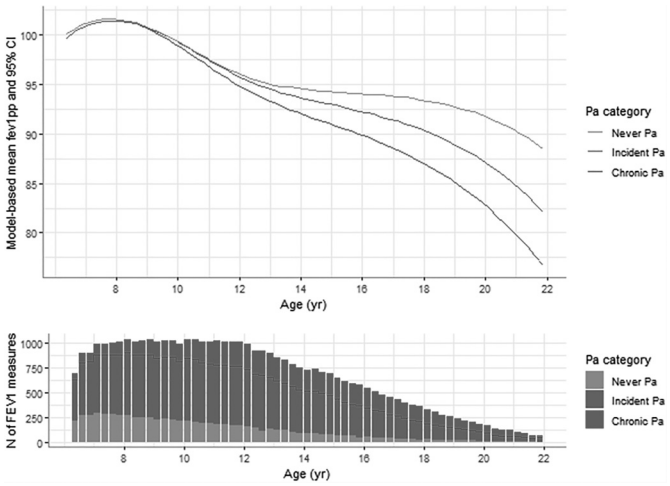

Figure 1. The top panel shows the model estimates of mean percentage predicted forced expiratory volume in 1 second (FEV_1pp) according to age and *Pseudomonas aeruginosa* stage for the reference value of all covariates. The bottom panel shows the number of FEV_1 values available at each age for each *P. aeruginosa* stage

Conclusions: Chronic *P. aeruginosa* infection is associated with lower FEV_1pp and more-rapid decline, particularly after age 12, in children with CF, even after adjustment for relevant covariates and regardless of chronic *P. aeruginosa* definition. Measures to prevent chronic *P. aeruginosa* infection could slow FEV_1 decline and improve long-term outcomes.

Acknowledgements: This work was supported by Cystic Fibrosis Foundation OBSERV13K0 and CUTTIN18XX1, BAMSHA18XX0, and KNOWLE18XX0. We thank all the people with CF who participated in the Early Pseudomonas Infection Control Observational Study and all the site primary investigators and research coordinators.

Reference

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Cystic fibrosis–specific reference equations for lung function adjusted for mortality attrition for people with cystic fibrosis in the United States

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Background: Percentage predicted forced expiratory volume in 1 second (FEV_1pp) has been widely used as the primary marker of lung health in a variety of pulmonary diseases, but it compares people with cystic fibrosis (PwCF) with healthy people without CF. In clinical care, patients and providers may benefit from generally understanding the health of a person with CF relative to that of other PwCF of similar age, sex, height, and CF transmembrane conductance regulator *(CFTR)* mutations. Using U.S. CFF Patient Registry (CFFPR) data from 1994 to 2001, reference equations of CF-specific FEV_1 percentiles have been calculated for U.S. CF populations [1]. Data from the Canadian CF Data Registry (CFDR) as of 2002 allowed modified CF-specific FEV_1 percentiles according to mortality to afford comparison of an individual with CF with all PwCF (alive and deceased) of the same birth cohort [2,3]. There has been an unmet need to update these health measures using recent lung health and mortality data from the U.S. CF population.

Methods: Based on recent U.S. CF registry data (CFFPR 2010–2017), we built reference equations generating CF-specific FEV_1 percentiles for the U.S. population using quantile regression. Yearly mortality for every 2-year birth cohort (1950–2010) was estimated based on the Kaplan-Meier survival curve or modified crude estimates. Rshiny software was used to develop a user-friendly online application to provide CF-specific reference equations for lung function with or without adjustment for mortality attrition for PwCF in the United States.

Results: The estimated U.S. CF-specific FEV_1 percentiles showed similar trends as previous estimates. Women and girls had lower scores than men and boys (Figure 1). Women and girls born between 1960 and 2000 also had lower survivor probability in recent years but not those born before 1960 or after 2000. The Rshiny application provides a tool that can automatically generate U.S. CF-specific FEV_1 percentiles with or without modification according to mortality, as well as previous versions of these measures.

Conclusions: These analyses yield a user-friendly online resource that may benefit patients, clinicians, and CF researchers.

Acknowledgements: This work was supported by the Cystic Fibrosis Foundation Statistical Expertise and Network (StatNet) Award (POLINE18Y7).

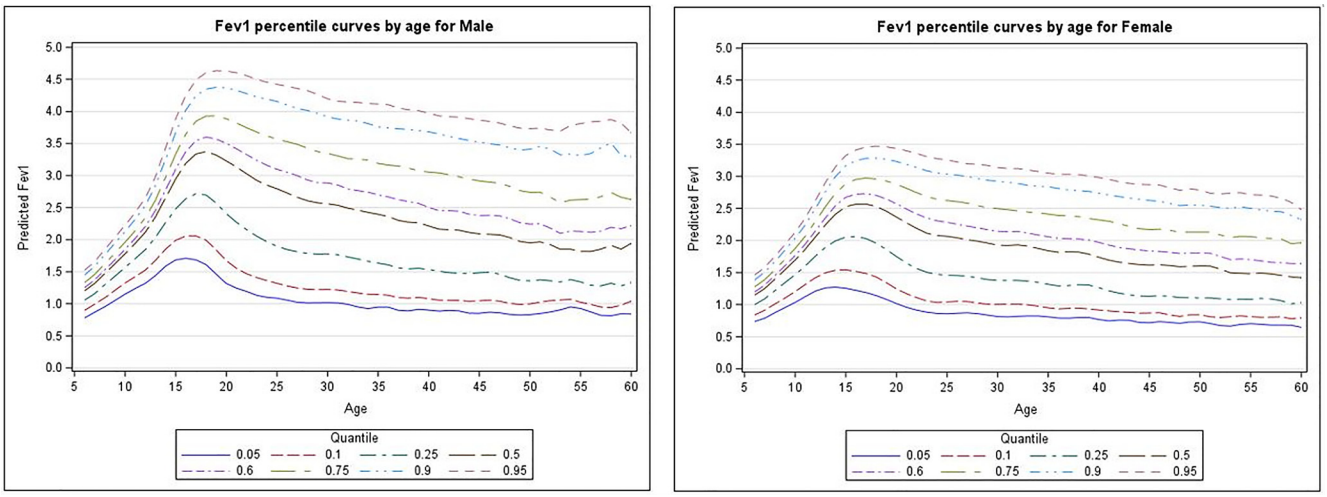

Figure 1 (abstract 42): Estimated reference equations for lung function for individuals with cystic fibrosis in the United States according to sex

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Cystic fibrosis transmembrane conductance regulator modulator–induced sweat chloride changes in the cystic fibrosis population from the Characterizing Cystic Fibrosis Transmembrane Conductance Regulator–Modulated Changes in Sweat Chloride Study: 2022 Update

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Background: We previously reported on the preliminary results from the Characterizing Cystic Fibrosis Transmembrane Conductance Regulator–Modulated Changes in Sweat Chloride (CHEC-SC) study [1], a large population-based epidemiological study characterizing the heterogeneity in sweat chloride response to cystic fibrosis (CF) transmembrane conductance regulator (CFTR) modulators and evaluating the association between sweat chloride and long-term clinical outcomes. With Food and Drug Administration approval of elexacaftor/tezacaftor/ivacaftor (ELE/TEZ/

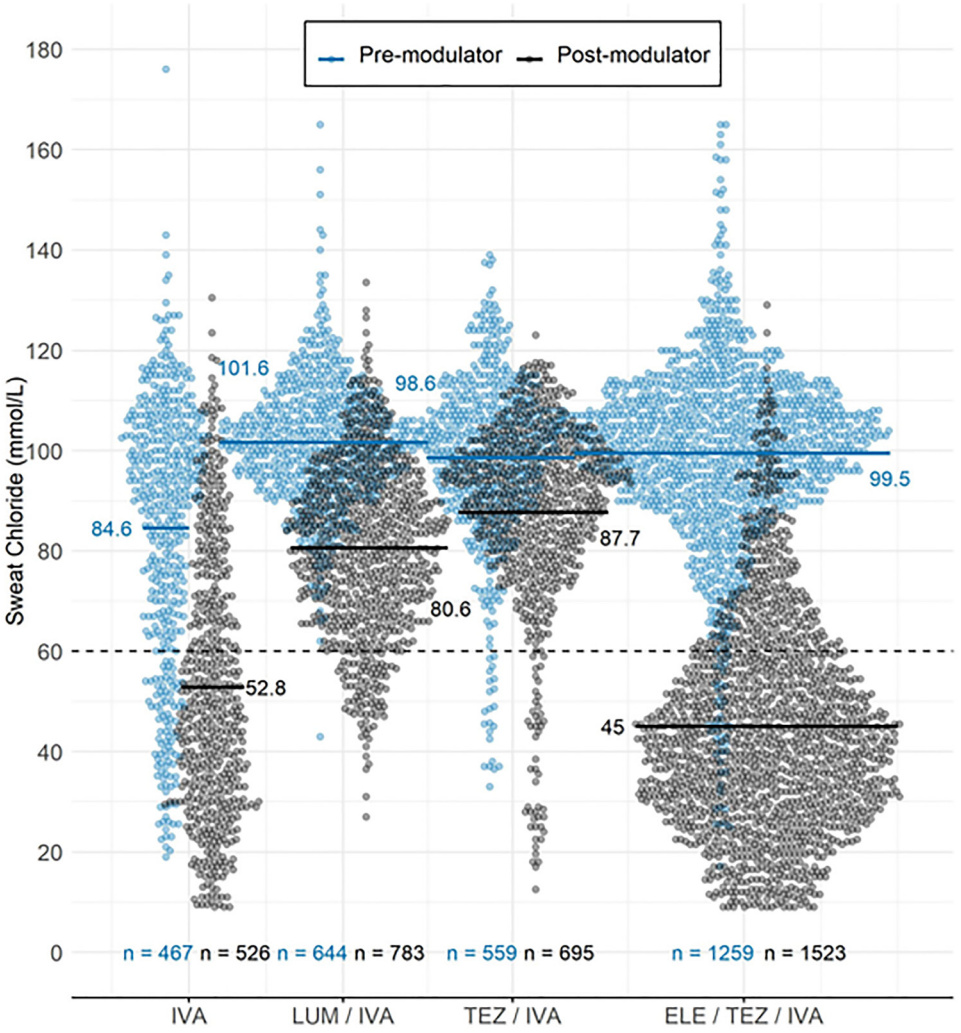

Figure 1 (abstract 43): Pre- and postmodulator sweat chloride in the Characterizing Cystic Fibrosis Transmembrane Conductance Regulator–Modulated Changes in Sweat Chloride study population. For each modulator (ivacaftor (IVA), lumacaftor/ivacaftor (LUM/IVA), tezacaftor/ivacaftor (TEZ/IVA), elexacaftor/tezacaftor/ivacaftor (ELE/TEZ/IVA)), pre- (blue circles) and postmodulator values (black circles) are shown, with corresponding means indicated by horizontal bars with numerical labels. Sample sizes are indicated below each set of modulator and time point pair.