A screening tool to identify risk for bronchiectasis progression in children with cystic fibrosis

Daan Caudri[1], Lidija Turkovic[2], Nicholas de Klerk[2], Tim Rosenow[2], Conor Murray[1], Ewout Steyerberg[3], Sarath Ranganathan[4], Peter Sly[5], Stephen Stick[1], and Oded Breuer[2]

[1]Princess Margaret Hospital for Children
[2]Telethon Kids Institute
[3]Erasmus MC
[4]Royal Children's Hospital, Melbourne
[5]University of Queensland

June 2, 2021

Abstract

Background: The marked heterogeneity in CF disease complicates selection of those most likely to benefit from existing or emergent treatments. Objective: We aimed to predict progression of bronchiectasis in preschool children with CF. Methods: Using data collected up to three years of age, in the Australian Respiratory Early Surveillance Team for CF (AREST CF) cohort study, clinical information, chest computed tomography (CT) scores and biomarkers from bronchoalveolar lavage were assessed in a multivariable linear regression model as predictors for CT bronchiectasis at age 5-6. Results: Follow-up at 5-6 years was available in 171 children. Bronchiectasis prevalence at 5-6 was 134/171 (78%) and median bronchiectasis score 3 (range 0-12). The internally validated multivariate model retained eight independent predictors accounting for 37% (Adjusted R2) of the variance in bronchiectasis score. The strongest predictors of future bronchiectasis were: pancreatic insufficiency, repeated intravenous treatment courses, recurrent lower respiratory infections in the first 3 years of life and lower airway inflammation. Dichotomizing the resulting prediction score at a bronchiectasis score of above the median resulted in a diagnostic odds ratio of 13 (95% CI 6.3-27) with a positive and negative predictive values of 80% (95%CI 72%-86%) and 77% (95% CI 69%-83%) respectively. Conclusion: Early assessment of bronchiectasis risk in children with CF is feasible with reasonable precision at a group level, which can assist in high-risk patient selection for interventional trials. The unexplained variability in disease progression at individual patient level remains high, limiting the use of this model as a clinical prediction tool.

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