

of peripheral vascular resistance; on the other hand the size of the intrapulmonary arteries in life does not directly correspond to the appearance in the specimen arteriogram and seems to reflect a functional state of vasoconstriction or of relative reduction in blood flow or blood volume.

For example, when right ventricular hypertrophy was present the micropaque angiograms showed dilatation whereas in life the intrapulmonary vessels were either normal, as in chronic bronchitis, or narrow, as in emphysema, and in neither case generally dilated.

Correlation of the specimen arteriogram with the angiogram and clinical features will help to elucidate the vascular damage in lung disease and the mechanism whereby this disease produces secondary effects on the heart.

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The Pulmonary Angiogram in Chronic Bronchitis and Emphysema

Introduction

Hitherto angiography has been contraindicated in cases of chronic bronchitis and emphysema because of a supposed increased hazard over other forms of angiography and because it was thought that the information obtained was unlikely to benefit the patient.

With the development of intensive therapy units specializing in acute cardiorespiratory illnesses and the more frequent recognition of pulmonary thrombo-embolic disease (Goodwin *et al.* 1963) it became apparent that a number of different syndromes existed which exhibited very wide variation in response to the accepted therapeutic practices. It became desirable, therefore, that the maximum information should be sought in order to assist in the management of the case, both during the period of quiescence and in anticipation of the next cardiorespiratory exacerbation. In these circumstances it was felt justifiable to proceed with the investigation in a number of selected cases.

In this series of 18 patients the highest pulmonary artery pressure at rest was 52 mmHg. No symptoms directly referable to the angiogram have occurred.

Classification

Considerable variation exists in the anatomical structure of the pulmonary arterial tree and in the functional behaviour of the pulmonary vascular structures as assessed by regional pulmonary blood flows. This variation is in accord with the clinical conception of a disease spectrum, the terms 'bronchitis' and 'emphysema' embracing a number of syndromes.

It is perhaps permissible to postulate three broadly based and overlapping groups on the present evidence in which there is some correlation between the clinical and radiological features. Two commonly accepted colloquial terms attributed by Scadding (1963) to Professor A C Dornhorst, the 'pink puffer' and 'blue bloater', will be used to identify two of the three groups. The clinical characteristics and the changes in the lung function studies have been described and established by Ogilvie (1959, 1964) and by Fletcher *et al.* (1963).

Group 1: Primary Emphysema, 'Pink Puffer', &c.

The patients are usually men who develop dyspnoea in middle life, not infrequently following an acute chest illness. The lungs are over-distended, leading to considerable difficulty in ventilation. Irreversible airway obstruction is present with a greatly increased residual volume. The patients are not cyanosed and are not subject to repeated attacks of right ventricular failure. They are greatly incapacitated. In this series the pulmonary artery pressures were normal at rest but showed an abnormal and prolonged rise in pressure following exercise.

Radiological features: The radiological features, although varying in degree, are consistent throughout the group.

The plain chest X-rays show the well-established features of primary pulmonary emphysema. The thorax is distended with compression of the heart and mediastinum; the diaphragm is low and flat and shows little movement. The pulmonary conus is frequently prominent and there may be moderate enlargement of the hilar vessels. The lung fields are hypertranslucent. The mid-lung vessels are symmetrically distributed, are small in character and tend to be more numerous in the upper zones. Tomograms demonstrate a rapid attenuation in size of the primary divisions of the pulmonary arteries. Tomograms are also useful in assessing the pulmonary veins, which are small in size and relatively few in number. The pulmonary angio-

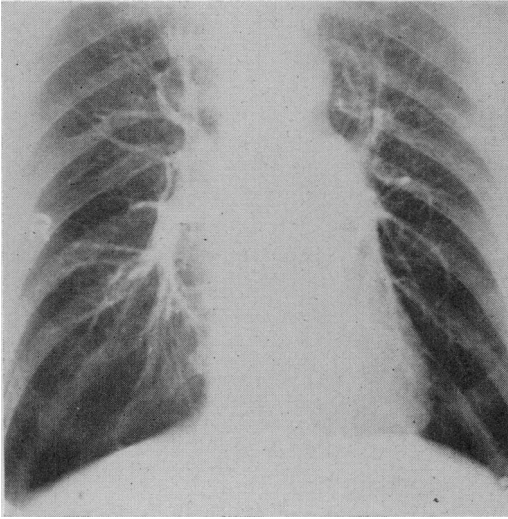


Fig 1 The arterial phase of a pulmonary angiogram in a case of primary emphysema showing rapid diminution in the calibre and number of the branches of the pulmonary arteries. The mid-pulmonary arteries tend to be rectilinear in character and show a wide angled type of branching

gram shows characteristic changes (Fig 1). There is a varied degree of enlargement of the pulmonary arteries and their primary divisions. These vessels diminish rapidly in diameter and give rise to a less than normal number of relatively small intrapulmonary arteries which then show a more gradual attenuation throughout the rest of the arborization. They are rectilinear in character or curvilinear where distorted by mechanical forces. The distal arterial tree divides in a divaricate manner, the branches being relatively few and ending in small, fine, straight terminal twigs.

There is slow penetration of the medium to the periphery of the lung and the vascularization is deficient, little developing in the way of background filling.

The veins appear late in the series and are small, poorly filled vessels. A differential exists in the rate and degree of filling between the upper and the lower zones. The upper lobe arteries and veins opacify in advance of, and to a greater extent than, those in the lower lobes. The distribution of these changes in the two lungs is roughly symmetrical and this feature is very characteristic of the group. In the earlier cases, prior to the extensive obliteration of the vascular bed, the branches are more numerous and the circulation through the upper lobes more nearly approaches the normal with good background filling and well-opacified veins.

Group 2: 'Blue Puffer', Winter Bronchitic, &c.

This is a broadly based group of patients whose clinical identity has not yet been established. The patients are frequently cyanotic and breathless; many infective episodes occur and in some cases the sputum is rarely free from infection. Most of the patients are disabled even during periods of remission and the worst cases are respiratory cripples. Pulmonary function tests are inconstant, demonstrating varying degrees of expiratory airway obstruction or alteration in the ventilation/perfusion ratios.

Vector cardiography demonstrates right ventricular hypertrophy in the majority of cases. As in the previous group a labile pulmonary arterial hypertension is present with a wide respiratory variation in the readings.

Radiological features: As might be anticipated the radiology is more varied than in the other groups.

The appearances of the chest X-ray are those, to a greater or lesser extent, of a mixed type of chronic bronchitic, emphysematous and infective elements being present in the lung fields (Scarrow 1964). In addition there are various degrees of disorganization of the normal, regular, radiating, diminishing branching pattern of the pulmonary vascular tree. There is usually some right heart enlargement, and dilatation of the proximal divisions of the pulmonary arteries.

The arterial phase of the angiogram (Fig 2) confirms this dilatation. The distribution of the

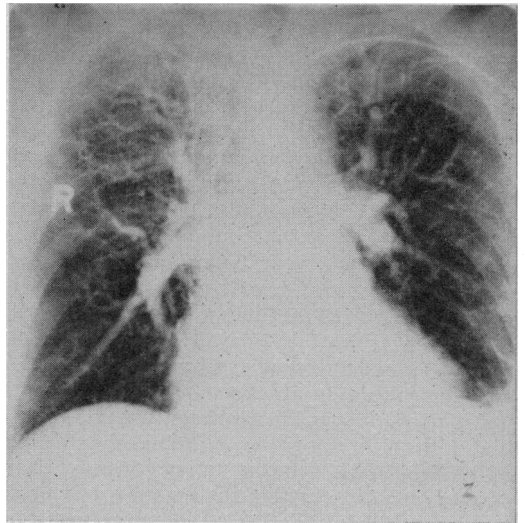


Fig 2 The arterial phase of a pulmonary angiogram in a chronic bronchitic of the Group 2 type. The lung fields are unevenly vascularized and the branches of the pulmonary arteries are tortuous and irregular

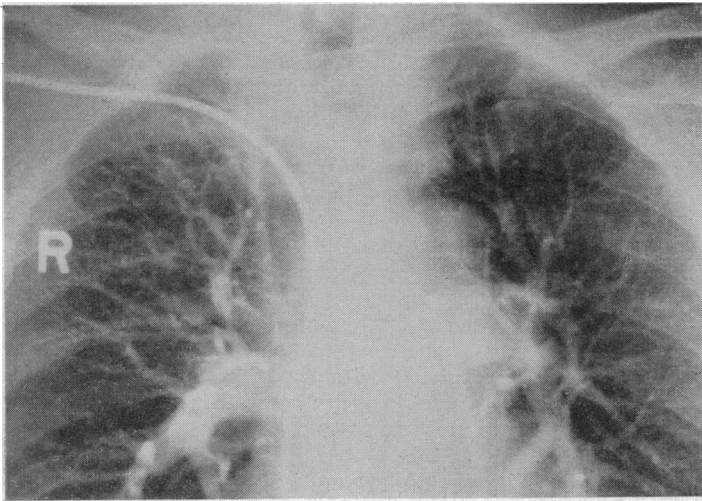


Fig 3 The arterial phase of a pulmonary angiogram in a less severe case of the Group 2 type. The vascular pattern in the right upper lobe can be seen to be in excess of, and more evenly distributed than, a comparative area of the left upper lobe. Greater background filling developed in the right upper lobe and the veins from this lobe filled in advance of those from the other lobes

mid-pulmonary vessels is uneven and these vessels in most areas are tortuous and irregular. Marked regional variations in the degree of vascularization are manifested by the distribution and rate of passage of the medium through the area. In the case illustrated, the right upper lobe arteries fill in advance of the other divisions and their main stem gives rise to a normal number of branches which arborize freely in a fastigate manner into relatively normal terminal twigs. These penetrate to the periphery of the lung field. Background filling develops and the veins fill freely.

The medium spreads much more slowly through the other lobes and their primary divisions give rise to apparently fewer branches. These divide early into sparse, tortuous and irregular terminal arborizations which do not penetrate to the periphery of the lung in great numbers. Background filling does not develop to any great extent and the veins are poorly opacified.

This case illustrates not only the unevenness of distribution of the arterial pattern but also the unevenness of function, the greatest perfusion being through the right upper lobe. This is a fairly constant finding in the group, possibly due to the unique anatomical arrangement of the primary division of the lobe.

The character of the vessels may be seen in Fig 3. This patient is not so advanced clinically or radiologically as the previous example. An uneven vascularization is again manifest in the angiogram, that of the right upper lobe differing from the rest of the lung fields. If the two upper zones are compared it can be seen that the medium and small arteries on the left are fewer, and more irregular and tortuous, than those on the right.

These appearances, with reference to those occurring in arterial trees in other parts of the body, are radiologically compatible with and

acceptable as evidence of arterial disease, even accepting the fact that the structure of the pulmonary arteries differs from their systemic counterparts, due to the differing tensions in the two circulations.

Group 3: 'Blue Bloater', Cor Pulmonale, &c.

Clinically these patients usually have a long history of chronic bronchitis with or without asthma. Repeated infective exacerbations occur and after a number of years such episodes are accompanied by cyanosis, CO₂ retention, right heart failure and peripheral oedema. It is the marked degree of CO₂ retention and peripheral oedema which characterizes these cases during their periods of clinical deterioration. The patient responds well to intensive therapy with controlled ventilation and on recovery is reasonably ambulant. Pulmonary function tests demonstrate disordered ventilation/perfusion ratios.

The vector cardiogram almost invariably demonstrates advanced right ventricular hypertrophy and blood gas analysis shows a persistently low oxygen tension and raised carbon dioxide content.

A labile pulmonary hypertension is again present, usually with a slightly raised resting pulmonary artery pressure.

Radiological features: Plain chest X-rays usually show a large volume thorax with low diaphragm and a narrow mediastinum. There is usually quite obvious right heart enlargement with dilatation of the primary divisions of the main pulmonary arteries. Some disorganization of the pulmonary vascular pattern is present but this tends to be local and, considering the clinical severity of the case, does not occur to the same degree as in Group 2. Tomograms frequently show large vessels in the lung fields.

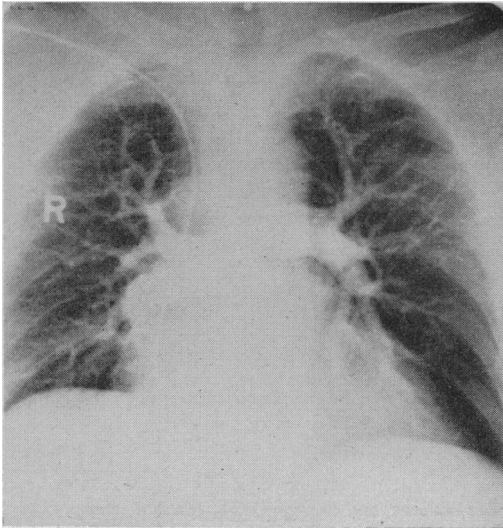


Fig 4 The arterial phase of a pulmonary angiogram in a chronic bronchitic in Group 3. The pulmonary arteries are dilated, the mid-pulmonary vessels are larger than in the other groups and the lung fields are more evenly vascularized. Occluded mid-pulmonary arteries can be recognized in the left mid-zone

The angiogram (Fig 4) confirms the dilatation of the pulmonary arteries which diminish in size gradually to give rise to normal or large mid-pulmonary trunks. There is a relatively even vascularization of the lung fields with regard to distribution, size and degree of filling of the arteries. There is a sparsity of branches arising from the mid-pulmonary trunks, but, nevertheless, venous filling appears simultaneously from all areas at a normal time and even rather early in the programme.

Occluded vessels can be recognized in the lung fields. These are usually vessels of the third and fourth generation but on occasion larger arteries are obliterated. Some, but not all, of these cases show clinical evidence of infarction during their periods of cardiorespiratory deterioration.

Summary

Although overlapping to some extent, in each group there is a preponderance of certain characteristics which may be summarized as follows:

Group 1: Those cases in which the primary disease is emphysema: (a) Relatively symmetrical distribution of the changes. (b) Rapid attenuation of the vessels into small rectilinear intrapulmonary arteries. (c) A divaricate type of branching ending in small straight terminal twigs, a poor capillary bed and a late venous filling. (d) The rate and degree of filling of the upper lobes is greater than, and in advance of, the filling in the lower lobes.

Group 2: Those cases in which significant pulmonary atherosclerosis is present in addition to chronic nonspecific lung disease: (a) A more gradual attenuation of the vessels into tortuous and irregular mid-pulmonary arteries. (b) Marked variation in the terminal arborization but most areas showing narrow irregular tortuous terminal twigs. (c) An uneven distribution of the vessels with variation in the rate and degree of regional blood flows.

Group 3: Those cases of chronic nonspecific chest disease in which ventilation perfusion disorders, cyanosis and oedema are features of the disease: (a) No abnormal diminution in the size of the intrapulmonary arteries and possibly enlargement of these vessels. (b) A more normal type of terminal arborization with a fastigate type of branching. (c) A more even pulmonary blood flow, the veins appearing evenly and at a more normal time. They may even appear early in the angiographic programme. (d) Evidence of occlusion of some branches of the pulmonary arteries.

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The Angiogram in Chronic Nonspecific Respiratory Insufficiency

In a normal pulmonary arteriogram, the size of the main vessels and the reduction in size after each branching can be seen and measured along a main or axial pathway, and a fairly even distribution of side branches is found. Visible vessels extend out evenly almost to the level of the acinus and a pattern similar to the centimetre pattern can be seen in the peripheral part (see Dr Jefferson's Figs 8 & 10, page 680). In the middle aged, the pattern is similar to that seen in children. In the one elderly patient, a man of 75, an angiogram carried out to exclude an aortic aneurysm showed relatively poor filling of the side branches; otherwise it appeared similar to a normal angiogram in a younger person.

The angiograms of the three middle-aged patients with cough and sputum and dyspnoea, and with the clinical features and respiratory function tests suggestive of chronic bronchitis