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Improved recognition of lung function decline as signal of cystic fibrosis pulmonary exacerbation: a Cystic Fibrosis Learning Network Innovation Laboratory quality improvement initiative

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ABSTRACT

Introduction Cystic fibrosis (CF) is a systemic autosomal recessive condition characterised by progressive lung disease. CF pulmonary exacerbations (PEx) are episodes of worsening respiratory status, and frequent PEx are a risk factor for accelerated lung function decline, yet many people with CF (PwCF) go untreated at the time of decline. The goal of this quality improvement (QI) initiative was to improve recognition, treatment and follow-up of PEx in PwCF.

Methods Using the Model for Improvement, the Cystic Fibrosis Learning Network (CFLN) initiated a QI innovation laboratory (iLab) with a global aim to decrease the rate of lung function decline in PwCF. The iLab standardised definitions for signals of PEx using a threshold for decline in forced expiratory volume in one second (FEV_1) and/or changes in symptoms. The FEV_1 decline signal was termed FIES (FEV_1-indicated exacerbation signal). Processes for screening and recognition of FIES and/or symptom changes, a treatment algorithm and follow-up in the presence of a signal were tested concurrently in multiple settings.

Specific aims The specific aim is to increase the per cent of PwCF assessed for a PEx signal at ambulatory encounters and to increase the per cent of recommendations to follow-up within 6 weeks for PwCF experiencing a PEx signal.

Results FIES recognition increased from 18.6% to 73.4% across all teams during the iLab, and every team showed an improvement. Of PwCF assessed, 15.8% experienced an FIES event (>10% decline in FEV_1 per cent predicted (FEV_1pp)). Follow-up within 6 weeks was recommended for an average of 70.5% of those assessed for FIES and had an FEV_1pp decline greater than 5%.

Conclusion The CFLN iLab successfully defined and implemented a process to recognise and follow-up PEx signals. This process has the potential to be spread to the larger CF community. Further studies are needed to assess the impact of these processes on PwCF outcomes.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Untreated pulmonary exacerbations (PEx) lead to worsening health and lower likelihood of returning to baseline lung health in people with cystic fibrosis (PwCF). CF Patient Registry data show that a substantial number of patient encounters with a 10% or greater decline in forced expiratory volume in one second per cent predicted (FEV_1pp), go untreated. There is currently no standard approach to assessing PEx in PwCF.

WHAT THIS STUDY ADDS

⇒ This project successfully standardised a definition for PEx signals based on a decline in FEV_1pp from baseline and implemented a process for assessing for signals at every patient encounter and following a specific treatment algorithm until resolution.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ This study demonstrated that a standardised PEx assessment and treatment pathway could be implemented in many different contexts and has the potential to improve PEx recognition, lead to more timely exacerbation treatment and preserve lung health for PwCF across the CF Care Center Network.

INTRODUCTION

Problem description

Cystic fibrosis (CF) is an autosomal recessive condition caused by mutations in the gene coding for the CF transmembrane conductance regulator (CFTR) protein, resulting in progressive lung disease. CF pulmonary exacerbations (PEx) are acute or subacute episodes of worsening respiratory status. PEx are associated with higher cost burden, lower quality of life and shortened survival in people with CF (PwCF), and frequent PEx episodes

are a risk factor for accelerated lung function decline.[1] The introduction of highly effective CFTR modulator therapy has reduced the incidence of PEx, although they continue to be reported in 10%–20% of PwCF depending on their age and baseline lung disease severity.[2]

Available knowledge

CF PEx are diagnosed clinically, yet no established consensus criteria on the definition of PEx exist. Clinical features commonly associated with a PEx diagnosis include increased cough, increased sputum production, change in chest examination and decline in lung function as measured by the forced expiratory volume in one second per cent predicted (FEV_1pp).

There is currently no standardised approach to assessing CF exacerbations based on symptoms. Several questionnaires assessing CF symptoms were developed for use in research studies, though none of them have been validated in clinical care or shown to be an accurate predictor of CF exacerbation outside of clinical trials. In addition, most questionnaires used in research are cost prohibitive for clinical use and vary in terms of diagnosis and response within and among centres.[3–5]

A decline in FEV_1pp of ≥10% from baseline represents an objective and clinically significant change in a patient's pulmonary function. However, analysis of the CF Foundation (CFF) patient registry data reveals between 15% and 30% of PwCF with >10% FEV_1pp decline below their baseline at a clinical encounter go untreated.[6–8] While not all these encounters necessarily represent PEx, data suggest that a substantial number of PEx go untreated. PwCF with untreated episodes of FEV_1pp decline are less likely to return to baseline than those who receive treatment with antibiotics.[6–9] In particular, those with higher baseline FEV_1pp have been shown to experience steeper rates of decline and are least likely to receive treatment for acute decline in lung function.[9 10]

Despite the importance of recognising and treating clinically significant FEV_1 decline, there are very few published care pathways that have shown improvement in the rate of treating FEV_1 decline. Kraynack *et al*[11] demonstrated that use of a PEx scoring tool that incorporated FEV_1 as one of the elements resulted in less variation in PEx diagnosis among different providers, but was not associated with clear improvement in median FEV_1 at their centre. Schechter *et al*[12] reported that incorporation of a quality improvement (QI) programme at a single care centre to ensure a consistent approach to the management of FEV_1 decline was associated with a significant improvement in median FEV_1 over time. Both programmes emphasised recognition of FEV_1 decline.

Rationale

Recognising the importance of addressing FEV_1 decline in PwCF, a subgroup of the CF Learning Network (CFLN) care centre teams[13] initiated a collaborative, multicentre QI project, known as an innovation laboratory (iLab), to standardise the recognition and treatment of FEV_1 decline.

The CFF has a rich history of investing and supporting the use of QI science to accelerate improvement in CF care and the CFLN is a collaborative, learning healthcare system sponsored by the CFF. At the time of this study, the CFLN was comprised of teams from 36 distinct accredited care centres and each team included interdisciplinary care team members and their associated patient and family partners (PFPs).[13] Those participating in the CFLN follow the Model for Improvement QI methodology and are supported by a centralised team of operational support, QI specialists and data analysts who facilitate team collaborations, QI tools and learning and manage centralised data collection, analysis and modelling.[13] The goal of the CFLN is for care teams and their PFPs to collaboratively improve care processes and patient outcomes using CFF Patient Registry data to inform decisions.[13 14] An iLab is a subset of CFLN teams who collaborate on a specific QI project with additional dedicated support of QI specialists and operational support to implement and rapidly test small changes in multiple contexts simultaneously. In addition, centralised reporting, data collection and analysis is embedded in the iLab structure to facilitate rapid learning and increase process reliability with a goal of reaching improved outcomes more quickly.[13 14]

Based on the available knowledge, it was hypothesised that standardised tools and reliable processes to assess, recognise and treat episodes of lung function decline in PwCF would reduce the magnitude of decline. Central to the rationale for this initiative and the mission of the CFLN, is inclusion of the patient and family perspective, which is demonstrated in online supplemental appendix A.

Global aim

The global aim of this iLab initiative was to *implement a standardised approach to PEx* that would result in a decreased rate of lung function decline in PwCF.

Specific aims

In the time frame of the iLab, the group focused its objectives on the following specific aims:

1. Increase the per cent of PwCF who were assessed for an exacerbation signal at each ambulatory care visit.
2. Increase the per cent of recommendations for follow-up within 6 weeks for PwCF who experienced an exacerbation signal.

METHODS

Context

This QI initiative commenced in August 2019 with a design meeting of key stakeholders, thought leaders and PwCF who were tasked with harmonising on the concept of an exacerbation signal. This was based on a defined drop in lung function FEV_1, subsequently known as an FEV_1-indicated exacerbation signal (FIES). FIES was defined as a >10% decline in FEV_1pp from baseline for

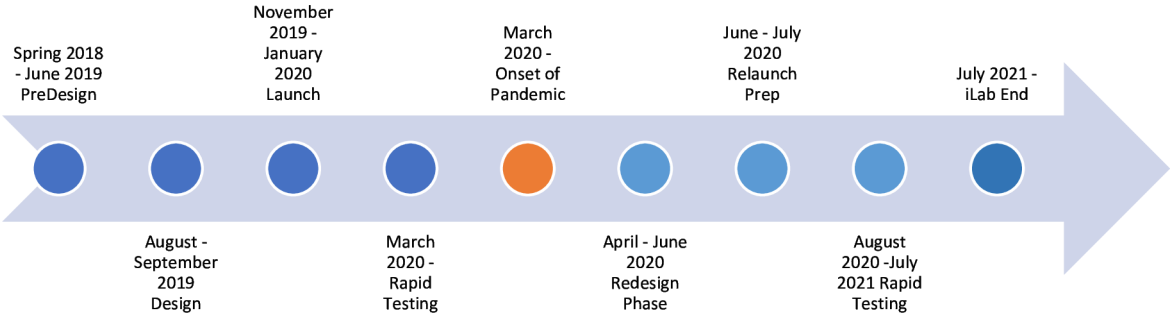

Figure 1 Timeline of FIES innovation laboratory

PwCF with baseline values at or greater than 50% FEV_1pp. For PwCF whose baseline FEV_1pp was less than 50%, FIES was defined as a 5% decline from baseline.

After establishing the FIES definition and foundational specific aims, the design team identified change ideas and developed interventions using a key driver diagram, (online supplemental appendix B), constructed an algorithm for the identification and treatment of FIES (online supplemental appendix C) and laid out the operational structure and timeline for the iLab (figure 1; online supplemental appendix D).

Criteria for teams to participate in the iLab included expertise in QI and a 12–18 month commitment from a triad of physician champion, QI leader and PFP. Participation initially required teams to attend weekly iLab huddles, conduct biweekly or monthly programme QI team meetings, submit weekly plan–do–study–act (PDSA) testing for designated interventions, engage in regular data tracking and submit specific process measures to iLab data managers.

The iLab officially launched in January 2020 and 21 out of 36 CFLN centre teams committed to participate. Each participating centre created a process map for the assessment of FIES at every eligible CF clinic encounter in their individual contexts (online supplemental appendix E). Shared measures were established, and initial testing commenced in March 2020. However, the COVID-19 pandemic began shortly thereafter, forcing pulmonary function lab and ambulatory clinic closures across the country and causing many clinics to adapt to telehealth visits.[15–17] With this unanticipated change in care, the FIES iLab was paused until August 2020 with an option to continue to test and submit data in the interim, as able.

The iLab officially relaunched in August 2020 with 11 of 21 original teams resuming and one new team joining for a total of 12 iLab teams participating through the end of the iLab. Many teams, who did not rejoin, reported diminished capacity to participate due to various effects of the pandemic in their contexts. With pulmonary function testing still not fully available at this point, the iLab pivoted accordingly by adapting the PEx algorithm to include a symptom-indicated exacerbation signal (SIES). This was adapted from the Fuchs definition of PEx[5] and used in the absence of available lung function measurement. This symptom assessment (online supplemental appendix F) provided a mechanism to continue using a standardised exacerbation signal for recognition of PEx at the point of care, even in the setting of telehealth without reliable home spirometry or pulmonary function testing in the ambulatory setting. Although teams updated processes to incorporate this shift (online supplemental appendix E), the use of SIES as an alternative exacerbation recognition tool was not uniformly adopted or tested by iLab teams.

The iLab's launch in January 2020 and relaunch in August 2020 followed a threefold strategy: (1) biweekly huddles with participating iLab teams, (2) biweekly data submission on all measures collected and managed using REDCap[18 19] electronic data capture tools and reviewed at each huddle and (3) biweekly strategic leadership huddles.

Each team developed and shared their own process for real-time recognition of FIES/SIES, completed coordinated tests of change on a weekly basis and documented them on PDSA worksheets and ramp summaries. These were reviewed in the iLab biweekly huddles to understand team engagement and to share and disseminate successful processes. The iLab collection instrument asked centres to include information for FIES or SIES for PwCF over 6 years old to avoid reacting to unreliable spirometry data.

Interventions

This improvement initiative incorporated several initial interventions:

- Development of centre-specific tools to calculate individual FEV_1 baselines for PwCF followed at the centre and the threshold for decline in FEV_1 that would trigger an action.
- Use of a standardised algorithm to guide actions based on the magnitude of FEV_1 decline from baseline FEV_1pp (FIES) or symptoms (SIES) in the absence of spirometry.
- Development and use of a centre-specific shared decision-making tool to coproduce treatment decisions.

The initial iLab's algorithm and key driver diagram (KDD) evolved through several iterations to account for the iLab's pivot during the COVID-19 pandemic and inclusion of SIES, demonstrating the flexible and adaptive nature of the iLab construct. The final KDD and

algorithm are shown in online supplemental appendices B and C, respectively.

Intervention 1

Each iLab team was asked to develop a process for identifying a baseline FEV_1pp value for each PwCF aged 6 and older who had at least one spirometry measurement in the preceding year, as part of their preclinic preparation. Teams were then asked to adopt or adapt a tool that would automatically calculate a threshold value indicating FIES that would inform subsequent actions. Tools were developed at the individual centre level and shared with the rest of the iLab through the cloud-based file sharing and storing platform, Dropbox (https://www.dropbox.com).

Intervention 2

The second intervention built on the adoption of the first intervention whereby teams would compare the FEV_1pp measured at the time of a clinical encounter to the calculated FIES threshold, to determine the presence of FIES (online supplemental appendix C). A treatment algorithm was introduced, and teams were asked to test its use and share data and learning back through the iLab REDCap survey, team huddles and Dropbox file sharing (online supplemental appendix C).

The algorithm was initially designed to suggest treatment pathways for FIES and was later expanded to direct treatment pathways for SIES if lung function testing was not possible. The algorithm was organised into coloured zones based on the severity of the decline in FEV_1pp or existence of any new pulmonary symptoms. The 'green zone' was established for PwCF with no substantial FEV_1 decline (<5%) or no new symptoms, and recommended continuation of routine therapies and routine follow-up. The 'yellow zone' pathway was indicated for PwCF who had a minor decline in FEV_1pp and no new symptoms. The yellow zone included declines from baseline between >5% and 9% for those with a baseline>50% FEV_1pp; or a decline between>3% and 4% for those with a baseline<50 FEV_1pp. This pathway included decision points for clinicians to consider increased or new therapies and recommended follow-up within 6 weeks to assess for a return to baseline or persistent/worsening decline.

For PwCF experiencing FIES or an SIES (any increase in symptoms), a 'red zone' pathway was defined and recommended antibiotic treatment, either oral or intravenous depending on severity, and recommended follow-up within 6 weeks. Given the many possible clinical scenarios, recommendation for follow-up within 6 weeks was the only standard feature in the algorithm for both yellow and red zones and offered a consistent means of measuring algorithm use.

Intervention 3

Teams were provided several models of shared decision-making tools that were codesigned with subject-matter experts including PwCF. The proposed intervention was to adopt or adapt one of the tools and create a process to implement at the point of care when FIES or SIES was identified. The use of the shared decision-making tools were not fully adopted by teams due to the interruption of the pandemic and the time constraints it placed on the iLab timeline.

Study of the interventions

Individual team PDSAs addressing real-time recognition of FIES were trialled and adapted early in the iLab using rapid tests of change. While individual centre processes for real-time recognition varied, they all incorporated the following elements: Previsit planning with identification of those PwCF eligible for FIES assessment, the establishment of their individual best baseline FEV_1 in the prior 12 months and their threshold for FIES. The FIES calculation process varied by teams reflecting individual centre context and ranged from hand calculation to use of Excel files or spirometry applications. All teams were asked to adopt use of the algorithm and establish a consistent method for ensuring early follow-up for those with identified FIES/SIES.

Measures

Initial process measures chosen to address the specific aims were:

1. Per cent of eligible encounters in which the patient was assessed for FIES.
2. Per cent of eligible encounters in which the patient was assessed for an FIES and experienced an FIES.

Additional measures were added at the time of the relaunch in August 2020 to accommodate those visits done in the absence of available spirometry testing (telehealth or pandemic restrictions on pulmonary function testing).

1. Per cent of eligible encounters in which the patient was assessed for SIES (in the absence of available spirometry testing).
2. Per cent of eligible encounters in which the patient was assessed for an SIES and experienced an SIES.

Once processes for real-time recognition of FIES were standardised, the iLab approached the subsequent aim of implementation of the FIES algorithm for treatment of exacerbation signal.

The use of the pulmonary algorithm was measured using a surrogate of recommendations for early follow-up.

Per cent of patients assessed to have FIES/SIES who received recommendation for follow-up within 6 weeks.

Data collection on algorithm use and clinician acknowledgement of exacerbation signals (as indicated by recommendation for follow-up within 6 weeks) began in December 2020. This was also submitted to the REDCap database and tracked bimonthly.

RESULTS

When the iLab first launched in January 2020, 6 adult centre teams and 15 paediatric centre teams joined. In total, 11 of the original teams and 1 additional team joined the relaunched iLab in August 2020, for a total of

12 participating centres. The final iLab cohort comprised of three adult and nine paediatric centre teams that ranged in size with four small centres (50–125 patients), two medium centres (125–200 patients) and six large centres (more than 200 patients). Those that elected not to continue reported diminished capacity to participate considering the various impacts of the ongoing pandemic in their contexts.

ILab teams met over a period of 18 months (January 2020–June 2021), with a 5-month interruption during the pandemic before the official relaunch in August 2020. There were a total of 30 huddles between February 2020 and July 2021 and attendance ranged between 67% and 100% of participating teams (median 92%). Distinct teams attended an average of 89% of huddles (range 63%–100%; median 90%).

Teams tested and refined interventions through PDSA cycles and shared their learning through Dropbox over the span of 26 submission weeks beginning in June 2020 through June 2021. There were 253 unique data submission, with 213 submissions coming after the relaunch. Teams submitted a total of 77 unique PDSAs over the course of the iLab and each team was successful in establishing a process that resulted in achievement of the initial iLab aim. All teams had developed standardised processes for real-time recognition of FIES/SIES shortly after the iLab official relaunch in August 2020 and this was sustained throughout the iLab. A median centreline value of 73.4% for real-time assessment and recognition of FIES was achieved by the end of the iLab (figure 2). Half of the iLab teams (6/12) reached a centreline value>70% median, with a quarter (3/12) reaching >85% median by the end of the iLab.

Assessment of FIES increased from 18.6% to 73.4% over the course of the iLab, and of those PwCF with FIES assessment, 16.7% on average experienced an FIES event (online supplemental appendix G). All teams improved in the assessment and recognition of FIES. The median magnitude of change achieved by the participating teams was 38.1%. The change magnitudes noticed in different teams ranged between 29.7% and 86.9% with a mean of 40.8%. Teams reported 21.3% of encounters had SIES assessment and 8% had an SIES event.

A total of 6944 clinic encounters were assessed for either FIES or SIES with some PwCF being assessed for both, *despite the intent of the iLab that SIES use only be reported in the absence of available spirometry testing. This limited some data analysis on the analysis of the first smart aim.*

On average, 70.5% of PwCF who were assessed for FIES and had an FEV_1pp decline outside of the green zone had recommendation for follow-up within 6 weeks in accordance with the treatment algorithm (figure 3. This finding was consistent from early on in the conduct of the iLab possibly indicating that iLab sites were early adopters of a 'close and early follow-up model of care' for PEx.

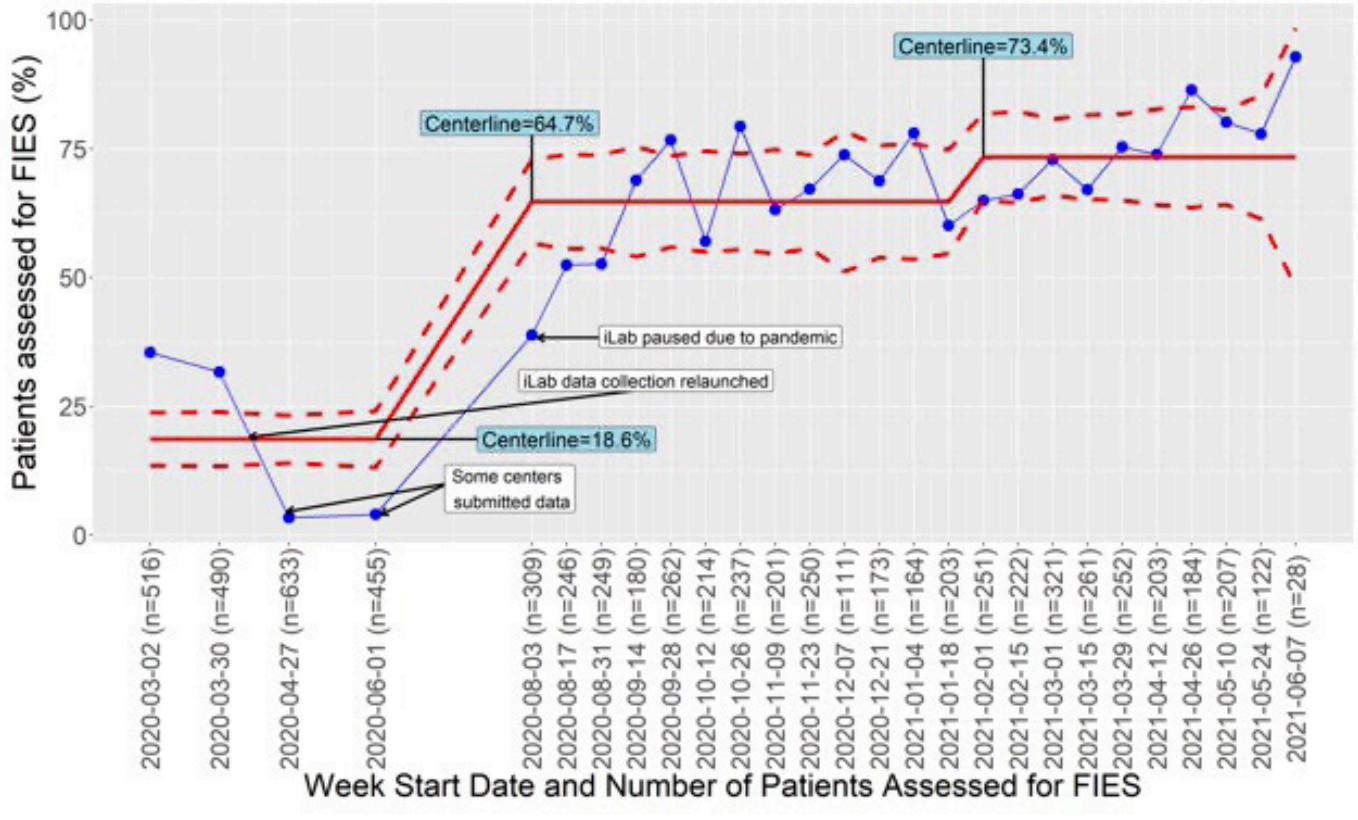

Figure 2 Percentage of eligible encounters with people with cystic fibrosis assessed for forced expiratory volume in one second indicated exacerbation signal (FIES).

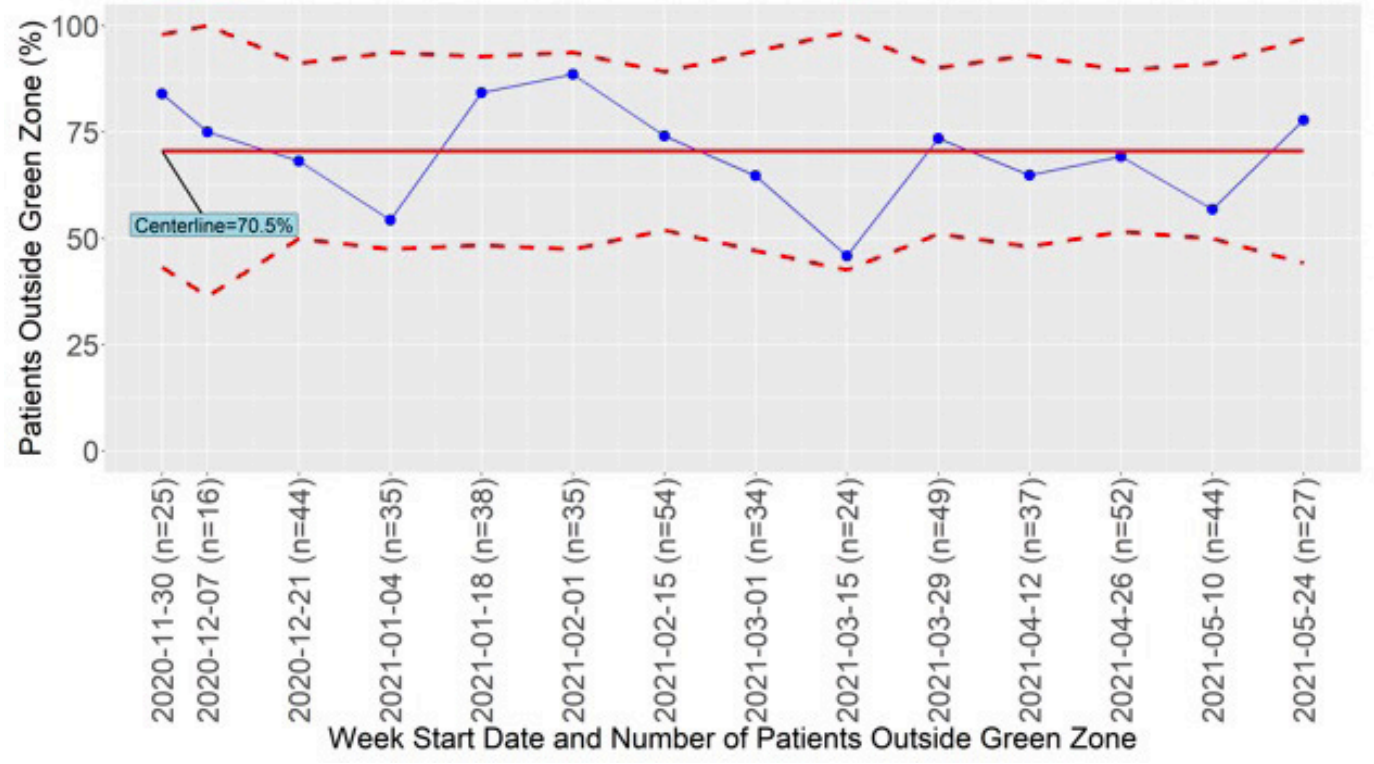

Figure 3 Percentage of people with cystic fibrosis who experienced forced expiratory volume in one second (FEV_1) per cent predicted decline outside of the green zone of the FEV_1-indicated exacerbation signal algorithm with a 6-week follow-up recommendation.

DISCUSSION

The FIES iLab was successful in developing and implementing a new tool for early identification of PEx in PwCF. Every team increased the percentage of PwCF assessed for exacerbations using a standardised exacerbation signal (FIES or SIES), supporting the global aim of decreasing lung function decline. Strengths of this initiative included the cohort of participating teams and the construct of the iLab itself. CFLN teams participating in the FIES iLab teams were interdisciplinary, inclusive of PFPs, experienced and supported in QI work and accustomed to the collaborative CFLN environment. The FIES iLab teams were adept in collaborating with each other; routinely participating in huddles, sharing their work through process mapping and consistently submitting data on PDSAs. The iLab construct provided the framework for rapid tests of change, shared learning, supported data collection and collation of results, and facilitated the rapid adaptation required with the interruption of a global pandemic. This process may be implemented by other CF teams, depending on their local resources and QI knowledge.

There were multiple barriers to the initial aim of assessing FIES during the pandemic. Most barriers were related to clinic and pulmonary function test laboratories closures in the context of governmental and institutional imposed COVID restrictions and the subsequent transition to telehealth, and heterogeneous adoption of home spirometry in telehealth by both PwCF and providers.[14–16] FIES assessment and recognition required an FEV_1 measurement. Even with telehealth visits and the option of FEV_1pp measurement obtained with home spirometers, there were challenges. During the pandemic, precipitous clinic closures, care team member redeployments and home spirometry device shortages created significant limitations on this option.[16 17] These limitations were especially challenging for paediatric CF centres in which, FEV_1pp measurement requires accurate height measures and age-appropriate coaching and education which was often not available.[17] In addition, there were multiple barriers to SIES implementation, including adoption of a standard symptom scoring tool and consensus on implementation strategies and differences in reporting between parents of children with CF and adults with CF. As a result, SIES administration was not standardised among centres, which limited the ability to further analyse this tool.

With the opening of society between mid-2020 and the beginning of 2021, clinic spirometry became more easily available and FIES recognition increased. PEx signals, which could be conflated with asthma exacerbation, poor technique, etc, required clinical assessment and validation to inform treatment decisions. Therefore, the proxy measure of follow-up within 6 weeks was chosen to demonstrate adherence to the algorithm and ensure that all acute or subacute declines in lung function were consistently pursued until resolution. Follow-up in response to FIES, outside of the green zone, was collected rigorously. However, no true baseline data were available to show the

magnitude of improvement in follow-up. Adoption of SIES assessment and follow-up across centres proved to be challenging due to lack of a consensus regarding the definition of exacerbation based on SIES responses.

Recall bias and inconsistencies of teams in reporting to REDCap survey also created some data limitations. Specifically, the treatment algorithm required centres to report follow-up for all patients 'outside of the green zone' (lung function decline >5% decline for PwCF with baseline values at or greater than 50% FEV_1pp and >3% absolute decline for those with baseline values less than 50% FEV_1pp). For this reason, interpretation of improvement in follow-up care only for those considered to have an FIES event (>10% decline in lung function) was not possible.

Additionally, the acute stress on medical teams during the pandemic, loss of team members, and competing priorities to address the need for rapid changes in healthcare delivery, impacted the time available for QI and diluted the focus of this initiative. Certainly, these factors contributed to the variability in success of the iLab between centres. Variations between centres based on geographic location, population served (adult vs children), adoption of telehealth as an alternative to clinic visits, access to home spirometers and timing to resuming in-person visits also played a part in the variation of results (online supplemental appendix H).

The global aim and primary outcome for the iLab was to decrease the rate of lung function decline in the iLab cohort of centres. This outcome was difficult to assess over a short period of time. However, the consistent assessment and recognition of exacerbation signals at every clinic encounter, in combination with high, consistent use of the treatment algorithm, were also identified as important iLab outcomes.

An additional factor influencing this initiative was the Food and Drug Administration approval of the highly effective modulator, tezacaftor/ivacaftor/elexacaftor, at the end of 2019. Many PwCF started this treatment during 2020 at the start of the iLab. This treatment led to significant decline in exacerbation rates and improved lung function.[20] The pandemic further influenced exacerbation rates, with multiple PwCF experiencing less exacerbations due to reduced social interactions and decreased exposure to all respiratory viruses, including COVID-19.[21] While these influences and challenges existed concurrently with the iLab, they highlight the remarkable dedication of iLab teams who continued to collaborate on this project, recognising its importance and impact beyond the COVID-19 pandemic and the introduction of new CF therapies.

After completing the iLab in July 2021, individual centres were encouraged to continue monitoring reliability of their ingrained processes. While those measures are outside the scope of this report, sustainability is an area of interest and could be explored in the future through analysis of the CF Patient Registry data. In addition, further studies are planned to fully assess the impact of this work on long-term lung function decline.

CONCLUSION

This study was successful in the use of an exacerbation tool for assessment and recognition of potential PEx, and routine application of a PEx algorithm for early follow-up of lung function decline. A template of the process or change package generated by this study can potentially be shared and spread to the entire CF Care Center Network. Implementation will require team education on FIES recognition and use, robust resources and QI expertise. Further study needed to evaluate the impact on patient outcomes, especially on FEV_1 decline, is underway.

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Patient and public involvement Patients and/or the public were involved in the design, or conduct, or reporting, or dissemination plans of this research. Refer to the Methods section for further details.

Patient consent for publication Not applicable.

Ethics approval There were no ethical concerns related to our study since the goal was to reliably implant best practice. This project was considered quality improvement and thus did not require human subject review.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available upon reasonable request. Additional tables and figures were produced relative to this manuscript which are available upon request. Raw data may be obtained from a third party and are not publicly available.

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