

# Giant Left Atrium due to Probable Rheumatic Disease and Atrial Functional Mitral Insufficiency

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## Introduction

Giant left atrium is a rare condition, and most cases described in the literature are associated with rheumatic disease.<sup>1</sup> However, some clinical descriptions suggest a nonrheumatic etiology for this condition, in which characteristic findings of rheumatic disease are not identified or anatomopathological findings do not suggest a specific etiology.

Atrial fibrillation is present in almost all cases and contributes to the maintenance of left atrial remodeling and persistent changes in structure and function.<sup>2</sup>

Although the mechanism of mitral valve disease may initially be associated with a primary abnormality of the valve apparatus, marked dilatation of the mitral annulus and other changes in valve morphology often led to the development of functional mitral regurgitation.<sup>3</sup>

## Case report

A 35-year-old man was referred for outpatient echocardiographic follow-up after an appointment with a cardiologist for complaints of chest pain. He reported no history of exertional dyspnea, orthopnea, or paroxysmal nocturnal dyspnea. Previous surgical correction of atrial septal defect at the age of 22. At that time, a chest radiograph showed marked enlargement of the cardiac silhouette and suggested the presence of a pericardial effusion. The electrocardiogram showed an atrial fibrillation rhythm. In previous consultations, he had been prescribed losartan, carvedilol, and spironolactone; the use of spironolactone was discontinued by the patient on his own initiative.

Two-dimensional transthoracic echocardiography showed a dilated left atrium with an anteroposterior diameter of 10.2 mm. The volume of the left atrium was estimated to be approximately 1,200 ml, with an indexed volume of 615 ml/m<sup>2</sup> (Figure 2). The mitral valve had a short posterior leaflet with limited mobility (hamstringing).

## Keywords

Left atrium; rheumatic heart disease; echocardiography

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The anterior leaflet was elongated with a displacement of its tip toward the left atrium (i.e., pseudoprolapse), resulting in significant coaptation failure and marked eccentric regurgitation (Figure 1). No thrombi were identified. Although the subvalvular apparatus did not show typical features of rheumatic disease (e.g., chordae tendineae without significant shortening or thickening) and no calcifications were observed, three-dimensional transthoracic examination revealed bicommissural fusion (Figure 3). The other valves showed no relevant morphofunctional changes. The left ventricle appeared significantly dilated, with a diastolic diameter of 86 mm, a diastolic volume of 427 ml, and a reduced ejection fraction (44%), which was estimated by using the Simpson method.

At a follow-up appointment, anticoagulant therapy was prescribed along with a request for evaluation for cardiac surgery.

## Discussion

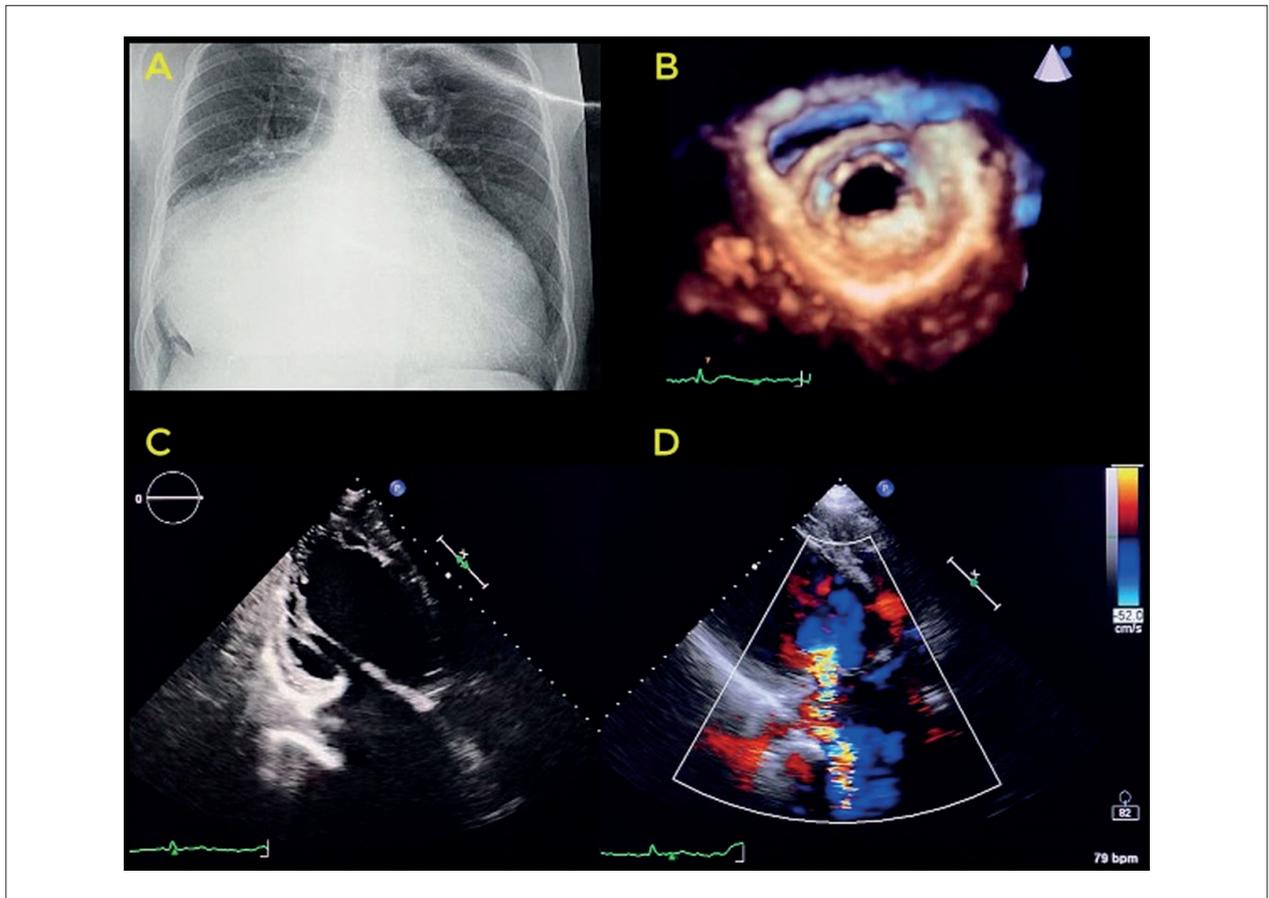
The definition of a giant left atrium is variable. It has been described on chest radiographs as extending to the right lateral aspect of the thoracic wall.<sup>1,4</sup> Furthermore, it may be defined by a cardiothoracic ratio of > 0.7 combined with an anteroposterior diameter of the left atrium greater than 80 mm, as assessed by transthoracic echocardiography.<sup>5</sup>

Imaging techniques such as cardiac magnetic resonance imaging and computed tomography may be needed to more accurately assess the size of the left atrium and its relationship to surrounding structures.<sup>6</sup>

Few cases of giant left atrium are asymptomatic; when symptomatic, they are usually related to either mitral valve disease or compression of the esophagus and airway because of enlargement of the posterior wall of the left atrium, resulting in dysphagia and respiratory dysfunction. Enlargement of the left atrium also facilitates thrombus formation and predisposes to thromboembolic events.<sup>7</sup>

Surgical management includes isolated mitral valve surgery or mitral valve surgery combined with left atrial volume reduction. The primary indication for volume reduction is the presence of intra- or extracardiac compressive symptoms.<sup>8</sup>

In the reported case, both two-dimensional and three-dimensional echocardiographic images show a short, narrow posterior leaflet with apparent bicommissural fusion. Chronic rheumatic heart disease, a significant cause of mitral valve disease in Brazil with a variable presentation on echocardiography, is often mentioned as a possible etiological factor.



**Figure 1** – A) Chest X-ray showing an enlargement of the cardiac area mainly due to the left atrium, which touches the right lateral border of the thoracic wall; B) Three-dimensional (3D) bidimensional echocardiogram (ventricular aspect) revealing a short/tractioned posterior mitral leaflet; C) Subvalvular mitral apparatus in the three-chamber apical view; D) Significant mitral regurgitation observed during color flow mapping.

The history of pathological interatrial communication requiring surgical correction raises the hypothesis that atrial dilatation may have occurred at some point. This condition predisposes to atrial fibrillation and may lead to functional mitral regurgitation.

A significant dilation of the left ventricle is also associated with a slight reduction in ejection fraction. However, the role of these changes as a cause or consequence of mitral valve regurgitation and consequently of left atrial remodeling cannot be determined in the pathological evolution described.

## Conclusion

Giant left atrium is an extremely rare condition. Echocardiography plays a pivotal role in identifying this condition, investigating its underlying causes, and determining appropriate management strategies. Although a rheumatic etiology predominates in this setting, several factors and causes should be considered as contributing to the excessive dilatation of the left atrium.

## Author Contributions

Conception and design of the research, acquisition of data, analysis and interpretation of the data, statistical analysis and writing of the manuscript: Feitosa INA; critical revision of the manuscript for intellectual content: Feitosa INA, Andrade ING.

## Potential Conflict of Interest

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

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There were no external funding sources for this study.

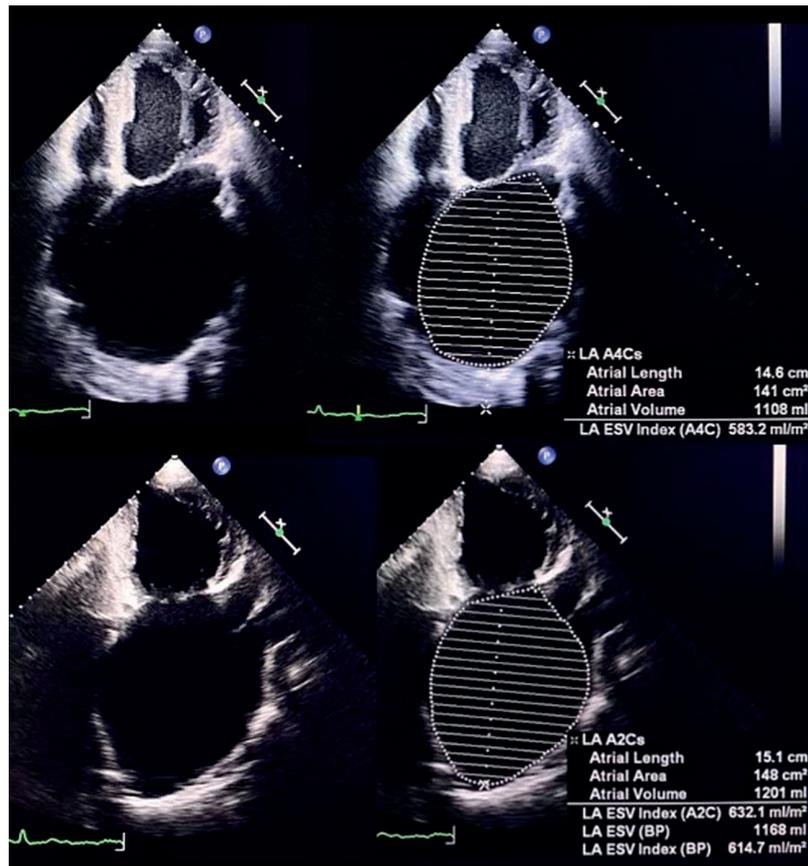
## Study Association

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

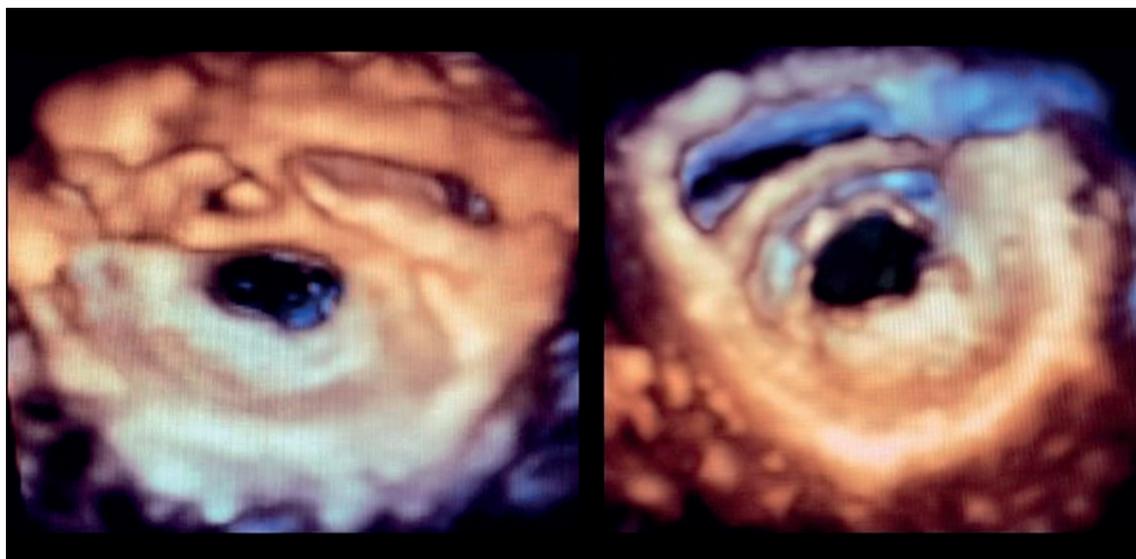
## Ethics Approval and Consent to Participate

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

## Case Report



**Figure 2** – Two-dimensional transthoracic echocardiography with measurements of the left atrial volume in apical 2- and 4-chamber views.



**Figure 3** – Three-dimensional transthoracic echocardiography of the mitral valve. On the left, the atrial aspect; on the right, the ventricular aspect.

### References

1. El Maghraby A, Hajar R. Giant Left Atrium: A Review. *Heart Views*. 2012;13(2):46-52. doi: 10.4103/1995-705X.99227.
2. van de Vegte YJ, Siland JE, Rienstra M, van der Harst P. Atrial Fibrillation and Left Atrial Size and Function: A Mendelian Randomization Study. *Sci Rep*. 2021;11(1):8431. doi: 10.1038/s41598-021-87859-8.
3. Honda Y, Watanabe N, Nishino S, Matsuura H, Nishimura M, Yano M, et al. Non-Rheumatic Giant Left Atrium: An Illustrative Case Successfully Treated by Surgical Intervention. *J Cardiol Cases*. 2021;24(2):79-83. doi: 10.1016/j.jccase.2021.01.012.
4. Hurst JW. Memories of Patients with a Giant Left Atrium. *Circulation*. 2001;104(22):2630-1. doi: 10.1161/hc4701.100775.
5. Piccoli GP, Massini C, Di Eusanio G, Ballerini L, Iacobone G, Soro A, et al. Giant Left Atrium and Mitral Valve Disease: Early and Late Results of Surgical Treatment in 40 Cases. *J Cardiovasc Surg (Torino)*. 1984;25(4):328-36.
6. Pandit BN, Aggarwal P, Subramaniyan S, Gujral JS, Nath RK. Largest Giant Left Atrium in Rheumatic Heart Disease. *J Cardiol Cases*. 2020;24(1):10-3. doi: 10.1016/j.jccase.2020.11.022.
7. Darwazah AK, El Sayed H. Giant Left Atrium Associated with Massive Thrombus Formation. *Thromb J*. 2013;11(1):5. doi: 10.1186/1477-9560-11-5.
8. Apostolakis E, Shuhaiber JH. The Surgical Management of Giant Left Atrium. *Eur J Cardiothorac Surg*. 2008;33(2):182-90. doi: 10.1016/j.ejcts.2007.11.003.



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