

<b>Table I. Cystic Fibrosis Foundation (CFF) Pulmonary Guidelines – Chronic Medications for Maintenance of Lung Health</b>					
Therapy	Indication	Dose	Level of Evidence	Net benefit	Grade*
Inhaled tobramycin	Age >6 years AND Moderate to severe lung disease (FEV1 % predicted 40-69%) AND persistent <i>Pseudomonas aeruginosa</i> on culture	300 mg via nebulization BID	Good	Substantial	A
	Age >6 years AND asymptomatic OR mild lung disease (FEV1% predicted >89%) AND persistent <i>Pseudomonas aeruginosa</i> on culture	300 mg via nebulization BID	Fair	Moderate	B
Other inhaled antibiotics†	Insufficient evidence to recommend for or against routine usage		Poor	Small	I
Recombinant human DNase	Age > 6 years AND Moderate to severe lung disease (FEV1 % predicted 40-69%)	2.5 mg nebulized once daily	Good	Substantial	A
	Age >6 years AND asymptomatic OR mild lung disease (FEV1% predicted >89%)	2.5 mg nebulized once daily	Fair	Moderate	B
Hypertonic Saline (7%)	Age >6 years	4 ml via nebulizer twice daily	Fair	Moderate	B
Inhaled corticosteroids	Age >6 years without asthma or ABPA – recommends against routine usage		Fair	Zero	D
Oral corticosteroids	Ages 6-18 years without asthma or ABPA, recommends against chronic use		Good	Negative	D
	Adult patients (>18 years of age) without asthma or ABPA, Insufficient evidence to recommend for or against routine usage		Poor	Zero	I
Oral NSAIDs	Age >6 years AND FEV1% predicted >60%)	Varied based on pharmacokinetics	Fair	Moderate	B
Leukotriene modifiers	Insufficient evidence to recommend for or against routine usage		Poor	Zero	I
Cromolyn	Insufficient evidence to recommend for or against routine usage		Poor	Zero	I
Macrolide antibiotics‡	Age >6 years	<40 kg, 250 mg thrice weekly >40 kg 500 mg thrice weekly	Fair	Substantial	B
Antistaphylococcal Antibiotics	Recommends against prophylactic use		Fair	Negative	D
Bronchodilators	Age > 6 years	Vary based on brand	Good	Moderate	B
Inhaled anticholinergics	Insufficient evidence to recommend for or against routine usage		Poor	Small	I
N-acetylcysteine	Insufficient evidence to recommend for or against routine usage		Poor	Zero	I

**\*U.S Preventative Service Task Force Grades**

† Note: Aztreonam lysine for inhalation (Oermann, Retsch-Bogart et al. 2010) was not FDA approved at time of CFF recommendations

‡ Note: At time of CFF recommendations, (Saiman, Anstead et al. 2010) data about use of macrolides in patients without *Pseudomonas aeruginosa* was not available