

Changes in lung clearance index in children with cystic fibrosis – guidance for clinical practice

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Abstract

Rationale The lung clearance index (LCI) is increasingly being used in the clinical surveillance of patients with cystic fibrosis (CF). However, there are limited data on long-term variability and clinically relevant changes in LCI during routine clinical surveillance. **Objectives** To evaluate long-term variability of LCI and propose a threshold for a clinically relevant change. **Methods** Children with CF aged 4-18 years performed LCI measurements every three months as part of routine clinical surveillance during 2011-2020 in two centers. The variability of LCI during periods of clinical stability was assessed using mixed-effects models and was used to identify thresholds for clinically relevant changes. **Results** Repeated LCI measurements of acceptable quality (N= 858) were available in 100 patients with CF. Variability of repeated LCI measurements over time expressed as coefficient of variation (CV%) was 7.4%. The upper limit of normal (ULN) for relative changes in LCI between visits was 19%. **Conclusion** We report the variability of LCI in children and adolescents with CF during routine clinical surveillance. According to our data, a change in LCI beyond 19% may be considered clinically relevant. These findings will help guide clinical decisions according to LCI changes.

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