Arch Jpn Chir 57(5), 434~442, Sept., 1988

症　　例

A Case Report of Coronary Sinus Septal Defect Associated With Left Superior Vena Cava, Sinus Venosus Defect and Partial Anomalous Pulmonary Venous Drainage

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Received for Publieation, June 9, 1988.

Summary

A 3-year-9-month old girl was operated on for atrial septal defect at the age of 2 years and 10 months through right antero-lateral thoracotomy. At surgery, a large sinus venosus defect with partial anomalous pulmonary venous drainage of the right lung, and coronary sinus septal defect with left superior vena cava were found. Patch closure of the defects, redirecting right pulmonary vein and coronary sinus into the left atrium, was performed. Postoperatively, she presented left heart failure and slight cyanosis of the lip. Beausce left superior vena cava draining into the left atrium also offers the great risk of brain embolism or abscess, the vessel was ligated about one year after the first operation.

Introduction

Atrial septal defect (ASD), including ostium secundum defect and partial endocardial cushion defect, is a common congenital heart disease. However, coronary sinus septal defect (CSSD), caused by incomplete formation of the left atrio-venous fold, is rare; occuring in 0.2–0.6 percent of ASD[1,15]. Only twenty one cases, including a case under discussion, have been reported in Japan (Table 1)[1,2,5~12,14~19,24~26]. We here report on a case, who was successfully operated on for CSSD and sinus venosus defect with partial anomalous pulmonary venous drainage (PAPVD) of the right lung. The relevant literatures are also reviewed.

Case Report

A 3-year-9-month old girl was admitted to our hospital at the age of 2 years and 10 months for reconstructive surgery of ASD.

Key words: Coronary sinus septal defect, Sinus venosus defect, Partial anomalous pulmonary venous drainage, Left superior vena cava.
索引語：冠静脈洞型心房中隔欠損，静脈洞型心房中隔欠損，部分肺動脈還流異常，左上大静脈.
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Table 1. Case reports in Japan

	Auther	Case	Symptom	Preoperative Diagnosis	LSVC	Associated Anomaly	Type	Operative Procedure	Result
1.	I. Ngai (1966)	12y.o. F	D.O.E. Palpitation	ASD	(−)	Hepatic vein -CS return		Explaration only	died
2.	T. Iijima (1977)	19y.o. M	(−)	ASD	(−)	(−)			alive
3.	H. Inoue (1978)	36y.o. M	D.O.E.	ASD	(−)	ASD (Primum, Central) Sinus Venosus Inferior Caval			alive
4.	Y. Moritani (1978)	16y.o. M	D.O.E. Palpitation	ASD	(−)	(−)			alive
5.	K. Kitano (1980)	57y.o. F	Dyspnea Heart Failure	MR+TR	(−)	PFO		(−)	died (Autopsy)
6.	K. Tsuchiya (1981)	5y.o. F	(−)	ASD	(−)	(−)			alive
7.	M. Okumori (1982)	42y.o. F	D.O.E. Leg edema	CSSD	(+)	(−)			alive
8.	M. Okumori (1982)	41y.o. F	Palpitation Fatiguability	CSSD TGA (Post-Blalock)	(+)	TGA			?
9.	M. Konishi (1983)	63y.o. M	D.O.E. Leg edema	ASD+Mr. (MV prolapse)	(−)	(−)			alive
10.	S. Kakimoto (1984)	60y.o. F	D.O.E. Palpitation	ASD	(−)	PFO			alive
11.	H. Mayumi (1984)	16y.o. F	(−)	VSD p.o. residual VSD ASD	(+)	VSD Ar			alive
12.	Y. Okada (1984)	43y.o. F	Palpitation D.O.E.	ASD p.o. CSSD	(−)	ASD			died
13.	S. Azuma (1984)	12y.o. F	Cyanosis Tachypnea	ASD PS	(−)	PFO Pure PS		 +Reconstruction of RVOT	alive
14.	K. Akiyama (1984)	64y.o. F	D.O.E.	ASD IHD (LAD 75%)	(−)	(−)		 +A-C Bypass	alive
15.	Y. Hamada (1984)	55y.o. F	D.O.E. Palpitation	CSSD	(+)	(−)			?
16.	K. Takahashi (1985)	15y.o. F	Brain abscess	ASD	(+)	PFO			alive
17.	Y. Fujiseki (1985)	8y.o. M	(−)	CSSD	(−)	(−)			alive
18.	M. Terada (1986)	4y.o. F	(−)	ASD PS	(−)	PFO Pure PS		 +Commissurotomy of PV-	alive
19.	Y. Hayashibe (1987)	47y.o. M	Chest oppression	ASD	(−)	(−)			alive
20.	H. Nakano (1987)	11y.o. M	(−)	ASD	(−)	(−)			alive
21.	Recent Report	3y.o. F	Cyanosis Heart Failure	ASD p.o. LSVC (CSSD)	(+)	ASD (Sinus Venosus) PAPVC			alive

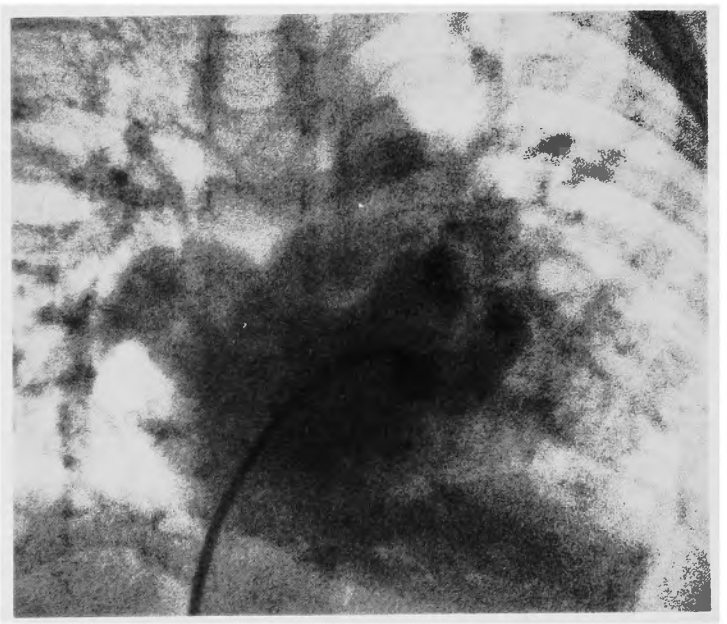

Fig. 1. Right ventriculogram, revealing left-to-right shunt at the atrial level.

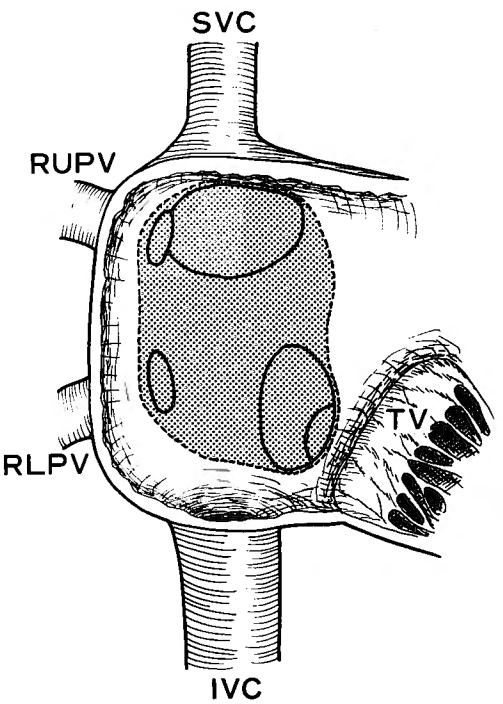

Fig. 2. Schema of the operative procedure. The defects were closed with a single patch, redirecting the drainage from right pulmonary vein and coronary sinus into the left atrium.

SVC: superior vena cava, IVC: inferior vena cava, RUPV: right upper pulmonary vein, RLPV: right lower pulmonary vein, TV: tricuspid valve.

On admission, her blood pressure was 110/70 mmHg and pulse rate was 98/min. A grade 3/6 systolic murmur and 2/6 diastolic rumbling were audible along left sternal border with the second heart sound splitting fixedly. Chest roentogenography revealed cardiomegaly (cardio-thoracic ratio: 0.55) with increased pulmonary vasculature. On the electrocardiogram, an incomplete right bundle branch block was noted. Upon cardiac catheterization, pulmonary and aortic pressure were 30 and 85 mmHg, respectively. Step-up of O_2 content was seen in the right atrium; calculated left-to-right shunt being 75%. The oxygen saturation of blood gas in the left ventricle was 90%. Angiography revealed left-to-right shunt at the atrial level (Fig. 1) and the diagnosis of ASD was established.

Repair of ASD was performed through right antero-lateral thoracotomy. At operation, right superior vena cava (RSVC) was too small, suggesting the presence of left superior vena cava (LSVC). A sinus venosus defect of 7×10 mm, associated with PAPVD of the right lung into the right atrium, was found. A defect of 7×9 mm was also seen on the postero-inferior part of the septum. A massive venous return from these defects was indicative of coronary sinus septal defect with LSVC. Because there were limitations of the operative time and exposure under artificial ventricular fibrillation through right antero-lateral thoracotomy, these defects

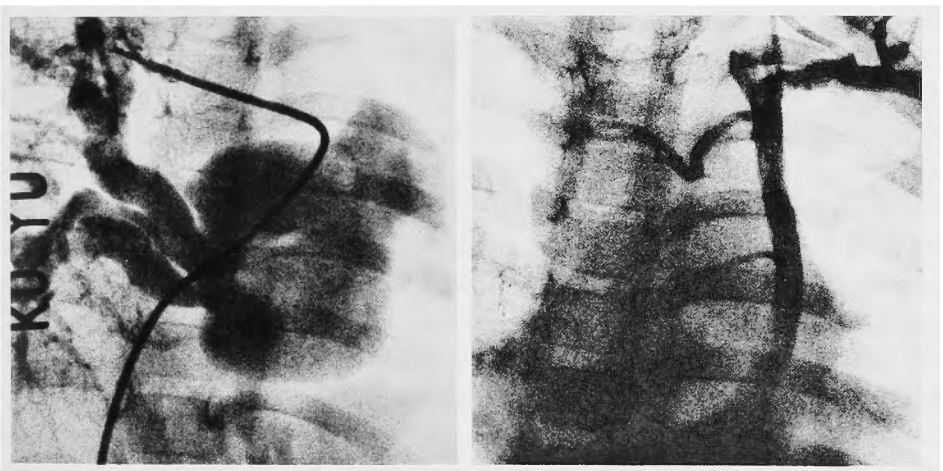

Fig. 3. Right pulmonary angiogram (left upper panel) and left superior vena cavogram (right upper panel), demonstrating pulmonary venous return draining into left atrium without mechanical obstruction, and left superior vena cava draining into left atrium.

RSVC: right superior vena cava, LSVC: left superior vena cava, RA: right atrium, LA: left atrium, IVC: inferior vena cava.

were closed with a single Rygg patch, redirecting the drainage from right pulmorary vein and coronary sinus into the left atrium. (Fig. 2).

Postoperatively, left heart failure and cyanosis of the lip were present, and 0.1 mg of digosin and 10 mg of furisemide were required daily. Echocardiography showed dilation of the left atrium and a large coronary sinus. Right pulmonary angiography demonstrated pulmonary venous return draining into left atrium without obvious mechanical obstruction. Contrast media injected into the left ante-cubital vein visualized LSVC draining into the left atrium: left innominate vein was not recognized (Fig. 3). Temporal balloon occlusion of LSVC raised the proximal pressure only to 5 mmHg, suggesting the presence of adequate collaterals.

Then we concluded that her left heart failure was not caused by mechanical pulmonary venous obstruction but by a massive venous drainage into the left atrium via LSVC. Because there was also the great risk of brain embolism and abscess, left antero-lateral thoracotomy was made about one year after the first operation. Temporal ligation of the LSVC produced a rise in the mean left internal juglar pressure of only 6 mmHg, thereby ligating the vessel. Her postoperative course was uneventful and she was discharged in good condition on the 7th day after the operation and has been doing well with no medication.

Discussion

Coronary sinus septal defect, first described by RAGHIB et al. in 1965[22] as a developmental anomaly consisting of termination of LSVC in the left atrium, an ASD and absence of coronary sinus, is a rare congenital heart disease. Embryologically, it is recognized as being caused by incomplete formation of the left atrio-venous fold (Fig. 4)[13,22]. Various types have been reported so far (Table 1): A) communication between the coronary sinus and left atrium (a focal defect in the coronary sinus septum: cases 1, 4, 11, 12, 13, 15, 20), B) coronary sinus septal defect (incomplete fusion of the atrial septum and atrio-venous fold: cases 2, 3, 5, 6, 9, 10, 14, 17, 18, 19, 21), C) unroofed coronary sinus (complete failure of development of left atrio-venous fold: cases 7, 8, 16). When the brachiocephalic anastomosis is incomplete, LSVC will remain; about half of the cases in foreign literature[14] are associated with LSVC, while only 5 of 21 cases in Japan.

There are no characteristic symptoms of the lesion. In most patients, operation is carried out for preoperative diagnosis of ASD, and the lesion is found at operation. However, it may not be always detected at operation, because of the defect being located in the left atrium. Additionally, detection and complete repair of the lesion may be difficult, when the operation is done under artificial ventricular fibrillation through right antero-lateral approach as in our case. Incomplete repair will remain as an intra-atrial shunt. Especially with LSVC, a large right-to-left shunt will offer the great risk of brain embolism and abscess, as reported by RAGHIB et al. in 1965[22], QUAEGEBEUR et al. in 1979[21] and TAKAHASHI et al. in 1985[24]. Therefore, it is of vital importance to doubt of the presence of the lesion in all the patient diagnosed preoperatively as ASD.

The presence of LSVC is suggestive of the lesion, and may be suspected by a vertical shadow

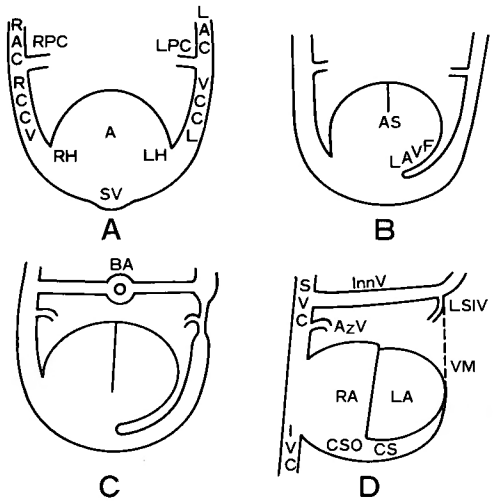

Fig. 4. Schema of normal development of coronary sinus and superior vena cava. A). Common cardinal veins (CCV) drain into the sinus venosus (SV) and primitive atrium (A) via the sinus venosus horn (H). B). With the atrium divided, left atrio-venous fold (LAVF) is growing inside to form a roof of left sinus venosus horn (LH). C). As the septation of the atrium and sinus venosus further develop, a communication between bilateral anterior cardinal veins (AC) occurs (brachio-cephalic anastomosis, BA). D). The atrial septum (AS) and left atrio-venous fold fuse together, then both atrium (RA, LA) and coronary sinus (CS) are created. Right superior vena cava (SVC) is derived from the caudal end of right anterior vein (RAC) and common cardinal vein (RCCV); The left innominate vein (InnV) from the cephalic portion of the left anterior cardinal vein (LAC) and brachio-cephalic anastomosis; The vein of Marshall (VM) from the left common cardinal vein (LCCV). PC: posterior cardinal vein, AzV: Azygos vein, LSIV: superior intercostal vein, CSO: coronary sinus orifice, IVC: inferior vena cava.

in the left upper mediastinum on chest roentogenogram[13] or detected on computed tomogram[19]. YEAGER et al.[27] and HAMADA et al.[6] in 1984 reported the usefulness of two-dimensional echocardiography, which will demonstrate a dilated coronary sinus and the defect between the coronary sinus and left atrium. Contrast echocardiography obtained by injecting the echoproducing microbubbles into the ante-cubital vein is also available[3,5]. On cardiac catheterization, bidirectional shunt at atrial level or low oxygen saturation of systemic arterial blood gas may be observed[13]. Venography obtained by catheter, accidentally or intentionally advanced into LSVC through the coronary sinus, also contributes to diagnosis[13,19]. Contrast media injected into the left ante-cubital vein will visualize the vessel draining into the left atrium. However, the preoperative detection is usually difficult. At operation, an absence of or small RSVC, or a massive venous return from a large coronary sinus indicates the possibility of LSVC, which can be confirmed by careful inspection of a heart.

Surgical options depend on the type of the defect and presence or absence of LSVC. The defect without LSVC will be corrected effectively by closure of coronary sinus ASD with suture or patch (Case 6, 14). Closure of the defect, redirecting the drainage from the coronary sinus into the right atrium, has been also reported (Case 2, 3, 4, 9, 10, 12, 13, 17, 18, 19, 20). In the

defect with LSVC, it is of surgical importance to redirect the return from the vessel into the right atrium[21,22,24]. Simple ligation of the vessel will be a more safe and accurate procedure (Case 7, 8, 16, 21). According to FEED et al. in 1973[4] and LEE et al. in 1979[13], a rise in proximal pressure only to less than 10 mmHg during temporal occlusion of the vessel, by balloon at catheterization or by ligation at operation, suggests that the vessel can be ligated. OOSAWA et al. in 1984[20], in their experimental study, described that RSVC with a diameter of more than 2/3 of the left would indicate the possibility. In a situation, in which the vessel can not be ligated, the following procedures are required: A) The defect is closed with suture or patch to redirect the drainage from the coronary sinus into the right atrium, as described previously (Case 11, 15). B) The roof of the coronary sinus is created with suture or prosthetic material[13]. C) The divided LSVC is reimplanted into the right atrium or RSVC[23], combined with closure of coronary sinus ASD as in case 7, 16 and 21.

In our case, studies before the first operation were not suggestive of the lesion. There was although low oxygen saturation of the blood gas in the left ventricle, which was under question at that time. At operation, a sinus venosus defect with PAPVC and a lesion with LSVC were found. Because the operation was done under artificial fibrillation through right antero-lateral thoracotomy, complete repair was difficult. Therefore, the defects were closed with a single patch, redirecting the drainage from the right pulmonary vein and coronary sinus into the left atrium, and LSVC was successfully ligated later. Our experience with this case strongly impress us with the diagnostic and surgical importance of this anomaly, lying concealed in ASD.

References

1) Akiyama K, Matsuyama Y, Imabun S, et al: A 64-year old case of combined coronary sinus ASD closure and coronary arterial bypass operation. Respiration and Circulation **32**: 855–859, 1984 (in Japanese).
2) Azuma S, Kakeuchi N, Imamura Y, et al: A surgical case of coronary sinus-left atrial communication combined with pure pulmonary stenosis. Jpn Ann Thorac Surg **4**: 191–195, 1984 (in Japanese).
3) Bourdillon PD, Foale RA, Somerville J: Persistent left superior vena cava with coronary sinus and left atrial connections. European J Cardiol **11**: 227–234, 1980.
4) Feed MD, Rosenthal A, Bernhard WF: Balloon occlusion of a persistent left superior vena cava in the pre-operative evaluation of systemic venous return. J Thorac Cardiovasc Sung **65**: 835–839, 1973.
5) Fujiseki Y, Saito S, Sagami R, et al: Two-dimensional echocardiographic diagnosis of coronary sinus ASD. Heart **17**: 748–752, 1985 (in Japanese).
6) Hamada Y, Ebihara H, Tanimoto Y, et al: Unroofed coronary sinus demonstrated by two-demensional echocardiography. Am Heart J **108**: 1558–1560, 1984.
7) Hayashibe Y, Yuge I, Ustumi K, et al: A surgical case of coronary sinus ASD. Respiration and Circulation **35**: 577–580, 1987 (in Japanese).
8) Iijima T, Taniguchi T, Onouchi K, et al: A case report of coronary sinus ASD. Jap J Thorac Surg **25**: 1523, 1977 (in Japanese).
9) Inoue H, Furuse A, Saigusa M: Coronary sinus atrial septal defect. Report of a case. Heart **10**: 725–729, 1978 (in Japanese).
10) Kakimoto S, Asada K, Sasaki S, et al: A surgical case of coronary sinus ASD. Jap J Thorac Surg **37**: 850–853, 1984 (in Japanese).
11) Kitano K, Ohkawa S, Ito Y, et al: An autopsy case of ASD of the coronary sinus type. Heart **12**: 102–106, 1980 (in Japanese).
12) Konishi M, Nobuoka W, Yokoyama S, et al: A case report of coronary sinus ASD. Jap J Thorac Surg **36**: 667–670, 1983 (in Japanese).

13) Lee ME, Sada RH: Coronary sinus septal defect. J Thorac Cardiovasc Surg **78**: 563–569, 1979.
14) Mayumi H, Tanaka J, Tokunaga K, et al: A case report of communication of the coronary sinus with both atria associated with persistent left superior vena cava. Jpn J Surg **14**: 1–15, 1984.
15) Moritani Y, Sakai Y, Taniguchi G, et al: A surgical case of coronary sinus ASD. Jap J Thorac Surg **31**: 389–391, 1978 (in Japanese).
16) Nagai I, Oda Y, Kawata K, et al: A case of the communication between L.A. and coronary sinus. Jap J Thorac Surg **19**: 363–365, 1966 (in Japanese).
17) Nakano H, Sato T, Chin Y, et al: Surgical management for unroofed coronary sinus: A case report. Jap J Thorac Surg **40**: 507–511, 1987 (in Japanese).
18) Okada Y, Ottomo M, Yokoyama K, et al: Diagnosis of coronary sinus-left atrial window with cineangiography and two-dimensional echocardiography: Report of a case. Heart **16:** 949–954, 1984 (in Japanese).
19) Okumori M, Hyuga M, Ogata S, et al: Raghib's syndrome: A report of two cases. Jap J Surg **12**: 356–361, 1982.
20) Oosawa M, Morikawa T, Kawamura T, et al: Persistent left superior vena cava draining into the left atrium with absence of the left innominate vein. Jap J Thorac Surg **31**: 656–660, 1978 (in Japanese).
21) Quaegebeur J, Kirklin JW, Pacifico AD, et al: Surgical experience with unroofed coronary sinus. Ann Thorac Surg **27**: 418–425, 1978.
22) Raghib G, Ruttenberg HD, Anderson RC, et al: Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus. Circulation **31**: 906–918, 1965.
23) Shumacker HB Jr., King H, Waldhausen JA: Surgical implication with special reference to caval drainage into the left atrium. Ann Surg **165**: 797–805, 1967.
24) Takahashi K, Maruyama A, Momokawa K, et al: A surgical experience of coronary sinus atrial septal defect with coronary sinus defect and termination of left superior vena cava in left atrium. J Jap Associ Thorac Surg **33**: 123–129, 1985 (in Japanese).
25) Terada M, Iida Y, Tsuchiya K, et al: A case of coronary sinus type atrial septal defect with pulmonary valve stenosis. Jap J Thorac Surg **39**: 829–831, 1986 (in Japanese).
26) Tsuchiya K, Iida Y, Fujiwara T, et al: A surgical case of coronary sinus ASD. Jap J Thorac Surg **34**: 806–809, 1981 (in Japanese).
27) Yeager SB, Chin AJ, Sanders SP: Subxiphoid two-demensional echocardiographic diagnosis of coronary sinus septal defects. Am J Cardiol **54**: 686-687, 1984.

和文抄録

静脈洞型心房中隔欠損，部分肺静脈還流異常，左上大静脈遺残を伴った冠静脈洞型心房中隔欠損症の一例

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症例は3歳9カ月・女児．2歳10カ月時に心房中隔欠損症の診断のもとに手術を受けた．手術時，部分肺動脈還流異常を伴った静脈洞型心房中隔欠損と左上大静脈遺残及び冠静脈洞型心房中隔欠損が認められた．右前側方開胸・人為心室細動下では手術時間及び術野に制限があるため完全修復は困難と考え，右肺静脈及び冠静脈洞を左房に導くようにこれらの欠損部をパッチにて閉鎖した．術後軽度の左心不全，口唇チアノーゼが出現，左房に還流する左上大静脈は脳梗塞，脳膿瘍の危険もあるため，初回手術約1年後これを結紮した．術後の経過は良好である．一治験例を報告すると共に文献的考察を加える．