BMI improved from 0.01 kg/m^2 (95% CI, −0.07–0.09 kg/m^2) to 0.33 kg/m^2 (95% CI, 0.23–0.43 kg/m^2). There was no change in BMI z-scores. Number of hospitalizations decreased from 2,656 to 1,957 ($p < 0.01$). Virtual outpatient consultations increased from 8% of toal outpatient consultations before pandemic onset to 47% during the pandemic. There was an increase in average number of consultations per patient from a median of 4 (interquartile range 2–5) to 5 (interquartile range 3–6) ($p < 0.01$).

Conclusions: In the 12 months after the onset of the COVID-19 pandemic, improvement was observed in the clinical outcomes of people with CF and the model of care delivery changed from the pre-pandemic period. Health care teams must consider how best to deliver care in light of improved outcomes observed during the COVID-19 pandemic.

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49

Medication adherence and lung function changes with elexacaftor/tezacaftor/ivacaftor treatment

C. Esther[1,2], R. De Vuyst[1], C. Kam[3], C. McKinzie[3]. *[1]Pediatric Pulmonology, University of North Carolina, Chapel Hill, NC; [2]Marsico Lung Institute, School of Medicine, University of North Carolina, Chapel Hill, NC; [3]Pharmacy, University of North Carolina Medical Center, Chapel Hill, NC*

Background: Introduction of triple combination therapy (TCT) with elexacaftor/tezacaftor/ivacaftor improves clinical measures in many people with cystic fibrosis (PwCF). Anecdotally, many PwCF respond to improved clinical status by reducing adherence to routine CF care, including inhaled medications, but the extent to which this occurs and clinical consequences of reduced adherence are unknown.

Methods: To address this question, we identified all PwCF undergoing CFTR modulator therapy from our pharmacy database. Medication adherence measured according to medication possession ratio (actual months filled/prescribed months) for modulators and inhaled medications hypertonic saline (HTS) and dornase alpha were assessed and compared with clinical outcome measures.

Results: Three hundred eleven PwCF had three or more CFTR modulator prescriptions filled from November 2016 to August 2021, of whom 235 had three or more fills for TCT. Medication adherence was high across all modulators (0.99 ± 0.10 for ivacaftor alone, 0.98 ± 0.09 for lumacaftor/ivacaftor, 0.97 ± 0.11 for tezacaftor/ivacaftor, 1.02 ± 0.20 for TCT). Lung function values were available for 156 PwCF before and after TCT initiation, demonstrating the expected increase in percentage predicted forced expiratory volume in 1 minute (FEV_1pp) values with treatment (76.2 ± 24.6 vs 84.1 ± 24.8, $p < 0.003$), although rate of change in FEV_1 per year remained negative after modulator therapy, with a trend toward a more-negative slope after treatment initiation (−0.7 ± 5.1 vs −1.5 ± 6.1, $p = 0.15$). Adherence to HTS and dornase also declined after TCT initiation within the group as a whole (negative slope in medication possession ratio with time) and was most evident in those who regularly filled these prescriptions through our pharmacy (n = 46 and n = 37, respectively), in whom adherence was greater than 80% and steady over time before TCT but fell to 69% and 73%, respectively, with a negative slope over time after TCT (Figure 1). Adherence to pancreatic enzymes (n = 52) followed a similar pattern, although changes in adherence to HTS or dornase after modulator therapy were not associated with lung function or changes in lung function over time.

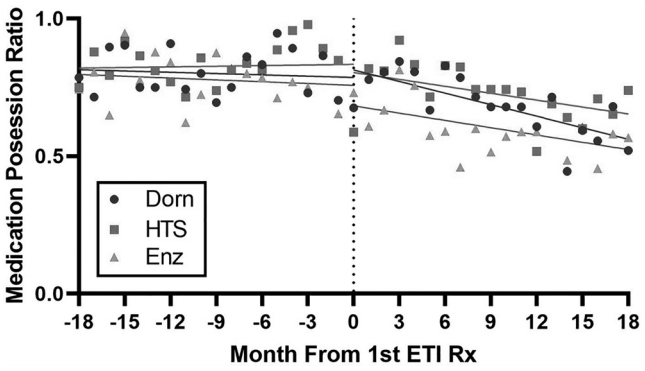

Figure 1. Medication possession ratio for dornase (Dorn), hypertonic saline (HTS), and pancreatic enzymes (Enz) relative to time before and after first prescription for elexacaftor/tezacaftor/ivacaftor (ETI)

Conclusions: In this real-world study, PwCF undergoing TCT were strongly adherent to modulator therapy, with substantial gains in lung function but continued decline in lung function over time. Although continued lung function decline on TCT could reflect the observed decrease in adherence to inhaled CF therapies, we did not observe a direct correlation between lung function and adherence. Further study will be needed to determine whether declines in lung function after TCT are sustained or reflect short-term changes in approach to CF care with highly effective modulator treatment.

50

Multi-center cohort study exploring the impact of the first U.K. COVID-19 lockdown on nebulizer use in adults with cystic fibrosis in the CFHealthHub learning health system

L. Lai[1], R. Sandler[2], S. Dawson[3], Z. Hoo[2], T. Daniels[4,5], M. Hutchings[2], M. Sperrin[1], M. Wildman[2,6], C. Echevarria[7]. *[1]Division of Informatics, Imaging and Data Sciences, University of Manchester, Manchester, UK; [2]Sheffield Adult Cystic Fibrosis Centre, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK; [3]Wolfson Adult Cystic Fibrosis Centre, Nottingham University Hospitals NHS Trust, Nottingham, UK; [4]York Hull Adult CF Centre, York and Scarborough Teaching Hospitals NHS Foundation Trust, UK; [5]York St John University, York, UK; [6]School of Health and Related Research, University of Sheffield, Sheffield, UK; [7]The Newcastle upon Tyne Hospitals NHS Foundation Trust, Translational and Clinical Research Institute, Newcastle University, Newcastle on Tyne, UK*

Background: Promoting adherence to preventative inhaled therapy in cystic fibrosis (CF) is challenging despite a clear association with better outcomes. Motivation to maintain adherence is one aspect of the complex process required for sustained behavior change. Fear is recognized as an acute motivator, and the COVID-19 pandemic created a significant fear stimulus, especially in people with underlying respiratory disease. The objective of this study was to explore changes in nebulizer use in response to the COVID-19 pandemic.

Methods: This 16-center cohort study involved more than 60% of adult CF centers in England through the CFHealthHub learning health system. CFHealthHub was established in 2015 to help promote adherence to nebulized therapy. It provides people with CF and their clinicians objective usage information through nebulizers with real-time electronic data capture capability. We explored the association between the first U.K. national lockdown (March 23, 2020, to May 28, 2020) and nebulizer use, using the mixed-effect multilevel model adjusted for potential confounders including age (fitted as a nonlinear term), sex, and deprivation index.

Results: We included 581 adults with CF participating in CFHealthHub between September 1, 2019, and August 21, 2020. Mean age was 34; 49% participants were female. The range of mean weekly nebulizations per patient increased from 9 to 12 to 12 to 14 in the week after the first U.K. national lockdown before decreasing to 11 to 12 throughout the rest of the study period (Figure 1). The association between lockdown and nebulizer