

## Idiopathic Dilatation of the Right Atrium in Asymptomatic Adolescent – Case Report

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### Introduction

Right atrial (RA) idiopathic dilatation is a rare condition, often asymptomatic, with controversial treatment, and may be restricted to clinical follow-up or surgical treatment. We describe the case of a teenager with echocardiographic findings consistent with this disease.

### Case Report

B.R.I.C., 16 years old, male, asymptomatic from the cardiovascular point of view, had routine tests for the purposes of doing physical activity, when significant increase in the cardiac area was observed on chest radiography. The electrocardiogram showed no abnormalities, so transthoracic echocardiography was conducted. The echocardiographic findings were a marked increase in the right atrium with atrial volume estimated at 270 mL (Figure 1). In the same patient, the left atrial volume was 30 mL. The tricuspid valve showed no structural abnormalities to suggest Ebstein's disease, however, we observed a marked dilatation of the tricuspid valve annulus, measuring 6.3 cm in the apical 4-chamber view. Surprisingly, tricuspid regurgitation was slight (Figure 2) and there was no significant pulmonary hypertension (systolic pulmonary artery pressure estimated at 30 mmHg from the tricuspid regurgitation). The right ventricle was slightly increased, but its systolic function was preserved. The left chambers also presented normal dimensions and systolic function with mild left atrial compression. To exclude the presence of intracavitary thrombi, transesophageal echocardiography was performed, which confirmed the findings of transthoracic echocardiography and revealed the integrity of the atrial septum and the absence of atrial appendix thrombi. Due to excessive RA enlargement, surgical treatment with partial resection of the right atrium was agreed, in an attempt to prevent atrial arrhythmias and potential thromboembolic events. During surgery, there was a marked RA dilatation, which presented paper-like thin walls.

### Keywords

Adolescent; Heart Atria/abnormalities; Heart Atria/surgery; Dilatation, Pathologic/diagnosis; Echocardiography.

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Partial resection was performed (Figures 3 and 4) and the postoperative period was uneventful. Histopathological examination of the resected segment revealed thinning of the myocardial layer with attenuation and focal absence of the myocardial layer and fatty degeneration (Figure 5). Echocardiogram scans performed in the early postoperative period and after one year showed mild residual dilatation of the right atrium with minimal tricuspid regurgitation. The patient remained asymptomatic with no clinical complications, maintaining annual cardiac monitoring until the present day. Recent echocardiogram scans, however, showed progressive dilatation of the right atrium.

### Discussion

RA idiopathic dilatation is a rare anomaly with etiology not yet defined. In most cases, the diagnosis is done in children, incidentally, from a chest X-ray with increased heart area. Rare cases have been reported of intrauterine<sup>1</sup> RA idiopathic dilatation or in patients with advanced age, in autopsy studies.<sup>2</sup> In general, it has a benign prognosis and may, however, be accompanied by atrioventricular block or atrial arrhythmia,<sup>3,4</sup> which in turn are a potential cause of thromboembolism. Anatomical and pathological findings consist of aneurysm restricted to the RA, which is made up of extremely thin walls, with histological studies showing extensive atrial fibrosis. The diagnosis of RA idiopathic dilatation is based on the typical echocardiographic findings,<sup>5,6</sup>. Differential diagnosis is mainly made with diseases that affect the right side of the heart, including the Ebstein's anomaly and its variations, Uhl disease and RV arrhythmogenic dysplasia. Ebstein's anomaly has a more apical implantation of the tricuspid valve, resulting in greater RA dimension, even surpassing the right ventricular size. However, there is no tissue replacement of the right chamber wall. On the other hand, in patients with RV arrhythmogenic dysplasia, the typical histological finding is the replacement of myocardial tissue for fibrofatty tissue on the ventricular wall; in Uhl's disease, there is no myocardial layer on the RV free wall, either total or partial, with preservation of valve anatomy on both occasions. Uhl's disease usually progresses to death in childhood, but cases where diagnosis was done in adulthood have been described. Treatment of RA idiopathic dilatation is controversial because of its rarity; studies with a larger number of patients (seven children) reported good prognosis for asymptomatic patients under clinical follow-up. For symptomatic patients who may present symptoms of right heart failure and/or recurrent arrhythmia, drug treatment may be useful for improving the patients' quality of life, serving as a bridge to the definitive surgical treatment, which consists of partial RA resection, and has been used for symptomatic

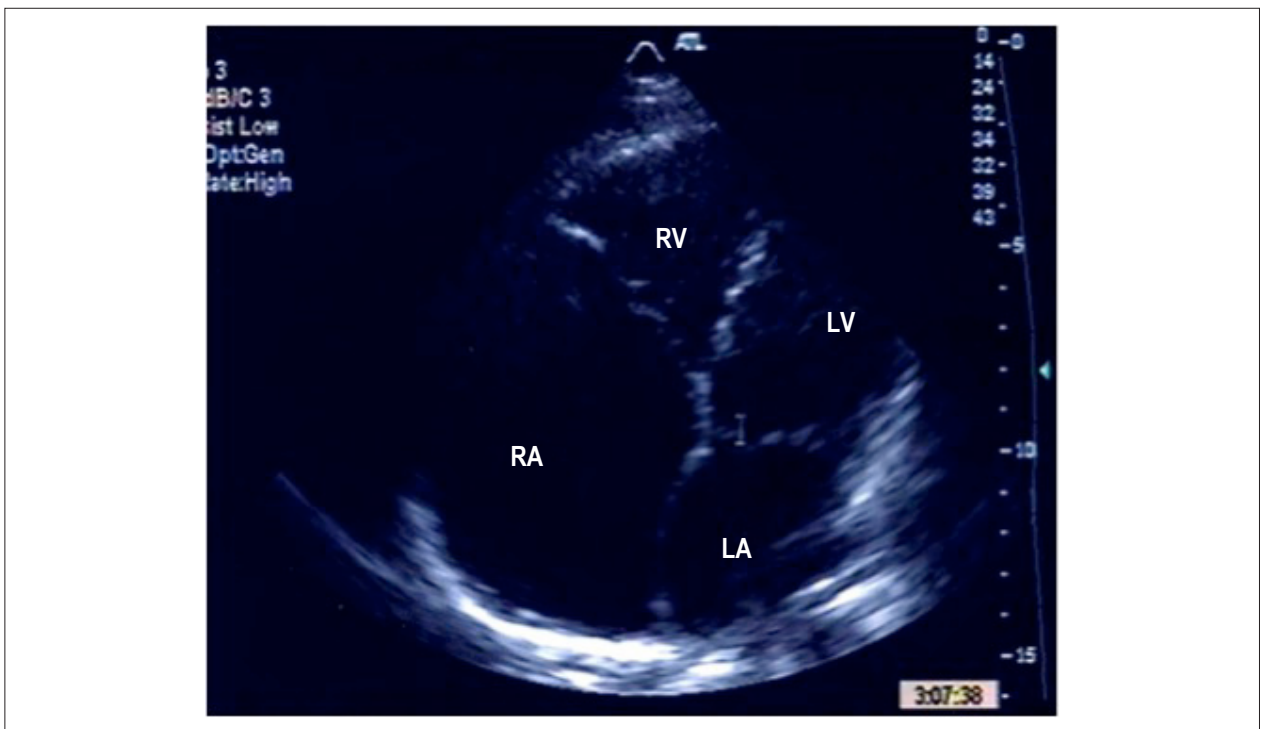


Figure 1 – Transthoracic echocardiography (TTE) showing severe right atrial dilation. RV: right ventricle; LV: left ventricle; RA: right atrial; LA: left atrial.

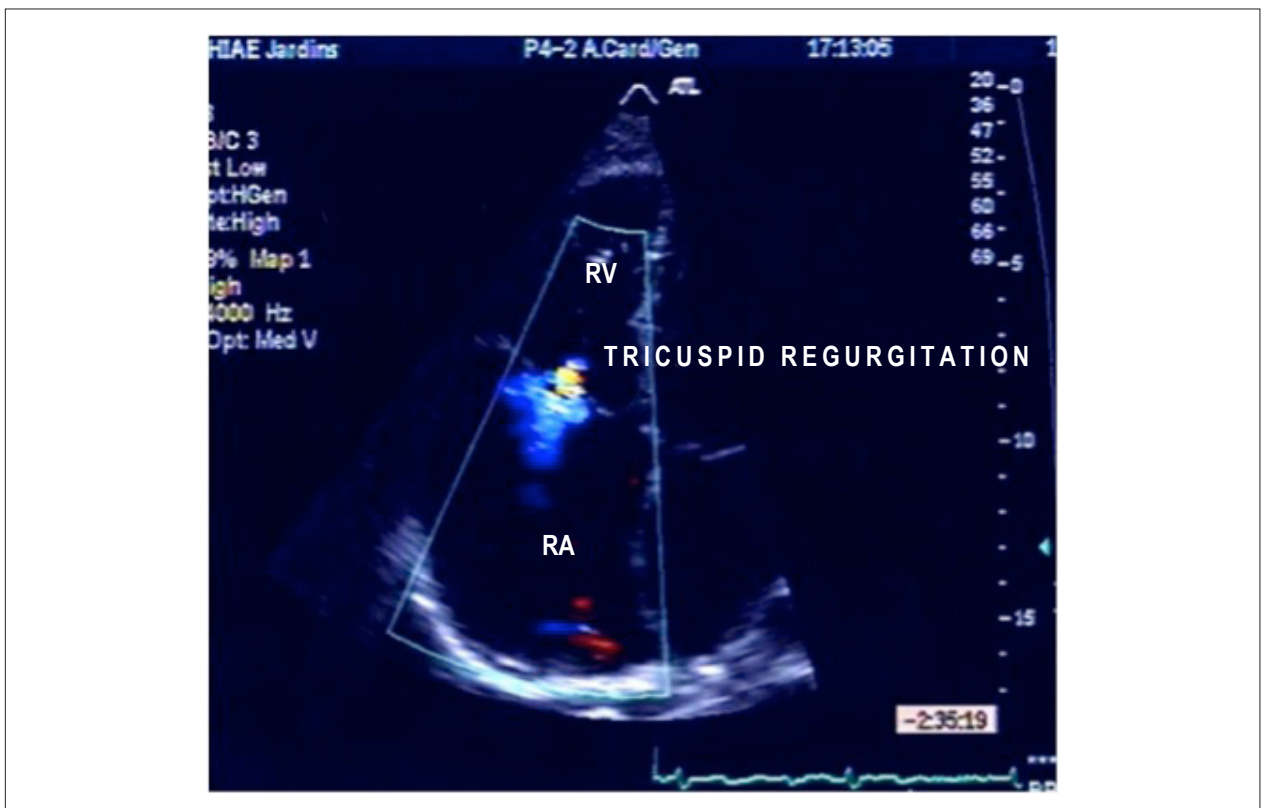
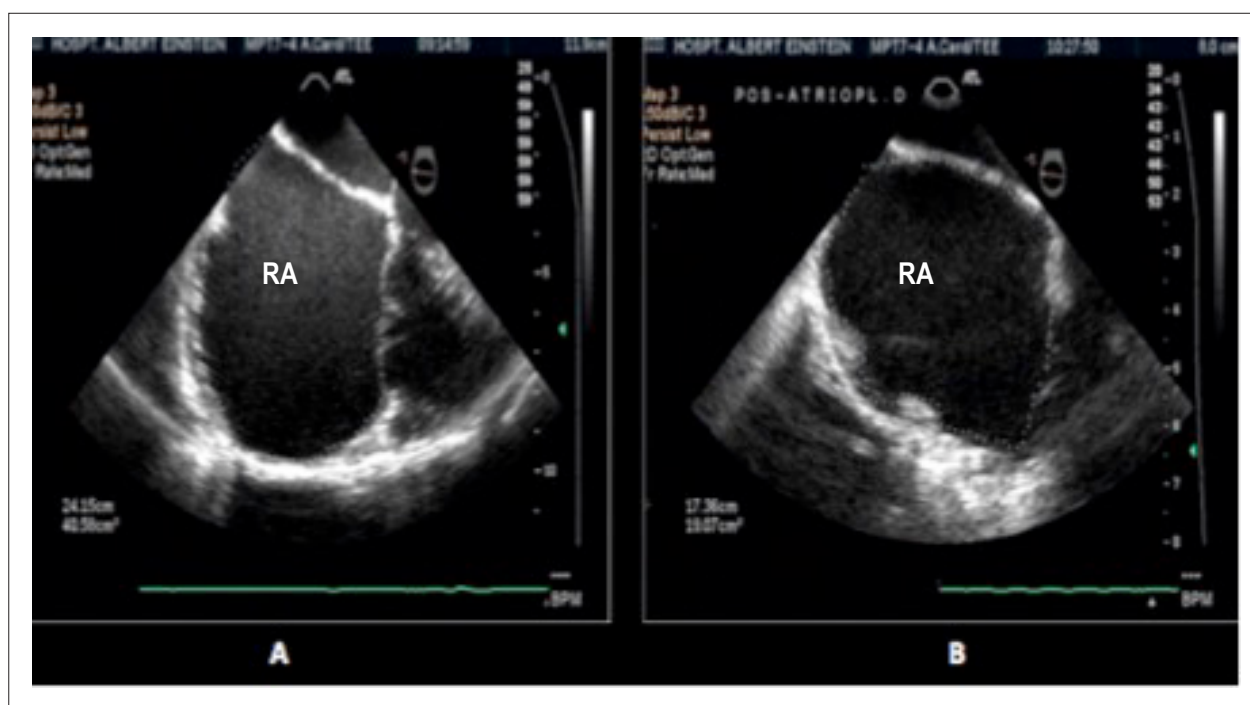
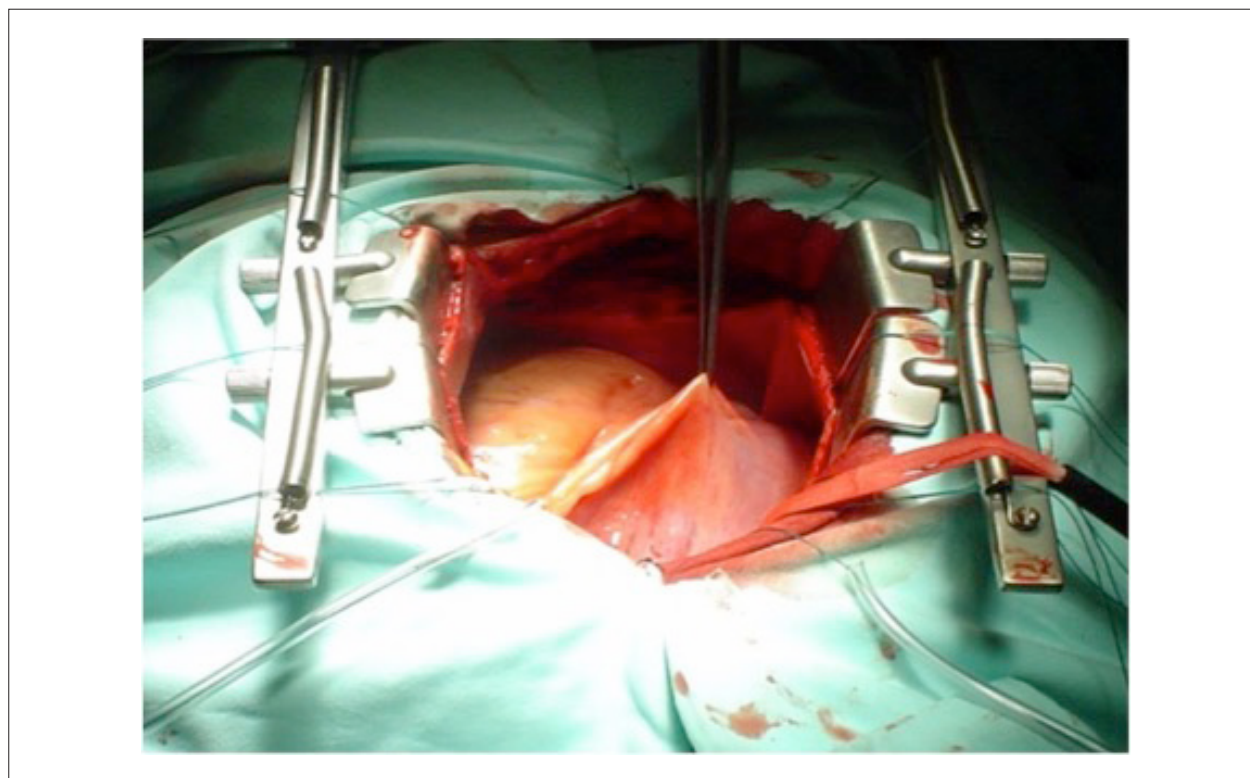


Figure 2 – TTE with significant right atrial increase and tricuspid annulus dilation causing slight regurgitation. RV: right ventricle; RA: right atrial.

## Case Report



**Figure 3** – Intraoperative transesophageal echocardiogram (TEE) conducted before partial resection of the right atrium (A) and post-procedural aspect, showing marked reduction in the sectional area of the right atrium (B).



**Figure 4** – Slender appearance of the right atrial wall before the partial resection.

patients with atrial fibrillation or flutter, with good results for the few cases reported with long-term follow-up.<sup>3,7</sup> In this situation, we opted for the surgical treatment preventively, since the patient maintained intense physical activity in an attempt to avoid embolism.

### Conclusion

Echocardiography is key for both the initial diagnosis and for proper patient management, particularly to avoid iatrogenesis.

### Authors' contributions

Research creation and design: Costa RCPL, Oliveira EM, Rodrigues ACT, Oliveira SA; Data acquisition: Oliveira EM, Rodrigues ACT, Oliveira SA; Analysis and interpretation of

data: Rodrigues ACT, Oliveira EM; Manuscript drafting: Costa RCPL, Rodrigues ACT; Critical review of the manuscript for important intellectual content: Costa RCPL, Vieira MLC, Morhy SS.

### Potential Conflicts of Interest

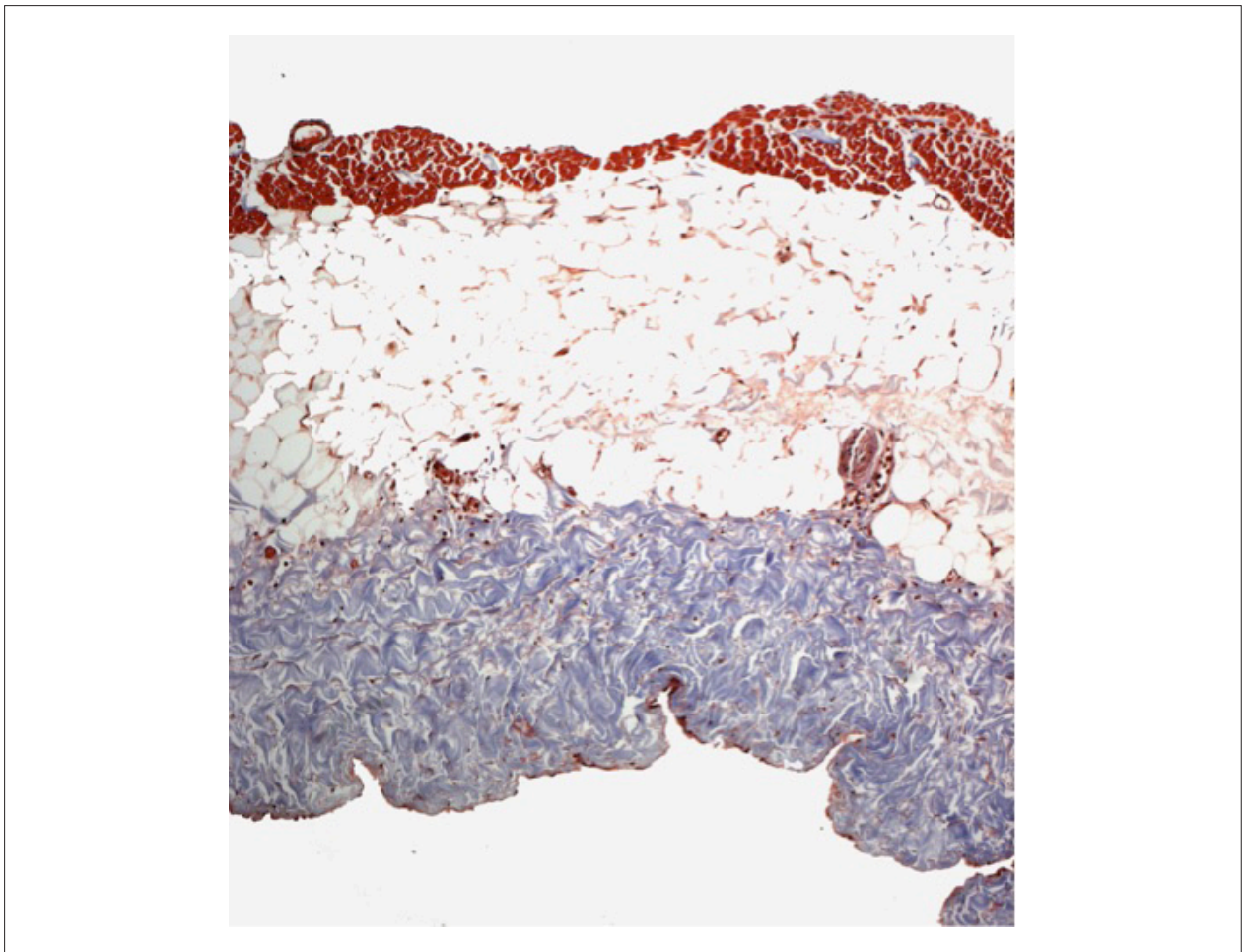
There are no relevant conflicts of interest.

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This study had no external funding sources.

### Academic association

This study is not associated with any graduate program.



**Figure 5** – Photomicrograph of the resected right atrial wall showing attenuation and focal absence of the myocardial layer (dark red color) and fatty infiltration. Trichromatic Masson staining objective lens magnification of 10X.

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