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Case Reports

Idiopathic Dilatation of the Right Atrium: Diagnosis and Management

Ljiljana Rankovic-Nicic, MD[1]; Milica Dragicevic-Antonic, MD[1]; Goran Loncar, MD, PhD[1,2]; Masa Petrovic, BS, MS[1,2]; Zelimir Antonic, MD[1]; Milovan Bojic, MD, PhD[1]

[1]Institute for Cardiovascular Diseases "Dedinje," Belgrade, Serbia
[2]University of Belgrade Faculty of Medicine, Belgrade, Serbia

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Abstract

Idiopathic dilatation of the right atrium is a rare condition with an unknown etiology. It is characterized by a significant enlargement of the right atrium without the presence of other valvopathies, intracardiac shunts, or pulmonary hypertension. This report presents the case of a 50-year-old woman with a significantly enlarged right atrium that was identified at birth; however, a definitive diagnosis was made later in life. The patient did not have any genetic diseases. Through the help of regular follow-up, anticoagulant therapy, previous radiofrequency ablation, and antiarrhythmic medications, she was able to carry a pregnancy to full term and live a regular life.

Keywords: Right atrium; dilatation; anomalies; congenital

Case Report

Presentation and Physical Examination

This report presents the case of a 50-year-old woman with a significantly enlarged right atrium (Fig. 1). The patient presented to the clinic for a regular follow-up. She reported occasional pressure in the chest and fatigue that occurred with less effort and lasted for a short duration. She had a New York Heart Association functional classification of II/III.

Medical History

The patient was initially diagnosed with Ebstein anomaly at birth; however, during her fourth year of life, it was established that she did not meet all the diagnostic criteria for Ebstein congenital heart defect; additionally, there were no other secondary causes (ie, valvulopathies, shunts, or pulmonary hypertension) for the significant enlargement of the right atrium. As a result, the patient was later diagnosed with an idiopathic dilatation of the right atrium.

The patient reported that during her childhood, she began to feel pronounced palpitations and the skipping of heartbeats and that, as a result of several arrhythmic events, she had been hospitalized on several occasions. During an 8-year period (2004-2012), the patient underwent 5 radiofrequency ablations for supraventricular re-entry tachycardia, the last being 11 years ago. At age 26 years, the patient carried a pregnancy to full term, with the last month of pregnancy spent in the hospital on heparin.

During her pregnancy, the patient underwent regular heart ultrasonography for both herself and the baby. The patient was also started on anticoagulant therapy, which she still takes regularly.

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Corresponding author: Masa Petrovic, BS, MS, University of Belgrade Faculty of Medicine, dr Subotića Starijeg 8, Beograd, Serbia 11000 (5rovicmasa@gmail.com)

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During the last 11 years, the patient has reported having no rhythm irregularities and was regularly monitored with ultrasonography. At the time of this report, the patient was on a treatment regimen that included warfarin, flecainide, and bisoprolol.

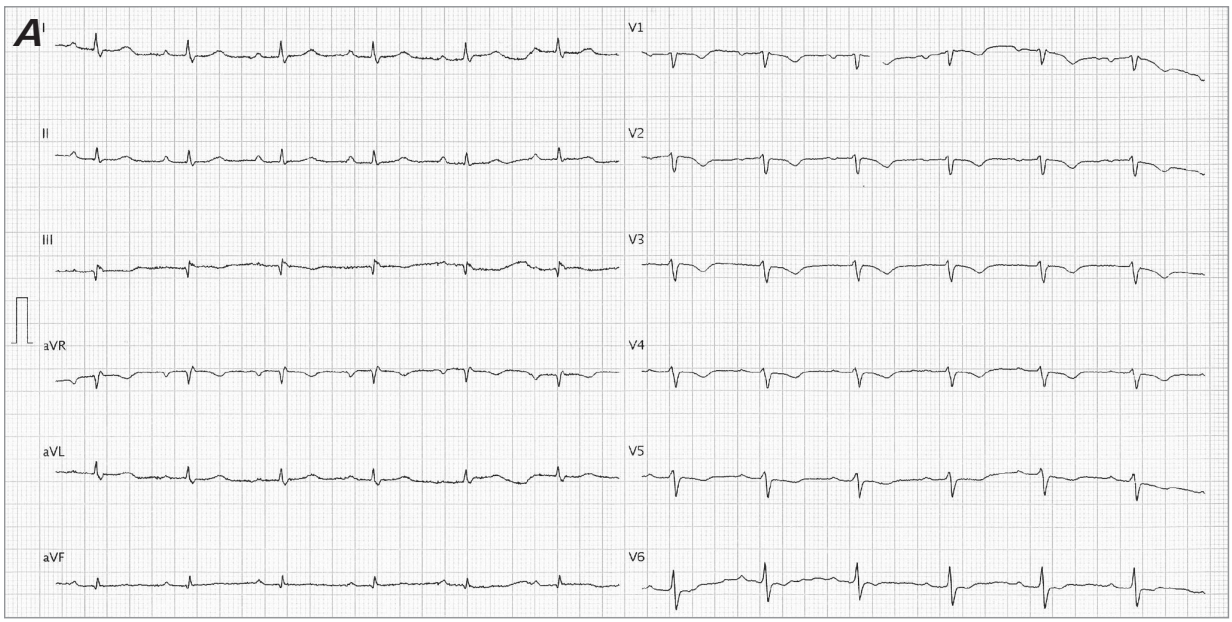
Recently, 24-hour Holter monitoring of electrocardiography recorded a sinus rhythm mean heart rate of 71/min (range, 63-90/min), and 1 supraventricular extrasystole was registered. No atrioventricular node disorders or significant pauses were registered. Her blood test results were within normal limits.

Key Points

- Idiopathic dilatation of the right atrium is a rare anomaly with a variable clinical presentation, from asymptomatic to cardiac failure.
- The use of various cardiac imaging modalities, such as MRI, CT, and echocardiography, has proven to play a pivotal role because the definitive diagnosis is reached through the exclusion of other anomalies.
- Long-term follow-up and imaging are important for progression to be monitored and intervention provided when necessary.

Abbreviations and Acronyms

CT computed tomography
MRI magnetic resonance imaging



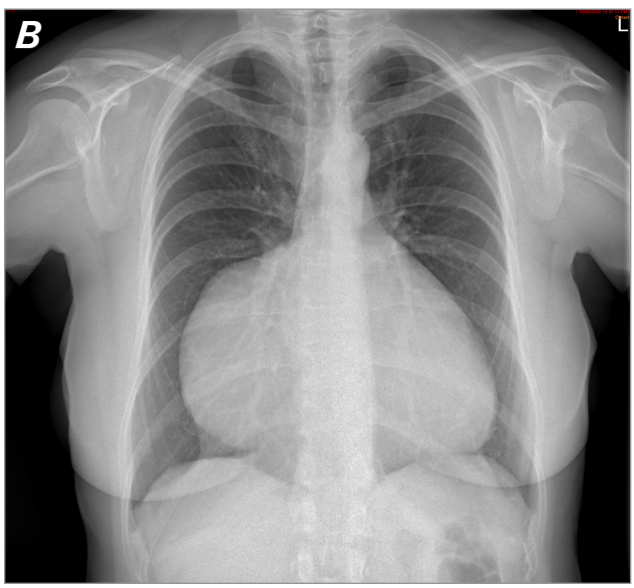


Fig. 1 A) Electrocardiogram showing sinus rhythm, a heart rate of 70/min, normogram, q in lead III, aVF, microvoltage, and negative T V1-V6. **B**) A chest radiograph with posterior-anterior projection showing an enlarged cardiac shadow.

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Differential Diagnosis

The differential diagnoses for this patient's presentation included Ebstein anomaly, Uhl anomaly, hypoplastic left heart syndrome, total anomalous pulmonary venous return, and atrial septal defect. One of the principal features of Ebstein anomaly is apical displacement of the septal leaflet of the tricuspid valve by at least 8 mm/m^2. In this patient, the displacement was 4.8 mm/m^2, and both leaflets were displaced, which eliminated Ebstein anomaly.

Technique

During follow-up, transthoracic echocardiography showed a significantly dilated right atrium measuring 160 mm × 120 mm, with dense spontaneous contrast observed without clearly organized thrombotic masses (Fig. 2). The septal leaflet of the tricuspid valve was positioned 9 mm lower than the anterior mitral leaflet, which did not meet the diagnostic criteria for Ebstein anomaly. In addition, mild to moderate tricuspid insufficiency was registered. Right ventricular systolic pressure measured 35 mm Hg. On the interatrial septum, a smaller shunt at the level of the fossa ovalis was observed. The left ventricle was small, with preserved contractility and good ejection fractions and without any kinetic irregularities. Cardiac computed tomography (CT) scans revealed an enlarged right atrium that measured 165 mm × 92 mm (Fig. 3).

To clarify the results of echocardiography, cardiac magnetic resonance imaging (MRI) scanning with late gadolinium enhancement was performed with a 1.5 T scanner (SIGNA Artist; GE) (Fig. 4). Black blood magnetic resonance and cine–magnetic resonance scanning revealed that the heart had shifted to the left side of the chest as a result of the significantly enlarged right atrium (149 cm^2, 81 cm/m^2, respectively; volume = 814 mL/m^2) pressing against the right ventricle and displacing the septal leaflet of the tricuspid valve by 7 mm in relation to the front leaflet of the mitral valve, with a small stream of regurgitation.

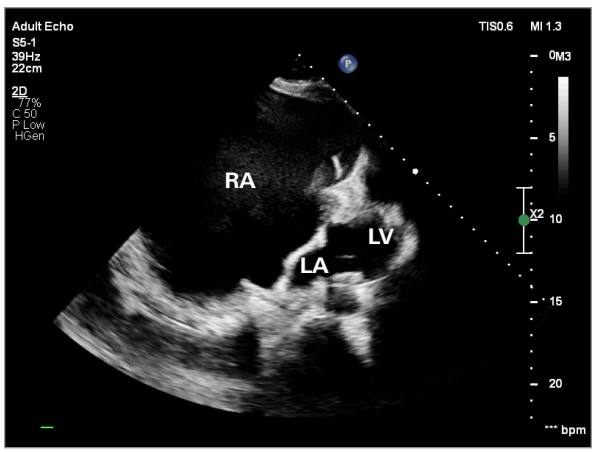

Fig. 2 Transthoracic echocardiogram 4-chamber view showing a markedly enlarged right atrium with spontaneous contrast.

LA, left atrium; LV, left ventricle; RA, right atrium.

The right ventricle had weakened systolic function (ejection fraction = 44%), and the left ventricle (ejection fraction = 55%) had normal systolic and diastolic volume of the cavum. The right ventricle had an end-diastolic volume of 67 mL/m^2 and end-systolic volume of 37 mL/m^2, and the left ventricle had and end-diastolic volume of 56 mL/m^2 and end-systolic volume of 25 mL/m^2. At the interatrial septum, a small communication of the foramen ovale aperture was present. The right ventricle was without signs of diffuse or focal fibrosis.

Outcome

In addition to her existing treatment regimen (warfarin, flecainide, and bisoprolol), the patient was prescribed 20 mg per day furosemide and 2 g per day potassium. Given the complexity of the case, the patient was referred to a specialized cardiac surgery clinic for adult congenital anomalies.

Latest Follow-Up

On the most recent follow-up, the patient reported decreased events of chest pressure, fatigue, and palpitations. She had a New York Heart Association functional classification of I/II.

Discussion

Idiopathic dilatation of the right atrium was first described in 1955 by Bailey in *Surgery of the Heart*.[1] It represents a rare condition, with only a few sporadic cases previously reported and an unknown etiology. Idiopathic dilatation of the right atrium is characterized by a significant enlargement of the right atrium without the presence of other valvopathies, intracardiac shunts, or pulmonary hypertension. This anomaly can be detected in any period of life, from the fetal period to adulthood.[1-4] Because early diagnosis during the fetal stage is possible, research has suggested that this

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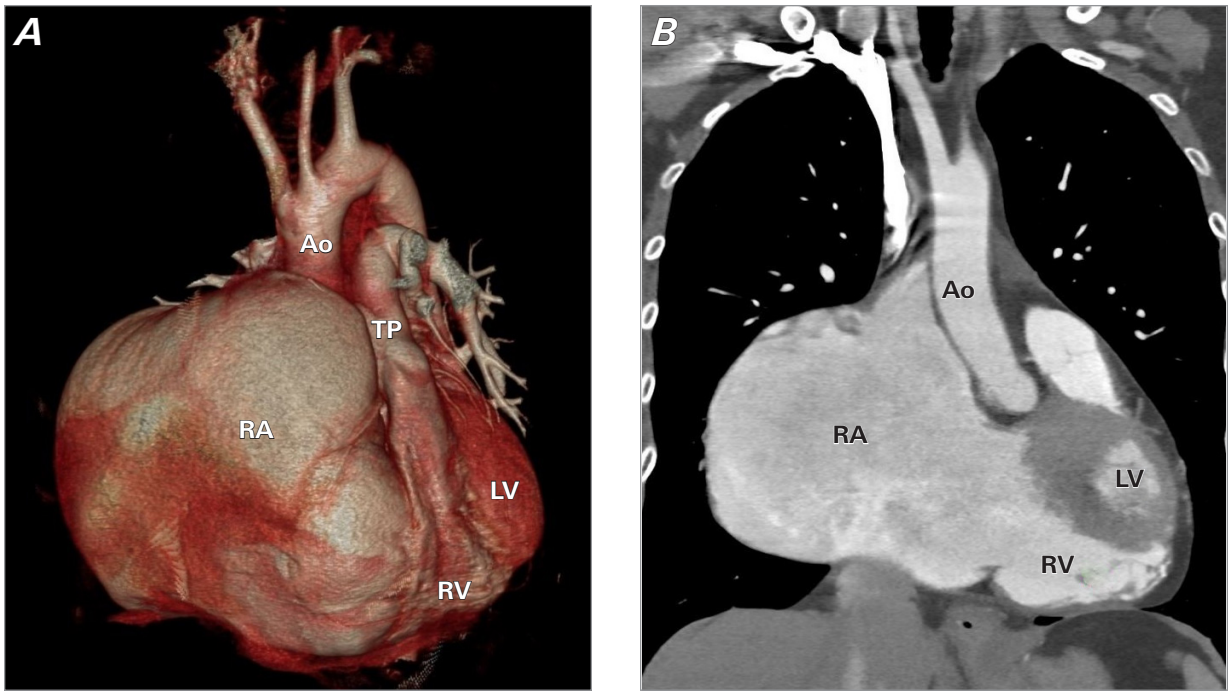

Fig. 3 A) Electrocardiography gated cardiac computerized tomography image with volume rendering reconstruction in the coronal view depicting a significantly enlarged right atrium. **B**) Computerized tomography image in the coronal view showing a markedly enlarged right atrium.

Ao, aorta; LV, left ventricle; RA, right atrium; RV, right ventricle; TP, pulmonary trunk.

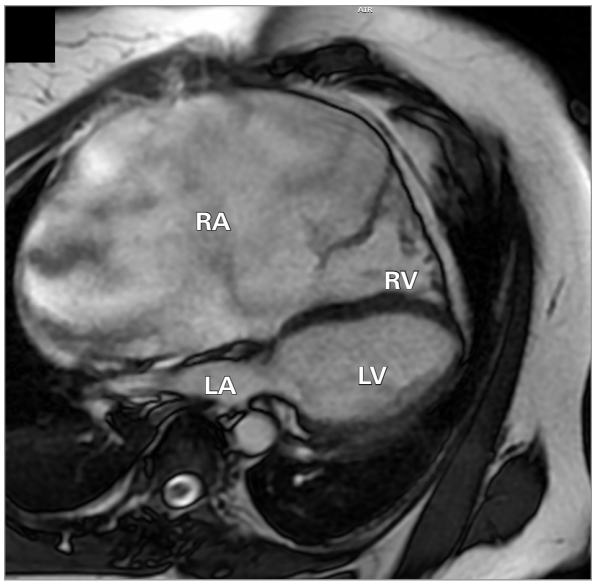

Fig. 4 Cardiovascular magnetic resonance image in 4-chamber view showing cardiomegaly, with a normal-size right ventricle.

LA, left atrium, LV-left ventricle; RA, right atrium; RV, right ventricle.

anomaly may be congenital; however, the pathogenetic mechanism is yet to be identified.[4] Patients with this anomaly have an overall increased risk of sudden cardiac arrest and typically present with arrhythmias and cardiac insufficiency; however, there have been some cases in which the patient is completely asymptomatic.[5-8] Despite the limited number of cases reported, data do not show any difference between sexes or racial groups.[9]

The diagnosis of this anomaly involved systematically ruling out more common causes that could contribute to the enlargement of the right atrium, such as Ebstein

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anomaly, tricuspid insufficiency, and atrial septal defect. The main diagnostic modality is echocardiography; however, other diagnostic modalities, such as transesophageal echocardiography, CT, and MRI, may also significantly aid in the assessment of the right atrium and entire heart to exclude other cardiac pathologies.[10] Although many patients may remain asymptomatic throughout their lives, some authors have suggested that other patients may experience arrhythmias, palpitations, chest pain, shortness of breath, fatigue, and syncope.[7,8]

The optimal treatment modality, whether surgical or medicinal, is still a widely debated topic. In the case of idiopathic dilatation of the right atrium, the use of antiplatelet drugs is widely accepted, whereas the use of anticoagulant treatment is indicated only in patients with the presence of atrial thrombosis or those who have a high risk of thrombus formation. In the case of rhythm disorders, antiarrhythmic drugs are typically used, and in some cases of refractory arrhythmias, radiofrequency ablation or the Maze surgical procedure is recommended.[7,8,11,12] Some authors have suggested that surgical treatment is indicated in adults with rapid enlargement of the atrium, while surgery in the asymptomatic pediatric patient population remains a topic for debate. Surgical techniques vary from atrial resection to atrial reduction plasty without cardiopulmonary bypass.[12,13] To prevent postresection dilation of the right atrium, Kalangos et al[14] presented a method of external reinforcement of the lateral wall of the right atrium with the use of an autologous pericardium.

In this patient, the enlarged right atrium was verified immediately after birth without the presence of any familial genetic disease; however, the definitive diagnosis was made later in life, when she was 4 years old. Through regular follow-ups and the use of anticoagulant therapy, previous radiofrequency ablations, and antiarrhythmic medications, she was able to carry a pregnancy to full term and live a regular life.

Although idiopathic dilatation of the right atrium is a rare anomaly, it should not be forgotten. The use of various cardiac imaging modalities, such as MRI, CT, and echocardiography, has proven to play a pivotal role because the definitive diagnosis is reached through the exclusion of other anomalies. Given the potential role of genetics, the screening of all relatives is highly recommended. In the future, as the field of cardiogenetics advances and more, similar cases are encountered in practice, current knowledge regarding this pathology may be expanded, allowing for better catered care for patients.

Article Information

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