Drainage of Right Superior Vena Cava Into Both Atria

Review of the Literature and Description of a Case Presenting with Polycythemia and Paradoxical Embolization

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SUMMARY **An unusual cause of polycythemia, cyanosis and paradoxical embolus is described in a 37-year-old man, consisting of a rare congenital anomaly of the superior vena cava (SVC). The right-sided SVC received pulmonary venous drainage from the right lung and drained, through two channels, into both atria with the left atrium receiving the larger of the two channels. The atrial septum was intact. Corrective surgery and postoperative cardiac catheterization are described and the literature concerning anomalies of the right superior vena cava is reviewed.**

THE OCCURRENCE of cyanosis in the absence of clinically detectable abnormalities of the cardiovascular system usually indicates pulmonary disease or unusual hemoglobinopathy.[1] Isolated abnormalities of the vena cava, however, can also present in this manner. In this report we describe a patient with connections of the superior vena cava (SVC) to both right and left atria, a condition not previously described in the English-language medical literature.

Case Report

A 37-year-old man was admitted to the University of Chicago Hospitals because of intermittent chest pain. His health had been satisfactory except for mild dyspnea on exertion from childhood and occasional left chest discomfort for several years. He had no risk factors for coronary artery disease other than a 40-pack-year history of smoking. He had a long-standing history of heavy alcohol intake. There was no family history of congenital heart disease.

On physical examination the patient was a normally developed man. There was mild clubbing of the fingers. Jugular venous pressure and peripheral pulses were normal. He showed mild left ventricular enlargement; the right ventricle (RV) was not enlarged. On auscultation, the second heart sound split normally. There was a grade 1/6 systolic ejection murmur at the left sternal edge. The remainder of the clinical examination was normal.

The ECG showed left ventricular hypertrophy by voltage criteria. Chest x-ray demonstrated mild cardiomegaly with normal lung fields. On room air, arterial P_{O_2} was 53 mm Hg, P_{CO_2} was 26 mm Hg, and pH was 7.42. The P_{O_2} rose to only 75 mm Hg on breathing near 100% oxygen by mask. The hematocrit was 57%. The white cell and platelet counts were normal.

Serum met- and sulfhemoglobinema were not present on spectroscopic examination, and hemoglobin electrophoresis was normal. Significant chronic lung disease was excluded by pulmonary function tests, which showed only minimally reduced vital capacity.

Shunting of blood from the right to left was suspected and was inadvertently confirmed on the third hospital day, when the flushing of an i.v. line resulted in transient right hemiparesis and aphasia. A pulmonary scan then demonstrated uptake of tracer by the thyroid and kidneys, documenting the presence of a right-to-left shunt.

Cardiac catheterization was performed through the right median antecubital vein and right femoral artery (table 1). The right-heart catheter passed readily from the SVC through the right atrium (RA) and the RV to the pulmonary artery (PA). Right atrial and right ventricular end-diastolic pressures were slightly increased. A step-up in oxygen saturation occurred at the level of the low SVC; this was attributed to an anomalous right upper lobe pulmonary vein, which could be entered with the catheter. There was also mild systemic arterial desaturation, with an oxygen saturation in the left ventricle of 92–96% (table 1). These results were indicative of a bidirectional shunt with a left-to-right flow of 1.9–2.5 l/min, and with a right-to-left shunt ranging from 0.5–1.3 l/min. PA pressures were normal. The findings were initially attributed to a sinus venosus atrial septal defect with partial anomalous pulmonary venous drainage. However, at no stage during the procedure did the catheter cross the atrial septum.

The coronary arteries were normal. Left ventriculography showed moderate prolapse of the posterior mitral valve leaflet and mild diffuse hypokinesis during a postpremature ventricular complex, with an ejection fraction of 63%. This mild left ventricular dysfunction could be secondary to chronic

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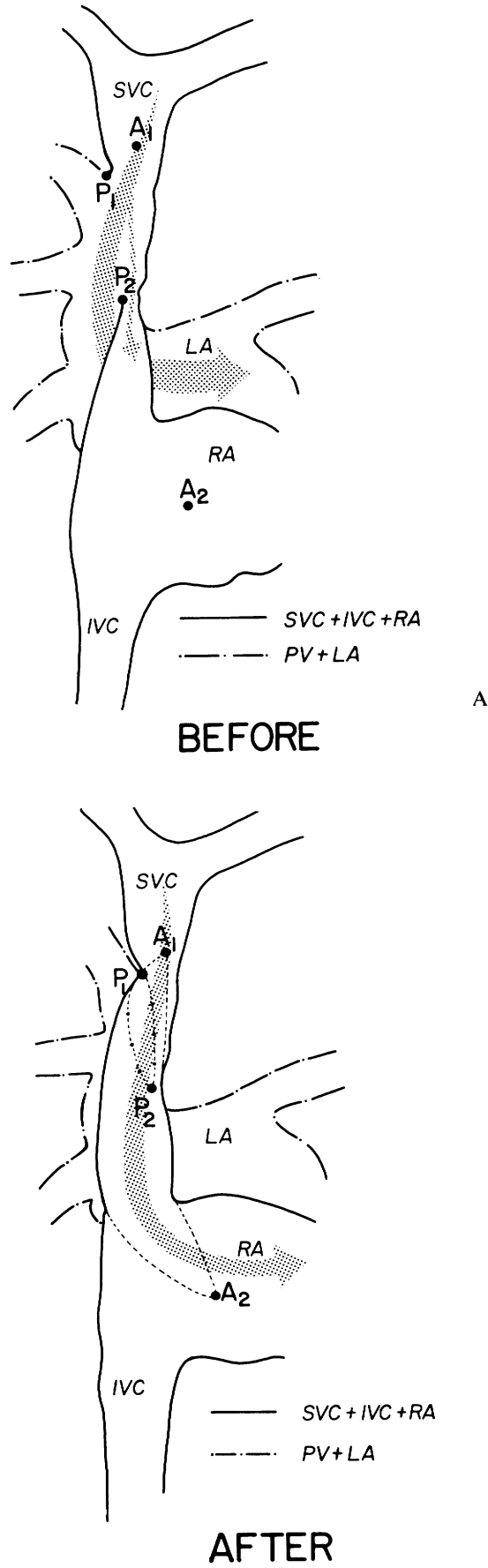

FIGURE 1. *(A) Before surgical correction, much of the superior vena cava (SVC) flow was directed posteriorly between points P_1 and P_2 and into the left atrium (LA), accounting for the right-to-left shunt. Pulmonary venous drainage entered this abnormal segment of the SVC, causing left-to-right shunting. (B) At surgery, a small posterior patch between P_1 and P_2 divided the SVC and forced all of its flow toward the right atrium (RA), while the pulmonary venous drainage was directed entirely toward the LA. A large anterior patch between A_1 and A_2 enlarged the stenotic SVC-RA junction. IVC = inferior vena cava; PV = pulmonary veins.*

heavy alcohol intake, mitral valve prolapse, or both. Pulmonary angiography revealed early filling of the right side of the heart.

The patient was taken for corrective cardiac surgery. At operation (fig. 1A), anomalous pulmonary veins draining into the low SVC were noted. The SVC was on the right side; an anomalous left-sided SVC was not present. During attempted SVC cannulation a stenotic SVC-RA junction was identified. Direct cannulation of the SVC was then performed at the innominate junction. The high azygos vein was ligated, and after onset of extracorporeal circulation the SVC was clamped above the pulmonary vein entrances.

An incision of the RA and SVC across the stenotic portion (figs. 1A, A_1 to A_2) was made. An atrial septal defect was not present. The major drainage of the SVC was into the left atrium (LA) between P_1 and P_2 (fig. 1A).

Once the anomalous SVC course was demonstrated, the repair involved separation of the SVC from the LA by means of a short, 3-cm Edwards patch

TABLE 1. *Preoperative Catheterization Data*

Hemodynamic and oxygen data	Oxygen saturation (%)	Pressure (mm Hg)
Superior vena cava High	69	—
Low	81	—
Inferior vena cava	78	—
Right atrium	79	10 (mean)
Right ventricle	77	27/10
Pulmonary artery	80–82	23/12
Pulmonary wedge	100	10 (mean)
Anomalous pulmonary vein	100	
Left ventricle	92–96	95/12
Flow (l/min)	Early*	Late*
Systemic blood flow	5.3	4.6
Pulmonary blood flow	5.9	6.6
Left-to-right shunt	1.9	2.5
Right-to-left shunt	1.3	0.5
Pulmonary/systemic flow	1.1/1	1.4/1

*Denotes initial values and those determined later during the catheterization procedure.

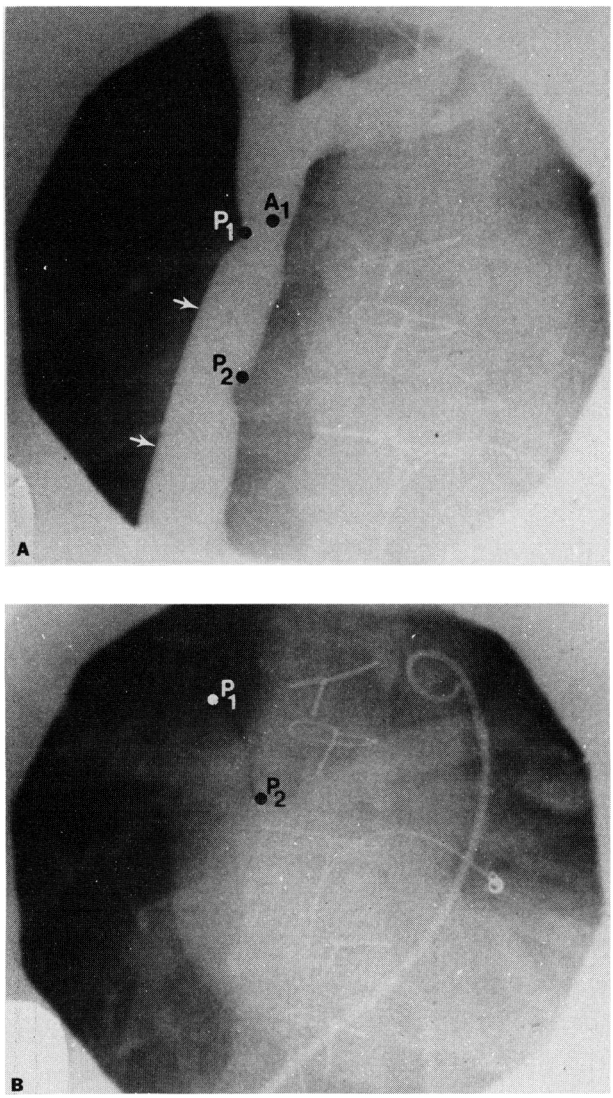

FIGURE 2. *After surgical correction, a superior vena cava injection (A) demonstrated unobstructed drainage into the right atrium. The levophase of a pulmonary artery injection shows (B) entrance of the right upper, middle and lower pulmonary veins and left upper pulmonary vein into the left atrium. The posterior baffle, which separates the anomalous superior vena cava from the left atrium, extends from P_1 to P_2. The anterior baffle, which enlarges the stenotic connection of the superior vena cava to the right atrium, begins at point A_1. Its edge is marked by arrows.*

posteriorly (P_1–P_2) and enlargement of the SVC-RA junction by another 10-cm Edwards patch anteriorly (A_1 to A_2). Oxygen saturations from the SVC and the PA at the end of operation revealed no step-up, and left atrial blood was fully saturated.

The postoperative course was uneventful.

Cardiac catheterization was repeated 6 weeks after surgery, this time through the right femoral vein and artery. Right- and left-sided pressures were within normal limits. There was no step-up in oxygen saturation across the right-heart chambers. The cardiac output was normal, at 6.5 l/min, with a cardiac index of 3.2 l/min/m^2. The hematocrit was 35%. The P_{O_2} was 89 mm Hg on room air. Injection of contrast into the SVC showed drainage of all SVC blood into the RA (fig. 2A). The levophase of a pulmonary angiogram showed normal pulmonary venous drainage into the LA without evidence of left-to-right shunting (fig. 2B). The patient had mild diffuse hypokinesis and a normal sinus beat ejection fraction of 55%. Prolapse of the posterior mitral valve leaflet was again noted.

One year later, the patient remained well, without dyspnea or chest pain that prompted his initial investigation.

Discussion

Cyanosis may occur as a result of abnormal systemic venous connections. The right or left SVC, the inferior vena cava, or the hepatic veins may drain alone or in combination into the LA.[2] This usually occurs in association with additional congenital malformations of the heart, most often atrial or ventricular septal defects, tetralogy of Fallot, pulmonary atresia, transposition of the great vessels and patent ductus arteriosus.[2] Of these, atrial septal defect is the most common. Anomalous systemic venous drainage may occur without other intracardiac abnormalities. Drainage of a persistent left SVC into the LA has been described as an isolated finding without an associated septal defect,[3, 4] as has drainage of the inferior vena cava into the LA.[5]

Seven cases of drainage of a right-sided SVC into the LA have been reported.[6–12] Some of the findings in these cases are summarized in table 2. The most common presentation was cyanosis in childhood. Symptoms of declining intellectual capacity,[6] mild effort intolerance and fatigue were also observed. Development was normal except for the case of Braudo et al.[9] Examination of the heart was often normal, although three patients had left ventricular enlargement and two had soft systolic murmurs. In only one, who also had a dilated SVC, was the jugular venous pressure elevated.[9] Chest x-ray and ECG were normal apart from left ventricular enlargement in three patients and evidence of an old inferior wall myocardial infarction in one patient. Diagnosis in all patients was by cardiac catheterization. All patients had systemic arterial desaturation and polycythemia. Four were operated on; two died as a result of the procedure. None of the patients had associated intracardiac anomalies. The drainage of the pulmonary veins was normal (or was not mentioned) in four of the patients. In two patients the right upper lobe veins drained into the area of junction between the right SVC and the LA.[9, 11] One patient[8] had connections between the right pulmonary veins and the SVC.

Our case is different in that connections of the SVC to both atria were present. Five similar patients have been reported in the French and German literature.[13–17] The clinical features are shown in table 3. Two patients presented in childhood; one died with se-

TABLE 2. *Reported Cases of Isolated Right Superior Vena Cava Draining to Left Atrium*

Authors	Sex	Age (years)	Presentation	Physical findings: Cyanosis	Clubbing	Cardiovascular	Operation	Additional factors
Tuchman et al[6]	M	15	Cyanosis from infancy; fatigue intellectual decline	Yes	No	Normal	Patient died; details not given	LVH at autopsy
Wood[7]	F	10	Cyanosis from infancy; mild effort intolerance	Yes	Not known	Enlarged LV; S_2 single	Not done	
Kirsch et al.[8]	F	2	Cyanosis from infancy	Yes	Yes	Normal	SVC divided and anastomosed to RA	Two small anastomoses between right pulmonary veins and SVC
Braudo et al.[9]	F	3	Cyanosis from birth; physical development below third percentile	Yes	Yes	Prominent neck veins; soft murmur; apical ejection click	SVC divided and anastomosed to RA	Right upper pulmonary vein drained to saccular dilatation of lower SVC
Park et al.[10]	F	34	Palpitations; dyspnea 1 yr	Yes	No	Normal	Not done	
Ezekowitz et al[11]	M	52	Dyspnea, light headedness; intellectual decline; cyanosis	Yes	Yes	Enlarged LV; S_2 single	Pericardial patch to divert blood from SVC to RA; patient died from septicemia	LVH and old inferior wall MI; apparent sinus venosus chamber leading to LA; Two right upper pulmonary veins entered this chamber
Vazquez-Perez and Frontera-Izquierdo[12]	M	7	Cyanosis from birth	Yes	No	Enlarged LV; soft systolic murmur	Not done	Reduced pulmonary blood flow on chest x-ray; left-axis deviation and LVH on ECG; PFO

Abbreviations: LV = left ventricle; SVC = superior vena cava; RA = right atrium; LVH = left ventricular hypertrophy; MI = myocardial infarction; LA = left atrium; PFO = patent foramen ovale.

vere cyanosis and congestive heart failure after unsuccessful surgery. Three patients presented relatively late in adult life and were managed medically.

Schematic diagrams of these five cases and the present report are shown in figure 3. They differ from each other in the location of entry of the right pulmonary veins and in the relative size of the caval connections to each atrium.

The hemodynamic consequences of a vena cava connected to both atria depend on the place of entry of the anomalous pulmonary veins, the relative size of the channels to right and left atria, and the pressures in the atria. Variations in these factors will result in different degrees of right-to-left and left-to-right shunting. This will dictate the clinical presentation. In our case, the larger terminal part of the SVC ran to the LA and the connection to the RA was smaller. However, pressures in both atria were essentially identical. Thus minor changes in atrial pressure could have resulted in significant alterations in shunt flow. In fact, respiration alone might alter bidirectional shunting. During cardiac catheterization, marked changes in right and left shunting were observed (table 1).

Kirsch et al.[8] and Braudo et al.[9] suggested that drainage of a right SVC to the LA was most probably caused by malposition of the right horn of the sinus venosus.[8, 9] Enjalbert et al.[16] proposed a similar mechanism for those cases with connections to both atria. However, because there is no evidence for abnormal structure or orientation of the interatrial septum, an alternative embryologic explanation can be suggested. Pulmonary venous channels form within the primitive lung tissue and connect secondarily to the common pulmonary vein, which forms as an outpouching of the posterior wall of the common primitive atrium. Inappropriate connections of one or all pulmonary veins to the right SVC or other vascular structures are not uncommon. Occasionally, a lung lobe has venous connections to both the LA and the SVC. Our case can be viewed as such a double connection, possibly from the right upper pulmonary vein, in which the channel to the SVC was large enough and directed such that systemic venous drainage could shunt to the LA. The normal SVC channel to the RA then became relatively hypoplastic. Thus, these connecting channels may be of pulmonary venous origin.

The importance of this case report lies in the recognition of another form of surgically correctable

TABLE 3. *Reported Cases of Right Superior Vena Cava with Connections to Both Right and Left Atria*

Author	Sex	Age (years)	Presentation	Physical findings	Operation	Additional factors
Nutzel[13]	M	47	Congestive heart failure for 3 years	Congestive heart failure; systolic murmur	Not done	Mitral valve stenosed and thickened; RVH
Hackensellner[14]	F	72	Thrombosis of abdominal aorta	Not given	Not done	Thrombosis of abdominal aorta; peritonitis; mitral stenosis; RVH
Breitfellner[15] (case A)	F	74	Congestive heart failure for 6 months	Not given	Not done	PFO; mitral annular calcification; RVH
Enjalbert et al.[16]	F	7	Cyanosis from birth	Cyanosis; clubbing thrill and systolic ejection murmur	Tricuspid valve dilatation; teflon patch to divert blood from the pulmonary veins to the LA.	PFO; abnormal tricuspid valve; gross tricuspid regurgitation developed, causing more marked cyanosis by shunting of blood through the abnormal connection between the RSVC and LA; narrowing at the point of entry of the RSVC into the RA; ultimately died in heart failure.
Laboux et al.[17] (case 1)	F	9	Not mentioned	3/6 pulmonary systolic murmur	Tied off abnormal connection to LA, allowing blood flow solely to the RA.	Incomplete right bundle branch block on ECG with moderate RVH.
Present case	M	37	Intermittant chest pain, mild dyspnea; paradoxical embolus	Mild clubbing; slight LV enlargement	See text	LVH by voltage on ECG; stenosis of SVC-RA junction.

For schematic representation of the anomalies, see figure 3. All patients had partial anomalous pulmonary venous drainage and connections between the RSVC and both atria.

Abbreviations: LV = left ventricular; RVH = right ventricular hypertrophy; PFO = patent foramen ovale; SVC = superior vena cava; RSVC = right superior vena cava; LA = left atrium; RA = right atrium; LVH = left ventricular hypertrophy.

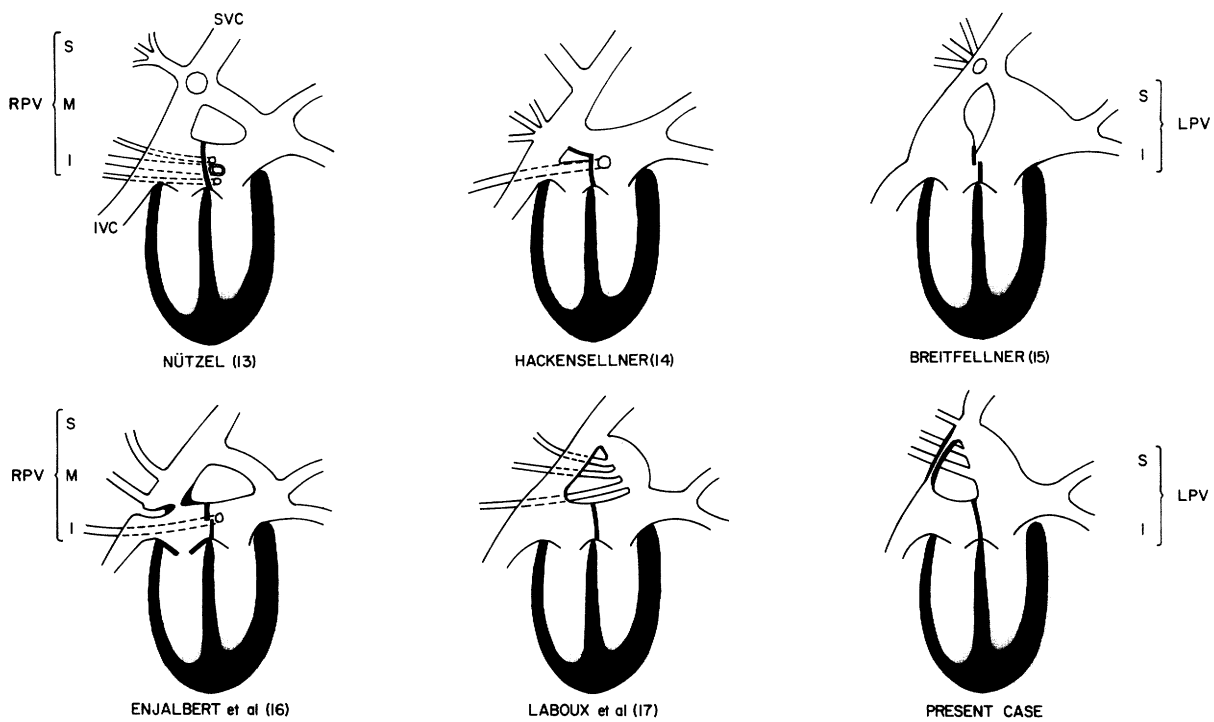

FIGURE 3. *Diagram of all reported cases of connection of the right superior vena cava to both atria. Note that there are abnormalities of pulmonary venous drainage from the right lung in all cases. RPV = right pulmonary veins; LPV = left pulmonary veins.*

heart disease that may produce cyanosis or significant left-to-right shunting of blood. All three patients[16, 17] (including the present case) with connections of the SVC to both atria were inaccurately diagnosed by cardiac catheterization. The diagnosis was assumed in each case to include an atrial septal defect of the sinus venosus type, though the atrial septum was not crossed. This preoperative error would have been avoided if an SVC angiogram had been performed at the time of catheterization. Accurate delineation of the caval connections is mandatory in any cyanotic patient with normal right-sided pressures, particularly if the atrial septum cannot be crossed. Nuclear angiography has been recently advanced as a means of diagnosing anomalous systemic venous drainage[10, 11] and this might have been applicable to our patient. Fortunately, intraoperative diagnosis and successful surgical correction are possible in this very rare congenital abnormality.

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