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Case Report
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Congenital Absence of Left Atrial Appendage: A Multimodality Evaluation

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Abstract

Left Atrial Appendage (LAA) contributes toward LA reservoir and contractile functions. However, the LAA is also the most common site for thrombus formation in Atrial Fibrillation (AF). The shape of the LAA is variable, but the absence is extremely rare. A man with no past cardiac history was admitted for catheter ablation of AF. Before the procedure, the patient underwent a Transoesophageal Echocardiography (TEE) examination, but neither cardiac thrombus nor LAA could be detected. Absence of LAA was confirmed by cardiac Computer Tomography (CT).

Introduction

Before the procedure, the patient underwent a Transoesophageal Echocardiography (TEE) examination [1], but neither cardiac thrombus nor LAA could be detected. Absence of LAA was confirmed by cardiac Computer Tomography (CT).

Case Presentation

A 69 year old man, with persistent AF, was referred to our centre for crioablation procedure. Two-dimensional (2D) and three-dimensional (3D) TEE was scheduled to exclude thrombus before the interventional procedure, using Vivid E9 Ultrasound system (General Electric Vingmed Ultrasound, Horton, Norway). Despite 2D TEE imaging at multiple angles, LAA could not be visualized and color Doppler imaging characteristic flow of LAA could not be demonstrated (Figure 1). The 3D TEE "en face" view demonstrated the absence of the anatomical orifice of LAA, below left upper pulmonary vein and lateral ridge. Cropping the 3D data set, LAA body wasn't identified; only a small residue like a rudimentary LAA was visible (arrows, Figure 2). The patient wasn't cleared for crioablation procedure, as complete occlusion of LAA with a thrombus remained a reason for its invisibility. Contrast-enhanced multidetector CT, using Dual Source technique (Somaton Definition Flash, Siemens, Erlangen, Germany), was performed for further delineation and confirmed the suspicion of congenital absence of the LAA with optimal spatial resolution (Figure 3), because LAA

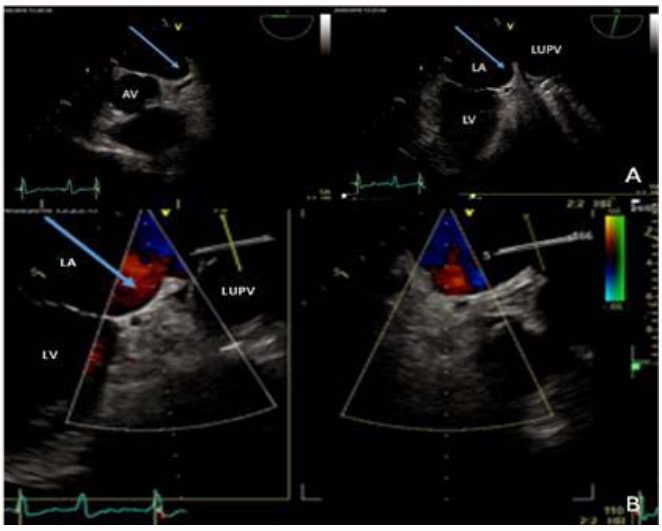

Figure 1: TEE images of the left atrium at the mid oesophageal level: on 2DTEE images (angles of 0 and 76 degrees) LAA was not visualized (arrows, panel *A*), even by Doppler (arrow, panel B). AV aortic valve; LA left atrium; LV left ventricle; LUPV left upper pulmonary vein.

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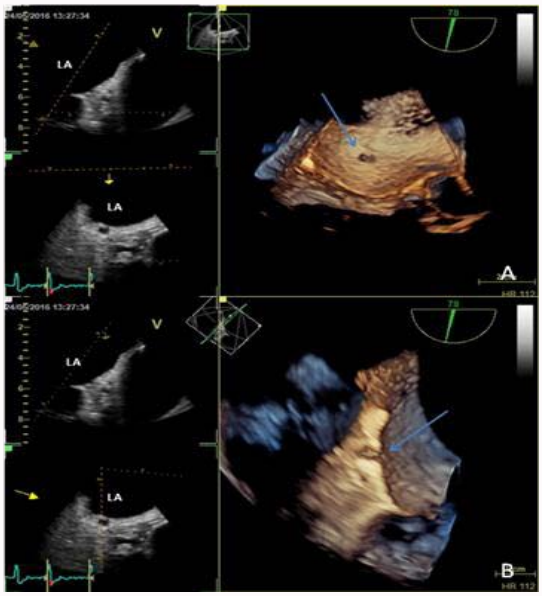

Figure 2: 3DTEE demonstrated the absence of LAA (panels A,B); a small residue was visible (arrows). LA left atrium.

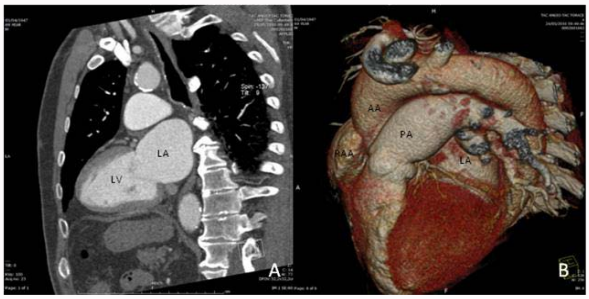

Figure 3: CT images on 2 chambers view (panel A) and 3D volume reconstruction (panel B) showed absence of LAA, with a clear definition of the right atrial appendage. LA left atrium; LV left ventricle; RAA right atrial appendage; PA pulmonary artery; AA ascending aorta.

body wasn't visible on the surface of the LA at its normal location. Anatomical reconstruction with Navx-/Ensite-system (Endocardial Solutions, St. Jude Medical, Inc. St. Paul, MN, USA) of LA during catheter ablation led to the same conclusions showing pulmonary veins in the usual anatomic pattern and LAA absence (Figure 4).

Conclusion

Congenital absence of LAA is a rare cardiac anomaly and only a few cases have been reporter in the literature [2-4]. The differential diagnosis for no visualization of the LAA during 2D TEE includes thrombus, variant anatomical features, poor echocardiographic windows, prior surgical ligation or insertion of an occluder device. Highly sophisticated imaging modalities, like multidetector CT and 3D TEE, could be performed to confirm the presence or absence of the LAA and to identify filling defects, because they offer optimal spatial resolution and in particular 3D TEE has the peculiar ability of an "en face" real time visualization of the LAA orifice, allowing the definition of anatomical details. Physiological consequences and the impact on cardio-embolic risk of a congenitally absent LAA are unknown and it seems more likely congenital anatomical variation.

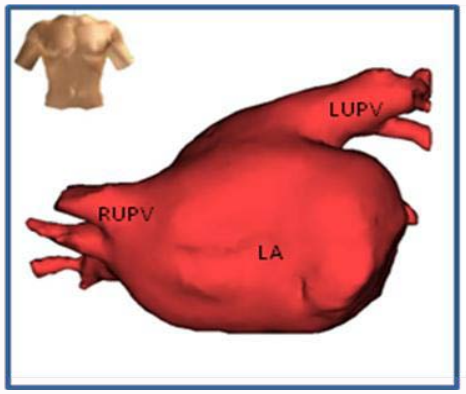

Figure 4: Anatomic reconstruction of the LA during catheter ablation. LA left atrium; LV left ventricle; LUPV left upper pulmonary vein; RUPV right upper pulmonary vein.

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