

APPENDIX 1

Outcome measures used

The D12 scale is an overall score for breathlessness severity that incorporates seven physical items and five affective items.[1] Participants complete the D-12 in reference to their experience of breathlessness “these days” at baseline and follow-up. D-12 consists of 12 descriptor items on a scale of none (0), mild (1), moderate (2), or severe (3). Total scores from the D-12 range from 0 to 36, with higher scores corresponding to greater severity. It has not been validated in ILD.

The King’s Brief Interstitial lung disease is a 15 item questionnaire consisting of three domains (breathlessness and activities, chest symptoms and psychological).[2] It has been validated in all ILD disease groups including IPF. A higher score indicates a higher quality of life (QoL). Scores range from 0-100 with the minimal important difference of 8 units of the total score.[3]

The St George’s Respiratory questionnaire [4] is a 50 item instrument designed to measure impact on overall health, daily life, and perceived wellbeing in patients with obstructive airways disease. Part 1 has a symptoms component (frequency and severity) with a 1, 3 or 12 month recall (several scales); Part 2 has a activities component looking at activities that cause or are limited by breathlessness and an impact component looking at social functioning, psychological disturbances resulting from airways disease and referring to current state as the recall (dichotomous (true/false) except last question (4 point Likert scale).[5] The MID for IPF in each of the SGRQ domains is Symptoms 8 units, Activity 5 units, -Impact 7 units and Total 7 units.[4] A lower score indicates a better quality of life. The generic SGRQ version has been validated in IPF.[4]

The Medical Research Council (MRC) dyspnoea scale (score range, 1-5, with higher scores indicating greater impairment) [6] is used to classify participants according to activity limitation. The MRC scale comprises five statements that describe almost the entire range of respiratory disability from none (Grade 1) to almost incapacity (Grade 5). It is self-administered by asking subjects to choose a phrase that best describes their condition. The MRC breathlessness scale does not quantify breathlessness itself. Rather, it quantifies the disability associated with breathlessness by identifying that breathlessness occurs when it should not (Grades 1 and 2) or by quantifying the associated exercise limitation (Grades 3–5). It has not been validated in ILD patients.

The 14-item Hospital Anxiety and Depression Scale (HADS) is a widely used tool for assessing psychological distress.[7] The HADS comprises seven items that tap anxiety (score range, 0-21) and seven items that tap depression (score range, 0-21), with higher scores corresponding to greater distress. Scores of 0-7 are classed as normal, 8-10 borderline abnormal and 11-21 abnormal. The HADs may be completed by both patient and informal caregiver. The HADs has not been validated in IPF. The MID in COPD is 1.5.[8]

The Carer Quality of Life Cancer (CQOLC) measures four conceptual domains of QoL: physical functioning, emotional functioning, family functioning and social functioning.[9] The CQOLC consists of 35 items that have a five-point Likert format that range from 0 (not at all), 1 (a little bit), 2 (somewhat), 3 (quite a bit) and 4 (very much): ten items relate to burden, seven to disruptiveness, seven to positive adaptation, three to financial concerns and eight single items to additional factors (disruption of sleep, satisfaction with sexual functioning, day-to-day focus, mental strain, informed about illness, protection of patient, management of patient's pain and family interest in caregiving). The CQOLC scale is scored by adding up the score on each item to yield a total score for the instrument and scores can range from 0-140. For all items and domains that measure QoL, a higher score represents a

better QoL.[9] There is no current tool to measure informal caregiver QoL in non-malignant respiratory disease. Therefore the CQOLC was used which has been validated in cancer patients.

The Zarit Burden Interview (ZBI) was developed to measure subjective burden among informal caregivers of adults with dementia.[10] Items were generated based on clinical experience with informal caregivers and prior studies resulting in a 22-item self-report inventory that examines burden associated with functional/behavioural impairments and the home care situation. The items are worded subjectively, focusing on the affective response of the informal caregiver.[11 12] Each question is scored on a 5 point Likert scale ranging from - never to nearly always present. Total scores range from 0 (low burden) to 88 (high burden). There is no validated tool to measure caregiver burden in ILD.

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