Congenital Absence of the Left Atrial Appendage: Role of Multimodality Imaging

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INTRODUCTION

Congenital absence of the left atrial appendage (abLAA) is extremely rare. Physiological function of the left atrial appendage (LAA) is not well understood, yet it is an important source for thromboembolism in atrial fibrillation (AF).

CASE PRESENTATION 1

A 57-year-old patient with paroxysmal AF (CHADSVASc2 score 4) presented to our emergency room with palpitations and shortness of breath. Medical history revealed apical hypertrophic cardiomyopathy and an implantable cardioverter-defibrillator that was recently placed due to ventricular tachycardia. The patient did not have a history of cardiac surgery. Upon presentation, blood pressure was 110/79 mm Hg and pulse was irregular and rapid at 144 bpm. The chest x-ray showed mild pulmonary edema. Electrocardiogram showed AF with rapid ventricular response at a heart rate of 137 (Figure 1). The patient was sent for urgent electrical cardioversion. Preprocedure transesophageal echocardiogram (TEE) could not locate the LAA (Figure 2). A three-dimensional (3D) reconstruction of the left atrium (LA) also failed to reveal the LAA or flushed thrombosis (Video 1).

Considering that there was no evidence of prior open-heart surgery or percutaneous exclusion of LAA, a differential diagnosis of hypoplastic or aplastic LAA, a fully thrombosed LAA, or unusual anatomy of LAA was entertained. After echocardiographic imaging of the LA for either anomalous LAA position or unusual anatomy, abLAA was ultimately suspected. A contrast-enhanced cardiac computed tomography (CCT) scan was performed to corroborate this suspicion because abLAA is a rare presentation and it was of ultimate importance to exclude an appendage thrombus prior to an elective cardioversion.

Cardiac computed tomography demonstrated no evidence of an LAA (Figure 3). Given no history of prior cardiac surgery, abLAA was presumed to be congenital. In addition, the CCT revealed an anomalous right coronary artery originating from the left coronary cusp through a slit-like ostium with an interarterial course (Figure 4).

VIDEO HIGHLIGHTS

Video 1: Case 1: 3D TEE midesophageal surgeon's view demonstrates the smooth echo texture of the upper lateral atrial wall (*) representing the abLAA.

Video 2: Representative 3D TEE example of a normal LAA from a comparison surgeon's view demonstrates a normal ostium of the LAA (#).

Video 3: Representative 3D TEE example of an LAA from a comparison surgeon's view after placement of an LAAO device (Watchman).

Video 4: Representative 3D TEE example of an LAA after surgical clipping with a residual small LAA stump (S) in a patient who underwent bioprosthetic MVR surgery. *BioP MV,* Bioprosthetic mitral valve.

Video 5: Representative 3D TEE example of an LAA after surgical exclusion with a layer of thrombus covering the residual stump and extending into the adjacent LA wall (?). *L,* left lower pulmonary vein ostium; *U,* left upper pulmonary vein ostium.

Video 6: Case 1: 3D volume-rendered CCT whole-heart image of the LA with the abLAA (*thick green arrow* indicates usual location of the LAA).

Video 7: Three-dimensional volume-rendered CCT whole-heart image with a normal appearing and normally located LAA *(arrow).*

View the video content online at www.cvcasejournal.com.

These additional findings were discussed with the referring physician, and a follow-up visit was planned for further testing.

The patient was successfully cardioverted without complications to sinus rhythm and was continued on intravenous heparin after the procedure as bridging to oral anticoagulation. Rivaroxaban was ultimately started upon discharge.

CASE PRESENTATION 2

A 71-year-old patient with history of paroxysmal AF and hyperlipidemia presented with palpitations and persistent chest heaviness that started abruptly. Preliminary cardiac workup included a transthoracic echocardiography (TTE) that demonstrated severe mitral regurgitation (estimated by color flow Doppler) that was associated with rheumatic heart disease and a visually estimated left ventricular ejection fraction of 55%. Cardiovascular magnetic resonance imaging was performed and confirmed the mitral regurgitation was severe, with a regurgitant fraction of 57%. There was no mitral stenosis. A coronary angiogram showed normal coronary arteries. The patient was referred to our institution for surgical mitral valve replacement (MVR). Upon

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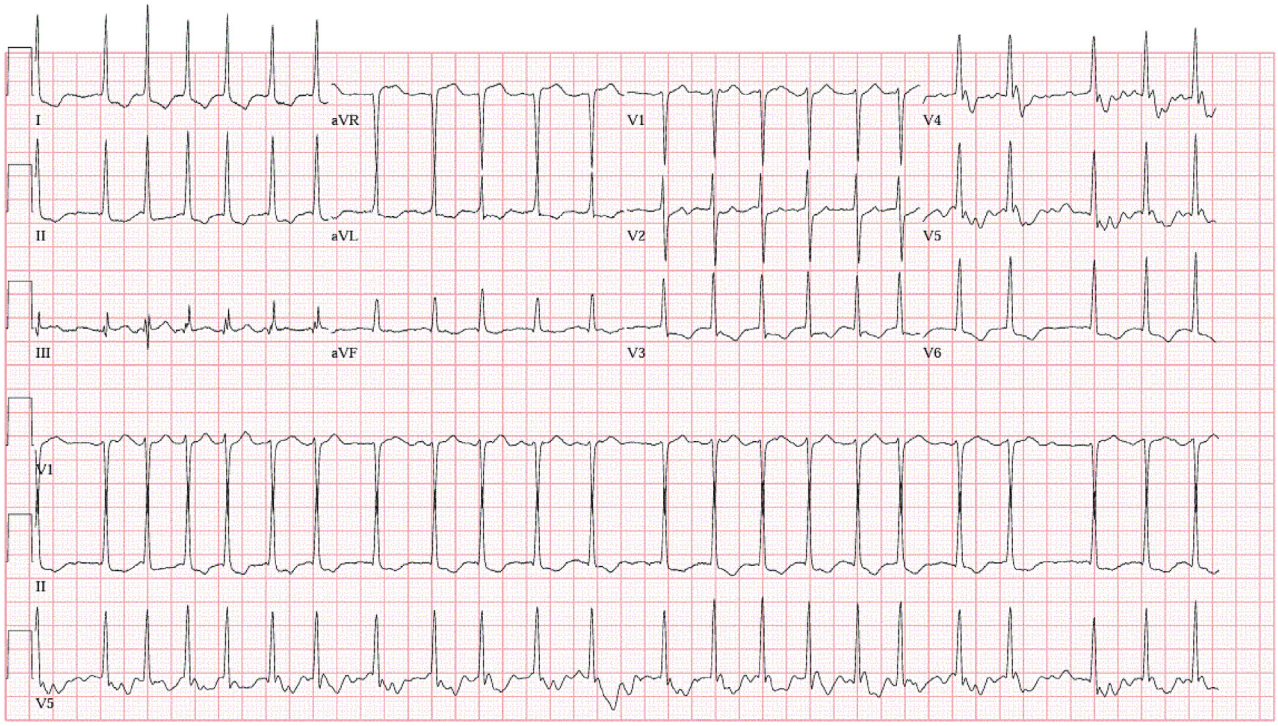

Figure 1 Twelve-lead electrocardiogram showed AF with rapid ventricular response at a heart rate of 137 bpm with ST depression and T-wave inversion in the lateral leads.

admission, a 12-lead electrocardiogram showed AF with rapid ventricular response; therefore, LAA ligation was also planned during open-heart surgery. Preoperative TEE failed to visualize the LAA (Figure 5). Congenital abLAA was visually confirmed during surgical MVR, and it is stated in the operative report as "an atretic LAA."

DISCUSSION

We hereby report 2 cases of congenital abLAA. There are only 23 reported cases of congenital abLAA (Table 1). This report also demonstrates the coexistence of a congenital abnormality,

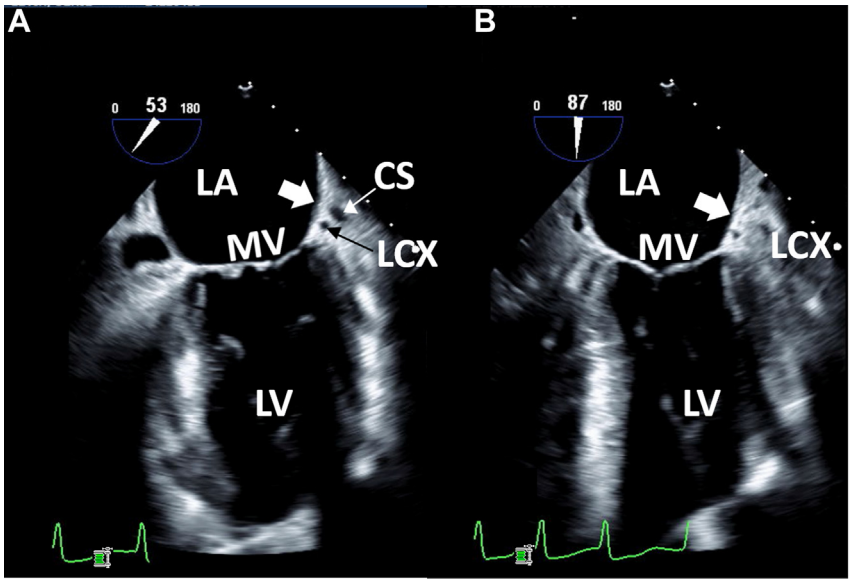

Figure 2 Case 1: two-dimensional TEE, midesophageal long-axis view, systolic view highlighting the mitral commissure (**A**; 53°) and conventional 2-chamber (**B**; 87°) view indicating the usual location of the LAA *(thick white arrow)*. The CS and LCx are well visualized in the atrioventricular groove indicating the usual anatomic location of the LAA. *CS*, Coronary sinus; *LCx*, left circumflex coronary artery; *LV*, left ventricle; *MV*, mitral valve.

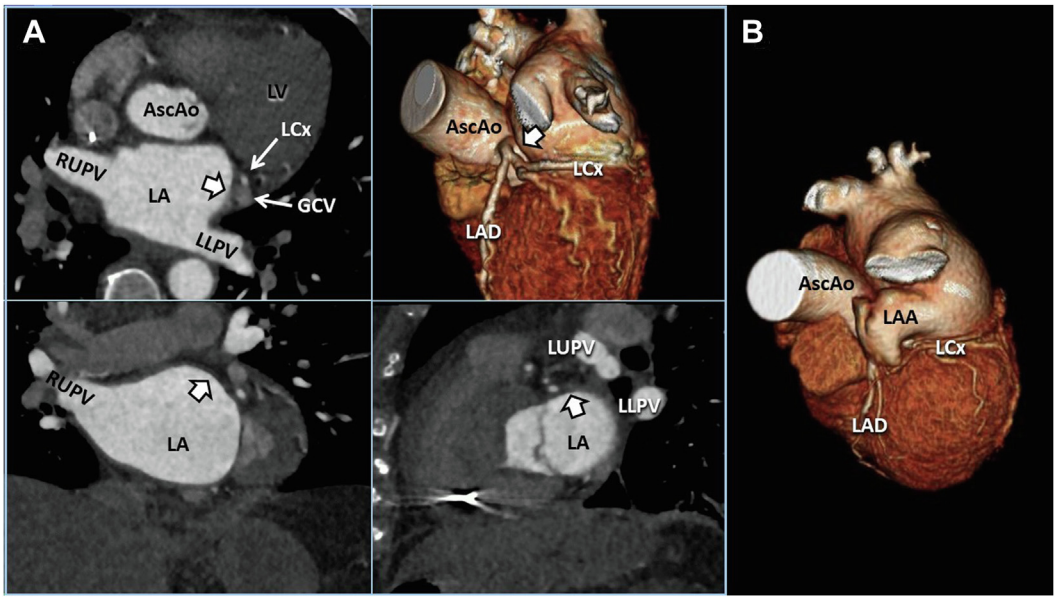

Figure 3 Case 1: CCT imaging demonstrates 3D cross-correlation of case 1 (**A**; *quad panel*) compared with a representative normal 3D volume-rendered example (**B**; *right single panel*). The multiplanar reformat quad display allows comprehensive demonstration of the abLAA *(thick white arrow)* in axial *(upper left)*, coronal *(lower left)*, and sagittal *(lower right)* orientations. *AscAo*, Ascending thoracic aorta; *GCV*, great cardiac vein; *LAD*, left anterior descending coronary artery; *LCx*, left circumflex coronary artery; *LLPV*, left lower pulmonary vein; *LUPV*, left upper pulmonary vein; *LV*, left ventricle; *RUPV*, right upper pulmonary vein.

specifically anomalous right coronary artery in one of the patients. Given the lack of high-risk imaging or clinical features for sudden cardiac death, no surgical intervention was recommended.[1] Overall, 20% of reported cases of abLAA are associated with other cardiac congenital abnormalities, warranting careful and thorough evaluation.[2-5]

Transthoracic echocardiography is the initial imaging study performed, frequently followed by TEE for a more detailed assessment

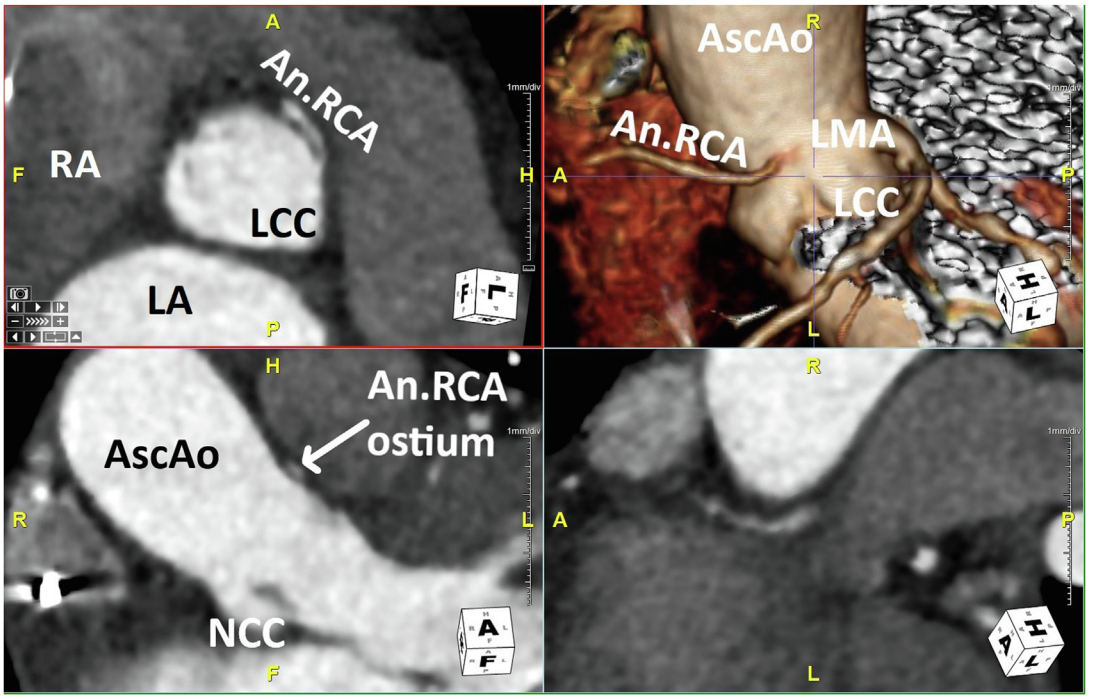

Figure 4 Case 1: CCT multiplanar reformat quad display demonstrates the anomalous RCA origin through a slit-like ostium (arises from the LCC) and interarterial course from multiple perspectives. *An.RCA*, Anomalous right coronary artery; *AscAo*, ascending thoracic aorta; *LCC,* Left coronary cusp; *LMA*, left main coronary artery; *NCC*, noncoronary cusp; *RA*, right atrium.

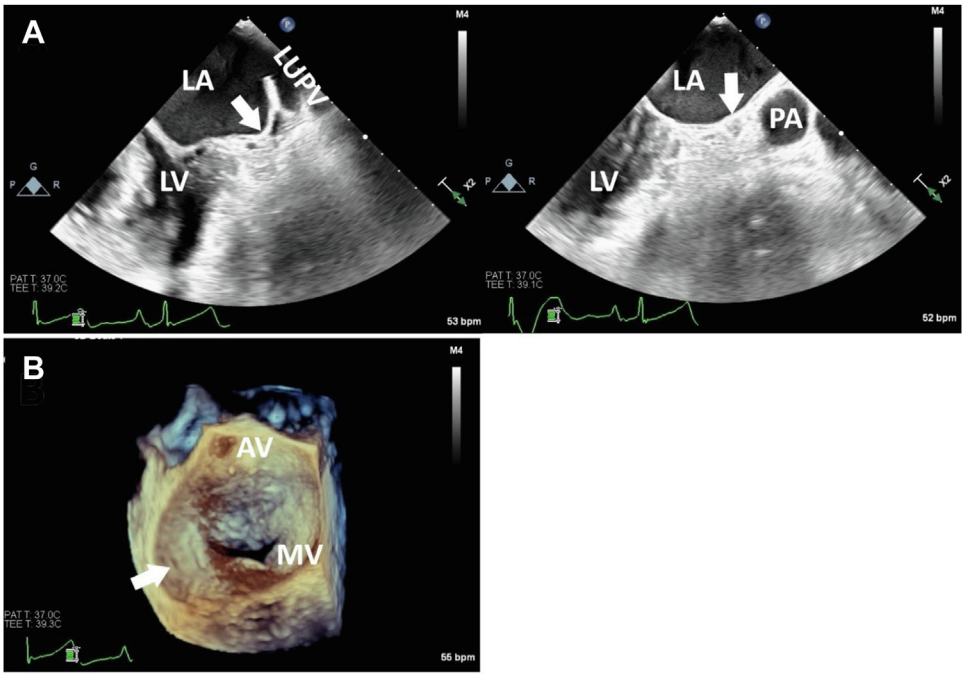

Figure 5 Case 2: two-dimensional TEE, midesophageal, long-axis systolic views at 45° (**A**; *top left*) and 90° (**A**; *top right*) demonstrates the abLAA *(arrows)*. The TEE 3D display, surgeon's view, demonstrates the en face view of the mitral valve from the perspective of the LA with the abLAA *(arrow)*. *AV*, Aortic valve; *MV*, mitral valve; *LUPV*, left upper pulmonary vein; *LV*, left ventricle; *PA*, main pulmonary artery.

of the cardiac valves or the LAA. This pattern was followed in both cases in this report whereby congenital abLAA was suspected; however, the diagnosis was not ascertained at that moment due to the rarity of the condition. The diagnosis was verified with CCT in the first case and with visual assessment during open-heart surgery in the second case. Anomalous coronary artery was also assessed by CCT in the first case.

The development of the LAA starts around the third week of gestational life. It is derived from the embryonic LA, whereas the remaining portion of the LA is formed from the branches of the primordial pulmonary veins.[6,7] Three different scenarios can cause the LAA to be absent from its usual anatomical location (to the left of the root of the pulmonary trunk). The first scenario is congenital anomaly of right juxtaposition of the atrial appendages, which places the LAA on the right side by the right atrial appendage and ascending aorta.[8] Second, spontaneous inversion of the LAA is observed, although rarely, as a complication of cardiac surgery with cardiopulmonary bypass surgery and vacuum application.[9,10] This should be suspected when a new LA mass is seen on intraoperative TEE.[10,11] And third is true congenital abLAA.

The LAA has attained significant importance in patients with AF since it serves as a trigger for the arrhythmia and also as the main source of thromboemboli with subsequent stroke, especially after the first 48 hours from onset.[12,13] Transesophageal echocardiography is frequently used as the imaging modality to evaluate LAA thrombus prior to cardioversion given its ease of clinical setting and lack of exposure to ionizing radiation. Appropriate visualization of an existing LAA, whether previously excluded or not, is of utmost importance to rule out the presence of thrombus prior to elective cardioversion. Understanding the highly variable normal echocardiographic anatomy of the LAA (Video 2) and the appearance of the LAA after placement of an LAA occlusion device (LAAO; Video 3) or surgical clipping (Video 4) or exclusion (Video 5) is important to recognize pathology. Importantly, CCT provides excellent images of the LAA, and 3D whole-heart cine data may be diagnostic (Videos 6 and 7).

Given the lack of studies regarding the best management of anticoagulation in cases with congenital abLAA, the approach is variable, with some practitioners favoring continuation as per the patient's CHADSVASC2 score, while others do not, arguing that the absence of the LAA stands in place for a totally excluded one (Table 1).

CONCLUSION

Congenital abLAA is a rare condition. A multimodality imaging approach is instrumental in arriving at the final conclusion in most cases given the usual limitations of any single modality. Other associated cardiac anomalies have been reported as per literature review. In case of congenital abLAA, anticoagulation management in AF is not addressed in the current guidelines, and the final decision will depend on the treating physician's discretion.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

Table 1 Summary of reported cases with abLAA[1-4]

	Age, years	Gender	Associated lesions	Indication	Modality	Anticoagulation
Case 1*	65	F	Anomalous RCA	DCCV	TEE, CCT	DOAC
Case 2*	71	F	None mentioned	MVR	TEE, surgery	Warfarin
Case 3	77	F	None mentioned	PVI	TEE, CCT	Warfarin
Case 4	78	M	None mentioned	LAAO	TEE, CCT	P2Y12 inhibitor
Case 5	79	M	None mentioned	Biventricular implantable cardioverter-defibrillator	TEE	?
Case 6	54	F	None mentioned	PVI	TEE, CCT	Yes
Case 7	70	M	None mentioned	PVI	TEE, CCT, angio	Yes
Case 8	67	M	None mentioned	Maze surgical ablation procedure	CCT	?
Case 9	50	M	None mentioned	DCCV	TEE, CCT, cardiovascular magnetic resonance imaging	Yes
Case 10	80	M	None mentioned	PVI	TEE, CCT	DOAC
Case 11	73	F	None mentioned	DCCV	TEE, CCT	?
Case 12	79	F	None mentioned	LAAO	TEE, CCT, angio	Yes
Case 13	58	M	None mentioned	Cardiac source of emboli	TEE, CCT	?
Case 14	62	F	Single left-sided and 2 right-sided pulmonary veins	PVI	CCT, TEE	?
Case 15	58	F	None mentioned	PVI	CCT, angio	DOAC
Case 16	68	M	None mentioned	PVI	CCT, angio	Warfarin
Case 17	76	F	Persistent left SVC	PVI	CCT, TEE, angio	?
Case 18	62	M	None mentioned	PVI	CCT, TEE, angio	?
Case 19	52	?	Persistent left SVC draining into the distal coronary sinus	PVI	TEE, CCT	?
Case 20	60	M	None mentioned	PVI	TEE, CCT, angio	?
Case 21	57	M	None mentioned	Wolf-Parkinson-White	TEE, CCT	?
Case 22	3mo	M	Complex (single ventricle)	Death	Autopsy	N/A
Case 23	42	M	None mentioned	Chest pain	CCT	N/A

angio, Invasive LA angiography; *DCCV*, direct-current cardioversion; *DOAC*, direct oral anticoagulant; *F*, female; *M*, male; *N/A*, not applicable; *PVI*, pulmonary veins isolation; *SVC*, superior vena cava; *Yes*, anticoagulation strategy undefined; *?*, not included in report.
*This report.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

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