Lung Perfusion Scans in Patients With Congenital Heart Defects

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In 63 patients with various congenital heart defects, lung perfusion was evaluated with technetium-99m macroaggregated albumin. Right lung perfusion abnormalities were documented in 34 patients (54%). A particularly high incidence occurred in patients who had undergone a systemic to pulmonary artery shunt operation as an initial palliative procedure or who had had right ventricular outflow reconstruction and in those with bilateral pulmonary artery stenosis.

Congenital or postoperative lung perfusion abnormalities in patients with a congenital heart defect may adversely affect their outcome and necessitate special interventions (1-3). Children with decreased lung perfusion, especially if it is unilateral, are usually asymptomatic and the above conditions are not easily detected by either chest radiology or two-dimensional echocardiography. The marked improvement in recent years (4,5) in our ability to directly relieve branch pulmonary artery stenosis, either surgically or with balloon angioplasty, has emphasized the need for an accurate and noninvasive method for quantitative evaluation of the relative pulmonary blood flow in patients with a congenital heart defect. Lung perfusion scintigrams using albumin macroaggregates labeled with technetium-99m have been shown to be an effective method for this assessment (6-10). In this report we describe our experience with this test in a heterogeneous group of patients with various congenital heart defects.

Methods

Study patients. From January 1989 to July 1990, 63 patients with a congenital heart defect were studied with lung perfusion scintigrams at the Beilinson Medical Center. There were 26 male and 37 female patients whose age ranged from 6 months to 40 years (mean ± SD 7.1 ± 6.6 years). The medical records, chest X-ray films, two-dimensional and Doppler echocardiograms and hemodynamic and cineangiographic data were reviewed.

Serial studies were helpful in evaluating the functional results of different transcatheter interventions for optimizing pulmonary blood flow. The quantitative relative perfusion radionuclide method was a more sensitive means of detecting cases of abnormal lung perfusion than was chest radiology.

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The indication for lung perfusion scintigraphy was the presence of any congenital heart defect or surgical intervention that could have affected the distribution of pulmonary blood flow to the lungs. Patients were classified into four groups: 1) patients who had undergone a surgical intervention to augment pulmonary blood flow (for example, systemic to pulmonary artery shunt operation, right ventricular outflow tract reconstruction, complete repair of tetralogy of Fallot); 2) patients who had undergone pulmonary artery banding or debanding, or both; 3) patients who had undergone direct anastomosis of systemic venous pathways to the pulmonary arteries (for example, bidirectional Glenn operation, modified Fontan operation); and 4) patients with congenital peripheral pulmonary artery stenosis. Informed consent for the procedure had been obtained from the parents or patient in each case.

Lung perfusion scintigraphy. Technetium-99m-labeled macroaggregated albumin in a dose of 0.5 to 2 mCi (18.5 to 74.0 MBq), adjusted to patient weight, was injected through an intravenous cannula inserted into a peripheral vein. Perfusion imaging was begun almost immediately after injection of the radiopharmaceutical agent with the patient supine. A standard multiple view scan was performed with use of a large field of view gamma camera (APEX SP-6, Elscint) equipped with a low energy, all purpose collimator. Four views (anterior, posterior and two posterior oblique views) were obtained. Each view was imaged for 500,000 counts and recorded on transparent multiformat mode. Lung perfusion was calculated as the relative ratio of the sum of radioactivity in each lung in the anterior and posterior views. Inequality of lung perfusion was assessed by an independent observer without reference to chest X-ray findings or hemodynamic and cineangiographic data. Abnormal lung perfu-
Lung perfusion scintigrams were defined as <45% or >58% right lung perfusion (Fig. 1) (6).

Chest X-ray studies. The patients' chest X-ray films, obtained within 6 months of the lung perfusion scans without an intervening procedure, were evaluated for pulmonary blood flow in both lung fields by an independent pediatric radiologist without reference to the calculated results of the lung perfusion scintigrams. The chest X-ray films were visually examined according to the method described by Amplatz and Castaneda-Zuniga (11). Arterial blood distribution in each lung was defined as normal, increased or decreased and compared with distribution in the contralateral lung field. Later, the results were correlated with calculated pulmonary flow distribution by perfusion lung scintigrams.

Cineangiograms. Thirty patients underwent cardiac catheterization and cineangiography within 6 months of the lung perfusion scintigram. Pulmonary artery anatomy was evaluated qualitatively and correlated with the results of the lung perfusion scintigram.

All results are presented as mean values ± SD.

Results

Overall results: augmentation of pulmonary blood flow (Table 1: Abnormal lung perfusion scintigrams were found in 34 patients (54%). Twenty had increased flow to the right lung (range 58% to 90%; mean 73 ± 9%) and 14 had decreased flow to the right lung (range 13% to 43%; mean 32 ± 10%).

Of the 44 patients who underwent an intervention to augment pulmonary blood flow, 25 (57%) had an abnormal lung perfusion scintigram. Increased flow to the right lung, which was present in 17 patients (range 65% to 96%; mean 74 ± 8%), was more common than decreased flow, which was present in 8 patients (range 13% to 43%; mean 31 ± 10%). Of the 27 patients who underwent a systemic to pulmonary artery shunt operation as the initial palliative procedure, 16 (59%) had an abnormal lung perfusion scintigram. Seventeen patients underwent a left-sided shunt operation and 9 had increased flow to the right lung, indicating a left pulmonary artery stenosis. Of the 10 patients who underwent a right-sided shunt operation, 4 had decreased flow to the right lung and 3 had increased flow to the right lung, suggesting stenosis of the right pulmonary artery either proximal or distal to the shunt insertion site (Fig. 2). Fifteen patients with tetralogy of Fallot underwent complete repair without a previous shunt. Eight (53%) had abnormal lung perfusion demonstrating peripheral pulmonary artery stenosis.

Results with other procedures. Eleven patients underwent pulmonary artery banding, five for a ventricular septal defect and six for various types of single ventricle physiology. Five (45%) of these patients had an abnormal lung perfusion scintigram (Fig. 3); two of these five were from the single ventricle group, which placed them at high risk for the modified Fontan operation. Five patients underwent either a bidirectional Glenn shunt (n = 2) or a modified Fontan operation (n = 3); two of these patients (one from each group) had an abnormal lung perfusion scintigram; one of these two had a previous pulmonary artery band operation. Of the three patients with congenital peripheral pulmonary artery stenosis, two had an abnormal lung perfusion scintigram.

Assessment of interventions: case reports. A repeat lung perfusion scintigram was helpful in evaluating the functional

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<th>Table 1. Results of Lung Perfusion Scintigraphy in 63 Patients</th>
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<td>Normal</td>
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<td>Augmentation of pulmonary blood flow (n = 44)</td>
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<td>Complete repair of ToF without a previous shunt (n = 15)</td>
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<td>Pulmonary artery banding (n = 11)</td>
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Unless otherwise indicated, data show number and percent of patients in each category. ToF = tetralogy of Fallot.
Figure 2. A, Right ventricular outflow angiogram demonstrating complete occlusion of the proximal right pulmonary artery (arrow) in a patient with pulmonary atresia and intact ventricular septum after a Waterston shunt operation as a newborn and right ventricular reconstruction and right pulmonary artery enlargement at age 2 years. B, Right pulmonary vein wedge injection demonstrating a hypoplastic distal right pulmonary artery (arrow). C, Lung perfusion scintigrams in four views in the same patient demonstrating severe hypoperfusion of the right lung (13%) compared with the left lung (87%). Abbreviations as in Figure 1.

Figure 3. Lung perfusion scintigram (posterior view) in a patient with multiple large apical ventricular septal defects after pulmonary artery banding, demonstrating severe underperfusion of the right lung (right lung 18%, left lung 82%).

results of different interventions that were performed to optimize pulmonary blood flow (Fig. 4 and 5).

Case 1. A 7-year-old asymptomatic girl with tetralogy of Fallot and a small right ventricle underwent right ventricular outflow tract reconstruction at age 1 year. At age 2 years, she underwent a modified left Blalock-Taussig shunt operation after she was found to have a hypoplastic left pulmonary artery. At age 5 years, she underwent complete repair with further enlargement of the right ventricular outflow tract, closure of the ventricular septal defect, and reconstruction of the left pulmonary artery. The shunt could not be reached from a midline sternotomy. A lung perfusion scintigram 1 year after this operation demonstrated severe underperfusion of the left (10%) compared with the right (90%) lung.

Because cardiac catheterization and angiography revealed severe narrowing of the proximal left pulmonary artery, the patient underwent balloon angioplasty of this artery, which increased in size from 3.7 to 8.4 mm. She also underwent coil embolization of the Blalock-Taussig shunt. A repeat lung perfusion scintigram demonstrated significant
improvement in left lung perfusion (36%) and right lung perfusion of 64% (Fig. 4).

Case 2. A 9-month-old girl with severe tetralogy of Fallot, hypoplastic pulmonary arteries and multiple systemic pulmonary artery collateral vessels underwent right ventricular outflow tract reconstruction. A lung perfusion scintigram demonstrated hyperperfusion of the right lung (72%) due to a large systemic to pulmonary collateral artery. At cardiac catheterization, this collateral vessel was coil embolized and a subsequent lung perfusion scintigram demonstrated almost equal perfusion of both lungs (right 48% and left 52%). The patient's clinical condition improved and the signs of congestive heart failure resolved.

Cases 3 and 4. Two patients with tetralogy of Fallot and pulmonary atresia aged 2 and 6 years, respectively, had severe left pulmonary artery stenosis, one after a modified left Blalock-Taussig shunt operation and one after right ventricular outflow tract reconstruction. Each underwent attempted left pulmonary artery balloon angioplasty without significant change in left pulmonary artery size or left lung perfusion.

Correlation with angiography. In 30 patients, the lung perfusion scintigram could be correlated with angiographic findings. In 27 patients (90%) there was a good correlation with anatomic findings and degree of abnormality. Thirteen patients had underperfusion of one lung, which correlated well with branch pulmonary artery stenosis. Two patients had increased flow to one lung that was demonstrated by cineangiography to be secondary to a systemic to pulmonary collateral artery in one and to a systemic to pulmonary artery shunt in the other. Twelve patients had a normal lung perfusion scintigram and normal pulmonary arteries on cineangiography. Three patients had a normal lung perfusion scintigram with abnormal cineangiographic findings. Two patients had right pulmonary artery stenosis distal to a modified right Blalock-Taussig shunt and one patient had moderate right pulmonary artery stenosis after pulmonary artery banding.

Correlation with chest X-ray findings. Both a chest X-ray film and a lung perfusion scintigram were available in 56 patients. Only 21 of the chest X-ray studies (37%) correlated well with the lung perfusion studies. When only the chest
Figure 5. A. Anterior view lung perfusion scintigrams in a patient with severe right pulmonary artery stenosis after complete repair of tetralogy of Fallot without a previous shunt operation, demonstrating severe underperfusion (18%) of the right lung. B. After successful balloon dilation of the right pulmonary artery, normal perfusion (46%) of the right lung.

X-ray films from those patients with abnormal lung perfusion were evaluated. correlation was much better, with agreement in 17 (63%) of 27 patients.

Discussion

Perfusion lung scanning is a known and safe means of assessing regional pulmonary perfusion in children with a congenital heart defect (6-10). The distribution of the radioactive aggregates between the two lungs is directly proportional to the division of pulmonary artery flow (12).

Safety. Up to 95% of the intravenously injected 10- to 50-μm radiotopes entering the pulmonary arteries exert their impact on the capillary or precapillary bed of the lung. These small particles temporarily occlude one of several hundred thousand capillaries but only 1 in 1,500 arterioles. This difference accounts for the >1,000-fold safety factor for perfusion lung scanning. The albumin particles eventually fragment into smaller components, pass through the capillary bed of the lung into the systemic circulation and are phagocytized by hepatic and splenic reticuloendothelial cells. We have used this method without complications even in cyanotic patients. The halflife removal rate of the radioactive aggregates from the lung is 4 to 8 h (13). The absorbed radiation dose resulting from 200 μCi of technetium-99m macroaggregated albumin injected into a newborn is 0.05 rad to the whole body and 0.65 rad to the lungs. This radiation dose diminishes rapidly as the patient grows and by age 1 year has decreased by nearly two-thirds (14).

Clinical value. The results of such a relatively noninvasive test enable one to plan further invasive studies such as cardiac catheterization and pulmonary artery cineangiography (15). The increased ability to restore equal lung perfusion either surgically or with transcatheter interventions such as balloon angioplasty of branch pulmonary arteries (4,5) or coil embolization of systemic to pulmonary artery collateral vessels has emphasized the usefulness of this relatively noninvasive, simple and easily reproducible method for assessing pulmonary blood flow.

Our results demonstrate that lung perfusion abnormalities are common (54%) in children with a congenital heart defect, especially after surgical intervention. Most of these patients were asymptomatic, even though they had a grossly abnormal lung perfusion scintigram. Similar results were described by Gates et al. (9), who found that 76 of the 31 children with a congenital heart defect had hyperperfusion of one lung. In the present study, lung scanning provided particularly useful information concerning the fate of the pulmonary arteries after a palliative systemic to pulmonary artery shunt operation in patients with various types of cyanotic heart disease and it allowed identification of those patients with pulmonary artery abnormalities who need special intervention either before or at the time of complete repair.

We used this method to assess transcatheter interventions in four patients. Ring et al. (4) described the use of this method in patients who underwent balloon angioplasty of branch pulmonary arteries, demonstrating that in 10 of their 13 patients, the mean pulmonary blood flow to the affected lung increased from 40 ± 4% to 51 ± 4%.

Lung perfusion abnormalities may be found in patients after a bidirectional Glenn or a modified Fontan operation, as in two of our five patients. Similar results have been described by del Toro et al. (10) and are more common when the patient has undergone a previous palliative operation.

Correlation with other methods. Correlation of roentgenographic and scintigraphic studies showed that the lung perfusion scintigram is more accurate than the other methods studied and that detection of pulmonary blood flow imbalance by inspection of chest X-ray films was possible only when there was grossly unequal pulmonary blood flow. In three of our patients, the lung perfusion scintigram failed to demonstrate significant pulmonary artery stenosis that was well visualized by cineangiography. The reason for this is unclear. This gives a sensitivity of 83% and specificity of 100%.

Conclusions. Our results and those of others demonstrate that the lung perfusion scintigram is a relatively easy, reliable and noninvasive method for the accurate determination of relative pulmonary blood flow, a value not easily
calculated by other means such as chest radiography and two-dimensional echocardiography. This method allows the detection of changes in pulmonary blood flow and enables one to plan more invasive studies or special transcatheter or surgical interventions.

References