Pulmonary Artery Agenesis

Diagnosis with Ventilation and Perfusion Scintiphotography

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SUMMARY

Pulmonary artery agenesis is one of several diagnostic possibilities which may present with similar chest roentgenographic features, including unilateral pulmonary hypoperfusion and nonvisualization of a pulmonary artery. We have applied ventilation and perfusion scintiphotography as an aid in differentiating agenesis from the other conditions with which it may be confused.

Four patients presenting with the chest roentgenographic features suggestive of pulmonary artery agenesis were evaluated, when possible, with complete pulmonary function studies, bronchography, cardiac catheterization, and pulmonary and aortic angiography. In all patients, the distribution of ventilation and perfusion were evaluated with the Anger scintillation camera, 133Xenon being used for the ventilation study and 131I-MAA for the perfusion study. In each patient, perfusion scintiphotography showed no blood flow to the affected lung, while ventilation scintiphotography disclosed preservation of homogeneous ventilation to that lung.

Absence of pulmonary artery flow to one lung with maintenance of relatively normal ventilation limits the broad range of diagnostic possibilities to three entities: pulmonary artery agenesis, thrombotic occlusion, and branch stenosis. The history and physical findings should give clues to the latter two diagnoses.

Additional Indexing Words:
Scans Pulmonary artery flow absent Asymmetrical hemithorax
Spirometric studies Unilateral pulmonary vascular abnormalities
Unilateral pulmonary parenchymal abnormalities

PULMONARY ARTERY agenesis has been known as an entity since its description in 1886 by Fraentzel. With the advent of the pulmonary angiogram as a common diagnostic procedure, many reports have appeared enumerating its clinical features, pathology, and embryology.1-3 However, the diagnostic approach to pulmonary artery agenesis has continued to present difficulties.4 History, physical examination, and chest roentgenogram have suggested the diagnosis, but more sophisticated procedures requiring hospitalization were necessary in establishing the diagnosis. These included bronchospirometry, bronchography, cardiac catheterization, and pulmonary and aortic angiography. Recently, however, simple, noninvasive radioisotopic technics have become available for defining the distribution of pulmonary blood flow and ventilation.5, 6 Such technics can be applied safely on an outpatient basis. Therefore, if the isotopic methods can serve as a guide to the selection of subsequent procedures, they offer a valuable adjunct to the initial diagnostic evaluation of a patient with suspected pulmonary artery agenesis.

Establishment of the diagnosis of pulmonary artery agenesis is important because surgical correction may be possible in selected
cases. Furthermore, this entity should be differentiated from other congenital or acquired abnormalities for which there is individualized treatment.

This paper reviews four cases in which a diagnosis of pulmonary artery agenesis was considered. In each case, pulmonary perfusion scintiphotography (using $^{131}$iodine-labelled albumin macroaggregates) and pulmonary ventilation scintiphotography (using $^{133}$xenon gas) were performed. In each instance, combined ventilation-perfusion studies provided valuable differential diagnostic information. This approach has not been applied previously, save one case in which perfusion studies were reported.

Methods

Subjects

All patients were referred because of abnormal roentgenograms obtained elsewhere. Referral diagnoses were multiple but did not include the possibility of pulmonary artery agenesis. When possible, the patients were admitted to the hospital for evaluation. Two patients refused hospitalization and could be evaluated only on an outpatient basis.

Lung Scintiphotography

The thyroid gland was blocked by the oral administration of saturated solution of potassium iodide. Perfusion scans were obtained following the intravenous injection of 400 $\mu$C of $^{131}$iodine-labelled macroaggregated albumin (MAA). Half the dose was given with the patient prone, the other half with the patient supine. The patient was then seated and scintiphotographs were obtained in the anterior, posterior, and both lateral positions with a Nuclear-Chicago Pho Gamma III Scintillation Camera.

Ventilation scintiphotography was performed using a 13.5-L Collins spirometer filled with a $^{133}$xenon gas-oxygen mixture. The seated subject, nose clip in place, was connected to the spirometer by a mouthpiece and initially breathed room air. The subject was turned toward the spirometer at the end of a normal expiration and asked to take a maximum inspiration which was held for 15 to 30 sec. Following this, the subject rebreathed from the spirometer until $^{133}$xenon equilibrium between the spirometer and the patient was achieved. The patient was then switched back to breathing room air to “wash out” the $^{133}$xenon. At 15-sec intervals the radioactivity from each lung was recorded and simultaneously a Polaroid picture was obtained.

Other Procedures

Static and dynamic lung volumes were determined by using a double-bell spirometer (Godart Pulmotest). Functional residual capacity (FRC) was determined by the helium-dilution method. The diffusing capacity for carbon monoxide ($D_{LCO}$) was determined by the single-breath method. Oxygen uptake and carbon dioxide output were calculated by standard methods. The minute ventilation ($V_{E}$) was measured directly from expired gas collected in Douglas bags. The alveolar oxygen tension was calculated from the alveolar air equation. Arterial blood gas values were determined with an Instrumentation Laboratories gas analyzer, model 113. Bronchography, right heart catheterization, pulmonary angiography, and aortic root angiography were done in the usual manner. The aorticopulmonary perfusion scintiphotographs were obtained following injection of 200 $\mu$C of $^{131}$I-MAA via the aortic catheter.

Report of Cases

Case 1

M.D., a 17-year-old male, was admitted to Georgetown University Hospital in April 1968 for evaluation of an abnormal chest roentgenogram noted during a preemployment physical examination. He denied cardiopulmonary symptoms. Physical examination showed a well-developed Negro male with normal vital signs. There was a slight shift of the trachea toward the left. Diaphragmatic excursions were equal. There were no adventitious pulmonary sounds. The breath sounds throughout the left lung field were somewhat diminished. The point of maximal cardiac impulse was at the fifth left intercostal

Figure 1

Posteroanterior chest roentgenogram in case 1.
space at the anterior axillary line. The remainder of the physical examination was unremarkable.

The electrocardiogram showed sinus rhythm with a vertical axis and an RSr in V1 compatible with leftward rotation of the heart. The chest roentgenogram showed shift of the trachea and mediastinum toward the left and decreased vascularity of the left lung.
artery was well visualized, but the left was not seen. There was minimal widening of the superior mediastinal shadow (fig. 1). A bronchogram showed normal bronchial anatomy with narrower branches on the left compatible with the lesser volume of that lung. Lung perfusion scintiphotographs showed no detectable blood flow to the left lung (fig. 2). The xenon ventilation study disclosed diminished but homogeneous ventilation of the left lung (fig. 2). Spirometric studies and arterial blood gases at rest, exercise, and during breathing of 100% oxygen were normal. Right heart catheterization disclosed normal pulmonary arterial pressures at rest and during exercise. A pulmonary angiogram showed no left pulmonary artery. The right pulmonary artery and its branches appeared normal and drained normally into the left atrium (fig. 3). An aortic angiogram demonstrated a right-sided aortic arch and large collateral vessels coming from the descending aorta supplying blood to the left lung (fig. 4A and B). This collateral circulation also was demonstrated by aortic perfusion scintiphotography (fig. 4C). These findings were compatible with the diagnosis of agenesis of the left pulmonary artery and a right aortic arch with aberrant aortic vessels supplying blood to the left lung.

Case 2

T.A., a 54-year-old Caucasian male, was referred to Georgetown University Hospital for evaluation in May 1963. He had been rejected for military service in 1943, "because there was something wrong with his lungs." At that time he was asymptomatic and had remained so except for the onset of mild dyspnea on exertion in 1961.

Physical examination disclosed a well-developed, middle-aged male with normal vital signs. His right hemithorax appeared smaller than the left, and breath sounds throughout the right lung field were somewhat diminished. The point of maximal cardiac impulse was inside the left midclavicular line. Findings on the remainder of the physical examination were within normal limits. The chest roentgenogram showed a decrease in volume of the right lung with mediastinal shift to that side. The left pulmonary artery was large, and vascular shadows to the left lung were increased. No right pulmonary artery could be seen, and vascularity to that lung was diminished (fig. 5). Spirometric studies and complete blood gas studies were within normal limits. Bronchography disclosed no abnormality other than bronchi of decreased caliber in the right lung. During right heart catheterization there was a slight rise in pulmonary artery pressure with exercise. A pulmonary angiogram disclosed one large pulmonary artery smoothly curving to the left with no evidence of a right pulmonary artery (fig. 6).

The patient was seen again in August 1968 at which time lung perfusion scintiphotography showed no evidence of blood flow to the right lung (fig. 2). Ventilation scintiphotography showed diminished but homogeneous ventilation to the right lung (fig. 2).

Case 3

J.R., a 30-year-old white female housewife, was referred for evaluation because of an abnormal chest roentgenogram. She had symptoms of mild bronchitis and had smoked a pack of cigarettes a day for 15 years.

Physical examination disclosed a thin, well-developed female with normal vital signs. Examination of the thorax disclosed symmetrical hemithoraces and a minimal scoliosis to the right. The breath sounds throughout the right lung were diminished. The chest roentgenogram showed mediastinal and tracheal shift to the right with decreased volume of the right lung, decreased vascular markings throughout the right lung, no evidence of a right pulmonary artery, and a large left pulmonary artery shadow. Lung perfusion scintiphotography showed no blood flow to the right lung. The ventilation study showed homogeneous distribution of xenon to the right lung. The patient refused further studies.

Case 4

C.W. was a 22-year-old Negro male who presented himself to find out the significance of an abnormal chest roentgenogram which had led to his rejection from military service in 1965. He was asymptomatic. Physical examination disclosed a well-developed young male with normal vital signs. The chest roentgenogram disclosed bilateral symmetrical hemithoraces with normal volume. Breath sounds were symmetrically decreased. The right hemithorax was smaller than the left. The chest roentgenogram disclosed mediastinal shift to the left with diminished vascular markings in the right lung. The patient refused further studies.
vital signs. The hemithoraces were symmetrical and diaphragmatic excursion was equal bilaterally. Breath sounds were diminished throughout the right lung. The remainder of the physical examination was unremarkable. The chest roentgenogram showed the mediastinum to be shifted to the right with no right pulmonary artery visible and decreased vascular markings throughout the right lung. The left pulmonary artery was prominent. Spirometric studies were normal. Lung perfusion scintiphotography showed no perfusion to the right lung with normal perfusion of the left lung. Ventilation scintiphotography showed preservation of homogeneous ventilation of the right lung. The patient refused further evaluation.

Results

Historical and Physical Features

All patients were referred for pulmonary evaluation because of an abnormal chest roentgenogram taken in the course of a medical evaluation. Effort dyspnea was present in only one and none had a history of hemoptysis, recurrent pulmonary infections, chest pain, or episodes of acute dyspnea.

Patients T.A. and J.R. showed asymmetrical hemithoraces, with the smaller hemithorax on the side of the pulmonary artery agenesis. Patient J.R. had a slight scoliosis to the side of the agenesis. Patient M.D. had readily palpable tracheal shift to the side of the agenesis. All patients had diminished breath sounds over the lung with the agenesis. No cardiac abnormality was evident on physical examination other than the displacement of the mediastinum, and there were no significant extrathoracic physical findings.

Laboratory Studies

The chest roentgenographic features of these four subjects were similar to those described in previously reported cases of
pulmonary artery agenesis. They included the following findings on the affected side: decreased lung size with a mediastinal shift, absence of a pulmonary artery, atypical vascular markings, and a higher than normal diaphragm.

The spirometric studies on three patients were normal. The arterial blood gases were normal in two of the patients tested. Cardiac catheterization disclosed calculated pulmonary vascular resistance to be normal in M.D. but slightly elevated in T.A.

Bronchograms of patients M.D. and T.A. showed no evidence of intrinsic tracheobronchial disease. Decreased caliber of the airways on the side of the agenesis was compatible with decreased lung volume of the affected lung. The pulmonary angiograms made of patients M.D. and T.A. were considered to be diagnostic of pulmonary artery agenesis (figs. 3 and 6).

The aortic angiogram in patient M.D. demonstrated a right aortic arch and aberrant aortic blood supply to the affected lung (fig. 4). Aortic injection scintiphotography showed the blood flow via this circulation to concentrate in the lateral basal region of the left

The 133Xenon ventilation studies in the four subjects showed that radioactivity in the lung, which was the area supplied by a relatively large aortic aberrant vessel (fig. 4).

Table 1 shows the percentages of radioactivity recorded from the lung with pulmonary artery agenesis in each patient during perfusion and ventilation scintiphotography. These data were obtained using the split crystal feature of the scintillation camera by aligning the crystal center with the patient's vertebral column. While perfusion scintiphotographs disclosed no radioactivity on the agenetic side, the quantitative counts disclosed a small, variable amount of activity. This activity represents a technical artifact due to mediastinal shift toward the agenetic side, allowing radionuclides in the normally perfused lung to be counted on the agenetic side. In addition, "scatter" from the perfused lung impinges upon the crystal area monitoring the nonperfused lung. These effects of shift and scatter were particularly marked in patient J.R. who had scoliosis and in whom 17% of total radioactivity appeared to come from the nonperfused lung.

The 133Xenon ventilation studies in the four subjects showed that radioactivity in the lung...
with the pulmonary artery agenesis averaged 42% of the total radioactivity for both lungs (table 1). The percentages of activity in the agenetic lung were slightly below those anticipated in normal lungs. Data were analyzed to determine whether this was due to the smaller volume of the agenetic lung or other factors. The relative distribution of ventilation to each lung was evaluated by comparing the distribution of total counts in each lung during the initial inspiration with distribution at equilibrium (thus providing an approximation of ventilation per unit of lung volume). Relative ventilation of each lung also was assessed by determining the half time (T%) of 133Xenon clearance from each lung. In all four cases, the lung not served by a pulmonary artery had a slightly lower calculated ventilation per unit of lung volume and a slightly longer T%. However, the differences were not statistically significant in this small sample. Thus, the decreased radioactivity seems primarily a reflection of the decreased total volume of the agenetic lung.

**Discussion**

The patient who proves to have pulmonary artery agenesis usually presents with an abnormal chest roentgenogram, few symptoms, and a variety of diagnostic possibilities. An ordered selection of potentially diagnostic procedures is essential. Simple, noninvasive procedures such as history, physical examination, and chest roentgenography have been used to guide selection of procedures which carry higher morbidity: for example, cardiac catheterization, angiography, and bronchography. Our experience indicates that pulmonary ventilation and perfusion scintigraphy are of such high value and low risk that they should be added to the initial diagnostic evaluation in patients with possible pulmonary artery agenesis. This procedure, by disclosing that one lung has absent perfusion with preservation of homogeneous ventilation, sharply limits the diagnostic possibilities.

Total unilateral absence of lung perfusion may occur in a substantial number of congenital and acquired conditions (table 2). However, when ventilation scintigraphy discloses that this lung is homogeneously ventilated, the diagnostic list narrows to three primary vascular abnormalities: pulmonary artery agenesis, severe pulmonary artery branch stenosis, and acquired occlusion of a main pulmonary artery (table 3).

Many of the entities in table 2 can be eliminated on the basis of history, and physical and roentgenographic features. In the few that cannot be eliminated unequivocally on these grounds, the results of combined perfusion-ventilation scintigraphy should lead to their discard. Total absence of

**Table 1**

<table>
<thead>
<tr>
<th>Perfusion (%)</th>
<th>Ventilation (%)</th>
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<tr>
<td>7</td>
<td>46</td>
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<tr>
<td>9</td>
<td>36</td>
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<tr>
<td>1</td>
<td>42</td>
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<tr>
<td>19</td>
<td>43</td>
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<tr>
<td>Mean</td>
<td>9</td>
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<table>
<thead>
<tr>
<th>I. Primary Unilateral Vascular Abnormalities</th>
<th>II. Primary Unilateral Parenchymal Abnormalities</th>
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<tbody>
<tr>
<td>A. Congenital</td>
<td>A. Congenital</td>
</tr>
<tr>
<td>1. Pulmonary artery agenesis</td>
<td>1. Agenesis of lobes or segments</td>
</tr>
<tr>
<td>2. Proximal pulmonary artery branch stenosis</td>
<td></td>
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<tr>
<td>B. Acquired</td>
<td>1. Infection</td>
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<tr>
<td>1. Thrombotic occlusion</td>
<td>2. Bronchiectasis</td>
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<tr>
<td>2. Secondary to parenchymal disease</td>
<td>3. Tumor</td>
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<td></td>
<td>4. Emphysema (including “unilateral hyperlucent lung”)</td>
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<td></td>
<td>5. Surgical resection</td>
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<td></td>
<td>6. Ruptured bronchus</td>
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<td></td>
<td>7. Pleural effusion</td>
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<tr>
<td></td>
<td>8. Fibrothorax</td>
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<tr>
<td></td>
<td>9. Pneumothorax</td>
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perfusion is uncommon in some of them (such as bronchiectasis and fibrothorax). Furthermore, even in those conditions in which pulmonary artery perfusion may appear totally absent, homogeneous ventilation is not preserved. For example, in patients with extensive unilateral parenchymal disease, there may be angiographic nonvisualisation of the pulmonary circulation to the involved lung.10 Our experience, however, has disclosed patchy defects on the ventilation scintigraphs rather than the homogeneous ventilation characteristic of agenesis, and that perfusion rarely appears to be totally absent on scintigraphy. The same differential value of scintigraphy applies to such entities as surgical resection, ruptured bronchus, pneumothorax, and fibrothorax in which both ventilation and perfusion are impaired.

Differential among the three entities in table 3 poses more difficulty, however. The physiologic situation in each can be the same; namely, total absence of blood flow with maintenance of homogeneous ventilation. Thus, these three entities must be distinguished on grounds other than scintigraphy.

Unilateral pulmonary artery branch stenosis of severe degree may closely mimic pulmonary artery agenesis. However, in this condition a characteristic murmur is usually audible and pulmonary angiography is often definitive. In most instances, the lung supplied by the stenotic vessel is not reduced in volume (as it usually is in agenesis) and mediastinal-tracheal shift is absent.

Chronic thrombotic occlusion of a main pulmonary artery is a rare event and most cases have had an acute episode suggesting, at least retrospectively, sudden occlusion.11 A history of acute onset of cardiorespiratory symptoms is absent in agenesis. The affected lung is of normal size in thrombotic occlusion unless it has been the site of infarction. In the latter case, bronchography should disclose the regional nature of the lung volume decrement. Angiography usually will display a branch which ends abruptly rather than the characteristic pattern in agenesis; namely, total absence of one branch with a smooth wall in the remaining pulmonary artery in the region from which that branch usually originates.

Bronchogenic carcinoma has resulted in total unilateral pulmonary artery occlusion in some instances.12,13 However, symptoms and roentgenographic signs (hilar mass, encroachment on bronchial lumen) usually prove adequate clues to the fact that these are not instances of primary thromboembolic occlusion. Further, substantial encroachment on bronchial lumen will result in abnormal ventilation scintigraphy.

Thus, once ventilation-perfusion scintigraphy has narrowed the possibilities to those listed in table 3, ample clues usually exist to differentiate among them. Under extraordinary circumstances, however, this may prove impossible without direct visualization at surgery. Each of the three lesions is potentially amenable to surgical correction. Chronic thrombotic occlusion of a main pulmonary artery has been successfully relieved.11 Agenesis has been corrected by anastomosis and grafting.1,7,8 Proximal coarctation of the pulmonary artery also is amenable to surgical attack. The indications for surgery are similar in the three conditions; namely, objective evidence of cardiorespiratory compromise. Therefore, in those rare instances in which differentiation among the three entities is impossible and cardiorespiratory compromise exists, surgical intervention may be required with alternate attack plans formulated, depending upon which of the three abnormalities is encountered.

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