

Surgical Correction of Total Anomalous Systemic Venous Drainage with Ventricular Septal Defect and Pulmonary Stenosis—Report of a Case

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Anomalous total systemic venous drainage is a very rare malformation. Few cases have been reported with or without surgical treatment.^{3,8)}

The first case of successful surgical correction of total anomalous systemic venous connection was reported by G. MILLER et al. in 1965.⁶⁾ The combination of this anomaly and infrahepatic interruption of the inferior vena cava (IVC) with azygos continuation, ventricular septal defect (VSD) and pulmonary stenosis (PS) must be extremely uncommon. The following case is reported because of its rarity and the successful surgical correction.

Case report

I.K., a girl born on Sept. 18, 1973, a production of a normal spontaneous delivery, suffered from cyanosis and feeding difficulty since her birth.

In Mar. 1977, the girl was referred to our hospital for diagnosis because of severe cyanosis. Examination revealed a medium pitched ejection type systolic murmur, maximal at the right sternal border and second intercostal space. The first heart sound was slightly prominent and the second sound was single. The electrocardiogram showed a mean QRS frontal phase axis of $+105^\circ$, negative p in lead I, II and aVf and right ventricular hypertrophy¹²⁾. The aorta was arched on the left. Pulmonary vascularity was decreased. The stomach air lay on the left. The bronchial tree showed bilateral left sidedness (Fig. 1). The liver was 1.5 cm below the right costal margin^{7,9,10)}.

The blood pressure was 100/60. The peripheral arterial oxygen saturation was 72.8% with a hematocrit of 62.9%. RBC was $698 \times 10^4/\text{mm}^3$. No Heinz or Howell-Jolly body was found in the peripheral blood smears.

Cardiac catheterization led to a preoperative diagnosis of dextrocardia [P (I), D, N], partial anomalous systemic venous drainage (LSVC into LA, hepatic veins into RA), large ASD, VSD, valvular and infravalvular PS, absence of IVC and hemiazygos connection. (Fig. 2)

Key words: Total anomalous systemic venous drainage, Mesocardia, Pericardia baffle, Atrial compartmentalization, Valvulotomy.

索引語: 総体静脈還流異常症, 中胸心, 心外膜片, 心房中隔形成術, 弁輪切開術.

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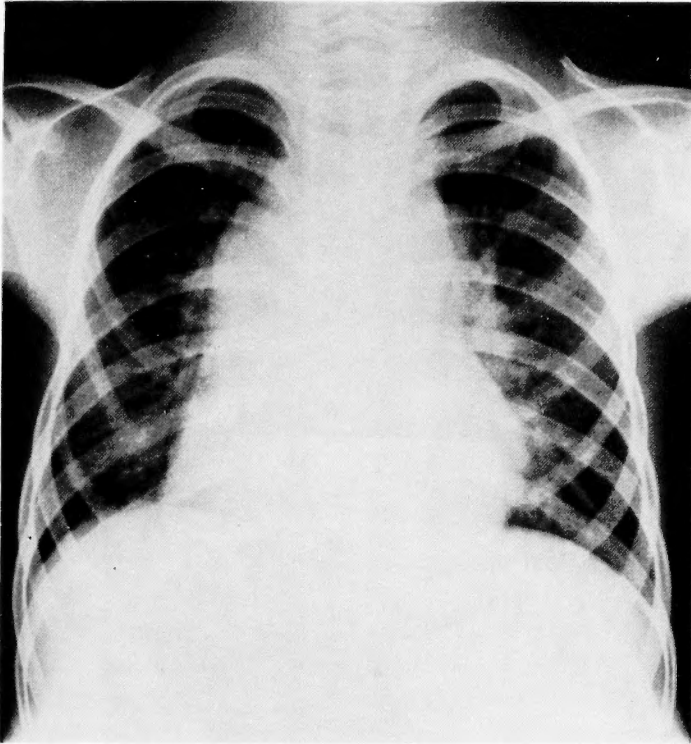


Fig. 1. Chest roentgengram shows moderate enlargement of the cardiac silhouette (CTR = 0.64), dextrocardia and decreased pulmonary vessels shadow.

Right Blalock-Taussig shunt operation was performed in February 1978 and cyanosis was greatly improved. Since May, 1980, however, cyanosis gradually advanced so that she was admitted again for radical operation.

Operative repair

The patient was operated on July 29, 1980. The heart was exposed through a median sternotomy incision.

The pericardium was taken for later autogenous patch. The right atrium with large perpendicular appendage lay posteriorly and on the right side of the heart. The right innominate vein joined with the left innominate and hemi-azygos veins and descended as a large SVC which entered the cephalad and left portion of the left atrium. The right SVC was absent. IVC was not recognized, though two hepatic veins entered the left atrium inferiorly. The right ventricle was anterior and positioned to the right with the left ventricle on the left and directed to the left. The external appearance was, therefore, like mesocardia.

The great vessels arose normally. Arterial cannulation was performed from right femoral artery with a No. 14 Bardic cannula.

The SVC cannula was introduced through the ASD and then cardiopulmonary bypass was instituted. The right Blalock-Taussig shunt artery established earlier was ligated. Core

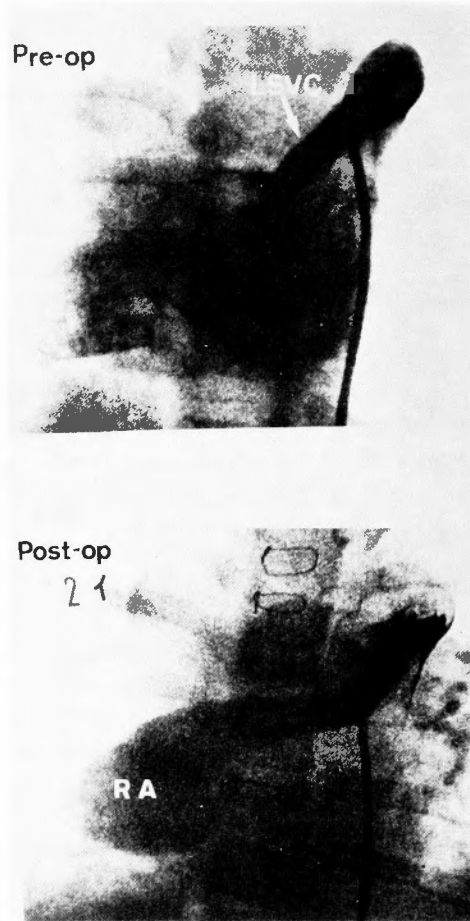


Fig. 2. (Upper): Pre-operative cineangiography revealed the hemiazygos vein joined the left SVC and entered the left atrium and the right atrium (through ASD).
(lower): Left SVC entered the right atrium after a successful atrial partition.

cooling was started. The patient's temperature was reduced to 28°C rectally and the heart was spontaneously fibrillated. Aorta was crossclamped and Young's solution was infused proximally.

The heart was brought to a standstill soon. Ice slush was poured around the heart. The right atrium was opened. Hepatic veins were then occluded with snares intermittently.

An atrial septal defect of a primum type, estimated at 1.5 cm in diameter was situated near the cephalic end of the septum.

The entire pulmonary and systemic veins opened into the left atrium. The coronary sinus ostium was to the left of the atrial septum (Fig. 3).

The tricuspid and mitral valves were free of anomalies. The atrial septum was widely excised. A pericardial baffle was then secured into position by continuous sutures so as to direct to the left SVC and the hepatic veins returned into the right atrium. All the pulmonary veins

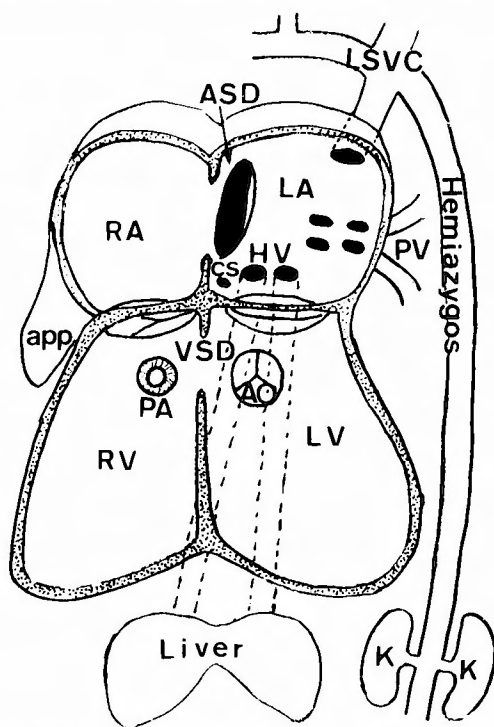


Fig. 3. Operative findings: mesocardia, D-loop, normal related great arteries. absence of IVC, hemiazygos vein connection, venous return of left SVC and hepatic veins into left atrium, ASD, VSD, valvular and infravalvular PS. RA: right atrium, App: right atrial appendage, RV: right ventricle, LA: left atrium, LV: left ventricle, PV: pulmonary veins, HV: hepatic veins, CS: coronary sinus, Ao: Aorta, PA: pulmonary vein. K: Kidney, LSVC: left superior vena caav, VSD: ventricular septal defect.

drained to the posterior of the baffle and directly through the mitral valve. The coronary sinus was left draining into the left side.

After constitution of the new atrial partition, the left ventricle was vented. Despite this vent, the left atrium was still inflated, so the right superior pulmonary vein was opened. Then the right ventricle was incised and VSD was confirmed.

VSD was infundibular form. situated below the crista supraventricularis, 1.5 cm × 1.5 cm in size and round in shape. It was patch closed (Fig. 3).

Infundibular muscle was cut away with scissors. There was adhesion between the anterior and posterior leaflets of the pulmonary valve. Commissurotomy was performed. The baffle of the pericardium was sutured to the inner surface of a Gore Tex patch of 1.5 cm in diameter in order to form a monocusp. Then the patch and monocusp were sutured into the incision site of PA and RV in order to enlarge the outflow tract. The right atrial incision was then sutured. Rewarming was commenced and good cardiac contraction spontaneously occurred. The incision wound of the right superior pulmonary vein was closed by sutures. The left ventricular vent was then removed.

There was no A-V block and left atrial pressure was about 15 mmHg.

The cardiopulmonary bypass was weaned gradually and successfully.

Pressures were measured. The left ventricular pressure was 75/5 mmHg, RV inflow 40/5 and PA 30/10.

The post-operative diagnosis was somewhat different from the pre-operative one. It was mesocardia. [P (A), D, N], i.e. polysplenia syndrome, situs ambiguus, D-loop and normal related great arteries.

Post-operative course

The patient was extubated after 14 hours. Blood loss was minimal. She recovered her health uneventfully. Post-operative cardiac catheterization was carried out on Sept. 12 1980, 45 days after operation. Except for a small residual VSD, the operation was rather successful as a whole (Fig. 4).

In the following up examination. (Nov. 19, 1982), her complexion was excellent without

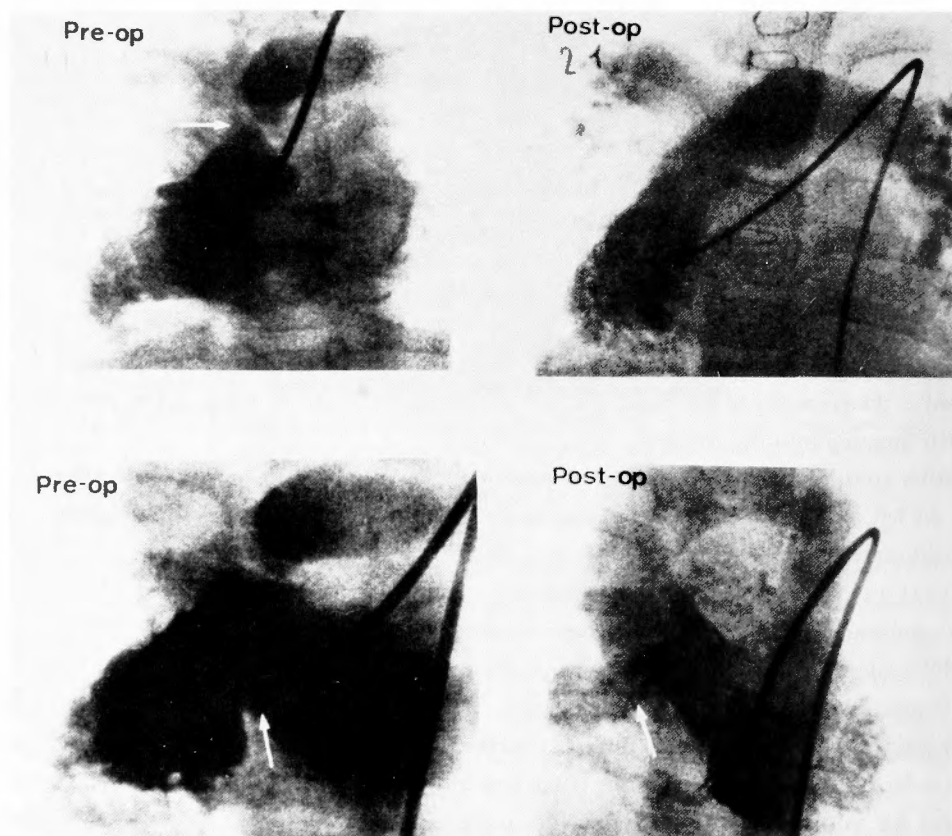


Fig. 4. (Left upper): Pre-operative cineangiography revealed valvular and infravalvular pulmonary stenosis.
(Right upper): Pulmonary stenosis was relieved after pulmonary valvulotomy and right ventricle outflow patch reconstruction.
(Left lower): Pre-operative left ventriculography revealed large VSD.
(Right lower): A small post-operative residual VSD could be found.

clinical cyanosis and she tolerated exercise normally.

There was no polycythemia. The girl obtained significant improvement showing no clinical symptoms and gradual improvement of clubbed fingers.

Discussion

The present case appears unique in that both total anomalous systemic venous drainage and infrahepatic interruption of IVC were present.^{1,2)} It also differs from the cases of successful correction previously reported in that no other cardiac malformation were present. To the best of our knowledge, this is the first case report.

The embryologic explanation of the anomaly is no better than speculation, but presumably the anatomic state reflects persistences of both anterior cardinal veins with abnormal development of the posterior and anterior cardinal veins on the left side. The right SVC is involute leaving a left SVC.^{5,11)}

Absence of IVC occurs when the right subcardinal vein fails to connect with the right vitelline veins. The hepatic veins then empty directly into the atrium, probably because of persistence of the left vitelline veins with atrophy of right vitelline veins. Systemic venous blood from below the diaphragm shunts directly into the right supra-cardinal vein (azygos vein). This vessel in turn empties into left supra-cardinal (hemiazygos) vein and then into left posterior and common cardinal vein (LSVC).

Communication of the coronary sinus and the left atrium may be considered as a form of levo-atrio-cardinal vein, because of persistence of the left sinus horn with partial atrophy of the right horn⁴⁾

Complete correction was achieved with excellent results by excision of the interatrial septum and reconstruction of the normal anatomy by using a pericardial baffle in order to separate the pulmonary drainage from the systemic venous flow.

The most difficult decision in correcting a complex case of systemic venous drainage is making choice of cannulation technique. In our case, hepatic veins were intermittently occluded to provide a bloodless operative field for the correcting procedure.

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和文抄録

総体静脈還流異常, VSD, PS を主体とする 複雑心奇型の一治験例

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チアノーゼを伴う体静脈還流異常・心室中隔欠損・肺動脈狭窄を主体とする複雑心奇型の一手術成功例を報告する。

症例は7才の女児で、主訴は生来のチアノーゼと Squatting である。患児は満期安産で生下時体重2900 grams. 生後二日目よりチアノーゼ出現、嘔吐及び哺乳困難を来たし、心疾患を指摘された。その後、内服治療を続けていたが、3才6カ月時精査のために当院に来院した。4才5カ月時右 Blalock-Taussing 手術を受け、チアノーゼは改善した。しかし6才8カ月時から再度チアノーゼが増強したため、6才10カ月時、今回の根治手術を行なった。

術前諸検査及び五回の心臓カテーテル検査で、心室中隔欠損、肺動脈弁狭窄及び円錐狭窄を認め、また、右上大静脈は左上大静脈と合流して一本となり左房に還流する複雑心奇形と診断した。

手術は中等度低体温で Young 氏液による心停止下

に行ない、脱血管は右房側より左上大静脈へ一本挿入、肝静脈は間歇的遮断を行なった。まず心外膜パッチで心房の compartmentalization を施行、即ち、左上大静脈・肝静脈・冠状静脈洞はすべて右房側に還流するようにパッチを縫合した。心室中隔欠損はⅢ型で、これをダックロンパッチで閉鎖し、最後に肺動脈狭窄に対して outflow patch による拡大術を行ない、手術を終了した。

術直後一時消化管出血を来したが、血行動態は比較的安定していた。術後48日目に心臓カテーテル検査を施行し、Small residual VSD を認めたが、体静脈系は右心房に、肺静脈は左心房にそれぞれ還流し、上大静脈と右心房間に圧差を認めなかった。肺動脈狭窄は十分解除され、右心室収縮圧は 1000 mmHg から 50 mmHg に低下していた。術後2カ月で退院し、二年を経過した現在元気で通学している。