The "absent" left atrial appendage...



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Introduction

Isolated congenital absence of the left atrial appendage (LAA) is a rare condition. The diagnosis results from a review of the past medical and surgical history in addition to the use of multimodality imaging, as several other conditions (thrombotic occlusion, surgical or percutaneous exclusion, variations in morphology and relative position of anatomic structures) may present with a similar finding.

Case report

A 62-year-old woman with a history of obesity, hypertension, and symptomatic persistent atrial fibrillation for which

she was treated with chronic oral anticoagulation elected to undergo catheter ablation. A preprocedural cardiac computed tomography showed a single left-sided and 2 right-sided pulmonary veins, no intracardiac thrombus, and no evidence of an LAA. Given no history of a prior cardiac procedure or surgery, absence of the LAA was presumed to be congenital. Transesophageal echocardiography and 3-dimensional computed tomography reconstruction of the left atrium in an electroanatomic mapping system (CARTO, Biosense Webster, Diamond Bar, CA) similarly demonstrated no LAA (Figure 1A and B). Her anatomy created no challenges for cryoballoon pulmonary vein isolation, which was successfully performed without complication.

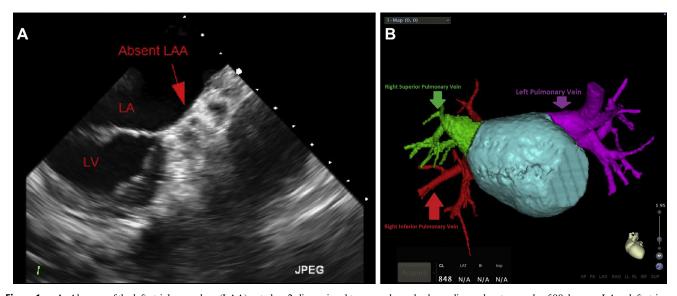


Figure 1 A: Absence of the left atrial appendage (LAA) noted on 2-dimensional transesophageal echocardiography at an angle of 90 degrees. LA = left atrium; LV = left ventricle. B: Anterior view of a 3-dimensional computed tomography reconstruction of the left atrium rendered in an electroanatomic mapping system (CARTO, Biosense Webster).

KEYWORDS Congenital; Atrial appendage; Atrial fibrillation; Absence; Ablation; Imaging; Cardiac CT; TEE (Heart Rhythm Case Reports 2017;3:494–495)

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Discussion

The appendage, unlike the pulmonary veins and posterior left atrium, derives from the primitive atrium, a structure that develops into a basal mesodermal layer, endocardial trabeculae, and pectinate muscles. The LAA accounts for $\sim 90\%$ of intracardiac thrombi associated with

KEY TEACHING POINTS

- Isolated congenital absence of the left atrial appendage is a rare condition.
- The diagnosis is best made after a comprehensive review of a patient's past medical and surgical history in addition to use of multimodality imaging, as other conditions (thrombotic occlusion, surgical or percutaneous exclusion, variations in morphology and relative position of anatomic structures) may present with a similar finding.
- It is plausible, although not known, that among patients with nonvalvular atrial fibrillation those with isolated congenital absence of the left atrial appendage may have a naturally occurring decreased risk for cardioembolic stroke.

nonvalvular atrial fibrillation. Given data demonstrating that occlusion of the LAA serves as a safe, alternative means of reduction of cardioembolic stroke risk when

compared to warfarin,^{2,3} it is plausible that congenital absence of the LAA conveys a similar naturally occurring risk reduction. However, it is unlikely that this will ever be meaningfully examined, given the rarity of the condition, with only a few reports of isolated congenital absence of the LAA noted in literature.^{4,5}

References

- Naksuk N, Padmanabhan D, Yogeswaran V, Asirvatham SJ. Left atrial appendage: embryology, anatomy, physiology, arrhythmia and therapeutic intervention. JACC Clin Electrophysiol 2016;2:403

 –412.
- Holmes DR Jr, Kar S, Price MJ, Whisenant B, Sievert H, Doshi SK, Huber K, Reddy VY. Prospective randomized evaluation of the Watchman Left Atrial Appendage Closure device in patients with atrial fibrillation versus long-term warfarin therapy: the PREVAIL trial. J Am Coll Cardiol 2014; 64:1–12
- Wiebe J, Franke J, Lehn K, Hofmann I, Vaskelyte L, Bertog S, Sievert H. Percutaneous left atrial appendage closure with the Watchman device. JACC Cardiovasc Interv 2015;8:1915–1921.
- Saleh M, Balakrishnan R, Castillo Kontak L, Benenstein R, Chinitz LA, Donnino R, Saric M. Congenital absence of the left atrial appendage visualized by 3D echocardiography in two adult patients. Echocardiography 2015; 32:1206–1210.
- Collier P, Cavalcante JL, Phelan D, Thavendiranathan P, Dahiya A, Grant A, Kwon D, Thamilarasan M. Congenital absence of the left atrial appendage. Circ Cardiovasc Imaging 2012;5:549–550.