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EDITORIAL

"PRIMARY PULMONARY HYPERTENSION IS LIKE CANCER, IN FACT IT IS WORSE THAN CANCER" (J. T. REEVES)

This collection of papers, in this special edition on pulmonary hypertension, in the *European Journal of Medical Research*, is dedicated to the memory of Dr. Jack (John) T. Reeves. His untimely death, after being hit by a car while riding his bicycle, now a little bit over a year ago, left a large gap in the Pulmonary Hypertension community. He was an influential and prominent teacher in the Cardiovascular Pulmonary Research Laboratory, here in Denver, in the 1970s and 1980s. His laboratory functioned as a "boot camp" for trainable young scientists. Jack was a very principled man, and one of his principles was the economy of the spoken or written word. He clearly liked to teach young fellows how to write a scientific paper. His appetite for work was insatiable, and his range of interests spanned high-altitude pulmonary edema, the design and function of the neonatal and adult lung circulation, eicosanoids and techniques to measure pulmonary artery pressure non-invasively. These interests took him to Mount Evans and Pikes Peak, in Colorado, and to the high plains of Tibet. He was interested in the mechanism of hypoxic pulmonary vasoconstriction, and he had once invested a considerable amount of time and effort to develop a model of appetite-suppressant, drug-induced pulmonary hypertension. His legacy was his enormous curiosity. He wanted to find things out and how they worked. To this end, he advocated to "randomize the first case". He frequently quoted the Harvard microbiologist, Hans Zinser, and smiled infectiously after saying: "One can see the whole world through a small window if it is just high enough". Quite likely, he would have enjoyed reading the papers that make up this special edition on pulmonary hypertension. The reader may ask why these papers have been selected for this edition. The answer is simply that they have been written, but, more importantly, they have been selected because all of the papers address very different aspects of the field of pulmonary hypertension research and open up new vistas. The case report on pulmonary hemangiomatosis makes the point that this is a disease of the lung capillaries, which has nothing in common with primary pulmonary hypertension. The case report on sarcoidosis makes the point that sarcoidosis indeed can exquisitely affect the lung vessels without participation of the interstitium, and can generate plexiform pulmonary vascular lesions. The retrospective data analysis by Agne Taraseviciute suggest that the patient population that is now being diagnosed with severe pulmonary hypertension is changing: whereas twenty years ago primary pulmonary hypertension was thought to be a disease of young women, it appears that now most of the patients with this diagnosis are indeed obese post-menopausal women. The date presented by Achcar is sobering, in that it demonstrates that chronic treatment with prostacyclin of patients with endstage angioproliferative vascular disease, treated with lung transplantation, did not alter the degree of the vascular disease. Zimmerman and colleagues, in their study of children with severe pulmonary hypertension, illustrate how a sophisticated and thorough assessment of the pulmonary vascular reactivity can uncover a residual endothelium-dependent vasodilator potential not discoverable by the use of conventional drugs – and that the presence of this endothelium-dependent vasodilation has prognostic implications. Lastly, Quaife and colleagues present exciting data based on the MRI exploration of the failing right ventricle in patients with severe pulmonary hypertension, and they suggest that the measurement of right ventricular wall stress may be superior as a marker of impaired performance of the volume and pressure-overloaded right ventricle.

Many papers written on pulmonary hypertension – in fact, there are too many to count – have an opening paragraph which says: "Severe pulmonary hypertension is a potentially life-threatening disease, and patients die from right heart failure". It is the opinion of this editor that the time has come to recognize that our patients do not die from the pulmonary artery pressure, and that the stretched and stressed right ventricle, which magically repairs itself following lung transplantation, has become a treatment target in its own right. It is becoming increasingly clear that it is not just the afterload, which is "seen" by the right ventricle. We can now see that the mechanical pump concept of the 19th and 20th centuries must be supplemented with the concepts of cell biology, balance of apoptosis and cell growth, inflammation and immune response. Understanding the pathobiology of right heart failure may, perhaps, have the largest impact on the morbidity and quality of life of our patients with severe angioproliferative pulmonary hypertension.

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