Longitudinal course of clinical lung clearance index in children with cystic fibrosis

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The lung clearance index (LCI) is sensitive to assess lung disease progression in children with CF in routine clinical care. An increased change in LCI should prompt further diagnostic intervention to determine the underlying pathological process. https://bit.ly/3ae9Rhp

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Abstract

Background Although the lung clearance index (LCI) is a sensitive marker of small airway disease in individuals with cystic fibrosis (CF), less is known about longitudinal changes in LCI during routine clinical surveillance. Here, our objectives were to describe the longitudinal course of LCI in children with CF during routine clinical surveillance and assess influencing factors.

Methods Children with CF aged 3–18 years performed LCI measurements every 3 months as part of routine clinical care between 2011 and 2018. We recorded clinical data at every visit. We used a multilevel mixed effect model to determine changes in LCI over time and identify clinical factors that influence LCI course.

Results We collected LCI measurements from 1204 visits (3603 trials) in 78 participants, of which 907 visits had acceptable LCI data. The average unadjusted increase in LCI for the entire population was 0.29 (95% CI 0.20–0.38) LCI units·year^{-1}. The increase in LCI was more pronounced in adolescence (0.41 (95% CI 0.27–0.54) LCI units·year^{-1}). Colonisation with either *Pseudomonas aeruginosa* or *Aspergillus fumigatus*, pulmonary exacerbations, CF-related diabetes and bronchopulmonary aspergillosis were associated with a higher increase in LCI over time. Adjusting for clinical risk factors reduced the increase in LCI over time to 0.24 (95% CI 0.16–0.33) LCI units·year^{-1}.

Conclusions LCI measured during routine clinical surveillance is associated with underlying disease progression in children with CF. An increased change in LCI over time should prompt further diagnostic intervention.