

The 5th WSPH: Advancing the Field of Pulmonary Hypertension – A Global Effort

The 5th World Symposium on Pulmonary Hypertension, held in Nice, France, in April was a gathering of 1138 delegates from 57 countries converging with one common goal: to advance the field of pulmonary hypertension. To carry out this mission, 129 members representing 21 countries comprising 12 task forces assessed the current state of understanding that has evolved since the previous World Symposium. They were charged with reviewing the body of literature that has emerged during the past 5 years and assessing the impact on current practice, and to set forth recommendations for needed changes and future directions. Indeed, the 3 days of presentations, deliberations, stated points and counterpoints – all aimed at trying to derive a consensus based on published data and expert opinions – embodied the work from the PH community worldwide. The global impact of PH was highlighted by emerging data characterizing this disease from parts of Asia and South America, the findings truly

emphasizing that PH does not have any boundaries and affects people of all racial and ethnic backgrounds. Indeed, the focused energy and dedication of the PH community represented in this gathering emphasized the commitment that we are all in this together.

Thus, it is with my sincere pleasure to present to you this issue featuring some of the highlights from the 5th WSPH. I am grateful to our guest editor Dr. Sean Gaine for all his efforts in bringing together the key members of 4 task forces in presenting the focal points of the meeting. My sincere thanks to all our authors – Drs. Nazzareno Galie, Marc Humbert, Michael McGoon, Gerald Simonneau, Rogerio Souza, and Fernando Torres – for their insights on the emerging data and controversies on the topics of epidemiology, PH registries, classifications, treatments, definitions, and diagnosis. This issue also brings you Part 2 of the Pulmonary Hypertension Clinical Centers initiative by Dr. Joel Wirth and Ms. Abby Poms focusing on the implementation of the program, the opportunities of the PHPN organization by Ms. Traci Stewart and Melisa Wilson, and a thought-provoking

discussion on our collective approach to treating our patients and assessing long-term outcome by Dr. Sean Studer. As well, we are very pleased to introduce a new section titled “Pulmonary Hypertension Grand Rounds”, a forum for fellows and junior faculty members to contribute to our Journal.

And finally, it is my sincere privilege to present to you a personal tribute to Dr. Robyn Barst by Dr. Greg Elliott. This was the first World Symposium without Dr. Barst in the “thick of the discussion,” voicing her thoughts, sharing her experiences and wisdom, giving us a greater perspective and, in turn, asking us to reach higher and do better. Her presence, however, was deeply felt during all the presentations and discussion, a true testimony to her everlasting contribution to the field of pulmonary hypertension.

Myung H. Park, MD

Associate Professor of Medicine
Director, Pulmonary Vascular Disease Program
University of Maryland School of Medicine

GUEST EDITOR'S MEMO

The World Cup, Olympic Games and PH World Symposium

Many major International sporting events take place every 4 years. Whether one is an avid sports enthusiast or not, one cannot but be touched in some way by the global reach of the events. Perhaps more important than the actual games themselves is the way they mark the passing of time and the link with important other events in our lives. The Atlanta Olympic Games in 1996 might have occurred during one's college years or be remembered for occurring the same year as a new arrival in the family. The World Symposia in Pulmonary Hypertension have a similar link in the

lives of the broad PH community. From the diverse locations (Evian, Venice, Dana Point, and Nice), to their International collegiality and 5-year cycle, there is an air of the Championship about the meetings.

The first World Symposium of the current era in Evian, France in 1998 was memorable. Perhaps for the first time, all the great names in PH research were present in one room and the genuine sense of international camaraderie was truly inspiring. The Evian meeting marked the drafting of a new treatment-based classification of PH that ultimately resulted in increased interest in PAH as a target for new drugs and to the subsequent development of many of the therapies we have today. The Venice

meeting in 2003 heralded a fresh new treatment algorithm, with the introduction of first oral therapy for PAH. In 2008 the meeting moved to Dana Point in California. The classification was further rationalized and the treatment algorithm was updated to include guidelines on combination therapy.

The World Symposium moved back to Europe last year and Nice witnessed a significant increase in the number of delegates. While the event takes place over three days, there is tremendous preparation during the preceding year. In all, 12 task forces were assembled with 129 PH experts from around the world. Work was done via emails and teleconferences to ultimately produce working

(Continued on page 16)

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

GUEST EDITOR'S MEMO

(continued from page 2)

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 dis-

covered 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.

Sean Gaine, MD, PhD

Director, National Pulmonary Hypertension Unit

Mater Misericordiae University Hospital
University College Dublin
Dublin, Ireland

Reference

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