ORIGINAL RESEARCH

Changes in Care during the COVID-19 Pandemic for People with Cystic Fibrosis

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Abstract

Rationale: Cystic fibrosis (CF) centers transitioned to telemedicine during the spring 2020 peak of the coronavirus disease (COVID-19) pandemic.

Objectives: We hypothesized that people with CF (pwCF) with more severe disease would be more likely to be seen in-person.

Methods: We used paired t tests to compare within-subject changes in body mass index (BMI) and percentage predicted forced expiratory volume in one second (FEV_1) and calculated relative risk (RR) to compare pulmonary exacerbations (PEx) between pwCF enrolled in the CF Foundation Patient Registry with at least one in-person clinic visit after March 15 in both 2019 and 2020.

Results: Overall, the proportion of clinical encounters that were in-person clinic visits decreased from 91% in 2019 to a low of 9% in April 2020. Among pwCF seen after March 15 in both 2019 and 2020, the mean (95% confidence interval [CI]) FEV_1 percentage predicted was 1.3% (0.1–2.4) predicted higher in 2020 for children 6 to <12 years of age, and 7.5% (7.1–7.9) predicted higher in 2020 among pwCF ≥12 years of age eligible for the highly effective CF transmembrane conductance regulator modulator, elexacaftor-tezacaftor-ivacaftor (ETI). There was no difference in FEV_1 percentage predicted for pwCF ≥12 years of age who were not eligible for ETI. Similarly, the mean (95% CI) BMI was 2.4 (2.0–2.8) percentile higher in 2020 for children 6 to <12 years of age and 5.2 (4.8–5.7) percentile higher in 2020 among children 12 to <18 years of age eligible for ETI. Mean (95% CI) BMI was 1.2 (1.2–1.3) (kg/m^2) higher for pwCF ≥18 years of age eligible for ETI, and 0.2 (0.1–0.3) (kg/m^2) higher for pwCF ≥18 years of age not eligible for ETI. The proportion of in-person clinic visits where any PEx was present was lower in 2020 compared with 2019, 25% compared with 38%, RR 0.82 (0.79–0.86).

Conclusions: The care of pwCF was substantially changed during the spring 2020 peak of the COVID-19 pandemic. Among pwCF seen in-person in both 2019 and 2020 after the spring peak of the COVID-19 pandemic, lung function and BMI were higher in 2020 for children 6 to <12 years of age and pwCF eligible for ETI.

Keywords: ambulatory care; pulmonary function; body mass index; pulmonary exacerbation

The coronavirus disease (COVID-19) pandemic resulted in widespread restrictions on social interactions and lockdowns in many areas of the world (1). These restrictions were associated with reductions in clinical encounters in many fields, including hospitalizations and emergency room visits for exacerbations of chronic diseases, acute medical events, and injuries (2–4). The reasons for these reductions are not entirely clear but may represent a desire to avoid the risk of exposure to infection, lost health insurance, increased thresholds for hospitalizations, or improvement in self-care during social distancing (3, 5). Similarly, reductions in outpatient clinic visits occurred in the United States during the initial peak of the pandemic in spring 2020, and at least some proportion of these in-person visits were replaced with encounters via telemedicine (6, 7).

The U.S. Cystic Fibrosis (CF) Foundation guidelines recommend that people with CF (pwCF) be seen in-person at an accredited CF center at least every 3 months after the first year of life (8–10). Mirroring the pattern of outpatient care in the United States at the start of the COVID-19 pandemic, CF centers rapidly transitioned to telemedicine during the spring 2020 peak of the pandemic. We used the CF Foundation Patient Registry (CFFPR) to describe the proportion of clinical encounters that were in-person, the recording of clinical information and interactions with members of the multidisciplinary CF team, and the characteristics and pulmonary exacerbations of pwCF beginning with the spring 2020 peak of the COVID-19 pandemic, compared with the same period in 2019. Our hypothesis was that pwCF seen in-person would have markers of more severe disease, including lower pulmonary function, lower body mass index (BMI), and be more likely to experience a pulmonary exacerbation (PEx).

Methods

Objective and Design

The objective of this report is to describe the characteristics of pwCF in the United States seen in-person during and after the initial peak of the pandemic in spring 2020, in comparison to 2019. In addition, we describe changes in care patterns that occurred throughout the last 9 months of 2020 in comparison to 2019. To address these objectives, we performed a retrospective study of clinical encounters in the CFFPR. The CFFPR contains demographic and clinical data collected at every encounter at CF Foundation (CFF)-accredited centers using a standardized data collection form (11). Encounter location was recorded as in clinic, in the hospital, or other. Encounters that occurred by phone or with video by phone or computer were added to the CFFPR during the spring peak of the pandemic, but given the variability in the timing of changes in data entry, we limited our analysis to data for in-person clinic visits and PEx treated with intravenous antibiotics in the hospital or at home. Clinical information includes anthropomorphic measurements, pulmonary function test results, microbiology results from respiratory secretions, treatments and medications, complications of CF, and whether a PEx was determined to be present at in-person clinic visits.

Study Population

People with CF are eligible to be included in the CFFPR if they receive care at one of the CFF's accredited U.S. care centers and they (and/or their legal guardians) provide informed consent, with approval from local institutional review boards (IRBs). This analysis was approved by the Indiana University IRB. For the current analysis, we reviewed data for all pwCF with at least one clinical encounter at a U.S. CF center after March 15 in 2019 or 2020, corresponding to the start of the spring 2019 peak of the COVID-19 pandemic. Individuals with CF transmembrane conductance regulator (CFTR)-related metabolic syndrome and CFTR-related disorders were excluded.

Variable Definitions

CF-related respiratory microbiology results, medications, and complications may be entered into the CFFPR at each clinical encounter, as is whether pwCF are seen by a respiratory therapist, dietician, and/or social worker. Median home zip code income and public insurance were used as markers of low socioeconomic status (12). Percentage predicted for forced expiratory volume in 1 second (FEV_1) measurements were calculated using the Global Lung Initiative reference equations (13). Baseline FEV_1 percentage ($FEV_1\%$) predicted as of March 15 was calculated based on the average of the two highest $FEV_1\%$ predicted values obtained at in-person clinic visits in the 12 months before and could include $FEV_1\%$ predicted obtained at the clinic visit. If only one $FEV_1\%$ predicted measurement was available, it was used as the baseline. No baseline value was assigned if there were no measurements of $FEV_1\%$ predicted in the past 12 months. The presence of depression and anxiety were based on self-report and results from annual Patient Health Questionnaire-9 and General Anxiety Disorder-7 screening entered into the CFFPR. BMI percentiles for children were based on the Centers for Disease Control and Prevention growth charts (14). PEx were defined separately according to *1)* the determination by the treating clinician at in-person clinic visits and *2)* treatment with intravenous antibiotics at home or in the hospital.

Statistical Analysis

Descriptive statistics were used to summarize the proportion of clinical encounters that occurred at in-person clinic visits, the proportion of clinical encounters with key clinical data and interactions with members of the multidisciplinary CF team, the clinical characteristics of pwCF seen in-person, and the proportion of pwCF who had a PEx. To address our hypothesis, we used paired *t* tests to compare within-subject differences in $FEV_1\%$ predicted and BMI, and calculated relative risk (RR) to compare the proportion of in-person clinic visits with a PEx for pwCF with at least one in-person clinic visit after March 15, 2019 and 2020. For this analysis, we excluded pwCF with prior lung transplantation. Although the use of elexacaftor/tezacaftor/ivacaftor (ETI) is recorded at clinical encounters, the exact start date is not. To account for ETI use, we separately report outcomes according to age and CF transmembrane conductance regulator (CFTR) protein mutations that determine eligibility for ETI (i.e., age ≥12 years and the presence of at least one copy of the *F508del* mutation).

Results

In-Person Clinic Visits

In 2019, 30,370 pwCF had at least one encounter at accredited U.S. CF centers between March 15 and December 31. In total, there were 105,049 in-person clinic visits among 30,097 pwCF who had at least one in-person clinic visit. In 2020, 30,163

pwCF had at least one encounter at accredited U.S. CF centers between March 15 and December 31. In total, there were only 46,633 in-person clinic visits among 22,811 pwCF who had at least one in-person clinic visit. Compared with 2019, all in-person clinic visits were reduced by 56%, and 24% fewer pwCF were seen at an in-person clinic visit in 2020. In 2019, in-person clinic visits accounted for 89–92% of all clinical encounters in any given month. In 2020, the proportion of clinical encounters that were in-person clinic visits decreased from a high of 90% in January and February to a low of 9% in April (Figure 1). The proportion of clinical encounters that were in-person clinic visits increased after the initial spring COVID-19 peak but did not return to 2019 levels.

Throughout 2019, more than 90% of clinical encounters had information recorded for height, weight, CF-related medications, and CF-related complications, and more than 75% had information recorded for respiratory microbiology and FEV_1 (*see* Table E1 in the online supplement). In 2020, the proportion of clinical encounters at which height, weight, microbiology, and FEV_1 were recorded varied greatly by month, correlating with the changes in the proportion of in-person clinic visits. However, reporting of CF-related medications and CF-related complications remained more stable over this time period (Table E1). Compared with 2019, the proportion of clinical encounters with interactions with respiratory therapists, dieticians, and social workers was lower in March and April, before increasing through September 2020 (Figure E1). The difference in interactions was greatest for interactions with respiratory therapists, reaching a low of 40% of levels seen in 2019. Interactions with respiratory therapists remained 15% below 2019 levels in December 2020, whereas

interactions with dieticians and social workers had returned to 2019 levels.

Characteristics of pwCF Seen In-Person

The characteristics of pwCF seen in-person during and after the spring peak of the pandemic were generally similar to pwCF seen during the same time period in 2019, though there was a lower proportion of pwCF seen in-person who were ≥18 years of age or had FEV_1 <70% predicted, and a higher proportion treated with any CFTR modulator or had FEV_1 ≥100% predicted (Table 1). The mean (SD) age was nearly two years lower in 2020, 20.9 (15.1) years, compared with 2019, 22.5 (15.4) years. There were no differences in the pwCF seen in-person according to Hispanic ethnicity or insurance status. The median income by zip code was not different in 2020, $64,188, compared with 2019, $65,307. The higher CFTR modulator use in 2020 was due to the introduction of ETI for pwCF ≥12 years of age and at least one copy of *F508del* at the end of 2019.

The characteristics of pwCF seen in-person after March 15 in both 2019 and 2020 (Table E2) were generally similar to the overall cohort, except the proportion of adult pwCF was lower due to the reduction in in-person clinic visits for adult pwCF in 2020. Among pwCF seen in-person in both time periods and without a history of lung transplantation, the mean (SD) FEV_1% predicted for pwCF seen in-person in 2020 was 85.0 (30.1), compared with 79.7 (27.6) in 2019; the mean (95% confidence interval [CI]) estimate of the difference is 5.3% (4.9–5.7) predicted. Mean (SD) FEV_1% predicted was statistically significantly higher for pwCF <12 years of age and pwCF ≥12 years of age with ≥1 copy of *F508del* (Table 2). There was no difference in FEV_1% predicted among pwCF ≥12 years of age

with no copies of *F508del* who were seen in-person in both time periods. The proportion of pwCF seen in-person during this time whose FEV_1 was >5% predicted below their baseline FEV_1 as of March 15 was lower in 2020 (22%) than in 2019 (39%).

The mean (SD) BMI for pwCF seen in-person after March 15 in both 2019 and in 2020 was higher in 2020 for adults, 24.7 (4.9) kg/m^2, compared with 2019, 23.6 (4.7) kg/m^2; the mean (95% CI) estimate of the difference is 1.08 (1.03–1.13) kg/m^2. Similarly, the mean (SD) BMI percentile in 2020 was higher for children, 59.1 (27.0) percentile, compared with 2019, 55.8 (26.5) percentile; the mean (95% CI) estimate of the difference is 3.3 (3.0–3.6). As with FEV_1% predicted, there were no differences between 2019 and 2020 in BMI percentile for children with CF ≥12 years of age and no copies of *F508del*, but BMI percentile was significantly higher in 2020 compared with 2019 for children with CF <12 years of age and children with CF ≥12 years of age with ≥1 copy of *F508del*. The largest increases were also seen among children with CF eligible for ETI (Table 2). The mean (SD) BMI was statistically significantly higher for adults with CF, regardless of eligibility for ETI, though the increase was again greater among adults with CF eligible for ETI (Table 2).

Pulmonary Exacerbations

The proportion of pwCF ≥12 years of age with at least one copy of the *F508del* CFTR mutation treated with intravenous antimicrobials at home or in the hospital for a PEx began to decrease substantially at the end of 2019 with the introduction of ETI (Figure 2). This decrease continued through the spring peak of the pandemic and was maintained through the end of 2020. The proportion of pwCF ≥12 years of age without any *F508del* CFTR mutations and pwCF <12 years of age (i.e., those not eligible for ETI) treated with intravenous antibiotics for a PEx began to decrease in 2020, before the spring COVID-19 peak. The lowest proportion of pwCF not eligible for ETI treated with intravenous antimicrobials was in April before rebounding slightly but staying below 2019 levels.

Similar to the trend of PEx treated with intravenous antimicrobials, yet in contrast to our hypothesis, the proportion of pwCF seen in-person during the spring peak of the pandemic (i.e., March 15 to May 31, 2020) who were diagnosed with a PEx (regardless of treatment) (25%) was lower than the
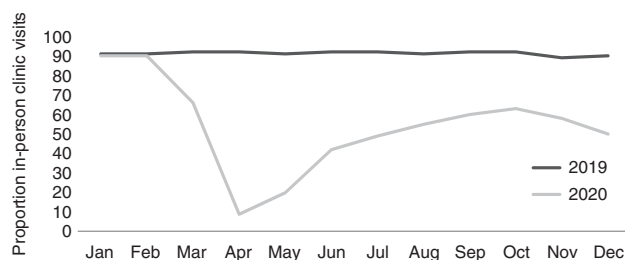

Figure 1. Proportion of clinical encounters recorded in the CFFPR that were in-person clinic visits. CFFPR = Cystic Fibrosis Foundation Patient Registry.

Table 1. Characteristics of people with cystic fibrosis seen in-person after March 15, 2019 and 2020

Characteristic		2019 N = 30,097, n (%)	2020 N = 22,811, n (%)
Age, yr	<2	1,109 (4)	924 (4)
	2–11	7,682 (25)	6,598 (29)
	12–17	4,769 (16)	4,065 (18)
	⩾18	16,537 (55)	11,222 (49)
Male sex		15,537 (52)	11,715 (51)
Race	White	27,723 (92)	20,893 (92)
	Black	1,402 (5)	1,140 (5)
	Other	972 (3)	778 (3)
Hispanic ethnicity		2,750 (10)	2,181 (10)
CFTR genotype	Homozygous *F508del*	13,462 (45)	10,302 (45)
	Heterozygous *F508del*	12,304 (41)	9,215 (40)
	Other/unknown	4,331 (14)	3,294 (15)
Pancreatic insufficient (on enzymes)		25,579 (85)	19,214 (84)
CF-related diabetes mellitus		6,598 (22)	4,576 (20)
Lung transplantation		1,586 (5)	992 (4)
Current year insurance status	Any public	15,012 (50)	12,049 (53)
	Only private	14,503 (48)	10,336 (45)
	Other/none/unknown	582 (2)	426 (2)
Any CFTR modulator use (at start of reporting period)		18,242 (61)	16,173 (71)
ETI use, ⩾12-yr-olds (start of reporting period)		216 (1)	9,285 (61)
Chronic inhaled steroid use		8,538 (28)	5,663 (25)
Baseline in-person clinic visit FEV$_1$ category, ⩾6-yr-olds*	⩾100% predicted	4,426 (18)	4,980 (27)
	70–99	11,610 (46)	8,726 (47)
	40–69	6,582 (26)	3,819 (21)
	<40	2,602 (10)	1,091 (6)
Microbiology history (any positive in year before)	*Pseudomonas aeruginosa*	12,330 (41)	8,648 (38)
	Staphylococcus aureus	19,385 (64)	15,112 (66)
	MRSA	6,906 (23)	5,345 (23)
	Stenotrophomonas maltophila	3,437 (11)	2,527 (11)
	Burkholderia cepecia complex	729 (2)	516 (2)
Any nontuberculous mycobacteria during reporting period		1,635 (5)	450 (2)
Allergic bronchopulmonary aspergillosis		1,571 (5)	997 (4)
Depression, ⩾12-yr-olds		5,061 (24)	3,496 (23)
Anxiety, ⩾12-yr-olds		4,425 (21)	3,248 (21)

Definition of abbreviations: CF = cystic fibrosis; CFTR = cystic fibrosis transmembrane conductance regulator; ETI = elexacaftor/tezacaftor/ivacaftor; FEV$_1$ = forced expiratory volume in 1 second; MRSA = methicillin-resistant *Staphylococcus aureus*; pwCF = people with CF.
*Baseline FEV$_1$ is based on the average of the best two FEV$_1$ measurements in the year before. Baseline FEV$_1$ for 2019 was based on only one measurement for 6.5% of pwCF; 1.6% had no measurements in the year before and used the in-person clinic measurement as baseline FEV$_1$. Baseline FEV$_1$ for 2020 was based on only one measurement for 4.8% of pwCF; 1.4% had no measurements and used the in-person clinic measurement as baseline FEV$_1$.

corresponding period in 2019 (38%), or a RR of 0.82 (0.79–0.86) in 2020 compared with 2019.

Discussion

The impact of the COVID-19 pandemic is seen in changes in the care of pwCF in the United States. In-person clinic visits were greatly reduced during the spring peak of the pandemic, and although many pwCF subsequently returned to in-person clinic visits, approximately 25% of pwCF seen in-person after March 15 in 2019 were not seen in-person at all after March 15 in 2020. In addition, pwCF were less likely to interact with members of the multidisciplinary team or have important CF-related clinical measurements recorded. Interactions with members of the multidisciplinary team and recording of CF-related clinical measurements decreased in concert with the reduction of in-person clinic visits. Respiratory therapists had the largest decrease in interactions with pwCF; this may be because pulmonary function test labs were closed, and/or respiratory therapists were more likely to be deployed to support the care of people hospitalized with COVID-19. In contrast to our hypothesis, pwCF seen in-person did not have evidence of worse lung disease, nutrition, or risk of PEx. The introduction of ETI had a large positive effect on disease outcomes and, therefore, may have masked lung function decline in those eligible. While we saw no evidence for worse lung disease even among pwCF under 12 years of age who were not eligible for ETI, we cannot say for certain that adults with more severe lung disease would have similar

Table 2. Mean (standard deviation) body mass index and forced expiratory volume in 1 second for people with cystic fibrosis seen in-person after March 15, 2019 and 2020 according to age and elexacaftor/tezacaftor/ivacaftor eligibility

		n	2019, Mean (SD)	2020, Mean (SD)	Mean (95% CI) differences	*P* Value
FEV$_1$	6–<12 yr of age	3,514	96.0 (29.0)	97.2 (29.3)	1.3 (0.1 to 2.4)	0.03
	≥12 yr of age, no *F508del*	1,523	76.9 (25.9)	76.6 (33.2)	−0.3 (−1.6 to 0.9)	0.60
	≥12 yr of age, ≥1 *F508del*	10,089	74.5 (25.1)	82.0 (28.7)	7.5 (7.1 to 7.9)	<0.001
BMI percentile	<12 yr of age	6,373	58.0 (25.7)	60.4 (26.6)	2.4 (2.0 to 2.8)	<0.001
	12–<18 yr of age, no *F508del*	649	54.8 (29.1)	55.5 (30.1)	0.7 (−0.3 to 1.7)	0.18
	12–<18 yr of age, ≥1 *F508del*	4,056	52.3 (26.9)	57.6 (26.9)	5.2 (4.8 to 5.7)	<0.001
BMI, kg/m^2	≥18 yr of age, no *F508del*	1,002	24.4 (5.4)	24.6 (5.6)	0.2 (0.1 to 0.3)	<0.001
	≥18 yr of age, ≥1 *F508del*	6,421	23.5 (4.5)	24.7 (4.8)	1.2 (1.2 to 1.3)	<0.001

Definition of abbreviations: BMI = body mass index; CI = confidence interval; FEV$_1$ = forced expiratory volume in 1 second; SD = standard deviation.

results. In addition, the CFFPR does not include sensitive markers such as chest computed tomographic or multiple breath washout that may be necessary to detect differences in disease severity.

The CFF recommends frequent clinical encounters to monitor for progression of lung disease, surveillance of respiratory microbiology, and monitoring for CF-related complications (8–10, 15). During the spring peak of the pandemic, this monitoring was disrupted. The impact of changes in care for pwCF may not be known for some time. A delay in the detection of newly acquired *Pseudomonas aeruginosa* could decrease the likelihood of successful eradication treatment attempts and increase the risk of subsequent PEx (16–19). pwCF, their caregivers, and parents of children with CF report negative impacts of the COVID pandemic on their emotional well-being (20). These changes in emotional well-being may not have been identified during the spring peak of the

pandemic in the absence of in-person clinic visits and reduced interactions with the multidisciplinary team, including social workers and psychologists. Respiratory symptoms are common presentations of PEx and may be reported during phone and telemedicine encounters (21), but respiratory signs (e.g., new crackles and declines in FEV$_1$% predicted are strongly associated with PEx and may have been missed in the absence of in-person clinic visits) (22). These risks should have been mitigated in pwCF who started ETI just before the start of the pandemic, in whom we would expect to see improved BMI, FEV$_1$% predicted, and respiratory symptoms (23, 24).

The potential negative impacts of changes in care may be addressed by supporting pwCF outside of in-person visits. Given the state of emergency, the U.S. Department of Health and Human Services relaxed restrictions to expand the options for applications used for telemedicine (25). The

CFF purchased and helped distribute devices to monitor pulmonary function at home. In addition, efforts have been made to allow for home collection of cultures of respiratory secretions (26). Although we did not detect differences in in-person clinic visits according to potential barriers to telehealth services, heightened efforts to address disparities in access to care are needed given the poorer outcomes among pwCF with disparities in social determinants of health (27), as well as poorer outcomes of COVID-19 infections among people with disparities in social determinant of health (28).

The reduction in PEx follows a similar reduction in exacerbations of chronic diseases, acute medical events, and injuries seen during the spring peak of the COVID-19 pandemic (2–4) but is also largely attributable to the effects of ETI (23). Notably, the largest decrease in intravenous-treated PEx was seen among pwCF eligible for ETI. Viral infections have been implicated in PEx (29–31), but the dramatic reduction in PEx during this period, corresponding with a reduction in circulating respiratory viruses (32), would argue that the contributions of viral infections to PEx have been underestimated. Even among pwCF not eligible for ETI, PEx began to decrease before in-person clinic visits did. pwCF have been considered at high risk for complications of infection with COVID-19 (33), and they may have changed behaviors to reduce their exposure to the virus and/or improve baseline health to better manage infection (20). pwCF may have also avoided hospitalizations by taking oral antibiotics or home intravenous antibiotics; numbers are small, but the reduction in intravenous antibiotics may be due primarily to a
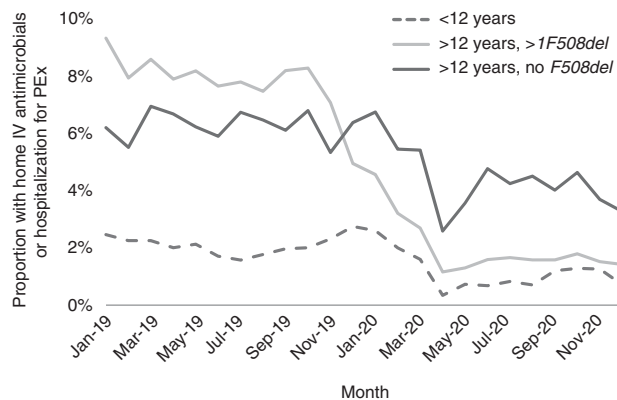

Figure 2. Proportion of pwCF with home intravenous (IV) antimicrobials or hospitalization for a PEx by month, age, and ETI eligibility. ETI = elexacaftor-tezacaftor-ivacaftor; pwCF = people with CF; PEx = pulmonary exacerbation.

reduction in hospitalizations, as compared with a smaller reduction in home intravenous antibiotics (data not shown).

Our report has several limitations. Our analysis is dependent on accurate data entry, and while error rates in data from the CFFPR, in general, are low (11), the pandemic may have altered these processes. It is possible that the high rates of reporting of CF-related medications and complications are attributable to carrying forward responses from previous encounters. Conversely, encounters with multidisciplinary team members may have occurred but were not recorded in the CFFPR. Although significant efforts are underway to closely monitor for anxiety and depression (34), responses from mental health screening questionnaires have only been completed and recorded in the CFFPRfor approximately 60–70% of pwCF ≥12 years of age. Telemedicine encounters were integrated into the CFFPR during the pandemic, but given some variability in the timing of uptake among centers, we focused solely on in-person clinic visits. This may have resulted in missed reports of PEx, but PEx treated via phone and/or telemedicine encounters may not contribute significantly to PEx treatments during the pandemic (35). The CFFPR does not capture potential causes for the reduction in PEx (e.g., avoidance of viral infections or improved adherence to chronic therapies during stay-at-home orders). It is possible that improvements in 2020 may in part be attributed to *1)* healthier pwCF attending in-person clinic visits during the spring 2020 peak of the pandemic and/or *2)* in-person clinic visits that occurred as part of the ETI initiation process and/or for monitoring of laboratory values while initiating ETI. However, the higher FEV_1 measurements during the spring 2020 peak of the pandemic were largely attributable to the group eligible for ETI, and the reduction in the number of in-person clinic visits was similar regardless of ETI eligibility (data not shown).

Initial reports of COVID-19 infections in pwCF have not demonstrated a significant risk for severe illnesses, although pwCF with older age, CF-related diabetes, and lower pulmonary function are at risk for hospitalization with COVID-19 (36, 37). Nonetheless, CF centers will need to be prepared to adapt their practices for future pandemic-related lockdowns and restrictions to in-person clinic visits, as well as the likelihood that some pwCF may prefer telemedicine visits over in-person visits (38). Telemedicine offers opportunities and presents unique challenges for patient care that is multidisciplinary, as in CF. Reports have indicated the potential for inequalities in access to telemedicine based on educational attainment, socioeconomic status, and language barriers (7, 39). This could adversely impact the care of pwCF with lower socioeconomic status, Hispanic ethnicity and other minorities, and/or more severe disease who are at risk for more severe outcomes (40, 41). Access to telemedicine services, with the inclusion of members of the multidisciplinary team, is needed to address changes in emotional well-being and support optimal self-care practices. The ability to monitor weight with high-accuracy home scales and pulmonary function with home spirometry devices would decrease the risk of "silent" progression of disease. Home spirometry results may be more variable, and efforts to provide remote support to enhance technique would be beneficial. Methods for home collection of respiratory secretion would reduce the risk of missed new infections with *P. aeruginosa*. CF centers are rapidly adapting to care during the COVID-19 pandemic, and the lessons learned during this time can be used to promote the health of pwCF in the future. ∎

References

1 Studdert DM, Hall MA. Disease control, civil liberties, and mass testing - calibrating restrictions during the Covid-19 pandemic. *N Engl J Med* 2020;383:102–104.

2 Bhambhvani HP, Rodrigues AJ, Yu JS, Carr JB, 2nd, Hayden Gephart M. Hospital volumes of 5 medical emergencies in the COVID-19 pandemic in 2 US medical centers. *JAMA Intern Med* 2021;181:272–274.

3 Blecker S, Jones SA, Petrilli CM, Admon AJ, Weerahandi H, Francois F, *et al.* Hospitalizations for chronic disease and acute conditions in the time of COVID-19. *JAMA Intern Med* 2021;181:269–271.

4 Simoneau T, Greco KF, Hammond A, Nelson K, Gaffin JM. Impact of the COVID-19 pandemic on pediatric emergency department use for asthma. *Ann Am Thorac Soc* 2021;18:717–719.

5 Woolhandler S, Himmelstein DU. Intersecting US epidemics: COVID-19 and lack of health insurance. *Ann Intern Med* 2020;173:63–64.

6 Patel SY, Mehrotra A, Huskamp HA, Uscher-Pines L, Ganguli I, Barnett ML. Trends in outpatient care delivery and telemedicine during the COVID-19 pandemic in the US. *JAMA Intern Med* 2021;181:388–391.

7 Katzow MW, Steinway C, Jan S. Telemedicine and health disparities during COVID-19. *Pediatrics* 2020;146:e20201586.

8 Cystic Fibrosis Foundation, Borowitz D, Robinson KA, Rosenfeld M, Davis SD, Sabadosa KA, *et al.* Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. *J Pediatr* 2009;155:S73–S93.

9 Lahiri T, Hempstead SE, Brady C, Cannon CL, Clark K, Condren ME, *et al.* Clinical practice guidelines from the cystic fibrosis foundation for preschoolers with cystic fibrosis. *Pediatrics* 2016;137: e20151784.

10 Yankaskas JR, Marshall BC, Sufian B, Simon RH, Rodman D. Cystic fibrosis adult care: consensus conference report. *Chest* 2004; 125:1S–39S.

11 Knapp EA, Fink AK, Goss CH, Sewall A, Ostrenga J, Dowd C, *et al.* The cystic fibrosis foundation patient registry. Design and methods of a national observational disease registry. *Ann Am Thorac Soc* 2016;13: 1173–1179.

12 Schechter MS. Nongenetic influences on cystic fibrosis outcomes. *Curr Opin Pulm Med* 2011;17:448–454.

13 Quanjer PH, Stanojevic S, Cole TJ, Baur X, Hall GL, Culver BH, *et al.* Multi-ethnic reference values for spirometry for the 3–95-yr age range: the global lung function 2012 equations. *Eur Respir J* 2012;40: 1324–1343.

14 Kuczmarski RJ, Ogden CL, Grummer-Strawn LM, Flegal KM, Guo SS, Wei R, *et al.* CDC growth charts: United States. *Adv Data* 2000:1–27.

15 Johnson C, Butler SM, Konstan MW, Morgan W, Wohl ME. Factors influencing outcomes in cystic fibrosis: a center-based analysis. *Chest* 2003;123:20–27.

16 Mayer-Hamblett N, Kronmal RA, Gibson RL, Rosenfeld M, Retsch-Bogart G, Treggiari MM, *et al.* Initial Pseudomonas aeruginosa treatment failure is associated with exacerbations in cystic fibrosis. *Pediatr Pulmonol* 2012;47:125–134.

17 Mayer-Hamblett N, Kloster M, Rosenfeld M, Gibson RL, Retsch-Bogart GZ, Emerson J, et al. Impact of sustained eradication of new Pseudomonas aeruginosa infection on long-term outcomes in cystic fibrosis. Clin Infect Dis 2015;61:707–715.

18 Cohen-Cymberknoh M, Gilead N, Gartner S, Rovira S, Blau H, Mussaffi H, et al. Eradication failure of newly acquired Pseudomonas aeruginosa isolates in cystic fibrosis. J Cyst Fibros 2016;15: 776–782.

19 Ratjen F, Moeller A, McKinney ML, Asherova I, Alon N, Maykut R, et al.; EARLY study group. Eradication of early P. aeruginosa infection in children <7 years of age with cystic fibrosis: the early study. J Cyst Fibros 2019;18:78–85.

20 Havermans T, Houben J, Vermeulen F, Boon M, Proesmans M, Lorent N, et al. The impact of the COVID-19 pandemic on the emotional well-being and home treatment of Belgian patients with cystic fibrosis, including transplanted patients and paediatric patients. J Cyst Fibros 2020;19:880–887.

21 Hoppe JE, Hinds DM, Colborg A, Wagner BD, Morgan WJ, Rosenfeld M, et al. Oral antibiotic prescribing patterns for treatment of pulmonary exacerbations in two large pediatric CF centers. Pediatr Pulmonol 2020;55:3400–3406.

22 Rabin HR, Butler SM, Wohl ME, Geller DE, Colin AA, Schidlow DV, et al.; Epidemiologic Study of Cystic Fibrosis. Pulmonary exacerbations in cystic fibrosis. Pediatr Pulmonol 2004;37:400–406.

23 Middleton PG, Mall MA, Drevinek P, Lands LC, McKone EF, Polineni D, et al. Elexacaftor-tezacaftor-ivacaftor for cystic fibrosis with a single Phe508del allele. N Engl J Med 2019;381:1809–1819.

24 Heijerman HGM, McKone EF, Downey DG, Van Braeckel E, Rowe SM, Tullis E, et al. Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: a double-blind, randomised, phase 3 trial. Lancet 2019;394:1940–1948.

25 Department of Health and Human Services. Notification of enforcement discretion for telehealth remote communications during the COVID-19 nationwide public health emergency 2020 [accessed 2021 February 3]. Available from: https://www.hhs.gov/hipaa/for-professionals/special-topics/emergency-preparedness/notification-enforcement-discretion-telehealth/index.html.

26 Lenhart-Pendergrass PM, Anthony M, Sariyska S, Andrews A, Scavezze H, Towler E, et al. Detection of bacterial pathogens using home oropharyngeal swab collection in children with cystic fibrosis. Pediatr Pulmonol 2021;56:2043–2047.

27 Schechter MS, Shelton BJ, Margolis PA, Fitzsimmons SC. The association of socioeconomic status with outcomes in cystic fibrosis patients in the United States. Am J Respir Crit Care Med 2001;163: 1331–1337.

28 Mackey K, Ayers CK, Kondo KK, Saha S, Advani SM, Young S, et al. Racial and ethnic disparities in COVID-19-related infections, hospitalizations, and deaths: a systematic review. Ann Intern Med 2021;174:362–373.

29 Somayaji R, Goss CH, Khan U, Neradilek M, Neuzil KM, Ortiz JR. Cystic fibrosis pulmonary exacerbations attributable to respiratory syncytial virus and influenza: a population-based study. Clin Infect Dis 2017;64: 1760–1767.

30 Etherington C, Naseer R, Conway SP, Whitaker P, Denton M, Peckham DG. The role of respiratory viruses in adult patients with cystic fibrosis receiving intravenous antibiotics for a pulmonary exacerbation. J Cyst Fibros 2014;13:49–55.

31 Asner S, Waters V, Solomon M, Yau Y, Richardson SE, Grasemann H, et al. Role of respiratory viruses in pulmonary exacerbations in children with cystic fibrosis. J Cyst Fibros. 2012;11:433–439.

32 Olsen SJ, Azziz-Baumgartner E, Budd AP, Brammer L, Sullivan S, Pineda RF, et al. Decreased influenza activity during the COVID-19 pandemic - United States, Australia, Chile, and South Africa, 2020. MMWR Morb Mortal Wkly Rep 2020;69:1305–1309.

33 Centers for Disease Control. People with certain medical conditions 2020 [updated 2021 February 1; accessed 2021 February 3]. Available from: https://www.cdc.gov/coronavirus/2019-ncov/need-extra-precautions/people-with-medical-conditions.html.

34 Quittner AL, Abbott J, Hussain S, Ong T, Uluer A, Hempstead S, et al. Integration of mental health screening and treatment into cystic fibrosis clinics: evaluation of initial implementation in 84 programs across the United States. Pediatr Pulmonol 2020;55:2995–3004.

35 Patel S, Thompson MD, Slaven JE, Sanders DB, Ren CL. Reduction of pulmonary exacerbations in young children with cystic fibrosis during the COVID-19 pandemic. Pediatr Pulmonol 2021;56:1271–1273.

36 McClenaghan E, Cosgriff R, Brownlee K, Ahern S, Burgel PR, Byrnes CA, et al. The global impact of SARS-CoV-2 in 181 people with cystic fibrosis. J Cyst Fibros 2020;19:868–871.

37 Cosgriff R, Ahern S, Bell SC, Brownlee K, Burgel PR, Byrnes C, et al. A multinational report to characterise SARS-CoV-2 infection in people with cystic fibrosis. J Cyst Fibros 2020;19:355–358.

38 Jaclyn D, Andrew N, Ryan P, Julianna B, Christopher S, Nauman C, et al. Patient and family perceptions of telehealth as part of the cystic fibrosis care model during COVID-19. J Cyst Fibros 2021;20:e23–e28.

39 Veinot TC, Mitchell H, Ancker JS. Good intentions are not enough: how informatics interventions can worsen inequality. J Am Med Inform Assoc 2018;25:1080–1088.

40 Schechter MS, McColley SA, Silva S, Haselkorn T, Konstan MW, Wagener JS, et al. Association of socioeconomic status with the use of chronic therapies and healthcare utilization in children with cystic fibrosis. J Pediatr 2009;155:634–639.e1–e4.

41 Rho J, Ahn C, Gao A, Sawicki GS, Keller A, Jain R. Disparities in mortality of hispanic patients with cystic fibrosis in the United States. A national and regional cohort study. Am J Respir Crit Care Med 2018; 198:1055–1063.