

Left atrial appendage occlusion in a patient with hereditary hemorrhagic telangiectasia and atrial fibrillation – a therapeutic option worth considering

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Hereditary hemorrhagic telangiectasia (HHT) is a dominantly inherited genetic disorder of blood vessel development characterized by epistaxis, mucocutaneous telangiectasias and arteriovenous malformations (AVMs). Recently, it was shown that HHT patients have a predilection to develop atrial fibrillation [1], which is probably due to frequent systemic AVMs, iron deficiency anemia and hypoxemia resulting in hyperdynamic circulation. Those patients cannot benefit from oral anticoagulant (OAC) therapy for stroke prevention, because of the unacceptably high bleeding risk [2]. Therefore left atrial appendage occlusion (LAAO) may be warranted in HHT patients with AF and high thromboembolic risk.

A 65-year-old woman with HHT was referred to our department for consideration of LAAO. The patient had developed gastric AVMs with several episodes of bleeding, which required repeated endoscopic treatment and blood transfusions. On the other hand, she had developed chronic AF with high thromboembolic risk according to CHA₂DS₂-VASc (her score was 3, giving her an estimated annual risk of stroke of 3.2%). The patient could not tolerate long-term OAC therapy. On that basis, LAAO was proposed by the Heart Team. Transesophageal echocardiography (TEE) and computed tomography angiography were performed to obtain measurements of the left atrial appendage (LAA) for occluder device sizing, as well as to exclude thrombus in the LAA (Figures 1 A, B). The procedure was conducted under general anesthesia under TEE guidance. Femoral vein access was obtained. The interatrial septum was punctured with a dedicated needle and a transseptal sheath was inserted into the left atrium. Subsequently, a transseptal sheath was replaced over a stiff wire by a delivery sheath and an Amplatzer AMULET (St Jude Medical) occluder was deployed in the LAA (Figures 1 C–E). The patient was awakened and transferred to the Cardiac Intensive Care Unit for 24-hour surveillance. Recovery was uneventful and the patient was discharged on the 5th day after LAAO. According to the standard protocol dual antiplatelet therapy was commenced, which consisted of aspirin 75 mg daily and clopidogrel 75 mg daily. Three months later TEE confirmed proper position of the device (Figure 1 F). At that time clopidogrel was laid off. As low-dose aspirin therapy was well tolerated, the patient stayed with it.

According to European Society of Cardiology Guidelines, LAAO may be considered in patients with AF and contraindications for long-term OAC

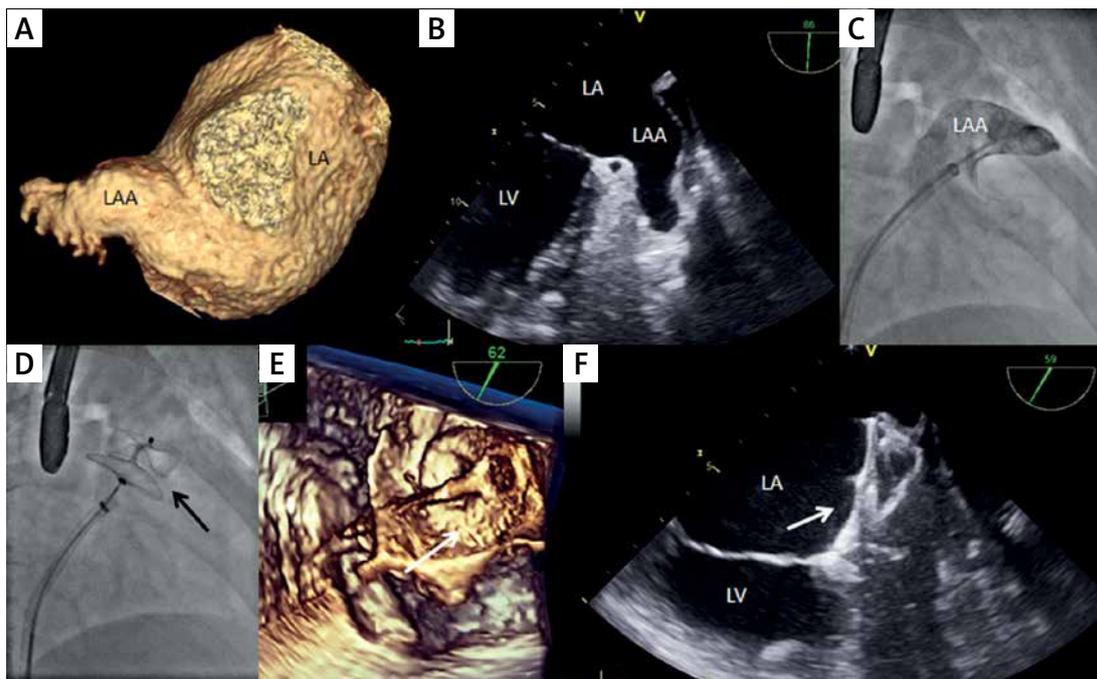


Figure 1. A – computed tomography angiography: 3D volume rendering of the left atrium (LA) and the left atrial appendage (LAA); B – transesophageal echocardiography: 2D imaging of the left atrium (LA), the LAA and the left ventricle (LV); C – fluoroscopy: LAA angiography; D – fluoroscopy: Amplatzer AMULET (arrow) deployed in the left atrial appendage, but still attached to a delivery system; E – transesophageal echocardiography: 3D imaging of Amplatzer AMULET (arrow) deployed in the left atrial appendage, but still attached to a delivery system – en face view; F – transesophageal echocardiography: 2D imaging of the Amplatzer AMULET (arrow) implanted in the left atrial appendage – 3-month follow-up

therapy [3]. To date, only a few LAAO in HHT patients have been reported [4]. Due to the fact that HHT patients tolerate antiplatelet agents better than OAC [2], short-term dual antiplatelet therapy required after Amplatzer AMULET implantation can usually be completed.

Conflict of interest

The authors declare no conflict of interest.

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