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Structural determinants of long-term functional outcomes in young children with cystic fibrosis

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Structural lung changes identified on a chest CT scan in children with cystic fibrosis under 6 years of age can identify those at risk of adverse long-term outcomes <http://bit.ly/39QH5jv>

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ABSTRACT

Background: Accelerated lung function decline in individuals with cystic fibrosis (CF) starts in adolescence with respiratory complications being the most common cause of death in later life. Factors contributing to lung function decline are not well understood, in particular its relationship with structural lung disease in early childhood. Detection and management of structural lung disease could be an important step in improving outcomes in CF patients.

Methods: Annual chest computed tomography (CT) scans were available from 2005 to 2016 as a part of the AREST CF cohort for children aged 3 months to 6 years. Annual spirometry measurements were available for 89.77% of the cohort (167 children aged 5–6 years) from age 5 to 15 years through outpatient clinics at Perth Children's Hospital (Perth, Australia) and The Royal Children's Hospital in Melbourne (Melbourne, Australia) (697 measurements, mean±SD age 9.3±2.1 years).

Results: Children with a total CT score above the median at age 5–6 years were more likely to have abnormal forced expiratory volume in 1 s (FEV₁) (adjusted hazard ratio 2.67 (1.06–6.72), p=0.037) during the next 10 years compared to those below the median chest CT score. The extent of all structural abnormalities except bronchial wall thickening were associated with lower FEV₁ Z-scores. Mucus plugging and trapped air were the most predictive sub-score (adjusted mean change –0.17 (–0.26 – –0.07) p<0.001 and –0.09 (–0.14 – –0.04) p<0.001, respectively).

Discussion: Chest CT identifies children at an early age who have adverse long-term outcomes. The prevention of structural lung damage should be a goal of early intervention and can be usefully assessed with chest CT. In an era of therapeutics that might alter disease trajectories, chest CT could provide an early readout of likely long-term success.