

# Large Left Atrial Appendage Associated with IVC Type Sinus Venosus ASD – A Rare Congenital Anomaly

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## Abstract

The large left atrial (LA) appendage associated with IVC Type PAPVC is a rare congenital anomaly and diagnosis in <4 year old was documented sparsely in the literature. The large LA appendage can cause serious complications such as arrhythmias, ventricular dysfunction, and stroke. A 4-year-old child with h/o recurrent respiratory tract infections and no other significant history was diagnosed as congenital acyanotic heart disease sinus venosus ASD type and was incidentally found to have a large LA appendage. Patient underwent PAPVC repair, her post-operative was uneventful and advised regular follow-up during discharge. Incidentally diagnosed large LA appendage without any symptoms or LA thrombus requires regular follow-up and further evaluation, those who are symptomatic should be treated either surgically or clipping in cath lab early.

**Key words:** Atrial septal defect, Inferior vena cava, Left atrial appendage

## BACKGROUND

The large left atrial (LA) appendage is a rare congenital anomaly and very rarely acquired. Large LA appendage is due to dysplastic pectinate muscles of atrium,<sup>[1]</sup> usually asymptomatic until the second to the third decade of age can present with complications of arrhythmias, stroke, and ventricular dysfunction<sup>[2]</sup> if symptomatic should be either surgically resected or to be clipped.<sup>[3,4]</sup>

## CASE PRESENTATION

A 3 year-old female child presented with h/o frequent respiratory tract infections, no other significant history and on examination had a murmur. Echo showed Siunus

venosus ASD ivc type where right lower pulmonary vein draining into IVC and a large LA appendage around 2.86 cm, LA velocity around 1.1 m/s, no LA thrombus, lv systolic and diastolic functions, and LA volumes are normal [Figures 1 and 2]. Blood investigations and coagulation profile were normal. Pulmonary venous anamoly closure was done surgically with short cross clamp time and short cardiopulmonary bypass time and extubated with in 2 h in ICU. Her post-operative was uneventful, discharged, and advised to have regular follow-up for LA appendage changes.

## DISCUSSION

Large LA appendage associated with IVC type PAPVC, probably first ever documented in the literature. The normal LA appendage size in adults is around 1.3–1.5 cm and the normal length of LA appendage in <4 year is yet to be documented. LA appendage grows with age and a decrease in ventricular contractility leads to elevated LA pressures. A decrease in the function of appendage contractility besides dysplastic pectinate muscles leads to further dilatation of appendage and switching from

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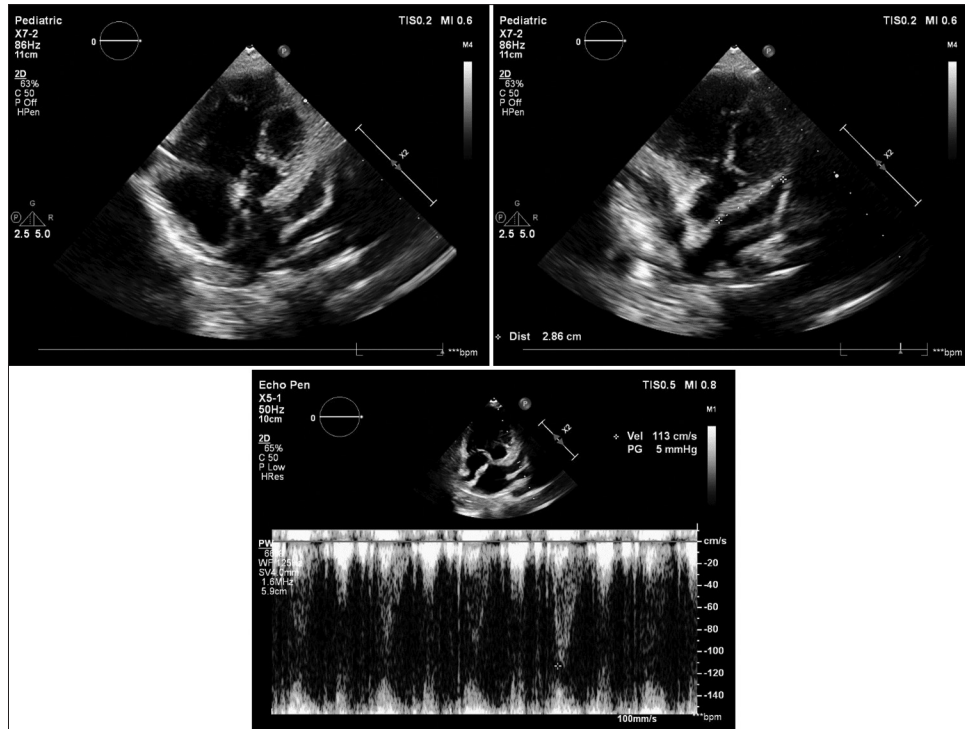


Figure 1: Transthoracic echocardiography images showing la appendage size and velocity

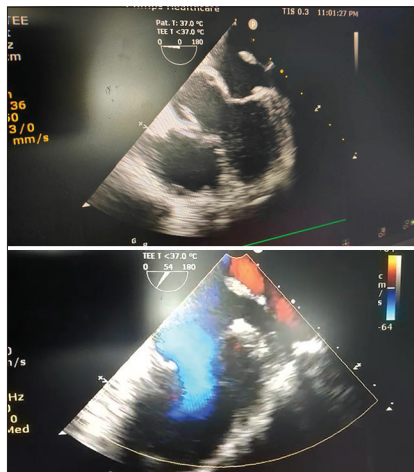


Figure 2: TEE

pump to reservoir function.<sup>[5]</sup> Enlarged LA appendage can cause complications such as arrhythmia, stroke, and ventricular dysfunction in second or third decade of age. Children can present early with the features of congestive heart failure<sup>[6]</sup> due to compression of pulmonary veins, distorted mitral annulus leading to mitral regurgitation, or symptoms of airway compression.<sup>[7]</sup> Symptomatic large LA appendage requires surgical intervention or clipping while asymptomatic patients can be managed conservatively with regular follow-up. Complications include thrombus formation and stroke in relation to the size of la appendage.<sup>[5,8]</sup> A correlation between

the size of LAA and the complications such as thrombus formation and stroke has not been documented till now.

## CONCLUSION

The large LA appendage in a PAPVC/IVC TYPE is a very rare anomaly probably the first to be reported in literature to the best of our knowledge. Symptomatic patients require intervention and asymptomatic patients without any thrombus can be managed conservatively with regular follow-up and further imaging of the Brain might require as there are chances of microemboli in asymptomatic patients.

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