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Case Report

Isolated Leftward Displacement of the Septum Primum (Septum Primum Malposition Defect): A Rare Congenital Anomaly

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Abstract

Septum primum malposition defect is a very rare congenital anomaly that usually associated with visceral heterotaxy. It results in partially or totally anomalous pulmonary venous drainage with a normal venous connection to the left atrium, depending on the degree of septal malposition. In this article we present a 35 days old female of an echocardiographic diagnosis with situs solitus and isolated leftward displacement of the septum primum.

Keywords: *Septum Primum Malposition; Atrial Septal Defect; Leftwards Septum Primum Displacement; Total Anomalous Pulmonary Drainage*

Introduction

Septum primum malposition defect is a rare congenital anomaly. It occurs when the cephalad crescent border of the septum primum remains unattached , because the superior limbic band of the septum secundum fails to develop . Clinically , it presents as a partial or a complete anomaly of the pulmonary veins drainage , despite their normal connecting to the left atrium . This anomaly is usually associated with other malformations , and with other syndroms especially visceral heterotaxy.

Case Report

A 35 days old girl infant, weight 2.2 kg, who presented with dyspnea, nursing difficulty, and cyanosis. These complaints have been noticed few days after birth with an exaggerating over time.

History: Vaginal birth, Full term, weight at birth was unknown. No dyspnea or cyanosis soon after birth according to parents.

On physical examination: Dyspnea RR: 55/min, intercostal retraction, pallor, failure to thrive. On auscultation: fixed splitting of the second heart sound along with an ejection systolic murmur grade 3/6 at the pulmonary area.

CXR: Cardiomegaly with bilateral pulmonary infiltrations.

Echocardiographic findings: Situs solitus, levocardia. An extreme left displacement of atrial septum with severe dilated right atrium and very small left atrium. The pulmonary veins were connected normally to the left atrium, but with a drainage to the right atrium across ASD. Severe dilated right ventricle with severe pulmonary hypertension, small VSD with right to left shunt across it.

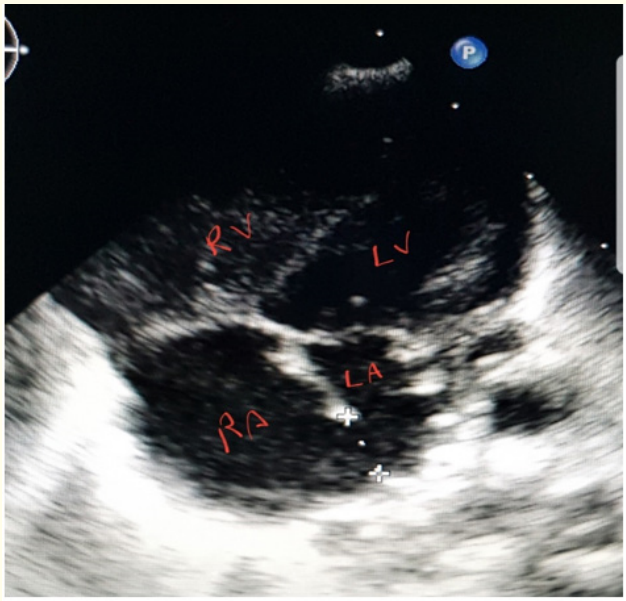

Figure 1: *Bidimensional transthoracic echocardiography from Subcostal 4- chamber view, left shifted septum premium.*

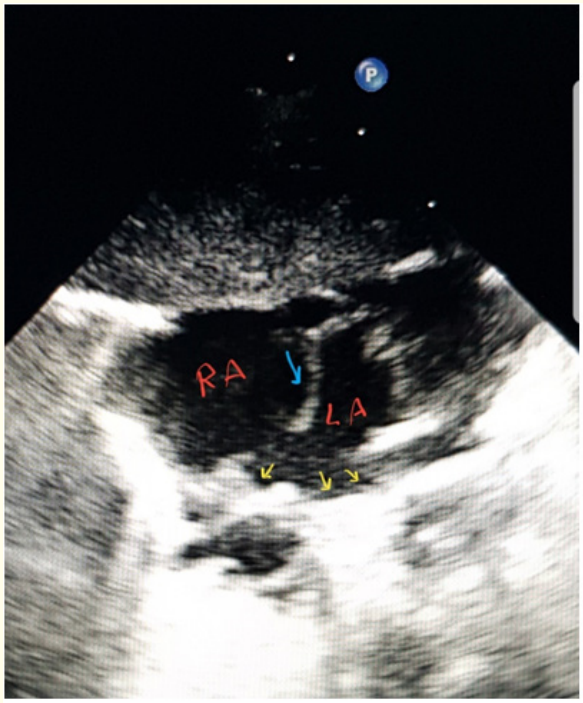

Figure 2: *Bidimensional transthoracic echocardiography from subcostal view. LA: left atrium, RA: right atrium, Blue arrow: left shifted septal primum, Yellow arrow: Pulmonary veins connected to LA.*

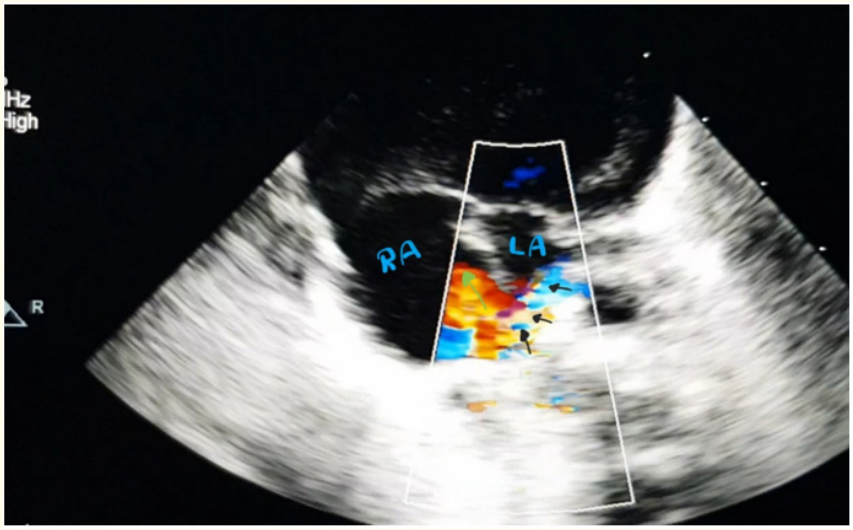

Figure 3: *Color transthoracic echocardiography from subcostal view, LA: small left atrium, RA: wide right atrium, Green arrow: Blood flow across ASD, black arrows: Blood flow across pulmonary veins. It shows the direct flow from the normally connected pulmonary veins to the RA, because of the leftward malposition of the ostium primum.*

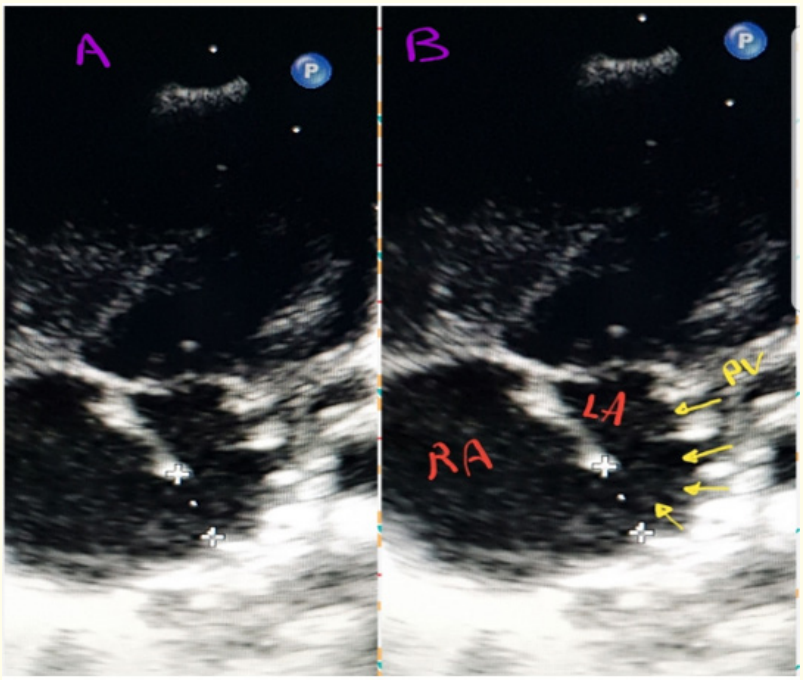

Figure 4: *A and B Bidimensional transthoracic echocardiographic images from subcostal view demonstrate the superposterior ASD++ between the left shifted septum primum and the posterior atrial wall. LA: left atrium, RA: right atrium, Yellow Arrows: Pulmonary veins.*

The patient was accepted in NICU because of a very severe pneumonia proved by chest CT scan. She had been putting on mechanical ventilator and had received treatment with wide broad antibiotics and medications to decrease the pulmonary hypertension.

Discussion

The septum primum malposition (SPM) defect is a very rare congenital anomaly [1,2]. Displacement of the septum primum-leftward in atrial situs solitus or rightward in atrial situs inversus is known as the SPM defect, and it appears to be responsible for anomalous pulmonary venous drainage. This abnormality occurs predominantly in patients with visceral heterotaxy and is usually accompanied by polysplenia, although it can be rarely seen with asplenia or a normally formed spleen as well [3]. Very few cases of SPM defect have been reported in the literature. The most recent ones are: the page uploaded by Trushar Gajjar on 29 January 2016, a case report of septum primum malposition defect in 18 years old male [3]. And the article published as: Cuttone F, Hadeed K, Lacour-Gayet F, Lucron H, Hascoet S, Acar P., *et al.* Isolated severe leftward displacement of the septum primum: anatomic 3D echocardiographic findings and surgical repair. *Interactive CardioVascular and Thoracic Surgery*. In this article, 3 cases of isolated severe leftward displacement of the septum primum have been discussed [4].

This anomaly can be diagnosed by transthoracic echocardiography, and the deviation of the septum primum is best demonstrated with the subxiphoid coronal, apical four-chamber, and parasternal long-axis views [5].

Embryologic review

Normally, the septum primum grows from sinus venous tissue adjacent to the inferior vena cava (IVC)-RA junction and parallel to the left venous valve. When it completes its normal growth, it is attached on the LA side of the superior limbic band. It is the valve of the foramen ovale for fetal circulation and it forms part of the interatrial septum for the postnatal heart [3].

The normal growth and attachment of the septum primum on the septum secundum are essential for the alignment of the common pulmonary vein with the cavity of the LA. If the superior limbic band of the septum secundum fails to develop, the cephalad crescent border of the septum primum remains unattached and could be carried leftward in cases of atrial situs solitus or rightward in cases of atrial situs inversus by the bloodstream of the fetal circulation which proceeds from the RA towards the LA.

Depending on the degree of septum primum displacement toward the LA, the scenario varies from partially to totally anomalous pulmonary venous drainage (TAPVD), presenting externally with a normal connection of the pulmonary veins with the morphological LA [4].

Conclusion

This article reports a case of severe leftward displacement of the septum primum, presented as an isolated cardiac malformation in a 35 days old girl infant. This anomaly is very rare, especially if it occurs in atrial situs solitus with no other cardiac defects. Transthoracic echocardiography with Doppler interrogation provides a reliable method for the diagnosis of this malformation.

Bibliography

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