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Brief Report

Transposition of the great arteries and cor triatriatum: a rare combination

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Abstract In this case report, we present a 5-month-old girl diagnosed with a unique combination of transposition of the great arteries and cor triatriatum sinistra. A 1-day-old female patient presented to our hospital with cyanosis since the early neonatal period. We confirmed transposition of the great arteries by echocardiography. The patient underwent arterial switch operation on day 8 and was discharged on day 35. After 5 months of the operation, the patient had a lower respiratory tract infection and was unable to gain weight. Echocardiography revealed mild neopulmonary regurgitation, minimal neoaortic regurgitation, and pulmonary arterial hypertension. In addition, a fibrous membrane was also seen dividing the left atrium. The patient was diagnosed with cor triatriatum and underwent successful resection of the membrane.

Keywords: Transposition of the great arteries; echocardiography; congenital heart disease

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Cor triatriatum is a rare cardiac malformation in which the left atrium is divided into two portions. The associated cardiac lesions are common and determine the clinical presentation. Its coexistence is important to identify because it can alter the haemodynamics and surgical outcome. We report the case of a 5-month-old girl who was diagnosed with transposition of the great arteries and cor triatriatum. Initially, the patient underwent arterial switch operation and thereafter resection of the fibrous membrane.

Case report

A 1-day-old female patient presented to our hospital with cyanosis since the early neonatal period. We confirmed transposition of the great arteries, atrial septal defect, severe tricuspid regurgitation, and persistent left superior vena cava by echocardiography. The patient underwent arterial switch operation on day 8 and was discharged on day 35. After 5 months of the arterial switch operation, the patient had a lower respiratory tract infection and was unable to gain weight (3 percentile). A grade III/VI systolic murmur was audible and the liver was palpable 1 cm below the right costal margin. The breath sounds were coarse on auscultation. Echocardiography revealed atrioventricular and ventriculoarterial concordance, mild neopulmonary regurgitation, minimal neoaortic regurgitation, and pulmonary arterial hypertansion. The left superior vena cava drained to the right atrium through an enlarged coronary sinus. In addition, a fibrous membrane was also seen dividing the left atrium into a proximal chamber that connected pulmonary veins and a distal chamber communicating with the mitral valve (Fig I). Pulse Doppler examination confirmed obstructed flow across the fibrous membrane as evidenced by continuous flow with diastolic velocity 2.5 m/sn. At catheterisation, we obtained the pressures in right pulmonary artery: systolic 52, diastolic 10, mean 28; in left pulmonary artery: systolic 43, diastolic 19, mean 27; in main pulmonary artery: systolic 57, diastolic 15, mean 29; in right ventricle: 63/0—12; in right atrium: mean 5; in coronary: sinüs 8; and in pulmonary capillary wedge: mean 23 mmHg. The patient was diagnosed

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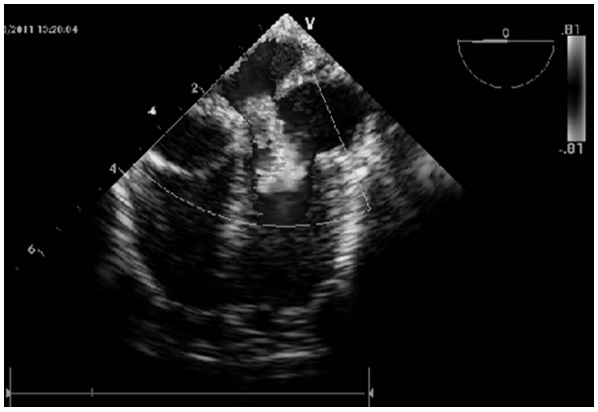

Figure. I.
Transoesophageal echocardiogram from the apical four-chamber view with colour Doppler.

with cor triatriatum and underwent successful resection of the membrane and achieved excellent haemodynamic parameters.

Discussion

Cor triatriatum is a rare congenital cardiac defect that can present with a wide range of symptoms and may be associated with other structural cardiac defects.[1] The associated cardiac lesions are common, present in 75% of cases, and determine the clinical presentation and outcome. The common associated lesions are atrial septal defect, partial or total anomalous pulmonary venous connection, patent ductus arteriosus, ventricular septal defect, atrioventricular septal defect, and coarctation of the aorta. There are only four cases that have been reported in the literature in association with corrected transposition of the great arteries.[2,3]

Transposition of the great arteries is considered to be the result of abnormal rotation during cardiac morphogenesis, whereas cor triatriatum is due to abnormal incorporation of the pulmonary veins into the left atrium. Similar to the other previously reported associated lesions, it is difficult to hypothesise an embryological basis for this rare coexistence.[4]

Pre-operative echocardiography is the only method to diagnose the coexistence of transposition of the great arteries and cor triatriatum. Moreover, with no prior information, the surgeon cannot waste precious time entering the left atrium to search for an undeclared rare entity. If not corrected during arterial switch operation membrane in the left atrium can lead to persistent obstruction to pulmonary venous inflow and increase in pulmonary arterial pressure, and can affect the clinical presentation despite a successful surgery. After 5 months of the arterial switch operation, our case had a lower respiratory tract infection and was unable to gain weight.

This case report aims to document the rare coexistence of transposition of the great arteries and cor triatriatum and to emphasise the need for careful echocardiographic examination in patients with congenital cardiac defect. Should associated lesion be overlooked, the outcome may be changed.

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Conflicts of Interest

None

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