Christine E Bear

List of Publications by Citations

108 papers	5,054 citations	31 h-index	69 g-index
116 ext. papers	5,663 ext. citations	8.3 avg, IF	5.44 L-index

#	Paper	IF	Citations
108	Purification and functional reconstitution of the cystic fibrosis transmembrane conductance regulator (CFTR). *Cell*, **1992**, 68, 809-18	56.2	865
107	Expression of the cystic fibrosis gene in non-epithelial invertebrate cells produces a regulated anion conductance. *Cell*, **1991**, 64, 681-91	56.2	485
106	Modulation of disease severity in cystic fibrosis transmembrane conductance regulator deficient mice by a secondary genetic factor. *Nature Genetics*, **1996**, 12, 280-7	36.3	343
105	Directed differentiation of human pluripotent stem cells into mature airway epithelia expressing functional CFTR protein. *Nature Biotechnology*, **2012**, 30, 876-82	44.5	292
104	ATPase activity of the cystic fibrosis transmembrane conductance regulator. *Journal of Biological Chemistry*, **1996**, 271, 28463-8	5.4	214
103	Directed differentiation of cholangiocytes from human pluripotent stem cells. *Nature Biotechnology*, **2015**, 33, 853-61	44.5	193
102	Cystic fibrosis transmembrane conductance regulator (CFTR) potentiator VX-770 (ivacaftor) opens the defective channel gate of mutant CFTR in a phosphorylation-dependent but ATP-independent manner. *Journal of Biological Chemistry*, **2012**, 287, 36639-49	5.4	192
101	The cystic fibrosis mutation (delta F508) does not influence the chloride channel activity of CFTR. *Nature Genetics*, **1993**, 3, 311-6	36.3	170
100	Purified cystic fibrosis transmembrane conductance regulator (CFTR) does not function as an ATP channel. *Journal of Biological Chemistry*, **1996**, 271, 11623-6	5.4	97
99	Orkambi and amplifier co-therapy improves function from a rare mutation in gene-edited cells and patient tissue. *EMBO Molecular Medicine*, **2017**, 9, 1224-1243	12	76
98	Acellular lung scaffolds direct differentiation of endoderm to functional airway epithelial cells: requirement of matrix-bound HS proteoglycans. *Stem Cell Reports*, **2015**, 4, 419-30	8	73
97	VX-809 and related corrector compounds exhibit secondary activity stabilizing active F508del-CFTR after its partial rescue to the cell surface. *Chemistry and Biology*, **2014**, 21, 666-78		69
96	Structural basis for alginate secretion across the bacterial outer membrane. *Proceedings of the National Academy of Sciences of the United States of America*, **2011**, 108, 13083-8	11.5	65
95	Cystic fibrosis gene modifier SLC26A9 modulates airway response to CFTR-directed therapeutics. *Human Molecular Genetics*, **2016**, 25, 4590-4600	5.6	62
94	A small-molecule modulator interacts directly with deltaPhe508-CFTR to modify its ATPase activity and conformational stability. *Molecular Pharmacology*, **2009**, 75, 1430-8	4.3	61
93	Determination of CFTR chloride channel activity and pharmacology using radiotracer flux methods. *Journal of Cystic Fibrosis*, **2004**, 3 Suppl 2, 119-21	4.1	59
92	Efficient generation of functional CFTR-expressing airway epithelial cells from human pluripotent stem cells. *Nature Protocols*, **2015**, 10, 363-81	18.8	55

91	A novel procedure for the efficient purification of the cystic fibrosis transmembrane conductance regulator (CFTR). *Biochemical Journal*, **1997**, 327 (Pt 1), 17-21	3.8	52
90	A chemical corrector modifies the channel function of F508del-CFTR. *Molecular Pharmacology*, **2010**, 78, 411-8	4.3	51
89	Phenotypic profiling of CFTR modulators in patient-derived respiratory epithelia. *Npj Genomic Medicine*, **2017**, 2, 12	6.2	46
88	Phosphorylation-activated chloride channels in human skin fibroblasts. *FEBS Letters*, **1988**, 237, 145-9	3.8	44
87	Transducing Airway Basal Cells with a Helper-Dependent Adenoviral Vector for Lung Gene Therapy. *Human Gene Therapy*, **2018**, 29, 643-652	4.8	39
86	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,	13.6	39
85	A heteromeric complex of the two nucleotide binding domains of cystic fibrosis transmembrane conductance regulator (CFTR) mediates ATPase activity. *Journal of Biological Chemistry*, **2004**, 279, 41664-9	5.4	38
84	Functional rescue of c.3846G>A (W1282X) in patient-derived nasal cultures achieved by inhibition of nonsense mediated decay and protein modulators with complementary mechanisms of action. *Journal of Cystic Fibrosis*, **2020**, 19, 717-727	4.1	37
83	The Walker B motif of the second nucleotide-binding domain (NBD2) of CFTR plays a key role in ATPase activity by the NBD1-NBD2 heterodimer. *Biochemical Journal*, **2007**, 401, 581-6	3.8	36
82	Functional Rescue of F508del-CFTR Using Small Molecule Correctors. *Frontiers in Pharmacology*, **2012**, 3, 160	5.6	35
81	Proton-dependent gating and proton uptake by Wzx support O-antigen-subunit antiport across the bacterial inner membrane. *MBio*, **2013**, 4, e00678-13	7.8	34
80	Correctors of the Major Cystic Fibrosis Mutant Interact through Membrane-Spanning Domains. *Molecular Pharmacology*, **2018**, 93, 612-618	4.3	33
79	Molecular basis for the ATPase activity of CFTR. *Archives of Biochemistry and Biophysics*, **2008**, 476, 95-100	4.1	32
78	Synergy of cAMP and calcium signaling pathways in CFTR regulation. *Proceedings of the National Academy of Sciences of the United States of America*, **2017**, 114, E2086-E2095	11.5	31
77	Is a Genetic Modifier of Cystic Fibrosis That Regulates Attachment to Human Bronchial Epithelial Cells. *MBio*, **2017**, 8,	7.8	31
76	Comprehensive mapping of cystic fibrosis mutations to CFTR protein identifies mutation clusters and molecular docking predicts corrector binding site. *Proteins: Structure, Function and Bioinformatics*, **2018**, 86, 833-843	4.2	31
75	ORKAMBI-Mediated Rescue of Mucociliary Clearance in Cystic Fibrosis Primary Respiratory Cultures Is Enhanced by Arginine Uptake, Arginase Inhibition, and Promotion of Nitric Oxide Signaling to the Cystic Fibrosis Transmembrane Conductance Regulator Channel. *Molecular Pharmacology*, **2019**, 96, 515-525	4.3	31
74	Insights into the mechanisms underlying CFTR channel activity, the molecular basis for cystic fibrosis and strategies for therapy. *Essays in Biochemistry*, **2011**, 50, 233-48	7.6	31

73	Purification and characterization of recombinant cystic fibrosis transmembrane conductance regulator from Chinese hamster ovary and insect cells. *Journal of Biological Chemistry*, **1995**, 270, 17033-43	5.4	31
72	Facilitating Structure-Function Studies of CFTR Modulator Sites with Efficiencies in Mutagenesis and Functional Screening. *Journal of Biomolecular Screening*, **2015**, 20, 1204-17		30
71	Genetic, cell biological, and clinical interrogation of the CFTR mutation c.3700 A>G (p.Ile1234Val) informs strategies for future medical intervention. *Genetics in Medicine*, **2014**, 16, 625-32	8.1	29
70	Probing conformational rescue induced by a chemical corrector of F508del-cystic fibrosis transmembrane conductance regulator (CFTR) mutant. *Journal of Biological Chemistry*, **2011**, 286, 24714-25	5.4	29
69	Direct interaction of a small-molecule modulator with G551D-CFTR, a cystic fibrosis-causing mutation associated with severe disease. *Biochemical Journal*, **2009**, 418, 185-90	3.8	29
68	Activity of a novel antimicrobial peptide against Pseudomonas aeruginosa biofilms. *Scientific Reports*, **2018**, 8, 14728	4.9	29
67	A Therapy for Most with Cystic Fibrosis. *Cell*, **2020**, 180, 211	56.2	28
66	The intact CFTR protein mediates ATPase rather than adenylate kinase activity. *Biochemical Journal* , **2008**, 412, 315-21	3.8	28
65	Perturbation of the pore of the cystic fibrosis transmembrane conductance regulator (CFTR) inhibits its atpase activity. *Journal of Biological Chemistry*, **2001**, 276, 11575-81	5.4	28
64	The CF Canada-Sick Kids Program in individual CF therapy: A resource for the advancement of personalized medicine in CF. *Journal of Cystic Fibrosis*, **2019**, 18, 35-43	4.1	28
63	A novel method for monitoring the cytosolic delivery of peptide cargo. *Journal of Controlled Release*, **2009**, 137, 2-7	11.7	27
62	Chloride channel activity of ClC-2 is modified by the actin cytoskeleton. *Biochemical Journal*, **2000**, 352, 789-794	3.8	27
61	The investigational Cystic Fibrosis drug Trimethylangelicin directly modulates CFTR by stabilizing the first membrane-spanning domain. *Biochemical Pharmacology*, **2016**, 119, 85-92	6	27
60	Non-CFTR chloride channels likely contribute to secretion in the murine small intestine. *Pflugers Archiv European Journal of Physiology*, **2001**, 443 Suppl 1, S103-6	4.6	26
59	Phosphorylation-induced conformational changes of cystic fibrosis transmembrane conductance regulator monitored by attenuated total reflection-Fourier transform IR spectroscopy and fluorescence spectroscopy. *Journal of Biological Chemistry*, **2004**, 279, 5528-36	5.4	24
58	Molecular Mechanism of Action of Trimethylangelicin Derivatives as CFTR Modulators. *Frontiers in Pharmacology*, **2018**, 9, 719	5.6	22
57	Coupling of ATP hydrolysis with channel gating by purified, reconstituted CFTR. *Journal of Bioenergetics and Biomembranes*, **1997**, 29, 465-73	3.7	22
56	The major cystic fibrosis causing mutation exhibits defective propensity for phosphorylation. *Proteomics*, **2015**, 15, 447-61	4.8	21

55	Anti-Infectives Restore ORKAMBI Rescue of F508del-CFTR Function in Human Bronchial Epithelial Cells Infected with Clinical Strains of. *Biomolecules*, **2020**, 10,	5.9	21
54	Lipophilicity of the Cystic Fibrosis Drug, Ivacaftor (VX-770), and Its Destabilizing Effect on the Major CF-causing Mutation: F508del. *Molecular Pharmacology*, **2018**, 94, 917-925	4.3	21
53	Generation of Induced Progenitor-like Cells from Mature Epithelial Cells Using Interrupted Reprogramming. *Stem Cell Reports*, **2017**, 9, 1780-1795	8	21
52	An essential role for ClC-4 in transferrin receptor function revealed in studies of fibroblasts derived from Clcn4-null mice. *Journal of Cell Science*, **2009**, 122, 1229-37	5.3	21
51	Molecular basis for the chloride channel activity of cystic fibrosis transmembrane conductance regulator and the consequences of disease-causing mutations. *Current Topics in Developmental Biology*, **2004**, 60, 215-49	5.3	21
50	Sphingosine-1-Phosphate Is a Novel Regulator of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Activity. *PLoS ONE*, **2015**, 10, e0130313	3.7	21
49	Emerging preclinical modulators developed for F508del-CFTR have the potential to be effective for ORKAMBI resistant processing mutants. *Journal of Cystic Fibrosis*, **2021**, 20, 106-119	4.1	21
48	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	6.4	19
47	Conformational defects underlie proteasomal degradation of Dent's disease-causing mutants of ClC-5. *Biochemical Journal*, **2013**, 452, 391-400	3.8	19
46	Cell volume changes and the activity of the chloride conductance path. *Annals of the New York Academy of Sciences*, **1989**, 574, 294-308	6.5	19
45	Testing gene therapy vectors in human primary nasal epithelial cultures. *Molecular Therapy - Methods and Clinical Development*, **2015**, 2, 15034	6.4	18
44	Functional rescue of DeltaF508-CFTR by peptides designed to mimic sorting motifs. *Chemistry and Biology*, **2009**, 16, 520-30		18
43	SLC6A14, an amino acid transporter, modifies the primary CF defect in fluid secretion. *ELife*, **2018**, 7,	8.9	17
42	Cholesterol Interaction Directly Enhances Intrinsic Activity of the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR). *Cells*, **2019**, 8,	7.9	17
41	ClC transporters: discoveries and challenges in defining the mechanisms underlying function and regulation of ClC-5. *Pflugers Archiv European Journal of Physiology*, **2010**, 460, 543-57	4.6	14
40	Quaternary structure of the chloride channel ClC-2. *Biochemistry*, **2000**, 39, 13838-47	3.2	14
39	Evidence for a channel for the electrogenic transport of chloride ion in the rat hepatocyte. *Hepatology*, **1985**, 5, 383-91	11.2	14
38	Current insights into the role of PKA phosphorylation in CFTR channel activity and the pharmacological rescue of cystic fibrosis disease-causing mutants. *Cellular and Molecular Life Sciences*, **2017**, 74, 57-66	10.3	13

37	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,	13.6	12
36	Preclinical Studies of a Rare CF-Causing Mutation in the Second Nucleotide Binding Domain (c.3700A>G) Show Robust Functional Rescue in Primary Nasal Cultures by Novel CFTR Modulators. *Journal of Personalized Medicine*, **2020**, 10,	3.6	12
35	Augmentation of Cystic Fibrosis Transmembrane Conductance Regulator Function in Human Bronchial Epithelial Cells via SLC6A14-Dependent Amino Acid Uptake. Implications for Treatment of Cystic Fibrosis. *American Journal of Respiratory Cell and Molecular Biology*, **2019**, 61, 755-764	5.7	11
34	Channel Gating Regulation by the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) First Cytosolic Loop. *Journal of Biological Chemistry*, **2016**, 291, 1854-1865	5.4	11
33	Activity of lumacaftor is not conserved in zebrafish Cftr bearing the major cystic fibrosis-causing mutation. *FASEB BioAdvances*, **2019**, 1, 661-670	2.8	11
32	Lung arginase expression and activity is increased in cystic fibrosis mouse models. *Journal of Applied Physiology*, **2014**, 117, 284-8	3.7	11
31	Purification and reconstitution of epithelial chloride channel cystic fibrosis transmembrane conductance regulator. *Methods in Enzymology*, **1999**, 294, 227-46	1.7	11
30	The CFTR Mutation c.3453G > C (D1152H) Confers an Anion Selectivity Defect in Primary Airway Tissue that Can Be Rescued by Ivacaftor. *Journal of Personalized Medicine*, **2020**, 10,	3.6	10
29	Studies of the molecular basis for cystic fibrosis using purified reconstituted CFTR protein. *Methods in Molecular Medicine*, **2002**, 70, 143-57		10
28	Synthesis and properties of molecular probes for the rescue site on mutant cystic fibrosis transmembrane conductance regulator. *Journal of Medicinal Chemistry*, **2011**, 54, 8693-701	8.3	9
27	Identification and validation of hits from high throughput screens for CFTR modulators. *Current Pharmaceutical Design*, **2012**, 18, 628-41	3.3	9
26	Techniques for studying biliary secretion: electrolytes in bile. *Hepatology*, **1984**, 4, 25S-30S	11.2	7
25	Attenuation of Phosphorylation-dependent Activation of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) by Disease-causing Mutations at the Transmission Interface. *Journal of Biological Chemistry*, **2017**, 292, 1988-1999	5.4	6
24	Synthesis and characterization of a photoaffinity labelling probe based on the structure of the cystic fibrosis drug ivacaftor. *Tetrahedron*, **2018**, 74, 5528-5538	2.4	6
23	Phenotyping Rare CFTR Mutations Reveal Functional Expression Defects Restored by TRIKAFTA. *Journal of Personalized Medicine*, **2021**, 11,	3.6	6
22	Conversion of human and mouse fibroblasts into lung-like epithelial cells. *Scientific Reports*, **2019**, 9, 9027	4.9	5
21	Effects of mutations in cAMP-dependent protein kinase on chloride efflux in Caco-2 human colonic carcinoma cells. *Journal of Cellular Physiology*, **1995**, 162, 64-73	7	5
20	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	8	5

19	Photochemically Activated Notch Signaling Hydrogel Preferentially Differentiates Human Derived Hepatoblasts to Cholangiocytes. *Advanced Functional Materials*, **2021**, 31, 2006116	15.6	5
18	Antisense oligonucleotide splicing modulation as a novel Cystic Fibrosis therapeutic approach for the W1282X nonsense mutation.. *Journal of Cystic Fibrosis*, **2021**,	4.1	5
17	An organoid model to assay the role of CFTR in the human epididymis epithelium. *Cell and Tissue Research*, **2020**, 381, 327-336	4.2	4
16	Identification of binding sites for ivacaftor on the cystic fibrosis transmembrane conductance regulator. *IScience*, **2021**, 24, 102542	6.1	4
15	Structural effects of extracellular loop mutations in CFTR helical hairpins. *Biochimica Et Biophysica Acta - Biomembranes*, **2018**, 1860, 1092-1098	3.8	3
14	Functional reconstitution and channel activity measurements of purified wildtype and mutant CFTR protein. *Journal of Visualized Experiments*, **2015**,	1.6	3
13	Generation of functional ciliated cholangiocytes from human pluripotent stem cells. *Nature Communications*, **2021**, 12, 6504	17.4	3
12	Finding new drugs to enhance anion secretion in cystic fibrosis: Toward suitable systems for better drug screening. Report on the pre-conference meeting to the 12th ECFS Basic Science Conference, Albufeira, 25-28 March 2015. *Journal of Cystic Fibrosis*, **2015**, 14, 700-5	4.1	2
11	50 years ago in the Journal of Pediatrics: the effect of N-acetylcysteine on the viscosity of tracheobronchial secretions in cystic fibrosis of the pancreas. *Journal of Pediatrics*, **2013**, 162, 85	3.6	2
10	Perspectives on the translation of in-vitro studies to precision medicine in Cystic Fibrosis. *EBioMedicine*, **2021**, 73, 103660	8.8	2
9	One-Step Formation of Protein-Based Tubular Structures for Functional Devices and Tissues. *Advanced Healthcare Materials*, **2021**, 10, e2001746	10.1	2
8	High-throughput functional analysis of CFTR and other apically localized channels in iPSC derived intestinal organoids		2
7	CFTR interactome mapping using the mammalian membrane two-hybrid high-throughput screening system.. *Molecular Systems Biology*, **2022**, 18, e10629	12.2	2
6	Stage-Specific Generation of Human Pluripotent Stem Cell Derived Lung Models to Measure CFTR Function.. *Current Protocols*, **2022**, 2, e341		0
5	Riociguat for the treatment of Phe508del homozygous adults with cystic fibrosis. *Journal of Cystic Fibrosis*, **2021**, 20, 1018-1025	4.1	0
4	A protocol for identifying the binding sites of small molecules on the cystic fibrosis transmembrane conductance regulator (CFTR) protein.. *STAR Protocols*, **2022**, 3, 101258	1.4	0
3	Cover Image, Volume 86, Issue 8. *Proteins: Structure, Function and Bioinformatics*, **2018**, 86, C1-C1	4.2	
2	Characterization of a CFTR construct with a C-terminal tetracysteine sequence and its use in the visualization of trafficking pathways. *FASEB Journal*, **2007**, 21, A243	0.9	

1	Sphingosine-1-Phosphate acutely modulates the CFTR (Cystic Fibrosis Transmembrane Regulator) transporter in an AMPK-dependent manner. *FASEB Journal*, **2010**, 24, 609.1	0.9