Cystic fibrosis: bench to bedside

LUNG CLEARANCE INDEX (LCI) AND FEV1 CORRELATE EQUALLY WITH TREATMENT BURDEN AS MEASURED BY CYSTIC FIBROSIS QUESTIONNAIRE-REVISED (CFQ-R)

doi:10.1136/thoraxjnl-2011-201054b.44

K O’Neill, J M Bradley, M Tunney, J S Elborn, Centre for Infection and Immunity, Queen’s University Belfast, Belfast, UK; Health and Rehabilitation Sciences Research Institute, University of Ulster, UK; School of Pharmacy, Queen’s University Belfast, Belfast, UK

Introduction LCI derived from multiple breath washout (MBW) measures the elimination of an inert marker gas during tidal breathing and is a sensitive measure of ventilation inhomogeneity in CF. LCI is more sensitive than FEV1 and FEF25\textendash}75 in detecting airways abnormalities and does not require a forced manoeuvre. The CFQ-R is a validated patient reported outcome used to assess health related quality of life (HRQoL) and patient perception of symptoms. There is a need to better understand the relationship between LCI, HRQoL and symptoms.

Objective To investigate the relationship between LCI, FEV1 % pred, HRQoL and symptoms as measured by the CFQ-R.

Methods These data are part of a larger study investigating the role of LCI as a tool to monitor lung function longitudinally. Patients were recruited from the adult and paediatric CF centres in Belfast Health and Social Care Trust. Inclusion criteria: clinical diagnosis of CF; clinically stable (exacerbation free = 4 weeks); informed consent. Age appropriate versions of the CFQ-R were used (patients >14 years, children aged 12 and 13, children aged 6–11). A parent questionnaire was completed in addition where appropriate (for children aged 6–13). The instrument yielded a score of 0–100 for each domain, with higher numbers indicating better function on various domains. Participants completed three MBW tests, using 0.2\% sulphur hexafluoride and a modified Innocor device. LCI was reported as the mean of at least 2 acceptable tests. Spirometry was performed to ATS/ERS standards.

Results Data were collected for 21 patients (15M:6F), age range 6–51 years, mean (SD) 26.4 (15.7). Mean (SD) FEV1 % pred was 77.1 (16.8). Mean (SD) LCI was 9.4 (2.5) (normal <7.5). LCI correlated negatively with FEV1 % pred (r = -0.62 p = 0.003). The domain of treatment burden was significantly correlated with LCI (r = -0.67 p = 0.001) and FEV1 % pred (r = -0.69 p = 0.001). However no correlation was observed with respiratory symptoms or any other domain of the CFQ-R.

Conclusion Patients with a greater treatment burden are more likely to have more severe lung disease. The severity of CF lung disease as determined by FEV1 % pred and LCI correlate equally with treatment burden. This further validates LCI as a useful measure of lung function.

REFERENCE


A COMPARISON OF THREE DIFFERENT SPECIMEN TYPES FOR THE DIAGNOSIS OF VIRAL RESPIRATORY INFECTIONS IN ADULTS WITH CYSTIC FIBROSIS

doi:10.1136/thoraxjnl-2011-201054b.46

W G Flight, K J Mutton, B Isakula, R J Bright-Thomas, A M Jones, Manchester Adult Cystic Fibrosis Centre, Manchester, UK; Department of Virology, Manchester Royal Infirmary, Manchester, UK

Introduction Respiratory viruses have been associated with increased symptoms and a decline in lung function in patients with cystic fibrosis (CF). The optimal means of diagnosis of respiratory viruses in CF is unclear. We compared the suitability of sputum,