on the mitral valve (Alfieri technique or MitraClip device).³

Given the low prevalence of the association between DOMV and BAV, as well as the limited evidence regarding optimal surgical management, we present this case to highlight the diagnostic value of advanced echocardiography and the feasibility of complex mitral valve repair in a young patient with progressive ventricular remodeling.

Case Presentation

A 24-year-old asymptomatic male with a known history of mitral valve prolapse and BAV was evaluated. Physical examination revealed a grade III/VI holosystolic murmur, while resting electrocardiography was normal. As part of the clinical reassessment, a new transthoracic two-dimensional (2D) echocardiographic evaluation was performed, which established the diagnosis of a DOMV with severe

mitral regurgitation. The left atrium demonstrated normal dimensions and indexed volume (36 mm and 32 mL/m²). The left ventricle exhibited eccentric hypertrophy (indexed LV mass 121 g/m²) with abnormal segmental wall motion, characterized by diffuse hypokinesia and globally reduced systolic thickening, with a left ventricular ejection fraction (LVEF) of 50%.

Anatomically, the mitral valve consisted of two orifices: an anterolateral orifice with two leaflet components and a posterolateral dominant orifice responsible for severe regurgitation. The mitral annulus was dilated, measuring 51 mm, and both papillary muscles (anterolateral, appearing bifid, and posteromedial) were visualized with a preserved subvalvular apparatus. Color Doppler analysis demonstrated laminar transmitral flow with an elevated regurgitant volume.

The BAV demonstrated preserved flow without significant regurgitation, although ascending aortic ectasia was noted. The tricuspid valve exhibited minimal regurgitation, and the pulmonary valve showed mild regurgitation, with an estimated pulmonary artery systolic pressure of 28 mmHg.

Three-dimensional transesophageal echocardiography (TEE) and computed tomography angiography (CTA) were performed (Figure 1). Three-dimensional TEE provided complementary anatomical information not fully appreciated on 2D imaging. The smaller anterolateral orifice consisted of two myxomatous leaflet components with mild regurgitation, whereas the larger dominant orifice exhibited redundant, multisegmental prolapsing leaflets responsible for severe mitral regurgitation. Severity was characterized by a vena contracta of 0.7 cm, a proximal isovelocity surface area (PISA) radius of 0.76 cm, an increased mitral E-wave velocity of 168 cm/s, a regurgitant volume of 87 mL per beat, a regurgitant fraction of 56%, and an effective regurgitant orifice area of 0.969 cm². The left atrial appendage was free of thrombus, with preserved flow velocities. The right ventricular inflow tract, superior vena cava, and tricuspid valve were normal, with a right ventricular ejection fraction of 52%.

The left ventricle was severely dilated, with an end-diastolic diameter of 65 mm and an end-systolic diameter of 53 mm, and systolic function at the lower limit of normal. CTA of the thoracic and abdominal aorta demonstrated aortic root dilation (41.6 mm) without evidence of coarctation (Figure 2).

Although preoperative echocardiography reported left atrial dimensions within normal limits, prior echocardiographic studies before referral had documented left atrial enlargement. The decision to perform a modified Maze surgical ablation combined with left atrial reduction atrioplasty and internal closure of the left atrial appendage was made intraoperatively,

Keywords

Mitral valve; Mitral Valve Insufficiency; Mitral Valve Annuloplasty

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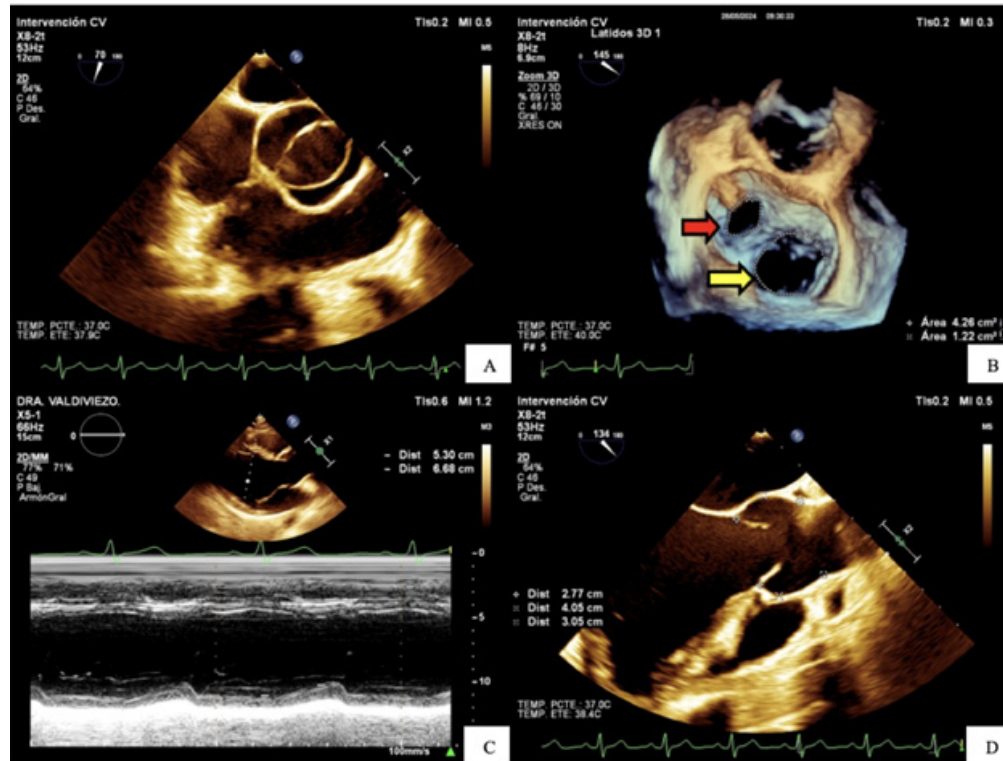


Figure 1 – Preoperative echocardiographic assessment. A) BAV, type 0 according to the Sievers classification. B) DOMV with asymmetric orifices: the larger posteromedial orifice (yellow arrow), responsible for severe mitral regurgitation, and the smaller anterolateral orifice (red arrow), with minimal regurgitation. C) M-mode TTE showing a dilated left ventricle with an end-systolic diameter of 53 mm, consistent with moderate systolic dysfunction. D) Parasternal long-axis view demonstrating ascending aortic ectasia, measuring 40.5 mm at the level of the sinuses of Valsalva. BAV: bicuspid aortic valve; DOMV: double-orifice mitral valve; TEE: transesophageal echocardiogram.

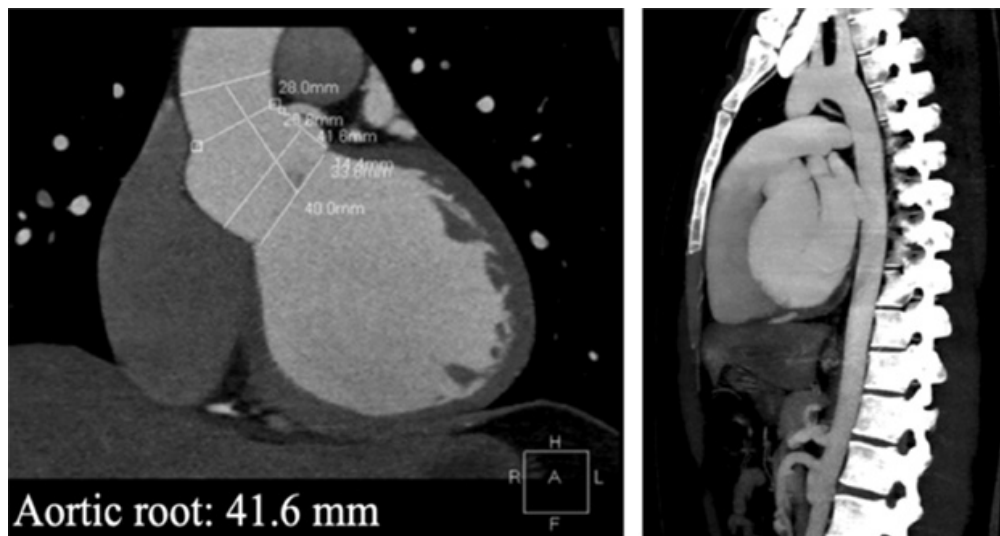


Figure 2 – Thoracic aorta CTA. Multiplanar reconstruction of CTA demonstrating aortic root dilation measuring 41.6 mm. No evidence of coarctation is observed throughout the thoracic aorta. CTA: computed tomography angiography.

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based on direct surgical assessment, with the aim of optimizing atrial geometry and facilitating reconstruction during a complex mitral valve repair.

Valve repair was preferred over replacement because of the patient's young age, preservation of the subvalvular apparatus, and its advantages in terms of improved long-term survival, preservation of left ventricular function, and avoidance of lifelong anticoagulation. The presence of redundant, myxomatous leaflets with multisegmental prolapse made repair technically feasible. Quadrantectomy of segments A2–A3 and P1–P2 was performed, followed by implantation of a size 33 semirigid annuloplasty ring to correct the excess valvular tissue responsible for severe regurgitation and restore adequate leaflet coaptation, while providing annular stabilization with preservation of the subvalvular apparatus. This strategy offers favorable long-term durability, which is closely related to the expertise of the surgical team.

At the first postoperative transthoracic echocardiogram (TTE) performed one month after surgery, an echogenic mass measuring 3.3×1 cm was identified at the roof of the right atrium. The patient was subsequently treated with rivaroxaban for three months. A second echocardiographic evaluation demonstrated regression of the lesion (2.9×0.6 cm), and complete resolution was documented at the third follow-up examination four months after surgery. The patient remained clinically stable throughout follow-up, without embolic events or documented arrhythmias. The progressive reduction and eventual disappearance of the lesion with anticoagulation support the diagnosis of a transient postoperative mural thrombus, likely related to atrial surgical manipulation.

A follow-up TTE performed four months after surgery showed a mildly dilated left ventricle, with an indexed left ventricular mass of 99 g/m^2 and a LVEF of 53%. The repaired posteroseptal mitral valve appeared anatomically normal following quadrangular resection and annuloplasty

ring implantation, with obliteration of the anterolateral orifice. Color Doppler imaging demonstrated turbulent transmitral flow, with an E-wave velocity of 116 cm/s and an A-wave velocity of 98 cm/s. The calculated mitral valve area was 2.49 cm^2 , effectively excluding mitral stenosis. Mild residual mitral regurgitation was observed. Pulmonary artery systolic pressure (PASP) was estimated at 23 mmHg, with no evidence of pulmonary hypertension (Figure 3).

Discussion

The incidence of DOMV is estimated to be between 0.05% and 0.06% in the general adult population, and it is correctly diagnosed in approximately 1% of patients with congenital heart disease. Clinically, DOMV is often asymptomatic, as normal flow may occur through both orifices; however, it can cause mitral stenosis or regurgitation if subvalvular anatomy is abnormal or if valve degeneration occurs.⁴

Based on echocardiographic findings, congenital DOMV can be anatomically classified into three types: (1) Complete bridge, in which a fibrous bridge completely divides the atrioventricular orifice (approximately 15% of cases); (2) Incomplete bridge, characterized by a small tissue strand connecting the anterior and posterior leaflets at the edge; and (3) Hole type (eccentric), the most common (85%), with a secondary orifice and subvalvular apparatus at the lateral commissure, as in this case.⁵

We report a case of eccentric hole-type DOMV with severe MR associated with BAV. Mitral valve repair is indicated in asymptomatic patients with severe primary MR and LV dysfunction, particularly when the LV end-systolic diameter is ≥ 40 mm or the LVEF is $\leq 60\%$ (Level of Evidence IB). The severity of MR is determined by qualitative, quantitative, semiquantitative, and structural echocardiographic criteria.^{6,7} In this context, individualized surgical planning is essential, particularly in young patients in whom valve repair may offer durable results while preserving ventricular function.

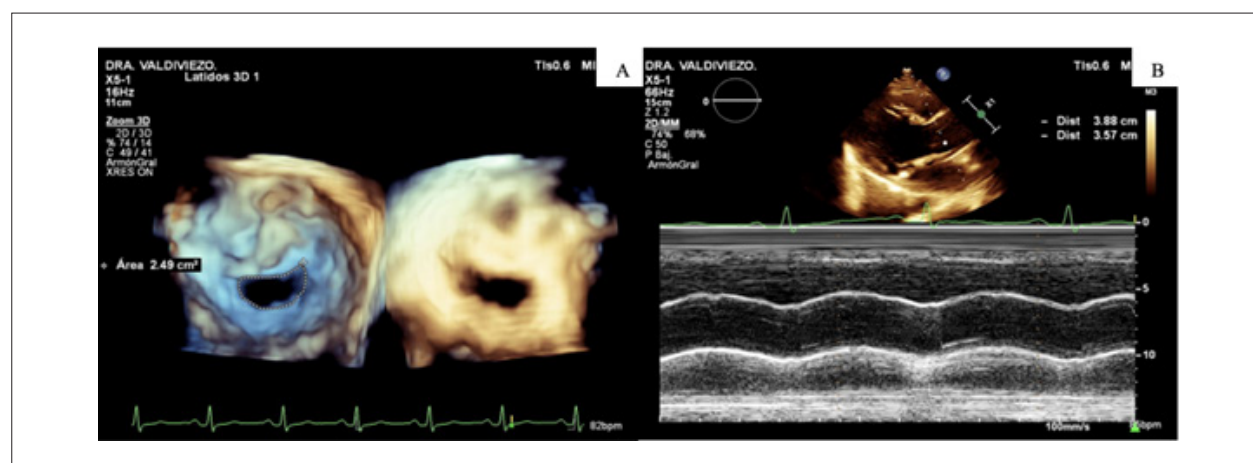


Figure 3 – Postoperative TTE. A) Three-dimensional reconstruction demonstrating successful repair of the DOMV, with a single functional orifice and a mitral valve area of 2.49 cm^2 . B) M-mode echocardiogram demonstrating a reduction in left ventricular end-systolic diameter compared with preoperative measurements. TTE: transthoracic echocardiogram; DOMV: double-orifice mitral valve.

The association of DOMV and BAV is rare. In 2021, the first case of a 5-year-old girl with Turner syndrome and DOMV associated with BAV was reported. A review of 24 patients with DOMV and BAV across 16 studies (1988–2020) showed ages ranging from 1 month to 67 years (mean, 25.6 years), with 62.5% male. Most cases were of the complete bridge type. MR was the most common dysfunction (41.7%), and aortic coarctation was the most complex malformation (45.8%). No cases involved genetic syndromes. Both DOMV and BAV may be associated with genetic syndromes, including Turner syndrome, suggesting a possible X-linked inheritance and supporting the hypothesis that multivalvular diseases may have a genetic component.⁸

Surgical repair of DOMV presents specific technical challenges that differ from those of conventional degenerative mitral valve repair. Accurate identification of the dominant regurgitant orifice is essential to avoid residual regurgitation or inadvertent functional mitral stenosis after reconstruction. In this case, the presence of redundant multisegmental prolapse required careful leaflet resection while preserving the integrity of the subvalvular apparatus. Annular stabilization had to be carefully balanced to restore adequate leaflet coaptation without excessively reducing the effective mitral valve area, particularly after obliteration of the accessory orifice. Precise intraoperative assessment and surgical expertise were crucial to achieving a durable and physiologically adequate repair.

Long-term considerations following complex DOMV repair include the risk of recurrent mitral regurgitation due to progressive annular dilation or residual leaflet pathology, functional mitral stenosis secondary to excessive tissue resection or restrictive annuloplasty, and atrial arrhythmias related to atrial remodeling procedures. In young patients, structured echocardiographic follow-up is essential to monitor ventricular remodeling, transmitral gradients, durability of the repair, and progression of associated aortopathy. Early detection of recurrent dysfunction may allow timely intervention and optimization of long-term outcomes.

This report describes a single case, which inherently limits generalizability. The duration of follow-up is relatively short, precluding definitive conclusions regarding the long-term durability of the repair and ventricular remodeling. Additionally, although the DOMV and the BAV may share potential developmental or genetic mechanisms, no formal genetic evaluation was performed in this patient. Larger series and studies with extended follow-up are required to better define optimal surgical strategies and long-term prognosis in this rare association.

Conclusion

This case illustrates a rare association of eccentric (hole-type) DOMV with BAV, successfully treated with complex mitral valve repair in a young patient. Advanced

echocardiography enabled precise anatomical assessment and individualized surgical planning. Early intervention and tailored repair strategies can achieve favorable outcomes even in asymptomatic patients with progressive ventricular remodeling, supporting preservation of the native valve in young individuals.

This case illustrates a rare association of eccentric (hole-type) DOMV with BAV in an asymptomatic young patient, successfully managed with complex mitral valve repair in accordance with current guideline recommendations. Advanced multimodality imaging was essential for precise anatomical characterization and surgical planning. Despite the technical challenges inherent to DOMV repair, preservation of the native valve can be achieved with favorable early outcomes when performed in experienced centers. Careful postoperative monitoring remains essential to assess the long-term durability of the repair, ventricular remodeling, and progression of associated aortopathy in this rare and complex valvular condition.

Author Contributions

Conception and design of the research, acquisition of data, analysis and interpretation of the data, statistical analysis, writing of the manuscript and critical revision of the manuscript for intellectual content: Cedeño PD, Herrera ECA, Mite CJT, Valenzuela EV, Bohorquez JJZ, Moreira GN.

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

All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.

Use of Artificial Intelligence

The authors did not use any artificial intelligence tools in the development of this work.

Availability of Research Data

The underlying content of the research text is contained within the manuscript.

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References

1. Alahmadi MH, Chowdhury YS. Double Orifice Mitral Valve. In: StatPearls [Internet]. Treasure Island: StatPearls Publishing; 2026 [cited 2026 May 25]. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK559298/>.
2. Pratap H, Gupta A, Acharya PK, Mahajan V. Double Orifice Mitral Valve: A Rare Cause of Isolated Severe Congenital Mitral Regurgitation. *Ann Pediatr Cardiol*. 2020;13(2):174-6. doi: 10.4103/apc.APC_133_19.
3. Lange M, Büttel H, Wichter T. Calcified Mitral Stenosis Imitates a MitraClip® and Forms a Double Orifice. *Eur Heart J Case Rep*. 2018;2(3):yty084. doi: 10.1093/ehjcr/yty084.
4. Torres-Martel JM, Barrera JC, Flores J, García-Tejada BA, Begazo-Paredes JE, García-Cruz E. Double Orifice Mitral Valve Associated with Bicuspid Aortic Valve: A Case Report. *Online J Cardiol Res Rep*. 2023;7(4):1-4. doi: 10.3352/OJCRR.2023.07.000667.
5. Alahmadi MH, Chowdhury YS. Double Orifice Mitral Valve. In: StatPearls. Treasure Island: StatPearls Publishing; 2026.
6. Vahanian A, Beyersdorf F, Praz F, Milojevic M, Baldus S, Bauersachs J, et al. Guía ESC/EACTS 2021 Sobre el Diagnóstico y Tratamiento de las Valvulopatías. *Rev Esp Cardiol*. 2022;75(6):524.e1-524.e69. doi: 10.1016/j.recesp.2021.11.023.
7. Otto CM, Nishimura RA, Bonow RO, Carabello BA, Erwin JP 3rd, Gentile F, et al. 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021;143(5):e72-e227. doi: 10.1161/CIR.0000000000000923.
8. Sun F, Tan X, Sun A, Zhang X, Liang Y, Ren W. Rare Double Orifice Mitral Valve Malformation Associated with Bicuspid Aortic Valve in Turner Syndrome: Diagnosed by a Series of Novel Three-Dimensional Echocardiography and Literature Review. *BMC Cardiovasc Disord*. 2021;21(1):377. doi: 10.1186/s12872-021-02184-2.



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