Michael W Konstan

List of Publications by Citations

202 papers	18,092 citations	69 h-index	132 g-index
209 ext. papers	20,674 ext. citations	6.6 avg, IF	6.28 L-index

#	Paper	IF	Citations
202	A CFTR potentiator in patients with cystic fibrosis and the G551D mutation. *New England Journal of Medicine*, **2011**, 365, 1663-72	59.2	1465
201	Lumacaftor-Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508del CFTR. *New England Journal of Medicine*, **2015**, 373, 220-31	59.2	910
200	Cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **1996**, 154, 1229-56	10.2	778
199	Effect of high-dose ibuprofen in patients with cystic fibrosis. *New England Journal of Medicine*, **1995** , 332, 848-54	59.2	632
198	Effect of VX-770 in persons with cystic fibrosis and the G551D-CFTR mutation. *New England Journal of Medicine*, **2010**, 363, 1991-2003	59.2	598
197	Inflammatory cytokines in cystic fibrosis lungs. *American Journal of Respiratory and Critical Care Medicine*, **1995**, 152, 2111-8	10.2	595
196	Bronchoalveolar lavage findings in cystic fibrosis patients with stable, clinically mild lung disease suggest ongoing infection and inflammation. *American Journal of Respiratory and Critical Care Medicine*, **1994**, 150, 448-54	10.2	447
195	Results of a phase IIa study of VX-809, an investigational CFTR corrector compound, in subjects with cystic fibrosis homozygous for the F508del-CFTR mutation. *Thorax*, **2012**, 67, 12-8	7.3	408
194	Genetic modifiers of lung disease in cystic fibrosis. *New England Journal of Medicine*, **2005**, 353, 1443-53	59.2	368
193	Risk factors for rate of decline in forced expiratory volume in one second in children and adolescents with cystic fibrosis. *Journal of Pediatrics*, **2007**, 151, 134-9, 139.e1	3.6	328
192	A CFTR corrector (lumacaftor) and a CFTR potentiator (ivacaftor) for treatment of patients with cystic fibrosis who have a phe508del CFTR mutation: a phase 2 randomised controlled trial. *Lancet Respiratory Medicine,the*, **2014**, 2, 527-38	35.1	309
191	Normal bronchial epithelial cells constitutively produce the anti-inflammatory cytokine interleukin-10, which is downregulated in cystic fibrosis. *American Journal of Respiratory Cell and Molecular Biology*, **1995**, 13, 257-61	5.7	297
190	Growth and nutritional indexes in early life predict pulmonary function in cystic fibrosis. *Journal of Pediatrics*, **2003**, 142, 624-30	3.6	292
189	Inflammation in cystic fibrosis lung disease: Pathogenesis and therapy. *Journal of Cystic Fibrosis*, **2015**, 14, 419-30	4.1	276
188	Significant microbiological effect of inhaled tobramycin in young children with cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **2003**, 167, 841-9	10.2	257
187	Association between respiratory tract methicillin-resistant Staphylococcus aureus and survival in cystic fibrosis. *JAMA - Journal of the American Medical Association*, **2010**, 303, 2386-92	27.4	253
186	Ataluren for the treatment of nonsense-mutation cystic fibrosis: a randomised, double-blind, placebo-controlled phase 3 trial. *Lancet Respiratory Medicine,the*, **2014**, 2, 539-47	35.1	242

185	Current understanding of the inflammatory process in cystic fibrosis: onset and etiology. *Pediatric Pulmonology*, **1997**, 24, 137-42; discussion 159-61	3.5	242
184	Compacted DNA nanoparticles administered to the nasal mucosa of cystic fibrosis subjects are safe and demonstrate partial to complete cystic fibrosis transmembrane regulator reconstitution. *Human Gene Therapy*, **2004**, 15, 1255-69	4.8	240
183	Safety, efficacy and convenience of tobramycin inhalation powder in cystic fibrosis patients: The EAGER trial. *Journal of Cystic Fibrosis*, **2011**, 10, 54-61	4.1	237
182	A two-year randomized, placebo-controlled trial of dornase alfa in young patients with cystic fibrosis with mild lung function abnormalities. *Journal of Pediatrics*, **2001**, 139, 813-20	3.6	235
181	Altered respiratory epithelial cell cytokine production in cystic fibrosis. *Journal of Allergy and Clinical Immunology*, **1999**, 104, 72-8	11.5	221
180	Excessive inflammatory response of cystic fibrosis mice to bronchopulmonary infection with Pseudomonas aeruginosa. *Journal of Clinical Investigation*, **1997**, 100, 2810-5	15.9	211
179	The role of inflammation in the pathophysiology of CF lung disease. *Clinical Reviews in Allergy and Immunology*, **2002**, 23, 5-27	12.3	180
178	Association between pulmonary function and sputum biomarkers in cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **2007**, 175, 822-8	10.2	175
177	Phase II studies of nebulised Arikace in CF patients with Pseudomonas aeruginosa infection. *Thorax*, **2013**, 68, 818-25	7.3	174
176	Whole-exome sequencing identifies rare and low-frequency coding variants associated with LDL cholesterol. *American Journal of Human Genetics*, **2014**, 94, 233-45	11	170
175	Factors influencing outcomes in cystic fibrosis: a center-based analysis. *Chest*, **2003**, 123, 20-7	5.3	169
174	Assessment of safety and efficacy of long-term treatment with combination lumacaftor and ivacaftor therapy in patients with cystic fibrosis homozygous for the F508del-CFTR mutation (PROGRESS): a phase 3, extension study. *Lancet Respiratory Medicine,the*, **2017**, 5, 107-118	35.1	158
173	Sustained Benefit from ivacaftor demonstrated by combining clinical trial and cystic fibrosis patient registry data. *American Journal of Respiratory and Critical Care Medicine*, **2015**, 192, 836-42	10.2	158
172	Long-term safety and efficacy of ivacaftor in patients with cystic fibrosis who have the Gly551Asp-CFTR mutation: a phase 3, open-label extension study (PERSIST). *Lancet Respiratory Medicine,the*, **2014**, 2, 902-910	35.1	150
171	Novel tobramycin inhalation powder in cystic fibrosis subjects: pharmacokinetics and safety. *Pediatric Pulmonology*, **2007**, 42, 307-13	3.5	150
170	Efficacy of the Flutter device for airway mucus clearance in patients with cystic fibrosis. *Journal of Pediatrics*, **1994**, 124, 689-93	3.6	148
169	Epidemiologic study of cystic fibrosis: design and implementation of a prospective, multicenter, observational study of patients with cystic fibrosis in the U.S. and Canada. *Pediatric Pulmonology*, **1999**, 28, 231-41	3.5	147
168	Clinical use of Ibuprofen is associated with slower FEV1 decline in children with cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **2007**, 176, 1084-9	10.2	144

167	Pulmonary exacerbations in cystic fibrosis. *Pediatric Pulmonology*, **2004**, 37, 400-6	3.5	141
166	Leukotriene B4 markedly elevated in the epithelial lining fluid of patients with cystic fibrosis. *The American Review of Respiratory Disease*, **1993**, 148, 896-901		140
165	Impact of pregnancy on women with cystic fibrosis. *Chest*, **2006**, 129, 706-11	5.3	134
164	Sputum biomarkers of inflammation in cystic fibrosis lung disease. *Proceedings of the American Thoracic Society*, **2007**, 4, 406-17		122
163	Ibuprofen attenuates the inflammatory response to Pseudomonas aeruginosa in a rat model of chronic pulmonary infection. Implications for antiinflammatory therapy in cystic fibrosis. *The American Review of Respiratory Disease*, **1990**, 141, 186-92		118
162	Tobramycin inhalation powder for P. aeruginosa infection in cystic fibrosis: the EVOLVE trial. *Pediatric Pulmonology*, **2011**, 46, 230-8	3.5	114
161	Presence of methicillin resistant Staphylococcus aureus in respiratory cultures from cystic fibrosis patients is associated with lower lung function. *Pediatric Pulmonology*, **2007**, 42, 513-8	3.5	111
160	IL-10 attenuates excessive inflammation in chronic Pseudomonas infection in mice. *American Journal of Respiratory and Critical Care Medicine*, **1999**, 160, 2040-7	10.2	108
159	Efficacy and safety of lumacaftor/ivacaftor combination therapy in patients with cystic fibrosis homozygous for Phe508del CFTR by pulmonary function subgroup: a pooled analysis. *Lancet Respiratory Medicine,the*, **2016**, 4, 617-626	35.1	100
158	Treatment complexity in cystic fibrosis: trends over time and associations with site-specific outcomes. *Journal of Cystic Fibrosis*, **2013**, 12, 461-7	4.1	95
157	Sweat chloride as a biomarker of CFTR activity: proof of concept and ivacaftor clinical trial data. *Journal of Cystic Fibrosis*, **2014**, 13, 139-47	4.1	95
156	Ibuprofen in children with cystic fibrosis: pharmacokinetics and adverse effects. *Journal of Pediatrics*, **1991**, 118, 956-64	3.6	94
155	A randomized double blind, placebo controlled phase 2 trial of BIIL 284 BS (an LTB4 receptor antagonist) for the treatment of lung disease in children and adults with cystic fibrosis. *Journal of Cystic Fibrosis*, **2014**, 13, 148-55	4.1	93
154	Effect of Pseudomonas infection on weight loss, lung mechanics, and cytokines in mice. *American Journal of Respiratory and Critical Care Medicine*, **2000**, 161, 271-9	10.2	91
153	Jointly modelling the relationship between survival and pulmonary function in cystic fibrosis patients. *Statistics in Medicine*, **2002**, 21, 1271-87	2.3	86
152	Safety and early treatment effects of the CXCR2 antagonist SB-656933 in patients with cystic fibrosis. *Journal of Cystic Fibrosis*, **2013**, 12, 241-8	4.1	84
151	Risk factors for rate of decline in FEV1 in adults with cystic fibrosis. *Journal of Cystic Fibrosis*, **2012**, 11, 405-11	4.1	83
150	Data from the US and UK cystic fibrosis registries support disease modification by CFTR modulation with ivacaftor. *Thorax*, **2018**, 73, 731-740	7.3	82

149	Oral, inhaled, and intravenous antibiotic choice for treating pulmonary exacerbations in cystic fibrosis. *Pediatric Pulmonology*, **2013**, 48, 666-73	3.5	82
148	Year-to-year changes in lung function in individuals with cystic fibrosis. *Journal of Cystic Fibrosis*, **2010**, 9, 250-6	4.1	81
147	Measuring and improving respiratory outcomes in cystic fibrosis lung disease: opportunities and challenges to therapy. *Journal of Cystic Fibrosis*, **2010**, 9, 1-16	4.1	80
146	Duration of treatment effect after tobramycin solution for inhalation in young children with cystic fibrosis. *Pediatric Pulmonology*, **2007**, 42, 610-23	3.5	79
145	Standardized procedure for measurement of nasal potential difference: an outcome measure in multicenter cystic fibrosis clinical trials. *Pediatric Pulmonology*, **2004**, 37, 385-92	3.5	79
144	The use of anti-inflammatory medications in cystic fibrosis: trends and physician attitudes. *Chest*, **1999**, 115, 1053-8	5.3	79
143	Inflammation and anti-inflammatory therapies for cystic fibrosis. *Clinics in Chest Medicine*, **2007**, 28, 331-46	3.5	78
142	Lung function decline from adolescence to young adulthood in cystic fibrosis. *Pediatric Pulmonology*, **2012**, 47, 135-43	3.5	77
141	Disease progression in patients with cystic fibrosis treated with ivacaftor: Data from national US and UK registries. *Journal of Cystic Fibrosis*, **2020**, 19, 68-79	4.1	76
140	Emergence of linezolid-resistant Staphylococcus aureus after prolonged treatment of cystic fibrosis patients in Cleveland, Ohio. *Antimicrobial Agents and Chemotherapy*, **2011**, 55, 1684-92	5.9	74
139	Anti-PcrV antibody in cystic fibrosis: a novel approach targeting Pseudomonas aeruginosa airway infection. *Pediatric Pulmonology*, **2014**, 49, 650-8	3.5	73
138	Association of socioeconomic status with the use of chronic therapies and healthcare utilization in children with cystic fibrosis. *Journal of Pediatrics*, **2009**, 155, 634-9.e1-4	3.6	73
137	Classifying severity of cystic fibrosis lung disease using longitudinal pulmonary function data. *American Journal of Respiratory and Critical Care Medicine*, **2006**, 174, 780-6	10.2	73
136	The efficacy and safety of meropenem and tobramycin vs ceftazidime and tobramycin in the treatment of acute pulmonary exacerbations in patients with cystic fibrosis. *Chest*, **2005**, 128, 2336-46	5.3	72
135	Patterns of medical practice in cystic fibrosis: part II. Use of therapies. Investigators and Coordinators of the Epidemiologic Study of Cystic Fibrosis. *Pediatric Pulmonology*, **1999**, 28, 248-54	3.5	72
134	Effect of ibuprofen on neutrophil migration in vivo in cystic fibrosis and healthy subjects. *Journal of Pharmacology and Experimental Therapeutics*, **2003**, 306, 1086-91	4.7	71
133	Identifying treatments that halt progression of pulmonary disease in cystic fibrosis. *Pediatric Research*, **1997**, 41, 161-5	3.2	69
132	Psychometric evaluation of the Cystic Fibrosis Questionnaire-Revised in a national sample. *Quality of Life Research*, **2012**, 21, 1267-78	3.7	67

131	Clinical use of dornase alpha is associated with a slower rate of FEV1 decline in cystic fibrosis. *Pediatric Pulmonology*, **2011**, 46, 545-53	3.5	66
130	Relationship between inhaled corticosteroid therapy and rate of lung function decline in children with cystic fibrosis. *Journal of Pediatrics*, **2008**, 153, 746-51	3.6	65
129	Prolonged inflammatory response to acute Pseudomonas challenge in interleukin-10 knockout mice. *American Journal of Respiratory and Critical Care Medicine*, **2002**, 165, 1176-81	10.2	65
128	Pharmacological approaches for the discovery and development of new anti-inflammatory agents for the treatment of cystic fibrosis. *Advanced Drug Delivery Reviews*, **2002**, 54, 1409-23	18.5	64
127	Serum and lower respiratory tract drug concentrations after tobramycin inhalation in young children with cystic fibrosis. *Journal of Pediatrics*, **2001**, 139, 572-7	3.6	62
126	Elastin and collagen degradation products in urine of patients with cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **1995**, 152, 157-62	10.2	57
125	Lumacaftor/Ivacaftor Treatment of Patients with Cystic Fibrosis Heterozygous for F508del-CFTR. *Annals of the American Thoracic Society*, **2017**, 14, 213-219	4.7	57
124	Effect of dornase alfa on inflammation and lung function: potential role in the early treatment of cystic fibrosis. *Journal of Cystic Fibrosis*, **2012**, 11, 78-83	4.1	52
123	Use of nasal potential difference and sweat chloride as outcome measures in multicenter clinical trials in subjects with cystic fibrosis. *Pediatric Pulmonology*, **2002**, 33, 142-50	3.5	52
122	Impact of CFTR Modulation on Intestinal pH, Motility, and Clinical Outcomes in Patients With Cystic Fibrosis and the G551D Mutation. *Clinical and Translational Gastroenterology*, **2017**, 8, e81	4.2	50
121	Shifting patterns of inhaled antibiotic use in cystic fibrosis. *Pediatric Pulmonology*, **2008**, 43, 874-81	3.5	50
120	Ibuprofen therapy for cystic fibrosis lung disease: revisited. *Current Opinion in Pulmonary Medicine*, **2008**, 14, 567-73	3	49
119	Infant care patterns at epidemiologic study of cystic fibrosis sites that achieve superior childhood lung function. *Pediatrics*, **2007**, 119, e531-7	7.4	48
118	BIIL 284 reduces neutrophil numbers but increases P. aeruginosa bacteremia and inflammation in mouse lungs. *Journal of Cystic Fibrosis*, **2014**, 13, 156-63	4.1	47
117	Trends in the use of routine therapies in cystic fibrosis: 1995-2005. *Pediatric Pulmonology*, **2010**, 45, 1167-72	3.5	47
116	Study of a novel pancreatic enzyme replacement therapy in pancreatic insufficient subjects with cystic fibrosis. *Journal of Pediatrics*, **2006**, 149, 658-662	3.6	45
115	Risk factors for mortality before age 18 years in cystic fibrosis. *Pediatric Pulmonology*, **2017**, 52, 909-915	3.5	44
114	An international randomized multicenter comparison of nasal potential difference techniques. *Chest*, **2010**, 138, 919-28	5.3	44

113	Use of ibuprofen to assess inflammatory biomarkers in induced sputum: Implications for clinical trials in cystic fibrosis. *Journal of Cystic Fibrosis*, **2015**, 14, 720-6	4.1	43
112	Assessing time to pulmonary function benefit following antibiotic treatment of acute cystic fibrosis exacerbations. *Respiratory Research*, **2010**, 11, 137	7.3	43
111	Multiple antibiotic-resistant Pseudomonas aeruginosa and lung function decline in patients with cystic fibrosis. *Journal of Cystic Fibrosis*, **2012**, 11, 293-9	4.1	42
110	A phase I randomized, multicenter trial of CPX in adult subjects with mild cystic fibrosis. *Pediatric Pulmonology*, **2002**, 33, 90-8	3.5	42
109	KB001-A, a novel anti-inflammatory, found to be safe and well-tolerated in cystic fibrosis patients infected with Pseudomonas aeruginosa. *Journal of Cystic Fibrosis*, **2018**, 17, 484-491	4.1	41
108	Characterizing aggressiveness and predicting future progression of CF lung disease. *Journal of Cystic Fibrosis*, **2009**, 8 Suppl 1, S15-9	4.1	41
107	Antibiotic and anti-inflammatory therapies for cystic fibrosis. *Cold Spring Harbor Perspectives in Medicine*, **2013**, 3, a009779	5.4	40
106	Association of High-Dose Ibuprofen Use, Lung Function Decline, and Long-Term Survival in Children with Cystic Fibrosis. *Annals of the American Thoracic Society*, **2018**, 15, 485-493	4.7	38
105	Lessons learned from a randomized trial of airway secretion clearance techniques in cystic fibrosis. *Pediatric Pulmonology*, **2010**, 45, 291-300	3.5	37
104	Long-term effects of pregnancy and motherhood on disease outcomes of women with cystic fibrosis. *Annals of the American Thoracic Society*, **2013**, 10, 213-9	4.7	36
103	Safety and preliminary clinical activity of a novel pancreatic enzyme preparation in pancreatic insufficient cystic fibrosis patients. *Pancreas*, **2006**, 32, 258-63	2.6	36
102	Design and powering of cystic fibrosis clinical trials using rate of FEV(1) decline as an efficacy endpoint. *Journal of Cystic Fibrosis*, **2010**, 9, 332-8	4.1	35
101	Relation of sweat chloride concentration to severity of lung disease in cystic fibrosis. *Pediatric Pulmonology*, **2004**, 38, 204-9	3.5	35
100	Patterns of medical practice in cystic fibrosis: part I. Evaluation and monitoring of health status of patients. Investigators and Coordinators of the Epidemiologic Study of Cystic Fibrosis. *Pediatric Pulmonology*, **1999**, 28, 242-7	3.5	35
99	Pulmonary outcome prediction (POP) tools for cystic fibrosis patients. *Pediatric Pulmonology*, **2010**, 45, 1156-66	3.5	34
98	Tissue-specific Fc gamma and complement receptor expression by alveolar macrophages determines relative importance of IgG and complement in promoting phagocytosis of Pseudomonas aeruginosa. *Pediatric Research*, **1994**, 35, 68-77	3.2	34
97	Antibiotic treatment of signs and symptoms of pulmonary exacerbations: a comparison by care site. *Pediatric Pulmonology*, **2015**, 50, 431-40	3.5	33
96	Forced Expiratory Volume in 1 Second Variability Helps Identify Patients with Cystic Fibrosis at Risk of Greater Loss of Lung Function. *Journal of Pediatrics*, **2016**, 169, 116-21.e2	3.6	33

95	Pulmonary exacerbations in cystic fibrosis: young children with characteristic signs and symptoms. *Pediatric Pulmonology*, **2013**, 48, 649-57	3.5	33
94	Tobramycin inhalation powder manufactured by improved process in cystic fibrosis: the randomized EDIT trial. *Current Medical Research and Opinion*, **2013**, 29, 947-56	2.5	32
93	The role of inhaled corticosteroids in the management of cystic fibrosis. *Paediatric Drugs*, **2009**, 11, 101-13	4.2	31
92	Transfer of the human Alpha1-antitrypsin gene into pulmonary macrophages in vivo. *American Journal of Respiratory Cell and Molecular Biology*, **1998**, 18, 591-601	5.7	31
91	Pitfall in the use of genotype analysis as the sole diagnostic criterion for cystic fibrosis. *Pediatrics*, **1999**, 103, 823-6	7.4	31
90	IV-treated pulmonary exacerbations in the prior year: An important independent risk factor for future pulmonary exacerbation in cystic fibrosis. *Journal of Cystic Fibrosis*, **2016**, 15, 372-9	4.1	31
89	Treatment and demographic factors affecting time to next pulmonary exacerbation in cystic fibrosis. *Journal of Cystic Fibrosis*, **2015**, 14, 763-9	4.1	30
88	Anti-inflammatory therapies for cystic fibrosis-related lung disease. *Clinical Reviews in Allergy and Immunology*, **2008**, 35, 135-53	12.3	30
87	Safety of aerosolized INS 365 in patients with mild to moderate cystic fibrosis: results of a phase I multi-center study. *Pediatric Pulmonology*, **2001**, 32, 122-8	3.5	30
86	Coefficients of fat and nitrogen absorption in healthy subjects and individuals with cystic fibrosis. *Journal of Pediatric Pharmacology and Therapeutics*, **2007**, 12, 47-52	1.6	29
85	Considerations for the Conduct of Clinical Trials with Antiinflammatory Agents in Cystic Fibrosis. A Cystic Fibrosis Foundation Workshop Report. *Annals of the American Thoracic Society*, **2015**, 12, 1398-406	4.7	28
84	Outcome measures for clinical trials assessing treatment of cystic fibrosis lung disease. *Clinical Investigation*, **2012**, 2, 163-175		28
83	Effect of smaller droplet size of dornase alfa on lung function in mild cystic fibrosis. Dornase Alfa Nebulizer Group. *Pediatric Pulmonology*, **1998**, 25, 83-7	3.5	28
82	Therapies aimed at airway inflammation in cystic fibrosis. *Clinics in Chest Medicine*, **1998**, 19, 505-13, vi	5.3	28
81	Randomized trial of efficacy and safety of dornase alfa delivered by eRapid nebulizer in cystic fibrosis patients. *Journal of Cystic Fibrosis*, **2015**, 14, 777-83	4.1	27
80	Design and powering of cystic fibrosis clinical trials using pulmonary exacerbation as an efficacy endpoint. *Journal of Cystic Fibrosis*, **2011**, 10, 453-9	4.1	27
79	Socioeconomic status and the likelihood of antibiotic treatment for signs and symptoms of pulmonary exacerbation in children with cystic fibrosis. *Journal of Pediatrics*, **2011**, 159, 819-824.e1	3.6	27
78	Anti-inflammatory medications for cystic fibrosis lung disease: selecting the most appropriate agent. *Treatments in Respiratory Medicine*, **2005**, 4, 255-73		27

77	Efficacy and safety of ataluren in patients with nonsense-mutation cystic fibrosis not receiving chronic inhaled aminoglycosides: The international, randomized, double-blind, placebo-controlled Ataluren Confirmatory Trial in Cystic Fibrosis (ACT CF). *Journal of Cystic Fibrosis*, **2020**, 19, 595-601	4.1	26
76	Probability of IV antibiotic retreatment within thirty days is associated with duration and location of IV antibiotic treatment for pulmonary exacerbation in cystic fibrosis. *Journal of Cystic Fibrosis*, **2016**, 15, 783-790	4.1	26
75	Lumacaftor/Ivacaftor reduces pulmonary exacerbations in patients irrespective of initial changes in FEV. *Journal of Cystic Fibrosis*, **2019**, 18, 94-101	4.1	26
74	A multi-center controlled trial of growth hormone treatment in children with cystic fibrosis. *Pediatric Pulmonology*, **2012**, 47, 252-63	3.5	26
73	Non-viral gene transfer therapy for cystic fibrosis. *Expert Opinion on Biological Therapy*, **2003**, 3, 449-58	5.4	26
72	Relationship of Antibiotic Treatment to Recovery after Acute FEV Decline in Children with Cystic Fibrosis. *Annals of the American Thoracic Society*, **2017**, 14, 937-942	4.7	25
71	Probability of treatment following acute decline in lung function in children with cystic fibrosis is related to baseline pulmonary function. *Journal of Pediatrics*, **2013**, 163, 1152-7.e2	3.6	25
70	Standard care versus protocol based therapy for new onset Pseudomonas aeruginosa in cystic fibrosis. *Pediatric Pulmonology*, **2013**, 48, 943-53	3.5	25
69	Estimating effectiveness in an observational study: a case study of dornase alfa in cystic fibrosis. The Investigators and Coordinators of the Epidemiologic Study of Cystic Fibrosis. *Journal of Pediatrics*, **1999**, 134, 734-9	3.6	24
68	Cross-sectional and longitudinal studies of naturally occurring antibodies to Pseudomonas aeruginosa in cystic fibrosis indicate absence of antibody-mediated protection and decline in opsonic quality after infection. *Journal of Infectious Diseases*, **1995**, 172, 453-61	7	24
67	Behavioral and nutritional treatment for preschool-aged children with cystic fibrosis: a randomized clinical trial. *JAMA Pediatrics*, **2015**, 169, e150636	8.3	23
66	A pipeline of therapies for cystic fibrosis. *Seminars in Respiratory and Critical Care Medicine*, **2009**, 30, 611-26	3.9	22
65	Ultrase MT12 and Ultrase MT20 in the treatment of exocrine pancreatic insufficiency in cystic fibrosis: safety and efficacy. *Alimentary Pharmacology and Therapeutics*, **2004**, 20, 1365-71	6.1	22
64	Association between the introduction of a new cystic fibrosis inhaled antibiotic class and change in prevalence of patients receiving multiple inhaled antibiotic classes. *Journal of Cystic Fibrosis*, **2015**, 14, 370-5	4.1	21
63	Treatment Setting and Outcomes of Cystic Fibrosis Pulmonary Exacerbations. *Annals of the American Thoracic Society*, **2018**, 15, 225-233	4.7	21
62	Improvements in lung function and height among cohorts of 6-year-olds with cystic fibrosis from 1994 to 2012. *Journal of Pediatrics*, **2014**, 165, 1091-1097.e2	3.6	20
61	The pathologic characteristics of interstitial pneumonia cystic fibrosis. A retrospective autopsy study. *American Journal of Clinical Pathology*, **1989**, 91, 522-30	1.9	19
60	Amikacin liposome inhalation suspension for chronic Pseudomonas aeruginosa infection in cystic fibrosis. *Journal of Cystic Fibrosis*, **2020**, 19, 284-291	4.1	19

59	Early childhood wheezing is associated with lower lung function in cystic fibrosis. *Pediatric Pulmonology*, **2014**, 49, 745-50	3.5	18
58	Trends in the clinical characteristics of the U.S. cystic fibrosis patient population from 1995 to 2005. *Pediatric Pulmonology*, **2008**, 43, 739-44	3.5	17
57	Clinical use of tobramycin inhalation solution (TOBI) shows sustained improvement in FEV1 in cystic fibrosis. *Pediatric Pulmonology*, **2014**, 49, 529-36	3.5	16
56	Changing thresholds and incidence of antibiotic treatment of cystic fibrosis pulmonary exacerbations, 1995-2005. *Journal of Cystic Fibrosis*, **2013**, 12, 332-7	4.1	16
55	Beta 2 adrenergic receptor polymorphisms in cystic fibrosis. *Pediatric Pulmonology*, **2005**, 39, 544-50	3.5	15
54	Normalized T1 magnetic resonance imaging for assessment of regional lung function in adult cystic fibrosis patients--a cross-sectional study. *PLoS ONE*, **2013**, 8, e73286	3.7	15
53	Modeling long-term health outcomes of patients with cystic fibrosis homozygous for F508del-CFTR treated with lumacaftor/ivacaftor. *Therapeutic Advances in Respiratory Disease*, **2019**, 13, 1753466618820186	4.0	14
52	Inflammatory mediators in CF patients. *Methods in Molecular Medicine*, **2002**, 70, 409-31		14
51	Patients with Cystic Fibrosis and a G551D or Homozygous F508del Mutation: Similar Lung Function Decline. *American Journal of Respiratory and Critical Care Medicine*, **2017**, 195, 1673-1676	10.2	13
50	One-year safety and efficacy of tobramycin powder for inhalation in patients with cystic fibrosis. *Pediatric Pulmonology*, **2016**, 51, 372-8	3.5	13
49	Decline in lung function does not predict future decline in lung function in cystic fibrosis patients. *Pediatric Pulmonology*, **2015**, 50, 856-62	3.5	13
48	Efficacy response in CF patients treated with ivacaftor: post-hoc analysis. *Pediatric Pulmonology*, **2015**, 50, 447-55	3.5	12
47	Risk factors for onset of persistent respiratory symptoms in children with cystic fibrosis. *Pediatric Pulmonology*, **2012**, 47, 966-72	3.5	12
46	Efficacy and safety of a unique enteric-coated bicarbonate-buffered pancreatic enzyme replacement therapy in children and adults with cystic fibrosis. *Clinical Investigation*, **2013**, 3, 723-729		12
45	A comparative study of lysozyme and its secretion by tracheal epithelium. *Experimental Lung Research*, **1982**, 3, 175-81	2.3	12
44	Preliminary comparison of normalized T1 and non-contrast perfusion MRI assessments of regional lung disease in cystic fibrosis patients. *Journal of Cystic Fibrosis*, **2017**, 16, 283-290	4.1	11
43	BMI fails to identify poor nutritional status in stunted children with CF. *Journal of Cystic Fibrosis*, **2017**, 16, 158-160	4.1	11
42	Pulmonary function outcomes for assessing cystic fibrosis care. *Journal of Cystic Fibrosis*, **2015**, 14, 376-82	4.1	11

41	The HEALing (Helping to End Addiction Long-term) Communities Study: Protocol for a cluster randomized trial at the community level to reduce opioid overdose deaths through implementation of an integrated set of evidence-based practices. *Drug and Alcohol Dependence*, **2020**, 217, 108335	4.9	11
40	Reopening Schools Safely: The Case for Collaboration, Constructive Disruption of Pre-Coronavirus 2019 Expectations, and Creative Solutions. *Journal of Pediatrics*, **2020**, 223, 183-185	3.6	11
39	Transition to adulthood and adult health care for patients with sickle cell disease or cystic fibrosis: Current practices and research priorities. *Journal of Clinical and Translational Science*, **2018**, 2, 334-342	0.4	11
38	Lung function decline is delayed but not decreased in patients with cystic fibrosis and the R117H gene mutation. *Journal of Cystic Fibrosis*, **2018**, 17, 503-510	4.1	10
37	Pulmonary exacerbations and acute declines in lung function in patients with cystic fibrosis. *Journal of Cystic Fibrosis*, **2018**, 17, 496-502	4.1	9
36	Evaluating the Impact of Stopping Chronic Therapies after Modulator Drug Therapy in Cystic Fibrosis: The SIMPLIFY Clinical Trial Study Design. *Annals of the American Thoracic Society*, **2021**, 18, 1397-1405	4.7	9
35	Building global development strategies for cf therapeutics during a transitional cftr modulator era. *Journal of Cystic Fibrosis*, **2020**, 19, 677-687	4.1	8
34	Implementation of the first worldwide quality assurance program for cystic fibrosis multiple mutation detection in population-based screening. *Clinica Chimica Acta*, **2011**, 412, 1376-81	6.2	8
33	Efficacy and Safety of a New Formulation of Pancrelipase (Ultrase MT20) in the Treatment of Malabsorption in Exocrine Pancreatic Insufficiency in Cystic Fibrosis. *Gastroenterology Research and Practice*, **2010**, 2010, 898193	2	8
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