

BMJ Open Quality Improving lung function in adolescents with CF by tracking annual rate of lung function decline

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ABSTRACT

Background For patients with cystic fibrosis (CF), sustaining lung function through the adolescent years is crucial to slow the progressive decline that leads to significant morbidity and early mortality. This holds true for patients with high per cent predicted forced expiratory volume in 1 s (ppFEV₁), as they may receive less vigilant monitoring and treatment. Early identification of lung function decline followed by aggressive treatment can lead to preservation of lung function.

Intervention The Emory+Children's Pediatric Cystic Fibrosis Program implemented multiple quality improvement (QI) initiatives to identify and aggressively treat adolescent patients with a rapid decline in lung function. These initiatives included (1) lung zones to categorise and highlight lung function decline, (2) individual lung decline tables for quick reference, (3) a lung health algorithm to encourage uniformity, (4) a rapid decliner checklist to identify potential reasons for individual decline and (5) an automated individual patient-level data report and centre scorecard. We tested these interventions with plan–do–study–act cycles and refined as needed.

Results Implementation of these QI initiatives resulted in overall improvement in lung function and slowing of lung function decline among adolescents with CF. This improvement could be attributed to the more standardised and proactive approach to decreases in lung function and the increased clinician attention to patients with rapid decline, especially for patients with high baseline ppFEV₁.

INTRODUCTION

Cystic fibrosis (CF) is a chronic, life-limiting genetic condition that leads to progressive lung disease over time. Mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene cause mucus in the airways to be dehydrated, leading to various complications including infections, inflammation, lung injury and respiratory failure.¹ The clinical course is also characterised by acute pulmonary exacerbations, or intermittent worsening of signs and symptoms that are often accompanied by a decline in lung function.² It is vital to identify and treat these exacerbations as early as possible to slow the progression of lung disease, as they have a significant impact on mortality,³ quality of life⁴

and long-term lung function.⁵ Consistent and aggressive treatment of pulmonary exacerbation is associated with improved outcomes and preserving lung function.^{6 7} Additionally, progressive CF pulmonary disease may lead to steady decline in lung function over time. This decline may be more clinically challenging to identify but also results in advancing lung disease.

Although life expectancy is increasing for patients with CF, there remains a mortality peak in early adulthood due to progressive pulmonary disease with respiratory failure.⁸ This pulmonary decline may be slowed with aggressive monitoring of per cent predicted forced expiratory volume in 1 s (ppFEV₁) and intervention during the paediatric and adolescent years. Patients with a high baseline ppFEV₁ may receive less vigilant monitoring despite having rapid rates of lung function decline.^{9 10}

Data from the Emory+Children's Pediatric Cystic Fibrosis Program showed below-average lung function in 2014 and 2015 in our adolescent population. We undertook a quality improvement (QI) initiative to track lung function decline and implement interventions based on QI principles. Our global aim was to reduce the annual rate of decline in ppFEV₁ and improve median lung function among our adolescents with CF. Our SMART aim was to improve ppFEV₁ among adolescents 13–17 years of age from a baseline of 86% in 2016 to 90% (above the national average of 88%) in 3 years.

METHODS

Context

The Children's Healthcare of Atlanta and Emory University Cystic Fibrosis Care Center, located in Atlanta, Georgia, provides care for almost 700 patients with CF and is one of the largest specialised care centres for this disease in the USA. The centre is accredited by the Cystic Fibrosis Foundation and one of only two CF care centres in Georgia. At the time

this QI project was initiated in 2016, the Emory+Children's Pediatric Program provided clinical care for nearly 200 children and adolescents with CF. Of these, about 47% were male and 88% were white. Of note, the percentage of black patients followed up at our programme is approximately twice the national average for paediatric programmes. The most common mutation among our patients was F508del, with nearly 90% of patients having at least one copy. F508del is the most frequently identified CF mutation and is a protein processing mutation that results in little to no functional CFTR channel at the cell surface and classic CF disease.¹ In 2016, 24% of patients at our programme <18 years of age were infected with *Pseudomonas aeruginosa*; 21% were infected with methicillin-resistant *Staphylococcus aureus*; and 35% had one or more pulmonary exacerbations per year. These characteristics were fairly typical of the national CF population except for our exacerbation frequency, which was 10% higher than the national average of 25%. Among adolescents aged 10–17 years, approximately 15% had a diagnosis of CF-related diabetes (national average 13%). CF-related diabetes is a complication associated with a more rapid rate of lung function decline and increased mortality.¹¹ Interdisciplinary CF care at our clinic is provided by a team that consists of physicians, nurse practitioners, nurses, respiratory therapists (RTs), dietitians and social workers. Our programme has a strong focus on QI with biweekly full team QI meetings.

Patient and public involvement

Patients and the public were not involved in the design of these QI interventions. As part of this project, patients and families were educated about their individual baseline lung function and rate of decline. The project design and results were presented to patients and families at our annual CF education day in 2018.

Interventions

Study of the interventions

We followed the model for improvement¹² methodology by using a key driver diagram (figure 1) and plan–do–study–act (PDSA) cycles to implement and refine the following interventions to reduce the annual rate of decline in forced expiratory volume in 1 s (FEV₁) and improve ppFEV₁ for our adolescents with CF.

PDSA cycle 1 (January 2017)

We created and assigned *lung zones* to individual adolescent patients with CF based on their rate of lung function decline over time. An individual's baseline ppFEV₁ was calculated for each year as the average of the best value of each quarter, according to the method used for the CF Foundation's annual programme reports. We selected this definition to allow us to benchmark to the national averages calculated using this method. Additionally, our tracked data would match and predict the final data we received in our CF Foundation benchmarking report annually. Using each individual's yearly baseline ppFEV₁, we calculated the annual rate of decline (%) from 2013 to 2016. Patients with a history of lung transplant were excluded. A total of 69 eligible adolescents had data for the time period of 2013–2016 for whom we could calculate a 3-year rate of decline. Quarterly best ppFEV₁ values were manually extracted from our pulmonary function test (PFT) software by our CF RTs and entered into a spreadsheet that calculated baseline lung function and rate of decline. Mean annual rate of decline was -2.1% /year (SD 3.3), similar to published reports.¹³ However, significant variability in rates of decline was noted: 25% (n=17) had no decline in ppFEV₁, while the bottom quartile had rates of decline of -3.7% to -13.3% per year. A review of risk factors for rapid decline in the initial 69 adolescents with CF revealed that patients with CF-related diabetes diagnosed by 2013 had significantly faster

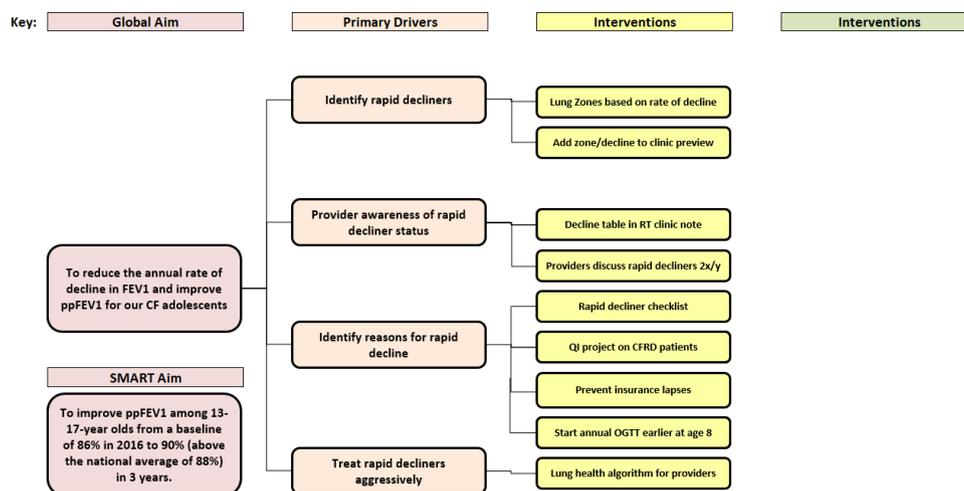


Figure 1 Driver diagram created to display factors that contribute to the achievement of our global aim. Further evaluation of rapid decliners revealed that CF-related diabetes and Medicaid insurance were risk factors for rapid decline in our adolescent population, which led to additional QI efforts. CF, cystic fibrosis; ppFEV₁, per cent predicted forced expiratory volume in 1 s; QI, quality improvement; RT, respiratory therapist; CFRD, cystic fibrosis related diabetes; OGTT, oral glucose tolerance test.

Year	FEV ₁ Liters	Baseline ppFEV ₁	Rate of decline in ppFEV ₁
2013	1.6	106%	
2014	1.8	102%	-4%
2015	1.9	94%	-8%
2016	2.0	90%	-4%

Lung Zone: D
 Best FEV₁ 2016: 2.1L, 91%
 Average annual rate of decline: -5%/year

Figure 2 Lung decline table included in the clinic note to highlight rate of decline and corresponding lung zone. FEV₁, forced expiratory volume in 1 s, ppFEV₁, per cent predicted forced expiratory volume in 1 s.

decline in ppFEV₁ (-5.1% vs -1.5%/year), as did patients with Medicaid insurance compared with private insurance (-3.5% vs -1.2%/year). These risk factors were therefore incorporated into our driver diagram and led to additional QI efforts at our programme. Lung zones were assigned based on approximate quartiles for our centre's 2013–2016 rate of decline in ppFEV₁. Many of our initiatives focused efforts on patients with accelerated decline in lung function (faster than average), who fall into the lowest two quartiles, the C and D zones, known as 'rapid decliners'.

- A: no decline (or improvement).
- B: decline of 0.01%–2.0%/year.
- C: decline of 2.01%–3.5%/year.
- D: decline >3.5%/year.

The CF interdisciplinary team has a weekly clinic preview meeting to discuss patients with upcoming appointments in the next week. We added the 2016 ppFEV₁ baseline, annual rate of decline and assigned lung zone to the weekly discussion sheet to highlight patients in zones C/D for provider awareness and team discussion at clinic preview.

PDSA cycle 2/3 (February and March 2017)

The CF RT created a *lung decline table* (figure 2), based off the lung function spreadsheet. This table was added to the RT clinic note for each patient, so the provider would not have to reference the lung function spreadsheet, making it easier to access and act on this information during clinic visits. We refined this cycle by encouraging providers to copy this table into their own clinic note to reference, also making it easier for an admitting hospital team to find the data and review.

PDSA cycle 4 (April 2017)

To encourage uniformity in care and focus attention on rapid decliners, we created a *lung health algorithm* for our providers (online supplemental figure 1), which includes an acute pulmonary exacerbation protocol. Based on the CF Foundation's benchmarking analysis of top-performing CF centres, it was found that more frequent follow-up and early, aggressive intervention of lung function decline were associated with improved outcomes.¹⁴ We used this to guide the creation of our algorithm to ensure rapid decliners received

appropriate, step-up investigations and treatments and closer follow-up. The algorithm was available for providers to use on all patients but focused on those in the C/D lung zones to align with the benchmarked best practices. The algorithm was discussed and agreed on by the full interdisciplinary team at a QI meeting.

PDSA cycle 5/6 (April 2017)

To identify potential reasons for individual lung function decline, we created a *rapid decliner checklist* (online supplemental figure 2). We determined that a checklist would be beneficial, as checklists play a vital role in improving patient safety and adherence to evidence-based best practices throughout the healthcare system.^{15 16} A few members of the team met to create the checklist by brainstorming potential contributing factors to rapid decline with corresponding corrective actions and then reviewed it with the entire team to obtain feedback for improvement and achieve consensus.

The multidisciplinary team reviewed the checklist every 6 months during clinic preview for all patients in the C/D lung zones. We refined this cycle in 2018 to review the rapid decliner checklist annually during a QI meeting to ensure optimal provider attendance and involvement. Prior to the meeting, the providers receive the list of the patients in the C/D lung zones and respond with comments about potential reasons for each patient's decline. The comments are reconciled into one document to discuss during the meeting and plan next step interventions for each patient.

PDSA cycle 7 (November 2019)

To improve our process of manually collecting annual baseline data to calculate the rate of decline, we partnered with our performance analytics team to create an *automated report and scorecard*. With this report, each patient's quarterly best ppFEV₁ is directly pulled from our CF registry data and the annual baseline is automatically calculated. The scorecard includes quarterly and year-to-date median ppFEV₁ metrics for the entire CF programme with corresponding run charts. The scorecard is shared quarterly with the multidisciplinary CF team.

MEASURES

Outcome measures

Since this QI initiative is focused on improving lung function, we chose our clinical outcome measures to be the cohort's annual and quarterly median ppFEV₁. FEV₁ is defined as the maximum amount of air one can forcefully exhale in the first second. This value is converted to the per cent predicted, based off the individual's age, height, weight and race.¹⁷ We tracked these outcome measures for several groups: group aged 13–17 years, group aged 12–21 years and a longitudinal cohort of 49 adolescents. We tracked individual annual baseline ppFEV₁ and individual average annual rate of decline in ppFEV₁ as part of this project. We also tracked the per cent of 'rapid

decliner' patients in the C and D zones each year as an additional outcome measure.

Process measures

Cycle-specific process measures are described in the PDSA results as follows.

RESULTS

PDSA cycle 1 (January 2017): calculated baseline lung function, created and assigned lung zones, added baseline, annual rate of decline and lung zone to the discussion sheet for clinic preview

To ensure our annual baselines could be accurately calculated from quarterly FEV₁ data, we tracked the per cent of patients with a quarterly FEV₁ missing. A quarterly FEV₁ is defined as the best FEV₁ that meets American Thoracic Society (ATS) standards during that quarter's clinic visit. We were then able to cross-reference any missing data with corresponding appointment data to validate. All (100%) of those patients with a missing quarterly FEV₁ truly did not have a PFT that quarter that met ATS standards. Of the adolescent patients for whom lung function data were available for the 2013–2016 time period (n=69), 100% had a rate of decline calculated and a lung zone assigned. Twenty of the 69 patients transitioned to adult care during early 2017 due to the retirement of a provider or moved away; therefore, 49 patients continued to receive care at our programme throughout 2017 and were exposed to the interventions (our longitudinal cohort). The baseline, annual rate of decline and lung zone were added to the discussion sheet for clinic preview 100% of the time. The cycle was also studied via qualitative feedback at our weekly multidisciplinary CF team clinic preview and QI meetings to obtain input on the usefulness of the intervention from the team. The RT reported this process was easily integrated into her clinic preview workflow. However, we found that adding the baseline, annual rate of decline and lung zone to the clinic preview discussion sheet was not as helpful as expected, as some providers do not consistently attend the clinic preview meeting, and there was not always time during clinic preview to discuss the lung decline and zone. Additionally, the team often forgot to bring the discussion sheet to the clinic, making it hard to use the data and address the patient's rate of decline during the visit. The RT continued to include this information on the discussion sheet, but we refined this process by creating a table to include in the RT note, so providers had access to it during clinic.

PDSA cycle 2/3 (February and March 2017): lung function decline table

Copying the lung function decline table into the RT's note for the team to review allowed the providers to access it during outpatient clinics. The RT notes contained the lung function decline table for 98% of patients in 2017 for our longitudinal cohort of 49 patients. The RT qualitatively reported this was straightforward and efficient to add to her workflow. When the CF RT was out, we noted

that the covering RTs did not consistently include the table. Thus, we worked toward developing a system where a single RT would serve as the back-up RT for CF clinic or the CF RT would prepare the tables before she was out. However, the location of the table in the RT note made it difficult for the outpatient providers and hospital providers to find the table during future encounters, as the RT notes are not as prominent as provider notes in our electronic medical record. Copying this table into the outpatient CF provider's note made it easier for clinic providers to find it and for an admitting team to review it on a hospital admission. Once the outpatient provider copies the table at the beginning of the year, it stays there for the remainder of the year, making it much easier to reference during clinic visits. The provider notes contained the lung function decline table 86% of the time in 2017. We did not require this, but most providers felt it was helpful and improved communication and completed it.

PDSA cycle 4 (April 2017): lung health algorithm

This algorithm encouraged consistency among providers by standardising education for patients and families, assessment for contribution factors of lung function decline, acute pulmonary exacerbation treatment, and chronic treatment and follow-up. Due to limited resources and our large patient population, we were not able to quantitatively track adherence to the algorithm. We studied this cycle by reviewing the lung health algorithm during QI meetings for qualitative feedback from the multidisciplinary team. We found that some aspects of the algorithm were difficult to implement, including rapid decliners not calling the clinic 5–7 days after starting antibiotics nor coming in for more frequent follow-up visits.

PDSA cycle 5/6 (April 2017): rapid decliner checklist

After the first year of this intervention, 54% of rapid decliner C/D zone patients had a checklist completed, and only one patient had two checklists completed. We found it difficult to review the checklist every 6 months during the interdisciplinary clinic preview meeting due to suboptimal provider attendance and the team forgetting to bring the checklists. Additionally, since the checklist review was dependent on patients having a scheduled appointment in the next week, then patients that cancelled their appointment would not have the opportunity to have their checklist completed until they rescheduled. Changing this process to review the checklist annually during the QI meeting resulted in greater provider attendance and ensured that all rapid decliners were discussed as a team (an improvement on PDSA 1). Allowing the providers to review the patients ahead of time allowed for efficient discussions.

PDSA cycle 7 (November 2019): automated report and scorecard

Creating a report that automatically pulls quarterly bests and baselines makes the process of calculating the annual

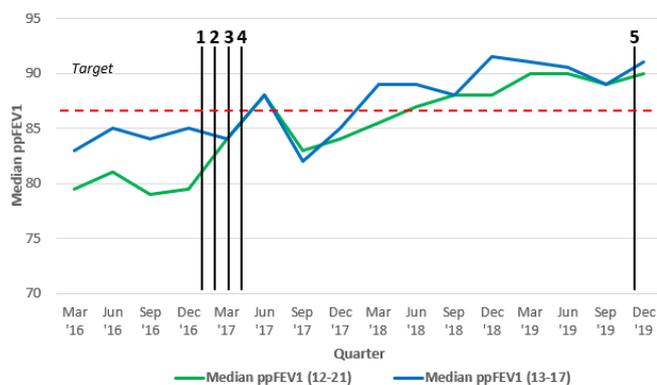


Figure 3 Median ppFEV₁ by quarter for adolescents. 1 (January 2017), SMART aim established; began putting baseline, rate of decline and lung zone into clinic preview document; began tracking quarterly median ppFEV₁ for 13–17 and 12–21; 2 (February 2017), lung decline tables added to respiratory therapist note; 3 (March 2017), lung decline tables added to provider note; 4 (April 2017), rapid decline checklist implemented, lung health algorithm finalised; 5 (November 2019), automated report and scorecard finalised. ppFEV₁, per cent predicted forced expiratory volume in 1 s.

rates of decline with corresponding lung zones faster due to the reduced manual component. The scorecard allows us to track our centre's quarterly and year-to-date median ppFEV₁ over time. The run charts promote easy data visualisation when sharing metrics with the CF team. The scorecard has run successfully for 83% of the quarters since initiation; we did not run the scorecard in quarter 2 of 2020 due to extremely limited in-clinic visits during the COVID-19 pandemic. Qualitative feedback on this report has been positive, and it has decreased the manual tracking burden on the CF team.

Quantitative results

To determine if our initiatives were successful, we tracked our outcome measure (median ppFEV₁) quarterly by age group (13–17 and 12–21 years of age) in a cross-sectional manner (figure 3). Our median ppFEV₁ in both age groups showed substantial improvement over time, reaching our target of 90% in 2019. We subsequently decided to expand these interventions outside of the original adolescent cohort to younger patients and to track the results by age group (including children 6–12 years of age).

We also looked at our longitudinal cohort of 49 patients 8–18 years of age as of 1 January 2013 who had at least one ppFEV₁ value each year from 2013 to 2016 and received care at our institution in 2017 (exposed to the QI interventions). In this group, the median ppFEV₁ increased from 81.3 to 83.3 in 2017 (figure 4). The mean rate of decline in ppFEV₁ was +0.7% in 2017, and 45% (n=22) of the patients improved their baseline ppFEV₁ with a median improvement of +3.6%. The mean annual rate of decline improved from –2.1%/year for 2013–2016 to –1.3%/year for 2014–2017, –1%/year for 2015–2018 and

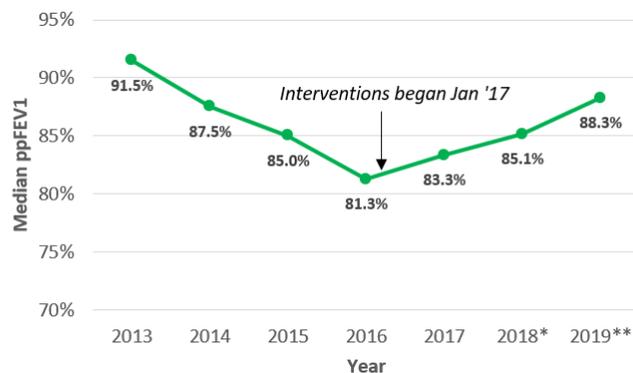


Figure 4 Annual median ppFEV₁ for the longitudinal cohort of 49 adolescents with cystic fibrosis. *n=46, **n=40. ppFEV₁, per cent predicted forced expiratory volume in 1 s.

–1%/year for 2016–2019. Even though the size of the cohort continued to drop due to transitions into adult care, we continued to calculate median ppFEV₁ and rates of decline for the longitudinal cohort in 2018 and 2019 and saw sustained improvements in median lung function for the remaining individuals. This longitudinal approach was unique because the same individuals were tracked over multiple years, and on average showed improvement in lung function instead of the gradual decline that would be expected with the natural course of CF lung disease progression. We also examined the per cent of patients in the C and D zones each year for this longitudinal cohort, and this measure of rapid decliners decreased overall from the start of the project (figure 5).

DISCUSSION

Among adolescent patients with CF, continually tracking individual annual baseline ppFEV₁ and rate of lung function decline led to improved identification of rapid decliners and implementation of more aggressive treatments. In our patient cohorts, these interventions resulted in improvements in median lung function and reduction in mean rate of decline. If maintained, these changes are

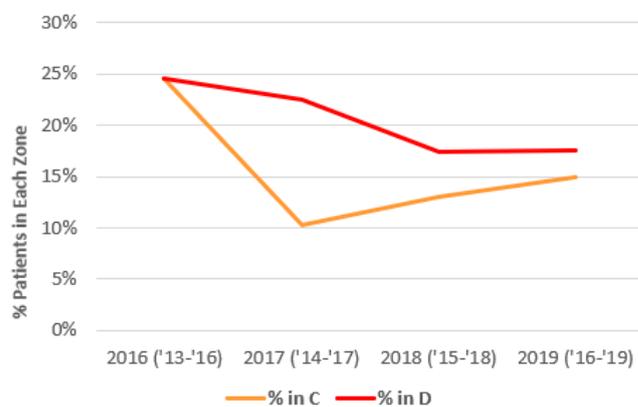


Figure 5 Per cent of adolescent patients with cystic fibrosis in the C and D lung decline zones (rapid decliners) each year (n=49, longitudinal cohort).

anticipated to result in slower progression of lung disease and increasing survival. These improvements may be attributable to a more standardised and proactive approach to decreases in lung function and increased clinician attention to patients with rapid decline, especially those with high baseline lung function whose decline could be more clinically silent. Of our interventions, we believe the lung function decline tables in the RT notes and annually discussing individualised reasons for rapid decline had the largest impact on improvement. This highlighted lung function decline for providers and allowed them to identify rapid decliners and intervene. A lesson learnt was that these interventions require provider and team buy-in and involvement to be successful. We were lucky to have excellent team buy-in from the beginning of our project. Our team was very motivated to improve after learning that our programme's lung function was below average in 2014 and 2015; we feel this buy-in contributed to our successes. We found benchmarking top-performing centres extremely beneficial, as it stimulated these QI initiatives and allowed us to choose appropriate goals. We also learnt the importance of coproduction with our patients and families. Although we did not have patient involvement at the time we designed this project, we now have two parent partners who are active members of our QI team. As a next step, we are expanding these interventions beyond the original cohort of teens to include all patients with at least 2 years of PFT data. Although few patients in our studied cohort were on highly effective CFTR modulator medications, the sustainability of these interventions will have to be modified due to the expansion in modulator therapy availability for CF and the corresponding increase in baseline lung function. Once patients reach their new baseline lung function on highly effective modulator therapy, we will be able to meaningfully track rate of decline again.

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Patient consent for publication Not required.

Ethics approval The Children's Healthcare of Atlanta's institutional review board determined the project was not a research activity and the initiative was given a not-human research determination.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Due to the nature of this research, participants of this study did not agree for their data to be shared publicly, so supporting data is not available. No data are available.

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Adolescent Lung Health Algorithm for CF Patients ≥ 12 years

Lung Decline Zone	Interventions
<p>A: No decline or improvement in ppFEV1</p> <p>B: ppFEV1 rate of decline* 0.01 – 2.0%/yr</p>	<p>Education</p> <ul style="list-style-type: none"> Educate on baseline[^] and best[§] FEV1 values <p>Assess for contributing factors</p> <ul style="list-style-type: none"> Routine monitoring of lung function Routine respiratory culture at each visit and AFB culture annually in sputum producers Routine annual IgE level to screen for ABPA Routine annual RT visit Routine weight monitoring and annual nutrition visit Routine annual OGTT in non-diabetic patients Routine blood glucose monitoring and every 3 month CFRD clinic visits in diabetic patients Routine annual social work visit Routine annual mental health screening <p>Acute pulmonary exacerbations</p> <ul style="list-style-type: none"> Increase airway clearance and hypertonic saline to 3-4 times per day and oral or IV antibiotics as indicated Instruct patient to contact nurse if not improving after 5-7 days of oral antibiotics <ul style="list-style-type: none"> If not improved, switch antibiotics if there is potential for covering different organism; if not, admit for IV antibiotics If change in FEV1 $\leq 5\%$, provider to use clinical judgement to decide need for additional follow-up PFTs If drop in FEV1 $> 5\%$, return to clinic 2-3 weeks after starting outpatient treatment for repeat PFTs <ul style="list-style-type: none"> Provider to give patient the FEV1 goal[†] needed to avoid admission ahead of time Admit for IV antibiotics if FEV1 not back to goal or new lung exam findings have not resolved If FEV1 not back to goal or new lung exam findings have not resolved with IV antibiotics, consider 3rd week of IV therapy with change in antibiotics and re-evaluate potential contributing factors using rapid decline checklist If FEV1 not back to goal or new lung exam findings have not resolved with IV antibiotics, consider prescribing 2 week oral antibiotic course after IV antibiotics (with continued 3-4x/day airway clearance while on antibiotics) to maintain pulmonary exacerbation treatment after discharge Hospital follow-up 4-6 weeks after discharge <p>Chronic treatments and Follow up</p> <ul style="list-style-type: none"> Guideline based treatments Standard follow-up every 3 months
<p>C: FEV1 PP rate of decline* 2.01 – 3.5%/yr</p>	<p>Education</p> <ul style="list-style-type: none"> Educate on baseline[^] and best[§] FEV1 values Educate on FEV1 rate of decline* Encourage patient to brainstorm reasons for decline

	<p>Assess for contributing factors</p> <ul style="list-style-type: none"> • Routine multidisciplinary monitoring and evaluations as above • <i>Use rapid decline checklist to assess potential contributing factors</i> • <i>Consider additional/more frequent evaluation based on checklist</i> <ul style="list-style-type: none"> ○ <i>GI, ENT, Endocrine, nutrition, social work, mental health, RT, PT</i> <p>Acute pulmonary exacerbations</p> <ul style="list-style-type: none"> • Routine exacerbation treatment as above • <i>Consider hospital follow-up clinic visit 2-4 weeks after discharge</i> <p>Chronic treatments and Follow up</p> <ul style="list-style-type: none"> • <i>Ensure using hypertonic saline and dornase alfa</i> • <i>Consider starting Azithromycin even if no Pseudomonas, especially if any bronchiectasis on Chest CT</i> • <i>Consider more frequent clinic follow-up every 6-8 weeks, particularly if FEV1 appears to be declining</i>
<p>D: FEV1 PP rate of decline* > 3.5%/yr</p>	<p>Education</p> <ul style="list-style-type: none"> • Educate on baseline[^] and best[§] FEV1 values • Educate on FEV1 rate of decline* • Encourage patient to brainstorm reasons for decline <p>Assess for contributing factors</p> <ul style="list-style-type: none"> • Routine multidisciplinary monitoring and evaluations as above • Use rapid decline checklist to assess potential contributing factors • Consider additional/more frequent evaluation based on checklist <ul style="list-style-type: none"> ○ GI, ENT, Endocrine, nutrition, social work, mental health, RT, PT • <i>Consider home outreach call by team member</i> <p>Acute pulmonary exacerbations</p> <ul style="list-style-type: none"> • Routine exacerbation treatment as above • <i>Hospital follow-up 2-4 weeks after discharge</i> <p>Chronic treatments and Follow up</p> <ul style="list-style-type: none"> • Ensure using hypertonic saline and dornase alfa • Consider starting Azithromycin even if no Pseudomonas, especially if any bronchiectasis on Chest CT • <i>Consider polysomnography if baseline FEV1 <65% to assess for nocturnal desaturations</i> • <i>Consider nocturnal non-invasive bilevel positive airway pressure for lung recruitment and optimization of airway clearance via collateral ventilation</i> • <i>Consider chronic suppressive alternating inhaled antibiotics and/or oral antibiotics, especially if cultures persistently positive for organisms</i> • <i>Consider home nursing</i> • <i>Follow-up every 6 weeks until FEV1 improves or stabilizes</i>

[^]Baseline FEV1: average of the best FEV1 in each quarter from the last annual year

[§]Best FEV1: best FEV1 from the last annual year

*FEV1 rate of decline: Average yearly change in baseline FEV1 % predicted

*Goal is at the discretion of the provider

CF Rapid Lung Function Decline Checklist

To be completed every 12 months for Lung Zones C and D

<u>Contributors to Rapid Decline</u>	<u>Yes</u>	<u>Maybe</u>	<u>No</u>	<u>Possible Actions</u>
<u>Sinopulmonary</u>				
Progressive pulmonary disease	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Ensure on dornase alfa, hypertonic saline, CFTR modulator; consider chest CT, Azithromycin, admission
New airway pathogen	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Consider bronchoscopy
Unusual airway pathogen	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Consider infectious disease consultation
Ineffective airway clearance	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional RT evaluation (e.g huff cough practice, different method)
Low physical activity/exercise capacity	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Physical therapy evaluation, exercise prescription
Allergic bronchopulmonary aspergillosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Check IgE and/or ABPA panel
Sinus Disease	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Sinus CT and/or ENT, allergy evaluation
Asthma	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Pre-post bronchodilator spirometry
<u>GI/Endocrine</u>				
Aspiration/GERD/Delayed gastric emptying	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	GI consultation, pH/impedance probe
Nutritional compromise	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional dietitian evaluation, appetite stimulant
CFRD/Impaired glucose tolerance	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	HbA1C, OGTT, endocrine consultation, CGM
<u>Psychosocial</u>				
Mental health (Depression, Anxiety, ADHD)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional SW/mental health coordinator evaluation and referrals
Substance use/abuse	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional SW/mental health coordinator evaluation
Smoke/irritant exposure	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional occupational history, SW evaluation
Non-adherence	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional SW/mental health evaluation, use adherence/MI tool
Missed appointments	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional SW evaluation
Financial barriers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Additional SW evaluation
Lack of disease understanding	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	Education activity/Transition program, pharmacist education