Pulmonary Hypertension and Pulmonary Vascular Disease

When a diagnosis of pulmonary hypertension is made, three interrelated questions must be answered: (1) what is the cause of the elevation in pulmonary artery pressure; (2) is pulmonary vascular disease present as a result of or as a cause of the pulmonary hypertension; and (3) what is the degree and extent of pulmonary vascular disease if present?

The first question can usually be answered by complete noninvasive and catheterization investigations except in a few rare causes of pulmonary hypertension such as primary pulmonary hypertension (idiopathic plexogenic pulmonary hypertension), pulmonary veno-occlusive disease and perhaps some forms of pulmonary fibrosis. It is in these latter cases that a lung biopsy plays an important part in making it a definitive diagnosis of the cause of pulmonary hypertension when other primary cardiac causes have been excluded.

Certain empirical conclusions can be made about the nature of the pulmonary vessels in some types of cardiac anomalies associated with pulmonary hypertension. Pulmonary venous obstruction is usually associated with medial hypertrophy of the arteries and veins with mild intimal sclerosis especially of the veins, and these arterial lesions are usually reversible when the pulmonary artery pressure falls after correction of the pulmonary venous obstruction. Plexogenic occlusive intimal disease is never found in pulmonary venous hypertension. Similarly, pulmonary hypertension associated with hypoxic lung disease and pulmonary fibrosis is usually associated with reversible medial hypertrophy and mild intimal sclerosis. On the other hand, the vascular disease associated with congenital heart disease with hyperkinetic pulmonary hypertension can progress at a variable rate to plexogenic or occlusive vascular disease which is probably not reversible after correction of the congenital cardiac defect. In fact, patients with obstructive pulmonary vascular disease may deteriorate and die sooner after "corrective" surgery without the safety of a pulmonary-to-systemic blow-off valve.

Certain correlations have been made between the degree of elevation of pulmonary vascular resistance and the degree and extent of pulmonary vascular disease in patients with congenital heart disease and pulmonary hypertension, and the likelihood of a drop in pulmonary artery pressure after corrective surgery. However, accurate calculation of pulmonary vascular resistance is notoriously difficult and a direct evaluation of the pulmonary vascular bed offers a more reliable index of operability in the individual patient.

Dr. Wagenvoort (see page 814) has presented this considerable experience in evaluating lung biopsies from patients with unexplained pulmonary hypertension, patients with congenital heart disease and pulmonary hypertension, and in patients with unexplained cardiac diagnoses. Examination of lung biopsies in the evaluation of pulmonary vascular disease is most valuable in patients with unexplained pulmonary hypertension and in evaluating the degree and extent of pulmonary vascular disease in patients with hyperkinetic pulmonary hypertension. However, there are some limitations to lung biopsy which must be borne in mind.

Although there were no reported deaths or serious complications in Wagenvoort's series, open lung biopsy is not without risk in patients with severe pulmonary hypertension and advanced pulmonary vascular disease, in whom anesthesia itself and secondary hemorrhage are not infrequent hazards. The size and site of the biopsied specimen are important considerations for an adequate evaluation of the pulmonary vessels. Nine of the specimens sent to Wagenvoort were inadequate for diagnosis, and as he mentioned, the location of the biopsy is important in certain diseases, since the pathology of the pulmonary vessels may be different in each lung or in different parts of the same lung, so that a single biopsy may give a misleading or incomplete evaluation of a whole pulmonary vascular bed.

A not inconsiderable factor in the proper evaluation of pulmonary vessels is the experience, skill, and patience of the pathologist who examines the
lung biopsies. Pathologists of Dr. Wagenvoort's caliber may not be available to most cardiologists.

A method which may compensate for some of the above shortcomings of open lung biopsy in the evaluation of the degree and extent of pulmonary vascular disease is magnification pulmonary wedge angiography. The angiographic appearances of the pulmonary microcirculation have been correlated with pathologic findings, so that the presence or absence of obstructive pulmonary vascular disease and plexogenic lesions can be diagnosed empirically.8 The advantages of wedge angiography over open lung biopsy are that both the hemodynamics and pulmonary vascular bed can be compared during the cardiac catheterization and the procedure is without the risks of open biopsy; multiple injections can be performed in each lung and at various sites in each lung so that a more complete evaluation may be obtained of the extent of the pulmonary vascular disease. Pulmonary venous obstruction,4 primary pulmonary hypertension,6 thromboembolic pulmonary hypertension4 and some forms of pulmonary disease7 all produce distinctive and diagnostic angiographic features and, after some experience, this technique may obviate the need for open lung biopsy in many instances. It remains to be shown whether pulmonary veno-occlusive disease can be diagnosed by wedge angiography.

In an era when so many types of congenital cardiac defects can be repaired, the only limiting factor to the fabrication of a normal cardiopulmonary circulation may be the state of the pulmonary vascular bed. Greater cooperation between the cardiologist and cardiovascular pathologist will lead to the refinement of diagnostic skills in determining the state of the pulmonary vascular bed and help to determine the optimum time for surgical intervention in cardiac defects associated with pulmonary hypertension.

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REFERENCES


Do No Harm—Cheaply

Respiratory therapy is one of those technical orphans that grew up eluding the cold eye of scientific inquiry. Despite recent attempts to correct this oversight,1,2 there are several reasons for suspecting it will continue to do so: the tremendous variation in how "therapy" is delivered to patients; the fact that this service is not "owned" by any single discipline; our woeful ignorance of the factors that make up and influence lung defense mechanisms; and not in the least, the profits these departments bring to hospitals. For over three decades, investigators have been issuing contradictory reports on the merits of this discipline. Since the Sugarloaf Conference,1 the number of publications has measurably increased, but not our insight. Most of these articles, including the current one in this issue of Chest (see page 610) suffer from a lack of suitable controls or from a design that prevents extrapolation to other studies.

For example, after almost a decade of malignement, there is now renewed (but still anecdotal) endorsement of IPPB as a volume modality,3,4 despite evidence that a spontaneous breath provides better ventilation to lung bases than a mechanical one.4 A "standard" regimen of chest physical therapy improves patients with lobar atelectasis,5 but not those with pneumonia.5 Delivering mists to the airways can enhance mucus clearance6 or induce bronchospasm.7 Cough both improves8 and disorganizes11 the flow of mucus from intermediate airways.

These differences should come as no surprise. Patients vary as much in their lung defenses12 as do the people ministering to them. In a survey of six large Chicago hospitals, including three university centers, it was found that the variation in methods of delivering routine postoperative care was as great as their charges (Table 1). One must presume