PERSISTENT LEFT SUPERIOR VENA CAVA DRAINING INTO
THE LEFT ATRIUM ASSOCIATED WITH
THE ATRIAL SEPTAL DEFECT AND PARTIAL ANOMALOUS
PULMONARY VENOUS DRAINAGE:
—Surgical Correction with Sepcially Designed Prosthetic Material—

TADAYOSHI AKASAKA, M.D., HIROSHI OHARA, M.D., YASUNORI OHKAWA, M.D.,
NAOAKI KOHGUCHI, M.D. AND KENJI ITOH, M.D.

Anomalous drainage of the persistent left superior vena cava into the left atrium with partial anomalous pulmonary venous return and the atrial septal defect is relatively rare condition, and there reported various kinds of surgical procedures in correction for this lesion. This report describes the original and simple surgical procedure in total correction of these lesions performed in our institute, using a large Dacron patch connected with a knitted Dacron vascular graft in its center. Surgical correction was accomplished by anastomosis of a Dacron graft with the orifice of the persistent left superior vena cava into the left atrium, widening of the atrial septal defect, and reconstruction of a new atrial septum with a Dacron patch. Postoperative cardiac evaluation revealed good condition of blood stream through a Dacron graft from the left superior vena cava to the right atrium, with corrected pulmonary venous drainage into the left atrium without pulmonary venous congestion and superior vena cava syndrome.

THE surgical management of the persistent superior vena cava draining directly into the left atrium without the communicating vein is also discussed.

CASE HISTORY

The patient, a 4-year-old girl, was pointed out heart murmur by her home doctor on catching a cold, and referred to our clinics. She had been known to have hyperperspiration, tachypnea on exertion and easy fatigability since 2 years before. Precordium was prominent on the

middle line of sternum. On auscultation, systolic ejection murmur of Levine 2/6 was audible at 2–3 intercostal space of the left sternal border, and diastolic inflow murmur of Levine 1/6 on the tricuspid orifice area. Second sound was fixedly splitting. Liver was palpable in 2 finger breadth. On chest X-P, cardiomegaly (CTR: 65%) and moderately increased pulmonary vascularity were demonstrated. ECG revealed normal sinus rhythm with QRS axis of 120, almost normal P wave in all leads, and incomplete right bundle block pattern. She was treated with digitaliza-

(Received on October 6, 1977; Accepted on November 11, 1977)

The Department of Thoracic and Cardiovascular Surgery, Kanagawa Children's Medical Center, Yokohama, Japan

Address for reprint request: Dr. Akasaka, T., Department of Thoracic and Cardiovascular Surgery, Kanagawa Children's Medical Center, Mutsukawa 2-138-4, Minami-ku, Yokohama, 232 Japan

TABLE I PREOPERATIVE AND POSTOPERATIVE CARDIAC CATHETERIZATION DATA

	Before operation	After operation	
	<i>O₂ Saturation</i>	<i>O₂ Saturation</i>	<i>Pressure (mmHg)</i>
R-SVC	64%	70%	
L-SVC	62		
IVC	79	71	
RA	77	70	
RV	88	70	42/8
M-PA	89	70	42/25 (32)
L-PA	86	70	25/20 (23)
LA	89		
R-PV	94		
FA	74	95	120/70 (85)

Flow ratio: 4.0 R-L Shunt: 21% L-R Shunt: 70%

(R-SVC: Right Superior Vena Cava, IVC: Inferior Vena Cava, RA: Right Atrium, RV: Right Ventricle, M-PA: Main Pulmonary Artery, R-PV: Right Pulmonary Vein, FA: Femoral Artery)

Fig.1. Venogram via the left antecubital vein did not reveal the coronary sinus, but vague shadow of the left atrium.

tion and diuretic therapy, and recommended to have further cardiac evaluation.

Right heart catheterization was performed 4 months later via the right saphenous vein (Table I). The catheter was inserted into the right

atrium, the right ventricle, and the main pulmonary artery, and then into the left atrium. It was also inserted easily into the left superior vena cava (LSVC), but was not introduced into the innominate vein via the LSVC. Oxygen step-up was recognized in the right atrium, suggesting of the atrial septal defect. Contrast media was injected into the main pulmonary artery and almost simultaneous opacification of the right atrium with the left atrium was visualized. Left pulmonary veins were drained normally into the left atrium, but it was questionable whether right pulmonary veins returned to the right atrium or left atrium.

On the day before scheduled operation, she was noted of her cyanosis on exertion and very mild clubbed fingers. The further evaluation was performed to study the cause of cyanosis. Contrast media was injected into the left antecubital vein for the purpose to make sure the cause of cyanosis. Venogram did not reveal clear opacification of the coronary sinus via the LSVC, but vague shadow of contrast media in the left atrium, which suggested that the LSVC drained into the left atrium and this was thought the main cause of cyanosis. No opacification of the communicating vein between the LSVC and the RSVC was also visualized in this venogram (Fig. 1). Dye-dilution test performed from the right and left antecubital vein revealed the presence of large amount of R-L shunt and small amount of L-R shunt on the curve from the left side, large L-R shunt from the right side. There-

Fig.2. Intracardiac findings and operative procedure.

- 2-A: large atrial septal defect with the bridge tissue across its center, partial anomalous right pulmonary venous drainage into the right atrium, and the LSVC draining directly into the left atrium.
- 2-B: prosthetic material prepared preoperatively.
- 2-C: anastomosis of Dacron graft to the LSVC and new atrial septum reconstructed with Dacron patch.
- 2-D: LSVC was drained into the right atrium through the implanted Dacron graft, and right pulmonary veins into the left atrium.

fore, according to the results of these test added to cardiac catheterization data and other physical findings, we preoperatively concluded direct

drainage of the LSVC into the left atrium, the atrial septal defect of secundum type, and possibility of anomalous drainage of right pulmonary

Fig. 3. Postoperative angiocardigraphy following injection of contrast media from the main pulmonary artery. No congestion of pulmonary venous return.

Fig. 4. Postoperative venogram via the left antecubital vein revealed the clear opacification of the well conditioned Dacron graft from the LSVC to the right atrium.

veins into the right atrium.

OPERATION

Operation was performed on January 20, 1977 through mediansternotomy. The right atrium, right ventricle and main pulmonary artery were enlarged for her age. The LSVC was found to return directly to the left atrium, at the

portion just cephalad of the left atrial appendage. The outer diameter of the RSVC was approximately two-thirds of the LSVC. Mild hypothermic perfusion was established by cannulation into the ascending aorta, the RSVC and the inferior vena cava. The heart was electrically fibrillated and the right atrium was opened obliquely. Intracardiac findings was illustrated in Fig. 2-A. There found a large atrial septal defect with a bridge tissue of septum across it which was excised, and through the enlarged atrial septal defect, the LSVC was recognized to drain directly into the left atrium, just posterior and cephalad of the left atrial appendage. The ostium of right pulmonary veins was recognized in the right side of the right atrium and left pulmonary veins in the left atrium. The coronary sinus located in the normal position and seemed normally sized. There seemed no other cardiac anomalies in atrio-ventricular valves, right ventricle, and left ventricle under direct vision and on digital examination. Surgical procedures was carried out, using a large Dacron patch connected with a knitted Dacron vascular graft in its center, which was prepared preoperatively as shown in Fig. 2-B. A knitted Dacron vascular graft was sutured around of the orifice of the LSVC in the left atrium, with 8 interrupted sutures. A new atrial septum was reconstructed with a Dacron patch in such a manner that right pulmonary veins returned to the left atrium without congestion. It was sutured to the lower margin of the atrial septal defect and to the cephalad free wall of the right atrium in the right and upper portion. When temporary occlusion of the LSVC was released, there flushed much venous blood from the opening window of a Dacron graft. Therefore, the LSVC was to be drained into the right atrium through the sutured Dacron graft. Electrical fibrillation was discontinued and spontaneous vigorous cardiac beating began. Weaning from the cardiopulmonary bypass was easy.

The patient made an uneventful recovery except for transient nodal bradycardia on ECG, which was well controlled with administration of small dosis of isoproterenol. We did not find clinically any sign of pulmonary venous congestion, superior vena cava syndrome and cerebral complications postoperatively. Cyanosis disappeared. Postoperative cardiac catheterization was performed one month later. No oxygen step-up in any part of right heart side and normal pulmonary artery pressure were recognized

Table I). Angiocardiography by injection of contrast media in the main pulmonary artery and in the left antecubital vein were shown in Fig. 3, 4 which suggested of well corrected arrangement of systemic and pulmonary venous return. She was discharged 5 weeks after the operation and followed as a outpatient.

COMMENT

Persistent superior vena cava is relatively common and has been reported from 0.5% to 4% of its incidence among the all patients of congenital heart diseases.³ Usually this anomalous vessel drains into the right atrium via the coronary sinus and as such has no physiological significance unless the patient is to have some intra-cardiac operation for associated heart anomalies with cardiopulmonary bypass. When the LSVC drains directly into the left atrium, this anomaly is usually the source of significant cyanosis with R-L shunt in the left atrium, and in this instance, surgical treatment should be considered. But even in this condition, a rare case without significant arterial oxygen unsaturation was reported.⁵ The incidence of the LSVC draining directly into the left atrium is reported 8% to 10% of all cases of the LSVC. This anomaly is usually associated with other important cardiovascular anomalies such as atrial septal defect, common atrium, endocardial cushion defect, and anomalous pulmonary venous return,⁹ but occasionally it may be found alone.^{1,4}

When the right superior vena cava exists and the innominate vein is of adequate size, a simple ligation of the LSVC draining into the left atrium would be sufficient. Although there reported that the LSVC draining into the left atrium can be safely ligated, even if the innominate vein is absent, because there are many run off routes via jugular collateral vessels,⁶ we believe that construction of blood route would be preferable so as to make LSVC blood drain into the right atrium, so we did not try to make temporary occlusion of the LSVC transoperatively.

There reported various kinds of surgical procedures for correction of this anomaly when the innominate vein is absent and adequacy of anastomosis between two upper systemic veins may be questionable. Shumacker¹⁰ reported this anomaly associated with Tetralogy of Fallot which they treated by transposing the LSVC to the right atrium, using the extracardiac transposed vessel by designing the extension flap from the left atrial appendage in the shape of the tube.

Rastelli⁸ reported that a tunnel was constructed from the posterior wall of the left atrium with a venous cannula for cardiopulmonary bypass through the LSVC used as a stent. De Leval² described several cases of construction of a pericardium tunnel from the LSVC to the right atrium, and in some cases associated with large atrial septal defect or common atrium, an intra-cardiac baffle of pericardium or prosthetic material was inserted in such a manner that the pulmonary veins drained into the mitral valve and systemic veins drained into the tricuspid valve, in experiences at Mayo Clinic. Kabbani⁴ reported in a cyanotic patient with absent RSVC and intact atrial septum, a cloth baffle replacement of partitioning pulmonary venous blood into the left ventricle after the interatrial septum excision, releasing complications of pulmonary and systemic venous congestion caused by placed pericardium at the first operation of this same patient.

We preferred a Dacron patch connected with a Dacron graft designed preoperatively for correction of this anomaly as shown in Fig. 2-C, than pericardial patch, because pericardial patch designed in a shape of tube frequently became affected by degeneration, fibrosis, calcification and complete disintegration after placement, and extracardiac transposed vein shaped with pericardium became later thrombosed as reported by Shumacker.¹⁰ More than anything else, our surgical procedure presented here is simple and easy to be performed for correction when appropriate size of prosthetic material is preoperatively prepared.

The postoperative course in our patient was neither complicated with superior vena cava syndrome nor congestion of pulmonary veins,⁷ and postoperative angiocardiography revealed well conditioned blood flow of the LSVC draining into the right atrium through the implanted Dacron graft.

The same surgical procedure of atrial repartitioning can be used for correction of anomalous systemic and/or pulmonary venous drainage, if the shape of designed prosthetic material will be reformed. There continues a possibility of the risk of thrombosis in the intraatrial implanted Dacron graft, therefore we have to follow this patient in future with greatest interest.

REFERENCES

1. DAVIS, W. H., JORDAAN, F. R. & SNYMAN, H. W.: Persistent left superior vena cava draining into

- the left atrium, as an isolated anomaly. *Am. Heart J.* **57**: 616, 1959.
2. De LEVAL, M. R., RITTER, D. G., McGOON, D. C. & DANIELSON, G. K.: Anomalous systemic venous connection. Surgical considerations. *Mayo Clin. Proc.* **50**: 599, 1975.
 3. FRASER, R. S., DVORKIN, J., ROSSALL, R. E. & EIDEM, R.: Left superior vena cava. A review of associated congenital heart lesions, catheterization data and roentgenologic findings. *Am. J. Med.* **31**: 711, 1961.
 4. KABBANI, S. S., FELDMAN, M., ANGELINI, P., LEACHMAN, R. D. & COOLEY, D. A.: Single (left) superior vena cava draining into the left atrium. *Ann. Thorac. Surg.* **16**: 518, 1973.
 5. MEADOWS, W. R. & SHARP, J. T.: Persistent left superior vena cava draining into the left atrium without arterial oxygen unsaturation. *Am. J. Cardiol.* **16**: 273, 1965.
 6. OSAWA, M., et al.: Persistent left superior vena cava and surgical management of its blood return. *J. Jap. Assoc. Thorac. Surg.* **24**: 1143, 1976.
 7. RAGHIB, G., RUTTENBERG, H. D., ANDERSON, R. C., AMPLATZ, K., ADAMS, P. & EDWARDS, J. E.: Termination of left superior vena cava in left atrium, and atrial septal defect, and absence of coronary sinus: A developmental complex. *Circ.* **31**: 906, 1965.
 8. RASTELLI, G. C., ONGLEY, P. A. & KIRKLIN, J. W.: Surgical correction of common atrium with anomalously connected persistent left superior vena cava. Report of a case. *Mayo Clin. Proc.* **40**: 528, 1965.
 9. RASTELLI, G. C., RAHIMTOOLA, S. H., ONGLEY, P. A. & McGOON, D. C.: Common atrium. Anatomy hemodynamics, and surgery. *J. Thorac. Cardio. Surg.* **55**: 834, 1968.
 10. SHUMACKER, H. B., KING, H. & WALDHAUSEN, J. A.: The persistent left superior vena cava. Surgical implications, with special reference to caval drainage into the left atrium. *Ann. Surg.* **165**: 797, 1967.