A 73-year-old woman with no history of cardiac surgery presented with symptomatic atrial fibrillation. A transesophageal echocardiogram was scheduled to exclude thrombus before cardioversion. No obvious intracardiac thrombus was identified, but the patient could not be cleared for cardioversion because the left atrial appendage (LAA) had not been visualized, raising concerns for either a flush occlusion with thrombus or a small ectopic appendage (Figure 1 and online-only Data Supplement Videos I-IV). Contrast-enhanced multidetector computed tomography revealed moderate bialtrial enlargement without intracardiac thrombus and confirmed a diagnosis of congenital absence of the LAA, a previously unreported finding (Figure 2 and online-only Data Supplement Videos V and VI).

The LAA typically develops in the third week of embryonic life, contains high concentrations of atrial natriuretic factor, and contributes toward left atrial reservoir and contractile functions. However, the LAA is also the most common site for thrombus formation in atrial fibrillation. The physiological consequences of a congenitally absent LAA are unknown.

The differential diagnosis for nonvisualization of the LAA during transesophageal echocardiogram imaging includes flush thrombus, variant anatomical features, poor echocardiographic windows, prior surgical ligation, or insertion of an occluder device. Either multidetector computed tomography or cardiac magnetic resonance imaging could be performed to confirm the presence or absence of the LAA and to identify filling defects. Multidetector computed tomography offers optimal spatial resolution and a rapid diagnosis, although delayed imaging is sometimes required to differentiate between slow-flow and thrombus. Cardiac magnetic resonance imaging with dedicated imaging planes through the LAA offers an alternative nonionizing radiation technique, with the caveat that small mobile thrombus (≤1 cm) could be missed because of limitations in temporal and spatial resolution.

Figure 1. Transesophageal electrocardiographic imaging of the left atrium. Multiple views were obtained at midesophageal level, but it was not possible to visualize the left atrial appendage (A-C). An echo-free space was seen in the usual location of the left atrial appendage, but administration of intravenous echo contrast revealed this structure to be distinct from the left atrial cavity (D). This structure corresponded to the great cardiac vein at cardiac CT. LA indicates left atrium.
Disclosures
None.

References

Figure 2. Multidetector computed tomography imaging of the left atrium. Left atrial anatomical characteristics were evaluated using multiplanar reconstruction axial images (A) and 3 dimensional volume-rendered images (B). The great cardiac vein was clearly visualized traversing the left atrium anterolaterally. LAA indicates left atrial appendage; RUPV, right upper pulmonary vein; LLPV, left lower pulmonary vein; LV, left ventricle; GCV, great cardiac vein; AscAo, ascending aorta; DescAo, descending aorta; PA, pulmonary artery.

Key Words: congenital • left atrial appendage • magnetic resonance imagingy • multidetector computed tomography • transesophageal echocardiography
Congenital Absence of the Left Atrial Appendage
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SUPPLEMENTAL MATERIAL
**Video Legends:**

**Video 1:** Trans-esophageal echo video clip of the left atrium.  
Caption: Mid-esophageal view at 0°.

**Video 2:** Trans-esophageal echo video clip of the left atrium.  
Caption: Mid-esophageal view at 75°.

**Video 3:** Trans-esophageal echo video clip of the left atrium.  
Caption: Mid-esophageal view at 90° following administration of contrast.

**Video 4:** Trans-esophageal echo video clip of the left atrium.  
Caption: Mid-esophageal view at 77° with contrast.

**Video 5:** MDCT multi-planar sequence (3mm axial slices) of the left atrium.  
Caption: For comparative purposes, a similar sequence of a typical LAA is shown in a highlight box.

**Video 6:** MDCT 3d volume rendered sequence of the left atrium.  
Caption: For comparative purposes, a similar sequence of a typical LAA is shown in a highlight box.