IMAGES in PAEDIATRIC CARDIOLOGY

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Abstract
Cor triatriatum sinister is a rare congenital cardiac malformation characterized by a membrane in the left atrium which separates the left atrium into the proximal and distal chambers.Association of cor triatriatum is extremely rare with situs inversus totalis. Here we report a rare case of cor triatriatum sinister with situs inversus totalis in a 5 month old female infant.

MeSH:
Cor triatriatum, Situs inversus, Patent ductus arteriosus, Infant

Introduction
Cor triatriatum is a rare congenital cardiac malformation with an estimated incidence of 0.1% of all congenital heart disease and it usually refers to the left atrium (cor triatriatum sinister). In cor triatriatum sinister (CTS) 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 (true left atrium) that connects to the left ventricle by means of the mitral valve.[1] In CTS, the obstructive nature of the membrane leads to creation of a pressure gradient, with an associated rise in pulmonary venous pressures.[2] Total situs inversus (TSI) is characterized by a heart on the right side of the midline while the liver and the gall bladder are on the left side.Here we report an extremely rare case of cor triatriatum sinister which is associated with TSI.

Case Report
A five month old female child was referred for cardiac murmur evaluation. History of suck rest suck cycle was present. No family history of congenital heart disease was noted. Physical examination revealed a pulse rate of 145 beats/minute, blood pressure of 94/50 mmHg, with baseline oxygen saturation of 99%. On cardiac examination, the second heart sound was normal and there was no evidence of pulmonary hypertension. Auscultation of the right chest revealed a 3/6 systolic murmur at the right parasternal border. Other systems were normal.Laboratory investigation showed a hemoglobin level of 10 g% and a white cell count of 16,000 mm^3. The chest X-ray demonstrated dextrocardia, cardiomegaly and normal pulmonary vascular markings. A gastric air bubble was noted on the right side, and a liver shadow on the left side was present. The electrocardiogram(ECG) revealed a sinus rhythm with dextrocardia. Abdominal ultrasound demonstrated the stomach and spleen on the right side and the liver on the left. The aorta and the inferior vena cava were inverted. Transthoracic echocardiography (TTE) demonstrated {I,L,I} (I, situs visceroatrialis inversus; L, L-loop of the ventricles; I, normally related inverted great arteries).

It also showed a fibromuscular membrane across the left atrium, dividing it into two compartments suggestive of cor triatriatum sinister (fig1. & fig2). The two chambers communicated through two restrictive orifices (gradient:16mmHg) (fig3). A small size patent ductus arteriosus (gradient 20/6mmHg) with left to right shunt was present (fig4.). Other cardiac anomalies like patent foramen

ovale (PFO), atrial septal defect (ASD) and anomalous venous connections were ruled out and the diagnosis of cor triatriatum sinister associated with TSI was reconfirmed.

Figure 1. Subcostal view of transthoracic echocardiography shows fibromuscular membrane in left atrium.

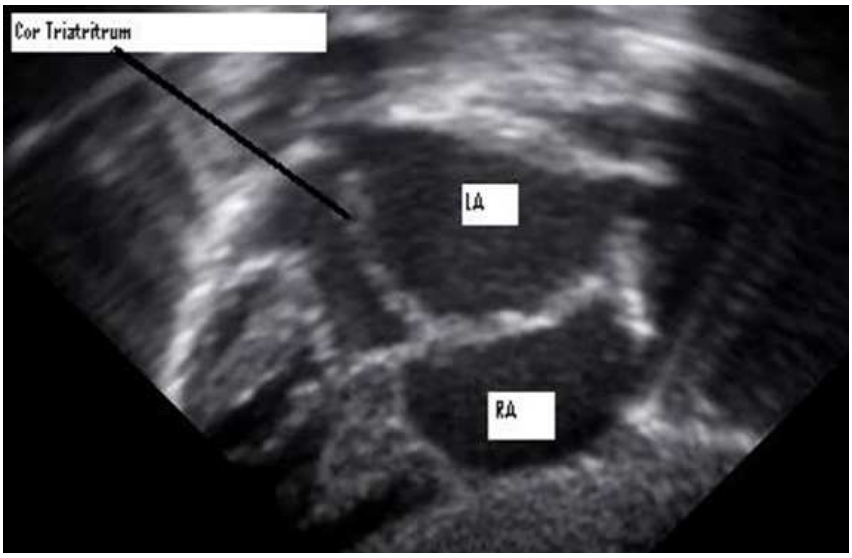


Figure 2. Long axis of parastenal view shows the fibromuscular membrane in left atrium.

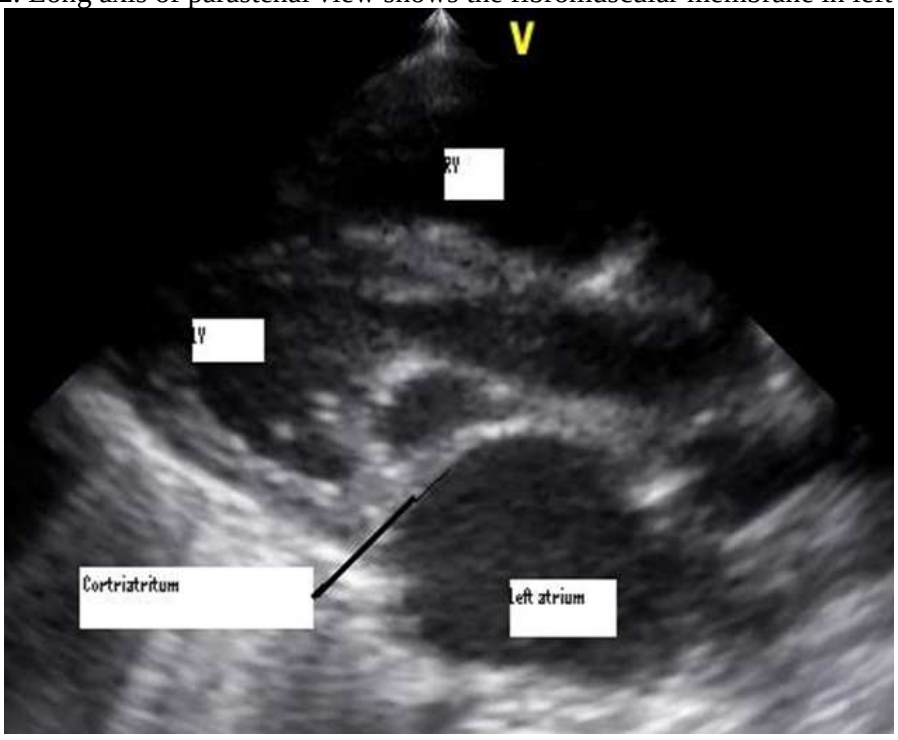


Figure 3. Apical four-chamber view shows the turbulent flow across the orifice of fibromuscular membrane.

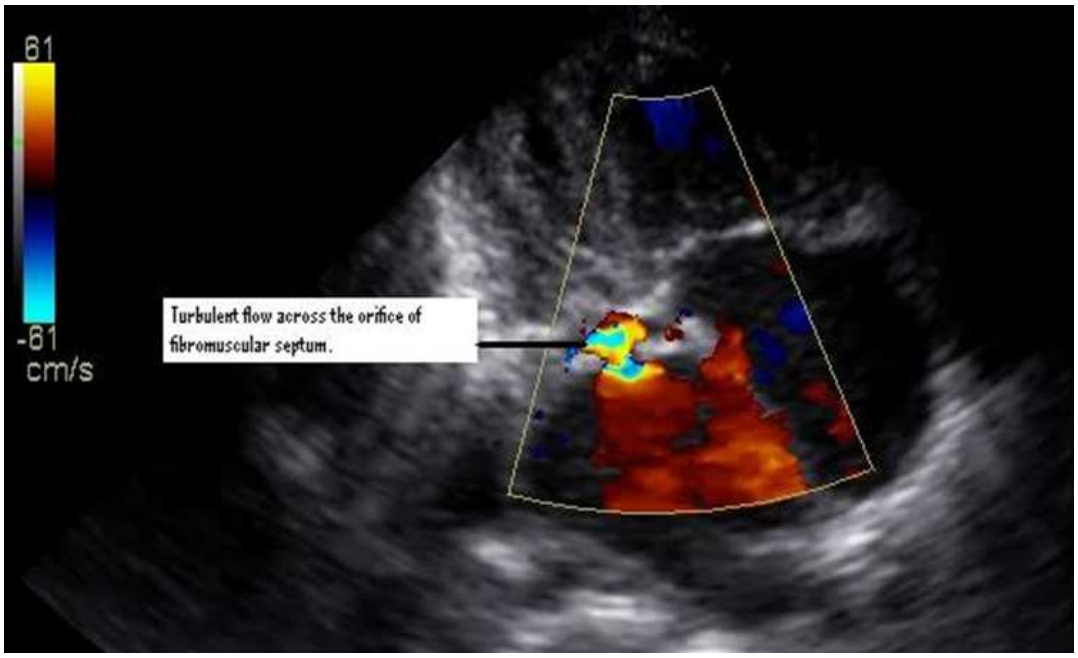
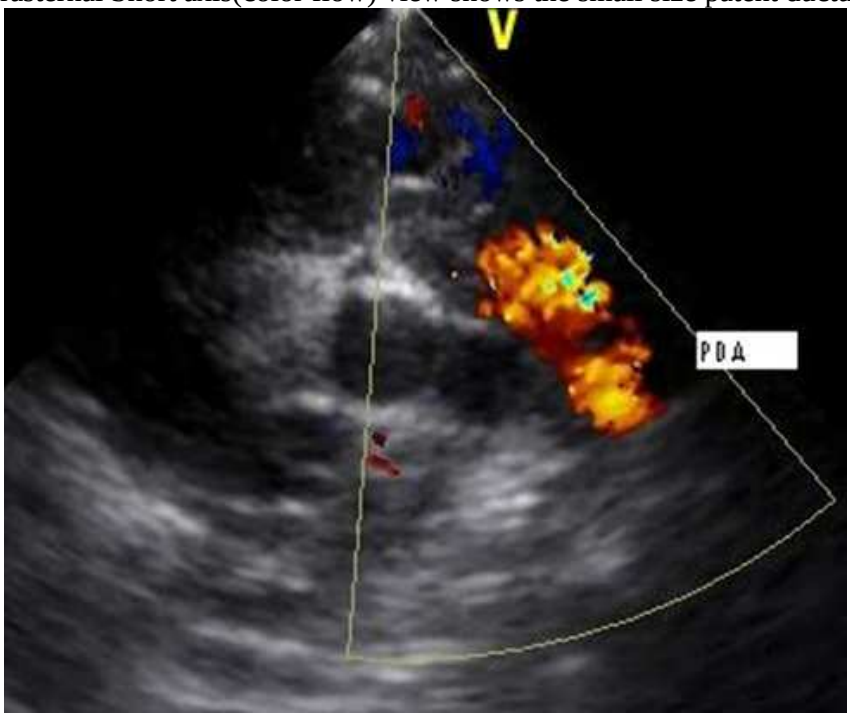


Figure 4. Parasternal Short axis(color flow) view shows the small size patent ductus arteriosus.



Discussion

Cor triatriatum was first described by Church in 1868, and the membrane of cor triatriatum appears as a linear echo bisecting the left atrium (CTS). The embryologic etiology of CTS remains debated, it may result from incomplete incorporation of the common pulmonary vein into the left atrium, abnormal overgrowth of septum primum, entrapment of the common pulmonary vein by the left horn of the sinus venosus preventing its incorporation into the left atrium,[1] or persistence of a left-sided superior vena cava which may impinge on the left atrium, resulting in the formation of an abnormal membrane.[2,3]

CTS was classified in 1949 by Loeffler,[4] according to the number and size of the orifices in the fibromuscular septum: Group 1 is defined as having no opening, Group 2 as having one or more small openings and Group 3 as having a single, large opening. Our case has two small restricted openings present in the fibromuscular membrane (CTS) which falls into group 2 classification. Cor triatriatum 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 defect, common atrioventricular canal and rarely asplenia and polysplenia. No genetic predisposition has been linked to this particular anomaly. Total situs inversus (TSI), termed as mirror image dextrocardia, is characterized by a heart on the right side of the midline while the liver and the gall bladder are on the left side. Patients are usually asymptomatic and have a normal lifespan. The association of CTSwith TSI which we report is extremely rare.

The clinical manifestations of CTS depend upon the size of the opening in the fibromuscular membrane and the presence of associated congenital cardiac defects.The presentation of CTS can mimic those of mitral stenosis, supravalvular mitral ring, left atrial thrombus or pulmonary venous stenosis. The common presenting symptoms are dyspnea, hemoptysis and orthopnea as a result of the obstructive function of the intra-atrial membrane.[5] Often, CTS is misdiagnosed as mitral valve disease, but can be erroneously diagnosed as primary pulmonary hypertension.[6] Several techniques have been used for diagnosis establishment such as transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), CT (computerized tomography) scan and magnetic resonance imaging (MRI).

Echocardiography is the most commonly used imaging technique for the diagnosis of cor triatriatum. The typical CTSappears as a membrane attached laterally to the junction of the left upper pulmonic vein and left atrial appendage, dividing the left atrium into 2 chambers. The proximal chamber receives blood from the pulmonary veins and the distal chamber contains the left atrial appendage and mitral valve. One or more fenestrations of varying sizes connect the 2 chambers. Similar findings were present in our case. Three-dimensional reconstruction of echocardiographic images has been used to better define the membrane and its relationship to surrounding structures. Both computerized tomography of the heart and cardiac magnetic resonance imaging (MRI) have been used for evaluating patients with suspected cor triatriatum. The use of CT scan however includes the hazards of radiation, while TEE includes the discomfort of scope intubation. TTE is the best method for evaluating the CTS.[7-9] Surgical resection of the accessory fibromuscular membrane has been successful in symptomatic patients with cor triatriatum even though successful balloon catheter dilatation of the communication between the two chambers has been described.[10] To the best of our knowledge, this is only the second report in the literature of a patient with total situs inversus presenting with CTS.[11]

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