Chronic Cor Pulmonale or Chronic Pulmonary Hypertensive Heart Disease?

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Paul D. White used the term "chronic cor pulmonale" or "chronic pulmonary heart disease" for the first time 30 years ago, but there still is not unanimous opinion about the meaning of these terms. It is not the purpose of this article to make an historic review of this subject or to tabulate what different writers have said in this field. The interested reader is referred to the excellent review written by Brill in 1957. Paul D. White considered an increased pulmonary vascular resistance produced by arteriolar narrowing and a reduction of the capillary bed as the main causes of cor pulmonale. He specifically excluded left ventricular failure, mitral stenosis and congenital heart diseases from other disorders producing increased pulmonary vascular resistance. Other authors, however, included these three etiologies without restriction, while still others emphatically rejected them. It has even been stated that cor pulmonale implies cardiac disorders secondary only to pulmonary parenchymal disease. Perhaps the term "cor pulmonale" no longer serves a useful purpose and should be discarded. The time has come to erase the confusion from what has been a problem of semantics primarily.

The conceptual differences around the causal factors of chronic "cor pulmonale" are probably due to the differences in approach when studying the lung. Usually the studies have been focused either on the airways or the vascular structures. Actually, the lung is a unit and should be considered as such, since the airways, vascular channels and lymphatics are integral parts of the whole, both structurally and functionally. If we proceed through analogy with the systemic circulation, confusion can be avoided. The term "hypertensive heart disease" is used when functional and/or structural changes due to persistent systemic arterial hypertension—have become definitely established in the left ventricle. It makes no difference whether the hypertension is "essential" or secondary as far as the cardiac changes are concerned. The pulmonary

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PULMONARY VASCULAR TREE

Figure 1: Schematic representation of the pulmonary vascular tree and of the limits of the arterial and venocapillary sections. R. A.=right atrium; R. V.=right ventricle; P. A.=pulmonary artery; Cap.=capillaries; P. V.=pulmonary veins; L. A.=left atrium; L. V.=left ventricle; Ao.=aorta.
circulation should be considered in the same way. Two factors always exist in chronic cor pulmonale: (a) hypertension of the pulmonary artery and (b) functional and/or organic changes in the right ventricle. The pulmonary arterial pressure becomes elevated (Fig. 1) when the resistance to blood flow increases either in the arterial, the precapillary or the capillary areas\(^5\) (or in two or in all of these three areas\(^6\)) (Fig. 2). The consequences are: (a) systolic overloading of the right ventricle (functional change) and (b) right ventricular hypertrophy with or without dilatation (organic changes) related to the magnitude and—persistence of the pulmonary arterial hypertension. From what has been said by different authors up to the present time, the term chronic cor pulmonale is used when right ventricular hypertrophy exists whether it is or is not accompanied by right ventricular heart failure. The condition is usually considered a consequence of sustained pulmonary arterial hypertension secondary to lung disease.

At this point another question arises. What is to be understood by lung disease? Are we going to consider isolated alterations of the parenchyma, the airways, the vessels or the lymphatics? Let us not forget that the lung like any other organ must be studied as a whole. "Heart disease" usually includes alterations of the endocardium, the myocardium, or the pericardium either individually or in combination. When no pathology of these last named structures can be proved, but electrocardiographic and/or clinical evidence of coronary insufficiency exists, the term "heart disease" is completely justified. The term is applicable irrespective of the portion involved and without regard to whether the abnormality is structural or functional. If this reasoning were to be applied to the lung, lung disease exists when any structural or functional alteration of any of its constituent parts is present and not necessarily when only alterations of the parenchyma or interstitial structures are shown. Looking at the problem in this light, any morphologic or functional alteration of the airways, the blood vessels or lymphatics within the lung should be considered in a definition of "lung disease."

When lung disease produces a persistent elevation of the pulmonary arterial blood pressure, right ventricular systolic overloading and later on hypertrophy is the natural consequence. This is seen commonly in diffuse and chronic lung disease. If several of the diseases which have been excluded from the etiology of chronic cor pulmonale are also considered, it will be seen that they cannot be treated in this way logically. Let us start with mitral stenosis which has been the cause of considerable

**PULMONARY VASCULAR TREE**

**FIGURE 2:** Schematic representation of the different areas of the pulmonary vascular tree
debate. Some of the patients with mitral stenosis develop a much higher and sustained pulmonary arterial blood pressure than those who have a disease accepted as causal of chronic cor pulmonale as is the case in pulmonary emphysema. Some patients with mitral stenosis show important histologic changes in the interalveolar walls, arterioles and small muscular arteries of the lung.6 These changes produce serious hemodynamic alterations in the pulmonary arterial section. This complex of changes will not always disappear when mitral stenosis has been successfully treated. The secondary vascular alterations in the lung in these patients produce pulmonary arterial hypertension. These changes which are predominantly localized to the vascular structures justify the term “vascular lung disease.”

If congenital heart disease with left-to-right shunt is studied, it will be seen that some cases are accompanied by significant and sustained pulmonary arterial hypertension. This happens, for example, in some cases of patent ductus arteriosus and in some with ventricular septal defect. It does not happen in most cases of atrial septal defect where the pressure elevations are only small.8 Histologic lung studies in cases of patent ductus arteriosus and ventricular septal defect reveal profound changes of the arterioles and small muscular arteries.9,10 These vascular changes can also justifiably be labeled as “vascular lung disease” as was done before when mitral stenosis was considered.

Any kind of lung disease, be it parenchymatous, interstitial, vascular or extrinsic, when it is diffuse and accompanied by important functional alterations, may elevate the pulmonary arterial pressure sufficiently to produce right ventricular hypertrophy. Once these two conditions are present—that is, pulmonary hypertension and right ventricular hypertrophy—one can speak interchangeably about chronic pulmonary hypertensive heart disease or chronic “cor pulmonale.”

**Etiology**

Many causes have been recognized as etiologic of chronic cor pulmonale,11,12 and at a given moment, it is hard to remember them all. For practical purposes and to obviate this difficulty, they parenchymatous and interstitial lung dis-

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**CHRONIC PULMONARY HYPERTENSIVE HEART DISEASE**

![Diagram of Chronic Pulmonary Hypertensive Heart Disease](image)

**Figure 3:** Schematic representation of the three etiologic groups of chronic pulmonary hypertensive heart disease and sites of action.
can be divided into three groups: (a) ease, (b) vascular lung disease, and (c) extrinsic lung disease (Fig 3).
A. Parenchymatous and Interstitial Lung Disease.
This group covers obstructive emphysema and fibrosis. Different types of emphysema have been considered: idiopathic or primary emphysema,\textsuperscript{18} old age emphysema\textsuperscript{13} and obstructive emphysema.\textsuperscript{14} Of these three, only the last form will produce pulmonary arterial hypertension and its consequent right ventricular hypertrophy. Nevertheless, not all obstructive forms of emphysema will lead to chronic cor pulmonale; this will happen in only about 20 per cent of all the cases according to the statistics cited by Harvey and Ferrer.\textsuperscript{15}
Chronic cor pulmonale occurs in about 40 per cent of patients with pulmonary fibrosis.\textsuperscript{16}
B. Vascular Lung Disease.
Within this group are found arteritis, thrombo-embolism and primary pulmonary hypertension, and vascular changes due to different causes such as: (a) sustained veno-capillary hypertension, (b) transmission of the systemic pressure to the arterial section of the lung as seen in some forms of heart disease with left to right shunt and (c) hypoxia.
C. Extrinsic Lung Disease.
Under the heading of extrinsic lung disease are considered all structural and functional alterations of the lung produced by diseases which restrict the thoracic movements or which compress the lung. The diseases which produce them are the following extrapulmonary ones: deformities of the thoracic cage, thoracic myopathies and extreme obesity, extreme adhesive pleuritis and pleural effusions, and bronchial or mediastinal tumor formations.
Conclusions

Whenever pulmonary arterial hypertension exists long enough—no matter how it was produced—and of such a degree as to cause hypertrophy of the right ventricle with or without dilatation, it is convenient to speak about “chronic pulmonary hypertensive heart disease.” The etiology of pulmonary arterial hypertension recognizes three types of chronic diffuse lung diseases: (a) parenchymatous and interstitial, (b) vascular, and (c) extrinsic.

Resumen
Este estudio lleva a la conclusión de que el término “cor pulmonale” ha dejado de ser útil. Consideraciones semánticas a la luz de nuestros conocimientos actuales llevan al autor a la búsqueda de un término más conveniente. Recuerda que la existencia de hipertensión arterial pulmonar es el requisito para las alteraciones cardíacas subsecuentes. Precursoras de la hipertensión arterial pulmonar son las enfermedades difusas del pulmón, con sus alteraciones estructurales. Una vez que éstas se han establecido son seguidas de hipertensión arterial pulmonar. Por estos razonamientos se propone el término “Cardiopatía hipertensiva pulmonar crónica.”

Resumé
Cette étude amène à la conclusion que le terme “coeur pulmonaire” n’a plus d’utilité actuelle. Des considérations sémantiques, à la lueur de nos connaissances présentes, rendent actuellement nécessaire la recherche d’un terme plus convenable. On doit garder présent à l’esprit que l’existence d’une hypertension artérielle pulmonaire est la condition primordiale expliquant les altérations consécutives du cœur. Les précurseurs de l’hypertension artérielle pulmonaire sont les maladies diffuses des poumons, avec leurs altérations fonctionnelles et structurales. Une fois que celles-ci sont bien établies, l’hypertension artérielle pulmonaire s’ensuit. Ensuite, la séquence des modifications cardiaques prend place. À la suite de ce raisonnement, le terme “cardiopathie due à une hypertension pulmonaire chronique” est proposé.

Zusammenfassung
Die Wegbahner für eine pulmonale arterielle Hypertonie sind diffuse Lungenerkrankungen mit ihren funktionellen und strukturellen Veränderungen. Haben diese erst einmal Platz gegriffen, dann folgt die chronische pulmonale arterielle Hypertonie. In der Folge davon können

References
1 White, P. D.: Heart Disease, 1. Mac Mil- lan, New York, 1931.

EXPERIMENTAL LIGATION OF THE PULMONARY ARTERY

The results of anatomopathologic and microbiologic studies of the lung after ligation and section of the left pulmonary artery in 30 dogs were presented. The animals were grouped into two series of 15. The authors gave no antibiotic to the first group, but administered a single dose of 40,000 U. procain penicillin-G 24 hours before ligation to the second group. The most important histologic alterations in the first group were: necrosis (53.3 per cent), acute inflammation (60 per cent), intra-alveolar edema (69.4 per cent) and intra-alveolar hemorrhage (70 per cent). In three of 15 dogs, they demonstrated Clostridium perfringens in association with zones of necrosis. The left lung alterations in dogs of the second series were less accentuated, and focal necrosis of the pulmonary arteries was demonstrated in just two of the 15 dogs (13.3 per cent). Clostridium perfringens was not found in any of this group. In the left lung, the lesions were advanced or very advanced in 11 of 15 dogs of the first series, and in three of 15 dogs of the second. This means that there was a significant reduction in the extension of the lung injury with penicillin administration. It is concluded that Clostridium perfringens could participate actively in the pathogenesis of the homolateral lung lesion at the ligation site.


MYOCARDIAL CONTUSION DUE TO BLUNT CHEST TRAUMA

Ten cases with myocardial contusions associated with blunt trauma to the chest, with electrocardiographic and clinical features, were seen by the authors. Patients with closed trauma to the chest and who, within a short period of time develop pre- cordial pain and arrhythmia, should be followed closely with serial electrocardiographic studies. Myocardial contusion should be suspected when the patient is young and there is no question of previous heart disease. The majority of these injuries are mild, transient and result in slight or no permanent damage.

The treatment of patients who have sustained chest trauma with myocardial contusion is essen- tially that of patients with myocardial infarction, with one important exception. The patient should not receive anticoagulants, as one of the features of this lesion is hemorrhage into the heart muscle. Rest is essential.

In mild cases, the symptoms usually subside and the electrocardiogram returns to normal in a short time. Quinidine is used in selected patients with arrhythmias. In the more severe cases with electrocardiographic evidence of marked myocardial dam- age, bed rest is necessary, the length of time depending on the patient's tolerance to increased effort and the ECG findings. Digitalis is used when cardiac failure develops. The severe cases should be watched closely during the second week for signs of cardiac tamponade for which immediate surgery is necessary.

All surgeons who see patients after trauma should be aware of this entity, give the patient full benefit of medical treatment, but proceed with the indicated surgical treatment in the overall management of the patient.