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Title: Lung function decline in a modern cystic fibrosis cohort

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Body: Introduction: Though the starting point for lung function measured by spirometry in children with cystic fibrosis (CF) has improved, the annual rate of decline has not changed significantly during the critical period of adolescence. The aim of this study was to describe factors associated with longitudinal decline in lung function in a contemporary cohort of children with CF. Methods: Best annual lung function data from children attending the CF service of the Royal Children's Hospital Melbourne were reviewed to determine rate of decline in FEV₁ up until time of transfer to an adult centre. Mixed multi-level modeling was used to determine the influence of age, sex, genotype (homozygous F508del), CF related diabetes mellitus (CFRD), Pseudomonas aeruginosa (PsA) infection, and body mass index (BMI) on lung function decline. Results: Longitudinal lung function data (range 5–20 years) were obtained for 98 patients with CF (55 male) on a median 12 (range 3–16) occasions. Overall, FEV₁ declined by a mean of -0.12 z-score each year (p<0.001). Homozygous F508del genotype (-0.10, p<0.01), CFRD (-0.15, p<0.01) and mucoid PsA infection (-0.09, p<0.01) were all independently associated with an increased rate of decline in FEV₁ z-score. Taken together, these three factors resulted in a cumulative reduction of -0.25 FEV₁ z-score (p<0.001). Conclusion: Genotype, CFRD and PsA infection are all associated with an increased rate of decline in lung function during adolescence. How these findings relate to underlying lung structural changes, and whether PsA eradication success can influence rate of decline in future cohorts, warrants further investigation.