Unsuccessful Lung Scan Due to Major Right-to-Left Shunt Through a Sinus Venosus Septal Defect

A. J. Brendel, B. Larnaudie, B. Lambert, F. Leccia, J. L. Barat, D. Ducassou, and F. Fontan

University Bordeaux II, Bordeaux, France

In a patient with a prior history of cerebral abscess and cerebral ischemia, an unsuccessful perfusion lung scan led to a radionuclide angiocardiogram using an arm vein injection. This showed a total right-to-left (R-L) shunt from the superior vena cava (SVC) to the left atrium. Repeat radionuclide study, through a leg vein, demonstrated a moderate R-L shunt and an interpretable lung scan could be obtained. Catheterization and contrast cineangiogram did not provide the exact diagnosis, the preoperative conclusion being anomalous drainage of the SVC into the left atrium, with atrial septal defect (ASD) and partial anomalous pulmonary venous connection to the SVC. The operative diagnosis was high atrial (sinus venosus) septal defect. This example of major but clinically unsuspected R-L shunt emphasizes the value of performing a perfusion lung scan, preferably in conjunction with radionuclide angiocardiography in patients with a prior history of unexplained cerebral abscess or systemic ischemia. Implications of the site of an ASD on quantitation of L-R shunts by radionuclide methods are also discussed.

J Nucl Med 26:1029–1034, 1985

Isolated atrial septal defect without pulmonary hypertension is commonly known as resulting in left-to-right (L–R) shunt. We recently studied a patient with a clinically unsuspected major right-to-left (R–L) shunt from the superior vena cava, through a sinus venosus defect, demonstrated by radionuclide angiocardiography (RAC), following an unsuccessful perfusion lung scan. This R–L shunt explained the patient's prior history of cerebral abscess and cerebral ischemia.

CASE REPORT

A 61-yr-old man had been operated upon 26 yr before for a cerebral abscess located deep in the left parietal lobe. At that time, no cause for the abscess could be found. Postoperatively the patient had significant neurological sequellae predominantly involving the right arm. In 1981 he was admitted to a neurology department for the sudden onset of altered consciousness and diplopia suggesting a cerebral ischemia. A computed tomographic scan showed an ischemic lesion in the left thalamus. Angiography of cervical and cerebral arteries was normal. Cardiac clinical examination, chest x-ray, standard electrocardiogram (ECG), continuous ECG monitoring, and two-dimensional echocardiogram were normal. The patient was discharged with persistent diplopia.

On November 19, 1983, pulmonary embolism was suspected after the sudden onset of cyanosis and dizziness, and the patient was referred to this institution for a perfusion lung scan. This was performed by injection of 4 mCi of technetium-99m (^{99m}Tc) human albumin microspheres in a left arm vein, because access to the right arm veins was difficult due to the right arm spasticity. The images showed very poor and heterogeneous activity in the lungs and most of the activity was seen in the systemic circulation: kidneys, spleen, thyroid, and brain where a defect corresponding to the site of the previously operated abscess could be seen (Fig. 1). These results suggested a major R-L shunt. A RAC was immediately performed using a bolus of 10 mCi of [^{99m}Tc]pertechnetate injected through the same left arm vein. Sequential anterior images showed the progression of the radiotracer into the left innominate vein, a right sided superior vena cava (SVC), the left heart and the aorta, without visible activity in the right heart or the lung fields. These images suggested the possible diagnosis of anomalous connection of the right SVC to the left atrium and the patient was then admitted to the cardiology division of this institution.

Physical examination on admission revealed a normally developed man with cyanosis and moderate clubbing of the

Received Jan. 4, 1985; revision accepted June 4, 1985.

For reprints contact: A. J. Brendel, MD, PhD, Nuclear Medicine Division, Hôpital Universitaire Pellegrin, F-33076 Bordeaux-Cédex, France.

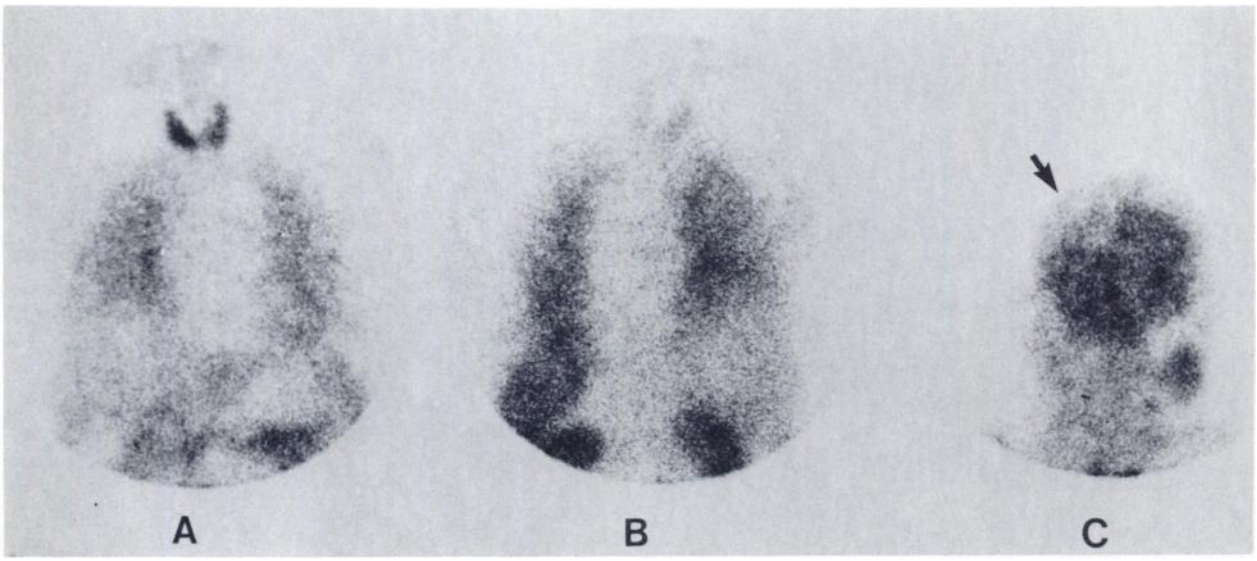

FIGURE 1
Perfusion lung scan obtained after left arm vein injection of [^{99m}Tc]human albumin microspheres. Anterior (A) and posterior (B) views show little activity in lungs and predominant activity in systemic circulation: kidneys, spleen, thyroid and brain (C) which is viewed in posterior view. Defect corresponding to site of previously treated cerebral abscess is shown (arrow)

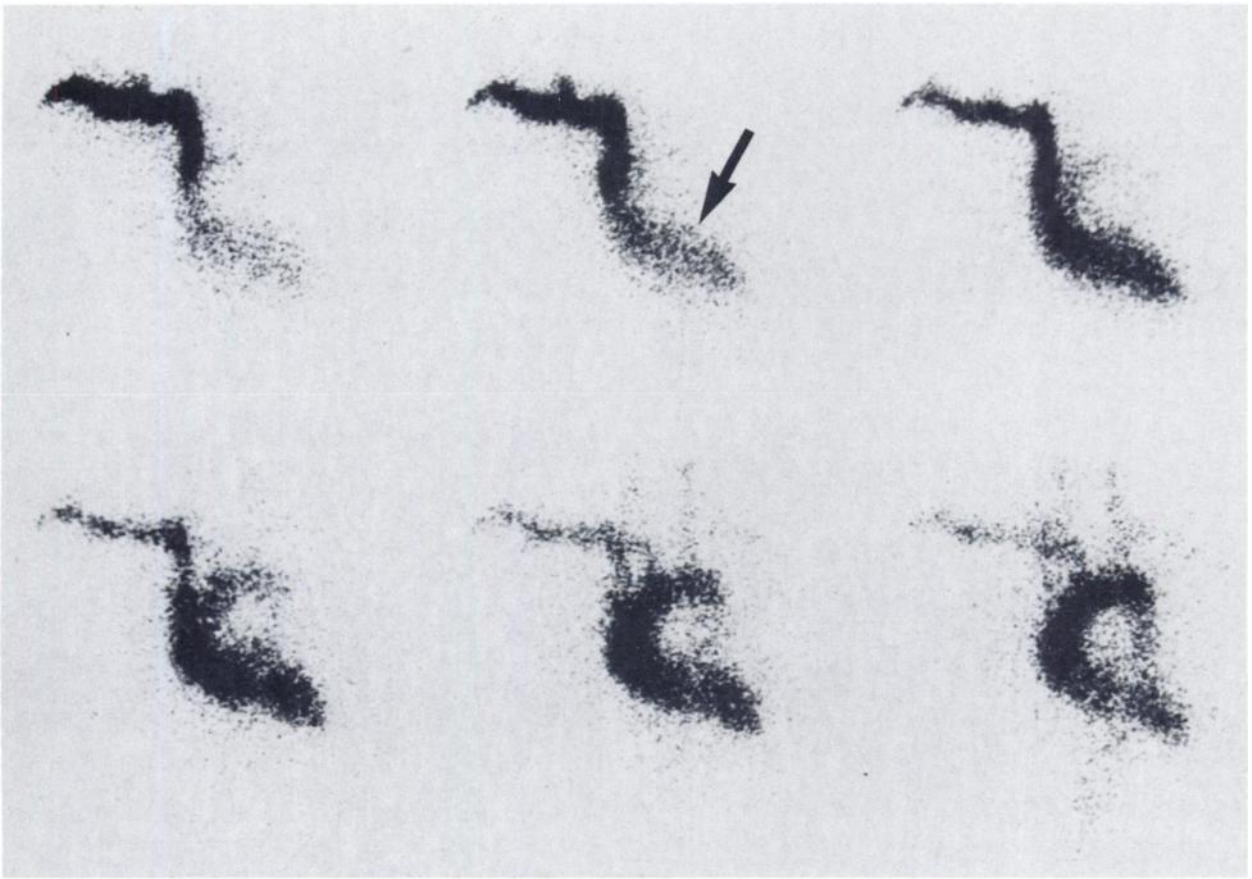

FIGURE 2
Radionuclide angiocardiogram (anterior view) obtained after right arm vein injection of 9 mCi of [^{99m}Tc]DTPA. Frame duration is 0.9 sec. Radiotracer passes directly from superior vena cava to left heart (arrow) and aorta. No activity can be seen in right heart and lung fields

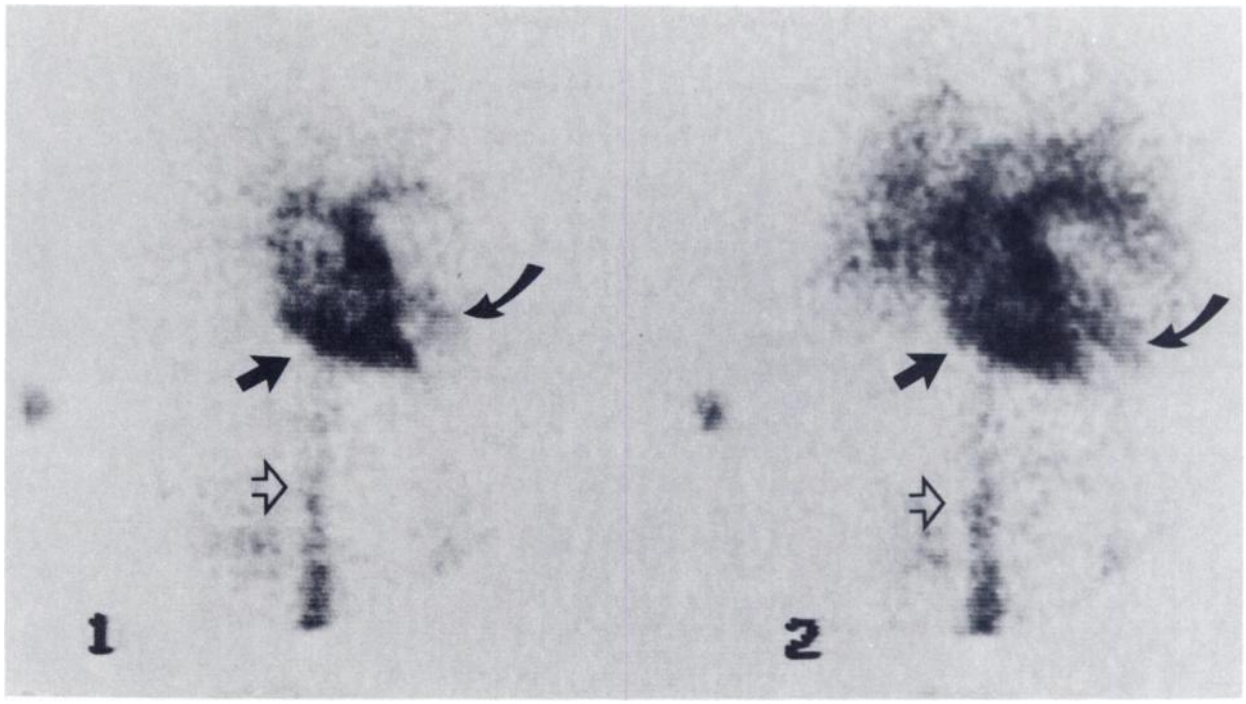

FIGURE 3
Radionuclide angiocardiogram (anterior view) obtained after leg vein injection of 5 mCi of [^{99m}Tc]human albumin microspheres. Frame duration is 3 sec. Radiotracer passes from inferior vena cava (open arrows) to right heart (arrows), pulmonary arteries and lung fields. Moderate R-L shunt is demonstrated (curved arrows)

fingers and toes. Cardiopulmonary auscultation was normal. Chest radiograph revealed a tortuous aorta with normal-sized heart and unremarkable lung fields. An ECG was normal. The hemoglobin was 15.5 g per 100 ml, hematocrit 47% and RBC count 5.01 x 10^6 per mm^3. Arterial blood gases on room air showed a PO_2 of 32 mm Hg, a PCO_2 of 28 mm Hg and a pH of 7.46. A two-dimensional echocardiogram was normal but it was of poor quality due to calcification of costal cartilages and narrowing of intercostal spaces. The patient was treated with heparin.

On the day after admission, new radionuclide studies were requested to obtain an interpretable perfusion lung scan and also to verify that the SVC visualized after the left arm injection was not a persistent left SVC connected to the left atrium, since the diagnosis of right SVC connected to the left atrium is very unusual (*1,2*). A RAC was performed using a right arm vein injection of 9 mCi of technetium-99m diethylenetriaminepentaacetic acid ([^{99m}Tc] DTPA). This study confirmed the presence of a right SVC from which the radiotracer passed entirely into the left heart (Fig. 2), as was previously noted. A perfusion lung scan was subsequently obtained by using a right leg vein injection of a bolus of 5 mCi of [^{99m}Tc]human albumin microspheres. The dynamic images showed the progression of the radiotracer through the inferior vena cava, the right atrium and right ventricle, the pulmonary arteries and both lungs. A moderate R-L shunt was demonstrated (Fig. 3). The static lung scan showed mildly reduced perfusion of the whole left lung compared to the right and also a focal perfusion defect in the posterior part of the left lung base.

A pulmonary angiogram was performed on the following day, using a catheter introduced through a left arm vein. The catheter passed directly from the SVC into the left atrium where the injection of contrast medium showed the left heart with reflux into the right atrium suggesting the presence of an atrial septal defect (ASD). The catheter was then pushed into the right heart and the main pulmonary artery where normal pressures were measured. The pulmonary angiogram showed a thrombus, 25 mm long, in the left inferior lobe artery, which was not causing complete luminal obstruction. Bilateral ascending lower limb contrast venography was normal.

Repeat catheterization with contrast cineangiography was performed 6 days later. The working diagnosis at this study was anomalous connection of the right SVC to the left atrium associated with ASD. The catheter was introduced into a femoral vein and passed into the SVC and then a right superior pulmonary vein indicating partial anomalous pulmonary venous connection to the SVC. Contrast material was then injected. It passed almost entirely into the left heart. A small fraction of it was visualized in the right atrium and ventricle (Fig. 4). Since the catheter was thought to have reached the SVC through the previously diagnosed ASD and the left atrium, the visualization of the right heart chambers was considered to be the consequence of L-R shunting through the atrial septum. This shunt was confirmed by slight visualization of the right atrium during the left heart filling phase, after injection of contrast material into the main pulmonary artery.

The preoperative diagnosis was: (a) anomalous connection of right SVC to the left atrium producing a major R-L shunt

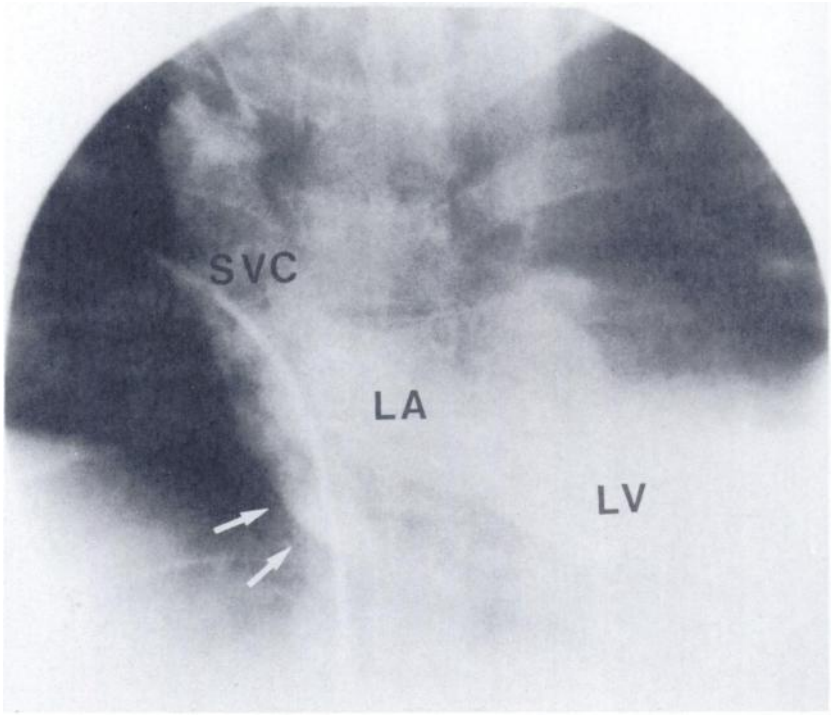

FIGURE 4
Selected image from contrast cineangiogram. Tip of catheter is in end of upper lobe pulmonary vein connected to superior vena cava (SVC). Most of contrast material passes into left atrium (LA) and left ventricle (LV). Small fraction (arrows) passes into right atrium

with cyanosis; (b) partial anomalous pulmonary venous connection to the SVC; (c) moderate bidirectional shunt through an ASD; and (d) pulmonary embolism of the left lung base. Even though it was not hemodynamically significant, this embolism was considered responsible for the decompensation of the patient's clinical status.

At operation the SVC was found to be connected to the cephalad portion of the right atrium. There was an anomalous connection of the right superior pulmonary vein with the caudal portion of this vessel, and an interatrial communication located cephalad to the fossa ovalis, near the site of the anomalous connection (high septal defect or sinus venosus defect). The SVC was overriding the septal defect in such a degree that it opened primarily into the left atrium. An outpouching was noted at the right cephalad portion of the left atrium. This could have favored preferential flow from the SVC to the left atrium. The repair consisted of resecting the outpouching and closing the ASD with a Dacron patch. The patient did well after operation. A radionuclide angiogram performed 4 mo later, using a right arm injection, was normal (Fig. 5). A repeat perfusion lung scan was also normal.

DISCUSSION

The sinus venosus defect is located in the uppermost portion of the atrial septum, the SVC straddling the defect (*3*). Its frequency is estimated to be from 8 to 10% of all ASD (*4–6*). This anomaly is always associated with partial anomalous pulmonary venous connection to the SVC, in particular the right upper lobe pulmonary vein entering the SVC just above the defect (*4*), as it was in our patient. The syndrome is considered to be an anomaly of pulmonary venous development while the atrial septum forms normally (*4*).

A sinus venosus defect usually results in a L-R shunt due to the ASD plus the partial anomalous pulmonary venous connection. However "as a result of the location of the defect, the SVC almost invariably overrides the atrial septum and drains into both atria" (*4*). The superior free border of the interatrial septum acts as a "dividing crest" to direct a portion of the SVC blood into the left atrium (*3,4*). Therefore the transatrial shunt is usually bidirectional.

In our patient, the contrast cineangiogram showed that most of the contrast material injected into the SVC was directed into the left atrium, whereas only a small part of it entered the right heart. It is unlikely that the pulmonary embolism had favored the R-L shunting since the pressures in the right heart and pulmonary artery were normal. The outpouching of the left atrium may have favored the dominant R-L shunt. The shunt was in fact bidirectional since a small L-R shunt was demonstrated by the contrast cineangiogram.

The RAC using an arm vein injection seemed to show exclusive flow from the SVC to the left heart. The dynamic images so obtained were comparable to those published by Park (*1*) and by Ezekowitz (*2*) in two patients with confirmed isolated drainage of the SVC into the left atrium. However, in their patients, no lung activity from labeled microspheres was noted, whereas a little pulmonary activity was present in our patient (Fig. 1) due to passage of some microspheres into the right heart and pulmonary artery, as suggested by the

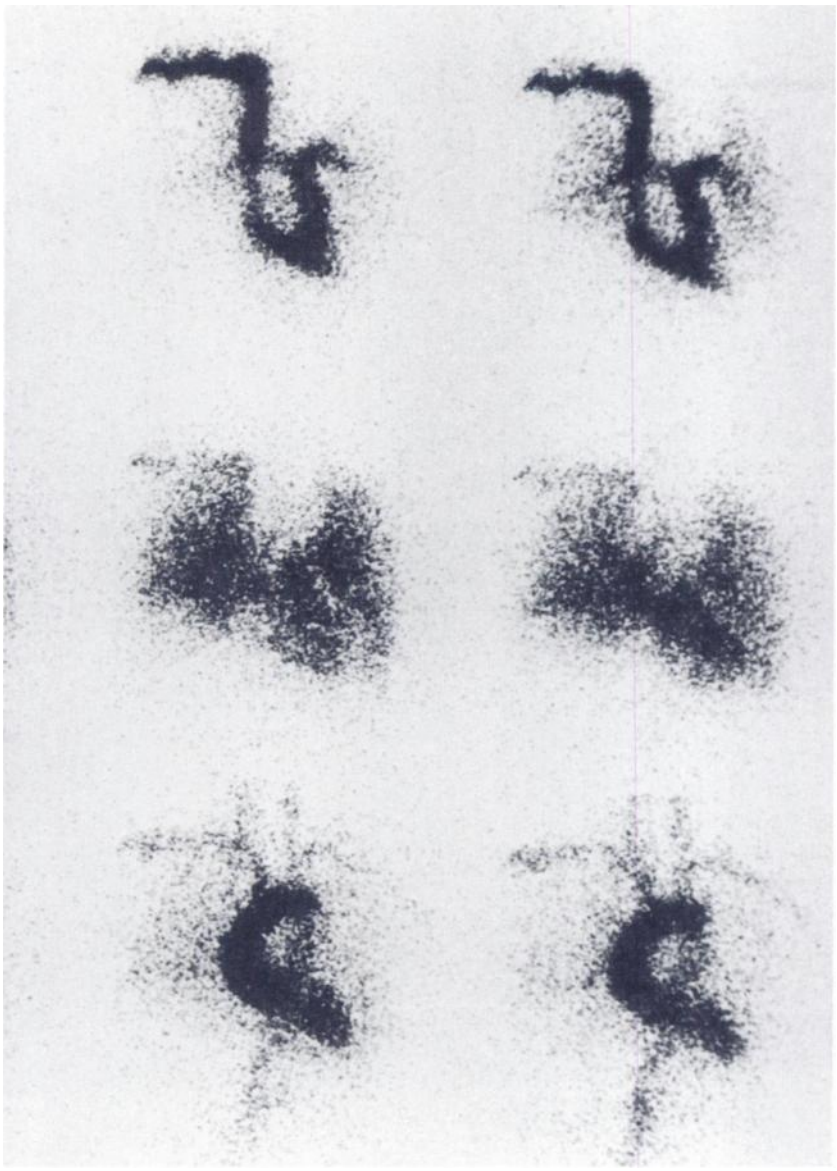

FIGURE 5
Selected images of postoperative radionuclide angiogram (anterior view) obtained after right arm vein injection of 10 mCi of [^{99m}Tc]DTPA. Frame duration is 0.9 sec. Appearances are normal

results of the contrast cineangiography, but not by the RAC. The radionuclide studies using a leg vein injection also showed a small R-L shunt from the inferior vena cava (IVC). All these facts are consistent with the observations of Swan (*4*), indicating preferential R-L shunting of superior as opposed to inferior caval blood in sinus venosus defects.

In this patient both the radionuclide studies and cardiac catheterization failed to provide the exact diagnosis since their results suggested an anomalous drainage of the SVC into the left atrium, associated with an ASD. Compared to the noninvasive radionuclide methods, the only additional diagnostic information provided by cardiac catheterization in this patient was demonstration of the presence of partial anomalous pulmonary venous drainage to the SVC, which prior to surgery seemed momentarily to suggest the true diagnosis. The radionuclide methods by themselves diagnosed the hemodynamic features of significant value, which were a massive R-L shunt from the SVC, and a smaller R-L shunt from the IVC. These shunts are likely to explain the prior history of cerebral abscess and of probable cerebral embolus that the patient had suffered several years before. In that respect the radionuclide studies were useful in directing the patient to catheterization and then to surgery to prevent the onset of new systemic accidents. It should be recommended that patients with otherwise unexplained cerebral abscess or ischemic accidents should undergo a perfusion lung scan, preferably including the recording of the vascular phase, to rule out a R-L shunt. If such a shunt exists, activity from labeled microspheres will be seen in

the systemic circulation, and the lung activity may be poor. When the pulmonary activity is very poor (unsuccessful lung scan) as it was in our patient, the amount of R-L shunt must be very major, either due to diffuse arteriovenous lung fistulas or intracardiac shunt. In these patients the finding of marked R-L shunting from SVC injection and very much less shunting from IVC injection should direct attention to drainage anomalies involving the SVC. Anomalous connection of the right SVC (*1,2*) or of a persistent left SVC (*8*) to the left atrium were previously described. Our patient demonstrates an alternative explanation.

Another implication can be extracted from our observation. The radionuclide techniques are widely used for quantitation of L-R shunts (*9*). The presence of an associated R-L shunting which is common in ASD of different types (*7*), may preclude an accurate quantitation (*9*). Small R-L shunts from the IVC are common in patients with ostium secundum type ASD, whereas R-L shunts from the SVC, when present, are of smaller magnitude (*4*). Therefore the R-L shunt component of an ASD of the ostium secundum type is likely to have little influence on the quantitation of the L-R shunt since the radiotracer is usually injected through an arm vein and reaches the heart by way of the SVC. This is not the case for the sinus venosus type ASD in which most of the R-L shunt component arises from the SVC (*4*). This could explain some of the discrepancies which are sometimes noted between the L-R shunt values obtained by the radionuclide methods and the oxymetry methods in atrial septal defects

ACKNOWLEDGMENT

The authors thank Dr. Michael J. Kelly for review of the manuscript and helpful comments.

REFERENCES

1. Park HM, Smith ET, Silberstein EB: Isolated right superior vena cava draining into left atrium diagnosed by radionuclide angiocardiography. *J Nucl Med* 14:240-242, 1973
2. Ezekowitz MD, Alderson PO, Bulkley BH, et al: 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. *Circulation* 58:751-756, 1978
3. Meszaros WT: *Cardiac Roentgenology*, Springfield, IL, Ch. Thomas, 1969, pp 417-434
4. Swan HJC, Kirklin JW, Becu LM, et al: Anomalous connection of right pulmonary veins to superior vena cava with interatrial communications. Hemodynamic data in eight cases. *Circulation* 16:54-66, 1957
5. Rahimtoola SH, Kirklin JW, Burchell HB: Atrial septal defect. *Circulation* 37 (Suppl. V):2-12, 1968
6. Dupuis C, Kachaner J, Pernot C, et al: Communications interauriculaires. In *Cardiologie Pediatrique*, Paris, Flammarion Medecine-Sciences, 1981, pp 132-139
7. Swan HJC, Burchell HB, Wood EH: Presence of venoarterial shunts in patients with interatrial communications. *Circulation* 10:705-713, 1954
8. Sherafat M, Friedman S, Waldhausen JA: Persistent left superior vena cava draining into the left atrium with absent right superior vena cava. *Ann Thor Surg* 11:160-164, 1971
9. Treves S, Fogle R, Lang P: Radionuclide angiography in congenital heart disease. *Am J Cardiol* 46:1247-1255, 1980