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Sensitive markers to detect progression of lung disease in children with cystic fibrosis

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LCI detects progression of lung disease in children with CF. Preschool LCI is a strong predictor of lung disease in older children with CF. However, factors influencing progression remain diverse and are not comparable between different patient cohorts. https://bit.ly/2MCeJTW

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Cystic fibrosis (CF) is caused by mutations in the cystic fibrosis transmembrane conductance regulator (*CFTR*) gene with consecutive functional impairment of the CFTR anion channel in various epithelia, leading to reduced chloride secretion and increased viscosity of epithelial secretions with impaired mucociliary clearance in the airways [1]. This inherited basic defect is already present before birth, leading to organ manifestations such as exocrine pancreatic insufficiency in ∼85% or meconium ileus in ∼10% at birth [2, 3]. In contrast to these gastrointestinal manifestations, the lungs are structurally normal at birth [4]. Today, the majority of patients die in young adulthood due to chronic CF lung disease, which starts in the first months of life. Early onset of CF lung disease was demonstrated by bronchoalveolar lavage and chest computed tomography studies showing neutrophilic inflammation and structural changes in infants diagnosed following newborn screening (NBS) at the age of ∼4 months [5]. However, less invasive end-points are desirable for studies on the natural history of early disease progression, and as clinical study end-points and diagnostic tools. A better understanding of the onset and early progression of CF lung disease is essential, as it has recently been shown that early inflammation determines the extent of bronchiectasis at the age of 5 years [6]. Improved understanding and close follow-up of early CF lung disease could contribute to the development of new therapies to prevent and/or delay irreversible lung damage [7].

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