

A Surgical Case of Combined Valvular Disease Complicated by Absent Right Superior Vena Cava and Persistent Left Superior Vena Cava

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A 74-year-old man with combined valvular disease with a recent cerebral infarction was admitted. While undergoing thorough examination for valvular disease, absent right superior vena cava (RSVC) and persistent left superior vena cava (PLSVC) were recognized. Chest X-ray film suggested a right arch protrusion, and CT and venogram confirmed the diagnosis. During surgery, replacement of the mitral and aortic valves and annuloplasty of the tricuspid valve were performed. A blood draining cannula was inserted in retrograde fashion from the coronary sinus into the PLSVC, without any difficulties in the tricuspid valve repair. Due to bradycardic atrial fibrillation, we believed that it would be difficult to insert an endocardial electrode postoperatively, hence myocardial electrode was placed in the right ventricular wall. Absent RSVC combined with PLSVC is very rare, and a patient who underwent combined valve surgery with this rare anatomical abnormality is herein presented. (Ann Thorac Cardiovasc Surg 2003; 9: 401-5)

Key words: absent right superior vena cava, persistent left superior vena cava, combined valvular disease, cardiac surgery

Introduction

Combined valvular disease complicated with absent right superior vena cava (RSVC) and persistent left superior vena cava (PLSVC) is a very rare condition, and there has not been a reported case in Japan. This anomaly can exist alone, and is difficult to diagnose because the hemodynamics of patients with this condition are normal and there may be a lack of clinical symptoms. It is often discovered accidentally while undergoing thorough examination for heart disease, during pacemaker implantation, or catheter insertion. In the present report, we present a patient with combined valvular disease complicated by absent RSVC and PLSVC who underwent surgery, and discuss the diagnosis and surgical procedures.

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Case Report

The patient was a 74-year-old man, and at the age of 68 years, he visited a local doctor and a diagnosis of valvular disease and atrial fibrillation was made. At the age of 73 years, the patient began to notice palpitation and shortness of breath on exertion. After suffering from aphasia, the patient was admitted to our department with diagnosis of cerebral infarction.

At the time of admission, the patient was 156 cm tall, weighed 58 kg, and had blood pressure of 120/52 mmHg, heart rate of 52 beats/minute, and arrhythmia. From the left margin of the third intercostal space to the cardiac apex, a Levine III/VI systolic murmur and II/VI diastolic murmur were audible, but no edema was seen, and neurological findings showed no abnormalities besides aphasia. As to clinical laboratory findings, no abnormalities were seen by general hematological or biochemical tests. The electrocardiogram showed bradycardia, atrial fibrillation, left ventricular hypertrophy, and left axis deviation. Chest X-ray film revealed cardiac dilatation of 59% cardiothoracic ratio (CTR), an abnormal shadow in the right superior mediastinum, and protrusion of the right

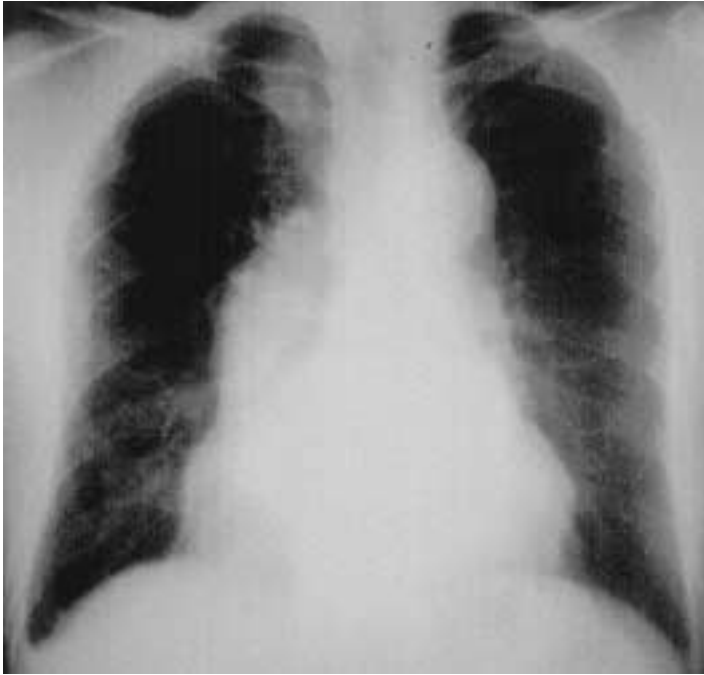


Fig. 1. Chest X-ray film showed cardiac dilatation of 59% CTR, an abnormal shadow in the right superior mediastinum, and a suggestion of protrusion of the right first arch.

first and second arches (Fig. 1). Chest computed tomography confirmed enlarged right and left atria, calcification of the mitral valve, absent RSVC and PLSVC connected to the right atrium. Cardiac catheterization showed that the pulmonary arterial pressure was 28/16 mmHg, and thus, pulmonary hypertension was not confirmed, but mitral stenosis with a valvular area of 0.72 cm², Sellers' grade III aortic valve insufficiency, and moderate tricuspid valve insufficiency were noted. In addition, the veno-

gram showed that the innominate vein was combined with PLSVC and was connected to the coronary sinus, while a contrast radiograph of the right atrium clarified that the superior margin of the right atrium was a blind ending (Fig. 2).

Based on these findings, the patient was diagnosed with combined valvular disease complicated by absent RSVC and PLSVC, and after waiting for three months to allow the patient to recover from aphasia, surgery was per-

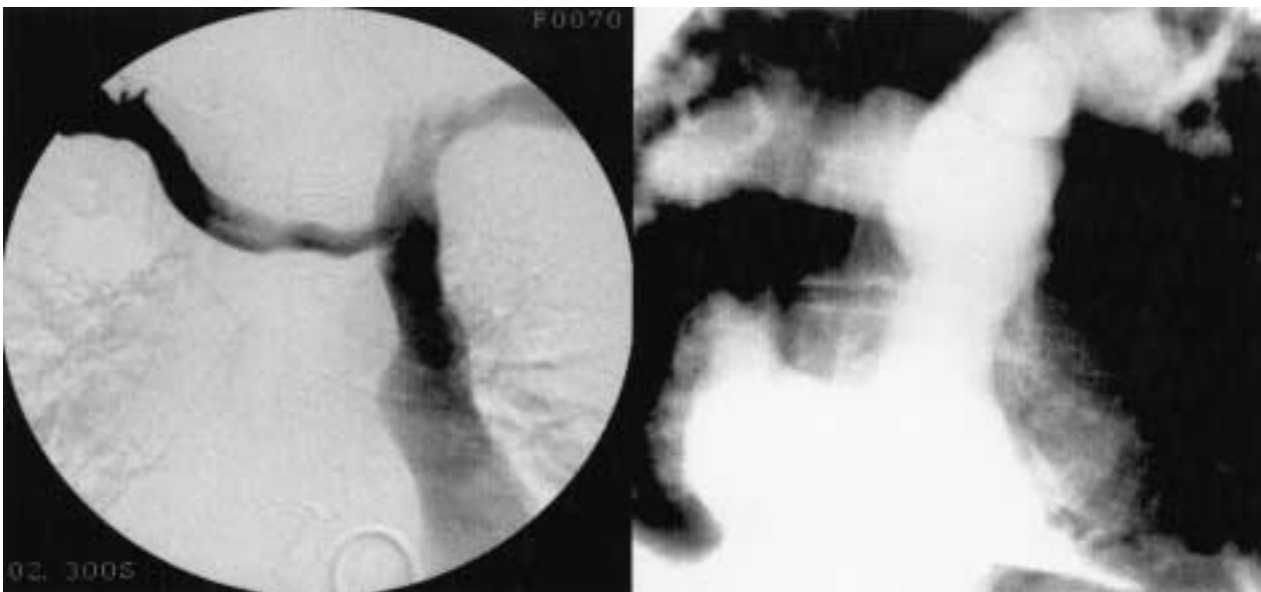


Fig. 2. Venogram showed that the innominate vein was combined with PLSVC and was connected to the coronary sinus (left), and contrast radiograph of the right atrium showed that the superior margin of the right atrium was a blind ending (right).

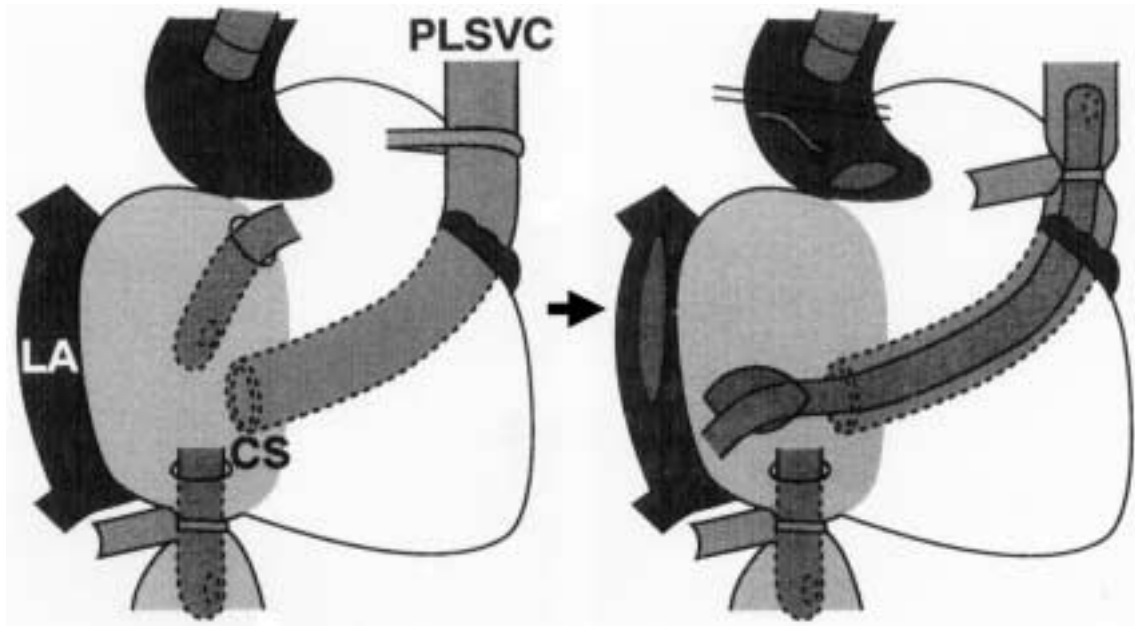


Fig. 3. Extracorporeal circulation was conducted by blood drainage via the right atrium and the inferior vena cava initially. After cardiac arrest, the right atrium was opened and a cannula was inserted through the coronary sinus to the PLSVC. PLSVC, persistent left superior vena cava; LA, left atrium; CS, coronary sinus.

formed. During surgery, after median sternotomy, the pericardium was cut, and absent RSVC and markedly enlarged atria, ventricles and pulmonary artery were observed. Extracorporeal circulation was conducted by supplying blood through the ascending aorta and draining it by inserting a cannula into the right atrium and the inferior vena cava. After cardiac arrest, the right atrium was opened, and a cannula was inserted through the coronary sinus to the PLSVC (Fig. 3). After opening the right side of the left atrium, a close examination of the mitral valve showed hardening and thickening of the anterior cusp and marked fusion of the anterior commissure. While conserving the posterior cusp, artificial valve replacement was performed using a 23-mm CarboMedics valve (CardioMedics Inc., Austin, TX, USA). All three cusps of the aortic valve were thin, and the tips of the cusps did not close properly. The aortic valve was replaced using a 21-mm CarboMedics valve. With regard to the tricuspid valve, annuloplasty was performed according to Kay's method, and in consideration of the possible future use of a pacemaker, a myocardial electrode was placed in the right ventricular wall. The cause of mitral stenosis was identified as rheumatic and aortic insufficiency as degenerative by the pathological findings. The postoperative course for the patient was fine, and the patient was dis-

charged without requiring implantation of a permanent pacemaker.

Discussion

PLSVC is a condition where the fetal left anterior cardinal vein did not regress and remained as the left superior vena cava. PLSVC is seen in 0.3% of all autopsy cases, and accompanies 3-10% of congenital heart malformation.^{1,2)} Of the various heart malformations, PLSVC complicated by absent RSVC with viscerotransposition, belonging to Sherman's type 3, is very rare,³⁾ and this condition accounts for only 1% of PLSVC cases, 0.09% of autopsied cases of congenital heart disease, and 0.07% of intravenous pacemaker implantation cases.^{4,6)}

The hemodynamics of patients with PLSVC connected to the right atrium is the same as that of healthy individuals, as a result, patients are mostly asymptomatic.^{1,7)} Therefore, it is often discovered accidentally while undergoing thorough tests for other heart diseases, intravenous pacemaker insertion, or during autopsy.⁵⁾ In the present patient, PLSVC was discovered during thorough tests for valvular disease, and a chest X-ray film showed an abnormal shadow in the right superior mediastinum suggestive of protrusion of the right first arch. The chest X-

Table 1. Adult cases of absent RSVC and PLSVC reported in Japan

Case No.	Author	Age, sex	Cardiac complication	Surgery
1	Nagatani (1959)	24, M	ASD, double aortic arch	Open heart
2	Nakagawa (1981)	60, F	SSS	PMI
3	Nakayama (1982)	38, M	SSS	PMI
4	Taniura (1983)	78, M	SSS	PMI
5	Nishizawa (1986)	41, M	–	–
6	Araki (1987)	50, M	Mitral regurgitation	Open heart
7	Nagayoshi (1988)	57, M	ASD, pulmonary artery aneurysm	Open heart
8	Fukata (1988)	55, M	SSS	PMI
9	Sakamoto (1990)	61, F	SSS	PMI
10	Watanabe (1992)	54, F	ASD	Open heart
11	Iwamoto (1994)	74, M	SSS	PMI
12	Yamamoto (1996)	72, M	Angina pectoris	–
13	Oi (1999)	80, F	–	–

ASD, atrial septal defect; SSS, sick sinus syndrome; PMI, pacemaker implantation.

ray findings associated with PLSVC include shadow in the upper left direction of the heart or double shadow,⁸⁾ but the X-ray findings seen in the present patient may be unique to combined valvular disease complicated by absent RSVC and PLSVC.

With regard to heart malformations, Bartram et al.⁵⁾ investigated 121 patients and confirmed heart malformations in 56 (46%). The most common malformation was atrial septal defect (16%), followed by endocardial cushion defect (11%) and Fallot's tetralogy (9%). However, the majority of these patients (n=65, 54%) did not have heart malformation, thus clarifying that absent RSVC and PLSVC can often be seen unaccompanied by other malformations.

To the best of our knowledge, there have been 23 reported cases of absent RSVC and PLSVC in Japan, and of these, 12 patients (52%) had heart malformations, 13 patients (57%, Table 1) were adults, and six patients (26%) underwent pacemaker implantation for the treatment of sick sinus syndrome. Only four adult patients (17%) underwent open heart surgery.⁹⁻¹²⁾ Of these four patients, atrial septal defect closure was performed on three patients (partial resection of a pulmonary artery aneurysm was also performed in one patient, and right aortic arch dissection in another), and mitral valve replacement was performed in one patient. Therefore, the present patient was the first case in Japan, who underwent surgery (replacement of the mitral and aortic valves and annuloplasty of the tricuspid valve) for combined valvular disease complicated by absent RSVC and PLSVC.

Some of the problems that surgeons might encounter during surgery include insertion of a blood draining cannula or intravenous insertion of a pacemaker. A cannula

can be inserted into the PLSVC in either way: a cannula can be inserted from the coronary sinus as was done in the present patient, or an L-shaped cannula can be directly inserted into the PLSVC. When PLSVC is complicated by absent RSVC, due to the dysfunction of the sinus node, the pacemaker near the coronary sinus is more active, thus causing coronary sinus rhythm to be predominant.¹³⁾ The former technique is disadvantageous from the standpoint of conserving coronary sinus rhythm,¹⁾ and in pediatric patients, a small surgical field in the right atrium is blocked.¹⁴⁾ The latter technique has been associated with thrombogenesis or a restriction in cannula size.¹²⁾ In the present patient, the former technique was selected because the duration of atrial fibrillation was long; there was no need to conserve coronary sinus rhythm; and cannula insertion was simple because the coronary sinus was markedly dilated, and the visual field was not blocked during the annuloplasty of the tricuspid valve. Furthermore, when implanting a pacemaker in patients with absent RSVC and PLSVC, it is technically difficult to insert an intracardiac electrode postoperatively because the electrode must be inserted at an acute angle,¹⁵⁾ and complications such as phlebotrombosis and electrode detachment have been reported. Numerous reports have recommended the use of a myocardial electrode,¹⁶⁾ and we therefore placed a myocardial electrode into the right ventricular wall during surgery.

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