Determinants of lung disease progression measured by lung clearance index in children with cystic fibrosis

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Lung clearance index measured during preschool years is a major determinant of school age LCI. These findings further support that the preschool years are critical for early intervention strategies.
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Abstract

The lung clearance index (LCI) measured by the multiple breath washout (MBW) test is sensitive to early lung disease in children with cystic fibrosis. While LCI worsens during the preschool years in cystic fibrosis, there is limited evidence to clarify whether this continues during the early school age years, and whether the trajectory of disease progression as measured by LCI is modifiable.

A cohort of children (healthy and cystic fibrosis) previously studied for 12 months as preschoolers were followed during school age (5–10 years). LCI was measured every 3 months for a period of 24 months using the Exhalyzer D MBW nitrogen washout device. Linear mixed effects regression was used to model changes in LCI over time.

A total of 582 MBW measurements in 48 healthy subjects and 845 measurements in 64 cystic fibrosis subjects were available. The majority of children with cystic fibrosis had elevated LCI at the first preschool and first school age visits (57.8% (37 out of 64)), whereas all but six had normal forced expiratory volume in 1 s (FEV_1) values at the first school age visit. During school age years, the course of disease was stable (-0.02 units·year^{-1} (95% CI -0.14–0.10). LCI measured during preschool years, as well as the rate of LCI change during this time period, were important determinants of LCI and FEV_1, at school age.

Preschool LCI was a major determinant of school age LCI; these findings further support that the preschool years are critical for early intervention strategies.