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EDITORIAL

Primary Ciliary Dyskinesia: Ready for Quality of Life Assessment

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Primary ciliary dyskinesia (PCD) is a chronic, inherited disease characterized by ciliary dysfunction leading to defects in mucociliary clearance, thus causing upper and lower respiratory problems such as sinusitis and bronchiectasis as well as infertility. Diagnostic workup includes a combination of different techniques such as nasal nitric oxide measurement, high-speed video microscopy analysis, electron microscopy, and genetic analysis because there is no single gold standard due to the genetic and phenotypic heterogeneity of the disease [1,2].

Due to the chronic nature of the disease, difficulty in expectorating respiratory secretions, chronic presence of symptoms such as productive cough, and requirement of daily treatment, this disease poses a burden on the patients and their families. PCD negatively impacts quality of life (QOL) because of disease complications as well as treatment burden. Moreover, treatment adherence decreases with time in patients, which might further deteriorate prognosis [3].

World Health Organization defines "health" as a complete state of physical, mental, and social well-being; thus, patient-reported outcomes, especially health-related QOL (HRQOL), are important components of health and need to be assessed as part of a routine care in chronic diseases such as PCD [4].

In the previous issue of the Turkish Thoracic Journal, Emiralioglu et al. described the translation procedure of PCD-specific HRQOL. Forward translation procedures followed international standards, where two independent and blinded translators fluent in English and Turkish participated. Backward translation into English was performed by a third independent translator. The final Turkish translations of the questionnaires were applied to five subjects from each age group: adult, adolescent, and pediatric PCD patients as well as five parents. After the subjects completed the questionnaire, a cognitive debriefing session was performed, where each item of the questionnaire was discussed by the subjects. Finally, all these information were used to achieve a final Turkish translation of the PCD-specific HRQOL that is ready for validation and reliability studies [5].

The PCD-QOL questionnaire developed by Lucas et al. has different domains for different age groups: pediatric, adolescent, and adult patients as well as parents. These domains mainly evaluate physical, emotional, and social aspects of PCD related to QOL. Moreover, there are different domains for various symptoms at different age groups. The total numbers of items in the questionnaires are 37 in the questionnaire for children, 43 in the one for adolescents, 49 in the one for adults, 41 in the parents' questionnaire [5-7]. Content validity of this questionnaire has been shown in English-speaking populations [6]. However, translations of QOL measures require cultural adaptation besides linguistic translation, and cognitive debriefing is one of the most important steps of this adaptation.

Assessment of QOL is an essential part of follow-up of children with chronic diseases such as PCD. Thus, it is important that we have a cultural adaptation of a Turkish PCD-QOL questionnaire. Next step shall include the demonstration of validity and reliability of this questionnaire in Turkish patient population.

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