cases. There is variability among SCCs, including rate of follow-up and repeat sweat Cl^- testing. Baseline data on CFSPID/CRMS infants in the large racially and ethnically diverse population of NYS indicate the need for ongoing evaluation to determine the conversion rate to CF.

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Cystic fibrosis in the United States by 2040: A population landscape analysis

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Background: The network of Cystic Fibrosis Foundation–accredited care programs partner with people with cystic fibrosis (CF) and their families to provide specialized disease management using a multidisciplinary approach. There are 128 pediatric, 119 adult, and 40 affiliate CF programs in the United States. In the past decade, the introduction of highly effective modulator therapies (HEMT) ivacaftor (IVA) and elexacaftor/tezacaftor/ivacaftor, in combination with other advances in CF care, has increased survival for persons living with CF, with approximately 57% of people with CF being aged 18 and older. In 2020, estimated median predicted survival was 50 for individuals born between 2016 and 2020, compared with 30 in the early 1990s. Given the potential for slower lung function decline with HEMT, it is likely that survival will increase, requiring CF care programs to serve a growing adult population. The objective of this analysis was to forecast the CF population in the United States through 2040 and predict the distribution of lung function and first transplants to inform planning for the future of CF care.
Methods: We implemented a simulation to predict the size and characteristics of the CF population from 2021 to 2040, simulating percentage predicted forced expiratory volume in 1 second (FEV_1pp), lung transplantation, and survival status. We used data from the Cystic Fibrosis Foundation Patient Registry to characterize the distribution of persons with CF according to age, sex, race, ethnicity, HEMT eligibility, and prescription status. We applied those distributions to a simulated 2020 prevalent population and projected CF births to 2040. The microsimulation used estimates of the probability an individual would transition between lung function categories defined according to FEV_1pp (high: >90%; moderate: 69–90%; low: 40–70%; severe: <40%), undergo lung transplant, or die derived from logistic regression models using longitudinal data for 2010 to 2020, with transition probabilities dependent on HEMT prescription status.
Results: By 2040, we estimate that adults with CF will comprise more than 70% of the total CF population. The proportion of adults classified as greater than 90% FEV_1pp will range from 35% to 40% over this period, with nearly 80% of children with FEV_1pp at this level or higher. Age at first transplant will increase over the next 20 years, with more than half of first transplants occurring in individuals aged 35 and older, compared with 37% reported in 2020.
Conclusions: Although CF has historically been characterized as a pediatric disease, we predict continued growth of the adult CF population over the next 20 years because of longer survival and a decline in CF births. Because of slower decline in lung function, age at time of lung transplant will increase, resulting in an older post-lung transplant population. It is likely that this growing, aging adult CF population will be more medically complex because of extra-pulmonary and age-related comorbidities, so coordination of care between CF care teams, subspecialists, and transplant centers will remain critical. The need to expand the workforce knowledgeable in the nuances of multidisciplinary adult CF care is more important than ever.
Acknowledgements: The authors would like to thank the Cystic Fibrosis Foundation for the use of their Patient Registry data to conduct this study. We would also like to thank the patients, care providers, and clinic coordinators at CF centers throughout the United States for their contributions to the Patient Registry.

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Effect of pregnancy on lung function in women with cystic fibrosis in the United States and United Kingdom: A registry-based study, 2003–2017

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Background: Women with cystic fibrosis (CF) are living longer, healthier lives and increasingly having children [1]. Lung function is known to decrease with age, but little is known about how pregnancy affects lung function decline. To address this, we assessed the impact of pregnancy on lung function trajectory in women with CF in the United States and United Kingdom.
Methods: This was a longitudinal registry study of 11,366 women aged 15 to 44 with CF contributing 331,724 lung function measures between 2003 and 2017 in the United States and 3,433 women contributing 24,394 measures in the United Kingdom over the same period. Pregnancy was recorded as a binary event in annual review records. We used previously developed mixed-effects models (random intercept and slope) to assess whether rate of decline of lung function changed after pregnancy, adjusting for clinically important covariates including age at diagnosis, genotype, and birth cohort [2,3].
Results: A total of 1,637 first pregnancies were recorded in the United States and 596 in the United Kingdom, with most occurring in the pre-modulator era (before 2013: United States, 65%, United Kingdom, 60%). Baseline clinical and demographic characteristics were similar for both population groups. After a record of pregnancy, the overall rate of lung function decline increased by 0.31 percentage points (95% CI, 0.23–0.39 percentage points) in percentage of forced expiratory volume in 1 second per year in the United States and 0.37 percentage points (95% CI, 0.18–0.56 percentage points) in the United Kingdom.
Conclusions: Our preliminary analysis suggests that pregnancy may be associated with greater decline of lung function of approximately one-third of a percentage point per year in the United States and the United Kingdom. Further robustness checks using alternative model specifications are required, along with studies in cohorts of women who have benefited from modulator therapy.
Acknowledgements: We acknowledge funding from the Research for Patient and Public Benefit Wales.

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