

55 years old patient with congenital heart malformation (RCD code: IV-1B.1a)

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Abstract

Cor triatriatum is a rare congenital anomaly where the left or right atrium is subdivided by a thin membrane, resulting in three atrial chambers. We present a case of 55 years old female who presented progressive dyspnea with peripheral cyanosis. In a transesophageal and computed tomography and CT threeatrial heart was seen, with a thin-walled membrane across the left atrium dividing it into two compartments, one receiving the pulmonary venous flow and a true left atrium that connects to the left ventricle through mitral valve. JRCDD 2015; 2 (3): 82–84

Key words: cor triatriatum, congenital heart malformation

Literature review

Cor triatriatum (or triatrial heart) is a congenital heart defect where the left atrium (LA) (*cor triatriatum sinistrum*) or right atrium (RA) (*cor triatriatum dextrum*) is subdivided by a thin membrane, resulting in three atrial chambers. Cor triatriatum is a rare congenital anomaly with a ratio of men to women of 1.5:1 [1]

Cor triatriatum represents 0.1–0.4% of all congenital cardiac malformations and usually refers to the LA [2–3]. In cor triatriatum sinister the LA is divided by a fibromuscular membrane into two distinct chambers: a posterior – superior chamber receiving the four pulmonary veins and an anterior – inferior chamber (true LA) which communicates with left ventricle (LV) through mitral valve [4]. In the majority of cases it is diagnosed in neonatal period or early infancy, whereas adult cases are very rare [5].

In the pediatric population, this anomaly may be associated with major congenital cardiac lesions such as tetralogy of Fallot, double outlet right ventricle, coarctation of the aorta, partial anomalous pulmonary venous connection, persistent left superior vena cava with unroofed coronary sinus, ventricular septal defect, atrioventricular septal (endocardial cushion) defect, and common atrioventricular canal. [6] In the adult, cor triatriatum is frequently an isolated finding.

Several classification schemes have been reported to describe cor triatriatum, the simplest was given by Loeffler in 1949 [7].

It is based on the number and size of fenestrations in the fibro-muscular membrane and it distinguishes three groups: group one is defined by the absence of connection between the two chambers, the accessory chamber might connect with the RA or some of the pulmonary veins might drain in anomalous fashion. In group two there are one or few small openings in the intra-atrial membrane. When the foramen is small, the obstruction is sufficient to create a pressure gradient within the atria, thus mimicking mitral stenosis. In group three, the accessory chamber communicates widely with the true atrium by a large single opening.

Case Presentation

55-years old woman admitted to hospital because of the congenital heart disease, particularly atrial septal defect (ASD). Since childhood she presented with signs and symptoms of a heart failure, which have deteriorated to New York Heart Association (NYHA) class III during last 2 years. Previously she was treated for hypertension and asthma. On physical examination central and peripheral cyanosis, clubbed fingers, and a systolic heart murmur in 4th intercostal septum were found.

Full blood count showed increased hematocrit (58%), with increased number of erythrocytes and hemoglobin level. N-terminal of the prohormone brain natriuretic peptide (NT-pro BNP) was

Conflict of interest: none declared.

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Figure 1. Electrocardiogram. Sinus rhythm, incomplete right bundle branch block, signs of right and left atria, and right ventricle overload



Figure 2. Transthoracic echocardiography. Dilated right heart. 26 mm defect in the interatrial septum (arrow)

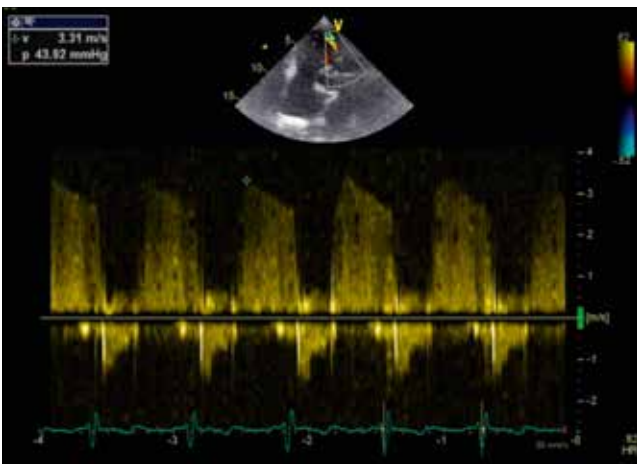


Figure 3. Transthoracic echocardiography. Continuous wave Doppler spectrum across the pulmonary valve. Calculated mean pulmonary artery pressure – 59 mm Hg



Figure 4. Transesophageal echocardiography. Left atrium subdivided by a thin membrane (arrow)



Figure 5. Computed tomography. Thin-walled membrane across the left atrium dividing it into two compartments

normal; in arterial blood gas oxygen saturation was decreased to 79,2%, with features of partial respiratory failure.

On electrocardiogram (ECG) normal sinus rhythm, incomplete right bundle branch block were found, with signs of both atria and right ventricle (RV) overload, and RV hypertrophy (Figure 1). In 6 minutes walk test she made 295 meters with blood oxygen desaturation from 76% to 66%. A transthoracic and transesophageal echocardiography (Figures 2–4) revealed dilated right heart, moderate tricuspid regurgitation and signs of pulmonary hypertension with right ventricular systolic pressure (RVSP) of 85 mm Hg. In the middle of an intraatrial septum two defects were found, one with right-to-left shunt, and another with left-to-right shunt. In transesophageal interatrial membrane was seen, what has given suspicion of cor triatriatum. Normal systolic function of LV was documented, with ejection fraction (EF) estimated >60%.

During cardiac catheterization mean pressure in right pulmonary artery (PA) was 53 mm Hg, mean pressure in RA was 4 mm Hg, and

10 mm Hg in LA. After Iloprost administration PA mean pressure decreased to 38 mm Hg.

Saturations in heart cavities suggested presence of intracardiac shunts (RA 83%, RV 86%, PA 87%, aorta 78%)

Coronary angiography showed normal coronary arteries.

In computed tomography (Figure 5) threeatrial heart was seen, with a thin-walled membrane across the LA dividing it into two compartments, one receiving the pulmonary venous flow and a true LA that connects to the LV through mitral valve. In addition, between RA and inflowing part of a LA, defect in intraatrial septum was seen, and there was no septum between RA and outflowing part of a LA. Confluens of coronary veins and pulmonary veins in the inflowing part of the LA was seen with bilateral common insertion of superior and inferior pulmonary veins.

These findings were confirmed in cardiac magnetic resonance. In addition confluens of the right pulmonary vein was seen in right part of LA (because of presence an intraatrial septum defect- blood flow directs to both atria).

Patient management and follow up

With these findings the patient was scheduled to the bosentan therapy in a dose 62,5 mg twice daily. After 4 months of ambulatory treatment patient was readmitted to cardiology ward for detailed follow-up examination and assessment of therapy results.

Patient made significant improvement and good tolerance of therapy. In 6 minutest walk test she made 400 m without shortness of breath. In transthoracic echocardiograph reduction of RV dimensions was seen, with decreased RVSP to 73 mm Hg. With these findings bosentan monotherapy was continued with a dose titrated to 125 mg BD.

It was decided not to undertake surgical intervention at the severe pulmonary hypertension period. The only therapeutic option is a lung transplantation with surgical correction this heart defect that however is associated with very high operative risk.

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