ESC European Society of Cardiology

European Heart Journal - Case Reports (2024) **8**, 1–5
https://doi.org/10.1093/ehjcr/ytae063

CASE REPORT
Medical management of arrhythmias

Weighing the risks and benefits of anticoagulation in atrial fibrillation: a case report of left atrial appendage absence in a patient referred for procedural occlusion

Roberto Lapetina Arroyo [1]*, Brian Monge Barrios[1], Hilton Franqui Rivera[1], Antonio Orraca Gotay[2], and Amanda Marrero Gonzalez[2]

[1]Department of Medicine, Cardiovascular Disease Division, University of Puerto Rico School of Medicine, P.° Dr. José Celso Barbosa, 00921 San Juan, Puerto Rico; and [2]Department of Medicine, Cardiovascular Disease Division, Pavia Santurce Hospital, San Juan, Puerto Rico

Received 10 June 2023; revised 20 January 2024; accepted 26 January 2024; online publish-ahead-of-print 30 January 2024

Background The absence of the left atrial appendage is an exceedingly rare structural variant that could have important implications for anticoagulation regimens in patients with atrial fibrillation.

Case summary We report the case of a 63-year-old Puerto Rican female with a history of hypertension, cerebral artery aneurysms, and type 2 diabetes mellitus who suffered multiple haemorrhagic strokes. The patient had never received anticoagulation therapy. During the indicated stroke work-up, the patient was found to have paroxysmal atrial fibrillation. Given the patients high risk for thrombo-embolism and contraindications to anticoagulation therapy, the patient was referred for left atrial appendage occlusion. Pre-procedural transoesophageal echocardiography failed to identify the left atrial appendage. Evaluation by way of cardiac computed tomography confirmed absence of the left atrial appendage. Left atrial appendage occlusion could not be carried out. The patient had been deemed being at high risk of bleeding, was not anticoagulated, and was instead closely followed. The patient has not had thrombo-embolic events nor has she experienced a haemorrhagic stroke recurrence at follow-up appointments.

Discussion To our knowledge, this is the first such case report that reports left atrial appendage absence in the setting of multiple haemorrhagic strokes. Given the rarity of the condition and lack of available guidelines, the most viable way to currently manage this patient population is on a case-to-case basis. However, we propose that absence of the left atrial appendage could confer a decreased risk of thrombo-embolic phenomena in patients with atrial fibrillation.

Keywords Case report • Left atrial appendage occlusion • Atrial fibrillation • Absent left atrial appendage • Anticoagulation • Stroke

ESC curriculum 2.1 Imaging modalities • 2.4 Cardiac computed tomography • 5.3 Atrial fibrillation • 8.6 Secondary prevention • 9.4 Thromboembolic venous disease

* Corresponding author. Tel: +787-758-2525, Email: roberto.lapetina@upr.edu
Handling Editor: Goksel Cinier
Peer-reviewers: Levent Pay; Kemal Emrecan Parsova
Compliance Editor: Franca Morselli
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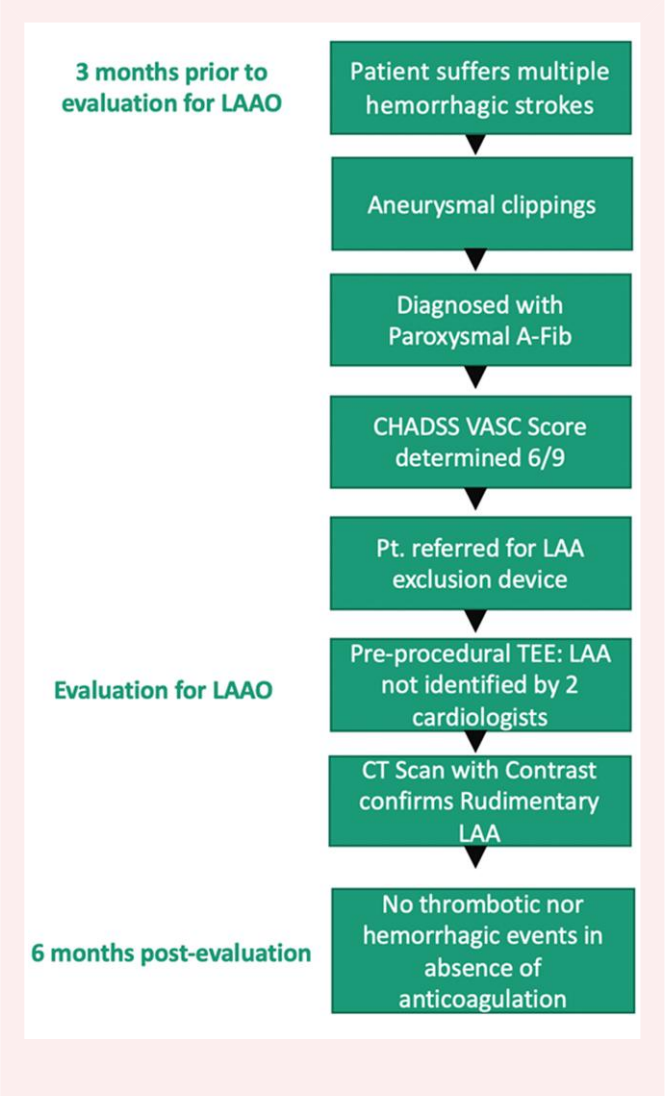
Learning points

- Congenital absence of the left atrial appendage (LAA) is an exceedingly rare anatomical variant that could impact anticoagulation regimens in patients with atrial fibrillation.
- The absence of the LAA may confer a decreased risk of thrombo-embolic events, and may represent a serendipitous finding in patients with atrial fibrillation.
- The risk of bleeding conferred by anticoagulation may outweigh the benefit of thrombo-embolic prophylaxis in patients with absent LAA and atrial fibrillation.

Introduction

Congenital absence of the left atrial appendage (LAA) is an extremely rare structural variant with largely unknown clinical implications. Knowledge of the condition is limited to a scarce number of case reports. Most of these cases describe LAA absence in the setting of concomitant atrial fibrillation.[1] Due to the paucity of available data, strategies for decreasing thrombo-embolic risk amongst atrial fibrillation patients with absent LAAs have not been clearly established.

Summary figure



Case presentation

A 63-year-old woman with a medical history of arterial hypertension and type 2 diabetes mellitus suffered multiple spontaneous haemorrhagic strokes. The patient had never received anticoagulation therapy. During her stroke work-up, the patient was found to have various cerebral aneurysms and paroxysmal atrial fibrillation was detected on ambulatory cardiac monitoring. The patient was subjected to surgical aneurysmal clipping. Following clipping, the patient was deemed at high risk of rebleeding and the neurosurgical team recommended absolute avoidance of anticoagulation. Given this patient's risk for thrombo-embolic events (*CHA2DS2-VASC* 6/9) and her poor candidacy for anticoagulation (*HAS-BLED 3/9*), the patient was referred to an electrophysiologist for insertion of an LAA occlusion device 3 months after her initial stroke episodes.

At the time of the electrophysiologist's evaluation, the patient was found to have only minimal aphasia without overt sensory or motor deficits. The patient's medication regimen consisted of lisinopril 10 mg daily, metformin 500 mg daily, and rosuvastatin 20 mg daily. The patient had never been treated with antiaggregant therapy. The pre-procedure Transeosphageal Echocardiography (TEE) failed to visualize the LAA. Both 2D and 3D TEE modalities were performed. TEE evidenced preserved ejection fraction, normal left atrial dimensions, mild mitral regurgitation, and no other congenital anomalies. The obtained TEE images were not available for the report. Insertion of LAA occlusion device could not be performed. Follow-up imaging with a contrast cardiac CT (CCT) showed an incidental rudimentary/diminutive LAA, within the spectrum of congenital absence of LAA. *Figures 1* and *2* are representative images from the CCT performed in this patient. *Figure 3* is a 3D multiple volume-rendered multiplanar 3D reconstructions of the heart in multiple anatomic views.

No changes were made to the patient's medication regimen following confirmation of LAA absence. The patient had not experienced thrombo-embolic events, nor haemorrhagic stroke recurrence at her 6 months follow-up visit with the electrophysiologist. Following this appointment, the patient received no additional follow-up care with an electrophysiologist. The patient established regular preventive care with a general cardiologist and is doing well.

Discussion

Classically, atrial fibrillation is thought to contribute to cardioembolic disease because inefficient atrial contraction foments poor blood mixing leading to atrial stasis.[2] It should be noted that atrial fibrillation burden has been suggested to positively correlate with atrial myopathy severity.[3] In turn, atrial myopathy is associated with atrial remodelling and endocardial dysfunction that may lead to cardioembolic stroke in the absence of fibrillating atria.[4] Recently, impaired intraventricular flow in the left ventricle has also been proposed as potentially underreported source of thrombogenesis.[5] Moreover, it is estimated that more than 90% of cardioembolic thrombi originate from the LAA in

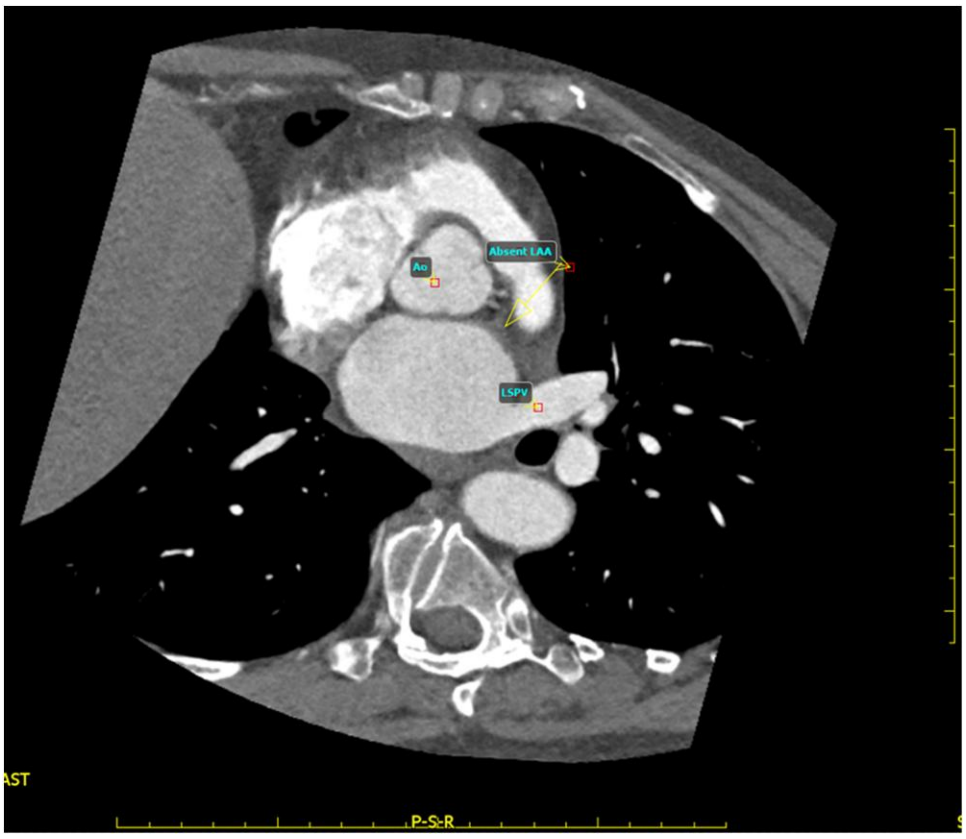


Figure 1 Retrospectively gated CCT arterial phase soft tissue window in an axial reconstruction at the expected level of the left atrial appendage, with non-visualization of the appendage.

patients with non-valvular atrial fibrillation.[6] The absence of the LAA may represent an important factor when weighing the risks and benefits of anticoagulation in atrial fibrillation.

Left atrial appendage occlusion devices have been shown to reduce the risk of ischaemic and bleeding events in patients with atrial fibrillation, as well as in high risk of embolic events who are not candidates for long-term anticoagulation therapy.[7] It is important to highlight that patients subjected to LAA occlusion procedures may not require post-procedure anticoagulation.[8] Given this reduction in risk with the procedural closure of the LAA, it would be reasonable to assume that congenital absence of this structure also confers a lower risk of cardioembolism.

The occurrence of haemorrhagic stroke in this patient was most likely due to a compound of multiple risk factors (T2DM, HTN, and cerebral aneurysms). However, it is important to note that intracranial haemorrhages are more prevalent in patients with atrial fibrillation due to their need for anticoagulation.[9] The atrial fibrillation patient presented in this case report was deemed a poor candidate for anticoagulation. We additionally suggest that, given her absent LAA, the risk represented by an anticoagulation regimen is unnecessary in the face of possibly marginal benefit from thrombo-embolic event prophylaxis. In the absence of a major bleeding episode, however, there would have been a stronger inclination to anticoagulate this patient. A reduced dose of a direct oral anticoagulant could have been considered as it has been shown to have a reduced probability of major bleeding when compared with higher dose regimens.[10]

Given the exceeding rareness of this anatomical variant, there is little consensus on anticoagulation regimens in this patient population. Elucidating this issue is difficult given that LAA absence has been documented, to our knowledge, in only 23 cases published in English, with notable clinical heterogeneity between reports.[1] However, it is important to note that no thrombo-embolic events have been reported in patients with absence of the LAA and concomitant atrial fibrillation.[1] Another important consideration pertains accurate diagnosis of LAA absence. Left atrial appendage absence is a benign condition that should be differentiated on TEE from flush appendage thrombus, anatomical variants, and obscuration from poor echocardiographic views.[11] Evaluation with a multimodality imaging using a combination of 2D/3D TEE followed by CCT has been the most widely employed approach.[1]

Currently, the most feasible alternative is to manage this unique patient population on a case-to-case basis. The risk reduction conferred by LAA occlusion procedures supports the idea that this congenital structural variation could be a serendipitous finding in atrial fibrillation patients. However, higher-tier studies are required to attest to the strength of this association. The most recent ESC Clinical Practice Guidelines for the management of atrial fibrillation delineate management strategies for various specific clinical settings,[12] but do not address the subset of patients found to have LAA absence. If a substantial body of literature is generated, we believe it is plausible that these patients could be included in future guidelines as a low risk atrial fibrillation subpopulation.

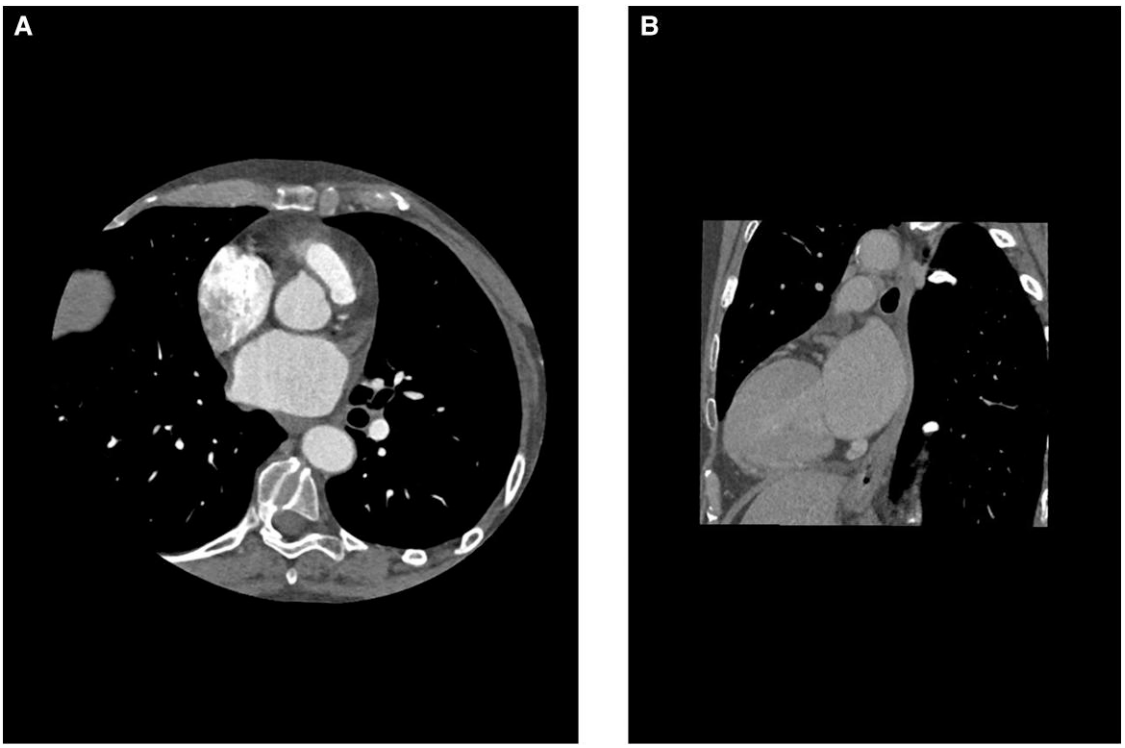


Figure 2 (*A* and *B*) Retrospectively gated CCT in soft tissue window delayed views in axial reconstruction (*A*) and sagittal reconstruction (*B*); there is a soft tissue density at the expected location of the left atrial appendage, with non-visualization of the appendage and no contrast opacification.

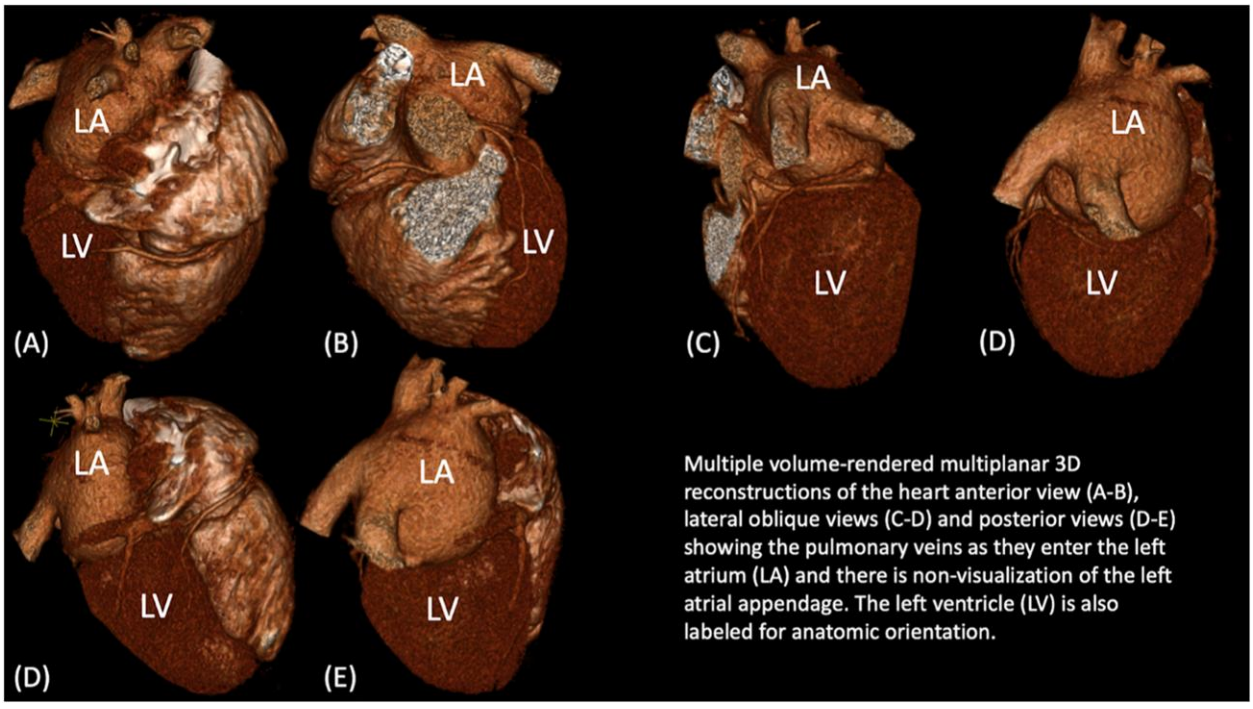
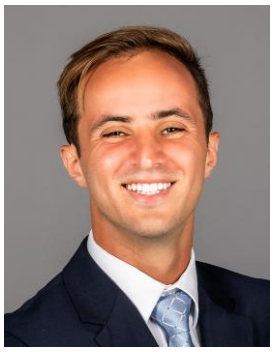


Figure 3 (*A–E*) Multiple volume-rendered multiplanar 3D reconstructions of the heart anterior view (*A* and *B*), lateral oblique views (*C* and *D*) and posterior views (*D* and *E*) showing the pulmonary veins as they enter the left atrium (LA) and there is non-visualization of the left atrial appendage. The left ventricle (LV) is also labelled for anatomic orientation.

Conclusion

Congenitally absent LAA is an exceedingly rare condition with important implications in patient management. Patients found to have LAA absence could have a lower risk for thrombo-embolic phenomena in the setting of atrial fibrillation. The risk of bleeding represented by anticoagulation therapy in these patients could outweigh the potential benefit provided from cardioembolic event prophylaxis.

Lead author biography



Roberto Lapetina Arroyo was born in Mayagüez, Puerto Rico on 6 March 1998. He is currently a fourth-year medical student from the University of Puerto Rico—School of Medicine. He will proximally be training to become an internal medicine physician, and has a strong interest in cardiology.

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: This work was not supported by any institution, public or private.

Data availability

The data underlying this article are available in the article and in its online supplementary material.

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