

10.13 Lung Clearance Index (LCI) in adult patients with cystic fibrosis (CF) in the era of CFTR modulators

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Introduction. Lung clearance index (LCI) describes ventilation inhomogeneity¹. It is typically measured by the multiple breath washout (MBW) test, which has been developed to detect early changes in the peripheral airways². The long term effects of highly-effective CFTR modulators on pulmonary outcomes of patients with cystic fibrosis (CF) remain under investigation. The aim of this pilot study was to assess the efficacy of MBW in the adult CF patients at the University Hospital Limerick.

Materials and Methods. LCI measurements were performed using the ExhalyzerD, controlled with Spiroware 3.2.1 software. All spirometry tests were carried out using a handheld EasyOne Air spirometer equated with the Quanjer (GLI) 2012 prediction equation. All MBW tests were completed prior to spirometry testing and the study subjects were clinically stable on assessment day.

Results. To date, we collected data from 23 independent assessments (Table1). Twenty-one spirometry assessments resulted in a normal FEV₁ value (>80%). The *mean*±*SD* of LCI value for corresponding assessments was 7.83±1.10. Although we observed a nonlinear correlation between LCI and ppFEF_{25-75%} (Figure1), additional tests will be required to establish more accurate correlation.

Conclusion. The MBW has potential for integration into the standard care of adult CF patients with mild lung disease.

Keywords: multiple breath washout, lung clearance index, cystic fibrosis, CFTR modulator.

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Table 1. Characteristics and lung function parameters.

Parameter	median; [range]
Age [years]	21 ; [16 – 27]
Height [cm]	169 ; [150 – 189]
Weight [kg]	69.8 ; [50.4 – 91.7]
BMI [kg/m ²]	24.1 ; [19.2 – 28.9]
LCI _{2.5} [TO]	7.62 ; [6.53 – 10.57]
ppFEV ₁ [%]	96 ; [58 – 113]
ppFEF _{25-75%} [%]	84 ; [23 – 140]

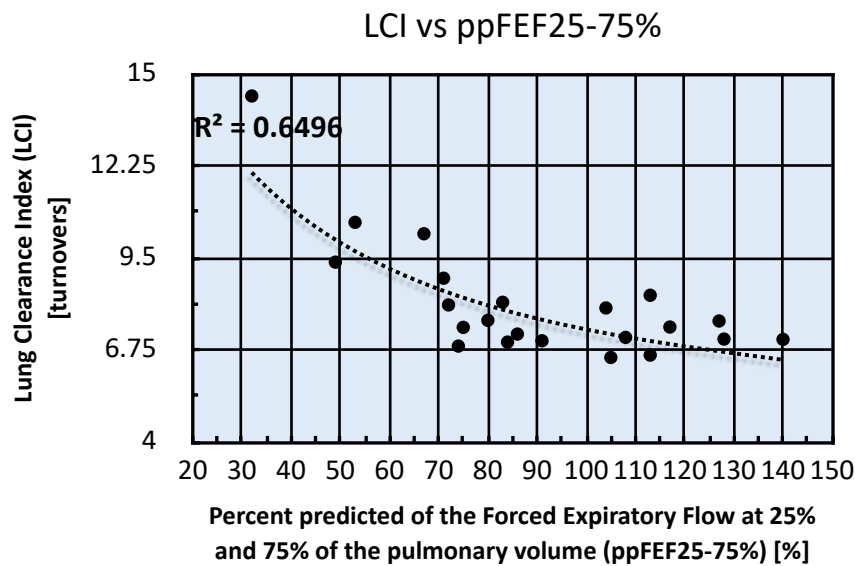


Figure 1. LCI vs ppFEF_{25-75%} in adults with CF (one test was excluded from this analysis).