

## IMAGE IN CARDIOLOGY

# An unusual finding in severe mitral regurgitation complicated with hypoxemia: The importance of three-dimensional echocardiography

## Um achado incomum numa insuficiência mitral grave complicada com hipoxemia: a importância da ecocardiografia 3D

Inês Ferreira Neves<sup>a,\*</sup>, Francisco Barbas de Albuquerque<sup>a</sup>, Ruben Ramos<sup>a</sup>, Rui Rodrigues<sup>b</sup>, Ana Galrinho<sup>a</sup>

<sup>a</sup> Department of Cardiology, Hospital de Santa Marta, Centro Hospitalar Universitário de Lisboa Central, ULS S. José, Lisbon, Portugal

<sup>b</sup> Department of Cardiac Surgery, Hospital de Santa Marta, Centro Hospitalar Universitário de Lisboa Central, ULS S. José, Lisbon, Portugal

Received 2 September 2024; accepted 10 November 2024

Available online 18 February 2025

A 64-year-old woman with severe mitral regurgitation (MR), pulmonary hypertension and patent foramen ovale (PFO) was referred for preoperative transthoracic echocardiography (TTE).

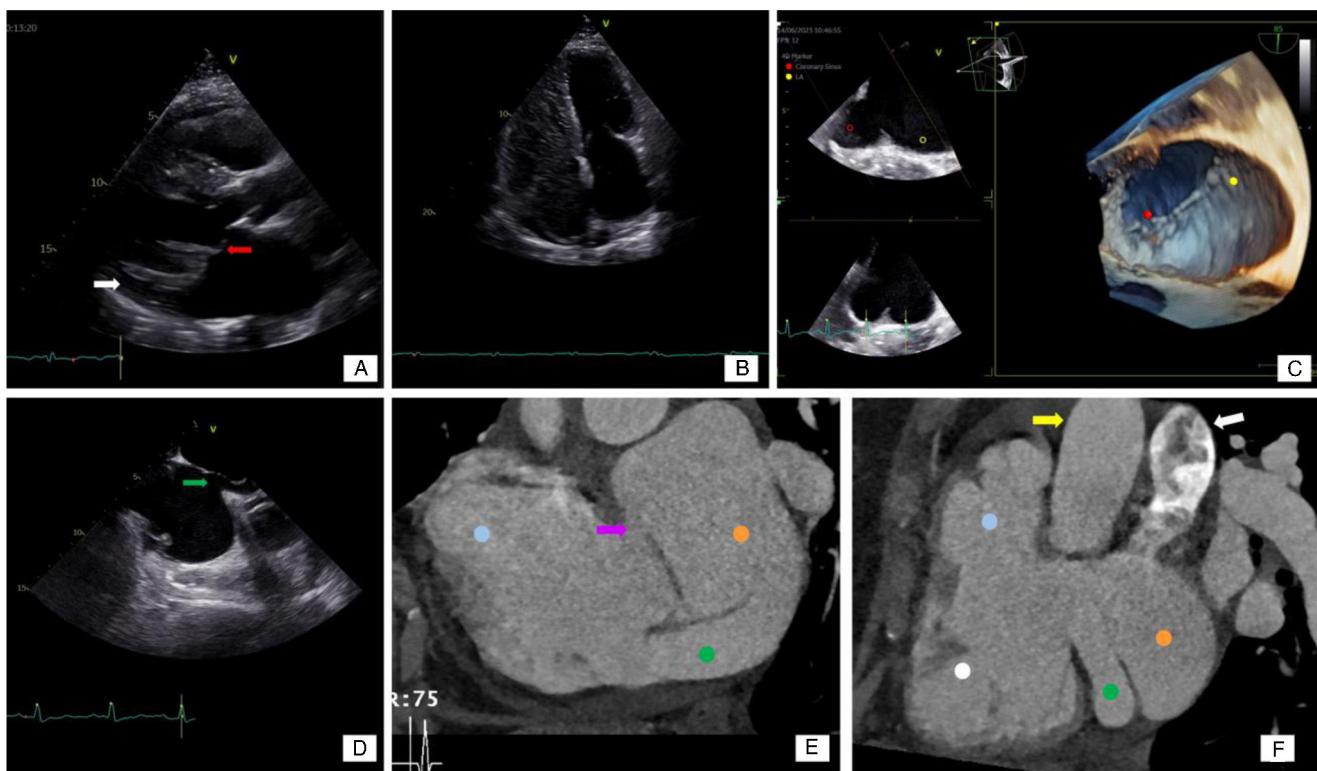
Besides her known conditions, the TTE showed a dilated coronary sinus (CS) and right ventricle that was not proportional to the MR. Transesophageal echocardiography (TEE) revealed prolapse of the mitral valve anterior leaflet (**Figure 1A**). The CS was markedly dilated, with no opacification after saline injection (**Figure 1B**). There was echo dropout in the left atrial wall and communication with the CS, raising the suspicion of unroofed CS (URCS) atrial septal

defect (ASD). Three-dimensional echocardiography showed absence of the CS wall (**Figure 1C**) and the already known PFO (**Figure 1D**). Coronary computed tomography angiography (CCTA) confirmed the diagnosis of URCS (**Figure 1E** and F). Right catheterization showed a cardiac index of  $11.47 \text{ l/min/m}^2$ , pulmonary artery pressure of 47 mmHg, and significant left-to-right shunt ( $\text{Qp}/\text{Qs} 2.2$ ). The patient underwent mitral valve replacement and ASD closure. The PFO was left open. Postoperative course was uneventful and the patient was still asymptomatic at one year.

URCS is a rare type of interatrial communication (less than 1% of lesions related to interatrial shunting).<sup>1</sup> Unlike a true ASD, the communication occurs through the ostium of the CS, resulting from the partial or complete absence of its roof.<sup>2,3</sup> The diagnosis requires a high level of suspicion.<sup>3,4</sup> Echocardiography remains the most important diagnostic modality.<sup>5</sup> CCTA can provide further information.<sup>1</sup>

\* Corresponding author.

E-mail address: [inessferreiraneves@gmail.com](mailto:inessferreiraneves@gmail.com)  
(I. Ferreira Neves).



**Figure 1** (A) Transthoracic echocardiography (TTE), parasternal long-axis view, showing the mitral valve (red arrow) and the enlarged coronary sinus (white arrow); (B) TTE, 4-chamber view, after injection of saline, without significant opacification; (C) three-dimensional echocardiography showing the coronary sinus (red dot) and left atrium (yellow dot) and revealing the absence of the coronary sinus wall; (D) transesophageal echocardiography showing the patent foramen ovale (green arrow); (E) coronary computed tomography angiography (CCTA) showing the right atrium (blue dot), left atrium (orange dot), the enlarged CS (green dot) with missing roof, communicating directly with the left atrium, and a communication between the right and left atria (purple arrow); (F) CCTA showing the right atrium (blue dot), right ventricle (white dot), left atrium (orange dot) and enlarged coronary sinus (green dot). The aorta (yellow arrow) and pulmonary artery (white arrow) can also be seen.

## Conflicts of interest

The authors have no conflicts of interest to declare.

## Appendix A. Supplementary data

Supplementary material associated with this article can be found in the online version at <https://doi.org/10.1016/j.repc.2024.11.013>.

## References

- Abazid RM, Goha A, Romsa JG, et al. Coronary sinus to left atrium fistula on computed tomography angiography: differentiation from unroofed coronary sinus with literature review. *J Cardiovasc Comput Tomogr*. 2021;15:e15-7.
- Adatia I, Gittenberger-de Groot AC. Unroofed coronary sinus and coronary sinus orifice atresia. Implications for management of complex congenital heart disease. *J Am Coll Cardiol*. 1995;25:948-53.
- Chen JL, Yu CG, Wang DJ, et al. Misdiagnosis of unroofed coronary sinus syndrome as an ostium primum atrial septal defect by echocardiography: a case report. *World J Clin Cases*. 2022;10:1592-7.
- Murli L, Ranjit MS, Shah P. Unroofed coronary sinus: an unusual interatrial communication and a rare childhood entity. *Ann Pediatr Cardiol*. 2019;12:64-5.
- Shi H, Yan J, Wang Q, et al. Surgical management of unroofed coronary sinus syndrome: a 20-year-single-center experience. *J Card Surg*. 2021;36:589-95.