The patient tells it! The importance of patient's quality of life perception in pulmonary arterial hypertension risk assessment

Silvia Ulrich [1] and Ekkehard Grunig[2]

Affiliations: [1]Clinic of Pulmonology, University Hospital Zurich, Zurich, Switzerland. [2]Centre for Pulmonary Hypertension, Thoraxklinik at Heidelberg University Hospital, Heidelberg, Germany.

Correspondence: Silvia Ulrich, Dept of Pulmonology, University Hospital Zurich, Raemistrasse 100, 8091 Zürich, Switzerland. E-mail: silvia.ulrich@usz.ch

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Assessment of quality of life in pulmonary hypertension is important to identify additional therapeutic needs, and patients with good QoL have better prognosis. QoL assessment in PH care can be done with validated short questionnaires, such as emPHasis-10. https://bit.ly/3s5jF3U

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Everyone wants quality of life (QoL), regardless whether a person is healthy or diseased. However, QoL means something different for every individual. QoL is not only influenced by the individual's preferences, wishes and expectations towards life, but also by the time of living, geographical, socioeconomic and political environment and, of course, the health state, which all contribute to the individual's resources to live with a high quality in happiness and satisfaction. The World Health Organization defines QoL as the individual's 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 [1]. Due to this complexity and subjectively differently weighted factors, and also the fact that different disciplines define QoL differently, measurement of QoL is challenging in health and disease [2]. For healthcare providers in medicine, it is crucial to understand patients self-reported QoL in order to improve treatment towards symptom relief, rehabilitation and better prognosis, and discard therapies that are of little benefit for patients. In order to shift limited resources to therapies which induce meaningful changes for patients, health authorities increasingly ask that novel medical therapies demonstrate the capability to improve patient-oriented outcomes such as QoL in high quality trials, instead of relying on biomedical endpoints [3]. Whereas generic questionnaires may be used to assess and compare QoL between different populations including healthy and diseased, health-related QoL (HRQoL) assessment tools take into account disease-specific factors and are thus more suited to measure meaningful changes for patients brought about with therapy [4].

The cardinal symptom of patients with pulmonary arterial hypertension (PAH) is progressive dyspnoea on exertion, which increasingly limits the ability of patients to take part in daily activities, and is associated with anxiety and a high socioeconomic burden [5]. More than a decade ago, it was shown that HRQoL is not only reduced in PAH, but also correlates with the severity of symptoms, the presence of anxiety and

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depression [6, 7] and traditional markers of disease severity, and is associated with a worse prognosis [8–10]. Psychosocial support is therefore recommended in PAH by European Society of Cardiology (ESC)/European Respiratory Society (ERS) pulmonary hypertension guidelines [11]. Subsequently, an increasing number of randomised controlled therapeutic trials in the field of PAH have assessed HRQoL by different instruments, and it has been shown that several drugs or their combination, rehabilitation programmes and oxygen therapy improve QoL in distinct PAH populations [12–17]. However, despite its importance for patients, QoL is not implemented into the risk assessment strategy as proposed by the latest ESC/ERS pulmonary hypertension guidelines, potentially due to a lack of standardisation [11]. Whereas early studies on HRQoL in PAH mostly used general instruments or questionnaires that were developed for comparably symptomatic patients with congestive heart disease, several PAH-specific instruments have subsequently been developed [9, 18–23]. However, the widespread use of some of these PAH-specific instruments was hindered by the extensive nature of some questionnaires, which did not suit limited consultation times, the need for user-licenses and, especially, the failing comprehensive validation or translation into different languages, cultural and geographic regions [24, 25]. The relatively short, quickly applicable and simple 10-item emPHasis-10 score (with a higher score demonstrating a higher symptom burden) has demonstrated good validity against other measures, a high re-test and internal consistency and is validated in several languages [23]. In this issue of the *European Respiratory Journal*, Borgese *et al.* [26] confirm the correlation of emPHasis-10 scores with other markers of disease severity, and that it serves as quantitative measure of patients' overall perception of the impact of PAH on their life in a large collective of US patients as retrieved by the Pulmonary Hypertension Association Registry (PHAR). Additionally, this registry-based real-life analysis suggest for the first time a minimal important difference of the emPHasis-10 score of −6 points, as a basis for further anchor-based validation [26, 27]. In a second article in this issue, Lewis *et al.* [28] describe in a large UK multicentre study that emPHasis-10 scores are independent prognostic markers in patients with idiopathic and connective tissue disease-associated PAH, thus providing an additional value on top of currently used parameters and revealed a minimal detectable difference of 9 points.

Thus, taking into account patients self-reported perception of HRQoL is of high value for healthcare providers in the risk assessment of patients with PAH and may be of especially high value in stratifying the large group of patients assigned to the intermediate risk group according to current guidelines [11, 29]. Listening to patients' needs is potentially the oldest, principal art of medicine. If we succeed in incorporating patients' comprehensively scored perception of their quality of life in risk assessment and, in a further step, individually risk-adapted management strategy, we will hopefully improve the QoL of patients living with PAH. The two presently published articles on the emPHasis-10 score are steps in this direction.

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