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EDITORIAL COMMENT

V/Q-SPECT Scintigraphy in Pulmonary Hypertension

Predictor of Mortality Versus Trigger for Transplant, A British Versus American Perspective*

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Despite a plethora of novel pulmonary vasodilators, patients with pulmonary hypertension (PAH) continue to succumb to their disease. Although the newer drugs have resulted in an improved intermediate-term prognosis for many patients (1), they are of limited use for others, and of no use whatsoever in certain PAH subtypes (e.g., group 3 or in those with sickle cell anemia).

The role of lung transplantation (LTX)—the one approach which in effect cures PAH—continues to be poorly defined, particularly with respect to its timing. For PAH patients failing medical management, lung transplantation (LTX) offers the sole meaningful chance for survival (2). (In patients with pulmonary hypertension plus right or left ventricular dysfunction, or pulmonary hypertension plus congenital heart defect, it is *heart and* lung transplantation that constitutes the sole therapeutic option that may lead to survival.) A recent consensus suggests certain triggers for referring PAH patients for transplantation and others for placing them on a waiting list (3), but as we will discuss later, there are difficulties with this approach. Most experts favor LTX for those PAH patients who are within 1 to 2 years of death (3), but such prognostication is difficult or impossible in the present state of clinical science because of unpredictability of the course of PAH, and this is difficult to operationalize in the United States because of the current approach to lung allocation.

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In this issue of *iJACC*, Chan et al. (4), of the UK National Pulmonary Hypertension Service, present what may be a useful insight into the pathophysiology and clinical assessment of PAH. Using ventilation and perfusion single-photon emission computed tomography (V/Q-SPECT) they appear to demonstrate the existence of a clinically and radiologically distinct group of PAH characterized by diffuse vascular damage as imaged by this modality. This may represent a previously unappreciated phase of PAH or the final common pathway of its multiple pathophysiologic subtypes, including idiopathic pulmonary hypertension (IPAH), PAH associated with systemic sclerosis and nonsclerodermatous connective tissue diseases (5), and others. This may even represent a distinct nosologic type of PAH. The importance of this finding is that these patients are characterized by a dramatically increased risk of death. Here, we review the authors' data and conclusions. Furthermore, from a U.S. perspective, which must take into account LTX and the optimization of organ allocation, we propose the more far-reaching conclusion that these V/Q-SPECT findings may have use as the trigger for LTX, which may improve patient survival.

To probe the meaning of V/Q-SPECT findings in adults with PAH, Chan et al. (4) studied 136 sequentially evaluated patients and correlated their V/Q-SPECT patterns with their outcomes and other work-up. The patients all had PAH diagnosed by heart catheterization, but no heart or parenchymal lung disease, and after V/Q-SPECT scintigraphy, they

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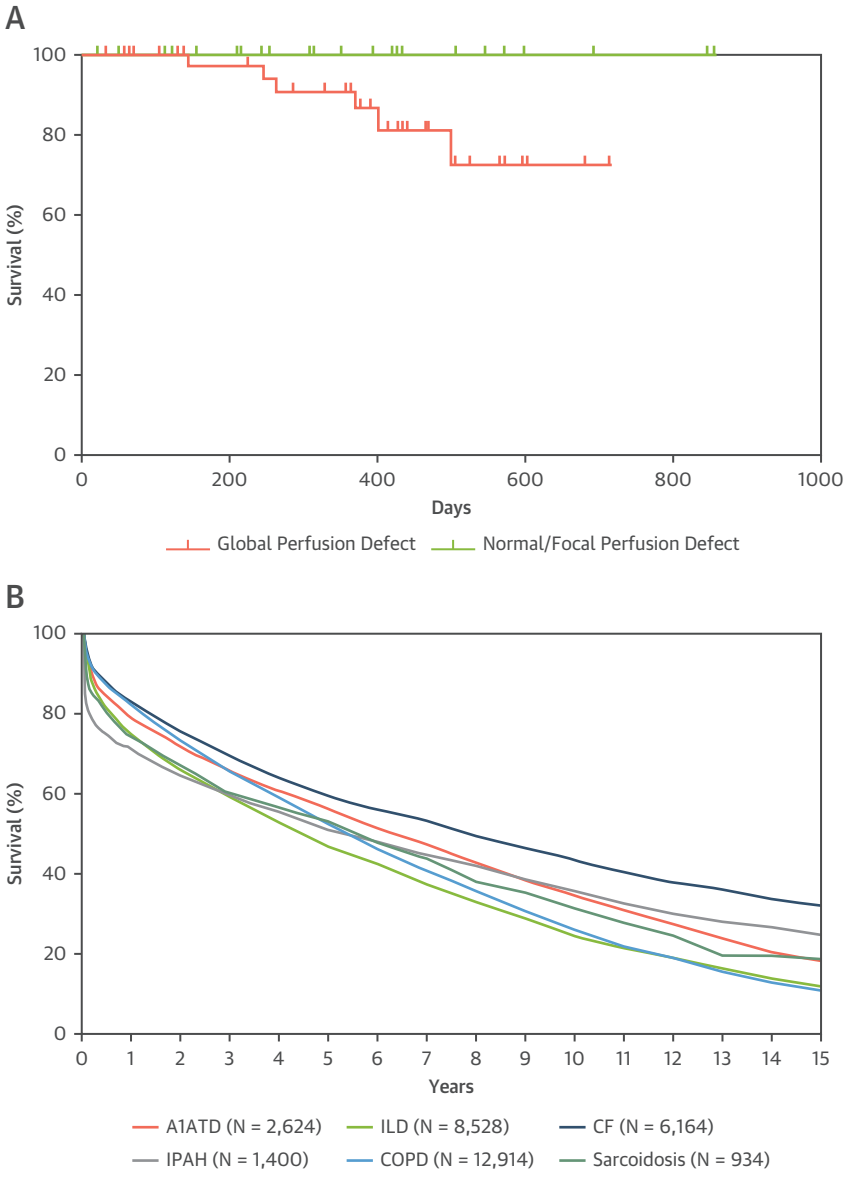
FIGURE 1 Pulmonary Arterial Hypertension: Prognosis Without and With Lung Transplant (Actuarial Data)



(A) Actuarial survival at the Royal Free Hospital, NHS Foundation Trust (London, United Kingdom), according to ventilation and perfusion single-photon emission computed tomography scan result. Presence of diffuse defects predicts worse mortality, with all mortality occurring in this group. All mortality was in the global perfusion defect subgroup. **(B)** Actuarial survival after lung transplantation by diagnosis group, historical data (The International Society for Heart and Lung Transplantation [ISHLT] registry) (transplants: January 1990 to June 2011). A1ATD indicates α1- antitrypsin deficiency-associated chronic obstructive pulmonary disease (COPD); COPD indicates non-A1ATD-associated COPD. CF indicates bronchiectasis associated with cystic fibrosis; ILD indicates interstitial lung disease, which includes idiopathic pulmonary fibrosis; and IPAH indicates idiopathic pulmonary arterial hypertension (6). Actuarial survival after transplantation for the diagnosis of pulmonary hypertension differs from all others in survival **(gray)** in that there is a distinct bimodal slope, with relatively very large early mortality followed by low mortality in extended follow-up.

underwent CT arteriography, and subsequently conventional pulmonary arteriography and targeted invasive pulmonary angiography, where relevant, to exclude distal thromboembolism. What caught the attention of the authors is that 75 patients showed significant perfusion deficits, but in fact had no evidence of thromboembolism on the corroborating angiographies.