Cardiology in the Young

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Non-obstructive membrane of left atrial appendage orifice: first paediatric report

Sakshi Sachdeva[1], Sushil Azad[2] and Sitaraman Radhakrishnan[2]

[1]Department of Cardiology, All India Institute of Medical Sciences, New Delhi, India and [2]Department of Pediatric Cardiology, Fortis Escorts Heart Institute and Research Centre, New Delhi, India

Abstract

Left atrial appendage membrane is a rare anomaly. Its implications on clinical course are unknown. We report the first paediatric case in association with a congenital heart disease.

Brief Report

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Author for correspondence:
Dr Sakshi Sachdeva, MD, Department of Cardiology, 7th Floor, Cardio Thoracic Centre, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, India, 110029.
Tel.: +91-9971363781.
Email: sakshisachdeva21@gmail.com

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Work was performed when corresponding author was affiliated to the Department of Pediatric Cardiology, Fortis Escorts Heart Institute and Research Centre, New Delhi, India

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Department of Pediatric Cardiology, Fortis Escorts Heart Institute and Research Centre, New Delhi, India

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Left atrial appendage is a structure of great interest to adult interventional cardiologists and electrophysiologists, given the risk of thrombus which might lead to systemic embolism in patients with atrial fibrillation.[1] In congenital heart diseases, left atrial appendage remains uninvolved and is often overlooked, with the exception of heterotaxy syndromes where the appendage morphology helps in diagnosing the type of isomerism. Left atrial appendage membrane is a very rare anomaly and has been reported in adults with structurally normal heart. We report a 10-month infant with non-obstructive partial membrane at the mouth of left atrial appendage in association with patent ductus arteriosus.

A 10-month infant presented with symptoms of significant post-tricuspid left to right shunt. He had tachypnoea, tachycardia, and signs of aortic run-off. There was cardiomegaly with hyper-dynamic apex and continuous murmur in left infra-clavicular region. Chest radiograph revealed cardiomegaly with features of pulmonary plethora. Trans-thoracic echocardiogram confirmed the diagnosis of moderate patent ductus arteriosus measuring 2.5 mm at pulmonary artery end, shunting left to right with peak systolic and diastolic gradient of 81 and 53 mmHg, respectively. On high para-sternal short-axis view, there was an unusual, thin and floppy, membrane-like structure between left pulmonary artery and ductus, which moved back and forth with each cardiac cycle. It was initially thought to be a ductal valve (Fig 1a; Supplementary video 1). However, upon focussed evaluation, on posterior tilt in para-sternal short-axis view, the structure was seen to lie at the orifice of left atrial appendage, partially occluding its opening into left atrium with no obstruction to flow, as seen on colour Doppler and pulse Doppler evaluation (Fig 1b and Fig 2; Supplementary videos 2 and 3). On detailed trans-thoracic imaging, this structure was diagnosed as a non-obstructive membrane at the orifice of left atrial appendage. Patient underwent successful patent ductus arteriosus device closure and is well at 2 years of follow-up. He has been kept under 2–3 yearly follow-up and parents informed about reporting if symptoms suggestive of thromboembolism or arrhythmia develop.

Membrane at the orifice of left atrial appendage is a very rare anomaly, with only few cases reported in literature within the last decade. In majority of cases, the membrane is non-obstructive,[2,3] but some cause obstruction to flow across the appendage.[4] Rarely the membrane can be fenestrated and cause turbulence of blood flow across it.[5] Half the cases reported were with atrial fibrillation and/or stroke and remaining were incidental findings.[6,7] According to the previous reports, there are two locations described for the appendage membrane. Membrane can lie either at the base of appendage, i.e. at the junction of left atrium and appendage, also called as the orifice/mouth of appendage.[4,5,7] The second location of membrane is within the cavity of appendage.[2,3,6] In our case, membrane was located at the orifice of appendage. Previous reports, which are all in adults, describe trans-oesophageal imaging for diagnosis of this entity. In view of good trans-thoracic imaging windows in the infant, trans-oesophageal imaging was not carried out in our patient. The entity can sometimes be over-diagnosed, so due care should be taken while identifying it from other structures like pectinate muscles, side-lobe artefacts, and partial resolution of appendage thrombi.[8]

Embryologically, appendage is the only part of the left atrium which arises from the wall of primary atrium. Remaining portion of atrium is formed from branches of primordial pulmonary veins.[9] Embryological origin and clinical significance of the membrane at the orifice of left atrial appendage is not yet clear. Whether the membrane predisposes to thrombus formation by reducing the blood flow velocity across appendage or it prevents embolism by reducing the orifice area of appendage is yet to be proved.[10,11]

We report the first paediatric case of membrane at the orifice of left atrial appendage, and the first in association with congenital heart disease. Once detected, it may have clinical implication in relation to arrhythmia or the risk of thrombus formation in appendage. Apart from this, the

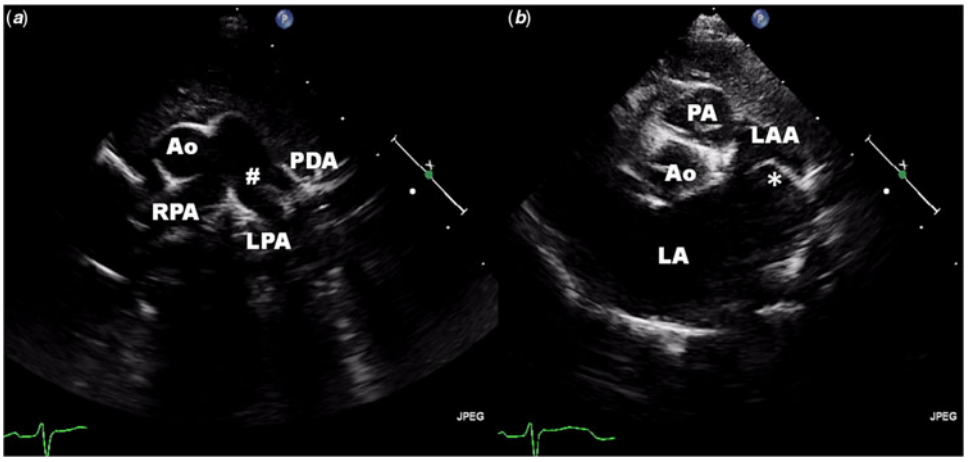


Figure 1. Trans-thoracic echocardiogram in para-sternal short-axis view (***a***) at the level of pulmonary artery showing a membrane (#) near junction of LPA and PDA mimicking ductal valve, and (***b***) at the level of left atrial appendage showing a non-obstructive membrane (*) at its base. Ao=Aorta; LA=left atrium; LAA=left atrial appendage; LPA=left pulmonary artery; PDA=patent ductus arteriosus; RPA=right pulmonary artery.

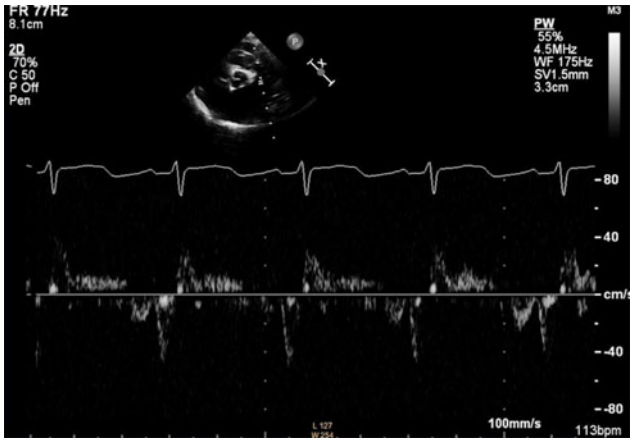


Figure 2. Pulse Doppler interrogation with sample volume kept across the orifice of left atrial appendage showing normal Doppler velocities.

appendage membrane can interfere with evaluation of patent ductus arteriosus, as in our patient. The recognition of the existence of this entity can help in quick assessment. As the appendage modulates the flow volume dynamics of the left atrium, once detected should these patients be kept in a close follow-up is an unanswered question.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951120001080

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Conflicts of Interest. None.

Ethical Standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (please name) and with the Helsinki Declaration of 1975, as revised in 2008.

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