

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