Pseudo Pulmonary Embolism in Complex Congenital Heart Disease

Gary J.R. Cook and Ignac Fogelman
Department of Nuclear Medicine, Guys Hospital, London, United Kingdom

Two children (aged 12 and 14 yr) with a history of complex congenital heart disease presented with symptoms and signs suggestive of pulmonary embolism. Initial ventilation-perfusion lung scans showed normal, approximately equal ventilation to both lungs. Global reduction of perfusion to the right lung was observed in one and was observed in the left lung in the other patient. It was not possible to exclude a large, central embolus in either case. Due to complex cardiac anatomy, however, which included bilateral cavopulmonary anastomoses in the first patient and SVC-right pulmonary artery and right atrial-pulmonary artery anastomoses in the second, repeat scans were performed within a short period in each case with different injection sites, including the contralateral arm and a leg injection, respectively. In both patients, these follow-up scans showed a different perfusion agent distribution to each lung when compared to the initial scans. These results demonstrated that there was no evidence of pulmonary embolism in either case.

Key Words: ventilation perfusion lung scan; congenital heart disease

J Nucl Med 1996; 37:1359–1361

Ventilation/perfusion (V/Q) scans in two children with complex congenital heart disease with symptoms suggestive of pulmonary embolism showed mismatch between perfusion and ventilation. A knowledge of the complex anatomy and a repeat scan using an injection site in a different limb prevented a false diagnosis of pulmonary embolus in each case.

CASE REPORT

Patient 1

A 12-yr-old boy with a history of complex congenital heart disease with multiple previous surgical corrections presented with a one-day history of right sided pleuritic chest pain and shortness of breath. A chest radiograph showed no focal lung abnormality and the pulmonary vasculature was considered to be within normal limits. A clinical diagnosis of pulmonary embolus was made and a V/Q scan was requested to confirm the diagnosis.

At birth the boy had a usual atrial arrangement (situs solitus) with concordant atrioventricular connections but discordant ventriculoarterial connections (transposition). Bilateral superior vena cavae (SVCs) existed where the left vena cavae drained into the coronary sinus and the right into the right atrium. Multiple muscular and a perimembranous ventriculoseptal defects (VSDs) existed with an obstructed left ventricular outlet and stenotic pulmonary valve. When the patient was a 4-yr-old, a left modified Blalock shunt was performed, followed by a modified right Blalock shunt at the age of 7. These procedures improved pulmonary blood flow by connecting the subclavian artery to the ipsilateral pulmonary artery with synthetic graft material. At the age of 11, a total

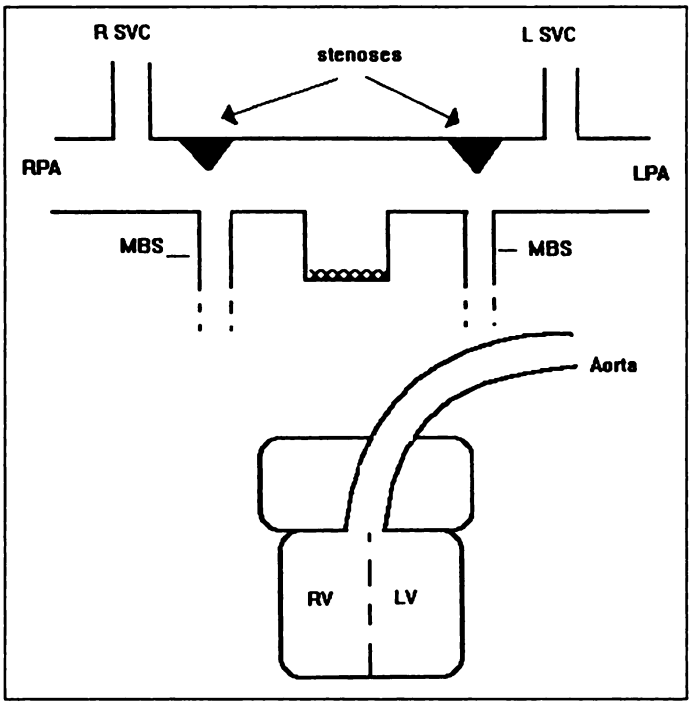

FIGURE 1. Diagrammatic representation of complex cardiopulmonary anatomy in Patient 1 involving transposition with bilateral cavopulmonary anastomoses, pulmonary trunk ligation and bilateral modified Blalock shunts with stenoses at anastomoses with pulmonary arteries. SVC = superior vena cava, PA = pulmonary artery, MBS = modified Blalock shunt, V = ventricle.

(bilateral) cavo pulmonary anastomosis was performed due to stenoses at the previous Blalock anastomoses (Fig. 1).

A V/Q scan was performed using ^{99m}Tc-DTPA aerosol for the ventilation images and 40 MBq ^{99m}Tc-macroaggregated albumin (MAA) injected in the left antecubital fossa for the perfusion images. Anterior, posterior and both posterior oblique images were acquired for perfusion and ventilation. Images showed normal ventilation to both lungs (quantified as L = 45%, R = 55%) (Fig. 2A). Perfusion to the right lung was globally reduced, however

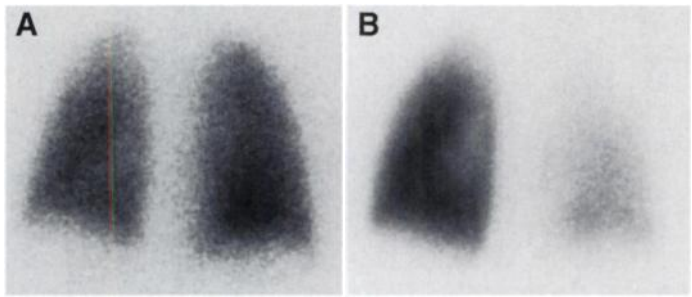

FIGURE 2. (A) Posterior ^{99m}Tc-DTPA aerosol ventilation image showing a normal pattern. Quantification showed distribution of activity of 45% in the left lung and 55% in the right lung. (B) Posterior ^{99m}Tc-MAA perfusion image after left arm injection. Severe global reduction in perfusion is seen in the right lung. Distribution activity was 83% in the left lung and 17% in the right lung.

Received May 31, 1995; revision accepted Aug. 25, 1995.
For correspondence or reprints contact: Dr. G.J.R. Cook, Department of Nuclear Medicine, Guys Hospital, London SE1 9RT, United Kingdom.

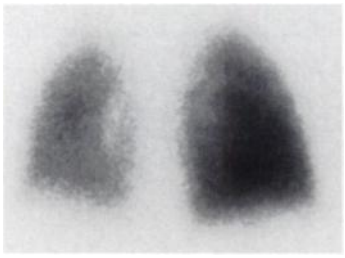

FIGURE 3. Posterior ^{99m}Tc-MAA perfusion image after right arm injection. Global reduction of ^{99m}Tc-MAA accumulation is seen in the left lung. Distribution activity: left 29%, right 71%.

(L = 83%, R = 17%) (Fig. 2B). A repeat study was performed 18 hr later with the ^{99m}Tc-MAA injected into the right hand. Images showed a reversal of the previous pattern with severely reduced perfusion to the left lung (L = 29%, R = 71%) (Fig. 3).

Thus, no perfusion deficit was present in either lung and no evidence of pulmonary embolism was discovered. Subsequent clinical examination revealed tenderness over the costochondral junctions and a clinical diagnosis of costochondritis was made. Symptoms settled with simple analgesia and the patient has remained asymptomatic over a follow-up period of 9 mo.

Patient 2

This 14-yr-old girl experienced shortness of breath 2 wk after undergoing a Fontan procedure (right atrial pulmonary artery anastomosis). A chest radiograph showed a small right pleural effusion with no other significant abnormalities. Although a clinical diagnosis of hyperventilation due to anxiety was made, a V/Q lung scan was requested because of the high risk factor for pulmonary embolism.

The girl had been born with tricuspid atresia with associated atrial septal defect (ASD), VSD and transposition. A SVC-right pulmonary artery shunt (Glenn shunt) had previously been fashioned prior to the Fontan procedure with an anastomosis made between the inferior vena cava (IVC)/right atrium and the medial part of the pulmonary artery. The ASD was closed (Fig. 4).

A V/Q scan was performed in the same manner as the previous case with an injection of ^{99m}Tc-MAA into the right arm. Ventilation was distributed approximately equally (R = 60%, L = 40%) (Fig. 5A). The perfusion scan showed a global

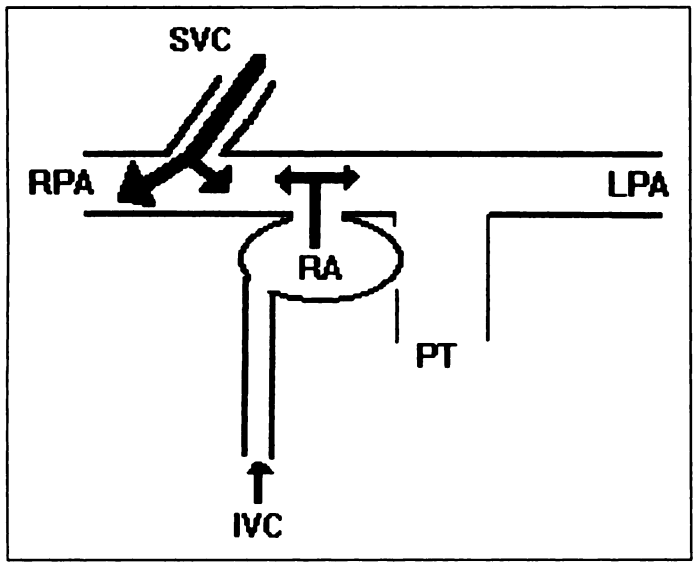

FIGURE 4. Diagrammatic representation of the cardiopulmonary anatomy of Patient 2. SVC right pulmonary artery anastomosis (Glenn shunt) and a medial RA pulmonary artery anastomosis (Fontan procedure) are present. Arrows show direction of blood flow returning from SVC and IVC. SVC/IVC = superior/inferior vena cava, PA = pulmonary artery, PT = pulmonary trunk, RA = right atrium.

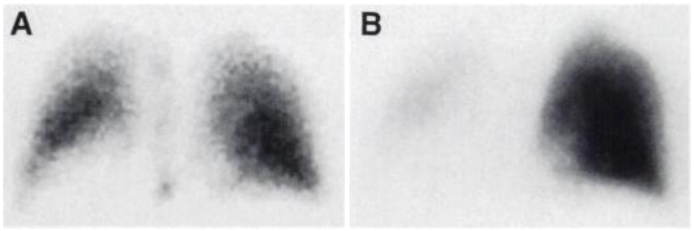

FIGURE 5. (A) Posterior ^{99m}Tc-DTPA aerosol image showing approximately equal ventilation to both lungs. Quantitation showed the distribution of activity to be approximately equal (R = 60%, L = 40%). (B) Posterior ^{99m}Tc-MAA image following injection through the right arm. There is globally reduced deposition of MAA in the left lung (R = 92%, L = 8%).

reduction of perfusion to the left lung (R = 92%, L = 8%) (Fig. 5B). A small matched defect in the right posterior costophrenic angle was seen in the position of the known pleural effusion. A repeat perfusion scan within 24 hr with an injection in the right foot showed a more even distribution (R = 58%, L = 42%) (Fig. 6).

DISCUSSION

A number of causes for perfusion-ventilation mismatch exist in addition to pulmonary embolus that must be kept in mind when interpreting V/Q scans (*1–3*). Considerable research has been performed on lung scanning in congenital heart and lung disease (*4,5*), but the site of administration of the perfusion agent is not ordinarily of importance because of atrial mixing and a common blood supply to both lungs.

In the present cases, the situation was further complicated by the fact that the patients were suspected of having pulmonary thromboembolism. In the first case, the complex anatomy, together with stenoses at the site of anastomoses of Blalock shunts, caused a difference in differential perfusion, depending on the side of injection. On injection of ^{99m}Tc-MAA into the left arm, the majority of activity was deposited in the left lung via the left SVC anastomosis to the left pulmonary artery. Mixing with blood supplied to the right lung was reduced by a proximal left pulmonary artery stenosis at the site of the previous modified Blalock anastomosis. A similar but mirror image pattern was observed when an injection was made into the right arm.

In the second patient, injection into the arm caused ^{99m}Tc-MAA to be deposited preferentially into the right lung via a surgical SVC-right pulmonary artery shunt. When the perfusion agent was injected via the leg, however, the more central anastomosis between the IVC/right atrium and pulmonary artery caused a more even distribution of ^{99m}Tc-MAA to occur (Fig. 4).

CONCLUSION

Awareness of alternative causes of V/Q mismatch and a knowledge of anatomy in patients with congenital heart disease is important when interpreting V/Q scans so as to avoid false-positive diagnoses of pulmonary embolism.

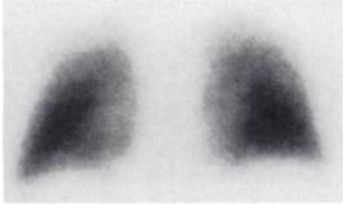

FIGURE 6. Posterior ^{99m}Tc-MAA image following injection into the right foot resulting in a more even distribution (R = 58%, L = 42%).

REFERENCES

1. Li DK, Seltzer SE, McNeil BJ. Ventilation-perfusion mismatches unassociated with pulmonary embolism: case report and review of the literature. *J Nucl Med* 1978;19: 1331–1333.
2. Finestone H, Colp C, Rackson M, Shams J, Gallagher R. Ventilation-perfusion imaging in sarcoidosis: potential for nonembolic segmental mismatch. *J Nucl Med* 1994;35:476–478.
3. Strickland NH, Hughes JM, Hart DA, Myers MJ, Lavender JP. Cause of regional ventilation-perfusion mismatching in patients with idiopathic pulmonary fibrosis: a combined CT and scintigraphic study. *Am J Roentgenol* 1993;161:719–725.
4. Ciofetta G, Gordon I, Peipsz A. Clinical applications of nuclear medicine. *Arch Dis Child* 1988;63:321–328.
5. Gordon I. Review article: Effect of nuclear medicine on pediatric imaging. *Br J Radiol* 1993;66:971–985.

Iodine-123-MIBG Imaging in Pheochromocytoma with Cardiomyopathy and Pulmonary Edema

Kazuyoshi Suga, Katsuhiko Tsukamoto, Kazuya Nishigauchi, Norihiko Kume, Naofumi Matsunaga, Tomoko Hayano and Takao Iwami

Departments of Radiology, and Second Internal Medicine, Yamaguchi University School of Medicine, Ube, Japan

We encountered a patient with pheochromocytoma associated with a catecholamine-induced cardiomyopathy that developed recurrently bilateral and unilateral pulmonary edema. The diagnosis of pheochromocytoma was made by elevated plasma catecholamine levels and intense tumor [^{123}I]MIBG uptake and was confirmed at the time of surgery. The patient showed reduced myocardial [^{123}I]MIBG uptake with left ventricular dysfunction, and endomyocardial biopsy findings were consistent with the diagnosis of catecholamine-induced cardiomyopathy. After tumor resection, plasma levels of catecholamine were normalized, and pulmonary edema never recurred, although cardiac dysfunction did not show an improvement on echocardiography. Myocardial and lung [^{123}I]MIBG uptake increased when compared to uptake levels on preoperative scans, but myocardial uptake was still below normal. These findings indicated that over-secreted catecholamines influenced both the heart and lungs. Pheochromocytoma can induce cardiac and lung injuries, and [^{123}I]MIBG scanning may contribute not only to tumor characterization but also to assessing and monitoring the influence of catecholamines on the heart and lungs.

Key Words: pheochromocytoma; cardiomyopathy; pulmonary edema; iodine-123-metaiodobenzylguanidine

J Nucl Med 1996; 37:1361–1364

Radiolabeled metaiodobenzylguanidine (MIBG), a structural analog of guanethidine, shares the same uptake and storage mechanisms as norepinephrine (*1–4*). For myocardial imaging, this agent is useful to assess cardiac adrenergic activity, and to explore neuronal norepinephrine uptake and storage functions (*4*). For tumor imaging, it can provide tissue characterization of adrenergic tumors, and positive tumor uptake has been documented in 87%–96% of patients with pheochromocytoma and neuroblastoma with extremely high specificity (*5–8*). Moreover, this agent is extracted in lung endothelium by the same active, sodium-dependent saturable transport system as norepinephrine (*9–11*).

In pheochromocytoma, over-secreted catecholamines can induce myocardial (*12–17*) and lung (*14,18,19*) injuries. MIBG scanning may become a unique procedure which allows detection of primary tumor as well as myocardial and lung abnormalities. In this paper, we present the findings of [^{123}I]MIBG scan performed before and after tumor removal in a patient with pheochromocytoma associated with cathecholamine-induced cardiomyopathy and lung edema.

Received May 23, 1995; revision accepted Oct. 8, 1995.

For correspondence or reprints contact: Kazuyoshi Suga, MD, Nuclear Medicine Office, Department of Radiology, Columbia-Presbyterian Medical Center, College of Physicians & Surgeons of Columbia University, 630 West 168th St., New York, NY 10032.

CASE REPORT

A 70-yr-old woman consulted her private physician complaining of a cough and a low-grade fever for 2 wk, and was administered an intravenous drip infusion of 500 ml glucose 5%. During treatment, she complained of severe dyspnea and heart palpitations, and was admitted to the emergency room of our hospital. Seven months earlier, she had been hospitalized for paroxysmal, acute bilateral pulmonary edema, which disappeared rapidly in 3 days. At that time, coronary angiography demonstrated normal coronary arteries, and endomyocardial biopsy showed a mild focal interstitial infiltrate of lymphocytes with scanty myocytolysis; hence, she was diagnosed with myocarditis due to viral infection. She had never demonstrated hypertension.

On this admission, chest auscultation revealed widespread moist rales in the right lung. Blood pressure was 142/86 mmHg and pulse rate was 110/min. Arterial blood gases showed hypoxia with PO_2 levels of 36.2 mmHg. Electrocardiogram showed sinus tachycardia, but no evidence of myocardial infarction or left ventricular hypertrophy. Chest radiograph and chest and CT showed right unilateral lung edema, accompanied by moderate cardiomegaly with a cardiothoracic index of 56%. Color Doppler echocardiography showed left ventricular hypokinesis with a fractional shortening of 44% and a contractility of 25%, without significant left ventricular hypertrophy. The patient's condition improved rapidly following bed rest and diuretic therapy. Chest radiograph and CT on the third day revealed that the right lung edema had disappeared. Lung perfusion scan with ^{99m}Tc macroaggregated albumin (MAA) performed on the fifth day did not show any abnormality in either lung. The patient was subsequently discharged. Review of the patient's history and the above findings led to the diagnosis of acute unilateral lung edema due to left ventricular failure.

Fifteen days after discharge, however, she again complained of sudden dyspnea while working in her garden and was hospitalized again. The main Clinical data obtained at this admission are summarized in Table 1. She again showed hypoxia and right unilateral lung edema on chest radiograph (Fig. 1); however, these improved rapidly on the second day, following bed rest. Thereafter, she occasionally became unstable with a wild fluctuation in blood pressure. Serum levels of catecholamines were significantly elevated. Abdominal ultrasound and x-ray CT revealed a large, solid mass measuring 60 × 60 mm in diameter in the ventral region of