Persistent Left Superior Vena Cava Draining into the Left Atrium, Atrial Septal Defect, and Absence of the Coronary Sinus

Recognition and Surgical Correction

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SUMMARY

The diagnosis of atrial septal defect (ASD), persistant left superior vena cava (LSVC) and absence of coronary sinus was established by cardiac cateterization in a 20 years old female. At surgery the coronary vein ostia were located in the left atrium. LSVC was ligated and a pericardial patch was sutured around the orifices of the coronary veins and extended to the ASD to redirect the coronary blood flow into the right atrium and close the defect. The postoperative cardiac catheterization and hemodynamic studies revealed good result.

Additional Indexing Words:
Complex congenital heart disease Left superior vena cava pressure

PERSISTENT left superior vena cava (LSVC) is the most common thoracic venous anomaly and is the most frequent cause of enlargement of the coronary sinus (CS). Although the true incidence of this anomaly is unknown,\(^1\) it has been stated to occur in 2.1–10\%\(^2\)-\(^4\) of patients with congenital heart disease and 0.5\% of the general population.\(^5\) Persist LSVC draining into the left atrium (LA) has been reported as rare.\(^6\)

Atresia and absence of the ostium of the CS in the right atrium (RA) is also rare, particularly so in the absence of a LSVC.\(^7\)-\(^15\) Among reported instances of termination of LSVC in the LA in which necropsy had been done, infero-posterior type of atrial septal defect (ASD) was the commonest associated congenital abnormality.\(^5\),\(^9\)

Infero-posterior type of ASD is also an uncommon type of ASD.\(^10\) The studies suggest that when LSVC joins the LA, this vascular anomaly is

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Received for publication May 23, 1980.
part of a developmental complex in which abnormal termination of the CS and a defect in the postero-inferior angle of the atrial septum are also a part.\textsuperscript{11)} The purpose of this communication is to report a case with infero-posterior type of ASD. Besides a review of literature, the clinical and hemodynamic studies, surgical correction as well as the postoperative results will be discussed.

**Case Report**

A 20 years old female was admitted to the Cardiovascular, Medical and Research Center in November 1979 because of palpitation and mild exertional dyspnea.

On physical examination she was a well-developed female with mild cyanosis but no clubbing. The blood pressure was \(120/70\) mmHg and the resting heart rate was \(84/\text{min}\). The first heart sound was normal; the pulmonic component of the second heart sound was loud, widely split and fixed. A grade 3/6 systolic ejection murmur was heard along the left upper sternal border and a grade 2/6 mid-diastolic murmur was audible at the apex.

The electrocardiogram showed normal sinus rhythm, P-R interval of 0.20 sec, right axis deviation and right ventricular hypertrophy. The chest roentgenogram revealed right ventricular enlargement, prominent pulmonary artery and increased pulmonary vascular markings. The right and left cardiac catheterization was performed by percutaneous technique from the right groin. The course of

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**Table I. Hemodynamic Data**

<table>
<thead>
<tr>
<th></th>
<th>Preop.</th>
<th>Postop.</th>
<th>Press. (mmHg)</th>
<th>O\textsubscript{2}Sat. (%)</th>
<th>Press. (mmHg)</th>
<th>O\textsubscript{2}Sat. (%)</th>
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<td></td>
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<td>72</td>
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<td>72</td>
<td></td>
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<tr>
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<td>a = 4</td>
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<td></td>
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<td></td>
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<tr>
<td>mid RA</td>
<td>v = 3</td>
<td>v = 2</td>
<td></td>
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<tr>
<td>low RA</td>
<td>m = 3</td>
<td>m = 2</td>
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<td></td>
<td></td>
</tr>
<tr>
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<td>60/0-2</td>
<td>30/0-2</td>
<td>91</td>
<td>78</td>
<td>76</td>
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<tr>
<td>PA</td>
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<td>L SVC</td>
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<tr>
<td>LV</td>
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<tr>
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<td>2 : 1</td>
<td>1 : 1</td>
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</table>

Legend: Press = pressure; \(O_2\)Sat = \(O_2\) saturation; SVC = superior vena cava; RA = right atrium; RV = right ventricle; PA = pulmonary artery; L SVC = left superior vena cava; PV = pulmonary vein; LA = left atrium; LV = left ventricle; AO = aorta; Qp/Qs = pulmonary/systemic flow.
the venous catheter was from inferior vena cava to the RA and right SVC. From RA, across ASD, LA and 4 pulmonary veins were entered.

From LA, adjacent to left upper pulmonary vein the catheter was also negotiated into a LSVC. The CS in the RA could not be probed. The O₂ saturation and hemodynamic studies were summarized in Table I. Cineangiography with injection of contrast material into main pulmonary artery showed normal PA and branches. On levophase all 4 pulmonary veins entered the LA. The RA was opacified through ASD. Injection into LSVC showed draining of this vessel into the LA (Fig. 1). Left ventricular and aortic root injections showed no other abnormalities.

The patient underwent operative repair of the congenital heart defects using cardiopulmonary bypass. A large defect was located at the infero-posterior part of atrial septum (Raghib's type). There was no CS in the RA. The orifices of coronary veins were located in the left atrium just below the right lower pulmonary vein. The orifice of LSVC was located in the roof of LA near the entrance of the left upper pulmonary vein. The LSVC was clamped and the pressure in the jugular veins were monitored for 10 min. As the pressure did not rise significantly, the LSVC was ligated.

A pericardial patch was sutured around the orifices of the coronary veins and extended to the atrial septal defect to redirect the coronary blood flow into the right atrium and close the defect (Fig. 2). The right and left heart catheterization were performed 4 weeks post operatively. The results were summarized in Table I. The patient was discharged in good condition.
Fig. 2. A) preoperative: drainage of the left superior vena cava (LSVC) and coronary veins (CV) into the left atrium (LA) and atrial septal defect (ASD) are demonstrated. B) postoperative: LSVC is ligated and a patch is sutured around the CV ostia and extended to the ASD to reroute the coronary blood flow into the right atrium and close the defect.

DISCUSSION

Persistent LSVC is not uncommon and its draining into the CS is estimated to occur in 0.5% of the general population. This anomaly may be associated with other intracardiac and extracardiac defects. In most cases, it is of no significant hemodynamic importance, however it can complicate the passage of the pacing catheter, Swan-Ganz catheter and the diagnostic approach from the left arm.

LSVC draining into the LA is uncommon, and it is mostly associated with other complex cardiac abnormalities. It can rarely occur as an isolated anomaly presenting with cyanosis, no cardiac murmur or a faint mild-diastolic murmur due to increased flow through the mitral valve and a soft pulmonary second sound due to diminished pulmonary blood flow and left ventricular
hypertrophy.6)−22) The commonest associated lesion with LSVC draining into the left atrium is an ASD usually of infero-posterior type (Raghib's type), which is also a rare type of ASD.4)10),17) In most instances these 2 anomalies are complicated by absence or atresia of CS as part of a developmental cardiac malformation.11)

Clinically, the clue to the diagnosis of this anomaly is mild to moderate cyanosis, findings of ASD, mid-diastolic flow rumble of mitral valve, and no evidence of pulmonary hypertension. The electrocardiogram is not likely to be helpful in identifying this complex, and is consistent with ASD, and/or other concomitant cardiac abnormalities.10),11)

The chest roentgenogram may show an unusually straight left heart and mediastinal borders.12)

The diagnosis can be established by cardiac catheterization. LSVC would be recognized if the study was done from the left arm. When performed from the femoral vein, the catheter can be manipulated into the LA, pulmonary veins and LSVC. In the RA, the catheter cannot be manipulated into the coronary sinus which is easily possible if LSVC drains into the RA. Oximetry shows a bidirectional shunt at the atrial level, causing systemic desaturation. Injection of contrast material into LSVC will show draining of this vessel into the LA. The pulmonary left ventricular and aortic root angiograms are helpful in identifying the associated anomalies.

In the operative repair of this complex anomaly 2 major points should be considered: 1—The presence or absence of the CS in the RA. The absence of CS indicates a congenital malformation in the sino-atrial region of the heart. This malformation is commonly associated with persistent LSVC, otherwise there are other associated complex cardiac abnormalities.7),15) 2—The persistent LSVC can permanently be ligated if an adequate innominate bridge connecting the left and right SVC has been demonstrated or monitoring of jugular venous and LSVC pressures fail to demonstrate a significant rise after clamping the latter. If the LSVC pressure rises to the critical level of 30 mmHg, central venous complications may occur and may lead to death.4),12),18),23) The risk appears to be minimal if the cava to be ligated is approximately the same size or smaller than the one to be left patent, and an innominate vein or collateral channels exist. The communication between the 2 superior venae cavae via collateral venous channels is adequate when proved by failure of the pressure to rise above 30 mmHg upon temporary occlusion of one cava.4) In our case, the jugular vein pressure rose only 3 mmHg, therefore it was permanently ligated. Ligation of LSVC draining into the left atrium has been reported,4),13),21) to avoid possible hazard of other methods used.4),12),19),20),22)
After ligation of LSVC or its diversion into the RA and closure of ASD, an obligatory right to left shunting via the coronary veins draining into the left atrium will remain. It is stated that leaving this shunt untouched should be physiologically insignificant, however to eliminate this relatively large right to left shunt which is as high as coronary blood flow, we created a baffle to redirect the coronary veins into the RA.

The embryological basis of this developmental complex has been described in detail. All the features of this complex anomaly result from a faulty development in the sino-atrial region of the heart. The left atriovenous fold of the left cardinal vein normally contributes to formation of the postero-inferior part of the atrial septum and coronary veins and its abnormal development results mostly in an infero-posterior type ASD, absence or atresia of the CS and the drainage of LSVC into the LA.

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