

CORRESPONDENCE

Further evidence for an association between LCI and FEV₁ in patients with PCD

We have taken keen interest in the current discussion on the meaning of the lung clearance index (LCI) in patients with primary ciliary dyskinesia (PCD). In patients with cystic fibrosis (CF), LCI from the nitrogen multiple-breath gas washout (N₂-MBW) test has become a sensitive marker for detection of ventilation inhomogeneity with a good correlation between LCI and FEV₁.¹ However, in patients with PCD, two studies failed to find a correlation for the FEV₁ and LCI.^{1 2} This difference in functional measures between CF and PCD was recently challenged by Boon *et al*,³ showing a robust correlation between LCI and FEV₁ in patients with PCD. In the carefully written accompanying editorial, several factors were given to explain, among others these, differences between studies.⁴

Our aim is now to add further data to this discussion. We performed N₂-MBW measurements and spirometry in 47 patients with PCD at a mean (SD; range) age of 14.7 (6.9; 4.0–42.3) years, using an Exhalyzer D (EcoMedics, Duernten, Switzerland) and Jaeger MasterScreen body plethysmograph (CareFusion, Höchberg, Germany) on the same day. z-Scores were defined of a healthy reference population for FEV₁⁵ and from an in-house healthy population (n=46) measured with the same equipment for LCI. We found a good correlation between LCI and FEV₁ z-scores (coefficient: -2.95 , 95% CI -3.92 to -1.97 , $p < 0.001$, R^2 0.45, r -0.67 , see figure 1). Using absolute values adjusted for age, gender and height, the correlation between LCI and FEV₁ remained significant in a robust way (coefficient: -3.24 , 95% CI -4.51 to -1.97 , $p < 0.001$, R^2 0.49, r -0.70). In our cohort, the association was independent of age (coefficient: -2.88 for < 14 years and -2.70 for > 14 years) and disease severity (coefficient: -3.36 for those with FEV₁ < -1 z-score and -3.42 for those with FEV₁ > -1 z-score). This suggests that it is neither the use of z-scores

nor the different age or disease severity that leads to the discrepant findings of previous studies.

Taken together, we confirm results from Boon *et al* of a good correlation between LCI and FEV₁ in patients with PCD. This association was independent of age or disease severity. Besides different pieces of equipment and tracer gases used,⁴ another reason for discrepancies between studies^{1 3} might still be the different genetic background leading to distinct PCD phenotypes between regions and centres.

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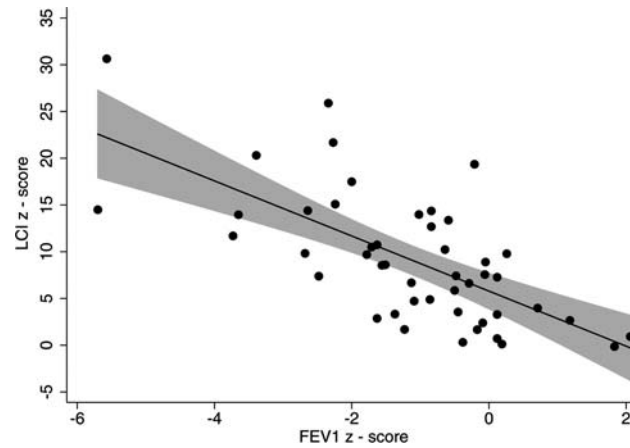


Figure 1 Association of lung clearance index (LCI) with FEV₁ in 47 patients with primary ciliary dyskinesia (PCD). LCI z-scores from the nitrogen multiple breath washout (N₂-MBW) versus FEV₁ z-scores in 47 patients with primary ciliary dyskinesia (PCD). Fitted values denote the black line. The grey background denotes the 95% CI.

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