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Early markers of cystic fibrosis structural lung disease: follow-up of the ACFBAL cohort

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In children with cystic fibrosis, airways disease severity on chest computed tomography at age 5 years increased the risk of bronchiectasis in adolescence and its extent was predicted by poorer nutrition, airway inflammation, and atelectasis <http://bit.ly/2Nnk8LW>

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ABSTRACT Little is known about early predictors of later cystic fibrosis (CF) structural lung disease. This study examined early predictors of progressive structural lung abnormalities in children who completed the Australasian CF Bronchoalveolar Lavage (ACFBAL) clinical trial at age 5-years and participated in an observational follow-up study (CF-FAB).

Eight Australian and New Zealand CF centres participated in CF-FAB and provided follow-up chest computed-tomography (CT) scans for children who had completed the ACFBAL study with baseline scans at age 5-years. CT scans were annotated using PRAGMA-CF scoring. Ordinal regression analysis and linear regression were used to investigate associations between PRAGMA-CF (Perth–Rotterdam Annotated Grid Morphometric Analysis for CF) outcomes at follow-up and variables measured during the ACFBAL study.

99 out of 157 ACFBAL children (mean±SD age 13±1.5 years) participated in the CF-FAB study. The probability of bronchiectasis at follow-up increased with airway disease severity on the baseline CT scan. In multiple regression (retaining factors at $p < 0.05$) the extent of bronchiectasis at follow-up was associated

with baseline atelectasis (OR 7.2, 95% CI 2.4–22; $p \leq 0.001$), bronchoalveolar lavage (BAL) \log_2 interleukin (IL)-8 (OR 1.2, 95% CI 1.05–1.5; $p=0.010$) and body mass index z-score (OR 0.49, 95% CI 0.24–1.00; $p=0.05$) at age 5 years. Percentage trapped air at follow-up was associated with BAL \log_2 IL-8 (coefficient 1.3, 95% CI 0.57–2.1; $p<0.001$) at age 5 years.

The extent of airway disease, atelectasis, airway inflammation and poor nutritional status in early childhood are risk factors for progressive structural lung disease in adolescence.