Cor Pulmonale:
A Semantic Consideration, with Brief
Notes on Diagnosis and Treatment

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There is a conspicuous lack of agreement as to what lesions or clinical conditions should be included in the term “cor pulmonale.” Varied and conflicting views prevail throughout the literature, current and old.1-18

Historically, the first use of the term cor pulmonale appears to have been in 1931 by Paul White;1 prior to this time “emphysema heart” was in common use. In the first edition of his textbook on heart disease White speaks of “pulmonary heart disease or cor pulmonale” and indicates preference for the latter term. Twenty years later (4th edition)2 he continues to speak of “cor pulmonale or pulmonary heart disease,” using these terms as synonyms. In discussing the etiology of cor pulmonale he stresses “chronically increased resistance in the pulmonary circulation due commonly to narrowing of the arterioles and capillary bed,” but specifically excludes from this category resistance due to “left heart failure, mitral stenosis, or congenital heart disease.”

In a recent review Fishman and Richards8 define cor pulmonale as “a heart which manifests dilatation, hypertrophy, or failure secondary to intrinsic disease of the lungs”; they specifically exclude from this definition “right ventricular hypertrophy, dilatation or failure secondary to disease of the heart, e.g. mitral stenosis.” On the other hand, Griffith4 includes right ventricular changes of mitral stenosis in the general category of cor pulmonale; he makes a distinction between “primary” cor pulmonale in which “pulmonary hypertension is the basic lesion” and “secondary” cor pulmonale “in which the basic lesion lies not in the pulmonary circuit but beyond, in the left side of the heart.”

Oram6 broadly defines cor pulmonale as “hypertrophy and eventual failure of the right ventricle resulting from disease of the lungs, or disorder of the pulmonary circulation.” However, when he lists the specific lesions causing disorders of the pulmonary circulation he omits all congenital cardiac lesions. Mack and Snider4 define cor pulmonale as “right ventricular hypertrophy resulting from disease involving the lung and pulmonary circulation” but exclude from this category “other causes of right ventricular hypertrophy, such as mitral stenosis, congenital heart disease, and left ventricular failure.” Hecht,7 however, includes pulmonary valvular stenosis as well as “pure mitral stenosis” in which “secondary vascular changes (in the pulmonary bed) develop, apparently the result of long-standing pulmonary vascular congestion, high capillary and arte-

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1. From the Department of Medicine, University of Oregon Medical School.
2. For a comprehensive discussion of this phase of therapy and a detailed description of specific measures the reader is referred to a recent article by Simon Dack.8
riolar pressure and precapillary and interalveolar edema." The latter author also feels that "For the sake of completeness elevation or right-sided pressure as the usual consequence of left ventricular failure should be mentioned."

Some authors restrict the use of the term cor pulmonale to right ventricular lesions resulting from disease of the lung only. While it is true that intrinsic lung disease is a common cause of right ventricular strain and failure, clinical and anatomical studies indicate that the development of cor pulmonale in such cases is dependent on associated involvement of the pulmonary vascular bed. Parenchymal lung disease per se, however extensive, may leave the right ventricle completely intact if there is no coincidental interference with the flow of blood through the pulmonary circuit. Some years ago when performing postmortem examinations in a tuberculosis hospital it was impressive to observe the frequency with which a normal right ventricle is seen in the presence of extensive diffuse pulmonary parenchymal disease. Later experience in clinicopathological conferences has confirmed this earlier impression (Figure 1).

Similar observations are frequently found in the literature. Spatt and Grayzel state "The mechanism of dilatation and hypertrophy of the right

FIGURE 1: Photograph of the cut surfaces of both lungs of a patient who died of pulmonary tuberculosis; there was extreme, diffuse parenchymal disease. The heart was entirely normal. (Autopsy by Dr. Nelson Niles of the Department of Pathology of the University of Oregon Medical School.)
ventricle (in diffuse pulmonary disease) seems fairly generally agreed upon. The pulmonary changes destroy and narrow blood capillaries in the lungs, causing pulmonary hypertension, which in turn increases the strain on the right heart, thus resulting in dilatation and hypertrophy.” Rubin states that in the absence of associated cardiovascular disease, a selective or preponderant enlargement of the right ventricle in emphysema is an uncommon finding. Florence McKeown in a postmortem study of 101 cases of emphysema found hypertrophied right ventricles in 39 and normal right ventricles in 62 cases. White and Brenner in a study of 100 consecutive unselected autopsies found that “the majority of cases with pulmonary disease had normal right ventricles. . . . There was relative little correlation between changes in the pulmonary vascular tree and thickness of the right ventricles except in the more marked cases. . . . Ordinarily, asthma, emphysema and pulmonary tuberculosis, even though of high degree, do not produce cor pulmonale.”

It is thus apparent that parenchymal lung disease alone does not necessarily produce strain or hypertrophy of the right ventricle. On the other hand, it is a common experience to find right ventricular hypertrophy whenever the pulmonary circulation is disturbed, even in the total absence of parenchymal lung disease. As examples of such pathological states one may cite primary pulmonary vascular sclerosis, essential pulmonary hypertension, and obstruction of the main stem of the pulmonary artery by tumor, aortic aneurysm or valvular stenosis. Hence, it would appear fair to state that in all instances where isolated right ventricular hypertrophy develops, the crucial lesion is a disturbed pulmonary circulation.

A disturbed pulmonary circulation may result from various pathological processes:

A. “Organic” Pulmonary Vascular Disease. Arteriosclerotic and/or thrombo-embolic narrowing of the pulmonary arteries and their branches (with secondary pulmonary hypertension) may be either primary as in primary pulmonary vascular sclerosis or secondary to some other pathological process. Examples of the latter are: (1) pulmonary parenchymal disease, (2) congenital cardiovascular disease, such as patent ductus arteriosus, septal defects, and other lesions with abnormal communication between the systemic and pulmonary circulation, (3) acquired cardiovascular lesions such as mitral stenosis and left ventricular failure, (4) peripheral vascular disease such as phlebothrombosis of the legs and pelvis, (5) blood dyscrasias such as polycythemia, primary or secondary, sickle cell anemia, leukemia.

B. Hypoxia and Hypercapnia. Hypoxemia and hypercapnia due to impaired pulmonary ventilation, such as may be seen in chronic bronchitis and emphysema, may produce pulmonary hypertension with or without demonstrable morphologic changes in the pulmonary vessels. The right ventricular hypertrophy which often occurs in such cases may be due to the hypertension alone or to the combined effect of the hypertension and the hypoxemia.

C. Essential Pulmonary Hypertension. Cases of pulmonary hypertension
proved by cardiac catheterization, have been observed in which no cause for the hypertension could be demonstrated either clinically or at autopsy. It may be assumed that in such cases the hypertension and the right ventricular hypertrophy resulted from arteriolar spasm of sufficient degree to interfere with the flow of blood through the lesser circulation.

D. Hypervolemia. Strain of the right ventricle may occur even in the absence of diffuse organic pulmonary vascular disease when the volume of blood propelled through the pulmonary circuit is increased. Such a condition obtains in the presence of septal defects and other congenital lesions with abnormal communications between the lesser and systemic circulations, in pulmonary arterio-venous aneurysms, and in polycythemia, primary or secondary.

E. Mechanical Obstruction. Obstruction to the right ventricular outflow tract may result from valvular lesions—especially pulmonic stenosis, coarctation of the pulmonary artery and compression of that vessel by tumor or aortic aneurysm.

F. Chest Deformities. Kyphoscoliosis and pectus excavatum are important examples. The exact mechanism by which these lesions cause strain on the right side of the heart is not completely established. Available evidence implicates pulmonary infection and emphysema secondary to compression of portions of the lung.12

Right ventricular response to disordered pulmonary circulation, regardless of etiology, is constant. If a hypertrophied right ventricle were isolated from the rest of the heart, the most discerning pathologist would be unable to tell whether the cause of the hypertrophy lay in diffuse lung disease, a congenital cardiac lesion, or pulmonary hypertension due to any of the anatomical or physiological abnormalities described above.

Within this broad range, the electrocardiogram also is non-specific, revealing an "RVH" (cor pulmonale) pattern but not its cause (Figure 2). To a more limited extent this is also true of x-ray film silhouettes, especially if only posteroanterior films are available (Figure 3).

Thus three facts stand out: (1) pulmonary parenchymal disease alone, if it fails to disturb the pulmonary circulation, does not produce right ventricular hypertrophy; (2) any condition which brings about increased resistance to the flow of blood through the pulmonary vascular bed may cause right ventricular strain, hypertrophy and eventual failure; (3) the clinical and pathological manifestation of such right ventricular hypertrophy and failure follow a fixed pattern—modified only by the symptoms obviously due to the associated disease—regardless of the condition which has disturbed the pulmonary circulation.

In the light of these considerations it would seem reasonable to regard "cor pulmonale" as a generic term comprising all types of right ventricular strain and hypertrophy due to a disordered pulmonary circulation regardless of etiology. In this sense it is analogous to "cor aortale" which refers to strain and hypertrophy of the left ventricle due to a disordered systemic (aortic) circuit, as in systemic hypertension, coarctation of the aorta and aortic valvular disease. The difficulty in arriving at a concise
FIGURE 3A
FIGURE 3A: Right ventricular hypertrophy due to: A, primary pulmonary vascular sclerosis in a 37 year old woman (autopsy), and B, mitral stenosis in a 37 year old man (surgery).

FIGURE 2: The electrocardiographic tracings on these two patients show striking similarity in the demonstration of right ventricular hypertrophy. In Case 1, the patient, a 56 year old woman, was found at autopsy to have intimal pulmonary arteriolar sclerosis; while in Case 2, a 36 year old woman, the cause for right ventricular hypertrophy was an interatrial septal defect.
definition of cor pulmonale lies in the fact that the type of heart disease covered by that term may occur in a large variety of unrelated clinical and pathological conditions. However, when it is made clear that all these "unrelated" conditions have one thing in common, a disturbed pulmonary circulation, the difficulty disappears, and cor pulmonale may then be defined simply as right ventricular hypertrophy, strain or failure due to disordered pulmonary circulation. "Pulmonary heart disease," "emphysema heart" and "pulmonary hypertensive heart disease," often used as synonyms of cor pulmonale are less generic in their connotation. Each of these terms refers to a more or less specific etiology of the right ventricular lesion; whereas "cor pulmonale" includes all these as well as other varieties of isolated right ventricular hypertrophy due either to pulmonary or extra-pulmonary factors.

**Diagnosis**

This concept of cor pulmonale focuses attention upon the essential pathologic physiology. In any given case consideration of the various pathologic processes which may disturb the lesser circuit points the way to a more definitive etiological diagnosis.

A history of chronic pulmonary disease is present in most instances. In cases secondary to mitral stenosis there is a background of rheumatic carditis dating from childhood. In primary pulmonary vascular disease or in primary pulmonary hypertension the history of illness is of relatively short duration—a few months to a year or two. The early manifestations are those of the underlying disease upon which are superimposed, gradually or abruptly, the symptoms of pulmonary hypertension. The latter include fatigue, dyspnea, cyanosis (often paroxysmal), dizziness, syncope and angina-like chest pains with atypical radiation. Once the symptoms become pronounced the course is rapidly downhill, terminating fatally within weeks or months. Sudden death is common.

Pertinent physical signs are dyspnea, apprehensiveness and cyanosis. Clubbing of the fingers may be present. The heart may or may not show enlargement to percussion. However, palpation will reveal a pronounced lower sternal thrust and increased systolic pulsations in the area of the conus arteriosus. A diastolic "shock" may be felt in the pulmonary area due to forcible closure of the pulmonic valves. The pulmonic second sound is greatly accentuated and systolic and diastolic murmurs along the left border of the sternum are common. Gallop rhythm may be present. In terminal stages congestive right-sided heart failure may appear; more often death occurs before gross signs of failure develop.

Radiographically, the most characteristic finding is a prominent pulmonary artery segment on the left border of the heart (Figure 3). The vascular markings in the peripheral portion of the lungs will vary with the underlying cause: they may be normal, greatly increased as in atrial septal defects and in mitral stenosis, or greatly diminished as in primary pulmonary vascular sclerosis and essential pulmonary hypertension. The heart may or may not appear enlarged; in the stage of hypertrophy without dilatation the right ventricle often looks normal on the posteroante-
rior film. With pronounced right ventricular hypertrophy and clockwise rotation of the heart, the cardiac silhouette is often enlarged to the left and may be wrongly interpreted as left ventricular hypertrophy. In point of fact this leftward extension of the cardiac shadow is due to enlargement of the right ventricle; at autopsy the left ventricle may be found even smaller than normal.

The electrocardiogram offers important aid in the diagnosis of cor pulmonale. The most definitive evidence of right ventricular hypertrophy is reversal of the QRS pattern in the precordial leads. The right precordial leads (V₁R, V₂R, V₁, V₂) exhibit tall R-waves with relatively small S-waves and the left precordial leads (V₅, V₆) have small R-waves and relatively deep S-waves. The onset of the intrinsicoid deflection is delayed in the right precordial leads and accelerated in the left, averaging from .08 to .05 seconds in the former and .02 to .03 seconds in the latter—likewise a reversal of the normal relationship. The T-waves are usually inverted in V₁ to V₄ and upright in V₅ and V₆; occasionally they are inverted in all the precordial leads, probably a reflection of associated left ventricular disease.

Less definitive patterns associated with cor pulmonale are incomplete RBBB and less frequently complete RBBB. The latter is more often seen in primary left ventricular disease but occasionally is encountered as a complication of cor pulmonale.

The standard leads usually exhibit marked right axis deviation, a nonspecific sign dependent upon the vertical position and clockwise rotation of the heart (Figure 2). However, since right axis deviation is the most constant single finding in the electrocardiogram of right ventricular hypertrophy, it must be regarded as an important component of the cor pulmonale pattern.

Tall, sharply-spiked P-waves (P-pulmonale) are often seen in standard leads II and III and in AVF, probably a reflection of right auricular hypertrophy or dilatation. Although not pathognomonic, this finding affords confirmatory evidence for the diagnosis of cor pulmonale.

Two points merit special emphasis: (1) the patterns associated with cor pulmonale are independent of etiology—similar or almost identical tracings may be obtained in atrial septal defect, in primary pulmonary vascular sclerosis or in mitral stenosis; (2) a completely normal tracing may be found in well-advanced cases of cor pulmonale; hence, the absence of electrocardiographic evidence of right ventricular hypertrophy does not exclude this lesion.

Treatment

Treatment is directed primarily toward the relief of the underlying cardio-pulmonary diseases and their complications. This implies early recognition and treatment of chronic broncho-pulmonary infection (obstructive emphysema, pneumonitis, bronchitis, broncietasis, asthma)*, and the early surgical correction of reversible cardiac lesions and thoracic

*For a comprehensive discussion of this phase of therapy and a detailed description of specific measures the reader is referred to a recent article by Simon Dack.*
cage deformities. For essential pulmonary hypertension or primary pulmonary vascular disease (sclerotic or thrombo-embolic) little of lasting therapeutic value is available, since the underlying pathological process is essentially irreversible. Temporary favorable results have been reported from the use of ganglionic blocking agents (hexamethonium bromide)$^{17}$ and from adrenolytic drugs (Priscoline).$^{7}$ Some relief has also been reported from various surgical procedures such as sympathectomy and vagotomy. The rationale for these procedures is the assumption that a reversible increment of "spasm" is superimposed on the organic vascular obstruction.

Management of congestive failure affecting primarily the right ventricle does not differ essentially from the treatment of any other type of congestive heart failure. Digitalis, salt restriction, diuretics and regulation of activity are usually effective, albeit to a less marked degree; also the results achieved are apt to endure for a shorter period of time.

SUMMARY

Since the term cor pulmonale was first introduced a quarter century ago, it has come to mean different things to different people. A more rational approach to the semantology of the term is suggested. On this basis, cor pulmonale is defined as "right ventricular hypertrophy due to a disordered pulmonary circulation," regardless of the cause. "Pulmonary heart disease," "emphysema heart" and "pulmonary hypertensive heart disease," are types of cor pulmonale; each refers to right ventricular hypertrophy of more or less specific origin. Brief notes on diagnosis and treatment are presented.

RESUMEN

Desde que el término cor pulmonale se introdujo hace un cuarto de siglo, ha significado diferentes cosas a también diferentes personas. Una interpretación más racional del término se sugiere. Sobre esta base el cor pulmonale se define como "hipertrofia ventricular derecha debida a trastornos de la circulación pulmonar" cualquiera que sea la causa. "Enfermedad cardíaca pulmonar," "Corazón enfisematoso" y "Enfermedad cardíaca hipertensiva pulmonar" son formas de cor pulmonale. Cada uno de estos términos se refiere a hipertrofia ventricular de origen más o menos específico. Se presentan resúmenes sobre diagnóstico y tratamiento.

RESUME

Depuis que le terme de "coeur pulmonaire" a été utilisé pour la première fois il y a un quart de siècle, il est arrivé à avoir des significations différentes selon les auteurs. I. C. Brill suggère qu’on revienne à une définition proche de la signification véritable de ce terme. Sur cette base, le coeur pulmonaire doit être défini comme "une hypertrophie ventriculaire droite, due à un trouble de la circulation pulmonaire," quelle qu’en soit la cause. Le coeur des affections pulmonaires, le coeur emphysématex, et le coeur de l’hypertension pulmonaire sont des types de coeurs pulmo-
nares; chacun d'entre eux est dû à une hypertrophie ventriculaire droite d'origine plus ou moins spécifique. L'auteur présente de brèves conclusions sur le diagnostic et le traitement.

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REFERENCES