

# Pulmonary hypertension in India: Need for organized approach

Pulmonary hypertension (PH) is a diverse set of disorders which result in elevation of pulmonary arterial pressure. Pulmonary arterial hypertension (PAH) is primarily a disease of pulmonary arterioles which increases pulmonary vascular resistance, eventually leading to right ventricular enlargement, right heart failure, and increased mortality.<sup>[1,2]</sup>

Although there are multiple medications available to achieve a low-risk status with reduced mortality, there is no curative therapy available as yet. Current management of PAH is a severity and risk-based approach with early initiation of combination and/or parenteral therapy for high-risk patients.<sup>[2,3]</sup> Even in the developed world, management of PAH is challenging due to complexity of pathology, need for invasive diagnostic studies, and high cost of therapies. The management of PAH in a developing country is immensely challenging.

In this issue of the journal, Jariwala *et al.* present the clinical experience and outcomes of PAH patients with World Health Organization (WHO) Functional Class (FC) II to IV treated with macitentan and followed for 12 months.<sup>[4]</sup> In this retrospective, observational cohort study, twenty patients were treated with macitentan. Before the initiation of therapy, 50% of the patients were in WHO FC II, 35% in class III, and 15% in class IV. Fifty-five percent of the patients were on preexisting PAH therapy. Among these patients treated with macitentan, there was a significant improvement in FC, exercise capacity as measured by 6-min walk distance, plasma N-terminal pro-brain natriuretic peptide, and echocardiographic parameters. The results of this study are consistent with observations on macitentan therapy in other populations.<sup>[2,5]</sup>

While the authors of the study deserve commendation for conducting this study, the study highlights inherent limitations of these efforts. This is a single-center, retrospective study with small number of patients monitored clinically. The number of patients included from a single center is significant, however, not enough to draw significant conclusions applicable to a large population.

PH is a rare disease with need for complex diagnostic workup and advanced therapies, which may not be available in all areas even in the developed world. These issues are understandably more challenging in the developing world. In their recent publication, Idrees *et al.* highlighted these issues and made recommendations

for collective but local approach.<sup>[6]</sup> While approach to diagnosis and management should be based on robust studies conducted anywhere in the world, these need to be tailored to the local populations which have unique social and economic determinants of health. It is likely that burden of PH is higher in developing countries due to diversity of underlying etiologies and large population involved. One can certainly conclude that burden of PH is high in India due to its large population base. There is no dearth of highly qualified experts in India. What is needed is to get organized and develop collaborative efforts similar to other countries.<sup>[6,7]</sup>

Our center participated in a large registry, and we have firsthand experience of direct and indirect benefits of participation in such efforts. In addition to robust data and outcomes, physicians across the United States had opportunity to network with world experts on PH.<sup>[7]</sup>

There appears to be beginning of such approach in India. Recently, Mehrotra *et al.*<sup>[8]</sup> published results from data based on a single-center registry and Harikrishnan *et al.*<sup>[9]</sup> outlined design for a Kerala, India, based regional registry. A national approach to address the challenges of managing PH in India is the need of the day. A national PH registry or a network of physicians with interest in PH with well-defined design and criteria can possibly collect data on one of the largest PH populations in the world. Funding for such opportunity is challenging, but initial time commitment from few physician leaders to develop such network will pave the way for collaboration with pharmaceutical industry. This will be a win-win situation for patients, physicians, and pharmaceutical industry. It is time for physicians in India to take the lead.

**Amina Pervaiz, Ghulam Saydain**

*Department of Medicine, Pulmonary Critical Care and Sleep  
Division, Wayne State University, Detroit, MI, USA  
E-mail: gsaydain@med.wayne.edu*

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