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Translation of the quality-of-life measure for adults with primary ciliary dyskinesia and its application in patients in Brazil

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ABSTRACT

Primary ciliary dyskinesia (PCD) is a genetic disorder that is typically inherited in an autosomal recessive manner. It is clinically characterized by recurrent respiratory infections. However, its repercussions for patient quality of life should not be overlooked. Studies have shown that PCD has a significant impact on the lives of patients, although there are as yet no PCD-specific markers of quality of life. To address that problem, researchers in the United Kingdom developed a quality-of-life questionnaire for patients with PCD. The present communication focuses on the process of translating that questionnaire into Brazilian Portuguese, through a partnership between researchers in Brazil and those in the United Kingdom, as well as its subsequent application in patients in Brazil.

Keywords: Quality of life; Kartagener syndrome; Surveys and questionnaires.

Primary ciliary dyskinesia (PCD) is a genetic disorder that is typically inherited in an autosomal recessive manner. It is characterized by recurrent respiratory infections caused by impaired mucociliary clearance, symptoms often appearing soon after birth.[1] Patients presenting with the triad of dextrocardia, bronchiectasis, and chronic rhinosinusitis are said to have Kartagener's syndrome. Up to 50% of patients with PCD have situs inversus due to abnormal motility of the nodal cilia, which are responsible for the left-right body orientation during embryonic development.[2]

The clinical spectrum of PCD is broad. However, when there are no laterality defects, the level of clinical suspicion of the disorder remains low. The clinical presentation includes neonatal respiratory distress syndrome, evolving to rhinosinusitis, chronic otitis media, recurrent pneumonia, and bronchiectasis. In this context, it is expected that PCD will have a significant impact on the quality of life of patients and their families.[3]

Studies on the psychological and cognitive aspects of PCD have contributed to a better understanding of the psychosocial needs of the affected patients. A pioneering study in this field[4] showed, through the use of questionnaires, that children with PCD had significantly higher scores for internalizing problems, as well as scoring higher for somatic complaints, symptoms of anxiety, and symptoms of depression, when compared with a control group. Parental distress and maternal stress were also significantly more common among parents of patients with PCD.[4]

The authors of a study[5] in which questionnaires were applied to 78 patients diagnosed with PCD (mean age at diagnosis, 9.4 years) concluded that patients with a higher "treatment burden" had a poorer quality of life.

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Over time, those patients lost interest in treating the disease, showing lower levels of treatment adherence. Therefore, early diagnosis and better treatment strategies—requiring greater physician knowledge about the condition and measures to encourage adherence to the treatments proposed—are paramount.[5]

A recent systematic review[6] on the psychosocial impact of PCD identified 14 studies conducted, variously, in the United States and in several European countries. That review showed that, over time, quality of life decreases in patients with PCD over the years, the disorder having significant effects on the physical aspects of quality of life (limitations in activities of daily living), as well as on its emotional aspects (frustration, anxiety and stress), and social aspects (stigmatization).[6]

The lack of PCD-specific quality of life markers led to the development of a quality-of-life measure for patients with PCD (the QOL-PCD), which was first devised for use in adult patients and later adapted for use in pediatric patients.[7,8] The questionnaire was developed through individual and group interviews with specialists, adult patients with PCD, and parents of pediatric patients with PCD. The questionnaire was later refined following cognitive interviews. The instrument contains 37, 43, 41, and 48 items in its child, adolescent, caregiver, and adult versions, respectively. The English-language version of the QOL-PCD for adults has recently been validated, representing an important step for its use in research and clinical practice.[9]

The objective of this brief communication was to present the process of translating the QOL-PCD from English into Portuguese, conducted through a partnership between Brazilian and British researchers from the referral center for PCD at the University of Southampton, in the United Kingdom. The process of translating and applying the questionnaire is described here. Our aim was to make an instrument in Portuguese available for the psychosocial evaluation of patients with PCD in Brazil and, as a consequence, enable more integrated care to be provided to those living with the disease, as well as to provide an important research tool for this population.

The QOL-PCD was initially translated from English into Portuguese by two researchers, working separately. Both were fluent in English and were native speakers of Brazilian Portuguese. Subsequently, there was a discussion among those two researchers and one of the creators of the original questionnaire. In that discussion, they compared the meaning of the Portuguese translations with the original meaning in English in order to reach an agreement and produce a consensus version. Another researcher, also fluent in English and a native speaker of Brazilian Portuguese, with no access to the original questionnaire, then translated the Portuguese-language version back into English (i.e., performed a back translation). There was then another discussion, among the two researchers who did the translation, the researcher who did the back translation, and the developer of the original questionnaire in English, in order to make the translation as faithful as possible to the original version. In this process, cultural differences between Brazil and the United Kingdom were taken into account, and, in order to adapt the Brazilian version to the social and cultural reality of the country, a few changes were made. Thus,

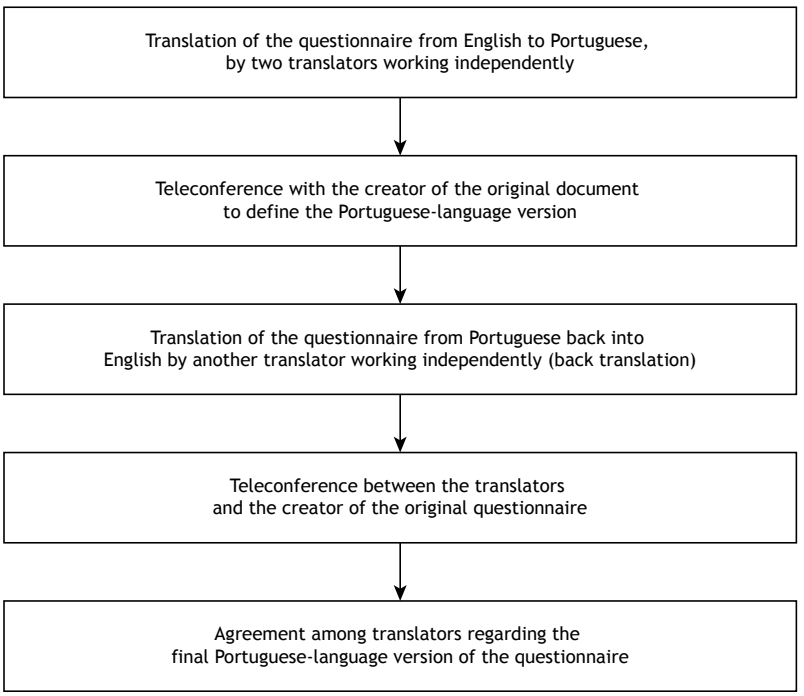


Figure 1. Algorithm for the process of translating the questionnaire from English into Portuguese.

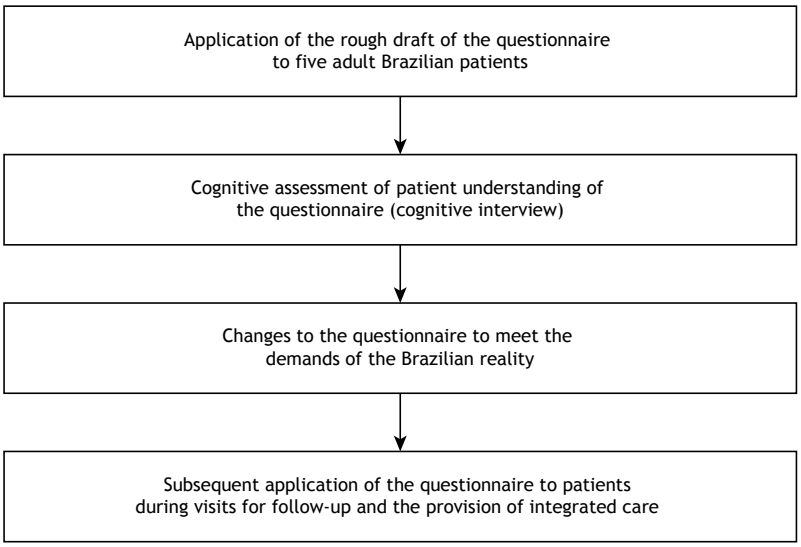


Figure 2. Algorithm for the application of the rough draft of the Portuguese-language version of the quality-of-life questionnaire for patients with primary ciliary dyskinesia, the cognitive interview, and the creation of the final version of the questionnaire.

we created four questionnaires in Portuguese, one for each target population: children, adolescents, adults, and caregivers (Figure 1).

The QOL-PCD for adults includes questions related to three domains of patient quality of life: everyday life; school, work, or activities of daily living; and symptoms. The questions address how patients feel in their everyday life, how they are affected by the disease and the treatment, and whether or not they experience impairment in performing their activities of daily living. The possible responses vary depending on to what the given block of questions refers, such as the frequency or intensity of a certain situation. Greater frequency and more pronounced repercussions for the life of a patient translate to poorer quality of life. The questionnaire can be accessed in full in the online supplement (Appendix 1) on the JBP website (http://jornaldepneumologia.com.br/detalhe_anexo.asp?id=59).

These are examples of the questions in Portuguese:

- *"Na última semana, com que frequência você se sentiu cansado/preocupado/cheio de energia/exausto/triste?"*

- *"Atualmente, quanto tempo por dia você passa fazendo o seu tratamento? Seu tratamento tornou suas atividades diárias mais difíceis de serem realizadas?"*

- *"Pensando no seu estado de saúde na última semana, indique o quão verdadeiro é cada uma das frases para você: 'Eu me sinto confortável ao tossir na frente de outras pessoas'; 'Eu me sinto preocupado por estar em contato com pessoas doentes'; 'Sinto-me sozinho'"*

The questionnaire was applied to adult patients with PCD seen at the Bronchiectasis Outpatient Clinic of the University of São Paulo. All participating patients gave written informed consent. After completing the questionnaire, in compliance with the protocol for validation of the questionnaire outside the United Kingdom, a group of five patients answered the questions of the so-called "cognitive interview". In the individual interview format, the session aimed to evaluate how Brazilian patients process the questions and the response options (i.e., the clarity and objectivity of the questions). Thus, it was possible to evaluate patient understanding when reading the questions and whether or not the meaning was consistent with the intent of the researchers. On the basis of that evaluation, changes were made to the instructions given in the headers, as well as to the questions and response options (Figure 2).

These are examples of the questions asked in the cognitive interview conducted after patients completed the prototype questionnaire in Portuguese:

- *How clear are the response options? Is there a better way to formulate the question?*

- *Are there any questions that do not apply to you or apply to events that have not happened to you? If so, which ones are they?*

- *Are there any questions that sound confusing or are difficult to answer? If so, which ones are they?*

- *Have we forgotten anything important?*

With the results of the cognitive interview in hand, we were able to take into consideration the point of view of the patients with PCD regarding the QOL-PCD. Another discussion among the researchers involved led to new adaptations in the questionnaire to meet the Brazilian demand in the best possible way.

Researchers and clinicians who wish to use the questionnaires for research or clinical purposes should contact the copyright holders (Jane Lucas, Margaret

Leigh, Alexandra Quittner, or Sharon Dell; e-mail to jlucas1@soton.ac.uk) for an agreement between the parties. Once permission is granted, the use of the questionnaires is free of cost.

In conclusion, the translation of the QOL-PCD into Portuguese and its final revisions followed a systematic and interactive approach in collaboration with the researchers who developed the original instrument in English. We believe that this communication is essential to encourage a more widespread use of the questionnaire and its translation into other languages. Here, we present the final version of the QOL-PCD in Portuguese for adults (Appendix 1, http://jornaldepneumologia.com.br/detalhe_anexo.asp?id=59). The use of this questionnaire is important for the appropriate clinical follow-up of patients with PCD. It also serves as an efficacy outcome measure in studies of therapeutic interventions and the natural history of the disease.

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