Elexacaftorâ€“Tezacaftorâ€“Ivacaftor for Cystic Fibrosi

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Citation Report

#	Article	IF	Citations
1	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
2	Entering the era of highly effective CFTR modulator therapy. Lancet, The, 2019, 394, 1886-1888.	6.3	6
3	Realizing the Dream of Molecularly Targeted Therapies for Cystic Fibrosis. New England Journal of Medicine, 2019, 381, 1863-1865.	13.9	34
4	Cystic fibrosis: triple therapy shows promising results. BMJ: British Medical Journal, 2019, 367, l6347.	2.4	6
5	Mycobacterium abscessus, an Emerging and Worrisome Pathogen among Cystic Fibrosis Patients. International Journal of Molecular Sciences, 2019, 20, 5868.	1.8	84
6	Elexacaftor/Ivacaftor/Tezacaftor: First Approval. Drugs, 2019, 79, 2001-2007.	4.9	57
7	TMEM16A Potentiators: Is There a Need for New Modulators in Cystic Fibrosis?. American Journal of Respiratory and Critical Care Medicine, 2020, 201, 888-889.	2.5	1
8	Towards next generation therapies for cystic fibrosis: Folding, function and pharmacology of CFTR. Journal of Cystic Fibrosis, 2020, 19, S25-S32.	0.3	20
9	Outcomes of a methicillinâ€resistant Staphylococcus aureus (MRSA) eradication protocol in pediatric cystic fibrosis (CF) patients. Pediatric Pulmonology, 2020, 55, 654-659.	1.0	3
10	Nutritional excess in cystic fibrosis: the skinny on obesity. Journal of Cystic Fibrosis, 2020, 19, 3-5.	0.3	10
11	New Drug Hailed as Major Breakthrough in Cystic Fibrosis. American Journal of Medical Genetics, Part A, 2020, 182, 8-9.	0.7	1
12	Cystic Fibrosis Lung Disease: An Overview. Respiratory Care, 2020, 65, 233-251.	0.8	94
13	Positive clinical outcomes following ivacaftor treatment in a cystic fibrosis patient with the genotype 3272â€“26AÂ>ÂG/Q493X. Journal of Cystic Fibrosis, 2020, 19, e3-e4.	0.3	3
14	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
15	Cystic fibrosis in the year 2020: A disease with a new face. Acta Paediatrica, International Journal of Paediatrics, 2020, 109, 893-899.	0.7	189
16	Functional Profiling of CFTR-Directed Therapeutics Using Pediatric Patient-Derived Nasal Epithelial Cell Models. Frontiers in Pediatrics, 2020, 8, 536.	0.9	11
17	Transition to adult care in cystic fibrosis: The challenges and the structure. Paediatric Respiratory Reviews, 2022, 41, 23-29.	1.2	5
18	Treatment of pulmonary exacerbations in cystic fibrosis. Current Opinion in Pulmonary Medicine, 2020, 26, 679-684.	1.2	11

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19	Editorial: The changing landscape of cystic fibrosis: new therapies, challenges and a global pandemic. Current Opinion in Pulmonary Medicine, 2020, 26, 668-670.	1.2	2
20	Highly Efficient Gene Editing of Cystic Fibrosis Patient-Derived Airway Basal Cells Results in Functional CFTR Correction. Molecular Therapy, 2020, 28, 1684-1695.	3.7	48
21	Monitoring early stage lung disease in cystic fibrosis. Current Opinion in Pulmonary Medicine, 2020, 26, 671-678.	1.2	16
22	Sexual and reproductive health in cystic fibrosis. Current Opinion in Pulmonary Medicine, 2020, 26, 685-695.	1.2	7
23	What Is the Current Status of Lung Transplantation?. Advances in Surgery, 2020, 54, 103-127.	0.6	2
24	Cystic Fibrosis and Genotype-Dependent Therapy: Is There a Need for a Sex-Specific Therapy?. , 2020, 4, 247028972093702.	0.8	1
25	Interstitial lung disease in infancy. Early Human Development, 2020, 150, 105186.	0.8	14
26	Targeting p53 in chronic lymphocytic leukemia. Expert Opinion on Therapeutic Targets, 2020, 24, 1239-1250.	1.5	20
27	Comparison of Organoid Swelling and In Vivo Biomarkers of CFTR Function to Determine Effects of Lumacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis Homozygous for the F508del Mutation. American Journal of Respiratory and Critical Care Medicine, 2020, 202, 1589-1592.	2.5	23
28	Sustained recovery of exocrine pancreatic function in a teenager with cystic fibrosis treated with ivacaftor. Pediatric Pulmonology, 2020, 55, 2493-2494.	1.0	12
30	Targeting the Heme Oxygenase 1/Carbon Monoxide Pathway to Resolve Lung Hyper-Inflammation and Restore a Regulated Immune Response in Cystic Fibrosis. Frontiers in Pharmacology, 2020, 11, 1059.	1.6	22
31	Steps toward Cell Therapy for Cystic Fibrosis. American Journal of Respiratory Cell and Molecular Biology, 2020, 63, 275-276.	1.4	3
32	Can lumacaftor-ivacaftor reverse glucose-tolerance abnormalities in cystic fibrosis?. Journal of Cystic Fibrosis, 2020, 19, 666.	0.3	2
33	The changing demography of the cystic fibrosis population: forecasting future numbers of adults in the UK. Scientific Reports, 2020, 10, 10660.	1.6	14
35	Current Treatment Options for Cystic Fibrosis-Related Liver Disease. International Journal of Molecular Sciences, 2020, 21, 8586.	1.8	22
36	Variations in Nutrition Practices in Cystic Fibrosis: A Survey of the DIGEST Program. Nutrition in Clinical Practice, 2021, 36, 1247-1251.	1.1	6
37	Small molecule drugs in cystic fibrosis. Archives of Disease in Childhood: Education and Practice Edition, 2022, 107, 379-382.	0.3	1
38	Vasculitis in Cystic Fibrosis. Frontiers in Pediatrics, 2020, 8, 585275.	0.9	4

#	Article	IF	Citations
39	Rats Race to Keep Pace in the Growing Cystic Fibrosis Model Space. American Journal of Respiratory and Critical Care Medicine, 2020, 202, 1212-1214.	2.5	1
40	Impact of Cross-Coupling Reactions in Drug Discovery and Development. Molecules, 2020, 25, 3493.	1.7	125
41	Targeting IgG Autoantibodies for Improved Cytotoxicity of Bactericidal Permeability Increasing Protein in Cystic Fibrosis. Frontiers in Pharmacology, 2020, 11, 1098.	1.6	7
43	Physiologically Based Pharmacokinetic Modeling of CFTR Modulation in People with Cystic Fibrosis Transitioning from Mono or Dual Regimens to Triple-Combination Elexacaftor/Tezacaftor/Ivacaftor. Pulmonary Therapy, 2020, 6, 275-286.	1.1	15
44	Acidic Submucosal Gland pH and Elevated Protein Concentration Produce Abnormal Cystic Fibrosis Mucus. Developmental Cell, 2020, 54, 488-500.e5.	3.1	24
45	Cystic fibrosis â€“ Ten promising therapeutic approaches in the current era of care. Expert Opinion on Investigational Drugs, 2020, 29, 1107-1124.	1.9	8
46	High-Dose Inhaled Nitric Oxide as Adjunct Therapy in Cystic Fibrosis Targeting Burkholderia multivorans. Case Reports in Pediatrics, 2020, 2020, 1-6.	0.2	16
47	New drug approvals for 2019: Synthesis and clinical applications. European Journal of Medicinal Chemistry, 2020, 205, 112667.	2.6	36
48	The Changing Face of Cystic Fibrosis and Its Implications for Screening. International Journal of Neonatal Screening, 2020, 6, 54.	1.2	10
50	Will Airway Gene Therapy for Cystic Fibrosis Improve Lung Function? New Imaging Technologies Can Help Us Find Out. Human Gene Therapy, 2020, 31, 973-984.	1.4	5
51	ENaC inhibition in cystic fibrosis: potential role in the new era of CFTR modulator therapies. European Respiratory Journal, 2020, 56, 2000946.	3.1	33
52	Correction of Airway Stem Cells: Genome Editing Approaches for the Treatment of Cystic Fibrosis. Human Gene Therapy, 2020, 31, 956-972.	1.4	19
53	Functional Genomics of ABCA3 Variants. American Journal of Respiratory Cell and Molecular Biology, 2020, 63, 436-443.	1.4	19
54	New therapies for people with CF in the CFTR modulator world. Journal of Cystic Fibrosis, 2020, 19, 669-670.	0.3	2
55	Oral antibiotic prescribing patterns for treatment of pulmonary exacerbations in two large pediatric CF centers. Pediatric Pulmonology, 2020, 55, 3400-3406.	1.0	8
56	Accelerated Approval or Risk Reduction? How Response Biomarkers Advance Therapeutics through Clinical Trials in Cystic Fibrosis. Trends in Molecular Medicine, 2020, 26, 1068-1077.	3.5	7
57	Cystic fibrosis and the gut. Frontline Gastroenterology, 2021, 12, 622-628.	0.9	5
58	Focusing on the penultimate step: increasing early lung transplant discussion in cystic fibrosis clinic to prepare patients for referral. BMJ Open Quality, 2020, 9, e001031.	0.4	1

#	Article	IF	Citations
59	Novel Anti-Inflammatory Approaches for Cystic Fibrosis Lung Disease: Identification of Molecular Targets and Design of Innovative Therapies. Frontiers in Pharmacology, 2020, 11, 1096.	1.6	30
60	Fluorescence assay for simultaneous quantification of CFTR ion-channel function and plasma membrane proximity. Journal of Biological Chemistry, 2020, 295, 16529-16544.	1.6	7
61	The Lung Life of a Cystic Fibrosis Patient: A Patient and Physician Perspective. Pulmonary Therapy, 2020, 6, 159-167.	1.1	0
62	Does newborn screening improve early lung function in cystic fibrosis?. Paediatric Respiratory Reviews, 2022, 42, 17-22.	1.2	3
63	CFTR trafficking mutations disrupt cotranslational protein folding by targeting biosynthetic intermediates. Nature Communications, 2020, 11, 4258.	5.8	23
64	Carbon monoxide-releasing molecules inhibit the cystic fibrosis transmembrane conductance regulator Cl^{â˜}channel. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2020, 319, L997-L1009.	1.3	3
65	Costly Genes. PLoS Genetics, 2020, 16, e1008889.	1.5	1
66	CFTR Modulators: Impact on Fertility, Pregnancy, and Lactation in Women with Cystic Fibrosis. Journal of Clinical Medicine, 2020, 9, 2706.	1.0	39
67	From Ivacaftor to Triple Combination: A Systematic Review of Efficacy and Safety of CFTR Modulators in People with Cystic Fibrosis. International Journal of Molecular Sciences, 2020, 21, 5882.	1.8	57
68	Carriers of a single CFTR mutation are asymptomatic: an evolving dogma?. European Respiratory Journal, 2020, 56, 2002645.	3.1	5
69	Multiple Reaction Monitoring Mass Spectrometry for the Drug Monitoring of Ivacaftor, Tezacaftor, and Elexacaftor Treatment Response in Cystic Fibrosis: A High-Throughput Method. ACS Pharmacology and Translational Science, 2020, 3, 987-996.	2.5	17
71	Treating Cystic Fibrosis with mRNA and CRISPR. Human Gene Therapy, 2020, 31, 940-955.	1.4	35
72	Towards the Development of AgoKirs: New Pharmacological Activators to Study Kir2.x Channel and Target Cardiac Disease. International Journal of Molecular Sciences, 2020, 21, 5746.	1.8	5
73	Cystic fibrosis year in review 2019: Section 1 CFTR modulators. Pediatric Pulmonology, 2020, 55, 3236-3242.	1.0	10
74	Integrative genomic meta-analysis reveals novel molecular insights into cystic fibrosis and Î”F508-CFTR rescue. Scientific Reports, 2020, 10, 20553.	1.6	7
75	â€˜Triple therapyâ€™ (elexacaftor, tezacaftor, ivacaftor) skin rash in patients with cystic fibrosis. Postgraduate Medical Journal, 2022, 98, 86-86.	0.9	9
76	Potential of the Electronic Nose for the Detection of Respiratory Diseases with and without Infection. International Journal of Molecular Sciences, 2020, 21, 9416.	1.8	25
77	The human respiratory tract microbial community structures in healthy and cystic fibrosis infants. Npj Biofilms and Microbiomes, 2020, 6, 61.	2.9	18

#	Article	IF	Citations
78	The era of CFTR modulators: improvements made and remaining challenges. Breathe, 2020, 16, 200016.	0.6	20
79	Targeted deubiquitination rescues distinct trafficking-deficient ion channelopathies. Nature Methods, 2020, 17, 1245-1253.	9.0	41
80	Effect of apical chloride concentration on the measurement of responses to CFTR modulation in airway epithelia cultured from nasal brushings. Physiological Reports, 2020, 8, e14603.	0.7	8
82	Transcriptomic and Proteostasis Networks of CFTR and the Development of Small Molecule Modulators for the Treatment of Cystic Fibrosis Lung Disease. Genes, 2020, 11, 546.	1.0	15
83	End-of-Life Care in Cystic Fibrosis: Comparing Provider Practices Based on Lung Transplant Candidacy. Journal of Palliative Medicine, 2020, 23, 1606-1612.	0.6	1
84	Phenotypes in Chronic Rhinosinusitis. Current Allergy and Asthma Reports, 2020, 20, 20.	2.4	32
85	Cystic fibrosis transmembrane conductance receptor modulator therapy in cystic fibrosis, an update. Current Opinion in Pediatrics, 2020, 32, 384-388.	1.0	20
86	Allele-Specific Prevention of Nonsense-Mediated Decay in Cystic Fibrosis Using Homology-Independent Genome Editing. Molecular Therapy - Methods and Clinical Development, 2020, 17, 1118-1128.	1.8	33
87	Patients eligible for modulator drugs: Data from cystic fibrosis registry of Turkey. Pediatric Pulmonology, 2020, 55, 2302-2306.	1.0	5
88	Correlation between Ivacaftor-induced CFTR Activation in Airway Epithelial Cells and Improved Lung Function: A Proof-of-Concept Study. Annals of the American Thoracic Society, 2020, 17, 1024-1027.	1.5	9
89	Control the platelets, control the disease: A novel cystic fibrosis hypothesis. Journal of Thrombosis and Haemostasis, 2020, 18, 1531-1534.	1.9	1
90	A helper-dependent adenoviral vector rescues CFTR to wild-type functional levels in cystic fibrosis epithelial cells harbouring class I mutations. European Respiratory Journal, 2020, 56, 2000205.	3.1	25
91	Antisense oligonucleotide-mediated correction of CFTR splicing improves chloride secretion in cystic fibrosis patient-derived bronchial epithelial cells. Nucleic Acids Research, 2020, 48, 7454-7467.	6.5	26
92	Effect of CFTR Modulators on Anthropometric Parameters in Individuals with Cystic Fibrosis: An Evidence Analysis Center Systematic Review. Journal of the Academy of Nutrition and Dietetics, 2021, 121, 1364-1378.e2.	0.4	45
93	Highlights from the 2019 North American Cystic Fibrosis Conference. Pediatric Pulmonology, 2020, 55, 2225-2232.	1.0	2
94	Human Nasal Epithelial Organoids for Therapeutic Development in Cystic Fibrosis. Genes, 2020, 11, 603.	1.0	40
95	Tezacaftor/ivacaftor in people with cystic fibrosis heterozygous for minimal function CFTR mutations. Journal of Cystic Fibrosis, 2020, 19, 962-968.	0.3	21
96	Cystic fibrosis drug trial design in the era of CFTR modulators associated with substantial clinical benefit: stakeholdersâ€™ consensus view. Journal of Cystic Fibrosis, 2020, 19, 688-695.	0.3	14

#	Article	IF	Citations
97	Building global development strategies for cf therapeutics during a transitional cftr modulator era. Journal of Cystic Fibrosis, 2020, 19, 677-687.	0.3	24
98	The Effect of Sodium Bicarbonate, a Beneficial Adjuvant Molecule in Cystic Fibrosis, on Bronchial Epithelial Cells Expressing a Wild-Type or Mutant CFTR Channel. International Journal of Molecular Sciences, 2020, 21, 4024.	1.8	17
99	Comment on "Effect of one-year lumacaftor-ivacaftor treatment on glucose tolerance abnormalities in cystic fibrosis patients". Journal of Cystic Fibrosis, 2020, 19, 839.	0.3	3
100	Non-Small-Cell Lung Cancer Signaling Pathways, Metabolism, and PD-1/PD-L1 Antibodies. Cancers, 2020, 12, 1475.	1.7	69
101	Recognizing genetic disease: A key aspect of pediatric pulmonary care. Pediatric Pulmonology, 2020, 55, 1794-1809.	1.0	2
102	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
103	A tripartite cooperative mechanism confers resistance of the protein kinase A catalytic subunit to dephosphorylation. Journal of Biological Chemistry, 2020, 295, 3316-3329.	1.6	2
104	New method for rapid and dynamic quantification of elastase activity on sputum neutrophils from patients with cystic fibrosis using flow cytometry. European Respiratory Journal, 2020, 55, 1902355.	3.1	4
105	Re-imagining cystic fibrosis care: next generation thinking. European Respiratory Journal, 2020, 55, 1902443.	3.1	12
106	Phenotyping of Rare CFTR Mutations Reveals Distinct Trafficking and Functional Defects. Cells, 2020, 9, 754.	1.8	23
107	CFTR Modulators: The Changing Face of Cystic Fibrosis in the Era of Precision Medicine. Frontiers in Pharmacology, 2019, 10, 1662.	1.6	287
108	Ivacaftor for the treatment of cystic fibrosis in children under six years of age. Expert Review of Respiratory Medicine, 2020, 14, 547-557.	1.0	6
109	Outcomes of pregnancy in women with cystic fibrosis (CF) taking CFTR modulators â€“ an international survey. Journal of Cystic Fibrosis, 2020, 19, 521-526.	0.3	57
110	Genomic research delivering on promises: From rejuvenation to vaccines and pharmacogenetics. Journal of Clinical Pharmacy and Therapeutics, 2020, 45, 585-589.	0.7	2
111	Genomically-guided therapies: A new era for cystic fibrosis. Archives De Pediatrie, 2020, 27, eS41-eS44.	0.4	2
112	Cystic Fibrosis Transmembrane Conductance Regulator Modulator Therapy: A Review for the Otolaryngologist. American Journal of Rhinology and Allergy, 2020, 34, 573-580.	1.0	10
113	Contemporary Concise Review 2019: Sleep and ventilation. Respirology, 2020, 25, 552-558.	1.3	2
114	Accumulation and persistence of ivacaftor in airway epithelia with prolonged treatment. Journal of Cystic Fibrosis, 2020, 19, 746-751.	0.3	9

#	Article	IF	Citations
115	Rationale and design of the HIT-CF organoid study: stratifying cystic fibrosis patients based on intestinal organoid response to different CFTR-modulators. Translational Medicine Communications, 2020, 5, .	0.5	10
116	Characterization of the mechanism of action of RDR01752, a novel corrector of F508del-CFTR. Biochemical Pharmacology, 2020, 180, 114133.	2.0	14
117	Chronic rhinosinusitis in patients with cystic fibrosisâ€"Current management and new treatments. Laryngoscope Investigative Otolaryngology, 2020, 5, 368-374.	0.6	18
118	Trikafta and Psychopathology in Cystic Fibrosis: A Case Report. Psychosomatics, 2020, 61, 735-738.	2.5	29
119	Academy of Nutrition and Dietetics: 2020 Cystic Fibrosis Evidence Analysis Center Evidence-Based Nutrition Practice Guideline. Journal of the Academy of Nutrition and Dietetics, 2021, 121, 1591-1636.e3.	0.4	45
120	Epithelial vectorial ion transport in cystic fibrosis: Dysfunction, measurement, and pharmacotherapy to target the primary deficit. SAGE Open Medicine, 2020, 8, 205031212093380.	0.7	2
121	The Resolution Approach to Cystic Fibrosis Inflammation. Frontiers in Pharmacology, 2020, 11, 1129.	1.6	4
122	iPSC-Derived Intestinal Organoids from Cystic Fibrosis Patients Acquire CFTR Activity upon TALEN-Mediated Repair of the p.F508del Mutation. Molecular Therapy - Methods and Clinical Development, 2020, 17, 858-870.	1.8	35
123	Phenotypic and molecular characteristics of CF patients carrying the I1234V mutation. Respiratory Medicine, 2020, 170, 106027.	1.3	1
124	Correcting CFTR: New Gene Editing Strategies for Rescuing CFTR Function ExÂVivo. Cell Stem Cell, 2020, 26, 476-478.	5.2	3
125	At the forefront of cystic fibrosis Basic Science research: 16th ECFS Basic Science Conference. Journal of Cystic Fibrosis, 2020, 19, 169-170.	0.3	1
126	Contraceptive use among women with cystic fibrosis: A pilot study linking reproductive health questions to the Cystic Fibrosis Foundation National Patient Registry. Contraception, 2020, 101, 420-426.	0.8	22
127	Triple Therapy for Cystic Fibrosis with a Phe508del CFTR Mutation. New England Journal of Medicine, 2020, 382, 684-684.	13.9	8
128	Variable cellular ivacaftor concentrations in people with cystic fibrosis on modulator therapy. Journal of Cystic Fibrosis, 2020, 19, 742-745.	0.3	16
129	Exploring the basic mechanisms in Cystic Fibrosis: Promoting data presentation and discussion at the 16th ECFS Basic Science Conference. Journal of Cystic Fibrosis, 2020, 19, S1-S4.	0.3	0
130	Regulation of CFTR Biogenesis by the Proteostatic Network and Pharmacological Modulators. International Journal of Molecular Sciences, 2020, 21, 452.	1.8	31
131	A Therapy for Most with Cystic Fibrosis. Cell, 2020, 180, 211.	13.5	47
132	Do ribosomal protein alterations affect ER stress response in CVID?. Clinical and Experimental Immunology, 2020, 200, 87-88.	1.1	0

#	Article	IF	Citations
133	Novel bacterial topoisomerase inhibitors derived from isomannide. European Journal of Medicinal Chemistry, 2020, 199, 112324.	2.6	11
134	Pediatric respiratory medicine. Canadian Journal of Respiratory, Critical Care, and Sleep Medicine, 2020, 4, S25-S27.	0.2	0
135	The Impact of the CFTR Gene Discovery on Cystic Fibrosis Diagnosis, Counseling, and Preventive Therapy. Genes, 2020, 11, 401.	1.0	19
136	The CFTR variant profile of Hispanic patients with cystic fibrosis: Impact on access to effective screening, diagnosis, and personalized medicine. Journal of Genetic Counseling, 2020, 29, 607-615.	0.9	4
137	Real-world evidence in cystic fibrosis modulator development: Establishing a path forward. Journal of Cystic Fibrosis, 2020, 19, e11-e12.	0.3	3
138	Cystic fibrosis 2019: Year in review. Paediatric Respiratory Reviews, 2020, 35, 95-98.	1.2	4
139	Progress in Model Systems of Cystic Fibrosis Mucosal Inflammation to Understand Aberrant Neutrophil Activity. Frontiers in Immunology, 2020, 11, 595.	2.2	12
140	Cystic Fibrosis, CFTR, and Colorectal Cancer. International Journal of Molecular Sciences, 2020, 21, 2891.	1.8	59
141	A new future for patients with cystic fibrosis. American Journal of Transplantation, 2020, 20, 1213-1214.	2.6	1
142	New drug treatments for cystic fibrosis. BMJ, The, 2020, 368, m118.	3.0	3
143	Lung Transplantation. Chest, 2020, 157, 757-758.	0.4	1
144	Toward a More Precise Future for Oncology. Cancer Cell, 2020, 37, 431-442.	7.7	21
145	The bidirectional relationship between CFTR and lipids. Communications Biology, 2020, 3, 179.	2.0	26
146	The preclinical discovery and development of the combination of ivacaftor + tezacaftor used to treat cystic fibrosis. Expert Opinion on Drug Discovery, 2020, 15, 873-891.	2.5	26
147	Impact of CFTR modulator use on outcomes in people with severe cystic fibrosis lung disease. European Respiratory Review, 2020, 29, 190112.	3.0	69
148	Familial Interstitial Lung Disease. Seminars in Respiratory and Critical Care Medicine, 2020, 41, 229-237.	0.8	10
149	Clinical response to lumacaftor-ivacaftor in patients with cystic fibrosis according to baseline lung function. Journal of Cystic Fibrosis, 2021, 20, 220-227.	0.3	24
150	How to determine the mechanism of action of CFTR modulator compounds: A gateway to theranostics. European Journal of Medicinal Chemistry, 2021, 210, 112989.	2.6	10

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151	Efficacy of elexacaftor/tezacaftor/ivacaftor in patients with cystic fibrosis and advanced lung disease. European Respiratory Journal, 2021, 57, 2003079.	3.1	48
152	Correction of CFTR function in intestinal organoids to guide treatment of cystic fibrosis. European Respiratory Journal, 2021, 57, 1902426.	3.1	71
153	Cystic fibrosis in low and middle-income countries (LMIC): A view from four different regions of the world. Paediatric Respiratory Reviews, 2021, 38, 37-44.	1.2	16
154	Genomic, transcriptomic, and protein landscape profile of CFTR and cystic fibrosis. Human Genetics, 2021, 140, 423-439.	1.8	3
155	Biliary disease and cholecystectomy after initiation of elexacaftor/ivacaftor/tezacaftor in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 506-510.	0.3	23
156	Azithromycin and tezacaftor/ivacaftor is associated with first-degree heart block in an adult with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, e19-e21.	0.3	4
157	Effect of highly effective modulator treatment on sinonasal symptoms in cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 460-463.	0.3	50
158	Emerging technologies for cystic fibrosis transmembrane conductance regulator restoration in all people with CF. Pediatric Pulmonology, 2021, 56, S32-S39.	1.0	8
159	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
160	CFTR modulator therapy for cystic fibrosis caused by the rare c.3700A>G mutation. Journal of Cystic Fibrosis, 2021, 20, 452-459.	0.3	19
161	Safety and Efficacy of Elexacaftor/Tezacaftor/Ivacaftor for 24 Weeks or Longer in People with Cystic Fibrosis and One or More F508del Alleles: Interim Results of an Open-Label Phase 3 Clinical Trial. American Journal of Respiratory and Critical Care Medicine, 2021, 203, 381-385.	2.5	116
162	Impact of novel CFTR modulator on sinonasal quality of life in adult patients with cystic fibrosis. International Forum of Allergy and Rhinology, 2021, 11, 201-203.	1.5	15
163	Projecting the impact of delayed access to elexacaftor/tezacaftor/ivacaftor for people with Cystic Fibrosis. Journal of Cystic Fibrosis, 2021, 20, 243-249.	0.3	35
164	Pulmonary Exacerbations in Adults With Cystic Fibrosis. Chest, 2021, 159, 93-102.	0.4	26
165	Development of elexacaftor â€“ tezacaftor â€“ ivacaftor: Highly effective CFTR modulation for the majority of people with Cystic Fibrosis. Expert Review of Respiratory Medicine, 2021, 15, 723-735.	1.0	23
166	Avatar acceptability: views from the Australian Cystic Fibrosis community on the use of personalised organoid technology to guide treatment decisions. ERJ Open Research, 2021, 7, 00448-2020.	1.1	7
167	Multisubstituted pyrazole synthesis via [3â€‰+â€‰2] cycloaddition/rearrangement/N H insertion cascade reaction of Î±-diazoesters and ynones. Chinese Chemical Letters, 2021, 32, 132-135.	4.8	22
168	1-BENZYLSPIRO[PIPERIDINE-4,1â€²-PYRIDO[3,4-b]indole] â€˜co-potentiatorsâ€™ for minimal function CFTR mutants. European Journal of Medicinal Chemistry, 2021, 209, 112888.	2.6	7

#	Article	IF	Citations
169	Long-term azithromycin use is not associated with QT prolongation in children with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, e16-e18.	0.3	6
170	International consensus statement on allergy and rhinology: rhinosinusitis 2021. International Forum of Allergy and Rhinology, 2021, 11, 213-739.	1.5	398
171	Defining the Clinical Utility of the Lung Clearance Index. Are We There Yet?. American Journal of Respiratory and Critical Care Medicine, 2021, 203, 937-939.	2.5	5
172	A current review of the safety of cystic fibrosis transmembrane conductance regulator modulators. Journal of Clinical Pharmacy and Therapeutics, 2021, 46, 286-294.	0.7	20
173	Lung transplant referrals for individuals with cystic fibrosis: A pediatric perspective on the cystic fibrosis foundation consensus guidelines. Pediatric Pulmonology, 2021, 56, 465-471.	1.0	4
174	The future of pediatric pulmonology: A survey of division directors, assessment of current research funding, and discussion of workforce trends. Pediatric Pulmonology, 2023, 58, 653-661.	1.0	4
175	Safety and efficacy of the cystic fibrosis transmembrane conductance regulator potentiator icenticaftor (QBW251). Journal of Cystic Fibrosis, 2021, 20, 250-256.	0.3	13
176	The Clinical Use of Lung MRI in Cystic Fibrosis. Chest, 2021, 159, 2205-2217.	0.4	29
177	Lumacaftor/ivacaftor therapy fails to increase insulin secretion in F508del/F508del CF patients. Journal of Cystic Fibrosis, 2021, 20, 333-338.	0.3	40
178	PEGylation of Recombinant Human Deoxyribonuclease I Provides a Longâ€Acting Version of the Mucolytic for Patients with Cystic Fibrosis. Advanced Therapeutics, 2021, 4, 2000146.	1.6	7
179	Therapeutic potential for coxibs-nitric oxide releasing hybrids in cystic fibrosis. European Journal of Medicinal Chemistry, 2021, 210, 112983.	2.6	4
180	Choline in cystic fibrosis: relations to pancreas insufficiency, enterohepatic cycle, PEMT and intestinal microbiota. European Journal of Nutrition, 2021, 60, 1737-1759.	1.8	18
181	Drug development for cystic fibrosis. Pediatric Pulmonology, 2021, 56, S10-S22.	1.0	6
182	Aerosolized lancovutide in adolescents (â‰¥12 years) and adults with cystic fibrosis â€“ a randomized trial. Journal of Cystic Fibrosis, 2021, 20, 61-67.	0.3	6
183	Diagnosis of cystic fibrosis in adulthood and eligibility for novel CFTR modulator therapy. Postgraduate Medical Journal, 2022, 98, 341-345.	0.9	2
184	Reduction of pulmonary exacerbations in young children with cystic fibrosis during the COVIDâ€19 pandemic. Pediatric Pulmonology, 2021, 56, 1271-1273.	1.0	31
185	Current and novel therapeutic strategies for the management of cystic fibrosis. Expert Opinion on Drug Delivery, 2021, 18, 535-552.	2.4	7
186	Early Diagnosis and Intervention in Cystic Fibrosis: Imagining the Unimaginable. Frontiers in Pediatrics, 2020, 8, 608821.	0.9	11

#	Article	IF	Citations
187	The short-term effects of ORKAMBI (lumacaftor/ivacaftor) on regional and distal lung structures using functional respiratory imaging. Therapeutic Advances in Respiratory Disease, 2021, 15, 175346662110467.	1.0	4
188	Novel Immunomodulatory Therapies for Respiratory Pathologies. , 2022, , 554-594.		5
189	Current trends in candidate selection, contraindications, and indications for lung transplantation. Journal of Thoracic Disease, 2021, 13, 6514-6527.	0.6	9
190	The PROSPECT Is Bright for CFTR Modulators. Annals of the American Thoracic Society, 2021, 18, 32-33.	1.5	5
191	Introducing the Adult Cystic Fibrosis Series. Chest, 2021, 159, 3-4.	0.4	2
192	Safety of research bronchoscopy with BAL in stable adult patients with cystic fibrosis. PLoS ONE, 2021, 16, e0245696.	1.1	3
193	Modulation of cAMP metabolism for CFTR potentiation in human airway epithelial cells. Scientific Reports, 2021, 11, 904.	1.6	5
194	Gender-affirming hormone therapy in cystic fibrosis â€“ A case of new Pseudomonas infection. Respiratory Medicine Case Reports, 2021, 32, 101353.	0.2	4
195	Lingering Identity as Chronically Ill and the Unanticipated Effects of Life-Changing Precision Medicine in Cystic Fibrosis: A Case Report. Journal of Patient Experience, 2021, 8, 237437352199697.	0.4	2
196	Animal Models and Their Role in Understanding the Pathophysiology of Cystic Fibrosisâ€“Associated Gastrointestinal Lesions. Annual Review of Pathology: Mechanisms of Disease, 2021, 16, 51-67.	9.6	5
197	Intracellular Trafficking of G Protein-Coupled Receptors to the Cell Surface Plasma Membrane in Health and Disease. , 2021, , 375-412.		2
198	Entering the era of highly effective modulator therapies. Pediatric Pulmonology, 2021, 56, S79-S89.	1.0	19
199	Rewriting CFTR to cure cystic fibrosis. Progress in Molecular Biology and Translational Science, 2021, 182, 185-224.	0.9	8
200	Production of CFTR-Î”F508 Rabbits. Frontiers in Genetics, 2020, 11, 627666.	1.1	7
202	CFTR modulators: transformative therapies for cystic fibrosis. Journal of Managed Care & Specialty Pharmacy, 2021, 27, 281-284.	0.5	5
203	Associations between peak oxygen uptake, lung function, and bronchiectasis in children with cystic fibrosis in the era of CFTR modulators. Pediatric Pulmonology, 2021, 56, 1490-1495.	1.0	4
204	Urinary stone disease prevalence and associations in cystic fibrosis. Urolithiasis, 2021, 49, 415-423.	1.2	6
205	The effectiveness and value of novel treatments for cystic fibrosis. Journal of Managed Care & Specialty Pharmacy, 2021, 27, 276-280.	0.5	4

#	Article	IF	Citations
206	â€œIl faut continuer Ã poser des questionsâ€•patient reported outcome measures in cystic fibrosis: An anthropological perspective. Journal of Cystic Fibrosis, 2021, 20, e108-e113.	0.3	4
207	Dysfunctional Inflammation in Cystic Fibrosis Airways: From Mechanisms to Novel Therapeutic Approaches. International Journal of Molecular Sciences, 2021, 22, 1952.	1.8	14
208	Rapid Improvement after Starting Elexacaftorâ€“Tezacaftorâ€“Ivacaftor in Patients with Cystic Fibrosis and Advanced Pulmonary Disease. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 64-73.	2.5	139
209	An evaluation of healthcare utilization and clinical charges in children and adults with cystic fibrosis. Pediatric Pulmonology, 2021, 56, 928-938.	1.0	7
210	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
212	Asthma in Cystic Fibrosis: Definitions and Implications of This Overlap Syndrome. Current Allergy and Asthma Reports, 2021, 21, 9.	2.4	7
213	Lumacaftor/ivacaftor in cystic fibrosis: effects on glucose metabolism and insulin secretion. Journal of Endocrinological Investigation, 2021, 44, 2213-2218.	1.8	20
214	Cystic Fibrosis. Pediatrics in Review, 2021, 42, 55-67.	0.2	46
215	CRISPR/Cas9 gene editing therapies for cystic fibrosis. Expert Opinion on Biological Therapy, 2021, 21, 1-14.	1.4	9
216	Cystic fibrosis patients of minority race and ethnicity less likely eligible for CFTR modulators based on CFTR genotype. Pediatric Pulmonology, 2021, 56, 1496-1503.	1.0	81
217	De nouveaux espoirs de traitements dans la mucoviscidose. Actualites Pharmaceutiques, 2021, 60, 46-50.	0.0	0
218	A formalized transition program for cystic fibrosis: A 10â€year retrospective analysis of 97 patients in Lyon. Pediatric Pulmonology, 2021, 56, 2000-2006.	1.0	1
219	Reflex zone stimulation reduces ventilation inhomogeneity in cystic fibrosis: A randomised controlled crossâ€over study. Pediatric Pulmonology, 2021, 56, 1558-1565.	1.0	0
224	Rescue from Pseudomonas aeruginosa Airway Infection via Stem Cell Transplantation. Molecular Therapy, 2021, 29, 1324-1334.	3.7	6
225	Cystic fibrosis transmembrane conductance regulator modulators for cystic fibrosis: a new dawn?. Archives of Disease in Childhood, 2021, 106, 941-945.	1.0	9
226	CFTR Modulator Therapy with Lumacaftor/Ivacaftor Alters Plasma Concentrations of Lipid-Soluble Vitamins A and E in Patients with Cystic Fibrosis. Antioxidants, 2021, 10, 483.	2.2	19
227	Therapeutic peptides for the treatment of cystic fibrosis: Challenges and perspectives. European Journal of Medicinal Chemistry, 2021, 213, 113191.	2.6	8
228	Soft, skin-interfaced sweat stickers for cystic fibrosis diagnosis and management. Science Translational Medicine, 2021, 13, .	5.8	65

#	Article	IF	Citations
229	Synthetic Approaches to the New Drugs Approved during 2019. Journal of Medicinal Chemistry, 2021, 64, 3604-3657.	2.9	30
230	The march towards CFTR modulator access for all people with CF: The end of the beginning. Journal of Cystic Fibrosis, 2021, 20, 185-187.	0.3	1
231	Modulators of CFTR. Updates on clinical development and future directions. European Journal of Medicinal Chemistry, 2021, 213, 113195.	2.6	39
232	Discrete choice experiment (DCE) to quantify the influence of trial features on the decision to participate in cystic fibrosis (CF) clinical trials. BMJ Open, 2021, 11, e045803.	0.8	3
233	MucoviscidoseÂ: un second souffle. Perfectionnement En PÃ©diatrie, 2021, 4, 5-9.	0.0	0
234	Stability Prediction for Mutations in the Cytosolic Domains of Cystic Fibrosis Transmembrane Conductance Regulator. Journal of Chemical Information and Modeling, 2021, 61, 1762-1777.	2.5	7
235	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
236	Airway Epithelial Inflammation In Vitro Augments the Rescue of Mutant CFTR by Current CFTR Modulator Therapies. Frontiers in Pharmacology, 2021, 12, 628722.	1.6	20
237	Would it have happened without Christmas? Thoracic emphysema and allergic bronchopulmonary aspergillosis in a juvenile cystic fibrosis patient with the G551D mutation receiving ivacaftor. ERJ Open Research, 2021, 7, 00758-2020.	1.1	2
238	Ex vivo model predicted in vivo efficacy of CFTR modulator therapy in a child with rare genotype. Molecular Genetics & Genomic Medicine, 2021, 9, e1656.	0.6	21
239	New insights into structure and function of bis-phosphinic acid derivatives and implications for CFTR modulation. Scientific Reports, 2021, 11, 6842.	1.6	9
240	Modulation of Ion Transport to Restore Airway Hydration in Cystic Fibrosis. Genes, 2021, 12, 453.	1.0	7
241	Discovery of CFTR modulators for the treatment of cystic fibrosis. Expert Opinion on Drug Discovery, 2021, 16, 897-913.	2.5	38
242	Nasal Epithelial Cell-Based Models for Individualized Study in Cystic Fibrosis. International Journal of Molecular Sciences, 2021, 22, 4448.	1.8	16
243	Elexacaftorâ€“tezacaftorâ€“ivacaftor: The new paradigm to treat people with cystic fibrosis with at least one p.Phe508del mutation. Current Opinion in Pharmacology, 2021, 57, 81-88.	1.7	12
244	Peripheral lung effect of elexacaftor/tezacaftor/ivacaftor in adult cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 160-163.	0.3	12
245	Aquagenic Wrinkling of the Palm: A Rare Diagnostic Clue of Cystic Fibrosis and the Response to CFTR-Modulating Therapy. Cureus, 2021, 13, e14425.	0.2	2
246	Mycobacterium abscessus in cystic fibrosis. Science, 2021, 372, 465-466.	6.0	11

#	Article	IF	Citations
247	Care of people with cystic fibrosis: What is the role of specialists in Endocrinology and Nutrition?. EndocrinologÃa Diabetes Y NutriciÃ³n (English Ed), 2021, 68, 215-217.	0.1	0
248	Interactions between ABCC4/MRP4 and ABCC7/CFTR in human airway epithelial cells in lung health and disease. International Journal of Biochemistry and Cell Biology, 2021, 133, 105936.	1.2	4
249	Gene Therapy: A Possible Alternative to CFTR Modulators?. Frontiers in Pharmacology, 2021, 12, 648203.	1.6	4
250	AtenciÃ³n a personas con fibrosis quÃstica: Â¿cuÃ¡l es el papel de los especialistas en EndocrinologÃa y NutriciÃ³n?. Endocrinologia, Diabetes Y NutriciÃ“n, 2021, 68, 215-217.	0.1	0
251	DNAJB12 and Hsp70 triage arrested intermediates of N1303K-CFTR for endoplasmic reticulum-associated autophagy. Molecular Biology of the Cell, 2021, 32, 538-553.	0.9	32
252	CF Fungal Disease in the Age of CFTR Modulators. Mycopathologia, 2021, 186, 655-664.	1.3	12
253	Real-World Outcomes of Ivacaftor Treatment in People with Cystic Fibrosis: A Systematic Review. Journal of Clinical Medicine, 2021, 10, 1527.	1.0	18
254	Cystic Fibrosis Lung Disease in the Aging Population. Frontiers in Pharmacology, 2021, 12, 601438.	1.6	9
255	Targeted therapy for cystic fibrosis. Pulmonologiya, 2021, 31, 226-236.	0.2	10
256	Prenatal Ultrasound Suspicion of Cystic Fibrosis in a Multiethnic Population: Is Extensive CFTR Genotyping Needed?. Genes, 2021, 12, 670.	1.0	4
257	A multi-centre, randomized, controlled trial on coaching and telemonitoring in patients with cystic fibrosis: conneCT CF. BMC Pulmonary Medicine, 2021, 21, 131.	0.8	17
258	Lives changed: A new era for people with cystic fibrosis. Journal of Paediatrics and Child Health, 2021, 57, 968-970.	0.4	1
259	Treatment of Pulmonary Disease of Cystic Fibrosis: A Comprehensive Review. Antibiotics, 2021, 10, 486.	1.5	15
260	On the Corner of Models and Cure: Gene Editing in Cystic Fibrosis. Frontiers in Pharmacology, 2021, 12, 662110.	1.6	16
261	Fertility, Pregnancy and Lactation Considerations for Women with CF in the CFTR Modulator Era. Journal of Personalized Medicine, 2021, 11, 418.	1.1	22
262	Potential of Intestinal Current Measurement for Personalized Treatment of Patients with Cystic Fibrosis. Journal of Personalized Medicine, 2021, 11, 384.	1.1	9
263	Pseudomonas aeruginosa infection, but not mono or dual-combination CFTR modulator therapy affects circulating regulatory T cells in an adult population with cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 1072-1079.	0.3	12
264	Challenges in the use of highly effective modulator treatment for cystic fibrosis. Journal of Cystic Fibrosis, 2021, 20, 381-387.	0.3	19

#	Article	IF	Citations
266	The associations between pediatric weight status and cystic fibrosisâ€related diabetes status and healthâ€related quality of life among children and young adults with cystic fibrosis: A systematic review. Pediatric Pulmonology, 2021, 56, 2413-2425.	1.0	2
267	Novel reaction to new cystic fibrosis medication Trikafta. Clinical Case Reports (discontinued), 2021, 9, e04116.	0.2	8
268	CFTR Modulators: Does One Dose Fit All?. Journal of Personalized Medicine, 2021, 11, 458.	1.1	11
269	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
270	Maternal and fetal outcomes following elexacaftor-tezacaftor-ivacaftor use during pregnancy and lactation. Journal of Cystic Fibrosis, 2021, 20, 402-406.	0.3	52
271	CFTR modulator drug desensitization: Preserving the hope of long term improvement. Pediatric Pulmonology, 2021, 56, 2546-2552.	1.0	14
272	Ivacaftor Reduces Inflammatory Mediators in Upper Airway Lining Fluid From Cystic Fibrosis Patients With a G551D Mutation: Serial Non-Invasive Home-Based Collection of Upper Airway Lining Fluid. Frontiers in Immunology, 2021, 12, 642180.	2.2	18
273	Computed Tomographic Changes in Patients with Cystic Fibrosis Treated by Combination Therapy with Lumacaftor and Ivacaftor. Journal of Clinical Medicine, 2021, 10, 1999.	1.0	9
274	Personalized Medicine Based on Nasal Epithelial Cells: Comparative Studies with Rectal Biopsies and Intestinal Organoids. Journal of Personalized Medicine, 2021, 11, 421.	1.1	19
275	Body composition and weight changes after ivacaftor treatment in adults with cystic fibrosis carrying the G551 D cystic fibrosis transmembrane conductance regulator mutation: A double-blind, placebo-controlled, randomized, crossover study with open-label extension. Nutrition, 2021, 85, 111124.	1.1	28
276	Digital technology for delivering and monitoring exercise programs for people with cystic fibrosis. The Cochrane Library, 0, , .	1.5	1
277	Clinical Pharmacokinetic and Pharmacodynamic Considerations in the Drug Treatment of Non-Tuberculous Mycobacteria in Cystic Fibrosis. Clinical Pharmacokinetics, 2021, 60, 1081-1102.	1.6	4
278	Partial Rescue of F508del-CFTR Stability and Trafficking Defects by Double Corrector Treatment. International Journal of Molecular Sciences, 2021, 22, 5262.	1.8	45
279	Pregnancy in women with Cystic Fibrosis in the 21st century. Journal of Cystic Fibrosis, 2021, 20, 375-376.	0.3	2
280	Catamenial haemoptysis in females with cystic fibrosis: a case series with review of management strategies. Respirology Case Reports, 2021, 9, e00755.	0.3	4
281	Efficacy of Elexacaftor/Tezacaftor/Ivacaftor in Advanced Cystic Fibrosis Lung Disease. Annals of the American Thoracic Society, 2021, 18, 1924-1927.	1.5	20
282	A systematic cochrane review of corrector therapies (with or without potentiators) for people with cystic fibrosis with class II gene variants (most commonly F508DEL). Paediatric Respiratory Reviews, 2021, 38, 33-36.	1.2	1
283	Sex differences in treatment patterns in cystic fibrosis pulmonary exacerbations. Journal of Cystic Fibrosis, 2021, 20, 920-925.	0.3	10

#	Article	IF	Citations
284	The Future of Highly Effective Modulator Therapy in Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2021, 203, 1453-1455.	2.5	2
285	Cardiovascular complications in cystic fibrosis: A review of the literature. Journal of Cystic Fibrosis, 2022, 21, 18-25.	0.3	25
286	A home run for human NaCT/SLC13A5/INDY: cryo-EM structure and homology model to predict transport mechanisms, inhibitor interactions and mutational defects. Biochemical Journal, 2021, 478, 2051-2057.	1.7	6
287	New Therapies to Correct the Cystic Fibrosis Basic Defect. International Journal of Molecular Sciences, 2021, 22, 6193.	1.8	9
288	Cystic fibrosis. Lancet, The, 2021, 397, 2195-2211.	6.3	316
290	Cystic Fibrosis-Related Diabetes (CFRD). , 0, , .		1
291	Pulmonary Complications in Cystic Fibrosis: Past, Present, and Future. Chest, 2021, 160, 1232-1240.	0.4	8
292	Effects of Lumacaftorâ€“Ivacaftor on Lung Clearance Index, Magnetic Resonance Imaging, and Airway Microbiome in Phe508del Homozygous Patients with Cystic Fibrosis. Annals of the American Thoracic Society, 2021, 18, 971-980.	1.5	65
294	Interval versus constant-load exercise training in adults with Cystic Fibrosis. Respiratory Physiology and Neurobiology, 2021, 288, 103643.	0.7	8
295	CFTR modulator therapy improves cystic fibrosis-related diabetes. But how?. Journal of Diabetes and Its Complications, 2021, 35, 107887.	1.2	0
298	Enhanced delivery of peptide-morpholino oligonucleotides with a small molecule to correct splicing defects in the lung. Nucleic Acids Research, 2021, 49, 6100-6113.	6.5	13
300	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
301	Chronic rhinosinusitis in cystic fibrosis: a review of therapeutic options. European Archives of Oto-Rhino-Laryngology, 2022, 279, 1-24.	0.8	12
302	Sweat Chloride Testing and Nasal Potential Difference (NPD) Are Primary Outcome Parameters in Treatment with Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Modulators. Journal of Personalized Medicine, 2021, 11, 729.	1.1	12
304	Validation of nasospheroids to assay CFTR functionality and modulator responses in cystic fibrosis. Scientific Reports, 2021, 11, 15511.	1.6	6
305	CFTR Modulators to the Rescue of Individuals with Cystic Fibrosis and Advanced Lung Disease. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 7-9.	2.5	4
306	Pharmacological Modulation of Ion Channels for the Treatment of Cystic Fibrosis. Journal of Experimental Pharmacology, 2021, Volume 13, 693-723.	1.5	24
307	Elexacaftorâ€“Tezacaftorâ€“Ivacaftor Therapy for Cystic Fibrosis Patients with The F508del/Unknown Genotype. Antibiotics, 2021, 10, 828.	1.5	14

#	Article	IF	Citations
308	CFTR function and clinical response to modulators parallel nasal epithelial organoid swelling. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2021, 321, L119-L129.	1.3	19
309	Magnetic Resonance Imaging Detects Progression of Lung Disease and Impact of Newborn Screening in Preschool Children with Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 943-953.	2.5	41
310	Combination CFTR modulator therapy in children and adults with cystic fibrosis. Lancet Respiratory Medicine,the, 2021, 9, 677-679.	5.2	4
311	A Precision Medicine Approach to Optimize Modulator Therapy for Rare CFTR Folding Mutants. Journal of Personalized Medicine, 2021, 11, 643.	1.1	20
312	A small molecule that induces translational readthrough of CFTR nonsense mutations by eRF1 depletion. Nature Communications, 2021, 12, 4358.	5.8	59
313	Effectiveness of Elexacaftor/Tezacaftor/Ivacaftor Therapy in Three Subjects with the Cystic Fibrosis Genotype Phe508del/Unknown and Advanced Lung Disease. Genes, 2021, 12, 1178.	1.0	15
314	Prevalence, Characteristics and Preoperative Predictors of Chronic Pain After Double-Lung Transplantation: A Prospective Cohort Study. Journal of Cardiothoracic and Vascular Anesthesia, 2022, 36, 500-509.	0.6	10
315	Disease burden in people with cystic fibrosis heterozygous for F508del and a minimal function mutation. Journal of Cystic Fibrosis, 2022, 21, 96-103.	0.3	2
316	Clostridioides difficile colonization and infection in a cohort of Australian adults with cystic fibrosis. Journal of Hospital Infection, 2021, 113, 44-51.	1.4	4
317	A new era for people with cystic fibrosis. European Journal of Pediatrics, 2021, 180, 2731-2739.	1.3	40
318	The rescue of F508del-CFTR by elexacaftor/tezacaftor/ivacaftor (Trikafta) in human airway epithelial cells is underestimated due to the presence of ivacaftor. European Respiratory Journal, 2022, 59, 2100671.	3.1	23
319	A case of self-limited drug induced liver injury under treatment with elexacaftor/tezacaftor/ivacaftor: When it is worth taking the risk. Journal of Cystic Fibrosis, 2021, 20, 712-714.	0.3	9
320	Update in Pediatrics 2020. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 274-284.	2.5	0
321	Hypertonic saline in people with cystic fibrosis: review of comparative studies and clinical practice. Italian Journal of Pediatrics, 2021, 47, 168.	1.0	7
322	Elexacaftor/tezacaftor/ivacaftor outpatient desensitization. Annals of Allergy, Asthma and Immunology, 2022, 128, 104-105.	0.5	5
323	Triple Therapy for Cystic Fibrosis Phe508delâ€“Gating and â€“Residual Function Genotypes. New England Journal of Medicine, 2021, 385, 815-825.	13.9	140
324	Use of elexacaftor/tezacaftor/ivacaftor in liver transplant patients with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 227-229.	0.3	15
325	Pregnancy in cystic fibrosis: Review of the literature and expert recommendations. Journal of Cystic Fibrosis, 2022, 21, 387-395.	0.3	28

#	Article	IF	Citations
326	Optimism with Caution: Elexacaftorâ€“Tezacaftorâ€“Ivacaftor in Patients with Advanced Pulmonary Disease. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 371-372.	2.5	2
327	Review of CFTR modulators 2020. Pediatric Pulmonology, 2021, 56, 3595-3606.	1.0	30
328	Theratyping cystic fibrosis in vitro in ALI culture and organoid models generated from patient-derived nasal epithelial conditionally reprogrammed stem cells. European Respiratory Journal, 2021, 58, 2100908.	3.1	39
329	Management of Individual Patient Expectations When Starting with Highly Effective CFTR Modulators. Journal of Personalized Medicine, 2021, 11, 811.	1.1	1
330	Cystic Fibrosis Transmembrane Conductance Regulator Modulators During Pregnancy: A Case Series. Cureus, 2021, 13, e17427.	0.2	6
332	Impact of Cystic Fibrosis Transmembrane Conductance Regulator Therapy on Chronic Rhinosinusitis and Health Status: Deep Learning CT Analysis and Patient-reported Outcomes. Annals of the American Thoracic Society, 2022, 19, 12-19.	1.5	37
333	Reply to Kuek etÂal.: Optimism with Caution: Elexacaftorâ€“Tezacaftorâ€“Ivacaftor in Patients with Advanced Pulmonary Disease. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 372-374.	2.5	10
334	Efficacy and safety of inhaled ENaC inhibitor BI 1265162 in patients with cystic fibrosis: BALANCE-CF 1, a randomised, phase II study. European Respiratory Journal, 2022, 59, 2100746.	3.1	5
335	The Impact of Highly Effective CFTR Modulators on Growth and Nutrition Status. Nutrients, 2021, 13, 2907.	1.7	27
336	Ivacaftor withdrawal syndrome: A potentially life-threatening consequence from a life-saving medication. Journal of Cystic Fibrosis, 2021, , .	0.3	4
337	The Intestinal Microbiome and Cystic Fibrosis Transmembrane Conductance Regulator Modulators: Emerging Themes in the Management of Gastrointestinal Manifestations of Cystic Fibrosis. Current Gastroenterology Reports, 2021, 23, 17.	1.1	8
338	A comparison of clinic and home spirometry as longtudinal outcomes in cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 78-83.	0.3	25
339	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.	1.5	38
341	Disease characterization of people with cystic fibrosis and a minimal function mutation: Data from the Italian registry. Pediatric Pulmonology, 2021, 56, 3232-3241.	1.0	0
342	CFTR Modulator Therapy and Its Impact on Lung Transplantation in Cystic Fibrosis. Pulmonary Therapy, 2021, 7, 377-393.	1.1	11
343	Triple combination cystic fibrosis transmembrane conductance regulator modulator therapy in the real world â€“ opportunities and challenges. Current Opinion in Pulmonary Medicine, 2021, 27, 554-566.	1.2	17
344	The Effects of Conditioning and Lentiviral Vector Pseudotype on Short- and Long-Term Airway Reporter Gene Expression in Mice. Human Gene Therapy, 2021, 32, 817-827.	1.4	4
345	Familyâ€building and parenting considerations for people with cystic fibrosis. Pediatric Pulmonology, 2022, 57, .	1.0	9

#	Article	IF	Citations
346	Foretelling Early Lung Disease Progression in CF: The Combined Benefits of MRI and Newborn Screen. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 880-881.	2.5	0
347	Anti-Inflammatory and Anti-Oxidant Effect of Dimethyl Fumarate in Cystic Fibrosis Bronchial Epithelial Cells. Cells, 2021, 10, 2132.	1.8	7
348	Elexacaftor/tezacaftor/ivacaftor as rescue therapy in a patient with the cystic fibrosis genotype F508DEL / G1244E. Clinical Case Reports (discontinued), 2021, 9, e04713.	0.2	1
349	Cyclic nucleotide phosphodiesterase inhibitors as therapeutic interventions for cystic fibrosis. , 2021, 224, 107826.		14
350	CFTR modulator use in post lung transplant recipients. Journal of Heart and Lung Transplantation, 2021, 40, 1498-1501.	0.3	18
351	Challenges Faced by Women with Cystic Fibrosis. Clinics in Chest Medicine, 2021, 42, 517-530.	0.8	9
352	Respiratory Mycoses: A Call to Action to Recognize, Educate and Invest. Mycopathologia, 2021, 186, 569-573.	1.3	3
353	Non-invasive ventilation is associated with long-term improvements in lung function and gas exchange in cystic fibrosis adults with hypercapnic respiratory failure. Journal of Cystic Fibrosis, 2021, 20, e40-e45.	0.3	3
354	Extracellular phosphate enhances the function of F508del-CFTR rescued by CFTR correctors. Journal of Cystic Fibrosis, 2021, 20, 843-850.	0.3	3
355	Assessing Psychosocial Distress in Cystic Fibrosis: Validation of the â€˜Distress in Cystic Fibrosis Scaleâ€™. Journal of Clinical Psychology in Medical Settings, 2022, 29, 699-708.	0.8	1
356	Olfactory dysfunction in cystic fibrosis: Impact of CFTR modulator therapy. Journal of Cystic Fibrosis, 2022, 21, e141-e147.	0.3	15
357	Challenging the paradigm: moving from umbrella labels to treatable traits in airway disease. Breathe, 2021, 17, 210053.	0.6	8
358	Palliative Care in Pediatric Pulmonology. Children, 2021, 8, 802.	0.6	0
359	SLC26A9 SNP rs7512462 is not associated with lung disease severity or lung function response to ivacaftor in cystic fibrosis patients with G551D-CFTR. Journal of Cystic Fibrosis, 2021, 20, 851-856.	0.3	11
360	Elexacaftor co-potentiates the activity of F508del and gating mutants of CFTR. Journal of Cystic Fibrosis, 2021, 20, 895-898.	0.3	53
361	Optical Measurements of Sweat for in Vivo Quantification of CFTR Function in Individual Sweat Glands. Journal of Cystic Fibrosis, 2021, 20, 824-827.	0.3	8
362	Exasperation with the lack of pulmonary exacerbation treatment standardization. Journal of Cystic Fibrosis, 2021, 20, 901-903.	0.3	0
363	Two Steps Forward: Improving the Management of Cystic Fibrosis Pulmonary Exacerbations. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 1245-1247.	2.5	1

#	Article	IF	Citations
364	State of the Art on Approved Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Modulators and Triple-Combination Therapy. Pharmaceuticals, 2021, 14, 928.	1.7	21
365	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
366	Improved correction of F508del-CFTR biogenesis with a folding facilitator and an inhibitor of protein ubiquitination. Bioorganic and Medicinal Chemistry Letters, 2021, 48, 128243.	1.0	6
367	Short-term effect of elexacaftor-tezacaftor-ivacaftor on lung function and transplant planning in cystic fibrosis patients with advanced lung disease. Journal of Cystic Fibrosis, 2021, 20, 768-771.	0.3	23
368	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
369	Combined agonists act synergistically to increase mucociliary clearance in a cystic fibrosis airway model. Scientific Reports, 2021, 11, 18828.	1.6	1
370	Optimizing sexual and reproductive health across the lifespan in people with cystic fibrosis. Pediatric Pulmonology, 2022, 57, .	1.0	8
371	Rethinking physical exercise training in the modern era of cystic fibrosis: A step towards optimising short-term efficacy and long-term engagement. Journal of Cystic Fibrosis, 2022, 21, e83-e98.	0.3	17
372	Ethical Dilemma. Chest, 2022, 161, 773-780.	0.4	4
373	Cancer in Cystic Fibrosis: A Narrative Review of Prevalence, Risk Factors, Screening, and Treatment Challenges. Chest, 2022, 161, 356-364.	0.4	18
374	Miracles in my time: Reflections of a pediatric respiratory physician. Pediatric Pulmonology, 2021, 56, 3586-3591.	1.0	1
375	CFTR Rescue in Intestinal Organoids with GLPG/ABBV-2737, ABBV/GLPG-2222 and ABBV/GLPG-2451 Triple Therapy. Frontiers in Molecular Biosciences, 2021, 8, 698358.	1.6	5
376	Cysteamine Inhibits Glycine Utilisation and Disrupts Virulence in Pseudomonas aeruginosa. Frontiers in Cellular and Infection Microbiology, 2021, 11, 718213.	1.8	7
377	Pharmacokinetic interactions between ivacaftor and cytochrome P450 3A4 inhibitors in people with cystic fibrosis and healthy controls. Journal of Cystic Fibrosis, 2021, 20, e72-e76.	0.3	13
378	Cystic Fibrosisâ€"related Diabetes Is Associated with Worse Lung Function Trajectory despite Ivacaftor Use. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 1343-1345.	2.5	3
380	A Randomized Clinical Trial of Antimicrobial Duration for Cystic Fibrosis Pulmonary Exacerbation Treatment. American Journal of Respiratory and Critical Care Medicine, 2021, 204, 1295-1305.	2.5	45
381	The prevalence of aberrations in body composition in pediatric cystic fibrosis patients and relationships with pulmonary function, bone mineral density, and hospitalizations. Journal of Cystic Fibrosis, 2021, 20, 837-842.	0.3	14
382	Projecting the impact of triple CFTR modulator therapy on intravenous antibiotic requirements in cystic fibrosis using patient registry data combined with treatment effects from randomised trials. Thorax, 2022, 77, 873-881.	2.7	11

#	Article	IF	Citations
383	A mutational approach to dissect the functional role of the putative CFTR â€œPTM-CODEâ€• Journal of Cystic Fibrosis, 2021, 20, 891-894.	0.3	3
385	Survival and Lung Transplant Outcomes for Individuals With Advanced Cystic Fibrosis Lung Disease Living in the United States and Canada. Chest, 2021, 160, 843-853.	0.4	19
387	Retracing changes in cystic fibrosis understanding and management over the past twenty years. Journal of Cystic Fibrosis, 2022, 21, 3-9.	0.3	3
388	Cystic Fibrosis in 2021: â€œThe Times They Are A-Changinâ€• Archivos De Bronconeumologia, 2022, 58, 536-538.	0.4	1
389	Patient perspectives following initiation of elexacaftor-tezacaftor-ivacaftor in people with cystic fibrosis and advanced lung disease. Respiratory Medicine and Research, 2021, 80, 100829.	0.4	16
390	Cystic fibrosis mutation classes in pediatric otitis media â€” Fickle or faulty?. American Journal of Otolaryngology - Head and Neck Medicine and Surgery, 2021, 42, 103067.	0.6	2
391	Cystic Fibrosisâ€”Diagnosis, Genetics and Lifelong Effects. , 2022, , 146-160.		0
392	Cystic Fibrosis Cellular Treatments. , 2022, , 161-178.		0
393	Pediatric interstitial lung disease. , 0, 2, 18-32.		4
394	Providing Care for a Changing CF Population. Respiratory Medicine, 2021, , 105-116.	0.1	0
395	Sex Differences in Cystic Fibrosis Across the Lifespan. Physiology in Health and Disease, 2021, , 145-168.	0.2	1
396	Corrector therapies (with or without potentiators) for people with cystic fibrosis with class II CFTR gene variants (most commonly F508del). The Cochrane Library, 2020, 2020, CD010966.	1.5	27
397	Effect of highly effective modulator therapy on quality of life in adults with cystic fibrosis. International Forum of Allergy and Rhinology, 2021, 11, 75-78.	1.5	20
398	Serum sickness–like reaction following initiation of elexacaftor/tezacaftor/ivacaftor therapy. Pediatric Pulmonology, 2020, 55, 2846-2847.	1.0	7
399	Organoids as a Model for Intestinal Ion Transport Physiology. Physiology in Health and Disease, 2020, , 1-39.	0.2	2
400	Pharmacological approaches for targeting cystic fibrosis nonsense mutations. European Journal of Medicinal Chemistry, 2020, 200, 112436.	2.6	25
401	Gene therapy could offer an inclusive cure for cystic fibrosis. Nature, 2020, 583, S12-S14.	13.7	7
402	An open-source high-content analysis workflow for CFTR function measurements using the forskolin-induced swelling assay. Bioinformatics, 2021, 36, 5686-5694.	1.8	6

#	Article	IF	Citations
403	Evaluation of the Effectiveness of Inâ€line Immobilized Lipase Cartridge in Enterally Fed Patients With Cystic Fibrosis. Journal of Pediatric Gastroenterology and Nutrition, 2021, 72, 18-23.	0.9	9
404	Utilisation, expenditure and cost-effectiveness of cystic fibrosis drugs in Ireland: a retrospective analysis of a national pharmacy claims database. BMJ Open, 2020, 10, e040806.	0.8	6
405	Assessing Human Airway Epithelial Progenitor Cells for Cystic Fibrosis Cell Therapy. American Journal of Respiratory Cell and Molecular Biology, 2020, 63, 374-385.	1.4	23
406	Allosteric folding correction of F508del and rare CFTR mutants by elexacaftor-tezacaftor-ivacaftor (Trikafta) combination. JCI Insight, 2020, 5, .	2.3	159
407	Rescue of multiple class II CFTR mutations by elexacaftor+tezacaftor+ivacaftor mediated in part by the dual activities of elexacaftor as both corrector and potentiator. European Respiratory Journal, 2021, 57, 2002774.	3.1	92
408	Preclinical evaluation of the epithelial sodium channel inhibitor BI 1265162 for treatment of cystic fibrosis. ERJ Open Research, 2020, 6, 00429-2020.	1.1	14
409	First clinical trials of the inhaled epithelial sodium channel inhibitor BI 1265162 in healthy volunteers. ERJ Open Research, 2021, 7, 00447-2020.	1.1	6
410	Evaluation of both exonic and intronic variants for effects on RNA splicing allows for accurate assessment of the effectiveness of precision therapies. PLoS Genetics, 2020, 16, e1009100.	1.5	23
411	P.F508del editing in cells from cystic fibrosis patients. PLoS ONE, 2020, 15, e0242094.	1.1	11
412	Overcoming Immunological Challenges to Helper-Dependent Adenoviral Vector-Mediated Long-Term CFTR Expression in Mouse Airways. Genes, 2020, 11, 565.	1.0	2
413	Real-World Safety of CFTR Modulators in the Treatment of Cystic Fibrosis: A Systematic Review. Journal of Clinical Medicine, 2021, 10, 23.	1.0	69
414	Keep cystic fibrosis patients out of the hospital. Cleveland Clinic Journal of Medicine, 2020, , .	0.6	6
415	Novel therapeutic approaches for the management of cystic fibrosis. Multidisciplinary Respiratory Medicine, 2020, 15, 690.	0.6	12
416	Elexacaftor-Tezacaftor-Ivacaftor: The First Triple-Combination Cystic Fibrosis Transmembrane Conductance Regulator Modulating Therapy. Journal of Pediatric Pharmacology and Therapeutics, 2020, 25, 192-197.	0.3	53
417	Different CFTR modulator combinations downregulate inflammation differently in cystic fibrosis. ELife, 2020, 9, .	2.8	75
418	Demographic characteristics, clinical and laboratory features, and the distribution of pathogenic variants in the CFTR gene in the Cypriot cystic fibrosis (CF) population demonstrate the utility of a national CF patient registry. Orphanet Journal of Rare Diseases, 2021, 16, 409.	1.2	6
419	Chloride transport modulators as drug candidates. American Journal of Physiology - Cell Physiology, 2021, 321, C932-C946.	2.1	15
420	Isotope dilution LC-MS/MS quantification of the cystic fibrosis transmembrane conductance regulator (CFTR) modulators ivacaftor, lumacaftor, tezacaftor, elexacaftor, and their major metabolites in human serum. Clinical Chemistry and Laboratory Medicine, 2021, 60, 82-91.	1.4	8

#	Article	IF	Citations
421	A small molecule CFTR potentiator restores ATPâ€dependent channel gating to the cystic fibrosis mutant G551Dâ€CFTR. British Journal of Pharmacology, 2022, 179, 1319-1337.	2.7	7
422	Elexacaftor is a CFTR potentiator and acts synergistically with ivacaftor during acute and chronic treatment. Scientific Reports, 2021, 11, 19810.	1.6	42
423	Economic evaluation of reproductive carrier screening for recessive genetic conditions: a systematic review. Expert Review of Pharmacoeconomics and Outcomes Research, 2022, 22, 197-206.	0.7	3
425	Three-Dimensional Airway Spheroids and Organoids for Cystic Fibrosis Research. Journal of Respiration, 2021, 1, 229-247.	0.4	4
426	A new platform for high-throughput therapy testing on iPSC-derived lung progenitor cells from cystic fibrosis patients. Stem Cell Reports, 2021, 16, 2825-2837.	2.3	19
427	Airway clearance and exercise for people with cystic fibrosis: Balancing longevity with life. Pediatric Pulmonology, 2022, 57, .	1.0	5
428	Increasing life expectancy in cystic fibrosis: Advances and challenges. Pediatric Pulmonology, 2022, 57, .	1.0	41
429	Pharmacological targeting of endoplasmic reticulum stress in disease. Nature Reviews Drug Discovery, 2022, 21, 115-140.	21.5	162
430	Effects of Tham Nasal Alkalinization on Airway Microbial Communities: A Pilot Study in Non-CF and CF Adults. Annals of Otology, Rhinology and Laryngology, 2022, 131, 1013-1020.	0.6	1
431	Cystic fibrosis-related liver disease: Clinical presentations, diagnostic and monitoring approaches in the era of CFTR modulator therapies. Journal of Hepatology, 2022, 76, 420-434.	1.8	41
432	Effectiveness and safety of elexacaftor/tezacaftor/ivacaftor in patients with cystic fibrosis and advanced lung disease with the Phe508del/minimal function genotype. Respiratory Medicine, 2021, 189, 106646.	1.3	26
433	Spontaneous and iatrogenic hypoglycemia in cystic fibrosis. Journal of Clinical and Translational Endocrinology, 2021, 26, 100267.	1.0	3
434	Precision medicine: Rare diseases and community genetics. Digital Medicine, 2019, 5, 154.	0.1	1
435	Elexacaftor-tezacaftor-ivacaftorâ€"A Combination Therapy for Phe508del Cystic Fibrosis. US Respiratory & Pulmonary Diseases, 2019, 4, 15.	0.2	0
437	Lung Transplantation for Cystic Fibrosis. Respiratory Medicine, 2020, , 229-258.	0.1	0
440	Cardiovascular complications of cystic fibrosis. , 2020, , 108-117.		0
441	Lack of Kcnn4 improves mucociliary clearance in muco-obstructive lung disease. JCI Insight, 2020, 5, .	2.3	11
443	Systematic profiling of temperature- and retinal-sensitive rhodopsin variants by deep mutational scanning. Journal of Biological Chemistry, 2021, 297, 101359.	1.6	9

#	Article	IF	Citations
444	CFTR modulation with elexacaftor-tezacaftor-ivacaftor in people with cystic fibrosis assessed by the Î²-adrenergic sweat rate assay. Journal of Cystic Fibrosis, 2022, 21, 442-447.	0.3	10
445	Gene therapy for cystic fibrosis: new tools for precision medicine. Journal of Translational Medicine, 2021, 19, 452.	1.8	23
446	Knowledge-based approaches to drug discovery for rare diseases. Drug Discovery Today, 2022, 27, 490-502.	3.2	15
447	Pharmacological chaperones improve intra-domain stability and inter-domain assembly via distinct binding sites to rescue misfolded CFTR. Cellular and Molecular Life Sciences, 2021, 78, 7813-7829.	2.4	36
448	Perspectives on the translation of in-vitro studies to precision medicine in Cystic Fibrosis. EBioMedicine, 2021, 73, 103660.	2.7	10
449	The 2020 Joseph W. St Geme, Jr. Leadership Award Address: On Leadership and the Joy of Pediatrics. Pediatrics, 2021, , e2021053872.	1.0	0
451	Optimizations of In Vitro Mucus and Cell Culture Models to Better Predict In Vivo Gene Transfer in Pathological Lung Respiratory Airways: Cystic Fibrosis as an Example. Pharmaceutics, 2021, 13, 47.	2.0	14
452	Targeting cystic fibrosis inflammation in the age of CFTR modulators: focus on macrophages. European Respiratory Journal, 2021, 57, 2003502.	3.1	17
453	Cystic Fibrosis and Chronic Rhinosinusitis: Interventions on the Horizon. , 2020, , 151-169.		0
454	Molecular Physiology and Pharmacology of the Cystic Fibrosis Transmembrane Conductance Regulator. Physiology in Health and Disease, 2020, , 605-670.	0.2	1
455	Toxic epidermal necrolysis induced by cystic fibrosis transmembrane conductance regulator modulators. Contact Dermatitis, 2022, 86, 224-225.	0.8	4
456	Frequency of allele variations in the CFTR gene in a Mexican population. BMC Medical Genomics, 2021, 14, 262.	0.7	0
457	Current evidence on the effect of highly effective CFTR modulation on interleukin-8 in cystic fibrosis. Expert Review of Respiratory Medicine, 2022, 16, 43-56.	1.0	5
459	Effects of a Partially Supervised Conditioning Program in Cystic Fibrosis: An International Multicenter, Randomized Controlled Trial (ACTIVATE-CF). American Journal of Respiratory and Critical Care Medicine, 2022, 205, 330-339.	2.5	16
460	Overweight and Cystic Fibrosis: An Unexpected Challenge. Pediatric Pulmonology, 2021, , .	1.0	7
463	Lumacaftor/ivacaftor-associated health stabilisation in adults with severe cystic fibrosis. ERJ Open Research, 2021, 7, 00203-2020.	1.1	10
464	Review of Rapid Advances in Cystic Fibrosis. Missouri Medicine, 2020, 117, 548-554.	0.3	0
465	Sweat metabolomics before and after intravenous antibiotics for pulmonary exacerbation in people with cystic fibrosis. Respiratory Medicine, 2022, 191, 106687.	1.3	2

#	Article	IF	Citations
466	Apparent Yield Stress of Sputum as a Relevant Biomarker in Cystic Fibrosis. Cells, 2021, 10, 3107.	1.8	10
467	A restructuring of microbiome niche space is associated with Elexacaftor-Tezacaftor-Ivacaftor therapy in the cystic fibrosis lung. Journal of Cystic Fibrosis, 2022, 21, 996-1005.	0.3	34
468	Impact of Airway Inflammation on the Efficacy of CFTR Modulators. Cells, 2021, 10, 3260.	1.8	10
469	Lung transplantation for idiopathic pulmonary fibrosis enriches for individuals with telomere-mediated disease. Journal of Heart and Lung Transplantation, 2022, 41, 654-663.	0.3	19
470	The use of elexacaftor/tezacaftor/ivacaftor in patients with cystic fibrosis postliver transplant: A case series. Pediatric Pulmonology, 2022, 57, 411-417.	1.0	15
471	The effect of the cystic fibrosis care center on outcomes after lung transplantation for cystic fibrosis. Journal of Heart and Lung Transplantation, 2022, 41, 300-307.	0.3	5
472	Urinary sodium/creatinine ratio is a predictor for fractional sodium excretion and related to age in patients with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, e136-e140.	0.3	2
473	Emerging Nonpulmonary Complications for Adults With Cystic Fibrosis. Chest, 2022, 161, 1211-1224.	0.4	2
474	Small-molecule drugs for cystic fibrosis: Where are we now?. Pulmonary Pharmacology and Therapeutics, 2022, 72, 102098.	1.1	26
475	Generation of functional ciliated cholangiocytes from human pluripotent stem cells. Nature Communications, 2021, 12, 6504.	5.8	15
476	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
478	Disconcerting and Counterintuitive Findings from a Trial of Exercise in Cystic Fibrosis: Can Exercise Make Our Patients Worse?. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 269-270.	2.5	1
479	Impact of elexacaftor/tezacaftor/ivacaftor on vitamin D absorption in cystic fibrosis patients. Pediatric Pulmonology, 2022, 57, 655-657.	1.0	11
480	Effect of elexacaftor-tezacaftor-ivacaftor on body weight and metabolic parameters in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 265-271.	0.3	64
481	Synergy in Cystic Fibrosis Therapies: Targeting SLC26A9. International Journal of Molecular Sciences, 2021, 22, 13064.	1.8	14
482	What's in a name: the importance of lung transplant at Cystic Fibrosis Foundation Accredited Care Centers for patients with Cystic Fibrosis. Journal of Heart and Lung Transplantation, 2021, , .	0.3	0
483	Latest Advances in Gene Therapy in Management of Cystic Fibrosis Lung Disease, Literature Review. Journal of Biochemical Technology, 2021, 12, 67-70.	0.1	0
484	Exploring provider attitudes and perspectives related to men's health in cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 652-656.	0.3	4

#	Article	IF	Citations
485	Systems Approaches to Unravel Molecular Function: High-content siRNA Screen Identifies TMEM16A Traffic Regulators as Potential Drug Targets for Cystic Fibrosis. Journal of Molecular Biology, 2022, 434, 167436.	2.0	3
486	Small molecule protein binding to correct cellular folding or stabilize the native state against misfolding and aggregation. Current Opinion in Structural Biology, 2022, 72, 267-278.	2.6	21
487	The role of modulators in cystic fibrosis related diabetes. Journal of Clinical and Translational Endocrinology, 2022, 27, 100286.	1.0	15
488	Gestational and pregestational diabetes in pregnant women with cystic fibrosis. Journal of Clinical and Translational Endocrinology, 2022, 27, 100289.	1.0	3
489	MucoviscidoseÂ: une trithÃ©rapie. Pourlascience Fr, 2020, NÂ° 507 - janvier, 9b-9b.	0.0	0
490	Paciente pediÃ¡trico con fibrosis quÃstica. Revista Medica Sinergia, 2020, 5, e503.	0.0	0
491	Assays of CFTR Function In Vitro, Ex Vivo and In Vivo. International Journal of Molecular Sciences, 2022, 23, 1437.	1.8	13
492	Receptor-mediated activation of CFTR via prostaglandin signaling pathways in the airway. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2022, 322, L305-L314.	1.3	6
493	Cystic Fibrosis: A Disease in Transformation, yet More Work to Be Done!. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 487-489.	2.5	2
494	Medicina de precisiÃ³n en fibrosis quÃstica. Revista MÃ©dica ClÃnica Las Condes, 2022, 33, 44-50.	0.2	0
495	Frequency of CFTR variants in southern Brazil and indication for modulators therapy in patients with cystic fibrosis. Genetics and Molecular Biology, 2022, 45, e20200275.	0.6	4
496	Trikafta Rescues CFTR and Lowers Monocyte P2X7R-induced Inflammasome Activation in Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 783-794.	2.5	26
497	The Changing Face of Cystic Fibrosis: An Update for Anesthesiologists. Anesthesia and Analgesia, 2022, Publish Ahead of Print, .	1.1	3
498	Just breathe: a review of sex and gender in chronic lung disease. European Respiratory Review, 2022, 31, 210111.	3.0	32
499	COUNTERPOINT: In the Era of Cystic Fibrosis Transmembrane Regulator Protein Modulator Therapy, Are the Treatment Goals for Adults Now Different From Those for Children With Cystic Fibrosis? No. Chest, 2022, 161, 21-24.	0.4	2
500	Body Mass Index Recovery after Lung Transplant for Cystic Fibrosis. Annals of the American Thoracic Society, 2022, 19, 1130-1138.	1.5	6
501	Measuring the impact of an empiric antibiotic algorithm for pulmonary exacerbation in children and young adults with cystic fibrosis. Pediatric Pulmonology, 2022, , .	1.0	3
502	Open reading frame correction using splice-switching antisense oligonucleotides for the treatment of cystic fibrosis. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, .	3.3	16

#	Article	IF	Citations
503	The management of cystic fibrosis chronic rhinosinusitis: An evidencedâ€based review with recommendations. International Forum of Allergy and Rhinology, 2022, 12, 1148-1183.	1.5	11
504	POINT: In the Era of Cystic Fibrosis Transmembrane Regulator Protein Modulator Therapy, Are the Treatment Goals for Adults Now Different From Those for Children With Cystic Fibrosis? Yes. Chest, 2022, 161, 18-20.	0.4	2
505	Recruited Monocytes/Macrophages Drive Pulmonary Neutrophilic Inflammation and Irreversible Lung Tissue Remodeling in Cystic Fibrosis. SSRN Electronic Journal, 0, , .	0.4	0
507	Forskolin induced swelling (FIS) assay in intestinal organoids to guide eligibility for compassionate use treatment in a CF patient with a rare genotype. Journal of Cystic Fibrosis, 2022, 21, 254-257.	0.3	11
508	Physiology and pathophysiology of human airway mucus. Physiological Reviews, 2022, 102, 1757-1836.	13.1	78
509	Rescue of Mutant CFTR Trafficking Defect by the Investigational Compound MCG1516A. Cells, 2022, 11, 136.	1.8	11
510	Exon-skipping antisense oligonucleotides for cystic fibrosis therapy. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, .	3.3	17
511	Measuring the effect of elexacaftor/tezacaftor/ivacaftor combination therapy on the respiratory pump in people with CF using dynamic chest radiography. Journal of Cystic Fibrosis, 2022, 21, 1036-1041.	0.3	6
512	Worldwide rates of diagnosis and effective treatment for cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 456-462.	0.3	112
513	Geographic distribution and phenotype of European people with cystic fibrosis carrying A1006E mutation. Respiratory Medicine, 2022, 192, 106736.	1.3	0
514	Sustained effectiveness of elexacaftor-tezacaftor-ivacaftor in lung transplant candidates with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 489-496.	0.3	38
515	Acute ST-elevation myocardial infarction in two young women with cystic fibrosis and cystic fibrosis-related diabetes. Journal of Cystic Fibrosis, 2022, 21, e44-e47.	0.3	6
516	Men's sexual and reproductive health in cystic fibrosis in the era of highly effective modulator therapiesâ€“A qualitative study. Journal of Cystic Fibrosis, 2022, 21, 657-661.	0.3	7
517	Precision Medicine Based on CFTR Genotype for People with Cystic Fibrosis. Pharmacogenomics and Personalized Medicine, 2022, Volume 15, 91-104.	0.4	8
518	Small molecule SWELL1 complex induction improves glycemic control and nonalcoholic fatty liver disease in murine Type 2 diabetes. Nature Communications, 2022, 13, 784.	5.8	19
519	Changes in Care during the COVID-19 Pandemic for People with Cystic Fibrosis. Annals of the American Thoracic Society, 2022, 19, 1697-1703.	1.5	8
520	CFTR modulator therapy alters plasma sphingolipid profiles in people with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 713-720.	0.3	13
521	The Rapid Reduction of Infection-Related Visits and Antibiotic Use Among People With Cystic Fibrosis After Starting Elexacaftor-Tezacaftor-Ivacaftor. Clinical Infectious Diseases, 2022, 75, 1115-1122.	2.9	19

#	Article	IF	Citations
522	Novel CFTR modulator combinations maximise rescue of G85E and N1303K in rectal organoids. ERJ Open Research, 2022, 8, 00716-2021.	1.1	17
523	Mucosal Immunity in Cystic Fibrosis. Journal of Immunology, 2021, 207, 2901-2912.	0.4	8
524	The Canadian Glomerulonephritis Registry (CGNR) and Translational Research Initiative: Rationale and Clinical Research Protocol. Canadian Journal of Kidney Health and Disease, 2022, 9, 205435812210890.	0.6	1
525	Efficacy of Elexacaftor/Tezacaftor/Ivacaftor in Decreasing Severe Cystic Fibrosis-Related Exacerbations in Specialty Pharmacy Patients and Vulnerable Subgroups: A Retrospective, Pre-Post Study. SSRN Electronic Journal, 0, , .	0.4	0
526	Sleep and obstructive lung disease. , 2022, , .		0
528	Utilization of the Healthy Eating Index in Cystic Fibrosis. Nutrients, 2022, 14, 834.	1.7	3
529	Elexacaftor/Tezacaftor/Ivacaftor in Patients with Cystic Fibrosis Homozygous for the F508del Mutation and Advanced Lung Disease: A 48-Week Observational Study. Journal of Clinical Medicine, 2022, 11, 1021.	1.0	25
530	In vitro assessment of triple combination therapy for the most common disease-associated mutation in cystic fibrosis. European Respiratory Journal, 2022, 59, 2102380.	3.1	0
533	Cystic fibrosis: candidate selection and impact of the cystic fibrosis transmembrane conductance regulator therapy. Current Opinion in Organ Transplantation, 2022, 27, 198-203.	0.8	3
534	How the sweat gland reveals levels of CFTR activity. Journal of Cystic Fibrosis, 2022, 21, 396-406.	0.3	4
535	Cystic fibrosis patients on cystic fibrosis transmembrane conductance regulator modulators have a reduced incidence of cirrhosis. World Journal of Hepatology, 2022, 14, 411-419.	0.8	5
536	A small molecule high throughput screening platform to profile conformational properties of nascent, ribosome-bound proteins. Scientific Reports, 2022, 12, 2509.	1.6	2
538	Effects of Elexacaftor/Tezacaftor/Ivacaftor Therapy on CFTR Function in Patients with Cystic Fibrosis and One or Two F508del Alleles. American Journal of Respiratory and Critical Care Medicine, 2022, 205, 540-549.	2.5	78
539	Magnetic resonance imaging of cystic fibrosis: Multi-organ imaging in the age of CFTR modulator therapies. Journal of Cystic Fibrosis, 2022, 21, e148-e157.	0.3	4
540	Biologic characterization of ABCA3 variants in lung tissue from infants and children with ABCA3 deficiency. Pediatric Pulmonology, 2022, 57, 1325-1330.	1.0	3
541	Health care costs in a randomized trial of antimicrobial duration among cystic fibrosis patients with pulmonary exacerbations. Journal of Cystic Fibrosis, 2022, , .	0.3	2
542	Targeting the E1 ubiquitin-activating enzyme (UBA1) improves elexacaftor/tezacaftor/ivacaftor efficacy towards F508del and rare misfolded CFTR mutants. Cellular and Molecular Life Sciences, 2022, 79, 192.	2.4	11
543	The changing landscape of pediatric lung transplantation. Clinical Transplantation, 2022, 36, e14634.	0.8	11

#	Article	IF	Citations
544	The Changing Landscape of Nutrition in Cystic Fibrosis: The Emergence of Overweight and Obesity. Nutrients, 2022, 14, 1216.	1.7	22
545	Elastic mucus strands impair mucociliary clearance in cystic fibrosis pigs. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, e2121731119.	3.3	7
546	Efficacy and Safety of Triple Combination Cystic Fibrosis Transmembrane Conductance Regulator Modulators in Patients With Cystic Fibrosis: A Meta-Analysis of Randomized Controlled Trials. Frontiers in Pharmacology, 2022, 13, 863280.	1.6	4
547	Translation Reprogramming as a Novel Therapeutic Target in MAFLD. Advanced Biology, 2022, , 2101298.	1.4	1
548	Efficacy and safety of elexacaftor plus tezacaftor plus ivacaftor versus tezacaftor plus ivacaftor in people with cystic fibrosis homozygous for F508del-CFTR: a 24-week, multicentre, randomised, double-blind, active-controlled, phase 3b trial. Lancet Respiratory Medicine,the, 2022, 10, 267-277.	5.2	66
549	Elexacaftor, tezacaftor and ivacaftor: a case of severe rash and approach to desensitisation. BMJ Case Reports, 2022, 15, e247042.	0.2	6
550	The Effect of CFTR Modulators on Airway Infection in Cystic Fibrosis. International Journal of Molecular Sciences, 2022, 23, 3513.	1.8	23
551	Cystic fibrosis lung transplant recipients 10 years of age or younger: Predisposing factors for endâ€stage disease. Pediatric Pulmonology, 2022, , .	1.0	0
552	A survey: Understanding the health and perspectives of people with CF not benefiting from CFTR modulators. Pediatric Pulmonology, 2022, 57, 1253-1261.	1.0	13
553	A new path for CF clinical trials through the use of historical controls. Journal of Cystic Fibrosis, 2022, 21, 293-299.	0.3	3
554	The L467F-F508del Complex Allele Hampers Pharmacological Rescue of Mutant CFTR by Elexacaftor/Tezacaftor/Ivacaftor in Cystic Fibrosis Patients: The Value of the Ex Vivo Nasal Epithelial Model to Address Non-Responders to CFTR-Modulating Drugs. International Journal of Molecular Sciences, 2022, 23, 3175.	1.8	19
555	CFTR Modulator Therapy for Rare CFTR Mutants. Journal of Respiration, 2022, 2, 59-76.	0.4	5
556	Physiologicallyâ€Based Pharmacokineticâ€Led Guidance for Patients With Cystic Fibrosis Taking Elexacaftorâ€Tezacaftorâ€Ivacaftor With Nirmatrelvirâ€Ritonavir for the Treatment of COVIDâ€19. Clinical Pharmacology and Therapeutics, 2022, 111, 1324-1333.	2.3	18
557	Distinct proteostasis states drive pharmacologic chaperone susceptibility for cystic fibrosis transmembrane conductance regulator misfolding mutants. Molecular Biology of the Cell, 2022, 33, mbcE21110578.	0.9	19
558	The Effect of Elexacaftor/Tezacaftor/Ivacaftor on Hospitalizations and Intravenous Antibiotic Use. , 2022, 26, .		5
559	Magnetic resonance imaging detects improvements of pulmonary and paranasal sinus abnormalities in response to elexacaftor/tezacaftor/ivacaftor therapy in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 1053-1060.	0.3	39
560	Proteostasis Regulators in Cystic Fibrosis: Current Development and Future Perspectives. Journal of Medicinal Chemistry, 2022, , .	2.9	9
561	Genetic evidence supports the development of SLC26A9 targeting therapies for the treatment of lung disease. Npj Genomic Medicine, 2022, 7, 28.	1.7	7

#	Article	IF	Citations
562	CFTR Modulator Therapies: Potential Impact on Airway Infections in Cystic Fibrosis. Cells, 2022, 11, 1243.	1.8	14
563	TMEM16A (ANO1) as a therapeutic target in cystic fibrosis. Current Opinion in Pharmacology, 2022, 64, 102206.	1.7	17
564	Bone accrual and structural changes over one year in youth with cystic fibrosis. Journal of Clinical and Translational Endocrinology, 2022, 28, 100297.	1.0	3
565	Translating in vitro CFTR rescue into small molecule correctors for cystic fibrosis using the Library of Integrated Networkâ€based Cellular Signatures drug discovery platform. CPT: Pharmacometrics and Systems Pharmacology, 2022, 11, 240-251.	1.3	4
566	High-Throughput Functional Analysis of CFTR and Other Apically Localized Proteins in iPSC-Derived Human Intestinal Organoids. Cells, 2021, 10, 3419.	1.8	6
567	Nutritional status and lung function in children with pancreatic-sufficient cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 769-776.	0.3	11
568	Progress in precision medicine in cystic fibrosis: a focus on CFTR modulator therapy. Breathe, 2021, 17, 210112.	0.6	10
569	Channels and Transporters of the Pulmonary Lamellar Body in Health and Disease. Cells, 2022, 11, 45.	1.8	5
570	Precision medicine inÂbronchiectasis. Breathe, 2021, 17, 210119.	0.6	9
572	Structure and function of ABCA4 and its role in the visual cycle and Stargardt macular degeneration. Progress in Retinal and Eye Research, 2022, 89, 101036.	7.3	26
573	Rare Trafficking CFTR Mutations Involve Distinct Cellular Retention Machineries and Require Different Rescuing Strategies. International Journal of Molecular Sciences, 2022, 23, 24.	1.8	15
574	Drug exposure to infants born to mothers taking Elexacaftor, Tezacaftor, and Ivacaftor. Journal of Cystic Fibrosis, 2022, 21, 725-727.	0.3	26
575	Cystische Fibrose: Innovative Arzneimittel verlÃngern Lebenserwartung. , 0, , .		0
576	Evaluating barriers to and promoters of telehealth during the COVID-19 pandemic at U.S. cystic fibrosis programs. Journal of Cystic Fibrosis, 2021, 20, 9-13.	0.3	18
577	Idiopathic chronic pancreatitis treated with ivacaftor in a CFTR carrier with methylmalonic acidemia. Journal of Cystic Fibrosis, 2021, , .	0.3	0
578	Outcomes following lung re-transplantation in patients with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 482-488.	0.3	13
579	Immunoglobulin A Mucosal Immunity and Altered Respiratory Epithelium in Cystic Fibrosis. Cells, 2021, 10, 3603.	1.8	9
580	Burden of cystic fibrosis in children <12 years of age prior to the introduction of CFTR modulator therapies. BMJ Open Respiratory Research, 2021, 8, e000998.	1.2	4

#	Article	IF	Citations
581	Rescue of chloride and bicarbonate transport by elexacaftor-ivacaftor-tezacaftor in organoid-derived CF intestinal and cholangiocyte monolayers. Journal of Cystic Fibrosis, 2021, , .	0.3	9
582	Left behind: The potential impact of CFTR modulators on racial and ethnic disparities in cystic fibrosis. Paediatric Respiratory Reviews, 2021, , .	1.2	10
583	Mucus and mucus flake composition and abundance reflect inflammatory and infection status in cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 959-966.	0.3	8
584	Theratyping of the Rare CFTR Variants E193K and R334W in Rectal Organoid-Derived Epithelial Monolayers. Journal of Personalized Medicine, 2022, 12, 632.	1.1	15
585	Onset of systemic arterial hypertension after initiation of elexacaftor/tezacaftor/ivacaftor in adults with cystic fibrosis: A case series. Journal of Cystic Fibrosis, 2022, 21, 885-887.	0.3	12
586	Systemic bis-phosphinic acid derivative restores chloride transport in Cystic Fibrosis mice. Scientific Reports, 2022, 12, 6132.	1.6	2
587	Overview of CF lung pathophysiology. Current Opinion in Pharmacology, 2022, 64, 102214.	1.7	10
596	Atâ€home compounding preparation of slow desensitization of elexacaftor/tezacaftor/ivacaftor for delayed hypersensitivity rash. Pediatric Pulmonology, 2022, 57, 1779-1781.	1.0	6
597	DNA sequencing analysis of cystic fibrosis transmembrane conductance regulator gene identifies cystic fibrosisâ€associated variants in the Severe Asthma Research Program. Pediatric Pulmonology, 2022, 57, 1782-1788.	1.0	3
598	Established and novel human translational models to advance cystic fibrosis research, drug discovery, and optimize CFTR-targeting therapeutics. Current Opinion in Pharmacology, 2022, 64, 102210.	1.7	6
599	Losartan ameliorates TGF-Î²1â€“induced CFTR dysfunction and improves correction by cystic fibrosis modulator therapies. Journal of Clinical Investigation, 2022, 132, .	3.9	7
600	Etiologic Classification of Diffuse Parenchymal (Interstitial) Lung Diseases. Journal of Clinical Medicine, 2022, 11, 1747.	1.0	27
601	Safety, Tolerability, Pharmacokinetics and Pharmacodynamics of Single and Multiple Doses of ION-827359, an Antisense Oligonucleotide Inhibitor of ENaC, in Healthy Volunteers and Patients with Cystic Fibrosis: A Double-Blind, Placebo-Controlled, Dose-Escalation, Phase 1/2a Study. SSRN Electronic Journal, 0, , .	0.4	1
602	Short-Term Effects of Elexacaftor/Tezacaftor/Ivacaftor Combination on Glucose Tolerance in Young People With Cystic Fibrosisâ€”An Observational Pilot Study. Frontiers in Pediatrics, 2022, 10, 852551.	0.9	24
603	<scp>Nonâ€invasive</scp> prenatal diagnosis of single gene disorders by paternal mutation exclusion: 3â€‰years of clinical experience. BJOG: an International Journal of Obstetrics and Gynaecology, 2022, 129, 1879-1886.	1.1	5
604	Use of elexacaftor/tezacaftor/ivacaftor among cystic fibrosis lung transplant recipients. Journal of Cystic Fibrosis, 2022, 21, 745-752.	0.3	23
605	Survival difference between high-risk and low-risk CFTR genotypes after lung transplant. Journal of Heart and Lung Transplantation, 2022, , .	0.3	1
606	Pathological remodeling of distal lung matrix in end-stage cystic fibrosis patients. Journal of Cystic Fibrosis, 2022, 21, 1027-1035.	0.3	4

#	Article	IF	Citations
607	Telehealth Beyond Communicable Diseases. Chest, 2022, 161, 1127-1128.	0.4	0
608	Blood mRNA biomarkers distinguish variable systemic and sputum inflammation at treatment initiation of inhaled antibiotics in cystic fibrosis: A prospective non-randomized trial. PLoS ONE, 2022, 17, e0267592.	1.1	1
609	Effects of Elexacaftor/Tezacaftor/Ivacaftor Therapy on Lung Clearance Index and Magnetic Resonance Imaging in Patients with Cystic Fibrosis and One or Two F508del Alleles. American Journal of Respiratory and Critical Care Medicine, 2022, 206, 311-320.	2.5	49
611	The impact of FDA and EMA regulatory decision-making process on the access to CFTR modulators for the treatment of cystic fibrosis. Orphanet Journal of Rare Diseases, 2022, 17, 188.	1.2	24
612	CFTR dysfunction and targeted therapies: A vision from non-cystic fibrosis bronchiectasis and COPD. Journal of Cystic Fibrosis, 2022, 21, 741-744.	0.3	3
613	Outpatient Pharmacologic Management of Lung Transplant Candidates on the Waiting List. Thoracic Surgery Clinics, 2022, 32, 111-119.	0.4	1
614	The new triple combination in CFTR modulators: A new era in the management of cystic fibrosis. Pneumon, 2022, 35, 1-4.	0.6	0
615	Sustained inhibition of ENaC in CF: Potential RNA-based therapies for mutation-agnostic treatment. Current Opinion in Pharmacology, 2022, 64, 102209.	1.7	4
616	Animal models of cystic fibrosis in the era of highly effective modulator therapies. Current Opinion in Pharmacology, 2022, 64, 102235.	1.7	8
617	Elexacaftor-Tezacaftor-Ivacaftor as a Final Frontier in the Treatment of Cystic Fibrosis: Definition of the Clinical and Microbiological Implications in a Case-Control Study. Pharmaceuticals, 2022, 15, 606.	1.7	7
618	Limitations of standard cost-effectiveness methods for health technology assessment of treatments for rare, chronic diseases: a case study of treatment for cystic fibrosis. Journal of Medical Economics, 2022, 25, 783-791.	1.0	3
619	An Update on CFTR Modulators as New Therapies for Cystic Fibrosis. Paediatric Drugs, 2022, 24, 321-333.	1.3	18
620	Research letter: The impact of elexacaftor/tezacaftor/ivacaftor on adherence to nebulized maintenance therapies in people with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 1080-1081.	0.3	7
621	Elexacaftor/tezacaftor/ivacaftor in children aged 6â€“11 years with cystic fibrosis, at least one F508DEL allele, and advanced lung disease: A 24â€week observational study. Pediatric Pulmonology, 2022, 57, 2253-2256.	1.0	9
622	How Should the Effects of CFTR Modulator Therapy on Cystic Fibrosis Lung Disease be Monitored?. American Journal of Respiratory and Critical Care Medicine, 2022, , .	2.5	2
623	Dose adjustments of Elexacaftor/Tezacaftor/Ivacaftor in response to mental health side effects in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 1061-1065.	0.3	37
624	Recruitment of monocytes primed to express heme oxygenase-1 ameliorates pathological lung inflammation in cystic fibrosis. Experimental and Molecular Medicine, 2022, 54, 639-652.	3.2	4
625	Translational Research in Cystic Fibrosis: From Bench to Beside. Frontiers in Pediatrics, 2022, 10, .	0.9	3

#	Article	IF	Citations
626	New drugs in cystic fibrosis: what has changed in the last decade?. Therapeutic Advances in Chronic Disease, 2022, 13, 204062232210981.	1.1	0
629	CFTR and Gastrointestinal Cancers: An Update. Journal of Personalized Medicine, 2022, 12, 868.	1.1	8
630	Development and Internal Validation of a Prognostic Model of the Probability of Death or Lung Transplantation Within 2 Years for Patients With Cystic Fibrosis and FEV1Ââ‰ 50%ÂPredicted. Chest, 2022, 162, 757-767.	0.4	4
631	Current state of CFTR modulators for treatment of Cystic Fibrosis. Current Opinion in Pharmacology, 2022, 65, 102239.	1.7	19
632	Who are the 10%? - Non eligibility of cystic fibrosis (CF) patients for highly effective modulator therapies. Respiratory Medicine, 2022, 199, 106878.	1.3	15
633	Phenotypic drug discovery: recent successes, lessons learned and new directions. Nature Reviews Drug Discovery, 2022, 21, 899-914.	21.5	81
634	Treatment With LAU-7b Complements CFTR Modulator Therapy by Improving Lung Physiology and Normalizing Lipid Imbalance Associated With CF Lung Disease. Frontiers in Pharmacology, 0, 13, .	1.6	0
635	Insights gained in the pathology of lung disease through singleâ€cell transcriptomics. Journal of Pathology, 2022, 257, 494-500.	2.1	7
636	CFTR Modulators in People with Cystic Fibrosis: Real-World Evidence in France. Cells, 2022, 11, 1769.	1.8	17
637	Medical student acceptance on gene therapy to increase children's well-being with genetic diseases: a study in Indonesia. Future Science OA, 0, , .	0.9	0
638	Elexacaftor-Tezacaftor-Ivacaftor Treatment Reduces Abdominal Symptoms in Cystic Fibrosis-Early results Obtained With the CF-Specific CFAbd-Score. Frontiers in Pharmacology, 0, 13, .	1.6	33
639	No patient left behind! Therapeutic options for cystic fibrosis patients living with lung transplantation. Journal of Cystic Fibrosis, 2022, , .	0.3	0
640	Objective and patientâ€based measures of chronic rhinosinusitis in people with cystic fibrosis treated with highly effective modulator therapy. International Forum of Allergy and Rhinology, 2022, 12, 1435-1438.	1.5	7
641	Targeting of Glycosaminoglycans in Genetic and Inflammatory Airway Disease. International Journal of Molecular Sciences, 2022, 23, 6400.	1.8	5
642	Recombinant Adeno-Associated Virus-Mediated Editing of the G551D Cystic Fibrosis Transmembrane Conductance Regulator Mutation in Ferret Airway Basal Cells. Human Gene Therapy, 2022, 33, 1023-1036.	1.4	8
643	Prospectively evaluating maternal and fetal outcomes in the era of CFTR modulators: the MAYFLOWERS observational clinical trial study design. BMJ Open Respiratory Research, 2022, 9, e001289.	1.2	20
644	Elexacaftor-Tezacaftor-Ivacaftor improve Gastro-Oesophageal reflux and Sinonasal symptoms in advanced cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 807-810.	0.3	12
645	ELX/TEZ/IVA use in cystic fibrosis liver disease: Is the perspective of improved lung function worth the risk?. Journal of Cystic Fibrosis, 2022, 21, 881-884.	0.3	1

#	Article	IF	Citations
646	Emerging medicines to improve the basic defect in cystic fibrosis. Expert Opinion on Emerging Drugs, 0, , .	1.0	3
647	Triangulating variation in the population to define mechanisms for precision management of genetic disease. Structure, 2022, , .	1.6	4
648	Pharmacological Responses of the G542X-CFTR to CFTR Modulators. Frontiers in Molecular Biosciences, 0, 9, .	1.6	1
649	Management of pregnancy in cystic fibrosis. Breathe, 2022, 18, 220005.	0.6	7
650	Towards generalizable predictions for G protein-coupled receptor variant expression. Biophysical Journal, 2022, 121, 2712-2720.	0.2	1
651	One Size Does Not Fit All: The Past, Present and Future of Cystic Fibrosis Causal Therapies. Cells, 2022, 11, 1868.	1.8	12
652	Poor tolerability of cystic fibrosis transmembrane conductance regulator modulator therapy in lung transplant recipients. Pharmacotherapy, 2022, 42, 580-584.	1.2	11
653	Exercise versus airway clearance techniques for people with cystic fibrosis. The Cochrane Library, 2022, 2022, .	1.5	9
654	Chest computed tomography improvement in patients with cystic fibrosis treated with elexacaftor-tezacaftor-ivacaftor: Early report. European Journal of Radiology, 2022, 154, 110421.	1.2	15
655	Mucus-targeting therapies of defective mucus clearance for cystic fibrosis: A short review. Current Opinion in Pharmacology, 2022, 65, 102248.	1.7	5
656	Discovery and SAR of 4-aminopyrrolidine-2-carboxylic acid correctors of CFTR for the treatment of cystic fibrosis. Bioorganic and Medicinal Chemistry Letters, 2022, 72, 128843.	1.0	1
657	Rechallenge of Elexacaftor/Tezacaftor/Ivacaftor After Skin Rash in Two Pediatric Patients. Journal of Pediatric Pharmacology and Therapeutics, 2022, 27, 463-466.	0.3	7
658	Pharmacist to the Rescue: Overcoming Obstacles for Select Patients. Journal of Pediatric Pharmacology and Therapeutics, 2022, 27, 407-408.	0.3	0
659	Effectiveness and Safety of Cystic Fibrosis Transmembrane Conductance Regulator Modulators in Children With Cystic Fibrosis: A Meta-Analysis. Frontiers in Pediatrics, 0, 10, .	0.9	1
660	Lubiprostone is a Non-Selective Activator of cAMP-Gated Ion Channels and Chloride Channel Protein 2 (Clc-2) Has a Minor Role in its Prosecretory Effect in Intestinal Epithelial Cells. Molecular Pharmacology, 2022, 102, 106-115.	1.0	6
661	Hemoptysis and the Risk for Lung Transplant or Death without Transplant in Individuals with Cystic Fibrosis in the United States. Annals of the American Thoracic Society, 2022, 19, 1986-1992.	1.5	2
662	Cystic fibrosis related diabetes is not associated with maximal aerobic exercise capacity in cystic fibrosis: a cross-sectional analysis of an international multicenter trial. Journal of Cystic Fibrosis, 2023, 22, 31-38.	0.3	2
663	Trends in Growth and Maturation in Children with Cystic Fibrosis Throughout Nine Decades. Frontiers in Endocrinology, 0, 13, .	1.5	4

#	Article	IF	Citations
664	A Splice Switch in SIGIRR Causes a Defect of IL-37-Dependent Anti-Inflammatory Activity in Cystic Fibrosis Airway Epithelial Cells. International Journal of Molecular Sciences, 2022, 23, 7748.	1.8	1
665	A case of Elexacaftor-Tezacaftor-Ivacaftor induced rash resolving without interruption of treatment. Journal of Cystic Fibrosis, 2022, 21, 1077-1079.	0.3	6
666	Insulin-Like Growth Factor Binding Protein (IGFBP-6) as a Novel Regulator of Inflammatory Response in Cystic Fibrosis Airway Cells. Frontiers in Molecular Biosciences, 0, 9, .	1.6	5
667	Cystic fibrosis: a comprehensive review. Reviews in Biological and Biomedical Sciences, 2022, 4, 43-57.	0.1	0
668	Molecular basis for variations in the sensitivity of pathogenic rhodopsin variants to 9-cis-retinal. Journal of Biological Chemistry, 2022, 298, 102266.	1.6	7
669	The Changing Epidemiology of Cystic Fibrosis. Chest, 2023, 163, 89-99.	0.4	16
670	Limitations of the dichotomized 6-minute walk distance when computing lung allocation score for cystic fibrosis: a 16-year retrospective cohort study. Disability and Rehabilitation, 2023, 45, 2578-2584.	0.9	1
671	Efficacy and Safety of Elexacaftor/Tezacaftor/Ivacaftor in Children 6 Through 11 Years of Age with Cystic Fibrosis Heterozygous for F508del and a Minimal Function Mutation: A Phase 3b, Randomized, Placebo-controlled Study. American Journal of Respiratory and Critical Care Medicine, 2022, 206, 1361-1369.	2.5	50
672	Characterizing CFTR modulated sweat chloride response across the cf population: Initial results from the CHEC-SC study. Journal of Cystic Fibrosis, 2023, 22, 79-88.	0.3	4
673	Clinician attitudes and practices on pregnancy planning and care in cystic fibrosis. Journal of Cystic Fibrosis, 2022, , .	0.3	0
674	Treatment Preference Among People With Cystic Fibrosis. Chest, 2022, 162, 1241-1254.	0.4	16
675	Elexacaftorâ€“Tezacaftorâ€“Ivacaftor improves exercise capacity in adolescents with cystic fibrosis. Pediatric Pulmonology, 2022, 57, 2652-2658.	1.0	14
677	Small-molecule eRF3a degraders rescue CFTR nonsense mutations by promoting premature termination codon readthrough. Journal of Clinical Investigation, 2022, 132, .	3.9	11
678	Pseudomonas Aeruginosa Lung Infection Subverts Lymphocytic Responses through IL-23 and IL-22 Post-Transcriptional Regulation. International Journal of Molecular Sciences, 2022, 23, 8427.	1.8	3
679	Biologic drugs in treating allergic bronchopulmonary aspergillosis in patients with cystic fibrosis: a systematic review. European Respiratory Review, 2022, 31, 220011.	3.0	11
680	Randomized controlled phase 2 trial of hydroxychloroquine in childhood interstitial lung disease. Orphanet Journal of Rare Diseases, 2022, 17, .	1.2	18
681	A multimodal iPSC platform for cystic fibrosis drug testing. Nature Communications, 2022, 13, .	5.8	12
682	Congenital etiologies of exocrine pancreatic insufficiency. Frontiers in Pediatrics, 0, 10, .	0.9	4

#	Article	IF	Citations
683	Cystic Fibrosis Modulator Therapies. Annual Review of Medicine, 2023, 74, 413-426.	5.0	13
684	Physical activity and exercise training in cystic fibrosis. Paediatric Respiratory Reviews, 2022, , .	1.2	0
685	Time to change course and tackle CF related obesity. Journal of Cystic Fibrosis, 2022, 21, 732-734.	0.3	5
686	Super resolution microscopy analysis reveals increased Orai1 activity in asthma and cystic fibrosis lungs. Journal of Cystic Fibrosis, 2023, 22, 161-171.	0.3	2
687	Dornase alfa in Cystic Fibrosis: indications, comparative studies and effects on lung clearance index. Italian Journal of Pediatrics, 2022, 48, .	1.0	8
688	An optimized protocol for assessment of sputum macrorheology in health and muco-obstructive lung disease. Frontiers in Physiology, 0, 13, .	1.3	6
689	Targeting trafficking as a therapeutic avenue for misfolded GPCRs leading to endocrine diseases. Frontiers in Endocrinology, 0, 13, .	1.5	7
690	Is CF airway inflammation still relevant in the era of highly effective modulators?. Journal of Cystic Fibrosis, 2022, , .	0.3	0
691	Cystic fibrosis transmembrane conductance regulator modulators and the exocrine pancreas: A scoping review. Journal of Cystic Fibrosis, 2023, 22, 193-200.	0.3	4
692	Competitive fitness of Pseudomonas aeruginosa isolates in human and murine precision-cut lung slices. Frontiers in Cellular and Infection Microbiology, 0, 12, .	1.8	3
693	Physical activity and exercise training in cystic fibrosis. The Cochrane Library, 2022, 2022, .	1.5	7
694	Evidence for Early CFTR Modulator Treatment for Children with Cystic Fibrosis Keeps Growing. American Journal of Respiratory and Critical Care Medicine, 0, , .	2.5	0
695	Diagnostic agreement among experts assessing adults presenting with possible cystic fibrosis: need for improvement and implications for patient care. ERJ Open Research, 2022, 8, 00227-2022.	1.1	1
696	Advances in Preclinical In Vitro Models for the Translation of Precision Medicine for Cystic Fibrosis. Journal of Personalized Medicine, 2022, 12, 1321.	1.1	15
697	Clinical outcomes of digital health in adults with cystic fibrosis. Respiratory Medicine, 2022, 202, 106970.	1.3	0
698	Drug treatment of cystic fibrosis. Australian Prescriber, 2022, 45, 171-175.	0.5	0
699	Phase 1 Study to Assess the Safety and Pharmacokinetics of Elexacaftor/Tezacaftor/Ivacaftor in Subjects Without Cystic Fibrosis With Moderate Hepatic Impairment. European Journal of Drug Metabolism and Pharmacokinetics, 2022, 47, 817-825.	0.6	5
700	Modulator Therapy in Cystic Fibrosis Patients with cis Variants in F508del Complex Allele: A Short-Term Observational Case Series. Journal of Personalized Medicine, 2022, 12, 1421.	1.1	4

#	Article	IF	Citations
701	Nutritional and metabolic management for cystic fibrosis in a post-cystic fibrosis transmembrane conductance modulator era. Current Opinion in Pulmonary Medicine, 2022, 28, 577-583.	1.2	3
702	Infection control in cystic fibrosis: evolving perspectives and challenges. Current Opinion in Pulmonary Medicine, 2022, 28, 571-576.	1.2	0
703	Drugs, Drugs, Drugs: Current Treatment Paradigms in Cystic Fibrosis Airway Infections. Journal of the Pediatric Infectious Diseases Society, 2022, 11, S32-S39.	0.6	4
705	Specific circulating neutrophils subsets are present in clinically stable adults with cystic fibrosis and are further modulated by pulmonary exacerbations. Frontiers in Immunology, 0, 13, .	2.2	7
706	Pulmonary disorders in pregnancy: Bronchiectasis, cystic fibrosis, sarcoidosis and interstitial diseases. Best Practice and Research in Clinical Obstetrics and Gynaecology, 2022, , .	1.4	0
707	Trajectories of early growth and subsequent lung function in cystic fibrosis: An observational study using UK and Canadian registry data. Journal of Cystic Fibrosis, 2023, 22, 388-394.	0.3	2
708	The CFTR Amplifier Nesolicaftor Rescues TGF-Î²1 Inhibition of Modulator-Corrected F508del CFTR Function. International Journal of Molecular Sciences, 2022, 23, 10956.	1.8	7
709	Using the genome to correct the ion transport defect in cystic fibrosis. Journal of Physiology, 2023, 601, 1573-1582.	1.3	1
710	Experience of using lumacaftor/ivacaftor in children with cystic fibrosis in the Astrakhan region. Acta Biomedica Scientifica, 2022, 7, 101-108.	0.1	1
711	Advent of elexacaftor/tezacaftor/ivacaftor for cystic fibrosis treatment: What consequences on Aspergillus-related diseases? Preliminary insights. Journal of Cystic Fibrosis, 2022, , .	0.3	4
712	Non-respiratory health-related quality of life in people with cystic fibrosis receiving elexacaftor/tezacaftor/ivacaftor. Journal of Cystic Fibrosis, 2023, 22, 119-123.	0.3	15
713	Preclinical evaluation of the epithelial sodium channel inhibitor AZD5634 and implications on human translation. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2022, 323, L536-L547.	1.3	2
714	Modulator Combination Improves In Vitro the Microrheological Properties of the Airway Surface Liquid of Cystic Fibrosis Airway Epithelia. International Journal of Molecular Sciences, 2022, 23, 11396.	1.8	5
715	Mucins and CFTR: Their Close Relationship. International Journal of Molecular Sciences, 2022, 23, 10232.	1.8	9
716	Elexacaftor/Tezacaftor/Ivacaftor Accelerates Wound Repair in Cystic Fibrosis Airway Epithelium. Journal of Personalized Medicine, 2022, 12, 1577.	1.1	3
717	Impact of Elexacaftor/Tezacaftor/Ivacaftor Therapy on the Cystic Fibrosis Airway Microbial Metagenome. Microbiology Spectrum, 2022, 10, .	1.2	14
718	Drug-induced acne with elexacaftor/tezacaftor/ivacaftor in people with cystic fibrosis. Journal of Cystic Fibrosis, 2022, 21, 1066-1069.	0.3	8
719	The Elusive Role of Airway Infection in Cystic Fibrosis Exacerbation. Journal of the Pediatric Infectious Diseases Society, 2022, 11, S40-S45.	0.6	3

#	Article	IF	Citations
720	Bronchial artery embolization for hemoptysis in adult patients with cystic fibrosis: a single-center retrospective study. Acta Radiologica, 2023, 64, 1381-1389.	0.5	1
721	Newly Discovered Cutting-Edge Triple Combination Cystic Fibrosis Therapy: A Systematic Review. Cureus, 2022, , .	0.2	5
722	Downstream Alternate Start Site Allows N-Terminal Nonsense Variants to Escape NMD and Results in Functional Recovery by Readthrough and Modulator Combination. Journal of Personalized Medicine, 2022, 12, 1448.	1.1	2
723	Malassezia Folliculitis following Triple Therapy for Cystic Fibrosis. Medicina (Lithuania), 2022, 58, 1204.	0.8	2
724	Antimicrobial Stewardship in Cystic Fibrosis. Journal of the Pediatric Infectious Diseases Society, 2022, 11, S53-S61.	0.6	3
725	KCa3.1 potentiation stimulates Cl⁻ secretion in F508del and G551D CFTR corrected primary human bronchial epithelial cells. American Journal of Physiology - Cell Physiology, 0, , .	2.1	0
726	Role of physical activity and airway clearance therapy in cystic fibrosis: moving forward in a rapidly changing landscape. Thorax, 0, , thorax-2022-219429.	2.7	1
727	Mucus aberrant properties in CF: Insights from cells and animal models. Journal of Cystic Fibrosis, 2023, 22, S23-S26.	0.3	3
728	Innovative cystic fibrosis drug development: A perspective. Respirology, 2022, 27, 1015-1017.	1.3	0
729	Trikaftaâ€"Extending Its Success to Less Common Mutations. Journal of Personalized Medicine, 2022, 12, 1528.	1.1	2
730	Pharmacological chaperone-rescued cystic fibrosis CFTR-F508del mutant overcomes PRAF2-gated access to endoplasmic reticulum exit sites. Cellular and Molecular Life Sciences, 2022, 79, .	2.4	2
732	Sexual and reproductive health experiences and care of adult women with cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, 223-233.	0.3	3
733	Eradication of early MRSA infection in cystic fibrosis: a novel study design for the STAR-ter trial. ERJ Open Research, 2022, 8, 00190-2022.	1.1	4
734	Coronary artery disease in patients with cystic fibrosis â€“ A case series and review of the literature. Journal of Clinical and Translational Endocrinology, 2022, 30, 100308.	1.0	2
735	Phage therapy for pulmonary infections: lessons from clinical experiences and key considerations. European Respiratory Review, 2022, 31, 220121.	3.0	14
736	Exploring the impact of elexacaftor-tezacaftor-ivacaftor treatment on opinions regarding airway clearance techniques and nebulisers: TEMPO a qualitative study in children with cystic fibrosis, their families and healthcare professionals. BMJ Open Respiratory Research, 2022, 9, e001420.	1.2	3
737	Structure basis of CFTR folding, function and pharmacology. Journal of Cystic Fibrosis, 2023, 22, S5-S11.	0.3	5
738	Trends in and Outcomes of Deliveries Complicated by Cystic Fibrosis. Obstetrics and Gynecology, 2022, Publish Ahead of Print, .	1.2	2

#	Article	IF	Citations
740	CGM patterns in adults with cystic fibrosis-related diabetes before and after elexacaftor-tezacaftor-ivacaftor therapy. Journal of Clinical and Translational Endocrinology, 2022, 30, 100307.	1.0	5
741	Pseudomonas aeruginosa in the Cystic Fibrosis Lung. Advances in Experimental Medicine and Biology, 2022, , 347-369.	0.8	6
744	Congenital bilateral cataracts in newborns exposed to elexacaftor-tezacaftor-ivacaftor in utero and while breast feeding. Journal of Cystic Fibrosis, 2022, 21, 1074-1076.	0.3	16
745	ECFS standards of care on CFTR-related disorders: Updated diagnostic criteria. Journal of Cystic Fibrosis, 2022, 21, 908-921.	0.3	18
746	Spatial Heterogeneity of Excess Lung Fluid in Cystic Fibrosis: Generalized, Localized Diffuse, and Localized Presentations. Applied Sciences (Switzerland), 2022, 12, 10647.	1.3	0
748	The relationship between weight and pulmonary outcomes in overweight and obese people with cystic fibrosis: A retrospective observational study. Health Science Reports, 2022, 5, .	0.6	4
750	Curative Measures for Cystic Fibrosis: A Perspective on Current Stem Cellâ€“Based, Gene, and Small Molecule Therapies. Georgetown Medical Review, 0, , .	0.1	0
751	Multicenter prospective study showing a high gastrointestinal symptom burden in cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, 266-274.	0.3	16
752	Standards of care for CFTR variant-specific therapy (including modulators) for people with cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, 17-30.	0.3	22
753	Gene therapy for cystic fibrosis: Challenges and prospects. Frontiers in Pharmacology, 0, 13, .	1.6	12
754	Dynamic Perfluorinated Gas MRI Shows Improved Lung Ventilation in People with Cystic Fibrosis after Elexacaftor/Tezacaftor/Ivacaftor: An Observational Study. Journal of Clinical Medicine, 2022, 11, 6160.	1.0	6
756	Drug Repurposing for Cystic Fibrosis: Identification of Drugs That Induce CFTR-Independent Fluid Secretion in Nasal Organoids. International Journal of Molecular Sciences, 2022, 23, 12657.	1.8	6
757	Effects of elexacaftorâ€“tezacaftorâ€“ivacaftor discontinuation in cystic fibrosis. Respiratory Medicine and Research, 2022, 82, 100972.	0.4	3
758	Quantitation of cystic fibrosis triple combination therapy, elexacaftor/tezacaftor/ivacaftor, in human plasma and cellular lysate. Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences, 2022, 1213, 123518.	1.2	4
759	Instability of Mature ABCA3 Protein: Toward a New Classification of ABCA3 Mutations?. American Journal of Respiratory Cell and Molecular Biology, 2022, 67, 602-605.	1.4	1
760	Challenged Urine Bicarbonate Excretion as a Measure of Cystic Fibrosis Transmembrane Conductance Regulator Function in Cystic Fibrosis. Annals of Internal Medicine, 2022, 175, 1543-1551.	2.0	8
761	The Orphan Liver Disease. In Clinical Practice, 2022, , 287-306.	0.1	0
762	Successful pregnancy in a cystic fibrosis patient with a severe impairment of lung function receiving Elexacaftor-Tezacaftor-Ivacaftor. Respiratory Medicine Case Reports, 2022, 40, 101776.	0.2	5

#	Article	IF	Citations
763	Clinical efficacy of elexacaftor-tezacaftor-ivacaftor in an adolescent with homozygous G85E cystic fibrosis. Respiratory Medicine Case Reports, 2022, 40, 101775.	0.2	2
764	Dual CFTR modulator therapy efficacy in the real world: lessons for the future. ERJ Open Research, 2022, 8, 00464-2022.	1.1	3
765	Culture independent markers of nontuberculous mycobacterial (NTM) lung infection and disease in the cystic fibrosis airway. Tuberculosis, 2023, 138, 102276.	0.8	3
766	Treatment effects of Elexacaftor/Tezacaftor/Ivacaftor in people with CF carrying non-F508del mutations. Journal of Cystic Fibrosis, 2023, 22, 450-455.	0.3	5
767	S945L-CFTR molecular dynamics, functional characterization and tezacaftor/ivacaftor efficacy in vivo and in vitro in matched pediatric patient-derived cell models. Frontiers in Pediatrics, 0, 10, .	0.9	3
768	Modifier Factors of Cystic Fibrosis Phenotypes: A Focus on Modifier Genes. International Journal of Molecular Sciences, 2022, 23, 14205.	1.8	7
770	Essential Fatty Acid Deficiency in Cystic Fibrosis Disease Progression: Role of Genotype and Sex. Nutrients, 2022, 14, 4666.	1.7	2
771	Discontinuation versus continuation of hypertonic saline or dornase alfa in modulator treated people with cystic fibrosis (SIMPLIFY): results from two parallel, multicentre, open-label, randomised, controlled, non-inferiority trials. Lancet Respiratory Medicine,the, 2023, 11, 329-340.	5.2	41
773	Update on Clinical Outcomes of Highly Effective Modulator Therapy. Clinics in Chest Medicine, 2022, 43, 677-695.	0.8	3
774	Non-Modulator Therapies. Clinics in Chest Medicine, 2022, 43, 717-725.	0.8	3
775	The Impact of Highly Effective Modulator Therapy on Cystic Fibrosis Microbiology and Inflammation. Clinics in Chest Medicine, 2022, 43, 647-665.	0.8	13
776	Cystic Fibrosis in the Era of Highly Effective CFTR Modulators. Clinics in Chest Medicine, 2022, 43, xiii-xvi.	0.8	1
777	Update on Lung Transplantation for Cystic Fibrosis. Clinics in Chest Medicine, 2022, 43, 821-840.	0.8	1
778	Family Planning and Reproductive Health in Cystic Fibrosis. Clinics in Chest Medicine, 2022, 43, 811-820.	0.8	6
779	Novel Applications of Biomarkers and Personalized Medicine in Cystic Fibrosis. Clinics in Chest Medicine, 2022, 43, 617-630.	0.8	2
780	Genetics of Cystic Fibrosis. Clinics in Chest Medicine, 2022, 43, 591-602.	0.8	4
781	Toward a Broader Understanding of Cystic Fibrosis Epidemiology and Its Impact on Clinical Manifestations. Clinics in Chest Medicine, 2022, 43, 579-590.	0.8	2
782	Emerging Approaches to Monitor and Modify Care in the era of CFTR Modulators. Clinics in Chest Medicine, 2022, 43, 631-646.	0.8	1

#	Article	IF	Citations
783	Endocrine Complications of Cystic Fibrosis. Clinics in Chest Medicine, 2022, 43, 773-789.	0.8	3
784	Glycemia and Î²-cell function before and after elexacaftor/tezacaftor/ivacaftor in youth and adults with cystic fibrosis. Journal of Clinical and Translational Endocrinology, 2022, 30, 100311.	1.0	7
785	Identification of novel F508del-CFTR traffic correctors among triazole derivatives. European Journal of Pharmacology, 2023, 938, 175396.	1.7	5
786	Common Respiratory Disorders in Children. , 2022, , .		0
787	2022 Update of indications and contraindications for lung transplantation in France. Respiratory Medicine and Research, 2023, 83, 100981.	0.4	0
788	Impact of elexacaftor/tezacaftor/ivacaftor on depression and anxiety in cystic fibrosis. Therapeutic Advances in Respiratory Disease, 2022, 16, 175346662211442.	1.0	21
789	Acneiform Eruption Following Elexacaftor-Tezacaftor-Ivacaftor Treatment in Patients With Cystic Fibrosis. JAMA Dermatology, 0, , .	2.0	4
790	Realâ€life efficacy and safety of elexacaftor/tezacaftor/ivacaftor on severe cystic fibrosis lung disease patients. Pharmacology Research and Perspectives, 2022, 10, .	1.1	8
791	Cystic Fibrosis Patients with F508del/Minimal Function Genotype: Laboratory and Nutritional Evaluations after One Year of Elexacaftor/Tezacaftor/Ivacaftor Treatment. Journal of Clinical Medicine, 2022, 11, 6900.	1.0	13
792	Cystic Fibrosis Screen Positive, Inconclusive Diagnosis Genotypes in People with Cystic Fibrosis from the U.S. Patient Registry. Annals of the American Thoracic Society, 2023, 20, 523-531.	1.5	5
793	Rectal organoid-guided CFTR modulator therapy restores lung function in a cystic fibrosis patient with the rare 1677delTA/R334W genotype. European Respiratory Journal, 2022, 60, 2201341.	3.1	6
794	Impact of Digital Technologies on Clinical Care for Adults with Cystic Fibrosis. Seminars in Respiratory and Critical Care Medicine, 0, , .	0.8	0
795	Elexacaftor/tezacaftor/ivacaftor corrects monocyte microbicidal deficiency in cystic fibrosis. European Respiratory Journal, 2023, 61, 2200725.	3.1	15
796	Diagnosis of cystic fibrosis in adults: Australian Cystic Fibrosis Data Registry data, 2000â€“2019. Medical Journal of Australia, 0, , .	0.8	1
797	The influence of exocrine pancreatic function on the exposure and pharmacokinetics of ivacaftor in people with cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, 564-569.	0.3	2
798	Inflammation and Infection in Cystic Fibrosis: Update for the Clinician. Children, 2022, 9, 1898.	0.6	3
799	Assessing the health risk of living near composting facilities on lung health, fungal and bacterial disease in cystic fibrosis: a UK CF Registry study. Environmental Health, 2022, 21, .	1.7	2
800	A phase I study assessing the safety and tolerability of allogeneic mesenchymal stem cell infusion in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, 407-413.	0.3	3

#	Article	IF	Citations
801	CFTR Modulators: Current Status and Evolving Knowledge. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 186-195.	0.8	2
802	<scp>ISPAD</scp> Clinical Practice Consensus Guidelines 2022: Management of cystic fibrosisâ€related diabetes in children and adolescents. Pediatric Diabetes, 2022, 23, 1212-1228.	1.2	17
803	Exercise and Airway Clearance Techniques in Cystic Fibrosis. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 209-216.	0.8	2
804	Patient-derived cell models for personalized medicine approaches in cystic fibrosis. Journal of Cystic Fibrosis, 2023, 22, S32-S38.	0.3	7
805	Prebiotics for people with cystic fibrosis. The Cochrane Library, 2022, 2022, .	1.5	1
806	Comparison of mental health in individuals with primary ciliary dyskinesia, cystic fibrosis, and parent caregivers. Respiratory Medicine, 2023, 207, 107095.	1.3	1
807	SIMPLIFYing cystic fibrosis treatment in a post-modulator era. Lancet Respiratory Medicine,the, 2023, 11, 299-300.	5.2	1
808	Design of Crotoxin-Based Peptides with Potentiator Activity Targeting the Î”F508NBD1 Cystic Fibrosis Transmembrane Conductance Regulator. Journal of Molecular Biology, 2023, 435, 167929.	2.0	1
809	What Do Adults With Cystic Fibrosis Want From Their Doctors?. Chest, 2022, 162, 1225-1226.	0.4	1
810	Impact of elexacaftorâ€“tezacaftorâ€“ivacaftor on bacterial colonization and inflammatory responses in cystic fibrosis. Pediatric Pulmonology, 2023, 58, 825-833.	1.0	22
811	CFTR pharmacological modulators: A great advance in cystic fibrosis management. Archives De Pediatrie, 2022, , .	0.4	0
812	Respiratory Viruses and Cystic Fibrosis. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 196-208.	0.8	1
813	The CFHealthHub Learning Health System: Using Real-Time Adherence Data to Support a Community of Practice to Deliver Continuous Improvement in an Archetypal Long-Term Condition. Healthcare (Switzerland), 2023, 11, 20.	1.0	2
814	Exercise as an Airway Clearance Technique in people with Cystic Fibrosis (ExACT-CF): rationale and study protocol for a randomised pilot trial. NIHR Open Research, 0, 2, 64.	0.0	0
815	Cystic Fibrosis Related Diabetes (CFRD) in the Era of Modulators: A Scoping Review. Paediatric Respiratory Reviews, 2022, , .	1.2	4
816	Rescue by elexacaftor-tezacaftor-ivacaftor of the G1244E cystic fibrosis mutation's stability and gating defects are dependent on cell background. Journal of Cystic Fibrosis, 2023, 22, 525-537.	0.3	9
817	Longitudinal Magnetic Resonance Imaging Detects Onset and Progression of Chronic Rhinosinusitis from Infancy to School Age in Cystic Fibrosis. Annals of the American Thoracic Society, 2023, 20, 687-697.	1.5	6
818	Elexacaftor/Tezacaftor/Ivacaftor Disrupts Respiratory Tract Development in a Murine Fetal Lung Explant Model. American Journal of Respiratory Cell and Molecular Biology, 2022, 67, 723-726.	1.4	1

#	Article	IF	Citations
819	Normalisation of circulating neutrophil counts after 12 months of elexacaftor-tezacaftor-ivacaftor in patients with advanced cystic fibrosis. European Respiratory Journal, 2023, 61, 2202096.	3.1	7
820	ABC transporters: human disease and pharmacotherapeutic potential. Trends in Molecular Medicine, 2023, 29, 152-172.	3.5	15
821	Fundamental and translational research in Cystic Fibrosis â€“ why we still need it. Journal of Cystic Fibrosis, 2022, , .	0.3	0
822	Cystic fibrosis transmembrane conductance regulator in COPD: a role in respiratory epithelium and beyond. European Respiratory Journal, 2023, 61, 2201307.	3.1	9
823	Recruited monocytes/macrophages drive pulmonary neutrophilic inflammation and irreversible lung tissue remodeling in cystic fibrosis. Cell Reports, 2022, 41, 111797.	2.9	20
824	Beyond the Lungsâ€"Emerging Challenges in Adult Cystic Fibrosis Care. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 307-314.	0.8	3
825	A survey assessing the impact of COVIDâ€19 and elexacaftor/tezacaftor/ifavacaftor on both physical and mental health in adults with cystic fibrosis. Pediatric Pulmonology, 2023, 58, 662-664.	1.0	4
826	Challenges and opportunities in the development of novel antimicrobial therapeutics for cystic fibrosis. Journal of Medical Microbiology, 2022, 71, .	0.7	3
828	Moving the Dial on Airway Inflammation in Response to Trikafta in Adolescents with Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2023, 207, 792-795.	2.5	7
829	Drug-drug interactions with CFTR modulator therapy in cystic fibrosis: Focus on TrikaftaÂ®/KaftrioÂ®. Journal of Cystic Fibrosis, 2023, , .	0.3	2
831	Nutritional Care in Children with Cystic Fibrosis. Nutrients, 2023, 15, 479.	1.7	9
832	Cystic Fibrosis-Related Gut Dysbiosis: A Systematic Review. Digestive Diseases and Sciences, 2023, 68, 1797-1814.	1.1	13
833	Diagnosing Cystic Fibrosis in Adults. Seminars in Respiratory and Critical Care Medicine, 0, , .	0.8	2
834	Effects of elexacaftor/tezacaftor/ivacaftor therapy in children with cystic fibrosis â€“ a comprehensive assessment using lung clearance index, spirometry, and functional and structural lung MRI. Journal of Cystic Fibrosis, 2023, 22, 615-622.	0.3	13
835	Mutual Effects of Single and Combined CFTR Modulators and Bacterial Infection in Cystic Fibrosis. Microbiology Spectrum, 2023, 11, .	1.2	4
836	Current and Future Therapeutic Approaches of Exocrine Pancreatic Insufficiency in Children with Cystic Fibrosis in the Era of Personalized Medicine. Pharmaceutics, 2023, 15, 162.	2.0	3
837	Analyzing the use and impact of elexacaftor/tezacaftor/ivacaftor on total cost of care and other health care resource utilization in a commercially insured population. Journal of Managed Care & Specialty Pharmacy, 2022, 28, 721-731.	0.5	2
838	Impact of pharmacy services on time to elexacaftor-tezacaftor-ivacaftor initiation. Journal of Managed Care & Specialty Pharmacy, 2022, 28, 989-996.	0.5	2

#	Article	IF	Citations
839	EFFECT OF ELEXACAFTOR/TEZACAFTOR/IVACAFTOR ON ANNUAL RATE OF LUNG FUNCTION DECLINE IN PEOPLE WITH CYSTIC FIBROSIS. Journal of Cystic Fibrosis, 2023, 22, 402-406.	0.3	12
841	The Singapore National Precision Medicine Strategy. Nature Genetics, 2023, 55, 178-186.	9.4	9
842	Effects of CFTR modulators on serum biomarkers of liver fibrosis in children with cystic fibrosis. Hepatology Communications, 2023, 7, e0010-e0010.	2.0	9
843	Clinical change 2 years from start of elexacaftorâ€tezacaftorâ€ivacaftor in severe cystic fibrosis. Pediatric Pulmonology, 0, , .	1.0	10
844	Understanding the Experiences of How Mindfulness is Used by People with Cystic Fibrosis: Barriers and Enablers. Mindfulness, 0, , .	1.6	0
845	Improved glucose tolerance after initiation of Elexacaftor / Tezacaftor / Ivacaftor in adults with cystic fibrosis. Journal of Cystic Fibrosis, 2023, , .	0.3	12
846	Menopause in people with cystic fibrosis. Menopause, 2023, 30, 401-405.	0.8	1
847	Therapeutic beta-lactam dosages and broad-spectrum antibiotics are associated with reductions in microbial richness and diversity in persons with cystic fibrosis. Scientific Reports, 2023, 13, .	1.6	1
848	Antisense Oligonucleotide Therapeutics for Cystic Fibrosis: Recent Developments and Perspectives. Molecules and Cells, 2023, 46, 10-20.	1.0	9
849	From genetic variation to precision medicine. , 2023, 1, .		2
850	Longitudinal effects of elexacaftor/tezacaftor/ivacaftor on liver tests at a large single adult cystic fibrosis centre. Journal of Cystic Fibrosis, 2023, 22, 256-262.	0.3	17
851	Nutritional Status and Circulating Levels of Fat-Soluble Vitamins in Cystic Fibrosis Patients: A Cohort Study and Evaluation of the Effect of CFTR Modulators. Children, 2023, 10, 252.	0.6	7
852	Changes in cystic fibrosis transmembrane conductance regulator protein expression prior to and during elexacaftor-tezacaftor-ivacaftor therapy. Frontiers in Pharmacology, 0, 14, .	1.6	4
853	Transition des patients atteints dâ€™une maladie respiratoire chronique depuis la pÃ©diatrie vers les services pour adultes. Medecine/Sciences, 2023, 39, 58-63.	0.0	0
854	Lessons from other fields of medicine, Part 2: Cystic fibrosis. Handbook of Clinical Neurology / Edited By P J Vinken and G W Bruyn, 2023, , 119-130.	1.0	1
855	Autoimmunity in people with cystic fibrosis. Journal of Cystic Fibrosis, 2023, , .	0.3	0
856	Long-term effects of lumacaftor/ivacaftor on paranasal sinus abnormalities in children with cystic fibrosis detected with magnetic resonance imaging. Frontiers in Pharmacology, 0, 14, .	1.6	5
857	Outpatient management of pulmonary exacerbations in children with cystic fibrosis. Expert Review of Respiratory Medicine, 2023, 17, 295-304.	1.0	1

#	Article	IF	Citations
858	A grumbling concern: an international survey of gastrointestinal symptoms in cystic fibrosis in the modulator era. NIHR Open Research, 0, 3, 18.	0.0	2
859	Targeted protein posttranslational modifications by chemically induced proximity for cancer therapy. Journal of Biological Chemistry, 2023, 299, 104572.	1.6	9
864	How many billions is enough? Prioritizing profits over patients with cystic fibrosis. Pediatric Pulmonology, 2023, 58, 1595-1597.	1.0	8
865	Optimization of CFTR gating through the evolution of its extracellular loops. Journal of General Physiology, 2023, 155, .	0.9	3
866	Cystic Fibrosis Transmembrane Conductance Regulator Protein Modulators in Children and Adolescents with Different CF Genotypes - Systematic Review and Meta-Analysis. Current Reviews in Clinical and Experimental Pharmacology, 2024, 19, 93-110.	0.4	1
867	Characterization of CFTR mutations in people with cystic fibrosis and severe liver disease who are not eligible for CFTR modulators. Journal of Cystic Fibrosis, 2023, 22, 263-265.	0.3	1
868	Elexacaftor/tezacaftor/ivacaftorâ€"real-world clinical effectiveness and safety. A single-center Portuguese study. Jornal Brasileiro De Pneumologia, 0, , e20220312.	0.4	1
869	Effects of CFTR-modulator triple therapy on sinunasal symptoms in children and adults with cystic fibrosis. European Archives of Oto-Rhino-Laryngology, 2023, 280, 3271-3277.	0.8	7
870	Diagnosis and Management of Cystic Fibrosis Exacerbations. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 225-241.	0.8	2
871	Clinical Relevance of Fungi in Cystic Fibrosis. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 252-259.	0.8	2
872	Future therapies for cystic fibrosis. Nature Communications, 2023, 14, .	5.8	17
873	The Lung Transplant Candidate, Indications, Timing, and Selection Criteria. Clinics in Chest Medicine, 2023, 44, 15-33.	0.8	0
874	Effects of Elexacaftor/Tezacaftor/Ivacaftor on Cardiorespiratory Polygraphy Parameters and Respiratory Muscle Strength in Cystic Fibrosis Patients with Severe Lung Disease. Genes, 2023, 14, 449.	1.0	7
875	Pregnancy in Cystic Fibrosisâ€"Past, Present, and Future. Journal of Clinical Medicine, 2023, 12, 1468.	1.0	1
876	The Role of MMPs in the Era of CFTR Modulators: An Additional Target for Cystic Fibrosis Patients?. Biomolecules, 2023, 13, 350.	1.8	3
877	Regulatory T cell enhancement in adults with cystic fibrosis receiving Elexacaftor/Tezacaftor/Ivacaftor therapy. Frontiers in Immunology, 0, 14, .	2.2	3
878	The French Compassionate Program of elexacaftor-tezacaftor-ivacaftor in people with cystic fibrosis with advanced lung disease and no F508delCFTRvariant. European Respiratory Journal, 0, , 2202437.	3.1	26
879	Cystic Fibrosis: A Descriptive Analysis of Deaths in a Two-Decade Period in Brazil According to Age, Race, and Sex. Diagnostics, 2023, 13, 763.	1.3	4

#	Article	IF	Citations
880	The Clinical Association betweenAspergillus fumigatusand Respiratory Outcomes in Adolescents and Adults with Cystic Fibrosis. Annals of the American Thoracic Society, 2023, 20, 984-992.	1.5	4
881	Increase of liver stiffness and altered bile acid metabolism after triple <scp>CFTR</scp> modulator initiation in children and young adults with cystic fibrosis. Liver International, 2023, 43, 878-887.	1.9	7
882	â€œFind ways to work parenting into cystic fibrosisâ€: A PhotoVoice exploration of being a parent and having CF. Pediatric Pulmonology, 2023, 58, 1527-1534.	1.0	4
883	Impact of elexacaftor/tezacaftor/ivacaftor on bacterial cultures from people with cystic fibrosis. Pediatric Pulmonology, 2023, 58, 1569-1573.	1.0	7
884	Respiratory Infection and Inflammation in Cystic Fibrosis: A Dynamic Interplay among the Host, Microbes, and Environment for the Ages. International Journal of Molecular Sciences, 2023, 24, 4052.	1.8	3
885	Managing cystic fibrosis in children aged 6-11yrs: a critical review of elexacaftor/tezacaftor/ivacaftor combination therapy. Expert Review of Respiratory Medicine, 2023, 17, 97-108.	1.0	6
886	Incidence of transaminitis in adults with cystic fibrosis taking elexacaftor/tezacaftor/ivacaftor. Journal of the American Pharmacists Association: JAPhA, 2023, 63, 920-924.	0.7	4
888	Immunoreactive Trypsinogen in Infants Born to Women with Cystic Fibrosis Taking Elexacaftorâ€“Tezacaftorâ€“Ivacaftor. International Journal of Neonatal Screening, 2023, 9, 10.	1.2	4
889	Cystic fibrosis and primary ciliary dyskinesia: Similarities and differences. Respiratory Medicine, 2023, 209, 107169.	1.3	5
890	Safety and efficacy of vanzacaftorâ€“tezacaftorâ€“deutivacaftor in adults with cystic fibrosis: randomised, double-blind, controlled, phase 2 trials. Lancet Respiratory Medicine,the, 2023, 11, 550-562.	5.2	14
891	A new triple combination cystic fibrosis transmembrane regulator modulator. Lancet Respiratory Medicine,the, 2023, , .	5.2	0
892	The Impact of Highly Effective Cystic Fibrosis Transmembrane Conductance Regulator Modulators on the Health of Female Subjects With Cystic Fibrosis. Clinical Therapeutics, 2023, 45, 278-289.	1.1	5
893	Elexacaftor/tezacaftor/ivacaftor projected survival and long-term health outcomes in people with cystic fibrosis homozygous for F508del. Journal of Cystic Fibrosis, 2023, 22, 607-614.	0.3	29
894	Safety of elexacaftor/tezacaftor/ivacaftor dose reduction: Mechanistic exploration through physiologically based pharmacokinetic modeling and a clinical case series. Pharmacotherapy, 2023, 43, 291-299.	1.2	8
895	When triple therapy is not working: A reverse iceberg perspective. Journal of Cystic Fibrosis, 2023, 22, 367-369.	0.3	2
896	Revisiting Host-Pathogen Interactions in Cystic Fibrosis Lungs in the Era of CFTR Modulators. International Journal of Molecular Sciences, 2023, 24, 5010.	1.8	5
897	Early and sustained improvements of lung clearance index from two to sixteen weeks of elexacaftor/tezacaftor/ivacaftor therapy in patients with cystic fibrosisâ€”a real world study. Frontiers in Pharmacology, 0, 14, .	1.6	3
898	Elexacaftor-Tezacaftor-Ivacaftor: A Life-Changing Triple Combination of CFTR Modulator Drugs for Cystic Fibrosis. Pharmaceuticals, 2023, 16, 410.	1.7	17

#	Article	IF	Citations
899	Advanced Cystic Fibrosis Lung Disease and Lung Transplantation in the Era of Cystic Fibrosis Transmembrane Conductance Regulator Modulators. Seminars in Respiratory and Critical Care Medicine, 2023, 44, 260-268.	0.8	3
900	The Cystic Fibrosis Upper and Lower Airway Metagenome. Microbiology Spectrum, 2023, 11, .	1.2	7
901	Acute pancreatitis in pancreatic-insufficient cystic fibrosis patients treated with CFTR modulators. Journal of Cystic Fibrosis, 2023, 22, 777-779.	0.3	3
902	Phase 3 Open-Label Clinical Trial of Elexacaftor/Tezacaftor/Ivacaftor in Children Aged 2â€“5 Years with Cystic Fibrosis and at Least One F508del Allele. American Journal of Respiratory and Critical Care Medicine, 2023, 208, 59-67.	2.5	32
903	Efficacy and Safety of Elexacaftor-Tezacaftor-Ivacaftor in the Treatment of Cystic Fibrosis: A Systematic Review. Children, 2023, 10, 554.	0.6	9
904	Personalized medicine: Function of CFTR variant p.Arg334Trp is rescued by currently available CFTR modulators. Frontiers in Molecular Biosciences, 0, 10, .	1.6	1
905	Inequalities in cystic fibrosis. , 2023, , 116-128.		0
906	Post-approval studies with the CFTR modulators Elexacaftor-Tezacaftorâ€”Ivacaftor. Frontiers in Pharmacology, 0, 14, .	1.6	7
907	Effects of Lumacaftor/Ivacaftor on Cystic Fibrosis Disease Progression in Children 2 through 5 Years of Age Homozygous for F508del-CFTR: A Phase 2 Placebo-controlled Clinical Trial. Annals of the American Thoracic Society, 2023, 20, 1144-1155.	1.5	3
908	Real-world safety and effectiveness of elexacaftor/tezacaftor/ivacaftor in people with cystic fibrosis: Interim results of a long-term registry-based study. Journal of Cystic Fibrosis, 2023, 22, 730-737.	0.3	26
909	A paradigm shift in cystic fibrosis nutritional care: Clinicians' views on the management of patients with overweight and obesity. Journal of Cystic Fibrosis, 2023, 22, 836-842.	0.3	3
910	Use of elexacaftor/tezacaftor/ivacaftor combination in pregnancy. Archives of Gynecology and Obstetrics, 2024, 309, 9-15.	0.8	1
911	Differential effects of ELX/TEZ/IVA on organ-specific CFTR function in two patients with the rare CFTR splice mutations c.273+1G>A and c.165-2A>G. Frontiers in Pharmacology, 0, 14, .	1.6	3
912	Pharmacologic improvement of CFTR function rapidly decreases sputum pathogen density, but lung infections generally persist. Journal of Clinical Investigation, 2023, 133, .	3.9	38
913	Cystic Fibrosis Screen Positive, Inconclusive Diagnosis Genotypes in the Cystic Fibrosis Registry. Annals of the American Thoracic Society, 2023, 20, 512-513.	1.5	0
914	The effects of elexacaftor/tezacaftor/ivacaftor beyond the epithelium: spurring macrophages to fight infections. European Respiratory Journal, 2023, 61, 2300216.	3.1	0
915	Complications and Practice Variation in the Use of Peripherally Inserted Central Venous Catheters in People With Cystic Fibrosis. Chest, 2023, 164, 614-624.	0.4	1
916	Isolated abnormal FEF75% detects unsuspected bronchiolar obstruction in CF children. Pediatric Research, 0, , .	1.1	0

#	Article	IF	Citations
917	Pharmacogenetics of cardiovascular drugs. Current Opinion in Cardiology, 2023, 38, 207-214.	0.8	1
918	Management of neuropsychiatric symptoms in adults treated with elexacaftor/tezacaftor/ivacaftor. Pediatric Pulmonology, 2023, 58, 1920-1930.	1.0	11
919	Inflammation as a Regulator of the Airway Surface Liquid pH in Cystic Fibrosis. Cells, 2023, 12, 1104.	1.8	5
920	Antibiotic Therapy for Pulmonary Exacerbations in Cystic Fibrosisâ€"A Single-Centre Prospective Observational Study. Antibiotics, 2023, 12, 734.	1.5	0
921	Survival-Adjusted FEV1 and BMI Percentiles for Patients with Cystic Fibrosis before the Era of Triple CFTR Modulator Therapy in Germany. Respiration, 2023, 102, 1-1.	1.2	0
922	Use of CFTR modulators in special populations, part 3: Solid organ transplant. Pediatric Pulmonology, 2023, 58, 3393-3402.	1.0	1
923	Effectiveness of lumacaftor/ivacaftor in 1-year therapy of cystic fibrosis in adult patients. Meditsinskiy Sovet, 2023, , 102-108.	0.1	0
924	Effects of elexacaftor/tezacaftor/ivacaftor therapy on mental health of patients with cystic fibrosis. Frontiers in Pharmacology, 0, 14, .	1.6	8
966	Rare Disease Research. , 2023, , 123-143.		0
967	Emphysema and Cystic Lung Disease. , 2023, , 763-779.		0
975	Cystic fibrosis and other ion channel-related diseases. , 2023, , 135-149.		0
1082	Divergent Approaches Toward Drug Discovery and Development. , 2023, , 557-578.		0
1127	Editorial: Real-world experience with CFTR modulator therapy. Frontiers in Pharmacology, 0, 14, .	1.6	0