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A case summary of Cor Triatriatum Sinister with unusual presence of OS-ASD (between true LA & RA)

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Cor Triatriatum Sinister is a rare congenital abnormality, usually diagnosed in childhood; few cases remain asymptomatic and are diagnosed in adulthood.

Triatrial heart is a rare congenital abnormality, reported by Jeiger at the autopsy, in 0.4% of patients with congenital heart disease, and found in less than 0.1% of clinically diagnosed cardiopathies. Though it was first described by Church in 1868, as a left atrium divided by an abnormal septum, the name “cor triatriatum” was given by Borst in 1905. It involves usually the left atrium (cor triatriatum sinister) and rarely the right atrium (cor triatriatum dexter), in this review, we will be discussing cor triatriatum sinister (CTS).

No clear difference in the incidence of the disease has been noted among men or women and no related genetic disorders have been described in the literature. The anomaly may be associated with other congenital cardiac lesions in up to 80% of cases in pediatric population, most frequently with ostium secundum atrial septal defect and anomalous pulmonary vein return. Other associated lesions are listed as below-

Congenital cardiac lesions associated with CTS

- Atrial septal defect
- Anomalous pulmonary venous return
- Tetralogy of Fallot
- Bicuspid aortic valve
- Double outlet right ventricule
- Coarctation of the aorta
- Persistant left superior vena cava with unroofed coronary sinus
- Ventricular septal defect
- Common atrio-ventricular canal
- Hypo plastic mitral valve

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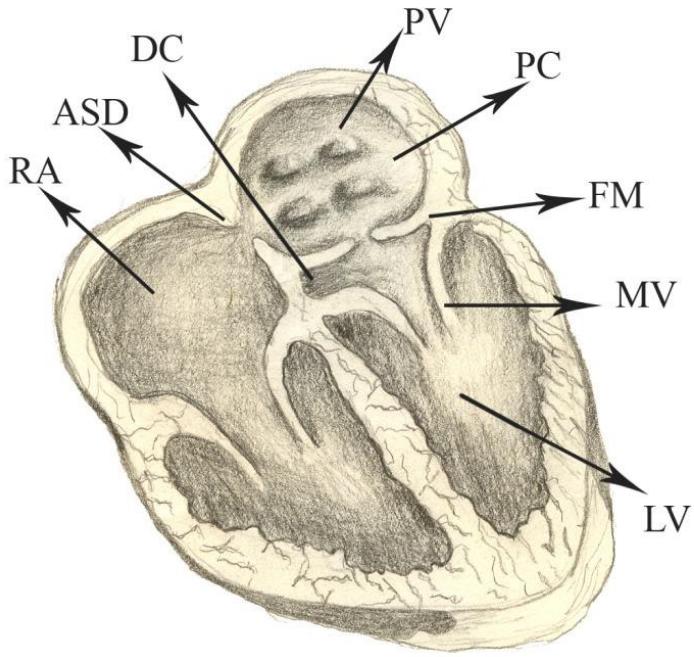
- Bicuspid right atrio-ventricular valve

Anatomical features and etiology

The atrium is divided into two distinct chambers, usually by a thick fibro-muscular septum, which could be membranous with transverse or horizontal orientation, band-like or funnel shaped. The proximal or superior chamber drains the pulmonary venous blood while the distal inferior chamber (or true atrium) is in contact with the atrio-ventricular valve and contains the atrial appendage and the true atrial septum. Several classification schemes have been reported to describe CTS, the simplest was given by Loeffler in 1949.

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 right atrium 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; and in group three, the accessory chamber communicates widely with the true atrium by a large single opening.

While the latter is mostly found in the adult population having this abnormality, the first two groups are usually diagnosed in highly symptomatic infants and children and are associated with increased mortality at a younger age



Clinical presentation

The physiologic consequences of CTS are directly related to the size of the orifice between the accessory and the true atrial chamber. When the foramen is small, the obstruction is sufficient to

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create a pressure gradient within the atria, thus mimicking mitral Stenosis. Relevant symptoms could also be related to associated cardiac abnormalities.

In infants and newborns, the manifestations of the disease are secondary to a relatively narrow opening with a subsequent rise in proximal left atrial pressure and pulmonary congestion. Dyspnoea ranges from mild forms to more severe presentations such as neonatal respiratory distress with an increased mortality risk.

Adults having the disease are usually asymptomatic, due to the presence of a large foramen with no intra-atrial pressure gradient. The appearance of symptoms occurs secondary to fibrosis and calcification of the accessory membrane orifice, though the latter is more often obstructive at a younger age before the degenerative changes take place. Symptoms include exceptional dyspnoea, orthopnoea and haemoptysis. Several cases of pulmonary edema during labour have also been reported in young adult women having initially undiagnosed CTS. The anomaly is sometimes revealed by atrial arrhythmias or cerebral and systemic embolic events, due to thrombus in an enlarged accessory atrial chamber.

On physical examination, cor triatriatum can be detected by auscultation. The murmur is typically diastolic with a loud P2 when pulmonary hypertension is present. The absence of an opening snap or a loud S1 marks the difference between CTS and mitral Stenosis. The intensity of the murmur depends on the velocity of the flow across the accessory membrane foramen. Signs of pulmonary congestion can also be found.

DIAGNOSIS

ECG is most likely normal. Right ventricular hypertrophy, right axis deviation and S1Q3 pattern are found with pulmonary artery hypertension. Atrial arrhythmia, such as atrial tachycardia or atrial fibrillation might be present.

Cardiac catheterization used to be the reference diagnosis technique before the echography era, it is seldom used nowadays. Typically, it shows a normal left ventricular hemodynamic profile with a trans-atrial gradient, suggested by a simultaneous measurement of an elevated pulmonary artery wedge pressure and a normal left ventricular end-diastolic pressure.

When performed, cardiac angiography may establish the diagnosis of CTS, showing a differential filling of the two distinct atrial chambers. Echocardiography evaluation of CTS is a non-invasive tool that can accurately delineate the morphology of the extra-membrane and the associated cardiac lesions. It is usually seen as a thin linear structure bisecting the atria on a four-chamber view, distally to the mitral valve and the left auricle. In the long-axis view, the superior part of the membrane is usually parallel to the aortic wall while its inferior part connects to the left atrial posterior wall. A single or more foramina can sometimes be seen within the membrane, showing an increased flow velocity on colour Doppler.

The measurements of the mean and maximal trans-membrane pressure gradients and maximal velocity are evaluated by spectral Doppler, in addition to the measurement of the pulmonary artery pressure. It has been reported that severe obstruction is indicated by maximum Doppler velocity greater than 2 m/s. 3-Dimensional Trans-Thoracic Echocardiography (TTE) may permit

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the measurement of the size of the orifice. When limitations in image quality of TTE prevent good morphological evaluation, transoesophageal echocardiography (TEE) may play a crucial role in confirming the diagnosis.

The 3D fully-sampled matrix array TEE (3D-MTEE) transducer was recently developed to allow real-time acquisition and online display of 3D images. In a study of 211 adult patients, Sugseng et al. reported an excellent visualisation by 3D-MTEE of the posterior cardiovascular structures such as the mitral valve (in up to 91 % of cases), the inter-atrial septum (84%) and the left atrial appendage (86%). 3D-MTEE was also successfully used in the diagnosis of CTS, being able to accurately visualise the extra-membrane and showing its exact morphology.

The area of the foramen can also be accurately measured and the relations of the membrane to other cardiac structures can be assessed especially to differentiate cor triatriatum sinistrum from supra-valvular ring, the latter being located below the left atrial appendage. In addition, the presence of associated cardiac lesions such as atrial septal defect, anomalous venous return and less frequently bicuspid aortic valve and dilated sinus venosus could be diagnosed offering a comprehensive anatomic and functional evaluation of these infrequent entities, that guides further surgical treatment options. illustrate a case of CTS, successfully diagnosed with echography.

Computed tomography scanning and Magnetic Resonance Imaging (MRI) can also effectively establish the diagnosis, easily showing the atrial accessory membrane, especially when three-dimensional volume rendering techniques with multi-slice gated computed tomography is performed. In addition, Cine-MRI clearly depicts the fenestration within the membrane with the associated flow turbulence, seen as a low intensity signal contrasted with the high-signal intensity of normal blood flow

Case – A 36 yrs old male referred to us for further evaluation as he had complains of sudden onset of breathlessness with palpitations for 07 days for which he was conservatively managed at other facility centre where his ECG showed atrial flutter and unstable hemodynamics.

Transthoracic Echocardiography

Transthoracic echocardiography revealed a left atrial membrane, with flow convergence at the membrane on color Doppler. A secundum ASD with continuous left-to-right shunting was also noted.. Transthoracic echocardiography also revealed pulmonary hypertension with severe dilation of RV and RA and moderate systolic dysfunction. It was originally unclear whether the left atrial membrane represented a supravalvular mitral membrane (SVMM) associated with Shone complex or a cor triatriatum sinister–type pathology. However, on further review, the mitral valve morphology was noted to be normal with two appropriately positioned papillary muscles, there was no evidence of a parachute mitral valve, and no sub- or supra-aortic stenosis was identified.

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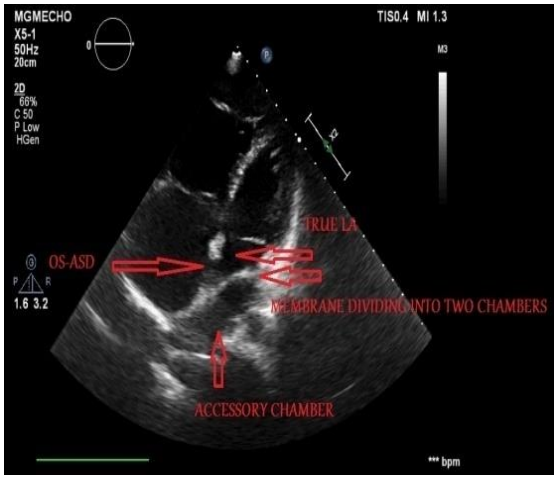
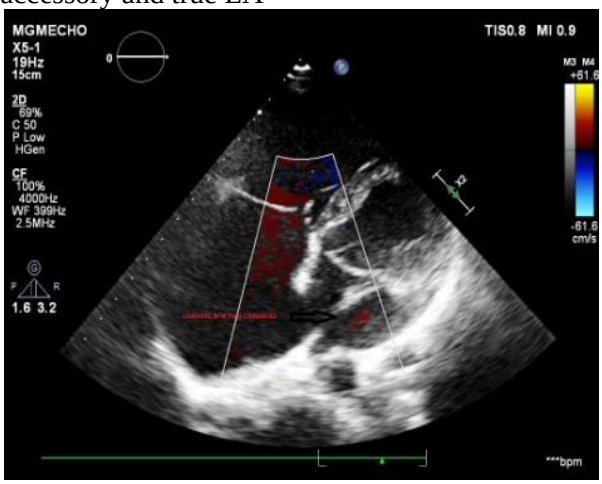
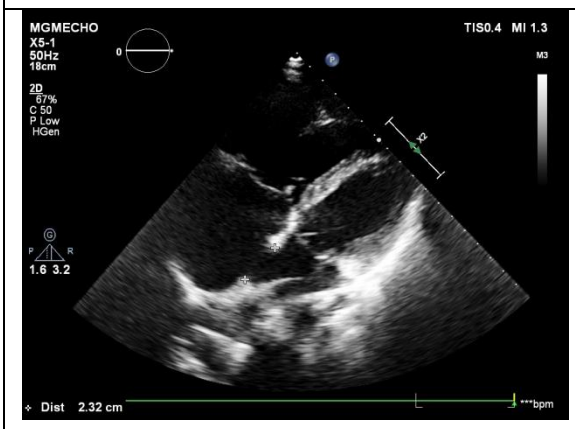
Trans Throsic view

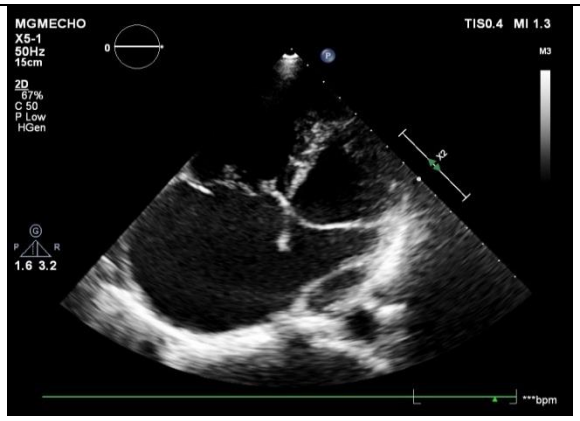


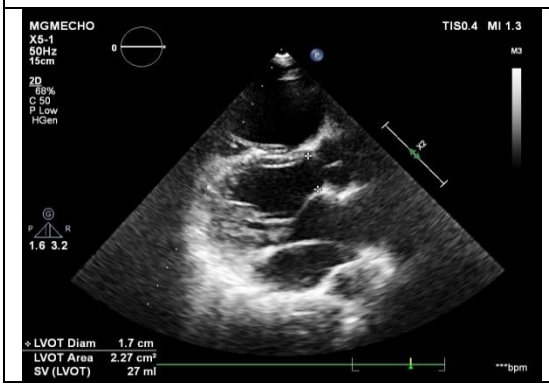
T T E A4 C View showing communication between accessory and true LA

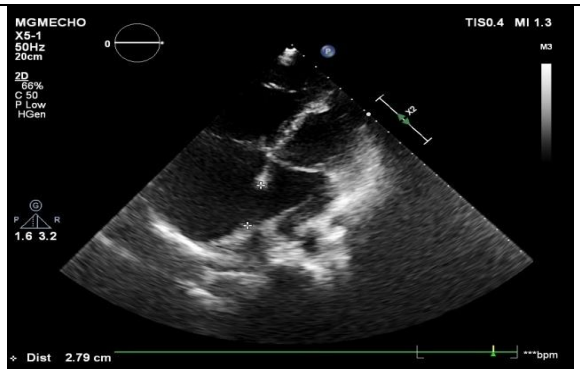




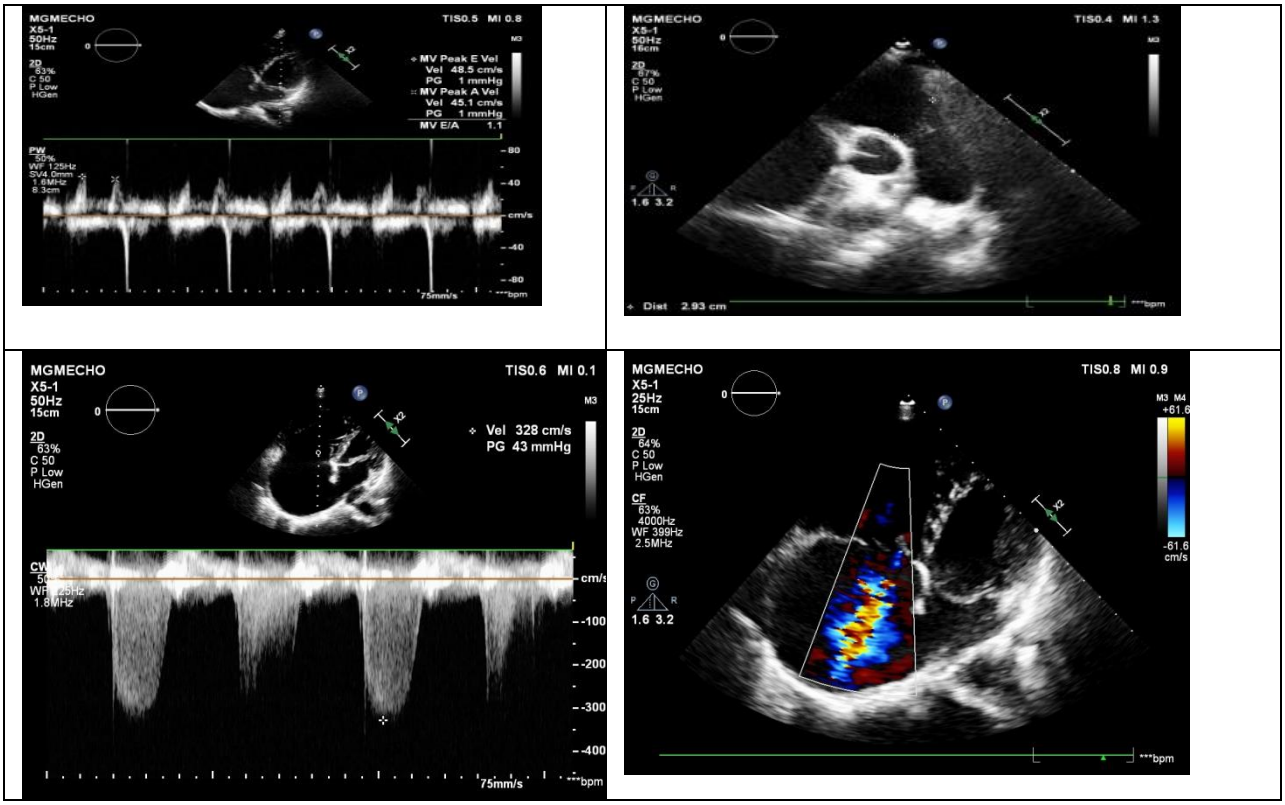
TTE A4c View demonstrating ASD b/w true LA and RA







ISSN: 0975-3583, 0976-2833 VOL15, ISSUE 6, 2024



Transesophageal Echocardiography

The patient underwent transesophageal echocardiography to confirm the diagnosis of cor triatriatum sinister (with the key pathognomonic finding of the left atrial appendage arising from the left atrial chamber, as opposed to the morphology of an SVMM. The cor triatriatum membrane was located immediately downstream to a secundum ASD. This ASD resulted in communication between the inferior chamber (True LA) and the right atrium. There was prominent left-to-right shunting. Four pulmonary veins were visualized connecting to the more superior pulmonary venous chamber of the left atrium.

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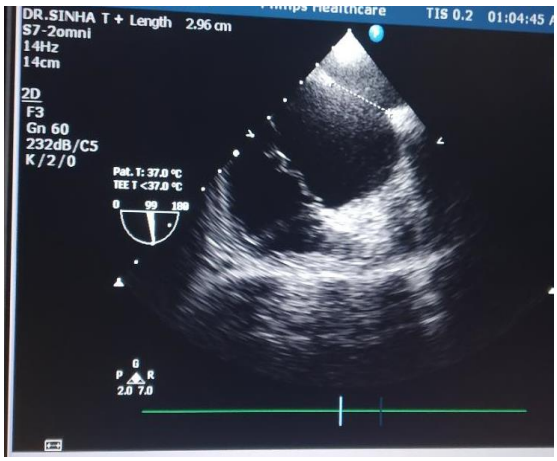
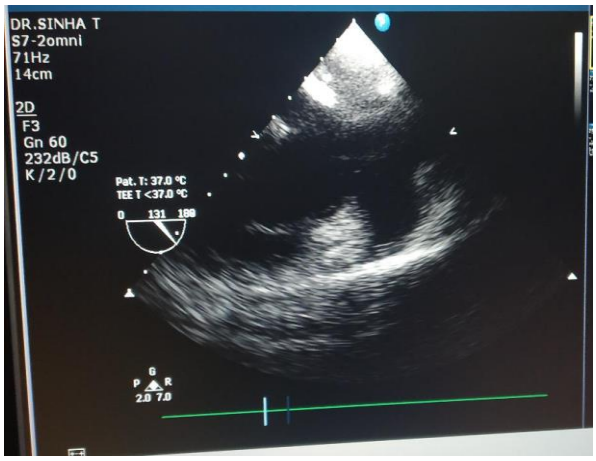


TEE View Demonstrating origin of LAA from True LA



TREATMENT

The medical treatment in CTS depends on symptoms. An incidental echographic finding of a left atrial extra-membrane with no pressure gradient in an asymptomatic patient does not require medical management. When exertional dyspnoea and signs of pulmonary congestion occur, diuretics, digoxin and preload reduction are the mainstay of medical therapy. In this case, surgical treatment is also usually indicated. Atrial arrhythmias should be adequately treated, since they may quickly lead to cardiac decompensation. In this instance, several successful cases of catheter ablation have also been described in literature even in unoperated CTS patients .

Surgical treatment is usually indicated in symptomatic children and adults having a significant intra-atrial obstruction. Historically, the first successful surgical repair was performed by Lewis et al. in 1956. Before the operation is undertaken, a careful evaluation of the associated abnormalities, especially atrial septal defect or abnormal venous return should be obtained by 3D TEE, computed tomography scanning or cardiac MRI. The operation is performed under hypovolemic cardio-pulmonary bypass using a right or left atrial approach. Usually most of the diaphragm is excised, the atrial septum is closed and the anomalous venous return is corrected if present. An immediate post operative TEE is usually carried out to control the surgical result.

Surgical outcome is usually favourable in well experienced centres, with nearly all patients becoming asymptomatic at follow-up and with an overall reported survival above 90% at five years .

CONCLUSION

Fully establishing the diagnosis cor triatriatum sinistrum is possible with no to minimally invasive cardiac imaging modalities, mainly three dimensional transthoracic and transoesophageal echocardiography. These valuable diagnostic tools help to optimise the management of this congenital heart disease whose rarity may frequently lead to misdiagnosis, but when adequately evaluated and treated the clinical outcome is excellent.

The End