CASE REPORT

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UNRESOLVED TACHYPNOEA: A PRESENTATION OF CONGENITAL COR-TRIATRIATUM

Fahisham Taib, Nur Atiqah Abdul Rahman, Mohd Rizal Mohd Zain

Abstract

Cor-triatrium is a rare cardiac anomaly. In literature, majority case reports on the condition focused on its late presentation in adulthood. It can be easily corrected by surgical intervention to avoid pulmonary congestion and subsequent pulmonary hypertension. We report a rare case of cor-triatrium with severe pulmonary hypertension in a 7-week-old baby who presented with persistent tachypnoea.

Keywords:

Cor-triatrium; Cardiac failure; Tachypnoea

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Introduction

Cor-triatrium is a rare cardiac malformation and may present with variation in clinical presentation. Its incidence is estimated at about 0.1% of all congenital heart disease. In this anomaly, the atrium is divided by a fibromuscular membrane into two distinct chambers: a posterior- superior chamber which receives the four pulmonary veins and an anterior-inferior chamber (true left atrium) which connected to the ventricle via atrioventricular valves. Normally, there is a small opening connecting these two chambers. There are two types: cor-triatrium sinister and cor-triatrium dexter for left and right atrium respectively and the later was extremely rare. We reported a case of cor-triatrium sinister in a 7-week-old baby who presented with respiratory symptoms.

Case report

Baby X was born normally at full term with the birth weight of 3.3 kg. He presented with a short (1 week) history of tachypnoea and interrupted feeding. He was commenced on the intravenous antibiotic for suspected lower respiratory tract infection despite negative infective markers. His respiratory rate was 70 breaths/minute at the initial presentation.

Following completion of antibiotic treatment, baby X maintained his respiratory rate at 65 breaths/minute with saturation of 97% in FiO₂ 0.35. There was no adventitious breath sound on auscultation, apex beat was palpable at the fourth intercostal space midclavicular line with the presence of loud second heart sound at the pulmonary area. His liver edge was palpable 1 cm below the right costal margin. His chest radiograph showed normal cardiac contour with the cardio-thoracic ratio of 0.51. There was haziness of the left upper zone with prominence pulmonary vascular markings. Echocardiography revealed situs solitus, levocardia with dilated right atrium (RA) and right ventricle (RV), presence of patent foramen ovale (PFO) size measured at 5.9 mm with bidirectional flow which connected between the upper chamber of the left atrium and right atrium, moderate to severe tricuspid regurgitation [TR] with pulmonary gradient (PG) of 80 mmHg with velocity of 4.7m/sec, dilated main pulmonary artery at 13 mm, aortic valve annulus of 9.2 mm and pulmonary venous (PV) annulus of 11.8 mm. Fibromuscular septum dividing left atrium (LA) into the upper and lower chamber connected through a small orifice measuring 4.8 mm was seen.

Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia

Corresponding author: Mohd Rizal Mohd Zain, Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia

Telephone no.: +6097676529; **Email:** drrizal@usm.my

He was subsequently transferred to the cardiac centre for surgical correction. The large PDA was found intraoperatively, which was ligated successfully. There was a stormy post-operatively period where he required a delay chest closure, complicated with pulmonary hypertension crisis requiring inotropic support and inhaled nitric oxide. He was discharged home on day 10 post-operatively. At 6 weeks follow up, no further evidence of pulmonary hypertension was noted on the echocardiogram. He was clinically well and gaining weight.

Discussion

Cor-triatritum is a rare cardiac anomaly with a prevalence of <0.1% of all congenital heart disease and has a ratio of male to female of 1.5:1 [1]. It can be corrected easily by surgical intervention. Clinical and radiological signs are typical of those with pulmonary arterial and venous hypertension [2]. In cor-triatritum sinister, the left atrium is divided by a fibromuscular membrane into two distinct chambers: a posterior - superior chamber receiving the four pulmonary veins and an anterior -inferior chamber that connects to the left ventricle by means of the mitral valve. Other associated anomalies are the unrooted coronary sinus, ventricular septal defect, coarctation of the aorta, atrioventricular septal defect, tetralogy of Fallot and rarely asplenia and polysplenia. Failure of incorporation of the common pulmonary vein into the left atrium lead to symptoms associated

to pulmonary venous obstruction and pressure loading of the right side of the heart [3]. Faltering growth, recurrent pneumonia, feeding difficulties, tachypnoea, heart failure, cyanosis and arrhythmias are some of the reported presenting features.

There are various embryologic hypotheses to explain the development of cor-triatritum – the entrapment and impingement theories [4]. Entrapment hypothesis postulated that the left horn of the sinus venosus entraps the common pulmonary vein and thereby prevents its incorporation into the left atrium. In the impingement theory, the persistent left superior vena cava on the left atrium induces the production of an abnormal left atrial membrane [5]. Often cor-triatritum presents late in adulthood due to fibrosis and calcification of the orifice in the anomalous septum, the development of mitral regurgitation, or atrial fibrillation.

Diagnostic accuracy should be achieved using transthoracic or transoesophageal echocardiography and cardiac catheterization. Correcting cor-triatritum should include finding its associated anomalies and delineating the complex congenital heart condition [6].

In our case, the patient has the anomaly in his left atrium thus is called cor-triatritum sinister (Figure 1) with type IIA1 based on Loeffler classification [8] (Table 1).

RA – Right atrium
RV – Right ventricle

LA – Left atrium
LV – Left ventricle

Figure 1. Apical chamber view of the echocardiography. The arrow pointed to a thin fibromuscular septum separating the left atrium

Table 1: Classification of cor-triatriatum

I	A. Patent foramen ovale in lower left atrial chamber
	1. With connection between right atrium and the upper left atrial chamber
II	Cor-triatriatum with small opening in anomalous intra atrial septum
	A. Patent foramen ovale
	1. In the upper left atrial chamber
	2. In the lower left atrial chamber
	3. Incomplete description of the position
	B. Closed foramen ovale
	1. In the upper left atrial chamber
	2. Between the upper and lower left atrial chamber
	3. In the lower left atrial chamber
	4. Incomplete description of the position
	C. Surgically operated cases
III	A. Patent foramen ovale in the lower left atrial chamber
	B. Closed foramen ovale in the lower left atrial chamber
	C. Closed foramen ovale: incomplete description of the position

His tachypnoea persisted without improvement after standard antimicrobial therapy. There was no evidence of heart failure on the chest

radiograph apart from plethoric lung field and normal cardiothoracic ratio (Figure 2).

Figure 2: AP view of the chest radiograph showing plethoric lung with cardiothoracic ratio of 51%

Besides pulmonary venous congestion features in cor-triatritum, the large PDA was the factor that contributed to the development of the severe pulmonary hypertension. The PDA finding was missed on the initial echocardiography due to reversal shunt of the flow which sometimes was difficult to appreciate on echocardiography. The learning point here is - if the patient has unresolved tachypnoea, cardiac aetiology should be considered. Our patient had a successful surgical correction of cor-triatritum, ligation of patent ductus arteriosus and closure of the patent foramen ovale at the tertiary cardiac centre.

Conclusion

Cor-triatritum should be considered as differential diagnosis in a baby presented with unresolved tachypnoea in general practice setting, following the exclusion of respiratory aetiology. Despite its rare condition, early identification and diagnosis would result in a good outcome for the patient.

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