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

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Abstract: 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, socio-economic 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].

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Editorial to the ERJ

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

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The world health organization defines quality of life (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 Everyone want's quality of life, 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. Due to this complexity and subjectively differently weighted factors and the fact, that also different disciplines define QoL differently, measurement of QoL is challenging in health and disease.² For care 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 which are of little benefit for patients. In order to shift limited resources to therapies which induce meaningful changes for patients, health authorities more and more ask that novel medical therapies have shown the capability to improve 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 for patients meaningful changes with therapy.⁴

The cardinal symptoms of patients with pulmonary arterial hypertension (PAH) is progressive dyspnea on exertion, which increasingly limits patients to take part in daily activities and is associated with anxiety and a high socioeconomic burden.⁵ Since more than a decade ago, it has been shown that HRQoL is not only reduced in PAH, but also correlates with traditional markers of disease severity and is associated with a worse prognosis.⁶⁻⁸ Subsequently, an increasing number of randomized-controlled therapeutic trials in the field of PAH assessed HRQoL by different instrument and it has been shown that several drugs or their combination, rehabilitation and oxygen therapy improve QoL in distinct PAH populations.⁹⁻¹⁴ However, despite its importance for patients, QoL is not implemented into the risk assessment strategy as proposed by the latest ERS/ERS pulmonary hypertension guidelines, potentially due to lacking standardization.¹⁵ 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 were subsequently developed.^{7,16-21} However, the wide-spread use of some of this PAH-specific instruments was hindered by the extensivity of some questionnaires which did not suit into limited consultation times, the need for user-licenses and especially the failing comprehensive validation or translation into different languages, cultural and geographic regions.^{22,23} 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.²¹ In this European Respiratory Journal, Borgese et al. 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).²⁴ Additionally, this registry-based real-life analysis suggest for the first time a minimal important difference of the emphasis-10 of -6 points, as basis for further anchor-based validation. In second article in this European Respiratory Journal, Lewis et al. describe in a large UK-multicenter study that emphasis-10 score 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 scores.²⁵ Thus, taking into account patients' self-reported perception of HRQoL is of high value for care providers in the risk assessment of patients with PAH and may be of especially high value to stratify the large group of patients assigned to the intermediate risk group according to current guidelines.^{15,26} Listening to the patients need is potentially the oldest and principal art of medicine, if we succeed in incorporating patient's comprehensively scored perception of their quality of life in risk assessment and, in a further step, individually risk-adapted management strategy, we hopefully achieve to improve PAH-patients QoL. The two presently published articles on the emphasis-10 score may be steps towards this direction.

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