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# Pulmonary alveolar microlithiasis : lung function in five cases[1]

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Pulmonary function was investigated in five patients with pulmonary alveolar microlithiasis. Four male cases occurred in two families. This contrasts with previous reports that females predominate in familial cases. Only one patient had respiratory symptoms. Total lung capacity was reduced in all patients and vital capacity was less than normal in four of five patients. The ratio of residual volume to total lung capacity and the one second forced expiratory volume was normal in all patients. Frequency of respiration and ventilatory equivalent were greater than normal and tidal volume was less than normal in the symptomatic patient. Minute volume of ventilation, $O_2$ consumption, and alveolar $O_2$ tension were normal in all patients. Arterial $O_2$ tension was less than normal in one patient and became less than normal during exercise in another patient. Arterial $CO_2$ was low in one patient and arterial *p*H was normal in all. The alveolar arterial $O_2$ gradient was greater than normal in all patients ; and the venous admixture-like effect was increased in three patients. The ratio of physiological dead space to tidal volume was greater than normal in two patients and the $O_2$ diffusing capacity was less than normal in one of three patients. Pulmonary function studies reported previously showed no specific pattern. All patients reported herein revealed a definite restrictive pattern with decreased lung volumes, absent airway obstruction, and uneven distribution of pulmonary capillary blood evidenced by an increased alveolar arterial $O_2$ tension gradient.

Pulmonary alveolar microlithiasis is a relatively rare disease of unknown aetiology, characterized by the presence of multiple microscopic stones within the pulmonary alveoli. The striking radiological picture often contrasts with mild or even absent symptoms except late in the course of the disease.

The radiological and clinical features of the disease have been well described (Sosman, Dodd, Jones, and Pillmore, 1957 ; Viswanathan, 1962 ; Yang and Lin, 1963). Detailed pulmonary function studies have received somewhat less attention.

The purpose of this communication is to present respiratory function data in five cases of pulmonary alveolar microlithiasis, and to review some results of pulmonary function tests reported in the literature.

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[2]Details of the radiological features of these patients are the subject of a separate communication (Balikian *et al.* in preparation).

## MATERIALS AND METHODS

Five patients (one female and four males) were studied. Patients 2 and 3 were brothers, as were patients 4 and 5. The diagnosis was made on the basis of a typical radiological picture of diffuse innumerable infiltrates of calcific density and was confirmed by lung biopsy in patient 1 as well as in one of each of the two pairs of brothers (patients 2 and 4). The age, sex, physical characteristics, and summary of the clinical findings are shown in Table I. The Figure shows a typical chest radiograph.[2]

Patient 1, a 15-year-old girl, was pale and had had a poor appetite since the age of 10 years. A paternal uncle had proven pulmonary tuberculosis, but the family history was otherwise negative. In view of a positive history of pulmonary tuberculosis a chest radiograph was taken at the age of 11 and revealed laminated deeply eosinophilic calcified nodules. She had no cough, sputum production, haemoptysis, dyspnoea or fever. Physical examination was completely negative. The haemoglobin was 14 g./100 ml., the haematocrit 40% and the leucocytes 8,000/cu. mm., with 55% polymorphonuclear leucocytes, 40% lymphocytes, and 5% monocytes. The erythro-

## TABLE I

AGE, SEX, PHYSICAL CHARACTERISTICS, AND SUMMARY OF CLINICAL FINDINGS

| Case | Age (yrs) | Sex | Height (cm.) | Weight (kg.) | Presenting Symptom | Cough | Dyspnoea | Clubbing | Cyanosis | Physical Examination | Chest Radiograph | Lung Biopsy |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1* | 15 | F | 126 | 25 | Pallor | None | None | None | None | Negative | Fine granular calcific densities throughout lungs | Alveolar microlithiasis |
| 2 | 17 | M | 168 | 65 | None. Discovered on routine chest radiographs | None | None | None | None | Few crepitant rales left base | Fine granular calcific densities throughout lungs | Alveolar microlithiasis |
| 3 (Brother of case 2) | 6 | M | 108 | 21 | None. Discovered on investigating family | None | None | None | None | Negative | Fine granular calcific densities throughout lungs | Not done |
| 4 | 15 | M | 135 | 30 | Dyspnoea on exertion | Productive cough several years | Exertional | Present | None | Underdeveloped. Bilateral basal crepitant rales | Fine granular calcific densities with nodular accentuation throughout lungs | Alveolar microlithiasis |
| 5 (Brother of case 4) | 17 | M | 153 | 44 | None. Discovered on investigating family | None | None | None | None | Negative | Fine granular calcific densities with nodular accentuation throughout lungs | Not done |

* Diagnosis was established at age 11 in 1963.

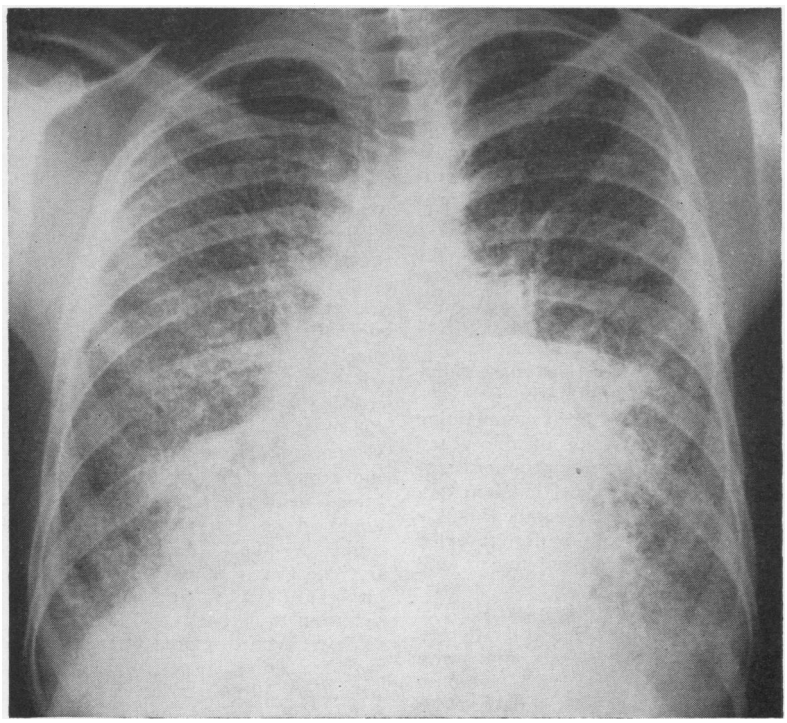

FIGURE. *Typical radiograph (case 2) showing fine granular calcific densities throughout both lungs.*

cyte sedimentation rate was 40 mm./hour, the serum calcium 9·7 mg./100 ml., phosphorus 5·4 mg./100 ml., alkaline phosphatase 8·2 shinawara units, albumin 3·3 g./100 ml., and globulins 2·7 g./100 ml. The urine examination was negative and the stools revealed ascaris ova. The P.P.D. (intermediate strength) was positive. Her parents and two brothers were living and well, and had negative chest radiographs. An open lung biopsy was done and showed numerous round laminated deeply eosinophilic calcified nodules. The pathological diagnosis was alveolar microlithiasis.

Patient 2, a 17-year-old boy, was to be enrolled in a Government school where all new students had a chest radiograph. This showed diffuse fine infiltrates of calcific density. He denied cough, sputum production, haemoptysis, dyspnoea, weight loss, fever or night sweats.

Physical examination revealed fine crepitations over the left lung base but was otherwise negative.

Haemoglobin was 14·6 g./100 ml., red blood cells 3,890,000/cu. mm., leucocytes 6,500/cu. mm., with 74% polymorphonuclear leucocytes, 18% lymphocytes, 4% monocytes, and 4% eosinophils. Erythrocyte sedimentation rate was 6 mm./hour. The stools showed ascaris ova and the urine examination was negative. P.P.D. (intermediate strength) was negative. A thoracotomy and lung biopsy were performed. The sections showed poorly expanded alveolar spaces occupied by round, laminated, deeply eosinophilic calcific nodules, with thickened alveolar septae but no inflammatory exudates. The histological diagnosis was alveolar microlithiasis. Chest radiographs were taken on the parents and all five siblings and were negative in all but the youngest brother (patient 3).

Patient 3, the 6-year-old brother of patient 2, was discovered accidentally during the radiological investigation of the family. He had no cough, sputum production, haemoptysis, dyspnoea, weight loss, fever or night sweats.

Physical examination revealed two small, freely movable lymph nodes at both angles of the jaw, each measuring 1 cm. in diameter. There was a grade I ejection type apical systolic cardiac murmur. The rest of the examination was negative. The chest radiograph was typical of alveolar microlithiasis.

Patient 4, a 15-year-old boy, had been complaining of dry cough and dyspnoea on exertion of nine years' duration. The cough was occasionally productive of scanty mucoid sputum. There was no haemoptysis, orthopnoea, nocturnal paroxysmal dyspnoea, ankle oedema, fever or night sweats. Examination revealed a thin underdeveloped boy with clubbed fingers and toes. The lungs revealed a few bilateral basal crepitations. The rest of the examination was negative. The haemoglobin was 15 g./100 ml., red blood cells 4,000,000/cu. mm., leucocytes 11,500/cu. mm. with 72% polymorphonuclear leucocytes, 18% lymphocytes, 2% monocytes, and 8% eosinophils. The erythrocyte sedimentation rate was 65 mm./hour. Urine examination was negative. The stools revealed ascaris ova. Venous pressure was 12 cm. of saline.

The P.P.D. (intermediate strength) was negative. A thoracotomy and lung biopsy were performed. The biopsy revealed numerous round, laminated, deeply eosinophilic, calcified intra-alveolar nodules: the alveolar walls were slightly thickened, but no inflammatory changes were noted. The histological diagnosis was pulmonary alveolar microlithiasis. Chest radiographs were taken on the parents and all seven siblings, and were negative in all but the oldest brother (patient 5).

Patient 5, the 17-year-old brother of patient 4, was discovered accidentally during the radiological investigation of the family. He was completely free of symptoms and his physical examination was entirely negative. His chest radiograph was typical of pulmonary alveolar microlithiasis.

The following pulmonary function studies were performed. Total lung capacity (T.L.C.) and residual volume (R.V.) were measured by helium dilution (Meneely and Kaltreider, 1949). The forced vital capacity (F.V.C.) and the one-second forced expiratory volume (F.E.V.) were measured with a 9-litre Collins respirometer. The predicted normal T.L.C. was obtained from the equations of Comroe, Forster, DuBois, Briscoe, and Carlsen (1962) and the predicted normal F.V.C. from the equations of Stewart (1922). Both of these sets of normal standards were found to agree closely to measurements made by us on normal Lebanese subjects of comparable age and sex.[3]

A Riley indwelling arterial needle was introduced into the brachial artery and expired gas and arterial blood were collected simultaneously (Lilienthal, Riley, Proemmel, and Franke, 1946). Patients 2, 4, and 5 were then exercised on a motor-driven treadmill and expired gas and arterial blood were similarly collected.

Expired $O_2$ and $CO_2$ were analysed in a Scholander microgas analyser (Scholander, 1947). Arterial $O_2$ tension ($P_{AO_2}$) was measured with an $O_2$ macroelectrode (Severinghaus and Bradley, 1958). Arterial $CO_2$ tension ($P_{ACO_2}$) was measured with a Severinghaus $CO_2$ electrode (Severinghaus and Bradley, 1958). Arterial *p*H was measured with a Radiometer micro glass electrode. The ventilation equivalent was calculated in the manner described by Comroe (1951). The alveolar $O_2$ tension ($P_{AO_2}$) was calculated from the alveolar air equation (Riley and Cournand, 1951). Physiological dead space ($V_D$) was calculated using the Bohr equation (Riley and Cournand, 1949). The venous admixture-like effect (Qva/Qt) was estimated from the line charts of Riley. A resting $O_2$ diffusing capacity ($D_{O_2}$) (Riley, Cournand, and Donald, 1951) was determined in patients 1 and 4 and a maximal $D_{O_2}$ (Riley, Shepard, Cohn, Carroll, and Armstrong, 1954) was determined in patient 2. In view of the young age of patient 3 (6 years), it was not possible to determine the T.L.C. or R.V. or to obtain arterial blood. $D_{O_2}$

[3]Unpublished data

could not be determined in patient 5 because he fainted during low $O_2$ breathing.

## RESULTS

The results of the pulmonary function studies are shown in Tables II and III. T.L.C. was greatly reduced in patient 4 and slightly reduced in the others. R.V. and the ratio of R.V. to T.L.C. was normal in all patients tested. F.V.C. was markedly decreased in patient 4, slightly decreased in patients 1, 2, and 5, and normal in patient 3. F.E.V. was normal in all patients.

Frequency of respiration and ventilatory equivalent were greater than normal, and tidal volume was less than normal, during rest and exercise in patient 4, but were normal in the others. Minute volume of ventilation, $O_2$ consumption, and alveolar $O_2$ tension were normal in all patients.

The alveolar arterial $O_2$ gradient was greater than normal (9 mm. Hg) in all patients. In general this gradient increased further during exercise.

$Pa_{O_2}$ was less than normal (78 mm. Hg) at rest in patient 4 and decreased further during exercise. $Pa_{O_2}$ was normal at rest in the others but decreased during exercise in patient 2. $Pa_{CO_2}$ was less than normal at rest in patient 1, and increased during exercise in patient 2. Arterial *p*H was normal in all patients. Arterial $O_2$ saturation was normal (92%) in all patients at rest. It decreased markedly in patient 4 and slightly in patient 2 during exercise.

Qva/Qt was slightly greater than normal in patient 1 and definitely greater than normal in patients 4 and 5. During exercise Qva/Qt increased further in patient 4 and returned towards normal in patient 5. The ratio of $V_D$ to tidal volume ($V_T$) was greater than normal at rest in patient 2, but returned to normal during exercise. Patient 4 had an abnormal $V_D/V_T$ during both rest and exercise. $Do_2$ was less than normal in patient 4 and was normal in patients 1 and 2.

## DISCUSSION

Four of our five cases occurred in two families. This tendency for familial occurrence has been well recognized (Rotem, Solomon, and Hertz-Frankenhuis, 1963; Yang and Lin, 1963; Oka, Shiraishi, Ogata, Goto, Yasuda, and Yanagihara, 1966); however, in contrast to Rotem's finding that females predominated in familial cases, our

TABLE II

LUNG VOLUMES AND FLOW RATES

| Case | T.L.C. (l.) | % of Predicted | R.V. (l.) | $\frac{\text{R.V.}}{\text{T.L.C.}} \times 100$ | F.V.C. (l.) | % of Predicted | F.E.V. (l.) | % of F.V.C. |
|---|---|---|---|---|---|---|---|---|
| 1 | 2·61 | 78 | 0·54 | 21 | 2·07 | 77 | 1·68 | 81 |
| 2 | 4·61 | 85 | 1·04 | 23 | 3·57 | 82 | 3·05 | 86 |
| 3 | — | — | — | — | 1·18 | 94 | 1·12 | 95 |
| 4 | 1·64 | 39 | 0·35 | 21 | 1·29 | 39 | 1·23 | 94 |
| 5 | 3·92 | 81 | 0·87 | 22 | 3·05 | 78 | 2·56 | 84 |

T.L.C.=Total lung capacity; R.V.=residual volume; F.V.C.=forced vital capacity; F.E.V.=one second forced expiratory volume.

TABLE III

VENTILATION, ARTERIAL BLOOD GASES, VENTILATION PERFUSION RELATIONSHIPS, AND $O_2$ DIFFUSING CAPACITY

| | Case | | | | | | |
|---|---|---|---|---|---|---|---|
| | 1 | 2 | | 4 | | 5 | |
| | Rest | Rest | Exercise | Rest | Exercise | Rest | Exercise |
| $V_T$ (tidal volume) (l.) | 0·352 | 0·341 | 1·168 | 0·255 | 0·645 | 0·536 | 1·425 |
| f (respiratory frequency/min.) | 18 | 25 | 33 | 41 | 55 | 15 | 58 |
| $\dot{V}_E$ (minute ventilation) (l./min.) | 6·34 | 8·52 | 38·53 | 10·46 | 35·50 | 8·04 | 82·63 |
| $\dot{V}o_2$ ($O_2$ consumption) (l./min.) | 0·206 | 0·261 | 1·711 | 0·219 | 0·895 | 0·275 | 2·324 |
| Ventilatory equivalent (l. $\dot{V}_E$/100 ml. $\dot{V}o_2$) | 3·08 | 3·26 | 2·25 | 4·78 | 3·97 | 2·92 | 3·56 |
| $PA_{O_2}$ (alveolar $O_2$ tension) (mm. Hg) | 114 | 104 | 98 | 107 | 110 | 106 | 117 |
| A–a gradient (alveolar–arterial $O_2$ gradient) (mm. Hg) | 23 | 13 | 18 | 33 | 60 | 26 | 34 |
| $Pa_{O_2}$ (arterial $O_2$ tension) (mm. Hg) | 91 | 91 | 80 | 74 | 50 | 80 | 83 |
| $Pa_{CO_2}$ (arterial $CO_2$ tension) (mm. Hg) | 32 | 41 | 46 | 39 | 38 | 39 | 33 |
| Arterial *p*H | 7·45 | 7·43 | 7·39 | 7·42 | 7·38 | 7·43 | 7·35 |
| $Sa_{O_2}$ (arterial $O_2$ saturation) (%) | 97 | 97 | 94 | 94 | 85 | 95 | 96 |
| Qva/Qt (venous admixture % of cardiac output) | 9 | 6 | 6 | 17 | 35 | 13 | 9 |
| $V_D/V_T$ (physiological dead space % or tidal volume) | 22 | 39 | 24 | 38 | 38 | 25 | 18 |
| $Do_2$ ($O_2$ diffusing capacity) (ml./mm. Hg/min.) | 18 | — | 72* | 7 | — | — | — |

* Maximal $O_2$ diffusing capacity.

four patients who exhibited a familial occurrence were all males.

The parents and all three siblings of patients 2 and 3 as well as the parents and all 7 siblings of patients 4 and 5 were free of the disease. Unfortunately other more distant members of the two families were not available for study so that the occurrence of other cases in those relatives cannot be ruled out.

It has been reported that most patients were over the age of 30 when the diagnosis was made (Sosman *et al.*, 1957; Viswanathan, 1962; Yang and Lin, 1963). Our patients were all under 20 years of age with a range of 6–17 years (Table I). This might suggest that the disease may not be too far advanced in our patients. Yet patient 4, though only 15 years old, had definite dyspnoea on exertion and showed marked disturbance of pulmonary function. It might be that in his case the disease was progressing at a more rapid rate than usual.

A review of the literature revealed that Finkbiner, Decker, and Cooper (1957), Thomson (1959), Viswanathan (1962), Rotem *et al.* (1963), Lebacq, Lauweryns, and Billiet (1964), and Oka *et al.* (1966) were the only workers to perform extensive pulmonary function tests on a total of seven patients. The majority of other investigators reported on isolated pulmonary function measurements.

The results of some pulmonary function tests obtained by other workers are shown in Table IV. Two out of five patients studied by others had reduced T.L.C. (Finkbiner *et al.*, 1957; Thomson, 1959; Lebacq *et al.*, 1964; Oka *et al.*, 1966). All our patients had some reduction in T.L.C., indicating a decrease in functioning lung volume.

Five out of eight patients studied by others had an increased ratio of R.V./T.L.C. (Finkbiner *et al.*, 1957; Sosman *et al.*, 1957; Thomson, 1959; Viswanathan, 1962; Varma, 1963; Lebacq *et al.*, 1964; Oka *et al.*, 1966), whereas all our patients had normal R.V./T.L.C. ratios, suggesting the absence of significant hyperinflation.

Twelve out of 16 patients reported in the literature had reduced vital capacities (Badger, Gottlieb, and Gaensler, 1955; Sosman *et al.*, 1957; Abdel-Hakim, El-Mallah, Hashem, and Abdel-Halim, 1959; Thomson, 1959; Tezok, Balci, Baris, and Kurt, 1965; Viswanathan, 1962; Rotem *et al.*, 1963; Lebacq *et al.*, 1964). Four of our five patients had reduced F.V.C. consistent with an encroachment on functioning lung volume. Airway obstruction, evidenced by decreased F.E.V., was found in four of 11 patients studied by others (Finkbiner *et al.*, 1957; Sosman *et al.*, 1957; Abdel Hakim *et al.*, 1959; Viswanathan, 1962; Rotem *et al.*, 1963; Varma, 1963; Lebacq *et al.*, 1964; Oka *et al.*, 1966). All our patients, on the other hand, had normal F.E.V., suggesting normal calibre and patency of the airways.

Only one of our patients (patient 4) had a low $Pao_2$ at rest which decreased further on exercise, suggesting a diffusion defect. This is in agreement with the findings in Viswanathan's (1962) patient. Similarly, only one of our patients (patient 1) had a decreased $Paco_2$ probably due to psychogenic hyperventilation. A normal $Paco_2$ in the majority of the patients studied may be of special significance since it has been suggested (Badger *et al.*, 1955) that a decrease in $CO_2$ may predispose to calcium deposition in the lung in alveolar microlithiasis.

In contrast to the finding of others (Finkbiner *et al.*, 1957; Sosman *et al.*, 1957; Thomson, 1959; Rotem *et al.*, 1963), all our patients had normal arterial $O_2$ saturation at rest.

Two of our patients (patients 2 and 4) had an increased ratio of physiological dead space to tidal volume. This is in agreement with the findings of Sosman *et al.* (1957) and of Viswanathan (1962), and suggests the presence of alveoli that are hyperventilated in relation to their pulmonary capillary blood flow.

The A-a $O_2$ gradient was greater than normal in all, and the Qva/Qt was increased in three of our patients. Both these findings suggest the presence of alveoli that are hypoventilated in relation to their pulmonary capillary blood flow leading to some decrease in $O_2$ tension in the mixed arterial blood.

Four out of six patients studied by others had reduced diffusing capacities (Finkbiner *et al.*, 1957; Thomson, 1959; Varma, 1963; Lebacq *et al.*, 1964; Oka *et al.*, 1966), whereas only one of our patients had a reduced diffusing capacity. This discrepancy may have resulted from differences in technique. We had used the $O_2$ diffusing capacity measurement while the other workers had used the more sensitive carbon monoxide method.

In conclusion, the pulmonary function studies reported in the literature show no consistent pattern. The results have ranged from the entirely normal to a marked reduction of lung volumes, arterial $O_2$ unsaturation, increased venous admixture, and impaired gas exchange. All our patients, on the other hand, showed a reduction in T.L.C. with no increase in R.V. Four of our five

## TABLE IV

### RESULTS OF PULMONARY FUNCTION TESTS REPORTED BY OTHER WORKERS

| Investigator | Subject | | T.L.C. (l.) | | R.V. (l.) | | R.V./T.L.C. (%) | F.V.C. (l.) | | F.E.V. (% of F.V.C.) | $PaO_2$ (mm. Hg) | | $PaCO_2$ (mm. Hg) | | $So_2$ (%) | | *p*H | | $V_D$ (% of tidal volume) | A–a Gradient (mm. Hg) | Qva/Qt (% cardiac output) | $D_{LCO}$ (ml./mm. Hg/min.) |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | Age (yrs) | Sex | Observed | Predicted Normal | Observed | Predicted Normal | | Observed | Predicted Normal | | Rest | Exercise | Rest | Exercise | Rest | Exercise | Rest | Exercise | | | | |
| Abdel-Hakim *et al.* (1959) .. | 30 | M | | | | | | 3·49 | N.G. | | | | | | | | | | | | | |
| Badger *et al.* (1955) .. | 45 | M | | | | | | 2·13 | 3·80 | | | | | | | | | | | Increased* | | |
| Finkbiner *et al.* 1957) .. | 31 | M | 3·8 | 5·43 | 1·59 | 1·1 | 42 | 2·18 | 4·27 | | | | 40 | 40 | 91·2 | 88·4 | 7·33 | 7·34 | | | | 10·2–12·6 |
| Lebacq *et al.* (1964) | 54 | M | 5·69 | 5·19 | 2·07 | 1·20 | 37 | 3·62 | 3·99 | 69 | | | 38 | | 96 | 96·5 | 7·44 | | | | | 19·6 |
| Oka *et al.* | 20 | F | 4·33 | 3·90 | 1·44 | 0·78 | 33 | 2·90 | 3·12 | 74 | | | | | | | | | | | | 19 |
| | 15 | F | 3·85 | 3·60 | 0·65 | 0·72 | 20 | 3·20 | 2·88 | 82 | | | | | | | | | | | | 19 |
| Rotem *et al.* (1963) .. | 6 | M | | | | | | 0·83 | 1·10 | 86 | | | | | 95 | 91 | | | | | | |
| Sosman *et al.* (1957) .. | 37 | M | | | | | | 3·94 | 3·86 | 63 | | | | | 89·6 | | | | | | | |
| | 29 | F | | | | | 36 | 2·52 | 3·15 | | | | 40 | | 97·1 | 96·4 | | | 28 | 7 | | |
| | 28 | M | | | | | | 1·8 | N.G. | | | | | | | | | | | | | |
| | 40 | M | | | | | | 2·9 | N.G. | | | | | | | | | | | | | |
| | 32 | F | | | | | | 1·2 | N.G. | | | | | | | | | | | | | |
| | 40 | M | | | | | | 2·3 | N.G. | | | | | | | | | | | | | |
| | 8 | M | | | | | | 2·39 | N.G. | | | | | | | | | | | | | |
| | 19 | F | | | | | | Decreased* | | | | | | | | | | | | | | |
| | 10 | F | | | | | | 1·65 | 1·94 | | | | | | | | | | | | | |
| | 17 | M | | | | | | 1·64 | N.G. | | | | | | | | | | | | | |
| | 59 | M | | | | | | 2·7 | N.G. | | | | | | | | | | | | | |
| Tezok *et al.* (1965) .. | 28 | M | | | | | | 3·2 | 3·27 | | | | | | | | | | | | | |
| Thomson (1959) | 35 | M | 4·6 | 5·7 | 1·97 | 1·5 | 45 | 2·67 | 3·6 | | | | | | 89 | 55 | | | | | 25 | 4·2–5·8 |
| Varma (1963) .. | 13 | M | 2·161 | N.G. | 0·53 | N.G. | 25 | 1·63 | N.G. | 96 | | | | | | | | | | | | 14 |
| Viswanathan (1962) .. | 43 | M | 4·148 | N.G. | 2·278 | N.G. | 55 | 1·87 | N.G. | | 72 | 62 | 46 | 42 | | | | | 44 | | | |

T.L.C. = Total lung capacity; R.V. = residual volume; F.V.C. = forced vital capacity; F.E.V. = one second forced expiratory volume; $PaO_2$ = arterial $O_2$ tension; $PaCO_2$ = arterial $CO_2$ tension; $So_2$ = arterial $O_2$ saturation; A–a = alveolar arterial $O_2$ gradient; Qva/Qt = venous admixture; $D_{LCO}$ = diffusing capacity; N.G. = not given.

* Actual values not given.

patients had a reduction in F.V.C. with a normal F.E.V. This is consistent with a restrictive pattern of lung disease and an absence of significant airway obstruction. Furthermore, all our patients (1, 2, 4, and 5) who had undergone arterial blood studies showed a definite increase in A-a $O_2$ gradient, and three out of four had an increase in venous admixture. These findings indicate the presence of alveoli that are perfused but not ventilated. This suggests that the microliths may have occluded several alveoli, thereby decreasing their ventilation without a concomitant reduction in perfusion. Our patients therefore exhibited a definite restrictive pattern with decreased lung volumes, absent airway obstruction, a definite unevenness of distribution of pulmonary capillary blood flow, as well as some evidence of uneven distribution of inspired gas (patients 2 and 4) and in the most advanced case (patient 4) a severe impairment of diffusion.

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