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Title: Cystic fibrosis lung disease assessed by lung clearance index

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Body: **AIM:** To detect whether the lung clearance index (LCI) is a sensitive and repeatable noninvasive measure of airway infection among children and adolescents with CF. **METHODS:** Seventy-three children with CF (mean age, 10.6) and 51 healthy age- matched children underwent multiple-breath washout testing. LCI within and between-test variability was assessed. Children with CF had spirometry performed, 32 children underwent HRCT scan, which was scored with modified Bhala score and 30 children performed a cardiopulmonary exercise test on cycle ergometer (Ergoline, Vmax Series V.20-1, Sensor medics). **RESULTS:** The mean (SD) LCI in healthy children was 6.9 (1.1). The LCI was higher in children with CF (10.3 [3.9]; $P < 0.0001$). LCI measurements were repeatable and reproducible. Forty-seven (64.4%) children with CF were chronically colonized with *Pseudomonas aeruginosa*. The LCI was higher in children with *Pseudomonas* (11.9 [3.9]) than in children without *Pseudomonas* (7.3 [0.9]) ($P < 0.0001$). The LCI correlated with FEV₁ % predicted ($R(2) = 0.492$, $P = 0.004$). The LCI correlated with Bhalla score ($R(2) = 0.827$, $P < 0.0001$). The LCI correlated with Peak Aerobic Capacity (V'O_{peak}) % predicted ($R(2) = 0.121$, $P = 0.003$). **CONCLUSIONS:** The LCI is elevated among children and adolescents with CF, especially in the presence of *Pseudomonas*. The LCI is a feasible, repeatable, and sensitive noninvasive marker of lung disease in children with CF that correlates with structural and functional lung impairment.