The Fifth World Symposium on Pulmonary Hypertension and Robyn J. Barst, MD

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"Stars of the first magnitude are rare, but that such a one will arise among women physicians, I have not the slightest doubt."
William Osler, MD

The Fifth World Symposium (5th WSPH) on Pulmonary Hypertension was held in Nice, France, from February 27 to March 1, 2013, almost 40 years after the first symposium was convened to develop an understanding of a rare disorder made visible by an epidemic of primary pulmonary hypertension (PH) caused by the prescription of Aminorex.

Nazzareno Galiè, MD, and Gerald Simonneau, MD, organizers of the 5th WSPH, dedicated the symposium to the memory of Robyn J. Barst, MD.[1] Drs Galiè and Simonneau appropriately called attention to Dr Barst's broad influence on the field. Indeed, the impact of Robyn's many contributions can be found throughout the 13 state-of-the-art papers published in December 2013 as a supplement to the *Journal of the American College of Cardiology*.

Robyn's career spanned an era of dramatic advances in the diagnosis and management of PH. During her career, primary PH (as it was called years ago) was transformed from a poorly understood disorder for which there was no effective treatment and little hope, to a well-defined disorder, diagnosed and treated by expert physicians like Robyn with an expanded armamentarium of medications and life-saving procedures such as lung transplantation.

I first met Robyn at a meeting of the National Institutes of Health (NIH) Primary Pulmonary Hypertension Registry investigators. I have never forgotten her bright eyes and her energy and enthusiasm for the work that we were about to undertake. We forged a close relationship that was built not only on our shared interest in PH, but also our love of medicine, family, and life in general. We both enjoyed early morning walks before the many meetings that we attended. I will forever picture Robyn at 5 in the morning, dressed in sneakers and walking shorts, ready for one of those walks during which we discussed anything and everything.

Robyn was no passive observer; rather, she was always at the center of the advances in the understanding and treatment of PH. As a young pediatric cardiologist Robyn contributed to the NIH Primary Pulmonary Hypertension Registry (NIH PPH Registry, 1981-1987), and as a senior clinician-investigator she served on the steering committee of the Registry to Evaluate Early and Long-term PAH Disease Management (REVEAL, 2006-2013). Both of these registries provided key epidemiologic data, and allowed the derivation of important prediction rules for survival. The 5th WSPH task force on pulmonary arterial hypertension (PAH) epidemiology and registries reported the changing demographics and survival of patients diagnosed with PAH based on registry data that Robyn influenced.

Robyn also contributed to important advances in our understanding of the genetic predisposition to PAH. Robyn collaborated closely with a team of scientists, including the late Jane Morse, MD, who discovered that mutations in the gene that codes for BMPR2 caused a heritable form of PAH. This discovery, reported almost simultaneously by 2 scientific teams, led to an entirely new understanding of heritable PH. The 5th WSPH task force on genetics and genomics of PAH reports some of the new discoveries, expanding the original contributions of many scientists, including Robyn Barst.


Robyn J. Barst, MD

Robyn was also at the epicenter of the dramatic revision of the clinical classification of PH, made by a task force at the Evian meeting in 1998. She had lived through the era during which clinicians differentiated primary PH from secondary PH, and, like most of us, recognized that this classification no longer accommodated our evolving understanding of PH. The revised clinical classification, proposed first at Evian, has become widely accepted and has provided a cornerstone for global communication among PH experts. As such, it was a centerpiece of Robyn's daily practice. She participated in each of the subsequent world symposia as clinicians and scientists refined the Evian clinical classification. However, nowhere was her influence more obvious than in the 5th WSPH updated clinical classification of PH. Key changes were made to embrace children and the field of pediatric PH, where persistent PH of the newborn was withdrawn from diagnostic Group 1 PAH; and, in agreement with the first task force on pediatric PH, a shared comprehensive classification for adults and children was created. Of course, Robyn was a driving force behind this particular committee.

The working group on definitions and

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diagnosis of PH wrestled with some of the most critical issues faced by PH experts: how should we define PH? Should the pulmonary circulation be challenged with intravenous fluids or exercise? How do we differentiate precapillary from postcapillary PH? As a cardiologist who performed diagnostic pulmonary artery catheterizations, Robyn possessed intimate knowledge of these critical issues, even though she could not participate in the discussions.

Not surprisingly, Robyn's contributions to the treatment of PAH, sustained over her long and distinguished career, emerge as her greatest influence on the field. Drs Galiè and Simonneau again fittingly highlighted Robyn's pivotal report, which demonstrated the efficacy of continuous intravenous epoprostenol for the treatment of primary PH. This landmark clinical trial, conducted by many pioneers in the field, changed the landscape of PH forever and enabled subsequent therapeutic advances. Of course, the epoprostenol breakthrough was only the beginning. Robyn continued to work tirelessly with others to explore new therapeutic advances. Like many of us, she had seen the devastation of PAH and was determined to overcome this disorder for her patients and their families.

Robyn was a principal investigator for pivotal clinical trials demonstrating the efficacy of endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, as well as trials to expand the routes of delivery of prostanoids and trials on combinations of therapeutic agents. Even after Robyn was diagnosed with cancer, she organized a trial of inhaled nitric oxide with the hope that this potent agent would prove to be yet another effective treatment for PAH. The 5th WPHS state-of-the-art task force statements on the updated treatment algorithm of PAH and the exploration of treatment goals of PH underscore her legacy as a clinician-investigator who, along with others, transformed the face of PAH forever. As Drs Galiè and Simonneau observed, the kingdom of the near dead will never be the same because of Robyn Barst.

Robyn's talent, passion, and dedication—known to all of us—shine brightly within the accomplishments of the 5th WSPH. I believe that Dr William Osler had Robyn J. Barst, MD, in mind when he predicted the appearance of a star of the first magnitude.

Reference

1. Galiè N, Simonneau G. The Fifth World Symposium on Pulmonary Hypertension. *J Am Coll Cardiol*. 2013;62(25) Suppl):D1-D3.

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documents for the meeting in Nice. Task force members had a chance to sit down face-to-face and refine the documents at the meeting before finally presenting the task force's recommendations to the broader audience attending the Symposium and submitting them for timely publication.[1]

In this edition of *Advances in Pulmonary Hypertension*, we asked some of the leading contributors to the task forces to summarize their work for our journal. They cover important topics such as classification, epidemiology, registries, definitions, diagnosis, and the new treatment algorithm. The Nice meeting will be remembered for many reasons: the announcement of newly discovered genes for heritable PAH; the imminent expansion of medical treatment into WHO Group 4 Chronic Thromboembolic PH;, the modification of the algorithm to take into account emerging evidence from new therapeutic endpoint, morbidity and mortality.

The next World Symposium is planned for 2018. It is worth looking for a moment to the future and considering what might 2018 be remembered for? Further significant advances in our understanding of the pathobiology of PH, new effective therapies, or perhaps a better understanding of how best to use the therapies we already have? Whatever may emerge over the next 5 years, there is comfort in reflecting on the advances since 1998 and the strong *esprit de corps* and common purpose that have developed in the international PH community which is symbolised by these gatherings every 5 years.

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Reference

1. Galie N, Simmoneau G. The Fifth World Symposium on Pulmonary Hypertension. J Am Coll Cardiol 2013;62 Suppl D: D1–D2