Edinburgh Medical Journal

February 1948

HEART FAILURE OF PULMONARY ORIGIN *

By JOHN McMICHAEL, M.D., F.R.C.P. Edin., F.R.C.P. Lond.

Professor of Medicine, University of London ; Director, Department of Medicine, Post-graduate Medical School of London

INTRODUCTION

IT is always a difficult task to lecture for an hour on a subject which has been the centre of one's interest, thought and research for a period of many years. While one would wish to deal with the subject completely and in all its aspects, one can only bring out the high lights as they appear to an active worker in the field. So long as research is proceeding energetically, the views expressed to-day may be considerably modified to-morrow. In trying to organise ideas on a rapidly changing subject, each investigator is inclined to emphasise certain facts at the expense or detriment of others. New light is likely to come rapidly in this subject from the steadily accelerating use of new and more accurate methods for the combined study of the heart pressures and output on the one hand, and pulmonary function on the other, most brilliantly exemplified to-day by the work of Cournand and Richards and their co-workers in New York. I would therefore ask you to regard what I have to say as an interim report on progress rather than a new chapter of a static textbook.

In this lecture I shall deal only with those conditions of heart failure which result from diseases primarily affecting the lungs or pulmonary arteries. Before going on to this subject, however, I should mention that the commonest cause of right heart failure is left heart failure. Attacks of left ventricular failure are nearly always accompanied by venous congestion in the systemic circulation, and in many cases of mitral stenosis the right ventricle is very grossly affected. Bloomfield and his colleagues (1946) have recently shown that, in severe mitral stenosis with heart failure, the systolic pressure in the pulmonary artery may rise to over 100 mm. mercury, but with recovery from the attack of failure it subsides to about 60 mm. mercury. This pressure is considerably above the normal upper normal limit of 30 mm. Hg. for the pulmonary systolic pressure. Why some cases of mitral stenosis develop an accompanying pulmonary hypertension

* A Honyman Gillespie Lecture given in the Royal Infirmary, 23rd October 1947.

and others do not, is a matter requiring further study. In uncomplicated essential hypertension without heart failure the pressure in the pulmonary artery is normal, but when heart-failure develops it rises considerably above normal. Many of the varieties of congenital heart disease also cause right heart strain but these also are beyond the scope of this talk.

Etiology and Incidence of Pulmonary Heart Disease

Right heart strain consequent upon primary disease in the lungs, particularly emphysema, has long been recognised by the pathologist who sees at post-mortem the hypertrophied and dilated right ventricle and the frequently atheromatous pulmonary artery, the latter change being ascribed to an accompanying pulmonary hypertension. In spite of the frequency of chronic bronchitis and emphysema cardiologists have often given it as their opinion that the disease is rare. Thus White and Jones (1928) saw only 21 cases of emphysema heart in 2314 cases of organic heart disease. Parkinson and Hoyle (1937) only found evidence of right ventricular hypertrophy on radiological examination in 18 out of 80 cases of emphysema. Scott and Garvin (1941), on the other hand, found the right ventricle more than 5 mm. thick at post-mortem in 80 per cent. of 48 cases of emphysema of varied etiology: most pathologists would endorse that finding. There are several reasons for this discrepancy of opinion: (1) Chronic bronchitis is a condition which is not seen nearly so much in teaching hospitals as in municipal hospitals, as these patients are frequently labelled "chronics"; (2) The primary disease is often present for many years without any obvious cardiovascular manifestations, and it is only in the late stages that these develop in a form likely to compel the interested attention of the cardiologist; (3) The development of right heart strain in these patients is not dramatic and in the early stages it is still a difficult matter to decide whether breathlessness is due to the pulmonary or cardiac component of the disease process. The absence of murmurs and other striking physical signs are further factors tending to divert these cases away from the teaching hospital.

There is a widespread impression that chronic bronchitis is commoner in males than females, and this is confirmed by my own experience. In the last eight years I have had 134 cases of chronic bronchitis under my personal care, and of these only 24 have been women, a sex incidence of about 9 males to 2 females. When one adds the occupational hazards of silicosis in the male population, and assuming the nearly equal sex incidence of the other and rarer causes of chronic cor pulmonale, the great preponderance in the male becomes obvious.

In addition to the chronic form of cor pulmonale resulting mainly from chronic affections of the lungs, there are subacute varieties resulting from secondary carcinoma in the lungs, and a now well-known acute

form resulting from pulmonary embolism. These varieties are indicated in the accompanying table.

TABLE

Classification of Causes of Pulmonary Heart Disease
(modified from Spain and Handler)

1. ACUTE.—Massive Pulmonary Embolism.
2. SUBACUTE.—(*a*) Lymphatic miliary carcinomatosis.
   (*b*) Embolic carcinomatosis.
3. CHRONIC.—A. *Lung disease.*
   (*a*) Emphysema Heart Disease
   due to (i) Bronchitis.
   (ii) Bronchiectasis.
   (iii) Cystic Disease.
   (*b*) Fibroid Lung
   due to (i) Tuberculosis.
   (ii) Scleroderma.
   (*c*) Pneumoconiosis
   due to (i) Silicosis.
   (ii) Anthraco-silicosis.
   B. *Vascular Disorders of the Lungs.*
   (*a*) Primary Pulmonary Hypertension.
   (*b*) Recurrent Pulmonary Embolism.
   C. *Kyphoscoliosis.*

I. EMPHYSEMA HEART

This is the commonest cause of pure right heart failure, and as it has been the subject of numerous investigations it will be described in some detail.

Pathogenesis and Pathological Physiology.—The simplest view of the nature of chronic emphysema heart disease is that it results from obstruction to the blood pressure through the lungs from the destruction to pulmonary capillaries. That the problem is not quite so simple may be argued from some quantitative considerations. Blumgart and Weiss (1928) have shown that the capacity of the lung vessels in health may be about three quarters of a litre of blood. The functioning part of the lung is essentially a capillary organ and the proportion of the lung blood in capillaries is very large, probably of the order of about 50 per cent. (de Burgh Daly). If we assume the average length of a pulmonary capillary to be one-tenth of a millimetre, then the cross-sectional area of the capillaries holding 375 c.c. of blood would be 37,500 sq.cm., or about 4 square yards. Now, the cross-sectional area of the pulmonary artery is about 8 sq.cm., a minute fraction of the cross-sectional area of the capillary bed. It follows that if the elimination of the pulmonary capillaries alone is to be held responsible for the creation of the high pressure in the pulmonary arterial tree, a quite enormous proportion of the capillaries would have to be destroyed. It seems highly probable that the obstruction of the lung circulation which develops in emphysema lies in the arterioles rather than in the capillaries.

These theoretical considerations have recently been put to the test by the development of techniques for the measurement of pulmonary arterial pressure in man. Cournand and his collaborators in New

York have been able to attach a sensitive manometer to the external end of a catheter, the tip of which lies in the right ventricle. The systolic pressure plateau recorded in this chamber is of course the same as the systolic pressure in the pulmonary artery. By this means they have found that the normal pulmonary arterial systolic pressure ranges from 18 to 30 mm. mercury. In patients with emphysema, before the development of venous congestion and heart failure, the pressure in the pulmonary artery is, in fact, often raised, thus confirming the long held opinions of clinicians and pathologists.

When we consider the degree of the pulmonary hypertension in relation to the severity of the causal pulmonary disease, however, it is clear that the exact mechanism by which the pulmonary pressure is raised still requires further study. Perhaps the best measure of the severity of anatomical damage to the lungs is the change in the percentage of residual air. In the normal subject the residual air constitutes 16-40 per cent. of the total lung volume (vital capacity plus residual air). In emphysema the residual air increases as the vital capacity declines, and, in severe cases, the residual air percentage may reach 70 per cent. or more of the total lung volume. If we take the figures obtained by Bloomfield and his co-workers (1946) and plot the residual air percentages against pulmonary systolic pressures, it is seen that there is no significant correlation (Fig.). There are some patients with emphysema who have a severe degree of damage judged by the residual air percentage, but in whom the pulmonary arterial pressure is not much raised or even within normal limits. On the other hand, high pulmonary pressures are found in cases with quite mild emphysema. Hickam and Cargill (1947) have demonstrated a feature of the pulmonary arterial pressure behaviour in emphysema which may be of much importance. When the normal human subject exercises mildly so as to double the resting output of the heart, the pulmonary arterial pressure does not rise. Similar exercise in emphysema with an increase in output of the heart will cause an additional large elevation of the already raised pulmonary pressure. It seems that the pulmonary vascular bed has become rigid and unable to adapt itself to carry at a normal pressure the increased pulmonary blood flow of exercise.

Obliteration of half the lung vascular bed by ligation of one pulmonary artery has long been known to produce no significant rise in pulmonary arterial pressure : this has been confirmed by Cournand and his colleagues in a study of patients submitted to pneumonectomy. It is thus clear that the cause of pulmonary hypertension in emphysema is unlikely to be the strictly anatomical capillary obliteration so often described. A further possible mechanism will be discussed later.

Besides the raised pulmonary arterial pressure there is another curious feature about emphysema heart disease which has been brought to light by heart-catheterisation studies ; when the heart fails, the output is often high normal, or even above normal. The only exception

to this seems to occur in the terminal phases when the blood pressure falls, and in these circumstances the cardiac output is low in proportion to the pressure. (Howarth, McMichael and Sharpey-Schafer, 1945; Richards, 1947.) The condition, therefore, falls into the group of *high output heart failure* previously defined (McMichael, 1946) which includes the heart failure of anæmia, Beri-beri (Dexter, 1947), and similar conditions determined by the presence of a "parasitic" circulation such as that seen in arteriovenous aneurysms and generalised

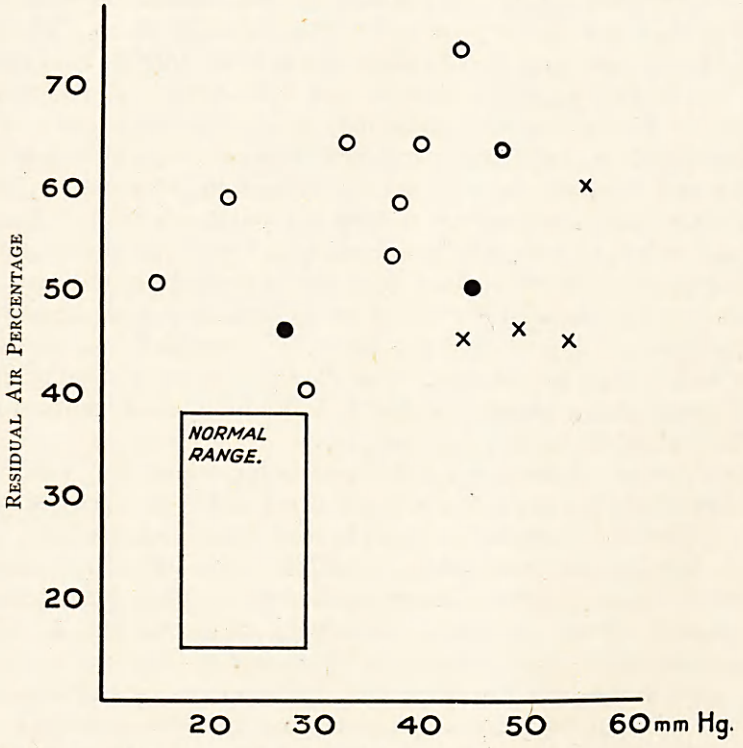

FIG.—The relationship of pulmonary systolic pressure and residual air percentage in emphysema, silicosis and pulmonary fibrosis (data from Bloomfield *et al.* 1946). In general the pulmonary systolic pressure is raised above the normal in this series. In emphysema there is a rough correlation, but cases are shown with residual air up to 60 per cent. where the pulmonary arterial pressure is within the normal range.

Paget's disease of bone (Edholm, Howarth and McMichael, 1946). The underlying mechanism of the raised output of the heart in emphysema and in anæmia may be similar, as in both conditions there is a deficiency of available oxygen in the arterial blood. The rise of venous pressure in these patients is best regarded as compensatory to maintain the cardiac output, and reduction of venous pressure by venesection may be harmful. The oxygen saturation of the arterial blood in patients with chronic emphysema and heart failure is nearly always below 80 per cent., and ranges widely below this value. We have seen patients with arterial oxygen saturation as low as 33 per cent.

Clinical Manifestations.—Most of the patients with emphysema heart disease are over forty. The breathless bronchitic whose dyspnœa persists between attacks has advanced to a stage at which emphysema is sufficiently pronounced to interfere with the normal oxygenation of his blood. Attacks of fainting and giddiness sometimes occur at this stage. Sooner or later evidence of right heart hypertrophy, and then failure with congestion and œdema, will complete the picture. The patient is dusky blue, his veins are congested, but in contrast to the patient with low output heart failure, whose extremities tend to be cold and blue, the hands, nose and ears are usually warm. The pulse is rapid (90-120) and is of good volume, as might be expected from the slightly raised cardiac output. A curious and interesting feature is the occasional presence of papillœdema and retinal hæmorrhages. Complicating exacerbations of bronchitis are often present and seem to be precipitating factors in determining failure. When there is accompanying asthma the patient is orthopnœic, but in some striking cases, without associated bronchial obstruction, he may be most comfortable in a completely recumbent position, even though venous engorgement is gross. Terminally hydrothorax and œdema may develop at the lung bases, a mechanical explanation of which is not easy to conceive. The cardiac output is usually falling at this stage and a general cardiac failure of a mal-oxygenated heart may be part of the terminal picture.

Apart from evidence of right hypertrophy, which may be difficult to detect clinically as the heart is often concealed by enlarged lungs, there is little to be made out on physical examination of the heart. Triple rhythm may sometimes be audible to the left of the sternum. A diastolic murmur of pulmonary insufficiency is rare. Radiologically, enlargement of the pulmonary artery is common, but it is only occasionally that right ventricular hypertrophy can be recognised with certainty in the X-ray picture. The electrocardiogram shows evidence of right ventricular preponderance, and this is often present before the other signs of right heart failure develop. Auricular fibrillation, which is an almost constant accompaniment of failure in mitral stenosis, is exceptional in emphysema heart, only occurring in about one case in ten.

Other Special Investigations.—The arterial blood may be grossly unsaturated with oxygen. Instead of the normal 95 per cent. saturation, values of 60-70 per cent. are common.

Examination of the oxygen unsaturation of a sample of arterial blood, which is a much simpler procedure than many clinicians imagine, may be a most useful diagnostic test. In most cases of left heart failure, even with œdema of the lungs, the oxygen saturation of the arterial blood seldom falls below 85 per cent. Unless the arterial oxygen saturation is below 80 per cent., it can be taken that emphysema is not sufficiently severe to play any part in the production of heart failure in that particular subject. Circulation time is often normal,

which is in contrast with the gross prolongations in low output heart failure. Polycythæmia is unusual, but it may sometimes be seen. It is worth while noting that arterial oxygen may fall much lower in severe congenital heart disease, while in emphysema, when the oxygen saturation is at its worst, there is often an exacerbation of acute bronchitis and infection may possibly interfere with bone-marrow activity.

The course is usually rapidly downhill. It is the exception for a patient to withstand more than two attacks of heart failure from emphysema. In this connection it is interesting to speculate on the possible influence of the coronary blood flow: systemic hypertension automatically increases the flow through the coronary arteries, but there is no such automatic regulation of the coronary supply in pulmonary hypertension. Müller (1937) has, in fact, shown that the right ventricle is much less capable of adapting to the increased work demanded by elevated resistance in the pulmonary arteries than the left ventricle is to a rise in systemic arterial pressure.

Treatment.—While digitalis and venesection increase the output of the heart in many cases of low output failure, these measures may have the opposite action, *i.e.* decrease in cardiac output, in many cases of chronic cor pulmonale (Howarth, McMichael and Sharpey-Schafer, 1947). This anomalous action of digitalis may be accounted for by the primary venous pressure reducing action of the drug which interferes with what may be a compensating mechanism maintaining cardiac output. In our experience venesection and digitalis have only raised the output of the heart in patients in whom the arterial pressure has fallen below 100 mm. mercury, and at this stage patients are often unlikely to make any satisfactory recovery. Digitalis and venesection, therefore, are unlikely to help and may even be dangerous. In most of these patients the attack of heart failure is precipitated by an exacerbation of bronchitis, and attention to this causal condition is more important than the use of standard cardiac remedies. An oxygen tent, in which the patient is kept continuously until his bronchitis has subsided, is the most valuable remedy. Bronchitis itself may be treated directly by ærosol penicillin inhalations.

Another Type of Circulatory Failure in Lung Disease

Acute Circulatory Failure from Sudden Reduction of Negative Intra-thoracic Pressure.—Patients with emphysema who develop superadded pneumothorax of the valvular variety, or who have a large bulla which may blow up in a similar manner, are likely to become acutely distressed with severe dyspnœa, gross air-hunger, and a pulse of poor volume and low pressure. In one such emphysematous patient who had a pneumothorax the intrathoracic pressure fluctuated round about zero, while the hydrostatic pressure in his right auricle remained extremely low. Filling of the heart, which normally takes place at an effective filling pressure of 7 cms. water (2 cms. hydrostatic

pressure plus 5 cms. negative intra-thoracic pressure), was grossly interfered with, and his cardiac output was 2·4 lit/min. (normal 5·3). In spite of the low output in this particular patient there was electrocardiographic and clinical evidence of right heart hypertrophy, showing that the raised pulmonary resistance is independent of the high cardiac output factor. This is a variety of acute circulatory failure which of course is easy to differentiate from chronic cor pulmonale with venous congestion.

II. Other Clinical Varieties of Pulmonary Heart Disease

1. Acute Cor Pulmonale.—This condition, first clearly defined by McGinn and White (1935) results from blockage of the pulmonary arteries by emboli, the latter usually coming from the leg veins, following operations in the surgical wards, or in chronically ill and bedridden patients in the medical wards, especially those past middle life. It may complicate many ordinary forms of heart disease. The acute attack is often preceded by symptoms of smaller emboli in the periphery of the lungs causing pleural pain or hæmoptysis, but the actual development of right heart strain results from blockage of the large arteries at the root of the lungs without any of the manifestations of peripheral pulmonary embolism.

The patient becomes suddenly aware of a feeling of suffocation and apprehension, perhaps with a sensation of substernal oppression, but there is no actual pain. He develops air hunger, is pale and may sweat, while the pulse becomes small in volume and the blood pressure falls. The venous pressure is raised, as seen by Lewis's method of inspection of the neck veins, but in the collapsed stage it may require careful inspection to make this out. On examination of the heart there may be evidence of exaggerated pulsation of the right ventricle and at times a gallop rhythm may be heard in this region. The electrocardiogram is often diagnostic. The standard leads may simulate a posterior myocardial infarction of Q_3 T_3 pattern, but precordial leads (C R or V leads) show inversion of T waves over the right ventricular region (Wood, 1941).

2. Recurrent Pulmonary Embolism.—Ljungdahl (1928) raised the question as to whether there was such a condition as chronic embolisation of the pulmonary arteries. He described two women, aged fifty-one and thirty-eight, who had suffered for years from a dry cough, slowly developing breathlessness and finally signs of heart failure. The right ventricle was enlarged and there was radiological enlargement of the pulmonary vascular shadow, but no other signs of valvular or other disease of the heart. At autopsy there was severe blocking of the pulmonary arteries with thrombi, with dilatation and hypertrophy of the right heart. The lungs appeared normal.

Three cases of this type have come under my notice in the past few years. Two, however, were somewhat complicated by other

factors (coronary disease in one, and empyema in the other), and I shall therefore only describe briefly the third case.

A woman, aged fifty-two, had been well until two years before admission to hospital. Following an attack of bronchitis she noted increasing shortness of breath on exertion, accompanied often by deep-seated pain under the right ribs (? hepatic congestion). This pain came on on effort and disappeared with rest. The dyspnœa steadily progressed and in the last two months she had difficulty in getting to her work as a typist. Four days before admission she came home from work feeling nauseated, with more severe pain, and dyspnœa worse than ever. She never had any hæmoptysis.

On examination she was restless and anxious with marked air-hunger. The lips were cyanotic, and the jugular veins were engorged 6 cms. above the sternal angle in the propped-up position. The apex beat was not easily palpable. A gallop rhythm was audible just internal to the mid-clavicular line and a systolic murmur was present over the tricuspid area. Blood pressure was 90/70. The legs showed extensive varicosities with thrombosis of the superficial veins and extensive scarring from old varicose ulcers. In addition a thrombosed cord could be felt in the middle of the left popliteal fossa. Sedimentation rate was 2 mm. in one hour and white cell count was 27,600.

A diagnosis of pulmonary embolism was made which was confirmed by characteristic electrocardiograms showing gross right preponderance and flat or inverted T waves in chest leads taken from the region of the right ventricle. Chest X-ray showed clear lung fields but the vascular shadows at the root of the lungs were considerably distended. The patient died suddenly two days after admission. Autopsy confirmed the diagnosis of pulmonary embolism. The heart weighed 480 gm. and there was gross hypertrophy of the right ventricle with no other demonstrable cause of heart failure, valves and coronary vessels being normal. Some of the emboli were very old and had become organised and recanalised. The source of emboli was traced in the popliteal and femoral veins.

In this case there had been some cause of obstruction of the pulmonary circulation acting for a considerable time to produce the degree of hypertrophy which was seen at post-mortem. No cause other than repeated pulmonary embolism could be found.

3. Subacute Cor Pulmonale from Secondary Carcinoma in the Lungs.—The term " subacute cor pulmonale " was first suggested by Brill and Robertson (1937) for this condition which develops in a few weeks or months. Greenspan (1934) reviewed the ways in which secondary carcinoma of the lungs might produce stresses on the right heart.

(1) Certain types of carcinoma may spread to the lungs as blood-borne emboli. Schmidt (1903) showed that tumour cells disintegrate in the lung vessels and surrounding thrombus becomes organised, with blockage of the lumen of the vessels.

(2) Greenspan records lymphangitis carcinomatosa of the lungs with accompanying obliterative endarteritis resulting from invasion of the lung vessels by carcinoma.

(3) Costedoat (1933) emphasised a suffocating form as distinct from an embolic form with a rapid pulse and dyspnœa and found no change in the lung vessels. This results from carcinomatous lymphangitis of the lungs.

A woman, aged sixty-seven, had carcinoma of the body of the pancreas. Miliary mottling was present in the lungs from January 1945, and the patient subsequently survived six months, during which time the mottling increased in size and density. Two weeks before her death the electrocardiogram showed definite signs of cor pulmonale, the arterial blood was 78 per cent. saturated with oxygen, and the cardiac output was 7 lit/min., a high figure for a small woman. Right auricular pressure was raised 2-3 cms. saline above the normal. This is an example of the suffocative form.

An example of the embolic form was seen recently in a woman who had had a miscarriage some four months before her death. She presented with heart failure, at first thought to be due to hyperthyroidism, but as time went on she developed clear signs of right heart stress. At post-mortem the lung vessels were extensively blocked by tumour emboli from a chorionepithelioma with secondary thrombosis.

4. PRIMARY PULMONARY HYPERTENSION.—There are now in the literature many instances of failure of the right ventricle with associated pulmonary vascular sclerosis usually described as cases of primary pulmonary hypertension. For a long time discussions on this subject were bedevilled by memories of "Ayerza's syndrome" in which a similar picture was said to be associated with polycythæmia and in the etiology of which syphilis was said to play a part. While this syndrome may occur it must be rare, and in South America competent observers are now inclined to stress its rarity. McCallum (1931) emphasises that the sclerosis affects the small arteries of the lungs (which were usually spared in mitral stenosis). Cases have been described in this country by de Nevasquez *et al.* (1940), East (1940) and Armstrong (1940).

A female, aged twenty-seven, developed dyspnœa over about a year. Investigation showed gross right ventricular enlargement with enlargement of the pulmonary artery. Pressure in the right auricle was 18 cms. saline above the sternal angle and the *mean* right ventricular pressure was 60 cms. above the same reference point. It is quite possible that her pulmonary systolic pressure was of the order of 80 mm. mercury. The illness was progressive and at post-mortem some twenty months after the onset of the illness, the small pulmonary arterioles were thickened and the right ventricle was grossly hypertrophied, but the picture was complicated by the presence of numerous pulmonary emboli. There was no bronchitis or emphysema.

Pulmonary embolism complicates very many instances of cardiac

failure, and where the primary cause lies in the pulmonary circulation the differential diagnosis between recurrent pulmonary embolism and primary pulmonary hypertension complicated by emboli may be a matter of considerable difficulty.

5. Cor Pulmonale Resulting from Fibroid Changes in the Lungs.—(a) *Non-tuberculous Fibrosis.*—The fibroid changes which accompany such conditions as bronchiectasis are frequently accompanied by the development of right heart failure in a manner similar to that seen in chronic bronchitis.

(b) *Tuberculosis.*—Nemet and Rosenblatt (1937) have given an excellent review of the condition of the right heart in tuberculosis. In 71 patients who came to post-mortem in the tuberculosis service of the Montefiore Hospital they found right ventricular hypertrophy in 24 instances. Generally speaking, however, the heart was not increased in weight, and, in fact, was often reduced. The thickness of the right ventricle in moderate enlargement was 5-7 mm. and reached as high as 9 mm. Clinically the patients were cyanotic, the veins were often congested, and the usual symptoms of cardiac failure were present. Some of these patients had miliary tuberculosis which would possibly be a factor in raising pulmonary arterial pressure. They emphasise the difficulty of making a diagnosis in the absence of radiological cardiac enlargement.

(c) *Silicosis.*—Cor pulmonale is generally recognised as a terminal event in silicosis, but there are no recent detailed studies of its mode of development. Bloomfield and his group include a few pressure measurements in silicosis in their report (Fig. 1). My own experience is limited to a case which showed endarteritis of the pulmonary arteries at post-mortem.

(d) *Pulmonary Fibrosis in Scleroderma.*—The association of that form of scleroderma which shows itself predominantly in the fingers (sclerodactyly) with Raynaud's disease and visceral affections of the œsophagus, lungs and heart, is now well recognised (Weiss, Stead, Warren and Bailey, 1943). These authors show that the heart itself may be directly affected. The lungs, however, often show a diffuse fibrous change affecting the walls of the alveoli. Some authors (for example, Kraus 1924) have reported right heart hypertrophy as a consequence of the pulmonary change. I have seen one instance of this condition. An elderly female patient had suffered for some ten years from Raynaud's disease and also dysphagia. The hands were characteristic of sclerodactyly and there were typical œsophageal lesions. The lungs showed extensive diffuse fibrosis with a heavy accentuation at the right apex. For the last five years of her life she had a right ventricular preponderance in the electrocardiogram, and had frequent admission to hospital with breathlessness. She died outside hospital and there was no post-mortem.

6. Cor Pulmonale in Kyphoscoliosis.—This is a form of chronic cor pulmonale not widely recognised in this country. Excellent

reviews have appeared in America, especially those of Boas (1923) and Chapman, Dill and Graybiel (1939). In France and Germany the condition has been well recognised for a long time. Corvisart (1806) was familiar with the fact that the right heart was often dilated in kyphoscoliosis. Traube (1878) found at autopsy in a hunchback dilatation of the right ventricle, the patient having had typical symptoms of right heart failure before death. Boas describes the mechanism of these changes as follows :—

" The deformed thorax is smaller than normal in size and the diaphragm is usually high. Moreover, because of the fact that thoracic distortion is well established before bodily growth is completed, the chest lags behind the rest of the body in development. As a result of these factors the lung volume is steadily encroached upon with ensuing atelectasis of large pulmonary fields."

By this means he thought the capillary bed of the lungs was reduced. Chapman and his colleagues found that the lung volume in these patients was in fact greatly reduced, but the mechanism by which the pressure in the pulmonary vascular bed was raised was uncertain. Our experience confirms the presence of changes in the right heart following on kyphoscoliosis. An example is seen in the following brief case record :

A patient, aged forty-four, with a gross dorsal kyphosis resulting from an old tuberculous spine was admitted to hospital in heart failure. He denied any significant bronchitis, and had never had a purulent sputum. Radiologically no fibroid changes were present in the lungs. He had gross venous congestion and right ventricular dominance was marked in the electrocardiogram. Cardiac output was 3·7 lit/min., but the blood pressure was 65/46 at the time, and he therefore showed reactions similar to those at the terminal stages of ordinary varieties of emphysema heart.

The patient died forty-eight hours after the observations were made. Autopsy showed gross right ventricular hypertrophy, but no significant bronchitis or pulmonary fibrosis.

Discussion and Conclusion

From the above survey it seems apparent that pulmonary heart failure develops largely as the result of hypertension in the pulmonary circulation. In a number of cases there is adequate evidence of pulmonary vascular obstruction—*the obstructive group*, but in others—*the anoxic group*—an anatomical cause of obstruction is absent. Some light may be shed on the problem by the recent work of Motley and his colleagues at the Bellevue Hospital. The inhalation of 10 per cent. oxygen in normal man immediately doubles the pulmonary arterial pressure without any accompanying change in cardiac output. This vaso-constrictive effect of low oxygen tension on the pulmonary arterioles has also been analysed by von Euler and Liljestrand, who have shown that it is not mediated by vaso-constrictor nerves, but is

rather a local reaction of the pulmonary arterioles and pre-capillaries to oxygen lack. The latter authors suggest that this mechanism regulates the blood-flow through various parts of the lung according to the efficiency of aeration in each area. It is by this means that the circulation through atelectatic and pneumonic lobes may be closed off. Poor ventilation of the lungs may induce this vaso-constrictive reaction and this may be the key to the problem of many forms of pulmonary heart disease. The difficulty of moving a kyphoscoliotic thoracic cage with consequent limited ventilation may have the same action on the pulmonary arterial pressure as the poor ventilation of emphysematous lungs, or lungs in which the bronchioles and alveolar ducts are strangled by peri-bronchial lymphatic carcinoma.

REFERENCES

ARMSTRONG, T. G. (1940), *Brit. Heart Journ.*, **2**, 201.

BOAS, E. P. (1923), *Amer. Journ. Med. Sci.*, **166**, 89.

BLOOMFIELD, R. A., LAUSON, H. D., COURNAND, A., BREED, E. S., and RICHARDS, D. W. (1946), *Journ. Clin. Invest.*, **25**, 639.

BLUMGART, H. L., and WEISS, S. (1928), *Journ. Clin. Invest.*, **6**, 103.

BRILL, J. C., and ROBERTSON, T. D. (1937), *Arch. Int. Med.*, **60**, 1043.

CHAPMAN, E. M., DILL, D. B., and GRAYBIEL, A. (1939), *Medicine*, **18**, 167.

CORVISART, J. N. (1806), " Essai sur les maladies et les lesions organiques du Coeur," *Observation XXIII.*, p. 114.

COSTEDOAT, A. (1933), *Presse Med.*, **41**, 745.

DALY, I. DE B. (personal communication).

DEXTER, L. (personal communication).

EAST, T. (1940), *Brit. Heart Journ.*, **2**, 189.

V. EULER, U. S., and LILJESTRAND, G. (1946), *Acta Physiol. Scand.*, **12**, 301.

EDHOLM, O. G., HOWARTH, S., and MCMICHAEL, J. (1945), *Clin. Sci.*, **5**, 249.

GREENSPAN, E. B. (1934), *Arch. Int. Med.*, **54**, 625.

HICKAM, J. B., and CARGILL, W. H. (1947), *Amer. Journ. Med.*, **3**, 504.

HOWARTH, S., MCMICHAEL, J., and SHARPEY-SCHAFER, E. P. (1947), *Clin. Sci.*, **6**, 187.

KRAUS, E. J. (1924), *Virchows Arch.*, **253**, 710.

LJUNGDAHL, M. (1928), *Dtsch. Arch. Klin. Med.*, **160**, 1.

MCCALLUM, W. G. (1931), *Bull. Johns Hopk. Hosp.*, **49**, 37.

MCGINN, S., and WHITE, P. D. (1935), *Journ. Amer. Med. Assoc.*, **104**, 1473.

MCMICHAEL, J. (1946), *Schweiz. Med. Wschr.*, **76**, 851.

MOTLEY, H. L., COURNAND, A., WERKO, L., HIMMELSTEIN, A., and DRESDALE, D. (1947), *Amer. Journ. Physiol.*, **150**, 315.

MÜLLER, E. A. (1937), *Püugers Arch.*, **238**, 638.

NEMET, G., and ROSENBLATT, M. B. (1937), *Amer. Rev. Tuberc.*, **35**, 713.

DE NEVASQUEZ, S., FORBES, J. R., and HOLLING, H. E. (1940), *Brit. Heart Journ.*, **2**, 177.

PARKINSON, J., and HOYLE, C. (1937), *Quart. Journ. Med.*, N.S., **6**, 59.

RICHARDS, D. W. (1947), *Amer. Journ. Med.*, **3**, 434.

SCHMIDT, M. B. (1903), *Die Verbreitungswege der Karzinome*, Jena G. Fischer.

SCOTT, R. W., and GARVIN, C. F. (1941), *Amer. Heart Journ.*, **22**, 56.

SPAIN, D. M., and HANDLER, B. J. (1946), *Arch. Int. Med.* **77**, 37.

TRAUBE, L. (1878), *Gesammelte Beitr. z. Path. u. Physiol.*, **3**, 354.

WEISS, S., STEAD, E. A., WARREN, J. V., and BAILEY, O. T. (1943), *Arch. Int. Med.*, **71**, 749.

WHITE, P. D., and JONES, T. D. (1928), *Amer. Heart. Journ.*, **3**, 302.

WOOD, P. (1941), *Brit. Heart Journ.*, **3**, 21.