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Left atrial appendage closure in patient with hemophilia C. An alternative or the only way of antithrombotic treatment in patient with rare bleeding disorder?

Short title: Left atrial appendage closure in patient with hemophilia C

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Atrial fibrillation (AF) prevalence in the adult population is estimated between 2% and 4%. AF is increasing the risk of stroke approximately 5 times. To prevent stroke we used anticoagulant therapy, which is recommended in patients with a CHA₂DS₂-VASc score of ≥ 2 in men or ≥ 3 in women and should be considered in patients with a CHA₂DS₂-VASc score of 1 in men or 2 in women. In patients with contraindications to chronic anticoagulant therapy, an alternative is percutaneous left atrial appendage closure (LAAC) [1, 2].

Below we present a description of left atrial appendage closure procedure in a patient with contraindications for long-term anticoagulant treatment due to congenital hemophilia C.

A 72-year-old man with hemophilia C (hereditary bleeding disorder characterized by deficiency in factor XI) and permanent atrial fibrillation (AF), without any kind of antithrombotic therapy was referred to our center by his cardiologist. His medical history included hypertension, ventricular arrhythmias, prostate cancer treated by prostatectomy 6 years ago and post-traumatic subdural hematoma 16 years ago.

The CHA₂DS₂-VASc score was estimated for 2 and his HAS-BLED was 3. After evaluation his case was presented at the Heart-Team consultation, and patient was qualified to LAAC.

Transesophageal echocardiography (TEE) revealed a thrombus of diameter 0.8 cm (Figure 1A) in left atrial appendage (LAA). Laboratory tests showed increase activated partial thromboplastin time (APTT) to 82.9 sec with correct international normalized ratio (INR) — 1.4. The LAAC procedure was postponed. The patient was consulted by a hematologist, who disqualified him from any kind of antithrombotic treatment. Patient was discharged from the hospital and next evaluation was scheduled after 2 months to check the presence of the thrombus.

After 2 months TEE was performed, which showed a presence of spontaneous contrast in left atrial appendage, which on the bottom had gelatinous consistency and was on the verge of clotting (Figure 1B). The APTT was also increased this time, and INR level was normal. Factor XI level was evaluated at 1.4%. It was decided to perform the procedure shortly without contrast injection for device positioning in left appendage (navigating with TEE only).

After 7 days we admitted patient again. This time during TEE no thrombus was found in LAA (Figure 1C). Only a presence of spontaneous contrast was noticed, which was not contraindication to LAAC procedure. It was decided to perform procedure. Due to heritage factor XI deficiency, the patient was consulted by a hematologist. According to hematologist's recommendation patient received 6 units of fresh frozen plasma (FFP) within 12 hours before the planned procedure. After transfusion factor XI level was 23.6% and APTT was 37.1 seconds and 2 more units of FFP was transfused in the operating room just before the procedure. After that preparation patient underwent successful percutaneous left atrial appendage closure using a 35mm Watchman FLX device (Figure 1D–F). The procedure went without complications. In the postoperative period the next 2 units of FFP was transfused. On the first day after LAAC level of factor XI was 28.9% and APTT was 26.3 seconds. No hemorrhagic complications during further hospitalization occurred. On 4th day after LAAC TEE confirmed the correct position of closure device. The same day patient was discharged from the hospital in stable condition.

To our knowledge it is the first reported case of percutaneous left atrial appendage closure in patient with hemophilia C. We found only 17 described cases of LAAC in patients with hemophilia (15 hemophilia A, 2 hemophilia B), none in Poland [3–5]. In our opinion LAAC after appropriate preparation is safe strategy in patient with hemophilia C and AF.

Article information

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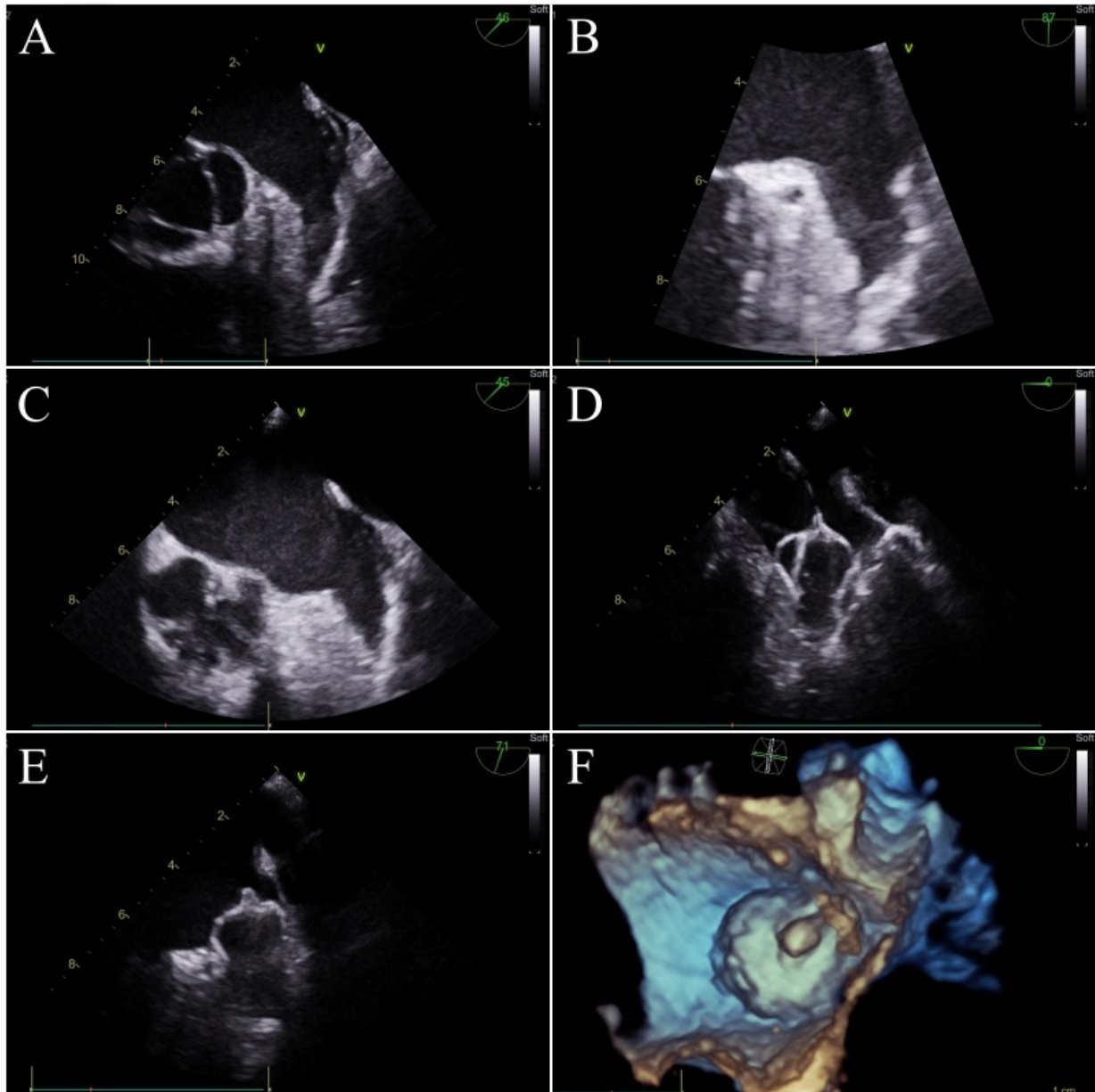


Figure 1. **A.** Thrombus of diameter 0.8 cm in LAA. **B.** Spontaneous contrast in LAA, which on the bottom has gelatinous consistency and is on the verge of clotting. **C.** Spontaneous contrast in LAA. **D.** 35 mm Watchman FLX device in LAA, before released, intraprocedural 2D TEE. **E.** 35 mm Watchman FLX device in LAA after released, intraprocedural 2D TEE. **F.** 35 mm Watchman FLX device in LAA after released, intraprocedural 3D TEE
 Abbreviations: 3D, 3 dimensions; LAA, left atrial appendage; TEE, transesophageal echocardiography