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List of Publications by Year
in descending order

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179 papers	20,225 citations	23567 58 h-index	11308 136 g-index
185 all docs	185 docs citations	185 times ranked	12224 citing authors

#	Article	IF	Citations
1	A CFTR Potentiator in Patients with Cystic Fibrosis and theG551DMutation. New England Journal of Medicine, 2011, 365, 1663-1672.	27.0	1,920
2	Cystic Fibrosis. New England Journal of Medicine, 2005, 352, 1992-2001.	27.0	1,354
3	Lumacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis Homozygous for Phe508delCFTR. New England Journal of Medicine, 2015, 373, 220-231.	27.0	1,308
4	Elexacaftorâ€“Tezacaftorâ€“Ivacaftor for Cystic Fibrosis with a Single Phe508del Allele. New England Journal of Medicine, 2019, 381, 1809-1819.	27.0	1,231
5	A revised airway epithelial hierarchy includes CFTR-expressing ionocytes. Nature, 2018, 560, 319-324.	27.8	878
6	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.	13.7	804
7	Effect of VX-770 in Persons with Cystic Fibrosis and the G551D-CFTRMutation. New England Journal of Medicine, 2010, 363, 1991-2003.	27.0	741
8	The future of cystic fibrosis care: a global perspective. Lancet Respiratory Medicine,the, 2020, 8, 65-124.	10.7	573
9	VX-445â€“Tezacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles. New England Journal of Medicine, 2018, 379, 1612-1620.	27.0	509
10	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.	5.6	466
11	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.	5.6	447
12	Tezacaftorâ€“Ivacaftor in Residual-Function Heterozygotes with Cystic Fibrosis. New England Journal of Medicine, 2017, 377, 2024-2035.	27.0	412
13	Cystic fibrosis. Nature Reviews Disease Primers, 2015, 1, 15010.	30.5	403
14	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.	10.7	372
15	Dual SMAD Signaling Inhibition Enables Long-Term Expansion of Diverse Epithelial Basal Cells. Cell Stem Cell, 2016, 19, 217-231.	11.1	313
16	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.	10.7	301
17	VX-659â€“Tezacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis and One or Two Phe508del Alleles. New England Journal of Medicine, 2018, 379, 1599-1611.	27.0	280
18	CFTR modulator theratyping: Current status, gaps and future directions. Journal of Cystic Fibrosis, 2019, 18, 22-34.	0.7	208

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19	Pseudomonas aeruginosa in Cystic Fibrosis Patients With G551D-CFTR Treated With Ivacaftor. Clinical Infectious Diseases, 2015, 60, 703-712.	5.8	198
20	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.	10.7	197
21	Muc5b overexpression causes mucociliary dysfunction and enhances lung fibrosis in mice. Nature Communications, 2018, 9, 5363.	12.8	175
22	Cigarette Smoke Induces Systemic Defects in Cystic Fibrosis Transmembrane Conductance Regulator Function. American Journal of Respiratory and Critical Care Medicine, 2013, 188, 1321-1330.	5.6	168
23	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.	7.1	168
24	Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in the Lower Airways in COPD. Chest, 2013, 144, 498-506.	0.8	163
25	A Pharmacologic Approach to Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in Smoking Related Lung Disease. PLoS ONE, 2012, 7, e39809.	2.5	159
26	Method for Quantitative Study of Airway Functional Microanatomy Using Micro-Optical Coherence Tomography. PLoS ONE, 2013, 8, e54473.	2.5	152
27	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.	5.6	147
28	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.	5.6	146
29	Triple Therapy for Cystic Fibrosis Phe508delâ€“Gating and â€“Residual Function Genotypes. New England Journal of Medicine, 2021, 385, 815-825.	27.0	140
30	Cystic Fibrosis: Emergence of Highly Effective Targeted Therapeutics and Potential Clinical Implications. American Journal of Respiratory and Critical Care Medicine, 2020, 201, 1193-1208.	5.6	137
31	Cystic Fibrosis Transmembrane Regulator Correctors and Potentiators. Cold Spring Harbor Perspectives in Medicine, 2013, 3, a009761-a009761.	6.2	135
32	A Functional Anatomic Defect of the Cystic Fibrosis Airway. American Journal of Respiratory and Critical Care Medicine, 2014, 190, 421-432.	5.6	135
33	Cigarette smoke and CFTR: implications in the pathogenesis of COPD. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2013, 305, L530-L541.	2.9	133
34	Characterization of Defects in Ion Transport and Tissue Development in Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)-Knockout Rats. PLoS ONE, 2014, 9, e91253.	2.5	133
35	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.	2.9	131
36	Human distal airways contain a multipotent secretory cell that can regenerate alveoli. Nature, 2022, 604, 120-126.	27.8	128

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37	Sweat chloride as a biomarker of CFTR activity: Proof of concept and ivacaftor clinical trial data. Journal of Cystic Fibrosis, 2014, 13, 139-147.	0.7	123
38	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.	3.2	113
39	Potential Role of High-Mobility Group Box 1 in Cystic Fibrosis Airway Disease. American Journal of Respiratory and Critical Care Medicine, 2008, 178, 822-831.	5.6	112
40	New and emerging targeted therapies for cystic fibrosis. BMJ, The, 2016, 352, i859.	6.0	112
41	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.	2.5	107
42	Cigarette Smoke Induces Systemic Defects in Cystic Fibrosis Transmembrane Conductance Regulator Function. American Journal of Respiratory and Critical Care Medicine, 2013, 188, 1321-1330.	5.6	95
43	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.	2.9	94
44	Development of an airway mucus defect in the cystic fibrosis rat. JCI Insight, 2018, 3, .	5.0	84
45	No Detectable Improvements in Cystic Fibrosis Transmembrane Conductance Regulator by Nasal Aminoglycosides in Patients with Cystic Fibrosis with Stop Mutations. American Journal of Respiratory Cell and Molecular Biology, 2007, 37, 57-66.	2.9	81
46	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.	2.9	79
47	Nasal Potential Difference Measurements to Assess CFTR Ion Channel Activity. Methods in Molecular Biology, 2011, 741, 69-86.	0.9	78
48	Lumacaftor/Ivacaftor Treatment of Patients with Cystic Fibrosis Heterozygous for F508delâ€CFTR. Annals of the American Thoracic Society, 2017, 14, 213-219.	3.2	78
49	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.	5.6	77
50	Overcoming the Cystic Fibrosis Sputum Barrier to Leading Adeno-associated Virus Gene Therapy Vectors. Molecular Therapy, 2014, 22, 1484-1493.	8.2	75
51	Detection of Cystic Fibrosis Transmembrane Conductance Regulator Activity in Early-Phase Clinical Trials. Proceedings of the American Thoracic Society, 2007, 4, 387-398.	3.5	73
52	Clinical development of triple-combination CFTR modulators for cystic fibrosis patients with one or two F508del alleles. ERJ Open Research, 2019, 5, 00082-2019.	2.6	72
53	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.	2.9	71
54	Cystic fibrosis transmembrane conductance regulator protein repair as a therapeutic strategy in cystic fibrosis. Current Opinion in Pulmonary Medicine, 2010, 16, 591-597.	2.6	69

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55	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.	2.9	69
56	Pharmaceuticals Targeting Nonsense Mutations in Genetic Diseases. BioDrugs, 2009, 23, 165-174.	4.6	68
57	An Autoregulatory Mechanism Governing Mucociliary Transport Is Sensitive to Mucus Load. American Journal of Respiratory Cell and Molecular Biology, 2014, 51, 485-493.	2.9	68
58	Suppression of CFTR premature termination codons and rescue of CFTR protein and function by the synthetic aminoglycoside NB54. Journal of Molecular Medicine, 2011, 89, 1149-1161.	3.9	67
59	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.	2.9	62
60	Progress in cystic fibrosis and the CF Therapeutics Development Network. Thorax, 2012, 67, 882-890.	5.6	60
61	Restoration of W1282X CFTR Activity by Enhanced Expression. American Journal of Respiratory Cell and Molecular Biology, 2007, 37, 347-356.	2.9	59
62	A small molecule that induces translational readthrough of CFTR nonsense mutations by eRF1 depletion. Nature Communications, 2021, 12, 4358.	12.8	59
63	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.	2.9	58
64	In vivo imaging of airway cilia and mucus clearance with micro-optical coherence tomography. Biomedical Optics Express, 2016, 7, 2494.	2.9	57
65	Breakthrough therapies: Cystic fibrosis (CF) potentiators and correctors. Pediatric Pulmonology, 2015, 50, S3-S13.	2.0	56
66	The epithelial sodium channel (ENaC) as a therapeutic target for cystic fibrosis. Current Opinion in Pharmacology, 2018, 43, 152-165.	3.5	56
67	Effect of ivacaftor on mucociliary clearance and clinical outcomes in cystic fibrosis patients with G551D-CFTR. JCI Insight, 2018, 3, .	5.0	56
68	Vaporized E-Cigarette Liquids Induce Ion Transport Dysfunction in Airway Epithelia. American Journal of Respiratory Cell and Molecular Biology, 2019, 61, 162-173.	2.9	54
69	An International Randomized Multicenter Comparison of Nasal Potential Difference Techniques. Chest, 2010, 138, 919-928.	0.8	50
70	Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction in Chronic Bronchitis and Other Diseases of Mucus Clearance. Clinics in Chest Medicine, 2016, 37, 147-158.	2.1	50
71	Influenza-mediated reduction of lung epithelial ion channel activity leads to dysregulated pulmonary fluid homeostasis. JCI Insight, 2018, 3, .	5.0	50
72	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.	2.8	49

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73	Advances in cystic fibrosis therapies. Current Opinion in Pediatrics, 2006, 18, 604-613.	2.0	47
74	Heme scavenging reduces pulmonary endoplasmic reticulum stress, fibrosis, and emphysema. JCI Insight, 2018, 3, .	5.0	47
75	Excess mucus viscosity and airway dehydration impact COPD airway clearance. European Respiratory Journal, 2020, 55, 1900419.	6.7	46
76	Brd4-p300 inhibition downregulates Nox4 and accelerates lung fibrosis resolution in aged mice. JCI Insight, 2020, 5, .	5.0	45
77	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.	2.5	44
78	Alterations in blood leukocytes of G551D-bearing cystic fibrosis patients undergoing treatment with ivacaftor. Journal of Cystic Fibrosis, 2016, 15, 67-73.	0.7	44
79	Growth in Prepubertal Children With Cystic Fibrosis Treated With Ivacaftor. Pediatrics, 2017, 139, .	2.1	44
80	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.	0.7	44
81	Intranasal micro-optical coherence tomography imaging for cystic fibrosis studies. Science Translational Medicine, 2019, 11, .	12.4	42
82	Elexacafator/tezacaftor/ivacaftor resolves subfertility in females with CF: A two center case series. Journal of Cystic Fibrosis, 2021, 20, 399-401.	0.7	42
83	The therapeutic potential of CFTR modulators for COPD and other airway diseases. Current Opinion in Pharmacology, 2017, 34, 132-139.	3.5	41
84	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.	4.1	40
85	Human Nasal Epithelial Organoids for Therapeutic Development in Cystic Fibrosis. Genes, 2020, 11, 603.	2.4	40
86	Lumacaftor/ivacaftor therapy fails to increase insulin secretion in F508del/F508del CF patients. Journal of Cystic Fibrosis, 2021, 20, 333-338.	0.7	40
87	Flexible, high-resolution micro-optical coherence tomography endobronchial probe toward in vivo imaging of cilia. Optics Letters, 2017, 42, 867.	3.3	39
88	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.	0.7	39
89	Klotho Inhibits Interleukin-8 Secretion from Cystic Fibrosis Airway Epithelia. Scientific Reports, 2017, 7, 14388.	3.3	36
90	A ferret model of COPD-related chronic bronchitis. JCI Insight, 2016, 1, e87536.	5.0	36

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91	Acquired defects in CFTR-dependent Î²-adrenergic sweat secretion in chronic obstructive pulmonary disease. Respiratory Research, 2014, 15, 25.	3.6	35
92	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.	5.6	35
93	A glycopolymer improves vascoelasticity and mucociliary transport of abnormal cystic fibrosis mucus. JCI Insight, 2019, 4, .	5.0	35
94	Dab2 is a key regulator of endocytosis and post-endocytic trafficking of the cystic fibrosis transmembrane conductance regulator. Biochemical Journal, 2012, 441, 633-643.	3.7	34
95	Novel outcome measures for clinical trials in cystic fibrosis. Pediatric Pulmonology, 2015, 50, 302-315.	2.0	34
96	Pilot evaluation of ivacaftor for chronic bronchitis. Lancet Respiratory Medicine,the, 2016, 4, e32-e33.	10.7	34
97	Impact of heterozygote CFTR Mutations in COPD patients with Chronic Bronchitis. Respiratory Research, 2014, 15, 18.	3.6	33
98	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.	1.8	33
99	Mutation of Growth Arrest Specific 8 Reveals a Role in Motile Cilia Function and Human Disease. PLoS Genetics, 2016, 12, e1006220.	3.5	33
100	Reduced Sodium Transport With Nasal Administration of the Prostasin Inhibitor Camostat in Subjects With Cystic Fibrosis. Chest, 2013, 144, 200-207.	0.8	32
101	Codon bias and the folding dynamics of the cystic fibrosis transmembrane conductance regulator. Cellular and Molecular Biology Letters, 2016, 21, 23.	7.0	32
102	Toward inclusive therapy with CFTR modulators: Progress and challenges. Pediatric Pulmonology, 2017, 52, S4-S14.	2.0	32
103	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.	5.6	32
104	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.	2.3	32
105	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.	3.2	32
106	Sinus Microanatomy and Microbiota in a Rabbit Model of Rhinosinusitis. Frontiers in Cellular and Infection Microbiology, 2017, 7, 540.	3.9	31
107	Effectiveness of ivacaftor in cystic fibrosis patients with non-G551D gating mutations. Journal of Cystic Fibrosis, 2019, 18, 102-109.	0.7	30
108	Changes in LCI in F508del/F508del patients treated with lumacaftor/ivacaftor: Results from the prospect study. Journal of Cystic Fibrosis, 2020, 19, 931-933.	0.7	30

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109	CFTR targeted therapies: recent advances in cystic fibrosis and possibilities in other diseases of the airways. European Respiratory Review, 2020, 29, 190068.	7.1	30
110	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.	0.7	30
111	Assessment of ciliary phenotype in primary ciliary dyskinesia by micro-optical coherence tomography. JCI Insight, 2017, 2, e91702.	5.0	30
112	Î”F508 CFTR Surface Stability Is Regulated by DAB2 and CHIP-Mediated Ubiquitination in Post-Endocytic Compartments. PLoS ONE, 2015, 10, e0123131.	2.5	29
113	Assessment of acquired mucociliary clearance defects using microâ€optical coherence tomography. International Forum of Allergy and Rhinology, 2017, 7, 920-925.	2.8	28
114	Regulatory domain phosphorylation to distinguish the mechanistic basis underlying acute CFTR modulators. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2011, 301, L587-L597.	2.9	26
115	Particle-Tracking Microrheology Using Micro-Optical Coherence Tomography. Biophysical Journal, 2016, 111, 1053-1063.	0.5	26
116	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.	5.6	26
117	Tobacco smoke exposure and socioeconomic factors are independent predictors of pulmonary decline in pediatric cystic fibrosis. Journal of Cystic Fibrosis, 2020, 19, 783-790.	0.7	26
118	Inhaled high molecular weight hyaluronan ameliorates respiratory failure in acute COPD exacerbation: a pilot study. Respiratory Research, 2021, 22, 30.	3.6	26
119	Roflumilast reverses CFTR-mediated ion transport dysfunction in cigarette smoke-exposed mice. Respiratory Research, 2017, 18, 173.	3.6	25
120	Pharmacological approaches for targeting cystic fibrosis nonsense mutations. European Journal of Medicinal Chemistry, 2020, 200, 112436.	5.5	25
121	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.	3.2	25
122	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.	2.5	23
123	IP-10 Is a Potential Biomarker of Cystic Fibrosis Acute Pulmonary Exacerbations. PLoS ONE, 2013, 8, e72398.	2.5	21
124	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.	5.6	21
125	Tobacco smoke exposure limits the therapeutic benefit of tezacaftor/ivacaftor in pediatric patients with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 612-617.	0.7	21
126	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.	2.0	20

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127	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.	3.3	20
128	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.	0.8	19
129	Pulmonary artery enlargement and cystic fibrosis pulmonary exacerbations: a cohort study. Lancet Respiratory Medicine,the, 2016, 4, 636-645.	10.7	19
130	Sensitivity of ivacaftor to drugâ€drug interactions with rifampin, a cytochrome P450 3A4 inducer. Pediatric Pulmonology, 2018, 53, E6-E8.	2.0	19
131	Seeing cilia: imaging modalities for ciliary motion and clinical connections. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2018, 314, L909-L921.	2.9	18
132	Co-cultured microfluidic model of the airway optimized for microscopy and micro-optical coherence tomography imaging. Biomedical Optics Express, 2019, 10, 5414.	2.9	18
133	Cystic Fibrosis Transmembrane Conductance Regulator: Roles in Chronic Obstructive Pulmonary Disease. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 631-640.	5.6	18
134	Objective Versus Self-Reported Adherence to Airway Clearance Therapy in Cystic Fibrosis. Respiratory Care, 2019, 64, 176-181.	1.6	17
135	Recovery of Acquired Cystic Fibrosis Transmembrane Conductance Regulator Dysfunction after Smoking Cessation. American Journal of Respiratory and Critical Care Medicine, 2015, 192, 1521-1524.	5.6	16
136	Variable cellular ivacaftor concentrations in people with cystic fibrosis on modulator therapy. Journal of Cystic Fibrosis, 2020, 19, 742-745.	0.7	16
137	Standardized Measurement of Nasal Membrane Transepithelial Potential Difference (NPD). Journal of Visualized Experiments, 2018, , .	0.3	15
138	Geometry-Dependent Spectroscopic Contrast in Deep Tissues. IScience, 2019, 19, 965-975.	4.1	15
139	Novel Correctors and Potentiators Enhance Translational Readthrough in CFTR Nonsense Mutations. American Journal of Respiratory Cell and Molecular Biology, 2021, 64, 604-616.	2.9	15
140	Pulmonary artery enlargement is associated with pulmonary hypertension and decreased survival in severe cystic fibrosis: A cohort study. PLoS ONE, 2020, 15, e0229173.	2.5	14
141	LPS decreases CFTR open probability and mucociliary transport through generation of reactive oxygen species. Redox Biology, 2021, 43, 101998.	9.0	14
142	Porcine nasal epithelial cultures for studies of cystic fibrosis sinusitis. International Forum of Allergy and Rhinology, 2014, 4, 565-570.	2.8	13
143	Moderate intensity exercise mediates comparable increases in exhaled chloride as albuterol in individuals with cystic fibrosis. Respiratory Medicine, 2015, 109, 1001-1011.	2.9	12
144	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.	0.7	12

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145	Understanding the Relationship Between Sweat Chloride and Lung Function in Cystic Fibrosis. Chest, 2013, 144, 1418.	0.8	11
146	Haemophilus influenzaepersists in biofilm communities in a smoke-exposed ferret model of COPD. ERJ Open Research, 2020, 6, 00200-2020.	2.6	11
147	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.	0.7	11
148	A multiple reader scoring system for Nasal Potential Difference parameters. Journal of Cystic Fibrosis, 2017, 16, 573-578.	0.7	10
149	Static mucus impairs bacterial clearance and allows chronic infection withPseudomonas aeruginosain the cystic fibrosis rat. European Respiratory Journal, 2022, 60, 2101032.	6.7	10
150	Evaluation of a novel CFTR potentiator in COPD ferrets with acquired CFTR dysfunction. European Respiratory Journal, 2022, 60, 2101581.	6.7	10
151	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.	2.9	9
152	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.	2.9	9
153	Ataluren/ivacaftor combination therapy: Two Nâ€ofâ€1 trials in cystic fibrosis patients with nonsense mutations. Pediatric Pulmonology, 2020, 55, 1838-1842.	2.0	9
154	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.	0.7	9
155	A Novel G542X CFTR Rat Model of Cystic Fibrosis Is Sensitive to Nonsense Mediated Decay. Frontiers in Physiology, 2020, 11, 611294.	2.8	9
156	Inhaled gene therapy of preclinical muco-obstructive lung diseases by nanoparticles capable of breaching the airway mucus barrier. Thorax, 2022, 77, 812-820.	5.6	9
157	Effects of ivacaftor on systemic inflammation and the plasma proteome in people with CF and G551D. Journal of Cystic Fibrosis, 2022, 21, 950-958.	0.7	9
158	Functional Anatomic Imaging of the Airway Surface. Annals of the American Thoracic Society, 2018, 15, S177-S183.	3.2	8
159	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.	3.2	8
160	Colocolonic intussusception in an adult cystic fibrosis patient. Journal of Cystic Fibrosis, 2019, 18, e11-e13.	0.7	8
161	The Effects of the Anti-aging Protein Klotho on Mucociliary Clearance. Frontiers in Medicine, 2019, 6, 339.	2.6	8
162	A little CFTR can change a lot: slowing cystic fibrosis progression. Lancet Respiratory Medicine,the, 2017, 5, 86-87.	10.7	7

#	Article	IF	Citations
163	Revealing the molecular signaling pathways of mucus stasis in cystic fibrosis. Journal of Clinical Investigation, 2019, 129, 4089-4090.	8.2	7
164	Use of ferrets for electrophysiologic monitoring of ion transport. PLoS ONE, 2017, 12, e0186984.	2.5	7
165	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.	2.8	7
166	Fibroblast Growth Factor Receptor 4 Deficiency Mediates Airway Inflammation in the Adult Healthy Lung?. Frontiers in Medicine, 2020, 7, 317.	2.6	6
167	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.	2.9	5
168	Riociguat for the treatment of Phe508del homozygous adults with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 1018-1025.	0.7	5
169	Cessation of smoke exposure improves pediatric CF outcomes: Longitudinal analysis of CF Foundation Patient Registry data. Journal of Cystic Fibrosis, 2021, 20, 618-624.	0.7	4
170	Poly (acetyl, arginyl) glucosamine disrupts Pseudomonas aeruginosa biofilms and enhances bacterial clearance in a rat lung infection model. Microbiology (United Kingdom), 2022, 168, .	1.8	4
171	Evaluating the predictive ability of sweat chloride. Journal of Cystic Fibrosis, 2014, 13, 118.	0.7	3
172	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.	1.8	3
173	Not simply the lesser of two evils. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2017, 314, L236-L238.	2.9	1
174	A simple test could extend cystic-fibrosis treatments to those left behind. Nature, 2020, 583, S5-S5.	27.8	1
175	Gaming Console Home-Based Exercise for Adults with Cystic Fibrosis: Study Protocol. International Journal of Caring Sciences, 2020, 13, 1530-1540.	0.0	1
176	Transparency and diversity in cystic fibrosis research â€“ Authors' reply. Lancet, The, 2020, 396, 602.	13.7	0
177	ACQUIRED CFTR DYSFUNCTION AND CHRONIC BRONCHITIS IN A NOVEL FERRET MODEL OF COPD. FASEB Journal, 2015, 29, 863.15.	0.5	0
178	Targeting the Underlying Defect in CFTR with Small Molecule Compounds. Respiratory Medicine, 2020, , 483-501.	0.1	0
179	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, , .	5.6	0