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# CRYOBALLOON ISOLATION OF PULMONARY VEIN IN PATIENT WITH PERSISTENT LEFT SUPERIOR VENA CAVA AND ATRESIA OF THE RIGHT SUPERIOR VENA CAVA: CASE REPORT

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*A clinical case of cryoballoon ablation of pulmonary veins in a patient with atrial fibrillation with congenital heart disease: persistent left superior vena cava, atresia of the superior vena cava. The methods of investigation at the pre-operative stage are described, which allow to plan the operation in advance, taking into account the peculiarities of the confluence of the main veins into the heart, and also some technical features of cryoballoon ablation surgery are emphasized.*

**Key words:** atrial fibrillation; cryoballoon ablation; persistent left superior vena cava; atresia of the superior vena cava

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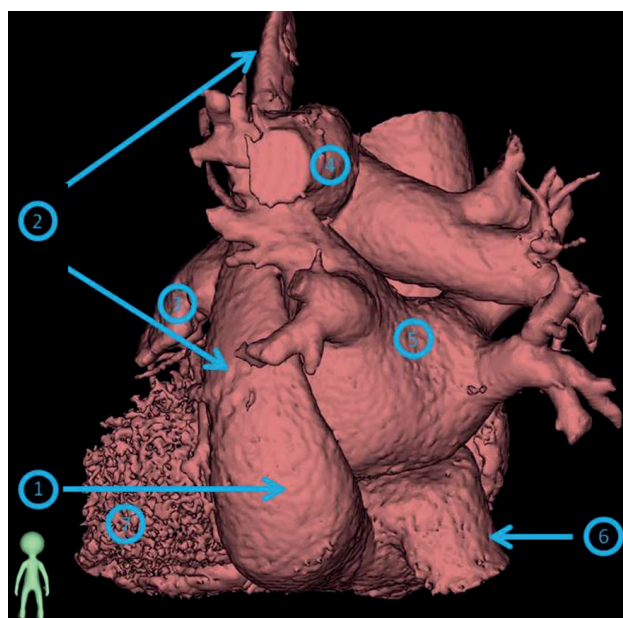
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Atrial fibrillation (AF) is the most common arrhythmia in the global population. «Gold standard» of catheter-based treatment is pulmonary vein isolation (PVI), for which radiofrequency and cryoballoon ablation (CBA) techniques are used. For this procedure, transvenous access to the right atrium (RA) is performed, followed by transseptal puncture to guide ablation catheters into the left atrium (LA).

When anomalies of the trunk vessels enter the heart, the standard surgical protocol requires modifications, leading to non-standard situations and complicating the PVI procedure. The most frequent variant of anomalies in the vessels providing inflow to the heart is the persistent left superior vena cava (PLSVC), which occurs in 2-5% of congenital heart disease cases. According to a systematic review and meta-analysis on prenatal echocardiography (Echo), this abnormality occurs in 0.2-0.6% in the general population [2]. It is worth noting that such a congenital heart defect is not clinically manifested in any way and is often an incidental finding during examination.

PLSVC formation is a consequence of impaired obliteration of the left anterior cardiac vein, which drains into the right atrium via the great cardiac vein and coronary sinus. The coronary sinus is dilated in such cases. The left vena cava is most often the accessory vein. In such cases, the width of the right and left superior vena cava may differ: when there is communication between them, the main right superior vena cava is often wider, whereas in the absence of communication, both veins are usually of the same width. In addition to no obliteration of the left anterior cardiac vein, right-sid-

ed obliteration of this vein may be observed. The result of this development would be atresia of the main right SVC while preserving the left one flowing into the coronary sinus (CS). The frequency of detection of a single left superior vena cava is about 2% [3].

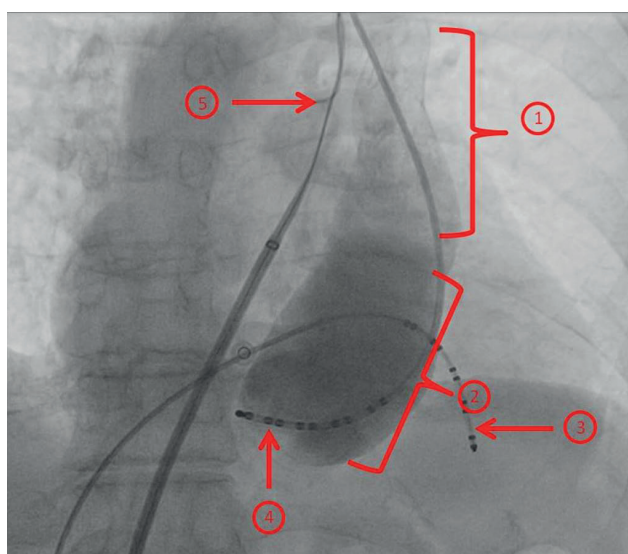


**Fig. 1. Three-dimensional model of heart chambers based on multispiral computed tomography with contrast. Note: 1 - dilated coronary sinus, 2 - left persistent superior vena cava, 3 - left atrial appendage, 4 - left pulmonary artery, 5 - left atrium, 6 - inferior vena cava.**

### Clinical case

A 66-year-old patient diagnosed with persistent form of AF was admitted to the cardiac rhythm disturbance department for catheter-based PVI. The patient complained of heart palpitations accompanied by dyspnea, weakness, dizziness. AF was first reported at the age of 48 years and had a paroxysmal form. After suffering a covid-associated viral infection, the arrhythmia became chronic. No attempts to restore sinus rhythm were made at the place of residence. As drug therapy, the patient is continuously taking metoprolol 25 mg once daily, amiodarone 200 mg once daily, and apixaban 5 mg twice daily. At the outpatient stage, the patient underwent all standard laboratory and instrumental investigations. During Echo in our center, enlarged inferior vena cava (23 mm and 11 mm on inspiration) and CS (17 mm) were noted. On the eve of surgery, the patient underwent contrast-enhanced multispiral computed tomography. Based on this, we performed an anatomic reconstruction of the heart chambers and identified atresia of the SVC and PLSVC, which flowed into the CS and passed between the auricle of the LA and the left PV. Using the software for processing multispiral computed tomography with contrast and three-dimensional reconstruction of the heart chambers, a model of the RA and LA with the main vessels flowing into them was constructed (Fig. 1). Before surgery, the patient underwent a transesophageal Echo, which excluded LA auricular thrombosis.

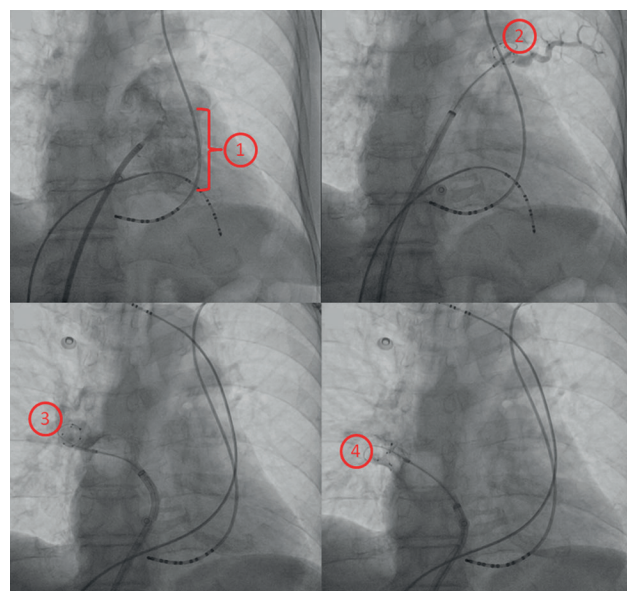
CBA was chosen as the method of catheter-based PVI. The procedure was performed under total intravenous anesthesia using fentanyl 0.05% and propofol 1% at a dose of 5 mg/kg/h. Venous access was performed using the Seldinger method. A ten-pole electrophysiology catheter was placed into the CS through the subclavian vein on the left side via the PLSVC. We performed contrast-enhanced CS, in which an increase in its size was noted (Fig. 2). A ten-pole guided electrophysiology catheter was placed through the femoral vein on the left side into the right ventricle. A Swartz intraducer was



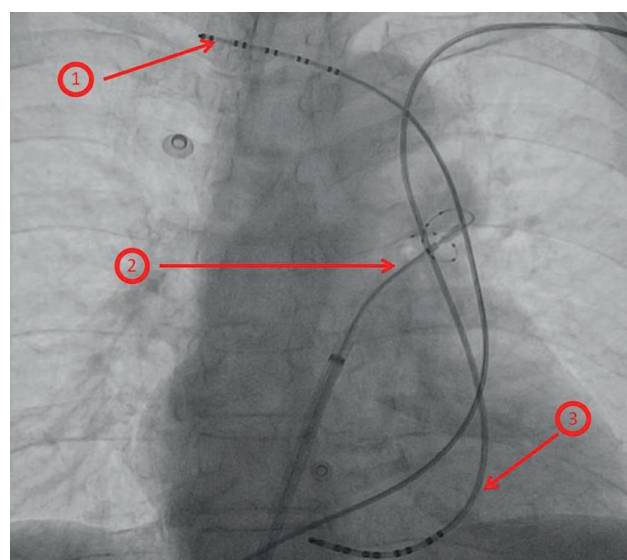
**Fig. 2. Coronary sinus contrast. Anteroposterior projection. Note: 1 - left persistent superior vena cava, 2 - dilated coronary sinus, 3 - electrode in the right ventricle, 4 - electrode in the coronary sinus.**

placed through the right femoral vein into the right atrium, through which the transseptal puncture needle was guided. Due to atresia of the superior vena cava, the long conductor was placed under the roof of the RA. Atrial septal puncture was performed under radiologic control and had a number of features.

First of all, the classic transseptal needle jump from the superior vena cava to the RA was absent. Also, CS dilatation altered the true boundaries of the atrioventricular sulcus. Heparin was then administered based on the



**Fig. 3. Radiologic picture of the left atrium and pulmonary veins. Anteroposterior projection. Note: 1 - left atrial contrast, 2 - occlusion and angiography of the left superior pulmonary vein, 3 - occlusion and angiography of the right superior pulmonary vein, 4 - occlusion and angiography of the right inferior pulmonary vein.**



**Fig. 4. Radiologic picture when a diaphragmatic nerve stimulation electrode is passed through the coronary sinus and left persistent superior vena cava. Anteroposterior projection. Note: 1 - diagnostic guided electrode in the region of the right subclavian vein, 2 - electrode in the coronary sinus, 3 - cryoballoon catheter in the upper pulmonary vein.**



patient's weight at a rate of 100 units per 1 kg of weight. After that, a Swartz introducer was inserted into the LA via a long conduit and then replaced with a guided delivery system, through which a cryoballoon catheter with a circular diagnostic electrode was guided. Using the Astrocad navigation system, anatomic reconstruction of the LA and PV was performed. Arctic Front Advance Pro was used as the cryoballoon catheter. Cryo-interventions lasting 180 seconds were performed according to the standard protocol. We observed no technical difficulties with the positioning of the Achieve eight-pole diagnostic electrode in the PV, as well as no problems with their occlusion with the cryoballoon (Fig. 3). For right PV exposures, a guided diagnostic catheter was guided through the coronary sinus into the region of the right subclavian vein to stimulate the right diaphragmatic nerve (Figure 4). At the end of the procedure, AF persisted and sinus rhythm was restored with 200 J electric pulse therapy. At the time of the patient's discharge, preservation of sinus rhythm was noted.

### DISCUSSION OF FINDINGS

According to the latest guidelines for the treatment of AF, PVI is recommended for all patients in the first stage, regardless of the form of arrhythmia [4]. Catheter-based interventions for cardiac rhythm in patients with PLSVC have been described in the literature [5-8]. However, there are limited publications on PV CBA in patients with PLSVC and SVC atresia, and all of them are described by foreign authors [9, 10].

The presence of this anomaly of venous blood flowing into the heart from the upper body suggests an additional risk of complications during PV CBA. First of all, the transseptal puncture procedure becomes non-

standard. Since there is no first jump of the transseptal needle when it is lowered to the atrial septum and the atrioventricular sulcus is disturbed due to dilated CS, there may be technical difficulties in providing access to the PV, as well as an increased risk of needle exit into the pericardium and cardiac tamponade. A number of authors recommend performing this procedure under echocardiographic control. Both transesophageal and intracardiac transducers for echocardiography can be used [11].

Also atresia of the SVC makes stimulation of the diaphragmatic nerve more difficult. Some authors, having encountered an anomalous cardiac structure with PLSVC and atresia of the SVC, controlled only the amplitude of diaphragm contraction during right PV exposures [12]. In our case, it was possible to achieve stimulation of the right diaphragmatic nerve using a guided diagnostic catheter.

### CONCLUSION

In presenting this clinical case, we would like to emphasize a number of important aspects. First, a complete preoperative examination of patients can detect cardiac abnormalities even preoperatively. Subsequently, this makes it possible to anticipate a number of technical difficulties and to think in advance about how to solve them. Secondly, we described a method of stimulation of the diaphragmatic nerve in SVC atresia. In this situation, we recommend the use of a guided diagnostic catheter, which can be passed through the CS and PLSVC to the area of the left subclavian vein. Third, we would like to point out that the standard transseptal puncture protocol undergoes significant modification due to SVC atresia and dilated CS. In this situation, it is advisable to perform atrial septal puncture under the control of transesophageal or intracardiac echocardiography.

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