OMCR

Oxford Medical Case Reports, 2020;2,69–74

doi: 10.1093/omcr/omaa011
Case Report

CASE REPORT

Giant right atrium: a long-term complication of rheumatic heart disease

Ashraf Abugroun[1,*], Mohamed Taha[2], Hussein Daoud[3] and Walid Ibrahim[4]

[1]Division of Geriatrics, Department of Internal Medicine, Wayne State University, Detroit, MI 48201, USA, [2]Department of Internal Medicine, University of Iowa Hospitals and Clinics, Iowa, IA 52242, USA, [3]Department of Internal Medicine, Advocate Illinois Masonic Medical Center, Chicago, IL 60657, USA, [4]Department of Cardiology, Jackson Memorial Hospital/University of Miami, FL 33136, USA

*Correspondence address. Department of Geriatrics, Wayne State University, 4201 St. Antoine St., Suite 5C, Detroit, MI 48201, USA. Tel: +1-313-577-5092; Fax: +1-313-745-4707; E-mail: Abugroun@live.com

Abstract

Giant right atrium (RA) is a rare entity often seen during childhood due to congenital anomalies. Limited literature has reported such finding in patients with rheumatic valvular heart disease. Here we present a case of a 68-year-old female with a history of rheumatic valve disease treated with a Starr Edwards mechanical ball-in-cage mitral valve replacement and tricuspid valve annuloplasty ring procedures. The patient developed heart failure and had multiple hospital admissions over three decades for heart failure exacerbations mostly triggered by medication and dietary non-compliance. She eventually developed a giant RA that filled most of her thorax. This case demonstrates an extreme form of cardiac remodeling caused by long-term rheumatic valvular heart disease.

INTRODUCTION

Giant right atrium (RA) is a rare entity often seen during childhood due to congenital anomalies. Limited literature has reported such finding in patients with rheumatic valvular heart disease. Pulmonary hypertension (PH) secondary to mitral valve dysfunction can lead to right ventricular (RV) failure and subsequent enlargement of the RA. While patients with a giant RA can remain asymptomatic, most patients present with symptoms of heart failure and are at higher risk for atrial fibrillation and thrombus formation. In this report, we discuss the case of a 68-year-old female with a history of rheumatic heart disease and poorly controlled heart failure who, over time, developed PH and, later, RV dilatation, functional tricuspid regurgitation (TR) and a giant RA.

CASE REPORT

A 68-year-old female was admitted to the hospital for acute decompensated heart failure. She had a history of rheumatic heart disease with severe mitral and tricuspid valvular disease complicated by heart failure and subsequent PH. Her mitral valve was replaced three decades earlier using a mechanical Starr-Edwards ball-in-cage valve. She also had a tricuspid annuloplasty ring repair for tricuspid insufficiency. Other comorbidities included hypertension, hyperlipidemia, permanent atrial fibrillation and a remote history of a cerebrovascular accident with residual left-sided weakness. The patient had a prior angiogram that showed normal coronaries and a globally depressed ventricular function. She also had an automatic implantable cardioverter defibrillator (AICD) placed for primary

Received: June 17, 2019. **Revised:** November 23, 2019. **Accepted:** January 29, 2020

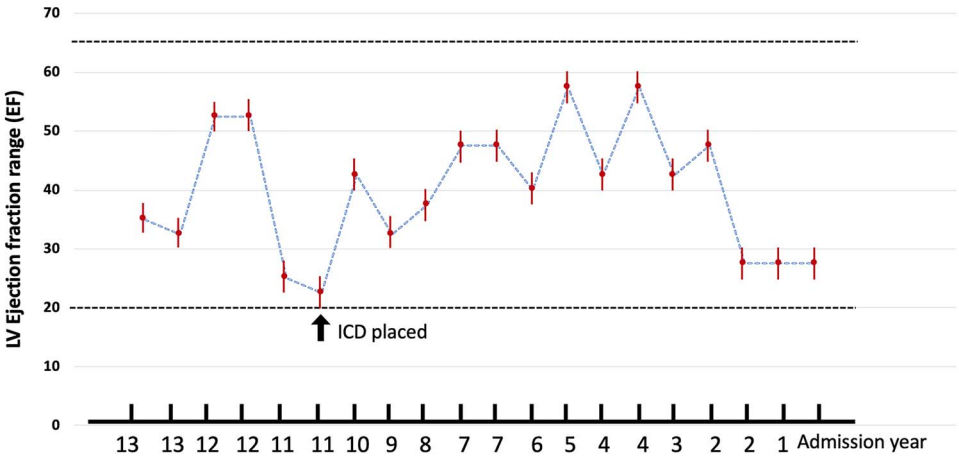

Figure 1: Upper and lower limits of the estimated left ventricular ejection fraction (LVEF) during hospital admissions over the past 13 years. Note: Duplicate number of years indicates repeated echo performed during the same year.

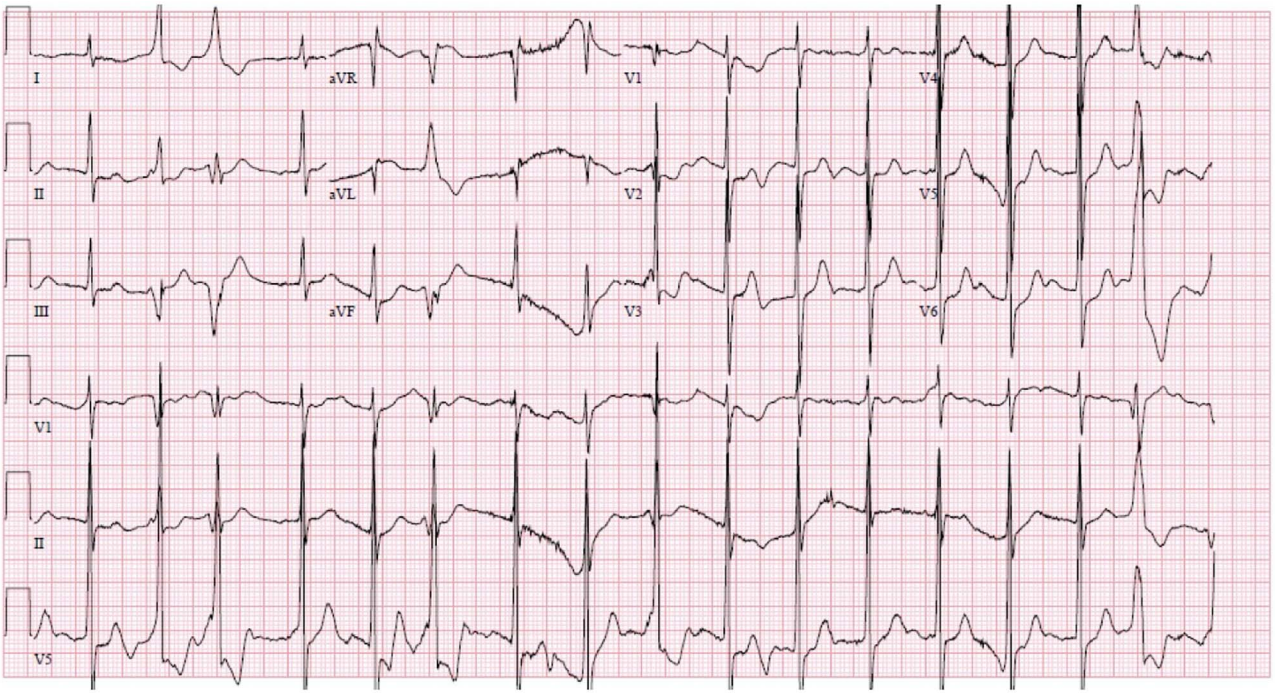

Figure 2: Electrocardiogram (ECG) showing a junctional rhythm with complete heart block, prolonged QT interval (522 ms) and LVH with repolarization abnormality.

prevention of sudden cardiac death given her low ejection fraction (Fig. 1).

Her home medications included lisinopril, metoprolol succinate, digoxin, metolazone, furosemide, spironolactone and warfarin. Over the past 15 years, she had frequent hospitalizations for heart failure exacerbation mostly due to dietary non-adherence and medication non-compliance (Fig. 1).

Upon hospital arrival, she reported progressive fatigue and decreased walking capacity from two blocks at baseline to less than half a block. She also reported mild lower extremity swelling but denied any chest pain, palpitations, orthopnea, paroxysmal nocturnal dyspnea or cough. Additionally, she experienced intermittent dizziness, particularly when changing position.

She was found to have a blood pressure of 130/58 mmHg and a heart rate of 55 beats per minute. On physical examination, visible neck pulsations and a raised jugular venous pressure were present. Furthermore, a loud S1 with a metallic sound, a soft systolic murmur heard throughout her chest and mild lower extremity edema were noted. An electrocardiogram showed left ventricular hypertrophy, a junctional rhythm, complete heart block and a prolonged QT interval of 522 ms (Fig. 2). X-ray and a computed tomography (CT) scan were notable for stable cardiomegaly (Fig. 3). A transthoracic echocardiogram showed a markedly dilated left and right atrium, dilated right ventricle with reduced systolic function, wide-open TR, markedly reduced left ventricular systolic function with an ejection fraction of 25–30%, severe diffuse hypokinesis with regional variations and a

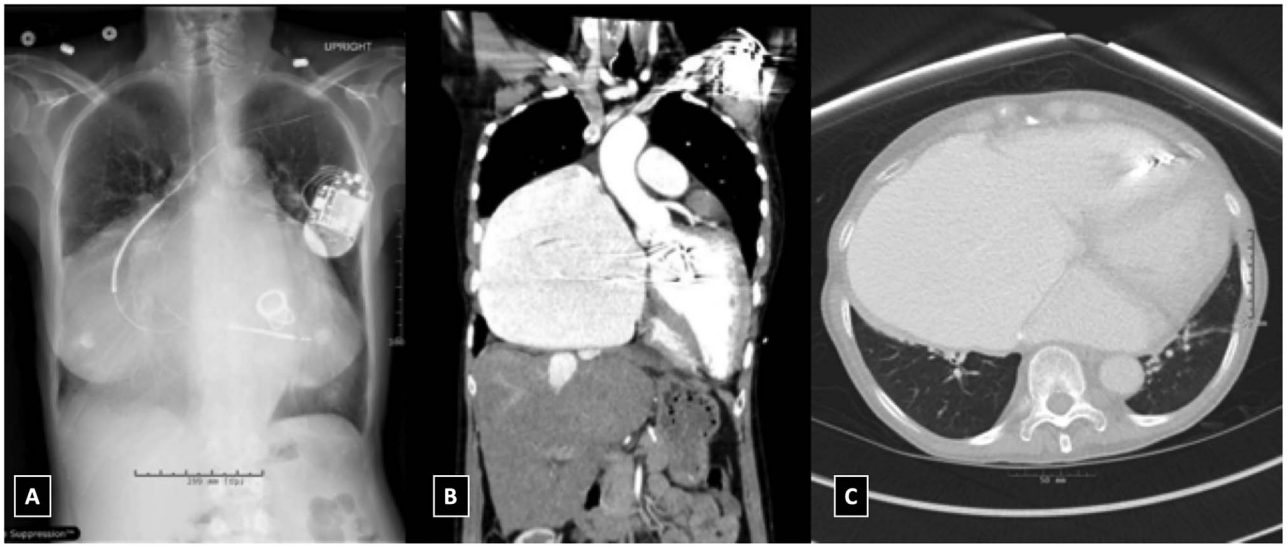

Figure 3: (**A**) Chest X-ray showing marked cardiomegaly with giant right ventricle filling most of the thoracic cage. (**B**) CT of the chest showing the heart and great vessels. (**C**) CT scan of the chest showing the four chambers of the heart.

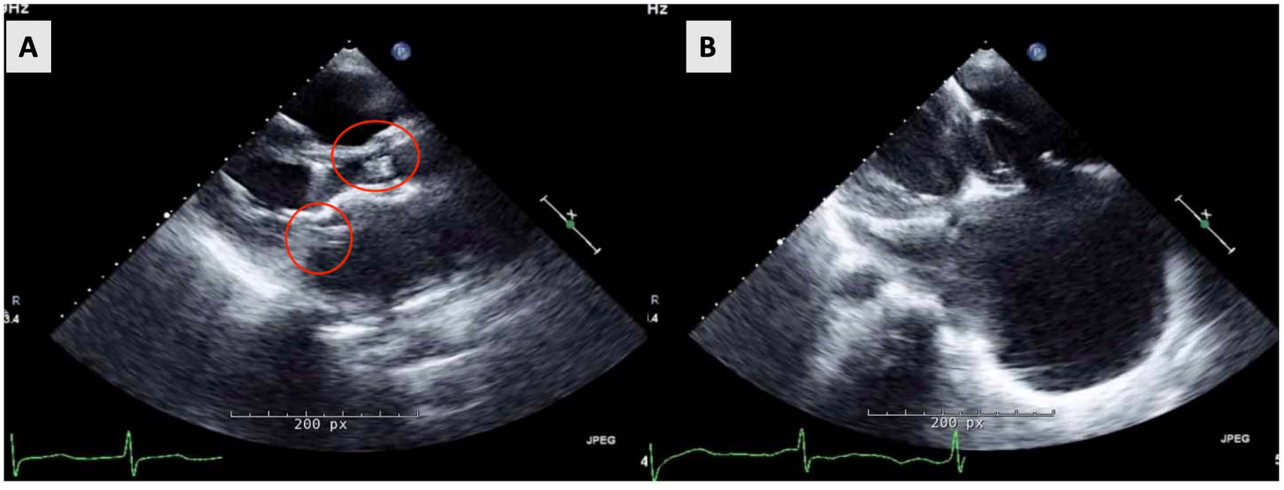

Figure 4: Transthoracic echo. (**A**) Long-axis view; thickening of aortic and mitral leaflets and dilated RV. (**B**) Four chamber view; marked dilated RA no thrombus seen.

mechanical mitral valve with a normal range of motion. The thickness of the ventricular septum was mildly increased with septal motion dyssynergy. The central venous pressure was 15 mmHg and the pulmonary artery systolic pressure (PASP) was 48 mmHg (Fig. 4; Table 1). The patient responded well to intravenous furosemide with improvement in her symptoms and was eventually discharged with the continuation of her goal-directed medical therapy regimen.

DISCUSSION

The majority of previously reported cases of giant atrial enlargement were inherited and mostly congenital and seen in pediatric patients [1]. We herein report a case of a giant RA seen in an adult patient with chronic rheumatic valve disease. The pathophysiology of a giant RA is not fully understood, as it is not seen in all patients with chronic valvular dysfunction who develop pressure and volume overload [2]. Interestingly, almost all previously reported cases of giant atrial enlargement in adults were in patients with a prior history of rheumatic heart disease (Table 2). Previous studies suggested a possible role of rheumatic pancarditis with chronic inflammation and fibrosis causing weakening of the atrial wall, thus, making it more susceptible to dilatation by increased intra-atrial pressure [3, 4]. The pathogenesis of giant atrial enlargement is, therefore, likely multifactorial and involves volume and pressure overload in the presence of a weakened atrial wall, leading to dilatation.

In our case, the patient had a history of rheumatic valvular heart disease with eventual mitral valve replacement and tricuspid valve annuloplasty. She later developed non-ischemic dilated cardiomyopathy and heart failure (Fig. 1). A prior angiogram showed normal coronaries; however, an echocardiogram noted regional variations in the wall motion. Hence, it is unclear if there was a prior myocardial infarction versus rheumatic myocarditis. While the PASP was elevated, it was possibly underestimated due to the presence of functional TR. Also, although the RV systolic pressure was not increased

Table 1: TTE parameters from hospital admission.

Left ventricle	
LV ID, ED, PLAX	(H) 5.4 cm
LV ID, ES, PLAX	(H) 4.2 cm
IVS thickness, ED	(H) 1.2 cm
LV PW thickness, ED	(H) 1.2 cm
IVS/LV PW ratio, ED	1.03
LV ejection fraction, 1-p	(L) 34%
A4C	
Mitral E-wave peak velocity	133 cm/sec
Mitral deceleration time	475 ms
LVOT	
LVOT ID, S	2.2 cm
LVOT ID	2.2 cm
LVOT peak velocity, S	0.72 m/s
LVOT VTI, S	10.5 cm
LVOT peak gradient, S	2 mmHg
Stroke volume (SV), LVOT DP	40 mL
Aortic valve peak velocity, S	1.9 m/s
Aortic valve mean velocity, S	1.27 m/s
Aortic valve VTI, S	28.6 cm
Aortic mean gradient, S	8 mmHg
Aortic peak gradient, S	14 mmHg
VTI ratio, LVOT/AV	0.37
Aortic valve area, VTI	1.4 cm^2
Aortic valve area, peak velocity	1.44 cm^2
Aortic regurg pressure half-time	498 ms
Aorta	
Aortic root ID, ED	3.1 cm
RVOT	
RVOT peak velocity, S	0.89 m/s
RVOT mean velocity, S	0.41 m/s
RVOT VTI, S	11.6 cm
RVOT peak gradient, S	3 mmHg
Mitral valve	
Mitral VTI at leaflet coaptation	45.4 cm
Mitral mean gradient, D	5 mmHg
Mitral valve area, LVOT continuity	0.9 cm^2
Mitral annulus VTI, D	45.4 cm
Tricuspid valve	
Tricuspid regurg peak velocity	2.9 m/s
Tricuspid peak RV-RA gradient	33 mmHg
Systemic veins	
Estimated CVP	15 mm
Right ventricle	
RV ID, ED, PLAX	6.0 cm
RV pressure, S, DP	48 mm
Pulmonic valve	
Pulmonic valve peak velocity, S	1 m/s
Pulmonic valve VTI, S	15.5 cm
Pulmonic acceleration time	71 ms
Pulmonic mean gradient, S	2 mmHg
Pulmonic peak gradient, S	4 mmHg
Pulmonic regurg velocity	1.65 m/s
Pulmonic regurg gradient	11 mmHg

CVP, central venous pressure; TTE, transthoracic echocardiogram; LV, Left ventricle; ID, Internal diameter; ED, End diastolic; ES, End systolic; VTI, Velocity time integral; SVC, Superior vena cava; IVC, Inferior vena cava; IVS, Interventricular septum; LVOT, left ventricular outflow tract; RVOT, Right ventricular outflow tract.

relative to the right atrial pressure, the atrial pressure was likely markedly elevated as a result of wide-open TR [5]. The absolute RV systolic pressure was, therefore, likely at least moderately increased. In the presence of poorly controlled heart failure with PH, the patient developed RV dilatation, functional TR and progressive dilation of the RA to a giant size that filled most of her thorax.

The patient's history of atrial fibrillation could have also contributed to her giant RA and functional TR. Past studies have shown that atrial fibrillation can lead to atrial remodeling and RA dilatation with subsequent loss of valve coaptation during systole in patients without PH [6, 7]. Though our patient's TR was functional, ICD implantation is also a known cause of TR through mechanisms such as valve leaflet impingement, perforation or adhesion [8].

Patients with an enlarged RA can be asymptomatic, but symptoms, when reported, are related to the underlying left and RV dysfunction and include dyspnea, fatigue and edema—as seen in our patient. In addition, patients with a giant RA are at higher risk for atrial arrhythmias, thrombus formation and left recurrent laryngeal palsy (Ortner's syndrome) [9]. Available surgical management options include size-reducing atrioplasty and tricuspid valve annuloplasty for TR. However, case selection based on comorbidities and risk factors is required to reduce postoperative morbidity and mortality, and the risk for arrhythmia remains elevated despite surgical intervention. Given the high risk for thrombus formation and embolization, all patients with a giant RA are mandated to have anticoagulation [10]. Our patient has been on lifelong anticoagulation, but surgical repair of the TR was not attempted as tricuspid repair or replacement was not expected to improve her RV dilation. Also, the patient was a very high-risk surgical candidate given her poor left ventricular systolic function and multiple comorbidities.

CONCLUSION

Giant RA is a rare complication of rheumatic heart disease, particularly in patients who eventually develop elevated cardiac pressures or volumes. Giant RA can be asymptomatic but may present with heart failure symptoms and carries a higher risk for adverse outcomes including arrhythmias and thromboembolic complications—which can significantly worsen prognosis and increase mortality. Early treatment of rheumatic heart disease and its complications is important in decreasing the probability of developing a giant RA. Medical management of a giant RA includes controlling RV and pulmonary pressures as well as anticoagulation. Surgical options include atrioplasty and tricuspid valve annuloplasty.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

None.

ETHICAL APPROVAL

No ethical approval required.

INFORMED CONSENT

The patient has given written consent for publication of this case report.

Table 2: Summary of previously reported cases of giant atrial enlargement.

Age/gender	Presentation	Structural heart abnormalities	Echo finding(s)	Clinical outcome(s)
45, F (7)	- Dyspnea - Ascites, hepatosplenomegaly, pretibial edema - Atrial fibrillation	- MV replacement (11 years earlier), severe TR and TS	- RA size: 12 × 14 cm - EF: 65% - PAP: 38 mmHg	- A 33-mm bioprosthesis was implanted for the TV - The RA volume was reduced by internal plication of RA from the SVC into the IVC
25, F (8)	- Engorged and pulsatile jugular vein with prominent 'V'-wave and large 'Y' descent, grade III/VI pan-systolic murmur at left lower parasternal area and pulsatile hepatomegaly	- Severe TR and trivial MR	- RA size: 12.1 × 10.5 cm - RA area approx. 97.5 cm^2	NA
58, F (9)	- Dyspnea, NYHA functional class IV, palpitations, and peripheral edema - Did not see a cardiologist for several years - Atrial fibrillation	- MS	- RA area: 80.6 cm^2 - RA volume: 621 ml - EF: 64%	- Furosemide and isosorbide dinitrate, digoxin and spironolactone
68, F (6)	- Dyspnea, NYHA functional class III, palpitations and ankle edema - Atrial fibrillation - For 30 years, on irregular follow-up and not compliant with injectable penicillin	- MS	- RA area: 13.1 cm × 7.05 cm - RA volume: 760 ml - EF: 60%	- IV furosemide, amiodarone, oral digoxin, warfarin and spironolactone - The patient did not consent to surgery and hence was discharged home after medical stabilization
19, M (10)	- NYHA functional class II	- Rheumatic heart disease with severe MS, moderate MR, severe TR and mild AR	- The right and left atrial area were 35 and 42 cm^2, respectively. The calculated right and left atrial volumes were 198 and 165 ml (normal range 22–52 ml), respectively	- MV replacement, TV annuloplasty and LA reduction
	- Pans-systolic murmur at the apex radiating to the axilla (grade III/VI) - Mean jugular venous pressure was elevated (prominent v-waves) with gross pedal edema	- Severe RV dysfunction with adequate LV function	- PAP: 115 mmHg	- Regular penicillin prophylaxis, diuretics, vasodilators and β blockers
65, F (11)	- Progressive dyspnea, lower extremity edema and palpitations - Neck veins were distended with a raised jugular venous pressure - Atrial fibrillation	- Rheumatic heart disease with severe TS	- Enlarged right atrium which had linear dimensions of 95 × 90 mm	- The patient refused the surgery
57, F (12)	- 24 years with dependent edema, ascites, pleural effusion, a large and pulsatile liver and progressive deterioration - Atrial fibrillation	- MS and severe TR	- PAP: 52 mmHg	- TV was replaced, mitral commissurotomy was performed and a portion of the redundant RA wall was resected - The patient expired 10 days postoperatively due to cardiogenic shock
32, F (12)	- 11 years with dependent edema, ascites, pleural effusion, a large and pulsatile liver and progressive deterioration - Atrial fibrillation	- MS and severe TR	- PAP: 59 mmHg	- The patient expired before surgical intervention could be performed
39, M (12)	- 26 years with dependent edema, ascites, pleural effusion, a large and pulsatile liver and progressive deterioration - Atrial fibrillation	- MS and severe TR	- PAP: 82 mmHg	- The MV was replaced and a TV annuloplasty was performed - Expired 4 months postoperatively from consequences of prosthetic MS resulting from interference of ball movement by the LV wall

F, Female; M, Males; TS, Tricuspid stenosis; MR, Mitral regurgitation; EF, Ejection fraction; LA, left atrium; TV, tricuspid valve; MV, mitral valve; MS, mitral stenosis; PAP, pulmonary artery pressure.

ACKNOWLEDGEMENTS

None.

REFERENCES

1. Hofmann SR, Heilmann A, Häusler HJ, Dähnert I, Kamin G, Lachmann R. Congenital idiopathic dilatation of the right atrium: antenatal appearance, postnatal management, long-term follow-up and possible pathomechanism. *Fetal Diagn Ther* 2012;**32**:256–61.
2. Darwazah AK, El Sayed H. Giant left atrium associated with massive thrombus formation. *Thromb J* 2013;**11**:5.
3. Willis HJ. Memories of patients with a giant left atrium. *Circulation* 2001;**104**:2630–1.
4. Roberts WC, Humphries JO, Morrow AG. Giant right atrium in rheumatic mitral stenosis: atrial enlargement restricted by mural calcification. *Am Heart J* 1970;**79**:28–35.
5. Parasuraman S, Walker S, Loudon BL, Gollop ND, Wilson AM, Lowery C *et al*. Assessment of pulmonary artery pressure by echocardiography—a comprehensive review. *Int J Cardiol Heart Vasc* 2016;**12**:45–51.
6. Yamasaki N, Kondo F, Kubo T, Okawa M, Matsumura Y, Kitaoka H *et al*. Severe tricuspid regurgitation in the aged: atrial remodeling associated with long-standing atrial fibrillation. *J Cardiol* 2006;**48**:315–23.
7. Najib MQ, Vinales KL, Vittala SS, Challa S, Lee HR, Chaliki HP. Predictors for the development of severe tricuspid regurgitation with anatomically normal valve in patients with atrial fibrillation. *Echocardiography* 2012;**29**:140–6.
8. Lin G, Nishimura RA, Connolly HM, Dearani JA, Sundt TM, Hayes DL. Severe symptomatic tricuspid valve regurgitation due to permanent pacemaker or implantable cardioverter-defibrillator leads. *J Am Coll Cardiol* 2005;**45**:1672–5.
9. Anandan PK, Shukkarbhai PJ, Cholenahally MN. Giant left and right atrium in rheumatic mitral stenosis and tricuspid regurgitation. *J Cardiovasc Echography* 2015;**25**:113–5.
10. Özen Y, Rabuş MB, Sarıkaya S, Günay D, Özgür MM, Kırali K. Giant Right Atrium. *Koşuyolu Heart J* 2016;**19**(3):208–10.