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A PROMINENT EUSTACHIAN VALVE MIMICKING COR TRIATRIATUM DEXTRUM

Cardiology

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ABSTRACT

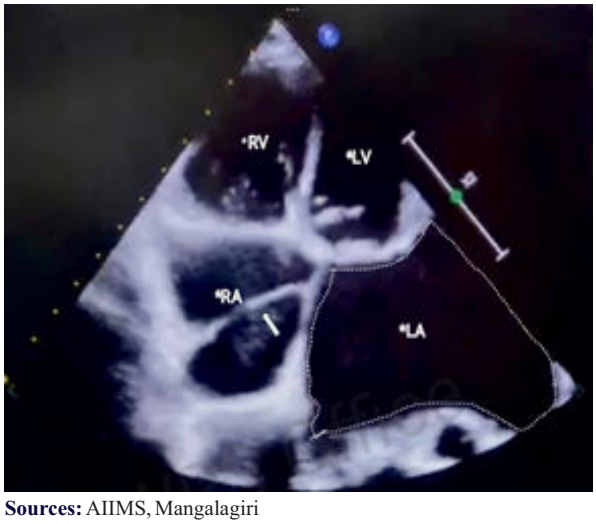
Persistent Eustachian valve (EV) is a rare congenital anomaly resulting from incomplete regression of the embryonic right sinus venosus valve. Although typically regressing during childhood, its persistence into adulthood is uncommon. We report a case of a young female with a history of rheumatic heart disease and prior percutaneous trans-mitral commissurotomy (PTMC) who presented with dyspnoea. Transthoracic echocardiography (TTE) revealed a large membranous structure consistent with a persistent EV, which mimicked Cor triatriatum dextrum (CTD) by dividing the right atrium (RA) into two chambers. Transesophageal echocardiography (TEE) could not be performed due to patient refusal, limiting detailed evaluation. While often asymptomatic, a persistent EV can lead to complications such as paradoxical thromboembolism, endocarditis, and difficulties with cardiac catheterization. Recognizing this anatomical variant is crucial for accurate diagnosis and management, particularly in interventional procedures or in the presence of thromboembolic events. Although some cases may require surgical intervention, this patient's condition did not necessitate active treatment. This case underscores the importance of distinguishing a persistent EV from CTD and understanding its clinical implications for appropriate management.

KEYWORDS

persistent eustachian valve (EV), right sinus venosus valve, cor triatriatum dextrum (CTD), paradoxical thromboembolism, endocarditis, cardiac catheterization.

CASE STUDY

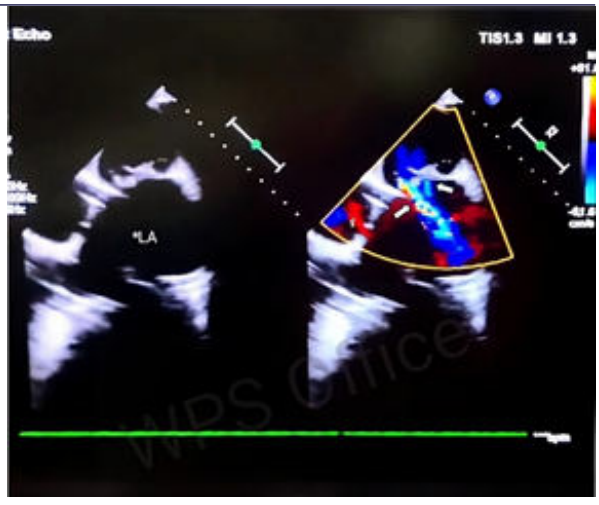
A young woman in her twenties with a history of rheumatic heart disease with mitral stenosis and childhood PTMC presented with worsening dyspnoea over past three months. Physical examination shows regular pulse, normal blood pressure, hyperdynamic apex, a soft S1, a grade 3/6 pansystolic murmur, a low-pitched rumbling mid-diastolic murmur, and a wide variable S2. Additionally, a pansystolic murmur in the fourth left intercostal space suggests tricuspid regurgitation (TR).



Sources: AIIMS, Mangalagiri

Figure 1: Echo cardiac image of heart in the four-chambered view (RA= right atrium, LA= left atrium, RV= right ventricle, LV= left ventricle). RA shows a prominent eustachian valve (white arrow). LA is dilated (dotted tracing)

TTE demonstrated a normally functioning left ventricle (LV) with an extensively dilated left atrium (LA) (Fig. 1) and a non-dilated LV. There was moderate mitral stenosis (MS) with a mitral valve area of 1.6 cm² and moderate-to-severe mitral regurgitation (MR) with two jets (Fig. 2), one of which appeared to originate from the base of the anterior mitral leaflet, likely due to a tear from the previous mitral valve procedure.



Sources: AIIMS, Mangalagiri

Figure 1: Colour Doppler focused on the left atrium (LA). Moderate-to-severe mitral regurgitation is seen with 2 separate jets (white arrows). Note that mitral valve area is decreased in size suggesting mitral stenosis. Atrial septal defect (possibly iatrogenic) is seen (asterisk).

Mild-to-moderate tricuspid regurgitation (TR) was observed, with a right ventricular systolic pressure (RVSP) of 45 mmHg. A small atrial septal defect (ASD), measuring 7-8 mm, with a left-to-right shunt, was likely due to prior septal puncture during PTMC.

The dilated right atrium (RA) contained an unusual structure that appeared to divide it into two chambers on 2D echocardiography. This membranous formation extended from the junction of the inferior vena cava (IVC) and the RA wall to the interatrial septum near the fossa ovalis. The IVC was compressive and non-dilated, measuring 22 mm.

A 7-mm discontinuity was noted at the junction of the cranial and caudal portions of the membrane, with an active jet passing through. The membrane was carefully differentiated from other congenital right atrial structures by assessing all its attachments and confirming these with various echocardiographic views.

DISCUSSION:

The EV, first described by Bartolomeo Eustachio in 1564 (E. M. Onorato, 2021), directs blood from the inferior vena cava (IVC) to the left atrium (LA) via the foramen ovale during fetal development, facilitating bypass of the underdeveloped fetal pulmonary circulation. In 1929, Yater further explored its function and variations (Wm, 1929). Normally, the EV regresses in childhood, but if persistent, it can cause varying degrees of obstruction in the right atrium, from partial separation to the complete division seen in CTD (Alboliras et al., 1987; E. Onorato et al., 2002).

Embryology: -

During early cardiac development, the right horn of the sinus venosus is flanked by two valves: the left and right venous valves. The left valve integrates into the septum secundum, while the right valve partially divides the RA. By the 9th to 15th weeks of gestation, the right valve regresses, with the cephalic portion forming the crista terminalis and the caudal portion becoming the EV of the IVC and the Thebesian valve of the coronary sinus. Persistent abnormalities of the right sinus venosus valve can lead to conditions ranging from partial septation of the RA by a prominent EV to a complete division of the RA, known as CTD (Doucette & Knoblich, 1963).

In some instances, the right sinus venosus valve may assume a windsock-like shape, potentially causing varied obstructions such as tricuspid valve interference, right ventricular outflow tract obstruction, IVC blockage, or affecting an existing ASD (Battle-Diaz et al., 1979; Jones & Niles, 1968; Rossall & Caldwell, 1957; Weinberg et al., 1985). Extensive fenestrations in the valve can lead to Chiari's network (Chiari, 1897).

The EV, a triangular structure with some amount of muscle tissue, originates at the lower part of the crista terminalis and extends along the IVC opening into the RA, reaching the fossa ovalis border. It forms the Eustachian ridge, which connects with the Thebesian valve, guarding the coronary sinus entrance (Standring, 2021).

Persistent Eustachian valves are present in about 67% of children but are rare in adults, appearing in only 0.20% of routine echocardiograms. The valve's size, shape, thickness, and texture vary widely, with an average length of 3.6 mm (range 1.5-23 mm) (Sarupria et al., 2014; Upadhyay et al., 2020). In this case, the valve measured approximately 24 mm.

CTD involves the persistence of the sinus venosus valve, creating a large obstructive flap that divides the RA into two chambers. The membrane typically extends from the IVC to the superior vena cava (SVC), separating the systemic venous portion of the right atrium from its vestibule and auricular complex. The upstream chamber receives blood from the great veins, while the downstream chamber includes the RA appendage (Yavuz et al., 2002). Newborns with CTD typically present with cyanosis within the first few days of life (Weyman, 1993). Echocardiographic examination often reveals an associated ASD with a right-to-left shunt. Surgical intervention is frequently necessary to ensure the infant's survival.

In this patient, a giant persistent EV was found without significant structural heart disease, and thus no active intervention was needed.

Complications:-

A prominent EV can redirect blood towards the interatrial septum. In patients with an ASD or patent foramen ovale (PFO), this may result in a right-to-left shunt, leading to cyanosis that requires surgical correction (Morishita et al., 1985). Endocarditis of the EV, typically seen in intravenous drug users, is treated with antibiotics or surgery based on the case's severity (Chan Pei Loon et al., 2017). The first documented case of EV endocarditis was reported by Edward et al. in 1986 (Edwards et al., 1986). It involved a patient who died of staphylococcal pneumonia despite antibiotic treatment.

The EV can also be a site for thrombus formation (E. M. Onorato, 2021). A thrombus arising in the venous system elsewhere in the body could also be lodged on the EV (Onwuanyi et al., 2003). Both the occasions potentially cause paradoxical thromboembolic events such as ischemic stroke (Nozue et al., n.d.) or pulmonary embolism (Onwuanyi et al., 2003). Myxomas can grow on the EV (Battle-Diaz et al., 1979).

In cardiac catheterization, a large EV increases the risk of difficult IVC cannulation (Sarupria et al., 2014) and may mimic the rim of an ASD, complicating closure procedures. During open procedures, the valve can be mistaken for an ASD rim, potentially resulting in incomplete closure and residual shunt. The EV can also hinder the proper placement of a PFO closure device in percutaneous procedures (E. Onorato et al., 2002). Persistent EV has been frequently observed in platypnea-orthodeoxia syndrome cases (Wm, 1929).

CONCLUSIONS/TAKE HOME MESSAGE

While the EV is typically regarded as a benign anatomical variant, CTD is considered more severe due to its associated complications and the intricate management and potential outcomes it entails. Clinicians must differentiate between EV and CTD by evaluating the structure's attachments within the RA. If needed, TEE should be employed to provide comprehensive imaging.

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