survived, and 55 of them have no sequelae. The most difficult patients are those who are under 3 years of age and are unconscious on admission, and none of those so far have made a complete recovery. Intrathecal streptomycin treatment has not been abandoned, but the minimum course is now reduced to 10 injections. The total duration of treatment is six months or longer, but most of this time the children are ambulant and either are treated at home or receive regular schooling in hospital. Systemic corticosteroids are used in children under 1 year of age or in those who are unconscious on admission; intrathecal hydrocortisone is of value in threatened or established spinal block.

The work on which the paper is based has been continuously in progress over 12 years. It would never have been possible without the continuous help and confidence of Professor R. S. Illingworth and without the selfless contribution of many colleagues in the region who referred their patients to us for treatment.

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UNILATERAL EMPHYSEMA

BY

R. F. FOUCHÉ, M.R.C.P.Ed., D.M.R.D.

Registrar, Radiodiagnostic Department, London Hospital

J. R. SPEARS, F.F.A.R.C.S.

Senior Registrar in Anaesthetics, London Hospital

AND

COLIN OGILVIE, M.D., M.R.C.P.

*Consultant Physician, United Liverpool Hospitals and
 Regional Thoracic Surgical Centre; Late Lecturer in
 Medicine, London Hospital*

An apparent increase in the transradiancy of one lung may be due to rotation, to diminished contralateral transradiancy, to giant cysts or bullae, to compensatory emphysema, or to obstructive emphysema resulting from incomplete bronchial occlusion. Macleod (1954) described a group of cases showing abnormal transradiancy of one lung in which none of these causes was apparent. The purpose of this paper is to report the radiological and physiological characteristics of seven patients conforming to Macleod's original description. These patients had all been referred with respiratory symptoms to the out-patient clinics of the London Hospital over a period of two years.

Methods

The study of each patient included clinical history and examination, postero-anterior radiographs in inspiration and expiration, fluoroscopy, whole-lung tomography, and either bronchography or bronchoscopy. The following physiological measurements were made:

1. The subdivisions of the lung volume, residual volume being measured by a closed-circuit helium-dilution technique (Bates and Christie, 1950).

2. The maximum breathing capacity expressed as the maximum voluntary ventilation at a respiratory rate of 80 per minute (M.V.V.80). This was repeated after an inhalation of 1% isoprenaline sulphate to assess the degree of reversible bronchial obstruction.

3. The maximum inspiratory and expiratory flow rates, derived from the spirometric tracing of a forced inspiration and expiration respectively.

4. Mixing efficiency, calculated from the helium-dilution curve (Bates and Christie, 1950).

FIG. 1.—Case 4. Postero-anterior radiograph showing increased transradiancy of right lung.

FIG. 2.—Case 2. Whole-lung tomogram showing diminished vascularity of right lung most marked in middle and lower zones. Note also low flat diaphragm on this side.

5. The diffusing capacity, measured by the single-breath carbon-monoxide technique (Ogilvie, Forster, Blakemore, and Morton, 1957).

6. The differential function of the two lungs was measured by bronchspirometry in one patient.

Results

The radiological findings are recorded in Table I and Figs. 1 and 2, and the physiological findings in Tables II and III and Figs. 3 and 4.

There were five men and two women, with an average age of 52 (range 38–70 years). All gave a long history of recurrent cough and exertional dyspnoea, and six had suffered attacks of pneumonia or pleurisy in earlier life, four of them in childhood. Except for one patient (Case 6) with severe disability, dyspnoea was of mild to moderate degree (grade 1 or 2). Diminished breath sounds over a part or the whole of the affected lung were noted in each patient.

TABLE I.—*Clinical and Radiological Findings in Seven Patients with Unilateral Emphysema*

Case No.	Age	Sex	Past Health	Transradiant Lung	Diaphragm on Abnormal Side	Resting Mediastinal* Position	Main Site of Narrowed Vessels	Bronchography
1	70	M	Pneumonia twice	Right	Low, diminished movement	Right	R.M.Z. R.L.Z.	—
2	51	M	“ three times in childhood	“	Low, flat, diminished movement	Central	“ “	R. lower lobe bronchial crowding. No obstruction or bronchiectasis
3	46	F	“ Born with bronchitis.”	“	“ “ “	Left	“ “	Normal apart from poor peripheral filling
4	46	F	Pleurisy at 6 years Pneumonia at 9 months. R. pneumonia at 35 years	“	Diminished movement	Right	Throughout R. lung	R. lower lobe bronchial crowding. No obstruction or bronchiectasis
5	38	M	Pneumonia in childhood. Off school with chest trouble from 5 to 7 years	“	Low, flat, diminished movement	Central	R.M.Z. R.L.Z.	—
6	55	M	Pneumonia at 25 years	Left	Diminished movement	“	L.U.Z. L.M.Z.	—
7	60	M	—	Right	“ “	Right	R.M.Z. R.L.Z.	—

* Mediastinal deviation toward the affected side on inspiration and away on expiration was noted in every case.

TABLE II.—*Pulmonary Function Tests in Seven Patients with Unilateral Emphysema*

Case No.	Vital Capacity (ml.)	Funct. Residual Capacity (ml.)	Residual Volume (ml.)	Total Lung Capacity (ml.)	R.V.: T.L.C. Ratio	Maximum* Breathing Capacity (l./min.) (M.V.V.80)	Swept Fraction	Maximum Inspiratory Flow Rate (l./min.)	Maximum Expiratory Flow Rate (l./min.)	Mixing Efficiency	Diffusing Capacity (% of Normal)
1	2,900	3,075	2,475	5,375	46%	45 (52)	18%	102	95	15%	41%
2	2,620	3,550	2,950	5,570	52%	75 (91)	34%	300	56	15%	80%
3	1,850	3,220	2,570	4,220	61%	49 (54)	27%	150	107	15%	110%
4	1,805	2,600	1,870	3,675	51%	52 (64)	31%	105	48	46%	83%
5	3,285	3,865	3,030	6,315	48%	62 (73)	21%	150	88	18%	83%
6	2,445	6,268	5,403	7,848	69%	26 (33)	13%	167	17	18%	55%
7	3,680	—	—	—	—	72 (81)	30%	221	92	—	77%
Mean ..	2,655	3,763	3,050	5,501	55%	54 (64)	25%	171	70	21%	76%

* Figures in parentheses indicate M.B.C. after isoprenaline.

FIG. 3.—Bronchspirometric studies in Case 2.

Chest radiographs (Figs. 1 and 2 and Table I) showed increased transradiancy of the right lung in six patients and of the left lung in one (Case 6). Calcified hilar glands were observed on the affected side in three patients, and in one (Case 6) a strand of fibrosis extended from the hilum into the upper lobe. In each patient there was diminished excursion of the diaphragm dome on the affected side, with depression and flattening in four cases. The position of the mediastinum at rest was variable, but in every case the mediastinum deviated toward the affected side on inspiration and away from it on expiration. Whole-lung tomography demonstrated a decrease in the calibre of the main pulmonary artery and of the peripheral vessels on the affected side, with (except in Case 6) an apparent increase in calibre on the opposite side. The greatest reduction in vessel size was observed in the middle and lower zones of the right lung in five patients, throughout the right lung in one patient, and in the upper and middle zones of the left lung in one (Case 6). Bronchography revealed crowding of the right lower lobe bronchi, without apparent obstruction or bronchiectasis, in two patients, and in a third the pattern was normal, except for poor peripheral filling.

Physiological studies (Table II) revealed an increase of residual volume at the expense of vital capacity, so that in each patient the ratio of residual volume to total lung capacity was much increased. The maximum

breathing capacity (M.V.V.₈₀) was reduced in every case, and the slight increase after a bronchodilator drug (5–16 l./min.) suggested that there was only a small element of reversible bronchial obstruction.

The low "swept fraction" (D'Silva and Kazantzis, 1954) indicated that the maximum breathing capacity was impaired relatively more than the vital capacity. The reduction in maximum expiratory flow rate was in every patient greater than the

reduction in maximum inspiratory flow rate. Mixing efficiency was grossly impaired in every patient, while the diffusing capacity was within the normal range in five patients and moderately impaired in two.

Discussion

The clinical and radiological patterns observed in these seven patients are similar to the description given by Macleod (1954) and by Dornhorst, Heaf, and Semple (1957). In all three series the majority of patients complained of exertional dyspnoea; diminished breath sounds and hyperresonant percussion note were found over the affected lung. Men were affected about three times more often than women. The three series also have in common the findings of increased transradiancy of one lung, diminished hilar and peripheral vascular shadows, and poor expiratory deflation in the absence of either gross bronchial obstruction or diffuse bullous changes. In several cases poor peripheral filling or localized bronchial crowding was observed in the bronchogram. A remarkably high incidence of pneumonia in early life was noted in the series of Dornhorst *et al.* as in ours, but Macleod does not mention the previous health of his patients. Dornhorst *et al.* recorded six attacks of pneumonia in early childhood in three of their four patients, while we recorded 11 attacks in six of our seven patients, seven occurring in early childhood.

The vascular pattern in our seven patients, as revealed by whole-lung tomography, is of some interest. The calibre of the hilar and peripheral vessels was diminished on the affected side and, except for one patient (Case 6), increased on the opposite side. The lack of this contralateral increase in calibre in Case 6 may be attributed to the fact that he had severe generalized emphysema, as indicated by the physiological studies (Table II). The finding of a small main pulmonary artery on the affected side need not necessarily imply a congenital maldevelopment of this vessel, for, as Swyer and James (1953) suggested, diminution of the peripheral vascular bed from disease in the lung might result in functional shrinkage of the proximal vessels. Some evidence in favour of a mechanism of this sort has recently been obtained from animal experiments in which part of the peripheral arterial tree of one lung was occluded by embolism (Fouché and D'Silva, 1960). Our experience of one patient in whom the whole of the left lung was compressed by two large bullae suggests that the size of the pulmonary artery at the hilum may help to distinguish "Macleod's syndrome" from increased transradiancy due to giant bullae compressing otherwise normal lung. In this patient, though the affected hemithorax appeared almost uniformly transradiant, there was no reduction in the size of the pulmonary artery on this side.

FIG. 4.—Ventilatory effects of unilateral bronchial occlusion.

reduction in maximum inspiratory flow rate. Mixing efficiency was grossly impaired in every patient, while the diffusing capacity was within the normal range in five patients and moderately impaired in two.

Bronchspirometric studies (Fig. 3) in one patient (Case 2) showed that the affected lung (right) was contributing 43% of the total ventilation (normal 55%) and 39% of total oxygen uptake (normal 55%). When hyperventilation of the normal lung was induced by clamping the opposite bronchial catheter for three minutes there was little change in the resting expiratory level. Hyperventilation was achieved entirely by an increase in tidal volume without any change in respiratory rate (Fig. 4). When the bronchial catheter on the normal side was occluded

TABLE III.—*Pulmonary Function Tests: Normal Values, Unilateral Emphysema, and Generalized Emphysema*

	Vital Capacity (ml.)	Funct. Resid. Capacity (ml.)	Resid. Vol. (ml.)	R.V.: T.L.C. Ratio	Maximum Breathing Capacity (in l. min.) (M.V.V. 80)	Swept Fraction	Maximum Inspiratory Flow Rate (l./min.)	Maximum Expiratory Flow Rate (l./min.)	Mixing Efficiency	Diffusing Capacity
Normal ..	3,500	2,700	1,500	30%	120	35%	300	400	70-90%	70-120%
"Unilateral transradiancy" (7 patients)	2,655	3,763	3,050	55%	54	25%	171	70	21%	76%
Emphysema (20 patients) ..	2,380	4,960	4,025	62%	30	20%	144	30	28%	(normal in 5) 60%

Tomography has also demonstrated that the reduction of vessel calibre is not uniformly distributed throughout the lung. In five of our seven patients the vessels of the middle and lower zones of the right lung were predominantly affected. It may be relevant that this is the site most often affected by bronchial obstruction in childhood, whether due to foreign body or to tuberculous hilar lymph nodes. Six of our seven patients had one or more attacks of "pneumonia" in earlier life, and the radiographs of three of them demonstrated calcified lymph nodes at the hilum on the affected side. These observations suggest that temporary bronchial obstruction in childhood may have damaged the parenchyma of the affected portion of lung, which in later life could result in overexpansion and impaired deflation because of loss of elastic recoil in expiration. An alternative explanation in those cases showing bronchial crowding is that there is "compensatory" emphysema of the parts not subjected to obstruction.

The physiological pattern in these seven patients differs only in degree from that observed in 20 patients (of the same age group and sex distribution) with classical chronic diffuse obstructive emphysema. The values obtained in "unilateral hypertransradiancy" are, with the exception of mixing efficiency, intermediate between the normal values and those found in generalized emphysema (Table III). The greater impairment of mixing efficiency in the unilateral disease could be attributed to a gross disparity in the rate of ventilation of the two lungs.

The diffusing capacity was impaired in one patient with severe disability and physiological evidence of generalized emphysema, and in another patient, who was the oldest in the series (70 years). It was within the normal range in five of the seven patients. This finding suggests that there must still be a considerable diffusion contribution from the affected lung or that the remainder of the capillary bed has successfully accommodated the increased blood-flow diverted to it. It is thus in keeping with the radiological observation of continued perfusion of parts of the affected lung and the increased vascularity of the normal lung. The fact that the lungs of these patients are able to maintain a normal diffusing capacity also suggests that the original fault must have arisen at an early age, when the remainder of the capillary bed was capable of accommodating a considerable increase in blood-flow. After pneumonectomy for carcinoma in middle age, the reduction in diffusing capacity is commensurate with the amount of lung tissue removed (McIlroy and Bates, 1956).

Bronchospirometric studies in one patient (Case 2) showed that there was a significant, though reduced,

contribution to function on the affected side. The period of suspended oxygen uptake which followed overdistension of the abnormal lung during hyperventilation suggests that blood-flow was temporarily diverted to the opposite side. This observation, taken in conjunction with the finding of a normal total diffusing capacity, indicates that reversible functional changes during hyperventilation on exercise may be a more important cause of disability than structural diminution in the capillary bed.

Summary

The clinical, radiological, and physiological characteristics of seven patients with increased transradiancy of one lung are described.

The patients all complained of dyspnoea on exertion. Diminished breath sounds and a hyperresonant percussion note were found over the affected lung. All but one of the patients gave a history of "pneumonia" in earlier life.

Radiographic studies revealed impaired expiratory deflation and decreased hilar and peripheral vascular shadows on the affected side. These changes were observed most often in the middle and lower zones of the right lung.

The physiological defect was distinguishable only in degree from that observed in generalized emphysema. The relatively greater impairment of mixing efficiency in the unilateral disease is attributed to a gross disparity in the rate of ventilation of the two lungs. The normal values obtained for diffusing capacity in five of the seven patients suggests that the unaffected portions of the pulmonary capillary bed had successfully accommodated the increased blood-flow diverted to them. In one patient studied by bronchspirometry, hyperventilation resulted in over-inflation and temporary suspension of oxygen uptake on the affected side.

It is suggested that transient bronchial obstruction in the course of a respiratory infection in earlier life may have been responsible for the changes observed in some of these patients.

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