

Understanding the perspective of patients with pulmonary arterial hypertension: looking beyond health-related quality of life

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To the Editor

Pulmonary arterial hypertension (PAH) is a rare and incurable disease characterized by increased pulmonary arterial pressure and pulmonary vascular resistance. Commonly reported symptoms include shortness of breath, tiredness, dizziness, and chest pain.¹ Over the last three decades, the health status and life expectancy of patients with PAH have significantly improved thanks to advances in treatment and clinical management. However, these positive changes have increased the salience of yet new issues related to the burden of living with PAH in the long term.² Accordingly, recent efforts have been made to integrate the perspective of patients within clinical practice, and to raise awareness of the importance of centering clinical care on improving patients' health-related quality of life (HRQoL).³ However, an approach to care centered on improving HRQoL may overlook aspects that are highly relevant for patients, which seems to be the reason why it is necessary to consider their perspective.

The construct 'HRQoL' comprises "the health aspects of quality of life (QoL)", and has generally been used to reflect on "the impact of disease and treatment on disability and daily functioning".^{4,5} Interest in HRQoL within the field of PAH grew in parallel to the development of PAH-specific medications, when efforts were mostly directed at increasing survival while assessing the impact these PAH-specific medications had on patients.⁶ However, the challenges faced in this field have since grown in complexity. As reported in recent patients' accounts and surveys,^{2,7} the lives of patients are impacted not only by PAH-related issues, but also by economic and psychosocial

factors not traditionally addressed by research and clinical practice focused on HRQoL.⁵ Accordingly, these new challenges call for new ways of caring for patients with PAH, and if the goal is to embrace the patient perspective and offer long-term holistic care, then the clinical focus should be extended beyond the health-related aspects of QoL.

Quality of life has been conceptualized in different ways. Nonetheless, there is consensus that it constitutes a broader construct than HRQoL and that it comprises all factors that impact upon an individual's life (i.e. related and unrelated to health, e.g. material comfort, personal safety, relationships, leisure, and spirituality).^{4,8} The World Health Organization defines QoL as "individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards, and concerns".⁹ QoL is subjective in nature, and such subjectivity has been essential in informing treatment decisions and long-term management in the context of chronic conditions,⁶ as well as in helping increase awareness that improvements in health status and survival do not always correspond to improvements in QoL.⁸ Therefore, a focus on increasing patients' QoL, rather than just HRQoL, may provide researchers and clinicians with a mind-set better suited to comprehending the perspective of patients with PAH and addressing current challenges, given that these transcend medically related aspects.

The field of PAH should devote more attention to grounding research and clinical practice on QoL/HRQoL theory in the interest of addressing its current challenges. First, this is essential to reach a

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precise and common understanding of the similarities and differences among commonly used constructs (e.g. health status, HRQoL, and QoL), and to align research and clinical practice with such understanding. This would also help researchers/clinicians differentiate between different types of patient-reported outcome (PRO) measures, including generic measures of health status (e.g. SF-36, EQ-5D, NHP),¹⁰⁻¹² measures developed to assess HRQoL in other cardiovascular conditions (e.g. CHFQ, MLHFQ),¹³ and measures specifically developed to assess HRQoL (e.g. emPHAsis-10, PAH-SYMPACT, CAMPHOR)^{14,15} and QoL (e.g. CAMPHOR)¹⁶ in PAH. Carefully constructed PRO measures typically inform readers about their conceptual underpinnings, and thus provide a clear idea of what is being measured. Nonetheless, health status, HRQoL, and QoL measures have been often used indistinguishably in the PAH and general literature,^{3,4,8,16} and such inconsistency has been known to lead to inaccurate interpretation of findings, and subsequently, to faulty clinical decision making.¹⁷ Second, research and clinical practice should be based on theoretical models that are useful in accounting for and addressing important issues. For example, most HRQoL models focus on issues related to health and functional status rather than on the self-management of disease,⁵ and this last aspect is, more than ever, of utmost importance to patients with PAH. In contrast, QoL models encompass a broader array of factors that can play important roles in helping patients manage PAH in the long term, such as individual (e.g. psychological flexibility, social support), social (e.g. awareness of PAH, access to care), and economic factors.^{2,4,8} Consideration of these factors can stimulate new research and open new avenues for implementing interventions directed at increasing the overall well-being of patients. This for example could contribute to attenuating the high rates of anxiety and depressive symptomatology afflicting patients with PAH, which have been found to range from 19% to 48% and 7.5% to 55%, respectively.¹⁸ Further, addressing these types of factors seems especially relevant in the context of an incurable condition such as PAH, where there is often limited room for improvements in health and functional status. Last, taking a theoretical stance that is congruent with the current challenges is important because it can define the nature and extent of responsibilities in clinical practice. There is a clear need for multi-disciplinary collaboration in the field of PAH,³ and doing so requires the acknowledgment that both

health-related and health-unrelated factors must be addressed to improve the well-being of patients and to transform clinical care accordingly; doing this is entirely congruent with the perspective of QoL models. In contrast, a continued focus on HRQoL may result in the neglect of important psychosocial determinants of the QoL of patients with PAH. After all, in the words of Robertson Davies “the eyes see only what the mind is prepared to comprehend”.

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