Cause of Regional Ventilation-Perfusion Mismatching in Patients with Idiopathic Pulmonary Fibrosis: A Combined CT and Scintigraphic Study

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OBJECTIVE. Regional ventilation and perfusion were studied in patients with idiopathic pulmonary fibrosis (cryptogenic fibrosing alveolitis) to seek an explanation for the mismatched ventilation/perfusion (V/Q) seen on scintigrams, which may suggest pulmonary embolic disease.

SUBJECTS AND METHODS. Eight patients with idiopathic pulmonary fibrosis were examined with inspiratory and expiratory CT scans. Planar and tomographic (single-photon emission computed tomography, SPECT) scintigraphy, using inhalation of krypton-81m gas (ventilation) and IV injection of ^{99m}Tc-albumin macroaggregates (perfusion), also was performed. The lungs were divided into quadrants (cranial, caudal, right, left) for analysis.

RESULTS. Cystic air spaces with a "honeycomb" appearance occupied more than 33% of the cross-sectional area in 75% of all lung quadrants (n = 16), more than 66% of the area in 44% of quadrants, and less than 33% of the area in the remaining 25% of quadrants. On expiratory CT scans, the cross-sectional area of the cystic spaces diminished significantly (unlike emphysematous spaces). Sixty-seven percent of lung quadrants, corresponding to those with marked or moderate involvement with cystic spaces, showed a mismatched V/Q pattern on scintigrams (absent perfusion, normal ventilation); 27% of quadrants had matched V/Q defects, and 6% did not show defects. Two patients had, in addition, large cystic spaces typical of emphysema, but the coexistent fibrosis prevented the gross air trapping seen in bullous emphysema.

CONCLUSION. The cystic air spaces that are often seen on CT scans of patients with idiopathic pulmonary fibrosis are unperfused (probably due to vascular obliteration) but are usually normally ventilated. This V/Q mismatch on scintigrams explains the large physiologic dead space seen at rest and on exercise and could suggest pulmonary embolism unless a CT scan is obtained. Conversely, the larger cystic spaces might be mistaken for emphysema unless V/Q scintigraphy is done.

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Small cystic air spaces (1–2 mm in diameter) in the periphery of the lung are a well-described feature of idiopathic pulmonary fibrosis [1, 2]. In the past, we have observed patients with large cystic air spaces that resemble emphysematous bullae on CT scans or on Gough thick lung sections at autopsy [3], but with atypical behavior, namely, normal findings on ventilation scans and no air-flow obstruction shown by pulmonary function testing. The presence of cystic air spaces (small or large), which are normally ventilated but unperfused, might explain several features commonly found in patients with idiopathic pulmonary fibrosis, such as the large physiological dead space on exercise [4, 5] and scintigraphic findings that suggest pulmonary embolic disease [3, 6, 7]. In addition, the functioning of larger cystic air spaces, usually considered to be "emphysematous" [8], remains to be defined.

We used CT to detect the presence and size of cystic air spaces in patients with idiopathic pulmonary fibrosis and V/Q scintigraphy (planar and single-photon emission computed tomography) to define the ventilatory function of these

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spaces. As a further check, we visualized the ventilatory excursion of the cystic spaces directly by repeating the CT scan during expiration.

Subjects and Methods

Eight patients with a diagnosis of idiopathic pulmonary fibrosis were studied with thoracic CT and V/Q scintigraphy. The research protocol was approved by the local ethics committee, and informed consent was obtained from each subject. The diagnosis was based on clinical criteria (basal crackles in all eight, finger clubbing in seven), radiologic grounds (reticulonodular shadowing in the lower zone in all eight), and the absence of an occupational cause for lung fibrosis [8, 9]. Histologic confirmation was achieved by open lung biopsy in five patients, by transbronchial biopsy in one patient, and from the differential cell count in the bronchoalveolar lavage sample in another patient. All patients but one were being treated with oral prednisolone; three were also taking cyclophosphamide, and two were taking azathioprine and cyclophosphamide. Their mean duration of exertional dyspnea was 69 months (range, 3–240 months). All were ex-smokers except for one current smoker. The average age was 61 years (range, 36–74 years). Results of routine pulmonary function tests (Table 1) were consistent with established idiopathic pulmonary fibrosis: vital capacity and carbon monoxide transfer factor were reduced. Because all eight patients had been cigarette smokers for many years, the ratio of forced expiratory volume in 1 sec to vital capacity (FEV_1/VC) was less than 75% in seven of eight (a ratio > 80% would be expected in uncomplicated diffuse lung fibrosis). Nevertheless, the residual volume (RV), a sensitive marker of diffuse airway obstruction, was normal or low in all subjects. Two patients (nos. 3 and 8) had evidence of localized bullous emphysema on CT scans. Although patient no. 8 had the lowest FEV_1/VC and the highest RV of the group, the general pattern of pulmonary function, even those two parameters, suggested idiopathic pulmonary fibrosis rather than emphysema. A progressive bicycle ergometer test showed arterial oxygen desaturation in six of eight patients when they exercised to their maximum capacity (Table 1).

Each patient had a chest CT examination in full inspiration and full expiration with 10-mm contiguous slices [10, 11]. The exposure time to image each 10-mm inspiratory or expiratory section was 1 sec. An established scanning technique was used [11], whereby the patient uses a hand signal to communicate when he has achieved full inspiration or expiration and the scan is then obtained. This ensures reproducibility of results and avoids positional overlap of contiguous slices. For patients examined in the second half of the study period, we had access to spiral CT technology. Two spiral data collections were sufficient to cover the whole chest, and the spiral mode was particularly useful for ensuring lack of overlap on contiguous sections. When spiral CT was used, the total scanning time was 24 sec.

The criteria used for differentiating the CT appearances of emphysema and idiopathic pulmonary fibrosis are based on published descriptions of the two diseases [1, 2, 8]. Emphysema is characterized by the appearance of relative transradiancy at the usual window settings, separation and thinning of lung vessels, and destruction of the parenchymal pattern. Emphysematous changes tend to be maximal in the upper lobes. Emphysematous bullae may arise within these areas and are thin walled. Idiopathic pulmonary fibrosis has a predilection for the lower lobes and usually occurs in a crescentic peripheral distribution. Small cystic air spaces resemble a honeycomb; the cysts have well-defined relatively thick walls and arise within this high-attenuation reticular background.

On the same day, all patients had ventilation (krypton-81m) and perfusion (^{99m}Tc-albumin macroaggregates) scans [12, 13]. Separate conventional planar images in anterior, posterior, and right and left oblique views were obtained for each of these radionuclides by using a gamma camera interfaced with an MDS A2 computer system, taking 64 10-sec images during a 360° rotation around the subject's thorax. ^{81m}Kr is a radioactive gas with an ultra-short half-life ($t_{1/2}$, 13 sec); when ^{81m}Kr is added continuously to the inspired air, during tidal breathing, an image is accumulated in 1–2 min that is equivalent to the summation of many small breaths of xenon-133 [14]. No equilibrium or washout images are obtained. In addition, axial tomographic (single-photon emission computed tomography [SPECT]) data were acquired with both radionuclides [15, 16] simultaneously by using the system in dual-isotope tomographic mode. The simultaneous acquisition ensured that the two physiologically different sectional lung images were spatially correlated. This allowed each ventilation image to be divided by the corresponding perfusion image to produce a series of sectional V/Q images. If the images had been collected sequentially, even small relative movements would have greatly impaired the appearance of the V/Q sectional images.

A low-energy general-purpose collimator, rated up to the energy of ^{81m}Kr, was used. Count rates in the ^{99m}Tc and ^{81m}Kr windows

TABLE 1: Pulmonary Function Data and Exercise Capacity in Eight Patients with Interstitial Pulmonary Fibrosis

Patient No.	FEV_1/VC	VC(l)	VC(%)	TLC(%)	RV(%)	D_LCO(%)	K_{CO}(%)	Exercise Capacity (W)	SaO_2 (Rest/exercise)
1	0.63	3.8	87	63	40	33	51	240 m[a]	97/87
2	0.55	4.3	115	90	81	49	57	105	98/93
3	0.56	2.6	74	68	65	49	71	90	96/90
4	0.72	3.8	80	79	60	50	66	150	96/95
5	0.67	2.1	56	55	62	41	70	120	96/84
6	0.55	2.9	71	75	110	49	67	60	94/93
7	0.76	3.9	91	98	111	51	58	105	96/92
8	0.43	2.9	71	83	115	39	75	200 m[a]	96/87
Mean	0.61	3.3	81	76	81	45	64	105	96/90
SD	0.11	0.8	18	14	28	7	8	30	1/4
SEM	0.04	0.3	6	5	10	2	3	12	0/1

Note.—Data in percent are percentages of predicted values. FEV_1 = forced expired volume in 1 sec, VC = vital capacity, TLC = total lung capacity, RV = residual volume, D_LCO = carbon monoxide diffusing capacity, K_{CO} = D_LCO per unit lung volume, exercise capacity = maximum power output in watts on a progressive bicycle ergometer test, SaO_2 = arterial oxygen saturation, SEM = standard error of the mean.

[a] Number of meters walked on the level (excluded from mean values).

were comparable, as were the effects of attenuation of the two similar energies by the lung. Scatter from the ^{81m}Kr into the ^{99m}Tc window was compensated for by viewing the delivery tube through scattering material before the imaging to give a down-scatter factor (of about 15%), multiplying the ^{81m}Kr images by this factor, and subtracting the results from the corresponding ^{99m}Tc images. No significant up-scatter was measured. Imaging the lung without breath-holding does not justify high-resolution reconstruction, and consequently a medium-resolution filter was chosen, which also had the effect of smoothing out much of the noise in both ventilation and perfusion images, making the V/Q images less noisy and easier to interpret.

The ventilation images were normalized to the set of perfusion images by multiplying by the ratio of the total counts in each set of slices. Division to produce the V/Q ratio was performed by integer image arithmetic. This involved multiplication of the counts for the ventilation images by a nominal factor of 100, division by the counts for the perfusion images, and then restoration of the true V/Q ratios by dividing those values by 100. Although the inevitable reconstruction background in the images was evident as noise, the ratio of backgrounds in the V/Q images gave results comparable with the true V/Q ratios that interfered with the appearance of the final display. The original noise background was therefore electronically masked out.

Each radiologist independently evaluated the paired inspiratory and expiratory CT scans, and then, at another review session, the V/Q scans. A scoring system was used in which each lung was divided into upper and lower zones (quadrants) above and below the carina. Inspiratory scans were assessed as showing none, 1–33%, 34–66%, and 67–100% of the lung parenchyma occupied by cysts. The expiratory CT scans were assessed as showing no change, less than 50% reduction, or more than 50% reduction in cyst size compared with the corresponding inspiratory CT scans.

For the planar scintigrams, V/Q scans were assessed, by using the same upper and lower zonal divisions for each lung, as normal, predominantly matched defects, and predominantly mismatched defects. The resolution of the SPECT images was not considered good enough in most cases for formal analysis by this scoring system to be meaningfully applied.

Disagreement between the opinions of the two radiologists was minimal (approximately 4–6%) and was resolved by joint reassessment of the cases concerned, after which a consensus opinion was reached. None of the disagreements concerned the percentage of cyst reduction determined by comparing inspiratory and expiratory CT scans. The differences in opinion arose in cases in which the percentage of lung involvement was considered to be borderline between 34–66% and 67–100%. This occurred in one lung quadrant on the planar V/Q scans (i.e., 1/32 or 3%) and in two lung quadrants on the inspiratory CT scans (i.e., 2/32 or 6%).

The CT scans were imaged at a window width of 1500 H centered at -500 H, settings that enable the mediastinal structures to be identified. Corresponding levels on the inspiratory and expiratory scans were selected for comparison by carefully correlating the anatomic structures on the two sets of scans from each patient. This is important because cystic spaces may change shape during respiration without altering volume; for a valid comparison, an intrapulmonary, bronchial, or mediastinal landmark must be used to match the slices as closely as possible [10]. Additional features, for example coronary artery calcification, were also noted and used to help match the corresponding levels of the inspiratory and expiratory scans.

Results

Figures 1A and 1B show that the small cystic air spaces associated with idiopathic pulmonary fibrosis appear deflated on the expiratory CT scan and appear well ventilated on the ^{81m}Kr scan (Fig. 1C). The multiple irregular perfusion defects (Fig. 1D) in the presence of normal ventilation give a V/Q mismatch. In addition, this patient had air spaces at the lung apices that were more characteristic of emphysema (Fig. 1E). On the right posterior oblique V/Q scans, a matched V/Q defect typical of emphysema can be seen (Figs. 1G and 1H).

Figure 2A shows cystic air spaces due to idiopathic pulmonary fibrosis, but rather unusually, these are most marked in the midzones bilaterally, instead of the more classic crescentic distribution of cystic air spaces predominantly in the periphery of the lower lobe in most patients with idiopathic pulmonary fibrosis [1, 2]. As seen in Figure 1, these cystic spaces deflate on expiration (Fig. 2B) compared with inspiration (Fig. 2A), and a functional V/Q image shows high-V/Q elements peripherally in the mid zones of both lungs (Fig. 2C). The functional image of V/Q was constructed for a single section at the same level (just cranial to the carina) by dividing the SPECT ventilation image by the perfusion image, as described in the Methods section. These areas of high V/Q in the lung correspond to the areas with the most cystic air spaces shown on the thin-section CT scan (Fig. 2A) from the same patient.

Two observers independently scored four axial sections (cranial, carinal, mid-caudal, caudal) at identical levels on the CT and SPECT V/Q reconstructions in four of eight patients (adequate SPECT V/Q images were unobtainable in the remaining four). Each axial section was divided into four quadrants (right upper, right lower, left upper, left lower; 64 total) and scored 0, 1, or 2 for the presence of cystic air spaces and V/Q mismatch (i.e., normal to high V/Q as in Fig. 2C). The CT and SPECT scores were the same in 87 (68%) of 128 comparisons (two observers). Substantial disagreement (difference of two points) occurred in only five (4%) of 128 comparisons.

Two patients (nos. 3 and 8) showed a mixed pattern consistent with emphysematous bullae and the cystic air spaces of idiopathic pulmonary fibrosis. Figure 3A shows emphysematous areas in the anterior segments of both lower lobes, with an area of the small cystic air spaces characteristic of idiopathic pulmonary fibrosis posteriorly in the right lower lobe. Figure 3B shows the different behavior of these two types of abnormalities on expiration, with less deflation of the emphysematous regions compared with the areas affected by idiopathic fibrosis.

We subdivided the 32 lung quadrants in the eight patients on the basis of the extent of their anatomic involvement with cystic air spaces as seen on inspiratory CT scans (Table 2). The lower quadrants are most severely affected, with 60% of them having more than two thirds of their cross-sectional area involved by the disease. Cyst deflation was visually assessed by comparing the expiratory CT scans with the corresponding inspiratory CT scans, as described in the Methods section. In 84% of the lung quadrants, the cystic spaces deflated to less than 50% of their inspiratory size. The remainder were seen to deflate, but to a lesser extent.

Table 2 also shows regional ventilation and perfusion data from the planar scintigrams in terms of their V/Q matching

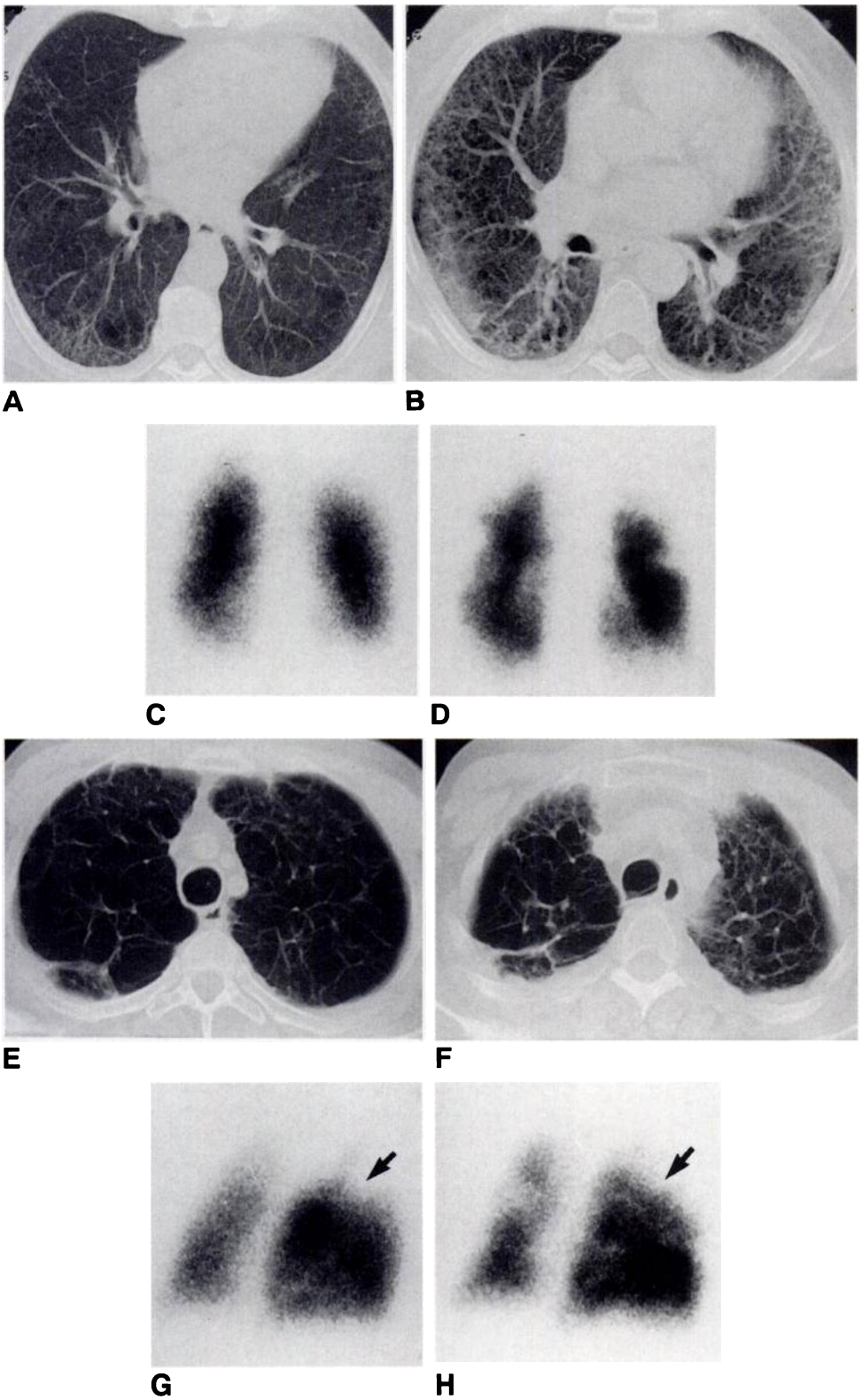

Fig. 1.—64-year-old man with interstitial pulmonary fibrosis.

***A*, Inspiratory CT scan of 10-mm section shows "fibrotic" cystic air spaces posteriorly in both lower lobes.**

***B*, Expiratory CT scan of same 10-mm section as *A* shows that spaces have disappeared or diminished, which indicates that they are ventilated.**

***C*, ^{81m}Kr ventilation scan, posterior view, shows normal ventilation in mid and lower zones where cystic air spaces were seen on CT scan.**

***D*, ^{99m}Tc-MAA perfusion scan, posterior view, shows perfusion defects where findings on ventilation scan were normal. Note V/Q mismatching.**

***E*, Inspiratory CT scan of 10-mm section through upper lobes shows larger "emphysematous" cystic air spaces.**

***F*, Expiratory CT scan of same section as *E* shows some deflation of emphysematous spaces bilaterally, but less than would be seen in normal lung.**

***G*, Right posterior oblique projection of ^{81m}Kr ventilation scan clearly shows defects in upper lobes (*arrow*) corresponding to emphysematous spaces shown on CT scan (these apical defects are also present, but less obvious, on *C*). Regional ventilation in lower and middle zones is normal.**

***H*, Right posterior oblique projection of the ^{99m}Tc-MAA perfusion scan shows defects (*arrow*) in upper lobes (also seen in *D*), namely, a matched V/Q defect in areas of emphysematous spaces.**

pattern. Sixty to eighty percent of the upper- and lower-zone quadrants had a mismatched V/Q pattern, with absent or reduced perfusion. In 25–30% of quadrants, the V/Q pattern was predominantly matched. Only 15% of the quadrants had no defects, and these quadrants were all in the upper zones. The predominantly high V/Q pattern is well illustrated in Table 3, where ventilation defects per quadrant are plotted against perfusion defects. Most of the quadrants lie in the upper right portion of the Latin square, indicating greater defects of local perfusion than ventilation.

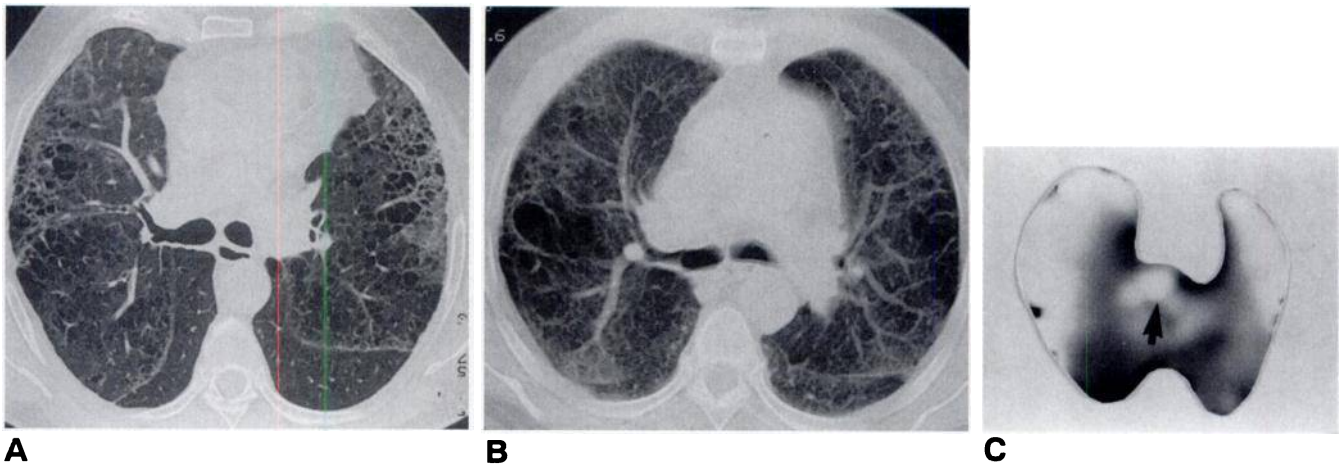

Fig. 2.—72-year-old man with interstitial pulmonary fibrosis.
***A*, High-resolution inspiratory CT scan of 2-mm section shows cystic air spaces predominantly in periphery of mid zone.**
***B*, Expiratory CT scan of same section as *A* shows disappearance of most cysts seen on inspiratory scan.**
***C*, Functional cross-sectional SPECT image of V/Q just above level of carina. High-V/Q regions (ventilated but unperfused) are white; normal V/Q regions are gray. Note high V/Q in periphery of both lungs and in trachea (*arrow*).**

Fig. 3.—74-year-old man with mixed emphysematous and idiopathic pulmonary fibrotic disease.
***A*, Inspiratory CT scan of 10-mm section shows extensive emphysematous spaces anteriorly in both lower lobes, in addition to cystic air spaces of idiopathic pulmonary fibrosis posteriorly in right lower lobe (*arrow*).**
***B*, Expiratory CT scan of same section as *A* shows that deflation of emphysematous areas of lung anteriorly is much less marked than deflation of normal lung or lung involved with cystic air spaces (*arrow*).**

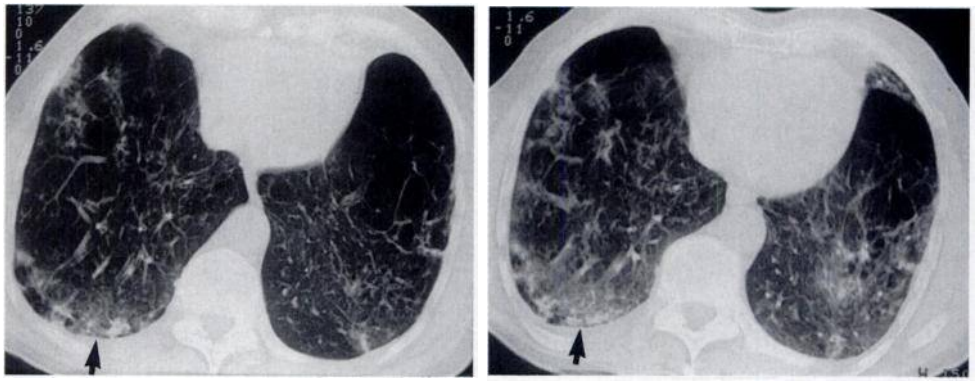

TABLE 2: Extent of Parenchymal Involvement by Cystic Air Spaces on Inspiratory CT Scans and Pattern of V/Q Matching on Planar Scintigrams in Eight Patients with Interstitial Pulmonary Fibrosis

Imaging Finding	Percentage of Quadrants	
	Upper Zone	Lower Zone
Zone area occupied by cysts on inspiratory CT scans		
1–33%	31.3	18.8
34–66%	37.5	18.8
67–100%	31.3	62.5
Ventilation/Perfusion (V/Q)		
Normal	12.5	0.0
Matched	31.3	25.0
Mismatched	56.3	75.0

Note.—Each patient's lungs were divided into quadrants (n = 32) on the basis of upper and lower zones (for right and left lungs), and frequency distribution is expressed as a percentage of the total number of quadrants in each zone (n = 16).

TABLE 3: Extent of Ventilatory Defects vs Extent of Perfusion Defects for All Lung Quadrants (n = 32) in Eight Patients with Interstitial Pulmonary Fibrosis

Extent of Ventilatory Defect (% of quadrant area)	Extent of Perfusion Defect (% of quadrant area)			
	0	1–33	34–66	67–100
0	2	6	7	3
1–33	0	3	6	0
34–66	0	0	1	2
67–100	0	0	0	2

Note.—Extent of ventilation and perfusion defects estimated from planar scintigrams. Numbers in body of table are numbers of quadrants seen for each ventilation and perfusion combination.

Discussion

In idiopathic pulmonary fibrosis, the commonest abnormality seen on regional ventilation and perfusion imaging is

a V/Q mismatched pattern (Fig. 1) with low or absent perfusion. The cause is vascular destruction by the fibrotic process rather than vasculitis [17]. The high V/Q pattern was confirmed on SPECT scans (Fig. 2C), on which regional ventilation and perfusion were measured simultaneously. SPECT also showed that the high-V/Q pattern was associated with regions of cystic air spaces shown on CT scans (Figs. 2A–2C), the "honeycomb" lung pattern. On CT scanning, 75% of lung quadrants had more than one third of their volume occupied by cystic air spaces (Table 2). Sixty-seven percent of lung quadrants showed a high-V/Q mismatch pattern on scintigrams. Furthermore, comparison of CT scans taken in full inspiration and expiration (Figs. 1A, 1B, 2A, 2B) showed a marked volume change in these cystic air spaces, suggesting that they had a normal or high compliance and patent airways.

It is difficult to match with accuracy the regions of high V/Q shown on a planar scintigram with the three-dimensional anatomy shown on a CT scan. That was the reason for doing SPECT scans at the same session as the planar images (the additional radiation exposure, being limited to an extra 10-min exposure to ^{81m}Kr gas, was trivial). Qualitatively, the three-dimensional SPECT images confirmed that high-V/Q regions were associated with regions of cystic air spaces seen on the CT scans. Nevertheless, the spatial resolution of the functional V/Q images (Fig. 2C) is not sufficiently good for precise correlation with CT scans. Therefore, a further check was done with inspiratory and expiratory CT scans that showed changes in cross-sectional area of the cystic spaces. We took great care to match the inspiratory and expiratory scans in terms of bronchial and mediastinal anatomy because the lungs move cranially on expiration. It is true that a change in cross-sectional area of a lung region on a CT scan does not necessarily reflect a change in its volume because of possible changes in shape [14]. With small cystic spaces (unlike giant bullae), reconstruction of volume change would be very inaccurate with 10-mm sections. Contiguous thin sections (2 or 3 mm) were not used because of the additional radiation exposure. In spiral CT mode, 2-mm reconstructions can be performed, but the spatial resolution is not sufficient for volumetric reconstruction. Granted these reservations, the combination of normal findings on ventilation scans during tidal breathing and normal deflation seen on serial CT scans is sufficient evidence that cystic spaces in patients with fibrotic lung disease ventilate normally.

V/Q mismatch (high V/Q) on ventilation and perfusion scintigrams has been reported in conditions other than pulmonary embolic disease [6]. The most common causes are carcinoma of the bronchus, radiation therapy, and vasculitis. V/Q mismatch in two cases of fibrotic sarcoidosis has been seen [7]. Single case reports [18, 19] of V/Q mismatch in idiopathic pulmonary fibrosis without CT scans, but with normal findings on a pulmonary angiogram in one instance [18], have been published. In a conference report [20], V/Q scanning in idiopathic pulmonary fibrosis was said to show ventilatory defects in 70% of cases, but no correlation with CT findings was done. The important point is that V/Q mismatch on scintigrams must be interpreted in conjunction with the findings on plain chest radiographs and, where appropriate, the thoracic CT scan. In our study, we found that the zones of V/Q mismatch corresponded to the presence of cystic air spaces (honeycomb lung) seen on CT. In addition, many of the more elderly patients have smoked for many years and may have air-flow obstruction. In such cases, matched V/Q defects may be seen.

Clearly, the V/Q mismatch pattern on scintigrams may suggest pulmonary embolism. In a patient with idiopathic pulmonary fibrosis who is being examined for unexplained breathlessness, and in whom a V/Q scan shows the classic mismatched pattern, the thoracic CT scan might be informative. If the CT scan showed cystic air spaces in the mismatched regions, this would adequately explain the perfusion defect; but if the lung parenchyma lacked cystic air spaces in the relevant area, the diagnosis of pulmonary embolism should be entertained.

An increased physiological dead space (V_D), both absolute and as a fraction of the tidal volume (V_T), is a feature of idiopathic pulmonary fibrosis [4, 20] and the Hamman-Rich syndrome [5]; V_D/V_T does not decrease on exercise as it does in healthy subjects. We have confirmed this in three of our subjects (mean V_D/V_T on exercise, 0.59; normal, <0.25) and in many others who were not part of this series. No convincing explanation has been offered for the high V_D/V_T on exercise in patients with idiopathic pulmonary fibrosis [4, 5]. In those who have gross lung restriction with a predicted tidal capacity of less than 50%, failure of the tidal volume to increase on exercise might be a contributing factor. This was not the case in our three patients, whose mean tidal capacity was 81% predicted and whose exercise V_T exceeded 1.5 l. The regional V/Q mismatch with high-V/Q units, as seen in this series, would be an adequate explanation.

Many of the patients with idiopathic pulmonary fibrosis have been cigarette smokers for many years, and they may have a moderate degree of air-flow obstruction. Can emphysematous areas be distinguished from the cystic air-space abnormalities of idiopathic pulmonary fibrosis? At the extremes, there is no problem. Generalized emphysema is associated with airway narrowing and collapse on expiration, a high residual volume (RV) due to air trapping, and a failure of the bullous spaces to deflate on expiration [20]. The combination of localized emphysema and pulmonary fibrosis leads to a more subtle picture. As in patient no. 3 (Figs. 1E–1H), pulmonary function tests may show only moderate air-flow obstruction (FEV_1/VC = 0.56) and no air trapping (low RV). In eight cases of combined idiopathic pulmonary fibrosis and emphysema, described by Wiggins et al. [8], FEV_1/VC and RV ranged from moderately obstructed with air trapping to completely normal. In fact, emphysematous lungs (and their associated bullae) have an increased compliance and could deflate more easily than normal lung were it not for the lack of parenchymal support that permits their airways to narrow and collapse, preventing normal expiration. The presence of fibrosis will tend to hold airways open and permit emphysematous regions to empty more completely than they otherwise would (Figs. 1E, 1F, 3A, and 3B).

Thus, the V/Q scan can be used to distinguish between emphysema and idiopathic pulmonary fibrosis in the mixed

case. In regions where emphysema predominates, the V/Q scan will tend to show a matched pattern with reduced ventilation (Figs. 1G and 1H) but in areas where fibrosis and cystic air spaces are dominant, a mismatched high-V/Q pattern will be seen. This may be a better discriminator than the CT findings alone.

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