Steven M Rowe, Msph

List of Publications by Year
in descending order

179 papers	20,225 citations	27035 58 h-index	12940 136 g-index
185 all docs	185 docs citations	185 times ranked	13277 citing authors

#	Article	IF	Citations
1	Effect of lumacaftor-ivacaftor on mucociliary clearance and clinical outcomes in cystic fibrosis: Results from the PROSPECT MCC sub-study. Journal of Cystic Fibrosis, 2022, 21, 143-145.	0.3	12
2	Ivacaftor restores delayed mucociliary transport caused by Pseudomonas aeruginosaâ€“induced acquired cystic fibrosis transmembrane conductance regulator dysfunction in rabbit nasal epithelia. International Forum of Allergy and Rhinology, 2022, 12, 690-698.	1.5	7
3	Inhaled gene therapy of preclinical muco-obstructive lung diseases by nanoparticles capable of breaching the airway mucus barrier. Thorax, 2022, 77, 812-820.	2.7	9
4	Clinical Effectiveness of Elexacaftor/Tezacaftor/Ivacaftor in People with Cystic Fibrosis: A Clinical Trial. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 529-539.	2.5	147
5	Cystic Fibrosis Transmembrane Conductance Regulator: Roles in Chronic Obstructive Pulmonary Disease. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 631-640.	2.5	18
6	Poly (acetyl, arginyl) glucosamine disrupts Pseudomonas aeruginosa biofilms and enhances bacterial clearance in a rat lung infection model. Microbiology (United Kingdom), 2022, 168, .	0.7	4
7	Static mucus impairs bacterial clearance and allows chronic infection withPseudomonas aeruginosain the cystic fibrosis rat. European Respiratory Journal, 2022, 60, 2101032.	3.1	10
8	Human distal airways contain a multipotent secretory cell that can regenerate alveoli. Nature, 2022, 604, 120-126.	13.7	128
9	Reply to: Change in Lung Function After Initiation of Elexacaftor-tezacaftor-ivacaftor: Do Not Forget Anatomy!. American Journal of Respiratory and Critical Care Medicine, 2022, , .	2.5	0
10	Evaluation of a novel CFTR potentiator in COPD ferrets with acquired CFTR dysfunction. European Respiratory Journal, 2022, 60, 2101581.	3.1	10
11	Effects of ivacaftor on systemic inflammation and the plasma proteome in people with CF and G551D. Journal of Cystic Fibrosis, 2022, 21, 950-958.	0.3	9
12	Changes in Glucose Breath Test in Cystic Fibrosis Patients Treated With 1 Month of Lumacaftor/Ivacaftor. Journal of Pediatric Gastroenterology and Nutrition, 2022, 75, 42-47.	0.9	3
13	Tobacco smoke exposure limits the therapeutic benefit of tezacaftor/ivacaftor in pediatric patients with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 612-617.	0.3	21
14	Clinical Effectiveness of Lumacaftor/Ivacaftor in Patients with Cystic Fibrosis Homozygous for F508del-CFTR. A Clinical Trial. Annals of the American Thoracic Society, 2021, 18, 75-83.	1.5	32
15	Lumacaftor/ivacaftor therapy fails to increase insulin secretion in F508del/F508del CF patients. Journal of Cystic Fibrosis, 2021, 20, 333-338.	0.3	40
16	Measuring the impact of CFTR modulation on sweat chloride in cystic fibrosis: Rationale and design of the CHEC-SC study. Journal of Cystic Fibrosis, 2021, 20, 965-971.	0.3	11
17	Inhaled high molecular weight hyaluronan ameliorates respiratory failure in acute COPD exacerbation: a pilot study. Respiratory Research, 2021, 22, 30.	1.4	26
18	PROMISE: Working with the CF community to understand emerging clinical and research needs for those treated with highly effective CFTR modulator therapy. Journal of Cystic Fibrosis, 2021, 20, 205-212.	0.3	39

#	Article	IF	Citations
19	Elexacafator/tezacaftor/ivacaftor resolves subfertility in females with CF: A two center case series. Journal of Cystic Fibrosis, 2021, 20, 399-401.	0.3	42
20	Novel Correctors and Potentiators Enhance Translational Readthrough in CFTR Nonsense Mutations. American Journal of Respiratory Cell and Molecular Biology, 2021, 64, 604-616.	1.4	15
21	A Phase 3 Open-Label Study of Elexacaftor/Tezacaftor/Ivacaftor in Children 6 through 11 Years of Age with Cystic Fibrosis and at Least One F508del Allele. American Journal of Respiratory and Critical Care Medicine, 2021, 203, 1522-1532.	2.5	146
22	LPS decreases CFTR open probability and mucociliary transport through generation of reactive oxygen species. Redox Biology, 2021, 43, 101998.	3.9	14
23	A small molecule that induces translational readthrough of CFTR nonsense mutations by eRF1 depletion. Nature Communications, 2021, 12, 4358.	5.8	59
24	Cessation of smoke exposure improves pediatric CF outcomes: Longitudinal analysis of CF Foundation Patient Registry data. Journal of Cystic Fibrosis, 2021, 20, 618-624.	0.3	4
25	Triple Therapy for Cystic Fibrosis Phe508delâ€“Gating and â€“Residual Function Genotypes. New England Journal of Medicine, 2021, 385, 815-825.	13.9	140
26	Riociguat for the treatment of Phe508del homozygous adults with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 1018-1025.	0.3	5
27	Empire-CF study: A phase 2 clinical trial of leukotriene A4 hydrolase inhibitor acebilustat in adult subjects with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 1026-1034.	0.3	9
28	Antisense oligonucleotide-based drug development for Cystic Fibrosis patients carrying the 3849+10Âkb C-to-T splicing mutation. Journal of Cystic Fibrosis, 2021, 20, 865-875.	0.3	30
29	The future of cystic fibrosis care: a global perspective. Lancet Respiratory Medicine,the, 2020, 8, 65-124.	5.2	573
30	Changes in Airway Microbiome and Inflammation with Ivacaftor Treatment in Patients with Cystic Fibrosis and the G551D Mutation. Annals of the American Thoracic Society, 2020, 17, 212-220.	1.5	113
31	Females with Cystic Fibrosis Demonstrate a Differential Response Profile to Ivacaftor Compared with Males. American Journal of Respiratory and Critical Care Medicine, 2020, 201, 996-998.	2.5	26
32	Excess mucus viscosity and airway dehydration impact COPD airway clearance. European Respiratory Journal, 2020, 55, 1900419.	3.1	46
33	Cystic Fibrosis: Emergence of Highly Effective Targeted Therapeutics and Potential Clinical Implications. American Journal of Respiratory and Critical Care Medicine, 2020, 201, 1193-1208.	2.5	137
34	Efficacy and Safety of the CFTR PotentiatorÂlcenticaftor (QBW251) in COPD: Results from a Phase 2 Randomized Trial . International Journal of COPD, 2020, Volume 15, 2399-2409.	0.9	32
35	Fibroblast Growth Factor Receptor 4 Deficiency Mediates Airway Inflammation in the Adult Healthy Lung?. Frontiers in Medicine, 2020, 7, 317.	1.2	6
36	G551D mutation impairs PKA-dependent activation of CFTR channel that can be restored by novel GOF mutations. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2020, 319, L770-L785.	1.3	5

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37	Transparency and diversity in cystic fibrosis research â€“ Authors' reply. Lancet, The, 2020, 396, 602.	6.3	0
38	Haemophilus influenzaepersists in biofilm communities in a smoke-exposed ferret model of COPD. ERJ Open Research, 2020, 6, 00200-2020.	1.1	11
39	Novel Therapy of Bicarbonate, Glutathione, and Ascorbic Acid Improves Cystic Fibrosis Mucus Transport. American Journal of Respiratory Cell and Molecular Biology, 2020, 63, 362-373.	1.4	9
40	Airway remodeling in ferrets with cigarette smoke-induced COPD using ÂµCT imaging. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2020, 319, L11-L20.	1.3	9
41	Changes in LCI in F508del/F508del patients treated with lumacaftor/ivacaftor: Results from the prospect study. Journal of Cystic Fibrosis, 2020, 19, 931-933.	0.3	30
42	Human Nasal Epithelial Organoids for Therapeutic Development in Cystic Fibrosis. Genes, 2020, 11, 603.	1.0	40
43	CFTR targeted therapies: recent advances in cystic fibrosis and possibilities in other diseases of the airways. European Respiratory Review, 2020, 29, 190068.	3.0	30
44	Ivacaftor Reverses Airway Mucus Abnormalities in a Rat Model Harboring a Humanized G551D-CFTR. American Journal of Respiratory and Critical Care Medicine, 2020, 202, 1271-1282.	2.5	35
45	Tobacco smoke exposure and socioeconomic factors are independent predictors of pulmonary decline in pediatric cystic fibrosis. Journal of Cystic Fibrosis, 2020, 19, 783-790.	0.3	26
46	Pulmonary artery enlargement is associated with pulmonary hypertension and decreased survival in severe cystic fibrosis: A cohort study. PLoS ONE, 2020, 15, e0229173.	1.1	14
47	Variable cellular ivacaftor concentrations in people with cystic fibrosis on modulator therapy. Journal of Cystic Fibrosis, 2020, 19, 742-745.	0.3	16
48	Ataluren/ivacaftor combination therapy: Two Nâ€ofâ€1 trials in cystic fibrosis patients with nonsense mutations. Pediatric Pulmonology, 2020, 55, 1838-1842.	1.0	9
49	Pharmacological approaches for targeting cystic fibrosis nonsense mutations. European Journal of Medicinal Chemistry, 2020, 200, 112436.	2.6	25
50	Brd4-p300 inhibition downregulates Nox4 and accelerates lung fibrosis resolution in aged mice. JCI Insight, 2020, 5, .	2.3	45
51	A Novel G542X CFTR Rat Model of Cystic Fibrosis Is Sensitive to Nonsense Mediated Decay. Frontiers in Physiology, 2020, 11, 611294.	1.3	9
52	A simple test could extend cystic-fibrosis treatments to those left behind. Nature, 2020, 583, S5-S5.	13.7	1
53	Targeting the Underlying Defect in CFTR with Small Molecule Compounds. Respiratory Medicine, 2020, , 483-501.	0.1	0
54	Gaming Console Home-Based Exercise for Adults with Cystic Fibrosis: Study Protocol. International Journal of Caring Sciences, 2020, 13, 1530-1540.	0.0	1

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55	CFTR modulator theratyping: Current status, gaps and future directions. Journal of Cystic Fibrosis, 2019, 18, 22-34.	0.3	208
56	Intranasal micro-optical coherence tomography imaging for cystic fibrosis studies. Science Translational Medicine, 2019, 11, .	5.8	42
57	The effect of CFTR modulators on a cystic fibrosis patient presenting with recurrent pancreatitis in the absence of respiratory symptoms: a case report. BMC Gastroenterology, 2019, 19, 123.	0.8	20
58	Elexacaftorâ€“Tezacaftorâ€“Ivacaftor for Cystic Fibrosis with a Single Phe508del Allele. New England Journal of Medicine, 2019, 381, 1809-1819.	13.9	1,231
59	Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: a double-blind, randomised, phase 3 trial. Lancet, The, 2019, 394, 1940-1948.	6.3	804
60	Geometry-Dependent Spectroscopic Contrast in Deep Tissues. IScience, 2019, 19, 965-975.	1.9	15
61	Clinical development of triple-combination CFTR modulators for cystic fibrosis patients with one or two F508del alleles. ERJ Open Research, 2019, 5, 00082-2019.	1.1	72
62	Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction and Radiographic Bronchiectasis in Current and Former Smokers: A Cross-Sectional Study. Annals of the American Thoracic Society, 2019, 16, 150-153.	1.5	8
63	Vaporized E-Cigarette Liquids Induce Ion Transport Dysfunction in Airway Epithelia. American Journal of Respiratory Cell and Molecular Biology, 2019, 61, 162-173.	1.4	54
64	Colocolonic intussusception in an adult cystic fibrosis patient. Journal of Cystic Fibrosis, 2019, 18, e11-e13.	0.3	8
65	Ivacaftor improves rhinologic, psychologic, and sleepâ€related quality of life in G551D cystic fibrosis patients. International Forum of Allergy and Rhinology, 2019, 9, 292-297.	1.5	49
66	Evaluation of 1,2,3â€Triazoles as Amide Bioisosteres In Cystic Fibrosis Transmembrane Conductance Regulator Modulators VXâ€770 and VXâ€809. Chemistry - A European Journal, 2019, 25, 3662-3674.	1.7	20
67	Objective Versus Self-Reported Adherence to Airway Clearance Therapy in Cystic Fibrosis. Respiratory Care, 2019, 64, 176-181.	0.8	17
68	Effectiveness of ivacaftor in cystic fibrosis patients with non-G551D gating mutations. Journal of Cystic Fibrosis, 2019, 18, 102-109.	0.3	30
69	The Effects of the Anti-aging Protein Klotho on Mucociliary Clearance. Frontiers in Medicine, 2019, 6, 339.	1.2	8
70	A glycopolymer improves vascoelasticity and mucociliary transport of abnormal cystic fibrosis mucus. JCI Insight, 2019, 4, .	2.3	35
71	Revealing the molecular signaling pathways of mucus stasis in cystic fibrosis. Journal of Clinical Investigation, 2019, 129, 4089-4090.	3.9	7
72	Co-cultured microfluidic model of the airway optimized for microscopy and micro-optical coherence tomography imaging. Biomedical Optics Express, 2019, 10, 5414.	1.5	18

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73	Sensitivity of ivacaftor to drugâ€drug interactions with rifampin, a cytochrome P450 3A4 inducer. Pediatric Pulmonology, 2018, 53, E6-E8.	1.0	19
74	Changes in Lung Clearance Index in Preschool-aged Patients with Cystic Fibrosis Treated with Ivacaftor (GOAL): A Clinical Trial. American Journal of Respiratory and Critical Care Medicine, 2018, 198, 526-528.	2.5	32
75	Ivacaftor-treated Patients with Cystic Fibrosis Derive Long-Term Benefit Despite No Short-Term Clinical Improvement. American Journal of Respiratory and Critical Care Medicine, 2018, 197, 1483-1486.	2.5	21
76	An Adeno-Associated Viral Vector Capable of Penetrating the Mucus Barrier to Inhaled Gene Therapy. Molecular Therapy - Methods and Clinical Development, 2018, 9, 296-304.	1.8	40
77	Development of an airway mucus defect in the cystic fibrosis rat. JCI Insight, 2018, 3, .	2.3	84
78	Functional Anatomic Imaging of the Airway Surface. Annals of the American Thoracic Society, 2018, 15, S177-S183.	1.5	8
79	Muc5b overexpression causes mucociliary dysfunction and enhances lung fibrosis in mice. Nature Communications, 2018, 9, 5363.	5.8	175
80	Standardized Measurement of Nasal Membrane Transepithelial Potential Difference (NPD). Journal of Visualized Experiments, 2018, , .	0.2	15
81	The epithelial sodium channel (ENaC) as a therapeutic target for cystic fibrosis. Current Opinion in Pharmacology, 2018, 43, 152-165.	1.7	56
82	VX-659â€“Tezacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles. New England Journal of Medicine, 2018, 379, 1599-1611.	13.9	280
83	VX-445â€“Tezacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles. New England Journal of Medicine, 2018, 379, 1612-1620.	13.9	509
84	A revised airway epithelial hierarchy includes CFTR-expressing ionocytes. Nature, 2018, 560, 319-324.	13.7	878
85	Seeing cilia: imaging modalities for ciliary motion and clinical connections. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2018, 314, L909-L921.	1.3	18
86	EMPIRE-CF: A phase II randomized placebo-controlled trial of once-daily, oral acebilustat in adult patients with cystic fibrosis â€“ Study design and patient demographics. Contemporary Clinical Trials, 2018, 72, 86-94.	0.8	33
87	Heme scavenging reduces pulmonary endoplasmic reticulum stress, fibrosis, and emphysema. JCI Insight, 2018, 3, .	2.3	47
88	Effect of ivacaftor on mucociliary clearance and clinical outcomes in cystic fibrosis patients with G551D-CFTR. JCI Insight, 2018, 3, .	2.3	56
89	Influenza-mediated reduction of lung epithelial ion channel activity leads to dysregulated pulmonary fluid homeostasis. JCI Insight, 2018, 3, .	2.3	50
90	Growth in Prepubertal Children With Cystic Fibrosis Treated With Ivacaftor. Pediatrics, 2017, 139, .	1.0	44

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91	A multiple reader scoring system for Nasal Potential Difference parameters. Journal of Cystic Fibrosis, 2017, 16, 573-578.	0.3	10
92	Identification of the amino acids inserted during suppression of CFTR nonsense mutations and determination of their functional consequences. Human Molecular Genetics, 2017, 26, 3116-3129.	1.4	69
93	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.	1.3	107
94	A little CFTR can change a lot: slowing cystic fibrosis progression. Lancet Respiratory Medicine,the, 2017, 5, 86-87.	5.2	7
95	Klotho Inhibits Interleukin-8 Secretion from Cystic Fibrosis Airway Epithelia. Scientific Reports, 2017, 7, 14388.	1.6	36
96	Toward inclusive therapy with CFTR modulators: Progress and challenges. Pediatric Pulmonology, 2017, 52, S4-S14.	1.0	32
97	The therapeutic potential of CFTR modulators for COPD and other airway diseases. Current Opinion in Pharmacology, 2017, 34, 132-139.	1.7	41
98	Tezacaftorâ€“Ivacaftor in Residual-Function Heterozygotes with Cystic Fibrosis. New England Journal of Medicine, 2017, 377, 2024-2035.	13.9	412
99	Assessment of acquired mucociliary clearance defects using microâ€optical coherence tomography. International Forum of Allergy and Rhinology, 2017, 7, 920-925.	1.5	28
100	The Cystic Fibrosis Transmembrane Conductance Regulator Potentiator Ivacaftor Augments Mucociliary Clearance Abrogating Cystic Fibrosis Transmembrane Conductance Regulator Inhibition by Cigarette Smoke. American Journal of Respiratory Cell and Molecular Biology, 2017, 56, 99-108.	1.4	79
101	Therapeutic benefit observed with the CFTR potentiator, ivacaftor, in a CF patient homozygous for the W1282X CFTR nonsense mutation. Journal of Cystic Fibrosis, 2017, 16, 24-29.	0.3	44
102	Not simply the lesser of two evils. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2017, 314, L236-L238.	1.3	1
103	Flexible, high-resolution micro-optical coherence tomography endobronchial probe toward in vivo imaging of cilia. Optics Letters, 2017, 42, 867.	1.7	39
104	Roflumilast reverses CFTR-mediated ion transport dysfunction in cigarette smoke-exposed mice. Respiratory Research, 2017, 18, 173.	1.4	25
105	Sinus Microanatomy and Microbiota in a Rabbit Model of Rhinosinusitis. Frontiers in Cellular and Infection Microbiology, 2017, 7, 540.	1.8	31
106	Assessment of ciliary phenotype in primary ciliary dyskinesia by micro-optical coherence tomography. JCI Insight, 2017, 2, e91702.	2.3	30
107	Use of ferrets for electrophysiologic monitoring of ion transport. PLoS ONE, 2017, 12, e0186984.	1.1	7
108	Lumacaftor/Ivacaftor Treatment of Patients with Cystic Fibrosis Heterozygous for F508delâ€CFTR. Annals of the American Thoracic Society, 2017, 14, 213-219.	1.5	78

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109	Codon bias and the folding dynamics of the cystic fibrosis transmembrane conductance regulator. Cellular and Molecular Biology Letters, 2016, 21, 23.	2.7	32
110	New and emerging targeted therapies for cystic fibrosis. BMJ, The, 2016, 352, i859.	3.0	112
111	Combination therapy with cystic fibrosis transmembrane conductance regulator modulators augment the airway functional microanatomy. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2016, 310, L928-L939.	1.3	58
112	Discovery of Clinically Approved Agents That Promote Suppression of Cystic Fibrosis Transmembrane Conductance Regulator Nonsense Mutations. American Journal of Respiratory and Critical Care Medicine, 2016, 194, 1092-1103.	2.5	77
113	Ataluren stimulates ribosomal selection of near-cognate tRNAs to promote nonsense suppression. Proceedings of the National Academy of Sciences of the United States of America, 2016, 113, 12508-12513.	3.3	168
114	Particle-Tracking Microrheology Using Micro-Optical Coherence Tomography. Biophysical Journal, 2016, 111, 1053-1063.	0.2	26
115	In vivo imaging of airway cilia and mucus clearance with micro-optical coherence tomography. Biomedical Optics Express, 2016, 7, 2494.	1.5	57
116	Pulmonary artery enlargement and cystic fibrosis pulmonary exacerbations: a cohort study. Lancet Respiratory Medicine,the, 2016, 4, 636-645.	5.2	19
117	Pilot evaluation of ivacaftor for chronic bronchitis. Lancet Respiratory Medicine,the, 2016, 4, e32-e33.	5.2	34
118	Dual SMAD Signaling Inhibition Enables Long-Term Expansion of Diverse Epithelial Basal Cells. Cell Stem Cell, 2016, 19, 217-231.	5.2	313
119	Alterations in blood leukocytes of G551D-bearing cystic fibrosis patients undergoing treatment with ivacaftor. Journal of Cystic Fibrosis, 2016, 15, 67-73.	0.3	44
120	Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in Chronic Bronchitis and Other Diseases of Mucus Clearance. Clinics in Chest Medicine, 2016, 37, 147-158.	0.8	50
121	A ferret model of COPD-related chronic bronchitis. JCI Insight, 2016, 1, e87536.	2.3	36
122	Mutation of Growth Arrest Specific 8 Reveals a Role in Motile Cilia Function and Human Disease. PLoS Genetics, 2016, 12, e1006220.	1.5	33
123	Increasing the Endoplasmic Reticulum Pool of the F508del Allele of the Cystic Fibrosis Transmembrane Conductance Regulator Leads to Greater Folding Correction by Small Molecule Therapeutics. PLoS ONE, 2016, 11, e0163615.	1.1	23
124	Therapeutic Approaches to Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in Chronic Bronchitis. Annals of the American Thoracic Society, 2016, 13 Suppl 2, S169-76.	1.5	25
125	Improved Clinical and Radiographic Outcomes After Treatment With Ivacaftor in a Young Adult With Cystic Fibrosis With the P67L CFTR Mutation. Chest, 2015, 147, e79-e82.	0.4	19
126	Breakthrough therapies: Cystic fibrosis (CF) potentiators and correctors. Pediatric Pulmonology, 2015, 50, S3-S13.	1.0	56

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127	Recovery of Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction after Smoking Cessation. American Journal of Respiratory and Critical Care Medicine, 2015, 192, 1521-1524.	2.5	16
128	Î”F508 CFTR Surface Stability Is Regulated by DAB2 and CHIP-Mediated Ubiquitination in Post-Endocytic Compartments. PLoS ONE, 2015, 10, e0123131.	1.1	29
129	Lumacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508delCFTR. New England Journal of Medicine, 2015, 373, 220-231.	13.9	1,308
130	Heme oxygenase-1-mediated autophagy protects against pulmonary endothelial cell death and development of emphysema in cadmium-treated mice. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2015, 309, L280-L292.	1.3	62
131	Novel outcome measures for clinical trials in cystic fibrosis. Pediatric Pulmonology, 2015, 50, 302-315.	1.0	34
132	Pseudomonas aeruginosa in Cystic Fibrosis Patients With G551D-CFTR Treated With Ivacaftor. Clinical Infectious Diseases, 2015, 60, 703-712.	2.9	198
133	Cystic fibrosis. Nature Reviews Disease Primers, 2015, 1, 15010.	18.1	403
134	Efficacy and safety of ivacaftor in patients with cystic fibrosis who have an Arg117His-CFTR mutation: a double-blind, randomised controlled trial. Lancet Respiratory Medicine,the, 2015, 3, 524-533.	5.2	197
135	Moderate intensity exercise mediates comparable increases in exhaled chloride as albuterol in individuals with cystic fibrosis. Respiratory Medicine, 2015, 109, 1001-1011.	1.3	12
136	Defective Innate Immunity and Hyperinflammation in Newborn Cystic Fibrosis Transmembrane Conductance Regulatorâ€“Knockout Ferret Lungs. American Journal of Respiratory Cell and Molecular Biology, 2015, 52, 683-694.	1.4	94
137	ACQUIRED CFTR DYSFUNCTION AND CHRONIC BRONCHITIS IN A NOVEL FERRET MODEL OF COPD. FASEB Journal, 2015, 29, 863.15.	0.2	0
138	Characterization of Defects in Ion Transport and Tissue Development in Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)-Knockout Rats. PLoS ONE, 2014, 9, e91253.	1.1	133
139	An Autoregulatory Mechanism Governing Mucociliary Transport Is Sensitive to Mucus Load. American Journal of Respiratory Cell and Molecular Biology, 2014, 51, 485-493.	1.4	68
140	Porcine nasal epithelial cultures for studies of cystic fibrosis sinusitis. International Forum of Allergy and Rhinology, 2014, 4, 565-570.	1.5	13
141	Acquired defects in CFTR-dependent Î²-adrenergic sweat secretion in chronic obstructive pulmonary disease. Respiratory Research, 2014, 15, 25.	1.4	35
142	Synthetic Aminoglycosides Efficiently Suppress Cystic Fibrosis Transmembrane Conductance Regulator Nonsense Mutations and Are Enhanced by Ivacaftor. American Journal of Respiratory Cell and Molecular Biology, 2014, 50, 805-816.	1.4	131
143	Sweat chloride as a biomarker of CFTR activity: Proof of concept and ivacaftor clinical trial data. Journal of Cystic Fibrosis, 2014, 13, 139-147.	0.3	123
144	Evaluating the predictive ability of sweat chloride. Journal of Cystic Fibrosis, 2014, 13, 118.	0.3	3

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145	Impact of heterozygote CFTR Mutations in COPD patients with Chronic Bronchitis. Respiratory Research, 2014, 15, 18.	1.4	33
146	Cystic Fibrosis Transmembrane Conductance Regulator Activation by Roflumilast Contributes to Therapeutic Benefit in Chronic Bronchitis. American Journal of Respiratory Cell and Molecular Biology, 2014, 50, 549-558.	1.4	71
147	Overcoming the Cystic Fibrosis Sputum Barrier to Leading Adeno-associated Virus Gene Therapy Vectors. Molecular Therapy, 2014, 22, 1484-1493.	3.7	75
148	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, 527-538.	5.2	372
149	A Functional Anatomic Defect of the Cystic Fibrosis Airway. American Journal of Respiratory and Critical Care Medicine, 2014, 190, 421-432.	2.5	135
150	Clinical Mechanism of the Cystic Fibrosis Transmembrane Conductance Regulator Potentiator Ivacaftor in G551D-mediated Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2014, 190, 175-184.	2.5	447
151	Ataluren for the treatment of nonsense-mutation cystic fibrosis: a randomised, double-blind, placebo-controlled phase 3 trial. Lancet Respiratory Medicine,the, 2014, 2, 539-547.	5.2	301
152	Cigarette smoke and CFTR: implications in the pathogenesis of COPD. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2013, 305, L530-L541.	1.3	133
153	Cigarette Smoke Induces Systemic Defects in Cystic Fibrosis Transmembrane Conductance Regulator Function. American Journal of Respiratory and Critical Care Medicine, 2013, 188, 1321-1330.	2.5	168
154	Cystic Fibrosis Transmembrane Regulator Correctors and Potentiators. Cold Spring Harbor Perspectives in Medicine, 2013, 3, a009761-a009761.	2.9	135
155	Reduced Sodium Transport With Nasal Administration of the Prostasin Inhibitor Camostat in Subjects With Cystic Fibrosis. Chest, 2013, 144, 200-207.	0.4	32
156	Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in the Lower Airways in COPD. Chest, 2013, 144, 498-506.	0.4	163
157	Understanding the Relationship Between Sweat Chloride and Lung Function in Cystic Fibrosis. Chest, 2013, 144, 1418.	0.4	11
158	Method for Quantitative Study of Airway Functional Microanatomy Using Micro-Optical Coherence Tomography. PLoS ONE, 2013, 8, e54473.	1.1	152
159	Optimizing Nasal Potential Difference Analysis for CFTR Modulator Development: Assessment of Ivacaftor in CF Subjects with the G551D-CFTR Mutation. PLoS ONE, 2013, 8, e66955.	1.1	44
160	IP-10 Is a Potential Biomarker of Cystic Fibrosis Acute Pulmonary Exacerbations. PLoS ONE, 2013, 8, e72398.	1.1	21
161	Cigarette Smoke Induces Systemic Defects in Cystic Fibrosis Transmembrane Conductance Regulator Function. American Journal of Respiratory and Critical Care Medicine, 2013, 188, 1321-1330.	2.5	95
162	Results of a phase IIa study of VX-809, an investigational CFTR corrector compound, in subjects with cystic fibrosis homozygous for theF508del-CFTRmutation. Thorax, 2012, 67, 12-18.	2.7	466

#	Article	IF	Citations
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