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**Title:** Structural lung disease, pulmonary infection and inflammation in infancy is associated with reduced spirometry at school-age in children with cystic fibrosis (CF)

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**Body:** Introduction: Pulmonary inflammation, infection and structural lung disease occur early in life in children with CF leading to lung function decline. We aimed to determine if these markers of CF lung disease in infancy altered spirometry outcomes at school-age. Methods: Infants (0-2 years) with CF diagnosed following newborn screening had annual bronchoalveolar lavage fluid collected to assess infection and inflammation and computed tomography (CT) scans to assess structural lung disease. Ninety-four children were followed up with lung function (FEV0.5) at 6-8 years of age and 59 children had lung function measured at both infancy and school-age. Results: Children with CF who were pancreatic insufficient had 9.6% (n=80) lower FEV0.5 compared with sufficient children (n=14; p = 0.03). The presence of air trapping on CT in infancy was associated with 14.5% lower FEV0.5 at school-age (p = 0.03). In children with longitudinal measurements of lung function infection with *P. aeruginosa* or *H. influenzae* in infancy was associated with a lower FEV0.5 (15.8% and 18.2%; respectively) compared with never infected (p = 0.04). The presence of free neutrophil elastase (NE) during infancy (n=23) was associated with a 13.5% reduction in FEV0.5 compared with CF children without NE (n=36; p = 0.01). Conclusions: In children with CF the presence of air trapping on CT, lower airway infection and free NE during infancy was associated with clinically-significant reductions in lung function in the early school years, highlighting the need for targeted interventions during infancy.