CLINICAL IMAGE

Navigating rare complex heart anatomy: percutaneous left atrial appendage closure in a patient with triatrial heart

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Triatrial heart, also known as cor triatriatum, is a rare congenital cardiac malformation with prevalence around 0.1% of diagnosed cases. It is characterized by a divided right or left atrium, resulting in 3 atrial chambers. It can occur as an isolated malformation or can be associated with different congenital cardiac anomalies. In this anatomic anomaly, the endocardium and fibromuscular tissue form a membrane within the left atrium (LA) (cor triatriatum sinister [CTS]) or the right atrium (cor triatriatum dexter). Classification based on the number and dimension of fenestrations in the fibromuscular membrane differentiates 3 triatrium heart anatomies. In the first category, the 2 atrial chambers are completely separated, in the second category, the intra-atrial membrane has 1 or several small fenestrations, and in the third group, the accessory chamber communicates widely with the true atrium through a single large fenestration. The malformation may be asymptomatic or may cause blood flow obstruction of varying degree, mitral valve stenosis, and pulmonary vein anomalies leading to symptomatic heart failure.[1-3]

A 76-year-old man with permanent atrial fibrillation (CHA_2DS_2VASc, 3; HAS-BLED, 3) was referred for an uncommon procedure of percutaneous LA appendage occlusion. The patient had a history of severe gastrointestinal bleeding due to stomach ulcers and treatment with non–vitamin K antagonist oral anticoagulants. To prevent bleeding episodes, he was permanently on enoxaparin.

Preprocedural transesophageal echocardiography revealed a fibromuscular membrane dividing the LA. The divided atrium consisted of the true LA (the chamber connected with the mitral valve and the atrial appendage) and the accessory LA (ALA; the chamber connected with the pulmonary veins) (FIGURE 1A and 1B). In this CTS variant, the membrane was partially fenestrated, providing communication between the true and accessory chambers, and therefore did not obstruct blood flow (FIGURE 1C). The membrane was not visible on transthoracic echocardiography.

Due to anatomic difficulties resulting from the presence of the fibromuscular membrane, trans-septal puncture required several attempts. The main concern was to avoid the membrane disruption. Safe puncture avoiding the atrial membrane contact was achieved using a Swartz guiding introducer (Abbott, Plymouth, Massachusetts, United States) and a BRK-1 transseptal needle (St. Jude Medical, Minnetonka, Minnesota, United States) (FIGURE 1D). Then, the guiding introducer was replaced with a Watchman double curved sheath (Boston Scientific, Marlborough, Massachusetts, United States), which enabled maneuvering and advancing the sheath to the ALA without a risk of the CTS membrane disruption (FIGURE 1E). In the final step, the Watchman Flex 35 mm occluder (Boston Scientific) was deployed in an optimal position (FIGURE 1F).

CTS presents a significant challenge for interventions that involve the LA. A limited number of procedures involving CTS can be found in the literature (3 cases of LA appendage closure and 4 cases of atrial fibrillation ablation).[4,5] The main difficulties are due to the anatomic abnormalities related to the condition, including the fibromuscular membrane. The main concern during the procedure is to avoid the membrane disruption. CTS complicates navigation during the procedure, making it considerably more difficult.[4]

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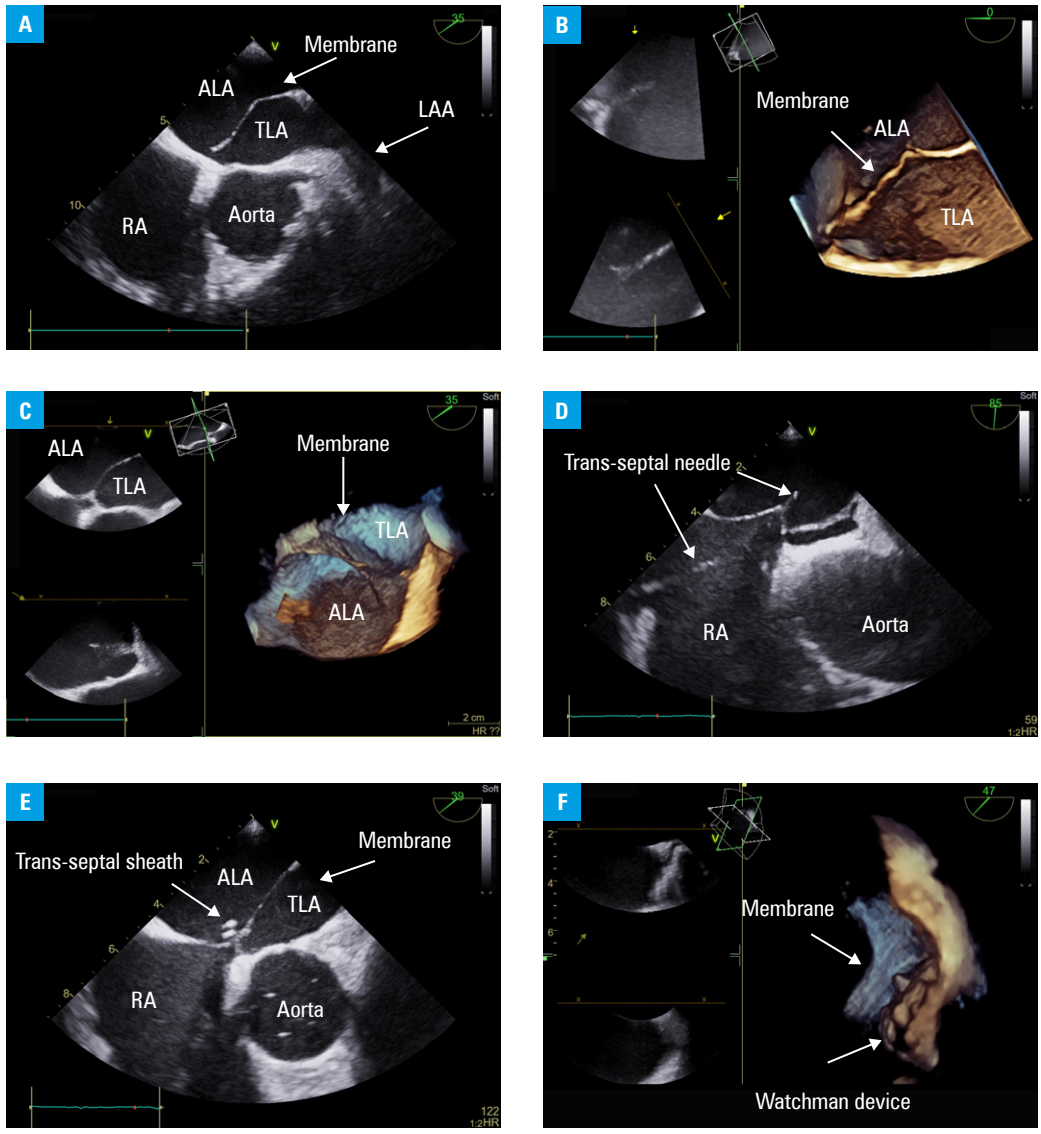

FIGURE 1 Transesophageal echocardiography during percutaneous left atrial appendage (LAA) closure; **A**–**C** – a membrane dividing the left atrium into the true left atrium (TLA) and the accessory left atrium (ALA); **D** – high-risk trans-septal puncture; **E** – trans-septal sheath safely bypassing the fibromuscular membrane; **F** – Watchman device in a correct position close to the fibromuscular membrane
Abbreviations: RA, right atrium

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