DOI: 10.15825/1995-1191-2020-4-168-172

HEART TRANSPLANT IN A PATIENT WITH PERSISTENT LEFT SUPERIOR VENA CAVA

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Objective: to present our own experience of heart transplantation in a patient with persistent left superior vena cava (PLSVC). A clinical case of successful orthotopic heart transplantation using the biatrial technique in a patient with PLSVC drainage into the right atrium is presented. The clinical effect achieved as a result of the treatment fully justifies the chosen surgical tactics and allows us to recommend the proposed tactics for treatment of such a rare anomaly. **Conclusion.** The clinical effect achieved as a result of the treatment fully justifies the chosen surgical tactics and allows us to recommend the proposed tactics for treatment of such a rare anomaly.

Keywords: congenital malformation, cardiovascular system, persistent left superior vena cava, heart transplantation.

Persistent left superior vena cava (PLSVC) is a rare congenital vascular anomaly. The incidence is 0.3–0.5% of the general population with a normal heart and in 4.5% of people with congenital heart disease [1]. In most cases (80–90%), PLSVC is associated with a right superior vena cava (SVC) [2], and may also be accompanied by other cardiac anomalies, such as abnormal pulmonary vein connections, coarctation of the aorta, tetralogy of Fallot, transposition of the great vessels, and patent ductus arteriosus (PDA) [3]. At the same time, cardiac arrhythmias are observed.

PLSVC usually flows into the right atrium (in 80–92%) through the dilated coronary sinus [5], but in 10–20% of cases, it flows into the left atrium [7]. The PLSVC can drain directly into the left atrium or coronary sinus, causing a right-to-left cardiac discharge. Most patients with PLSVC are asymptomatic. Only patients with abnormal drainage and right-to-left discharge have a clinical picture. Abnormal venous return via PLSVC can cause cardiac arrhythmias, decreased exercise tolerance, progressive fatigue, chest discomfort, palpitations, fainting, or cyanosis [6].

The presence of PLSVC is important for central venous catheter placement, pacemaker implantation, and cardiac catheterization. PLSVC is also a relative contraindication for retrograde cardioplegia during cardiac surgery [6].

To date, the choice of optimal tactics and the extent of the proposed surgical intervention remains an open question. In this paper, we present the experience of heart transplantation in a recipient with an abnormal left SVC performed by surgeons at the cardiac surgery department No. 1.

CASE STUDY

Patient K., 52 years old, has considered himself ill since 2004, when, without a previous coronary history, he suffered inferior wall acute myocardial infarction (AMI) with the development of early post-infarction angina pectoris. Coronary artery stenting was performed on the background of acute coronary syndrome. In 2008, examination revealed thrombophilia, anticoagulants were prescribed. In 2010, he underwent coronary artery bypass grafting of the right coronary artery and the circumflex branch off the left coronary artery. According to the Holter monitoring data, paroxysmal supraventricular tachycardia (PSVT) was registered. In 2014, a dual-chamber cardioverter-defibrillator was implanted. In 2018, battery depletion was detected and a dual-chamber cardioverter-defibrillator was reimplanted. On July 10, 2019, electrophysiological examination and radiofrequency ablation of the right isthmus and ectopic foci were performed. Control coronarography revealed shunt stenosis, balloon angioplasty with shunt stenting were performed on December 18, 2019 to the circumflex branch. From February 2020, there was an increased frequency of tachycardia attacks, cardioverter defibrillator activation, numerous syncopal episodes. The patient was admitted at the cardiology department of the Shumakov National Medical Research Center of Transplantology and Artificial Organs for examination under the program for potential heart recipients with a ***clinical diagnosis:*** *ischemic cardiomyopathy. Coronary artery disease: Stenosing coronary sclerosis. Postinfarction cardiosclerosis (2004, 2008). Operation: balloon-assisted vasodilation with right coronary artery stenting, anterior interventricular branch, circumflex artery bran-*

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ches of 2005, 2006, 2007. Left marginal artery, May 28, 2018. Coronary artery bypass grafting of the right coronary artery and circumflex artery in 2010. Shunt restenosis to circumflex artery 80%. Balloon angioplasty with stenting. Coronary artery bypass grafting of the right coronary artery on December 18, 2019.

Main complication: *Arrhythmias: Paroxysmal supraventricular tachycardia. Cardioverter defibrillator implantation in 2014 Battery depletion and reimplantation of cardioverter defibrillator in 2018. Radiofrequency ablation of the right isthmus and ectopic foci on July 10, 2029. Relative mitral regurgitation. Relative tricuspid valve insufficiency. Chronic heart failure, stage 2A circulatory insufficiency, NYHA Class III heart failure.*

Concomitants: *Multifocal atherosclerosis with coronary and carotid artery lesions. Consequences of ischemic stroke of April 2015. Thrombophilia. Chronic cholecystitis. Multiple small cysts in the liver parenchyma. Diffuse nodular goiter. Iodine-induced subclinical thyrotoxicosis.*

Objectively: *a state of moderate severity. Skin and visible mucous membranes of physiological color. There were no peripheral edemas. In the lungs, breathing was rigid, occurring in all parts, no wheezing. Respiratory rate – 17 breaths per min. Examination showed that the boundaries of relative cardiac dullness were not expanded, the cardiac impulse was not determined, the apical impulse was determined in the fifth intercostal space along the left midclavicular line. Clear heart sounds, regular rhythm, a single extrasystole. Heart rate was 72 beats/min. Blood pressure 100/70 mm Hg. The abdomen was soft and painless on palpation. The liver was not enlarged. Murphy's punch sign was negative. There were no dysuric disorders. There were no acute focal symptoms.*

General blood test, biochemical blood test, coagulogram without any peculiarities.

ECG at rest had normal sinus rhythm with a heart rate of 70/min. Myocardial scarring at the posterior diaphragmatic region of the left ventricle. Decreased blood supply in the scarring area, apical-lateral wall.

Frontal chest X-ray: moderate increase in pulmonary vascular pattern was noted.

Preoperative echocardiographic data: *Aorta: at the level of the annulus fibrosus 2.7 cm. Valsalva sinus 4.2 cm. Ascending aorta 3.9 cm. Left atrium: 3.9 cm (anteroposterior dimension); Left atrial volume 60 mL. Right atrium: Right atrial volume 48 mL. Right ventricle: 2.7 cm (anteroposterior dimension); Right ventricular anterior wall thickness 0.5 cm. Interventricular septum 1.2–1.3 cm. Left ventricular posterior wall 1.0 cm. End-diastolic volume 197 mL; End-systolic volume 123 mL; Stroke volume 54 mL. Ejection fraction 37%. Left ventricular local contractility: diffuse hypokinesis. Inferior wall akinesis, posterior part, interventricular septal dyskinesia. No pathological formations. Electrodes are in the right heart chambers. Valve apparatus: Aortic valve: 3-cusp leaflet: sealed with Pgr 9.2 mmHg. Regurgitation degree 1. Mitral valve: Leaflets: sealed, calcium at the base of the posterior valve of the MC. Regurgitation 1 degree. Tricuspid valve: no features of the leaflets. Regurgitation degree 1. Pulmonary artery: Leaflets: no particularities. Pulmonary artery trunk diameter 2.4 cm, Pgr 4.5 mmHg, degree 0–1 regurgitation. Systolic pressure 32 mmHg.*

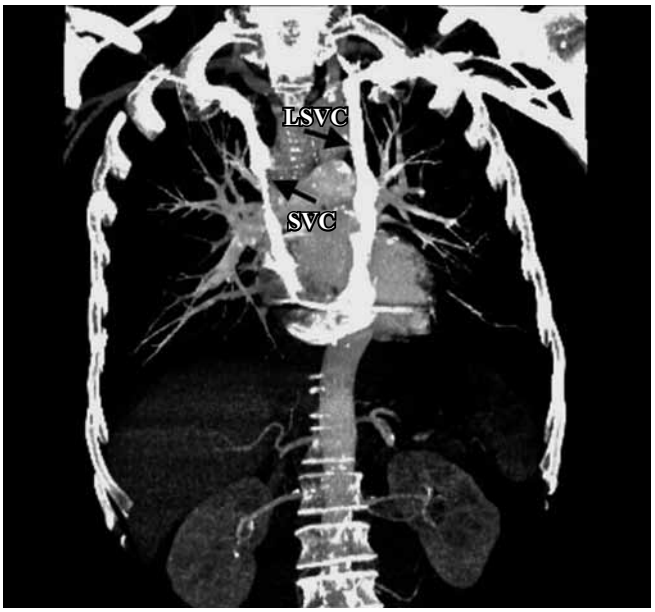

Fig. 1. Contrast-enhanced multidetector CT imaging of the heart. The right superior vena cava is typically located. Visualized is a persistent left superior vena cava, enveloping the left atrium along its posterior wall and flowing into the coronary sinus, while the transverse vein is not defined; SVC – superior vena cava. LSVC – left superior vena cava

OPERATION

Longitudinal median sternotomy. Cardiolysis of the aorta and the right heart was performed. The heart was enlarged. Cannulation of the aorta and vena cava. Artificial blood circulation was started, cardiolysis continued. The shunts from the aorta were ligated, stitched and cut off. The ascending aorta was clamped, the recipient's heart was excised with technical difficulties due to the pronounced adhesion process. PLSVC was revealed, which entered the pericardium at the left superior pulmonary vein level and flowed into the coronary sinus. The PLSVC was isolated from the adhesions, mobilized, and clamped with a turnstile. A left atrial cavity was formed. The cardiac graft was placed in the pericardial cavity. External cooling of the graft. The left atria of the donor and the recipient were anastomosed with a twisted suture, then the right atria of the donor and recipient. Given the long SVC stump of the donor heart, an end-to-end anastomosis was applied between the donor SVC stump and the recipient's PLSVC. The anastomosis was laid in the transverse sinus, behind the aorta and pulmonary artery. The donor and recipient aorta and the donor and

recipient pulmonary arteries were anastomosed with continuous twisted prolene suture 5-0. The clamp was removed from the aorta to prevent air embolism. Cardiac activity was restored after defibrillation. Drainage was removed from the left ventricular cavity. Cardiotonic support was selected. On the background of inotropic drugs, artificial blood circulation was completed routinely. Decannulation. Thorough hemostasis. The anterior mediastinal cavity was drained. The pericardial and the enclosed left pleural cavity were drained with a single drain. A pacemaker electrode was sutured to the right ventricle. Implantable cardioverter defibrillator with 2 electrodes was removed from a separate incision in the right subclavian region. The sternum was sutured with 6 wire sutures. Layer-by-layer suturing of the surgical wound and a wound in the right subclavian region.

Postoperative course: *The patient was extubated on postoperative day 1. The early postoperative period was accompanied by myocardial insufficiency requiring inotropic support, metabolic disorders requiring renal replacement therapy. On postoperative day 9 against the background of regressive myocardial and respiratory failure, and regression of metabolic disorders, the patient was transferred to the department. The further postoperative period was uneventful. He was discharged on day 23 in a satisfactory condition.*

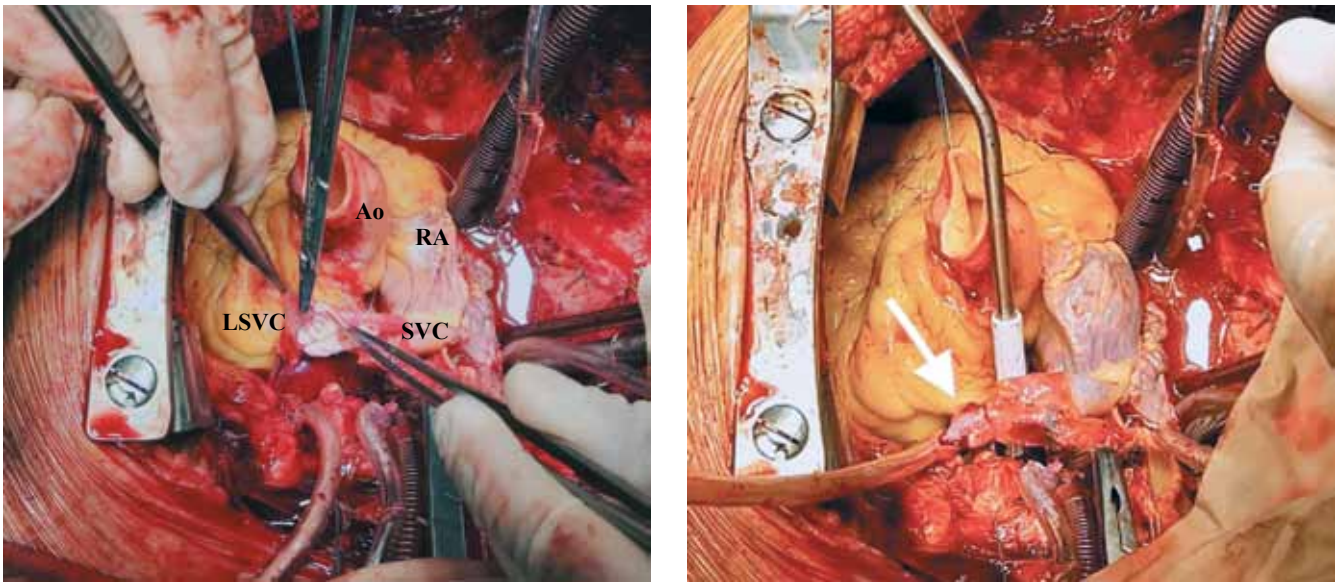

Fig. 2. An end-to-end anastomosis of the donor SVC to the recipient persistent LSVC was performed. The anastomosis is laid in the transverse sinus, behind the aorta and pulmonary artery. The white arrow indicates the patency and tightness of the anastomosis. Ao – aorta; RA – right atrium; SVC – donor superior vena cava; LSVC – persistent recipient left superior vena cava

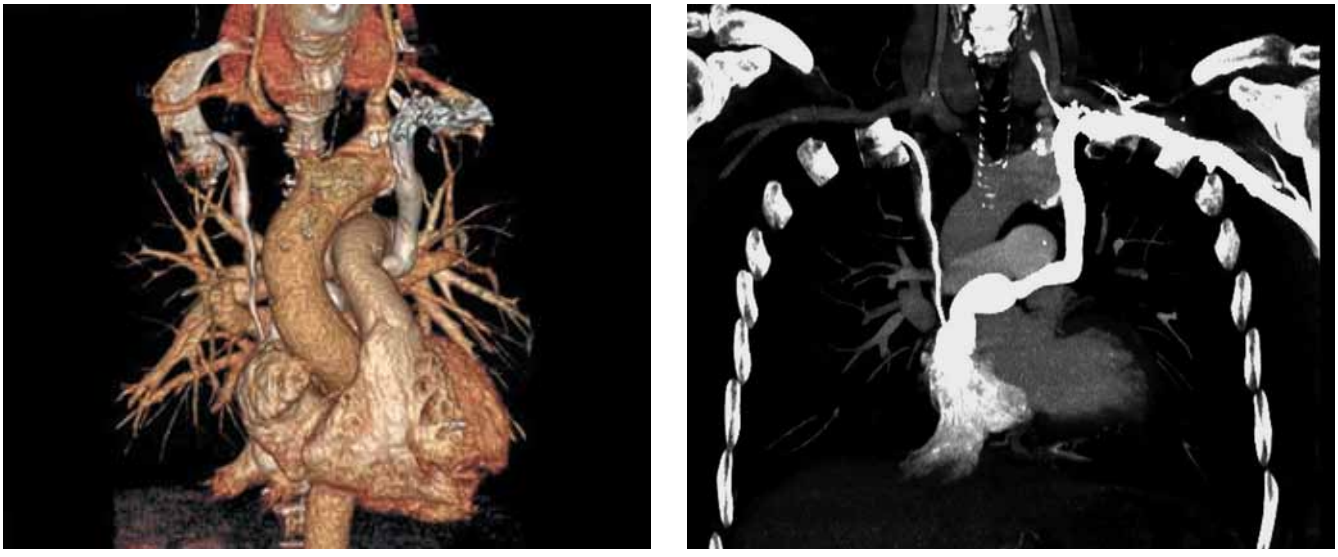

Fig. 3. Contrast-enhanced multispiral CT imaging of the heart. The right superior vena cava is typically located, and it is of small diameter. The persistent left superior vena cava is visualized, anastomosis of the recipient left superior vena cava and the donor superior vena cava is patent

Postoperative echocardiographic results: *Aorta: at the level of the annulus 2.2 cm. Sinuse of Valsalva 3.5 cm. Ascending aorta 3.2 cm. Left atrium: 4.5 cm (anteroposterior dimension) 6.5 × 3.8 (from the apical approach). Right atrium: 5.7 × 3.8 (from the apical approach); Right ventricle: 2.9 cm (anteroposterior dimension); Left ventricle: End-diastolic volume 114 mL; End-systolic volume 37 ml; Stroke volume 77 mL; Ejection fraction 68%. No pathological formations. Valve apparatus: Aortic valve: 3 leaflets, valves are sealed with Pgr/MGr 10.0/– mmHg. 0–1 degree regurgitation. Mitral valve: Leaflets: sealed. M-shaped leaf movement. No peak A. Degree 1 regurgitation. Tricuspid valve: Leaflets: no particularities. 1–2 degree regurgitation. Pulmonary artery: Leaflets: no particularities. Pulmonary artery trunk diameter 2.3 cm, Pgr 7.5 mmHg, degree 1 regurgitation. Systolic pressure 46.0 mmHg. Inferior vena cava 1.9 cm, collapses >50% on inspiration. Pulmonary hypertension group 1.*

DISCUSSION

In cardiac transplantation, PLSVC deserves special attention due to the peculiarities of venous return during the operation of the heart-lung machine and formation of the superior vena cava anastomosis.

If a well-developed unnamed vein is present, occlusion of the PLSVC by simple ligation creates sufficient venous drainage through the innominate vein. However, if the innominate vein is small or absent, PLSVC ligation may increase the risk of neurovascular complications [8].

Several surgical techniques during orthotopic heart transplantation can preserve PLSVC. These surgical approaches include end-to-end anastomosis of the left superior vena cava to the donor's right superior vena cava, end-to-end anastomosis of the PLSVC to the right atrial appendage (direct anastomosis or conduit anastomosis) [8–10].

This paper presents a case of orthotopic heart transplantation in a patient with PLSVC. For direct anastomosis of the PLSVC with the SVC stump of the graft, it was decided to use the biatrial heart transplantation technique, which made it possible to preserve, of sufficient length, the donor heart's SVC stump. When laying the anastomosis in the transverse sinus behind the aorta and the pulmonary artery, there were no significant kinks of the anastomosis, which was confirmed by postoperative examination and the absence of clinical symptoms of venous stasis. In this case, the absence of the innominate vein and complete ligation of the PLSVC could lead to irreversible neurovascular complications. This technique excluded the use of artificial conduits due to the sufficient length of the donor heart's SVC.

CONCLUSION

The clinical effect achieved from treatment fully justifies the chosen surgical tactics and allows us to recommend the proposed tactics for treatment of such a rare anomaly.

The authors declare no conflict of interest.

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The article was submitted to the journal on 2.09.2020