Isolated Drainage of the Superior Vena Cava into the Left Atrium in a 52-Year-Old Man

A Rare Congenital Malformation in the Adult Presenting with Cyanosis, Polycythemia, and an Unsuccessful Lung Scan

MICHAEL D. EZEKOWITZ, M.R.C.P., D. Phil., PHILIP O. ALDERSON, M.D., BERNADINE H. BULKLEY, M.D., PATRICK N. DWYER, M.D., LEVI WATKINS, M.D., DONALD L. LAPPE, M.D., H. LEON GREENE, M.D., AND LEWIS C. BECKER, M.D.

SUMMARY This report describes a 52-year-old black male with the isolated finding of an anomalous superior vena cava draining into the left atrium. The patient presented with dizziness, mild cyanosis, polycythemia and normal cardiac and pulmonary findings. The first major diagnostic clue in this confusing clinical presentation was an unsuccessful lung perfusion scan in which intravenous tracer consistently bypassed the lungs. This appears to be the first adult presenting with this rare anomaly. This condition should be suspected if cyanosis, clubbing, and a "normal" cardiac examination coexist and if the more common pulmonary and hematological causes of this triad have been excluded.

IT IS UNUSUAL TO FIND the superior vena cava entering the left atrium, particularly in the absence of other, more complex cardiac and extracardiac anomalies. DeLeval et al. reviewed 5,127 consecutive cases of congenital cardiac anomalies and found only 28 patients with the superior vena cava draining into the left atrium, all of whom had associated cardiac anomalies. Four cases of the superior vena cava entering the left atrium as an isolated finding have been described in the literature. All were in children and adolescents. Tuchman et al. described a 15-year-old male who succumbed 20 hours after surgical repair. Kirsch et al. reported a 2-year-old girl who did well following surgery. Paul Wood reported a 10-year-old girl with this abnormality identified at cardiac catheterization who did not undergo surgery, and Braudo described a 3-year-old girl who survived surgery. This report describes a 52-year-old man with isolated drainage of the superior vena cava into the left atrium — an anomaly virtually unheard of in the adult.

Case Report

A 52-year-old man was admitted to The Johns Hopkins Hospital because of longstanding shortness of breath and lightheadedness which became progressively severe in the months before his admission. Past medical history included a normal infancy and childhood without major illnesses; however, his mother noted him to be cyanotic from a young age. The patient's wife reported a 10-year history of decreasing memory and attentiveness. Twenty months before this admission, the patient had a well-documented, uncomplicated inferior myocardial infarction with subsequent vague, intermittent chest pain that did not appear to be ischemic in etiology. Although heart failure was never present, six months before admission digoxin and lasix were started because of shortness of breath and lightheadedness. He was found at another hospital to have a hematocrit of 64.5 with an erythropoietin level twice normal. A primary etiology for the polycythemia was not found.

Physical examination on admission to this hospital revealed a normally developed man with striking central cyanosis and clubbing of the fingers and toes. His blood pressure was 110/86 and respirations normal. All pulses were palpable and normal. The jugular venous pressure was not elevated. The cardiac apex was located in the sixth intercostal space just lateral to the midclavicular line, and was slightly thrusting in nature. On auscultation the only notable finding was a single second heart sound. No murmurs were heard and the lungs were clear. The retinae apart from minor venous engorgement were within normal limits. The abdomen was unremarkable and showed no liver or splenic enlargement. There was no stigmata of chronic liver disease. Apart from reduced cognitive function the central nervous system was normal.

Laboratory findings included an electrocardiogram showing normal sinus rhythm with significant Q waves in leads 2, 3, and aVF with T wave inversion consistent with the old inferior infarct. Chest radiograph revealed a tortuous aorta with normal-sized heart and unremarkable lung fields. The hemoglobin was 19.2, hematocrit 60, platelet count 266,000, white cell count 5,200, and blood smear normal. Serum met- and sulf-hemoglobinemia were not detected by spectroscopic examination, and hemoglobin electrophoresis was normal. Respiratory function tests showed a FVC of

From the Cardiovascular Division of the Department of Medicine and the Cardiac Surgery Division of the Department of Surgery, The Johns Hopkins Hospital, Baltimore, Maryland.

Address for reprints: Michael D. Ezekowitz, M.D., Department of Medicine, The University of Oklahoma Health Science Center, Cardiology Section, P.O. Box 26901, Oklahoma City, Oklahoma 73190.

Received May 1, 1978; accepted June 23, 1978.
Circulation 58, No. 4, 1978.
2.85 (69% of expected) and FEV1/FVC of 78%, and a normal carbon monoxide diffusion coefficient of 27.8 ml/m/mm Hg. Exercise electrocardiogram was normal and adequate (heart rate 175; target 156, 90% of maximum). Exercise-induced unifocal PVCs were seen. An echocardiogram was normal.

Blood Gases at Rest and Exercise

Arterial blood gases on room air, at rest, showed a pH of 7.40, Pco2 of 37 mm Hg and a Po2 of 62 mm Hg. After the anomaly was diagnosed, the effect of exercise of the upper and lower limbs on the arterial blood gases was determined by giving the patient 100% oxygen for 5 minutes before exercising the arms and legs in turn. After arm exercise the Po2 dropped from 151 to 60, with no change in Pco2 and pH. On exercising the legs the Po2 and Pco2 remained unchanged at 155 mm Hg and 34 mm Hg, respectively.

Isotope Studies

Since occult pulmonary embolism was a possible cause for the patient's systemic oxygen desaturation, a ventilation-perfusion lung scan was ordered. A 133Xe ventilation study was performed before the perfusion scan and was normal. Then, with the patient in the supine position, a standard 3 mCi dose of 99mTc-labelled human albumin microspheres (10-30 μ diameter, approximately 500,000 particles) was injected intravenously from the right antecubital fossa. The distribution of activity was initially imaged on a standard low-resolution persistence oscilloscope, and was localized to the abdomen rather than the lungs. An assay of radiopharmaceutical supplies revealed that the patient's dose was drawn from a multi-dose microsphere vial which had produced technically adequate lung scans in several other patients injected earlier in the day. A 35 mm abdominal image was obtained and revealed that the activity was localized primarily in the kidneys (fig. 1). An additional image taken over the head revealed intracerebral activity. The findings of systemic particle deposition suggested a right-to-left vascular shunt bypassing the pulmonary capillary bed. Therefore, the next day a radionuclide angiocardiogram was performed following a right antecubital intravenous bolus injection of 20 mCi of 99mTc-albumin. Images were obtained at the rate of 1/sec over the chest. The study showed activity first in the superior vena cava (6 sec) and then in the left ventricle (8 sec) and aorta (12 sec), without evidence of right heart or pulmonary perfusion (fig. 2). The precise anatomy of the right-to-left shunt could not be determined, so a cardiac catheterization was scheduled.

Cardiac Catheterization

A catheter introduced into the right femoral vein was passed through the inferior vena cava into the right atrium without any difficulty. Repeated attempts...
to enter the superior vena cava and to cross the atrial septum were unsuccessful. The catheter was advanced from the right atrium into the right ventricle and into both pulmonary arteries. Right sided pressures were normal and oxygen saturations were 65% in all right sided chambers, thus excluding a significant left-to-right shunt. Green dye curves were performed from the main pulmonary artery to the femoral artery and from the inferior vena cava to the femoral artery during Valsalva maneuver. The appearance time of all these curves was normal at 11 seconds, cardiac output was 6.2 l/min and the dye curves showed no early recirculation. These findings excluded a significant left-to-right or right-to-left shunt from these chambers. A second catheter from the right brachial vein was advanced to the superior vena cava where the saturation was 59%. It passed freely into the left atrium where the saturation was 71% and the mean pressure was 6 mm Hg. Green dye injected into the superior vena cava appeared early at the femoral artery at 6 seconds. A third catheter was passed retrogradely from the femoral artery to the ascending aorta where pressures of 100/64 mm Hg were recorded. No gradients were noted. The left ventricle was entered; pressure was 100/0–12 mm Hg; oxygen saturation 71%. A biplane angiogram was performed with dye injected into the superior vena cava (fig. 3). The superior vena cava, followed by the left atrium, left ventricle and then aorta, were opacified. There was no communication with any right-sided structure. A right anterior oblique ventriculogram showed good overall left ventricular function with inferior wall akinesis. The calculated ejection fraction was 56%, and the end-diastolic volume was 100 ml. Selected coronary arteriography demonstrated a total occlusion of the proximal right coronary artery with a good distal vessel. The left coronary system was normal and supplied left-to-right collaterals to the distal right coronary artery. The coronary sinus drained normally into the right atrium.

A diagnosis of isolated drainage of the superior vena cava into the left atrium was made, and the patient was submitted to surgery.

Surgery (fig. 4)

Monitoring lines were inserted, avoiding veins of the upper extremities. The chest was opened through a median sternotomy incision. The extrapericardial portion of the superior vena cava was in the normal position. The pericardium was opened and a portion was removed in anticipation of using the pericardium as a patch. The intrapericardial portion of the superior vena cava drained into an outpouching that externally had the appearance of a sinus venosus.
chamber. No sinus venosus defect was present. Two small superior pulmonary veins on the right drained into this chamber. The inferior vena cava and left pulmonary vein drained normally. The right atrium was opened. There was no atrial septal defect. The atrial septum was incised superiorly with the incision extending into the superior vena cava. A pericardial patch was fashioned so as to divert the superior vena caval blood into the right atrium and the pulmonary venous return into the left atrium. The right atrium was also patched with the pericardium to increase its size. A saphenous vein bypass graft of the right coronary artery was performed in the usual way after completion of the atrial construction. Difficulty was experienced weaning the patient off the bypass pump. After using the intraaortic balloon and inotropic agents, he was successfully separated from the bypass pump. His hemodynamic status improved. By the fifth postoperative day the intraaortic balloon was successfully removed. The patient, however, had sustained acute renal failure early in his postoperative course, requiring hemodialysis. While renal function was improving, he became septicemic and died three weeks after the operation.

Autopsy Findings

At autopsy the heart weighed 500 g. Its surface was covered by fibrinous epicarditis that showed no signs of infection. The right atrium and right ventricle were of normal size without hypertrophy or dilatation. The left ventricle was hypertrophied, but there was no evidence of left ventricular dilatation. A transmural inferior myocardial infarction 2 cm wide was present. There was no myocardial necrosis or hemorrhage. The old infarct was in the distribution of the severely narrowed right coronary artery. The remainder of the coronary tree was widely patent. A saphenous vein bypass graft had been placed in the posterior descending coronary artery and was patent. The reconstruction of the atria was intact. An adequately sized superior vena cava, which had been widened with a graft, drained into the right atrium. The interatrial septal patch was intact without evidence of leak. Aside from the abnormality of the superior vena cava drainage there was no other congenital anomaly of the heart. Specifically, there was no evidence of coarctation, patent ductus arteriosus or other intracardiac shunts. The coronary sinus was present at the usual location and there were no other anomalies of venous connection. Examination of the lungs revealed an extensive bronchopneumonia which was the likely cause of death. There was no evidence of pulmonary embolus or pulmonary arteriovenous fistulae. The liver was congested and showed central lobular necrosis (compatible with low output syndrome). The spleen was passively congested and had multiple small healing infarcts.

Discussion

This patient's presentation and the mode of diagnosis was unusual. He is the fifth reported case of isolated drainage of the superior vena cava into the left atrium, and the first adult described with this lesion. Findings in our patient and the four previously reported are summarized in table 1 and provide some understanding of the natural history of this rare anomaly. The findings are clearly consistent with adult survival, but whether life expectancy is shortened is not clear. We suspect that the quality of life may be adversely affected. There is evidence that the changes in cognitive functions were present in at least two of the five reported patients, presumably a result of chronic hypoxia. Our patient also complained of shortness of breath and dizziness, also probably on the basis of hypoxia. Resting hypoxia was not profound in any of these patients and might well have been overlooked. Interestingly, differential limb exercise in our patient revealed that even mild exercise of the upper limbs caused profound systemic hypoxia, whereas exercise of the lower limbs did not alter the Po2 level significantly. A potential danger in these patients results from the direct communication of the
superior vena cava with the systemic circulation, which poses the increased risk of systemic emboli. It is recommended that intravenous infusions utilizing veins of the upper body should be avoided if a patient with this syndrome is recognized.

Once having thought of the possibility of anomalous venous connection, the diagnosis of this condition is not difficult. In a 52-year-old man, causes of chronic cyanosis and clubbing with a normal heart are mainly respiratory. Chronic obstructive airway disease, emphysema, interstitial lung disease and the pulmonary cyanosis are easily excluded on the basis of chest x-ray and simple tests of respiratory function. In most instances recurrent pulmonary emboli present with pulmonary hypertension. Hematological causes of cyanosis which weigh heavily in the differential diagnosis usually have met- or sulfhemoglobinemia as a common denominator. The great majority of cases of met-hemoglobinemia are due to the action of chemical agents which increase the rate of auto-oxidation of hemoglobin in the red cells. Rare congenital defects due to the absence of the enzyme diaphorase which reduces met-hemoglobin, and the presence of an abnormal hemoglobin, hemoglobin M, have many clinical features in common with anomalous venous connection. Cyanosis is present from an early age; symptoms of dizziness, shortness of breath, and weakness are common, and mental retardation may even occur. Sulfhemoglobin often occurs in the presence of met-hemoglobin and is not uncommonly associated with a history of ingestion of sulphur-containing laxatives. Spectroscopic examination of the blood excluded both met- and sulfhemoglobinemia. Polycythemia in conjunction with mild hypoxia from whatever cause may present with cyanosis out of proportion to resting blood gases. Thus, included in the differential diagnosis are other causes of polycythemia not already mentioned. These are idiopathic polycythemia rubra vera, tumors and cysts of the kidney, various carcinomata (particularly of the liver), cerebral tumors (particularly cerebellar hemangioblastoma), adrenal adenoma, uterine myomatia, pheochromocytoma and Cushing's syndrome.

Cardiac causes of cyanosis at this age, apart from abnormal venous connection, should be readily excluded with the use of chest radiograph, electrocardiogram and echocardiography. Virtually all such conditions, including Ebstein's anomaly and Eisenmenger's syndrome, should show cardiomegaly and right-sided cardiac abnormalities. Congenital pulmonary arteriovenous fistulæ which may present with cyanosis can usually be recognized by the appearance of multiple lung opacities on the chest radiograph and the presence of continuous murmurs over the lung fields. The definitive diagnosis of anomalous insertion of the superior vena cava, however, can only be made by cardiac catheterization. Anomalous insertion of the superior vena cava into the left atrium is commonly associated with other cardiac and extracardiac anomalies which most often bring the patient to clinical recognition. Intracardiac
defects that may accompany superior vena caval drainage into the left atrium include atrial septal defect, single atrium, ventricular septal defect, single ventricle, Eisenmenger’s complex, Fallot’s tetralogy, pulmonary stenoses or pulmonary atresia and transposition of the great vessels. The extracardiac anomalies include coarctation of the aorta, pulmonary arterial venous fistulae, patent ductus arteriosus, malformations of the inferior vena cava and the presence of both right and left superior vena cava, either intercommunicating or as two separate entities draining into either or both atria.8 The finding of what appears to be an isolated anomalous superior vena caval drainage at catheterization should prompt one to exclude these other frequently associated anomalies.

The major challenge of isolated anomalous superior vena caval drainage, however, is its recognition, which previously has been largely dependent on cardiac catheterization. With more sophisticated noninvasive methods, however, one should be able to suspect the anomaly by nuclear tracer studies. In our patient, the first clue to anomalous venous drainage came with the attempted lung perfusion scan. An injection into the right brachial vein bypassed the lungs and imaged the brain and kidneys. A subsequent study confirmed the course of the tracer from the brachial vein to the superior vena cava, to the left ventricle and aorta.

Although his heart was normal by chest radiograph and echocardiogram, this convincing evidence for a right-to-left shunt prompted cardiac catheterization, which defined the precise anatomy. Thus, unlike the four previously reported patients, the diagnosis of this rare anomaly in our patient was made predominantly from the noninvasive studies.

References
Isolated drainage of the superior vena cava into the left atrium in a 52-year-old man: a rare congenital malformation in the adult presenting with cyanosis, polycythemia, and an unsuccessful lung scan.

M D Ezekowitz, P O Alderson, B H Bulkley, P N Dwyer, L Watkins, D L Lappe, H L Greene and L C Becker