Michael J Welsh

List of Publications by Citations

283 papers	37,632 citations	100 h-index	190 g-index
298 ext. papers	40,893 ext. citations	13.4 avg, IF	6.94 L-index

#	Paper	IF	Citations
283	Quorum-sensing signals indicate that cystic fibrosis lungs are infected with bacterial biofilms. *Nature*, **2000**, 407, 762-4	50.4	1230
282	Molecular mechanisms of CFTR chloride channel dysfunction in cystic fibrosis. *Cell*, **1993**, 73, 1251-4	56.2	1193
281	Cellular and molecular barriers to gene transfer by a cationic lipid. *Journal of Biological Chemistry*, **1995**, 270, 18997-9007	5.4	1126
280	Processing of mutant cystic fibrosis transmembrane conductance regulator is temperature-sensitive. *Nature*, **1992**, 358, 761-4	50.4	1081
279	Cystic fibrosis airway epithelia fail to kill bacteria because of abnormal airway surface fluid. *Cell*, **1996**, 85, 229-36	56.2	848
278	Neuroprotection in ischemia: blocking calcium-permeable acid-sensing ion channels. *Cell*, **2004**, 118, 687-98	56.2	816
277	A component of innate immunity prevents bacterial biofilm development. *Nature*, **2002**, 417, 552-5	50.4	815
276	Structure and function of the CFTR chloride channel. *Physiological Reviews*, **1999**, 79, S23-45	47.9	748
275	Adenovirus-mediated gene transfer transiently corrects the chloride transport defect in nasal epithelia of patients with cystic fibrosis. *Cell*, **1993**, 75, 207-16	56.2	662
274	Disruption of the CFTR gene produces a model of cystic fibrosis in newborn pigs. *Science*, **2008**, 321, 1837-41	33.3	595
273	Expression of cystic fibrosis transmembrane conductance regulator corrects defective chloride channel regulation in cystic fibrosis airway epithelial cells. *Nature*, **1990**, 347, 358-63	50.4	579
272	Phosphorylation of the R domain by cAMP-dependent protein kinase regulates the CFTR chloride channel. *Cell*, **1991**, 66, 1027-36	56.2	575
271	Reduced airway surface pH impairs bacterial killing in the porcine cystic fibrosis lung. *Nature*, **2012**, 487, 109-13	50.4	569
270	The acid-activated ion channel ASIC contributes to synaptic plasticity, learning, and memory. *Neuron*, **2002**, 34, 463-77	13.9	541
269	The DRASIC cation channel contributes to the detection of cutaneous touch and acid stimuli in mice. *Neuron*, **2001**, 32, 1071-83	13.9	512
268	Motile cilia of human airway epithelia are chemosensory. *Science*, **2009**, 325, 1131-4	33.3	504
267	Nucleoside triphosphates are required to open the CFTR chloride channel. *Cell*, **1991**, 67, 775-84	56.2	482

266	Acid-sensing ion channels: advances, questions and therapeutic opportunities. *Trends in Neurosciences*, **2006**, 29, 578-86	13.3	459
265	Discovery of five conserved beta -defensin gene clusters using a computational search strategy. *Proceedings of the National Academy of Sciences of the United States of America*, **2002**, 99, 2129-33	11.5	423
264	The mammalian sodium channel BNC1 is required for normal touch sensation. *Nature*, **2000**, 407, 1007-11	50.4	414
263	Mutations in CFTR associated with mild-disease-form Cl- channels with altered pore properties. *Nature*, **1993**, 362, 160-4	50.4	403
262	Cyclic AMP-dependent protein kinase opens chloride channels in normal but not cystic fibrosis airway epithelium. *Nature*, **1988**, 331, 358-60	50.4	392
261	Mechanism by which Liddle's syndrome mutations increase activity of a human epithelial Na+ channel. *Cell*, **1995**, 83, 969-78	56.2	375
260	Origins of cystic fibrosis lung disease. *New England Journal of Medicine*, **2015**, 372, 351-62	59.2	368
259	Chronic hyperalgesia induced by repeated acid injections in muscle is abolished by the loss of ASIC3, but not ASIC1. *Pain*, **2003**, 106, 229-239	8	358
258	Cystic fibrosis pigs develop lung disease and exhibit defective bacterial eradication at birth. *Science Translational Medicine*, **2010**, 2, 29ra31	17.5	356
257	Acid-sensing ion channel 1 is localized in brain regions with high synaptic density and contributes to fear conditioning. *Journal of Neuroscience*, **2003**, 23, 5496-502	6.6	355
256	Chloride and potassium channels in cystic fibrosis airway epithelia. *Nature*, **1986**, 322, 467-70	50.4	350
255	Acid-sensing ion channel-1 contributes to axonal degeneration in autoimmune inflammation of the central nervous system. *Nature Medicine*, **2007**, 13, 1483-9	50.5	316
254	Expression and characterization of the cystic fibrosis transmembrane conductance regulator. *Nature*, **1990**, 347, 382-6	50.4	314
253	Extracellular acidosis increases neuronal cell calcium by activating acid-sensing ion channel 1a. *Proceedings of the National Academy of Sciences of the United States of America*, **2004**, 101, 6752-7	11.5	310
252	Segregation of receptor and ligand regulates activation of epithelial growth factor receptor. *Nature*, **2003**, 422, 322-6	50.4	310
251	Heteromultimers of DEG/ENaC subunits form H+-gated channels in mouse sensory neurons. *Proceedings of the National Academy of Sciences of the United States of America*, **2002**, 99, 2338-43	11.5	308
250	Bactericidal activity of mammalian cathelicidin-derived peptides. *Infection and Immunity*, **2000**, 68, 2748-55	3.7	308
249	Adenovirus fiber disrupts CAR-mediated intercellular adhesion allowing virus escape. *Cell*, **2002**, 110, 789-99	56.2	301

248	The amygdala is a chemosensor that detects carbon dioxide and acidosis to elicit fear behavior. *Cell* , **2009**, 139, 1012-21	56.2	296
247	Basolateral localization of fiber receptors limits adenovirus infection from the apical surface of airway epithelia. *Journal of Biological Chemistry*, **1999**, 274, 10219-26	5.4	287
246	Seizure termination by acidosis depends on ASIC1a. *Nature Neuroscience*, **2008**, 11, 816-22	25.5	267
245	Impaired mucus detachment disrupts mucociliary transport in a piglet model of cystic fibrosis. *Science*, **2014**, 345, 818-22	33.3	263
244	Cloning and expression of a novel human brain Na+ channel. *Journal of Biological Chemistry*, **1996**, 271, 7879-82	5.4	263
243	An in vitro model of differentiated human airway epithelia. Methods for establishing primary cultures. *Methods in Molecular Biology*, **2002**, 188, 115-37	1.4	260
242	Binding of adeno-associated virus type 5 to 2,3-linked sialic acid is required for gene transfer. *Journal of Biological Chemistry*, **2001**, 276, 20610-6	5.4	257
241	Production of CFTR-null and CFTR-DeltaF508 heterozygous pigs by adeno-associated virus-mediated gene targeting and somatic cell nuclear transfer. *Journal of Clinical Investigation*, **2008**, 118, 1571-7	15.9	250
240	Synergistic and additive killing by antimicrobial factors found in human airway surface liquid. *American Journal of Physiology - Lung Cellular and Molecular Physiology*, **2000**, 279, L799-805	5.8	250
239	Inactivation of a Pseudomonas aeruginosa quorum-sensing signal by human airway epithelia. *Proceedings of the National Academy of Sciences of the United States of America*, **2004**, 101, 3587-90	11.5	241
238	A mouse model for the delta F508 allele of cystic fibrosis. *Journal of Clinical Investigation*, **1995**, 96, 2051-64	15.9	235
237	Complexes of adenovirus with polycationic polymers and cationic lipids increase the efficiency of gene transfer in vitro and in vivo. *Journal of Biological Chemistry*, **1997**, 272, 6479-89	5.4	217
236	Ripped pocket and pickpocket, novel Drosophila DEG/ENaC subunits expressed in early development and in mechanosensory neurons. *Journal of Cell Biology*, **1998**, 140, 143-52	7.3	216
235	The two nucleotide-binding domains of cystic fibrosis transmembrane conductance regulator (CFTR) have distinct functions in controlling channel activity. *Journal of Biological Chemistry*, **1995**, 270, 1711-7	5.4	214
234	Fear and panic in humans with bilateral amygdala damage. *Nature Neuroscience*, **2013**, 16, 270-2	25.5	213
233	Abnormal regulation of ion channels in cystic fibrosis epithelia. *FASEB Journal*, **1990**, 4, 2718-25	0.9	211
232	Repeat administration of an adenovirus vector encoding cystic fibrosis transmembrane conductance regulator to the nasal epithelium of patients with cystic fibrosis. *Journal of Clinical Investigation*, **1996**, 97, 1504-11	15.9	211
231	Cystic fibrosis transmembrane conductance regulator: a chloride channel with novel regulation. *Neuron*, **1992**, 8, 821-9	13.9	209

230	Safety and efficacy of repetitive adenovirus-mediated transfer of CFTR cDNA to airway epithelia of primates and cotton rats. *Nature Genetics*, **1994**, 6, 75-83	36.3	202
229	Loss of CFTR chloride channels alters salt absorption by cystic fibrosis airway epithelia in vitro. *Molecular Cell*, **1998**, 2, 397-403	17.6	200
228	Airway acidification initiates host defense abnormalities in cystic fibrosis mice. *Science*, **2016**, 351, 503-7	33.3	197
227	Electrophysiological and biochemical evidence that DEG/ENaC cation channels are composed of nine subunits. *Journal of Biological Chemistry*, **1998**, 273, 681-4	5.4	195
226	Neuropeptide FF and FMRFamide potentiate acid-evoked currents from sensory neurons and proton-gated DEG/ENaC channels. *Neuron*, **2000**, 26, 133-41	13.9	194
225	Loss of anion transport without increased sodium absorption characterizes newborn porcine cystic fibrosis airway epithelia. *Cell*, **2010**, 143, 911-23	56.2	193
224	Restoring Cystic Fibrosis Transmembrane Conductance Regulator Function Reduces Airway Bacteria and Inflammation in People with Cystic Fibrosis and Chronic Lung Infections. *American Journal of Respiratory and Critical Care Medicine*, **2017**, 195, 1617-1628	10.2	191
223	A molecular component of the arterial baroreceptor mechanotransducer. *Neuron*, **1998**, 21, 1435-41	13.9	184
222	Acid-sensing ion channel 2 (ASIC2) modulates ASIC1 H+-activated currents in hippocampal neurons. *Journal of Biological Chemistry*, **2004**, 279, 18296-305	5.4	182
221	Lack of high affinity fiber receptor activity explains the resistance of ciliated airway epithelia to adenovirus infection. *Journal of Clinical Investigation*, **1997**, 100, 1144-9	15.9	178
220	The porcine lung as a potential model for cystic fibrosis. *American Journal of Physiology - Lung Cellular and Molecular Physiology*, **2008**, 295, L240-63	5.8	176
219	Activity of abundant antimicrobials of the human airway. *American Journal of Respiratory Cell and Molecular Biology*, **1999**, 20, 872-9	5.7	176
218	Overexpression of acid-sensing ion channel 1a in transgenic mice increases acquired fear-related behavior. *Proceedings of the National Academy of Sciences of the United States of America*, **2004**, 101, 3621-6	11.5	163
217	Identification of revertants for the cystic fibrosis delta F508 mutation using STE6-CFTR chimeras in yeast. *Cell*, **1993**, 73, 335-46	56.2	162
216	Acidic pH increases airway surface liquid viscosity in cystic fibrosis. *Journal of Clinical Investigation*, **2016**, 126, 879-91	15.9	160
215	Enhanced locomotion caused by loss of the Drosophila DEG/ENaC protein Pickpocket1. *Current Biology*, **2003**, 13, 1557-63	6.3	159
214	Protons are a neurotransmitter that regulates synaptic plasticity in the lateral amygdala. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 8961-6	11.5	154
213	The Δ508 mutation causes CFTR misprocessing and cystic fibrosis-like disease in pigs. *Science Translational Medicine*, **2011**, 3, 74ra24	17.5	153

212	Development and analysis of recombinant adenoviruses for gene therapy of cystic fibrosis. *Human Gene Therapy*, **1993**, 4, 461-76	4.8	151
211	Identification and function of thermosensory neurons in Drosophila larvae. *Nature Neuroscience*, **2003**, 6, 267-73	25.5	150
210	The ion channel ASIC2 is required for baroreceptor and autonomic control of the circulation. *Neuron*, **2009**, 64, 885-97	13.9	149
209	Acid-sensing ion channel 1a is a postsynaptic proton receptor that affects the density of dendritic spines. *Proceedings of the National Academy of Sciences of the United States of America*, **2006**, 103, 16556-61	11.5	149
208	Loss of cystic fibrosis transmembrane conductance regulator function produces abnormalities in tracheal development in neonatal pigs and young children. *American Journal of Respiratory and Critical Care Medicine*, **2010**, 182, 1251-61	10.2	147
207	Chloride secretion by canine tracheal epithelium: I. Role of intracellular c AMP levels. *Journal of Membrane Biology*, **1982**, 70, 217-26	2.3	142
206	A low rate of cell proliferation and reduced DNA uptake limit cationic lipid-mediated gene transfer to primary cultures of ciliated human airway epithelia. *Gene Therapy*, **1997**, 4, 1173-80	4	141
205	Drosophila hygrosensation requires the TRP channels water witch and nanchung. *Nature*, **2007**, 450, 294-8	50.4	138
204	Contribution of Drosophila DEG/ENaC genes to salt taste. *Neuron*, **2003**, 39, 133-46	13.9	138
203	Chloride secretion by canine tracheal epithelium: II. The cellular electrical potential profile. *Journal of Membrane Biology*, **1982**, 70, 227-38	2.3	138
202	pH modulates the activity and synergism of the airway surface liquid antimicrobials β-defensin-3 and LL-37. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 18703-8	11.5	132
201	Cystic fibrosis. *Scientific American*, **1995**, 273, 52-9	0.5	132
200	Comparison of DNA-lipid complexes and DNA alone for gene transfer to cystic fibrosis airway epithelia in vivo. *Journal of Clinical Investigation*, **1997**, 100, 1529-37	15.9	132
199	Stimulation of CFTR activity by its phosphorylated R domain. *Nature*, **1997**, 389, 294-6	50.4	131
198	Acid-sensing ion channels contribute to synaptic transmission and inhibit cocaine-evoked plasticity. *Nature Neuroscience*, **2014**, 17, 1083-91	25.5	128
197	The ion channel ASIC1 contributes to visceral but not cutaneous mechanoreceptor function. *Gastroenterology*, **2004**, 127, 1739-47	13.3	123
196	Breathing Inhibited When Seizures Spread to the Amygdala and upon Amygdala Stimulation. *Journal of Neuroscience*, **2015**, 35, 10281-9	6.6	121
195	Incorporation of adenovirus in calcium phosphate precipitates enhances gene transfer to airway epithelia in vitro and in vivo. *Journal of Clinical Investigation*, **1998**, 102, 184-93	15.9	120

194	Antimicrobial peptides and proteins in the innate defense of the airway surface. *Current Opinion in Immunology*, **2001**, 13, 89-95	7.8	118
193	Adeno-associated virus-targeted disruption of the CFTR gene in cloned ferrets. *Journal of Clinical Investigation*, **2008**, 118, 1578-83	15.9	117
192	Detecting activity-evoked pH changes in human brain. *Proceedings of the National Academy of Sciences of the United States of America*, **2012**, 109, 8270-3	11.5	116
191	Interactions between subunits of the human epithelial sodium channel. *Journal of Biological Chemistry*, **1997**, 272, 27295-300	5.4	113
190	Pathology of gastrointestinal organs in a porcine model of cystic fibrosis. *American Journal of Pathology*, **2010**, 176, 1377-89	5.8	112
189	The amino-terminal portion of CFTR forms a regulated Cl- channel. *Cell*, **1994**, 76, 1091-8	56.2	108
188	TRPA channels distinguish gravity sensing from hearing in Johnston's organ. *Proceedings of the National Academy of Sciences of the United States of America*, **2009**, 106, 13606-11	11.5	107
187	Inhibition of chloride secretion by furosemide in canine tracheal epithelium. *Journal of Membrane Biology*, **1983**, 71, 219-26	2.3	107
186	A severe phenotype in mice with a duplication of exon 3 in the cystic fibrosis locus. *Human Molecular Genetics*, **1993**, 2, 1561-9	5.6	106
185	Processing and function of CFTR-DeltaF508 are species-dependent. *Proceedings of the National Academy of Sciences of the United States of America*, **2007**, 104, 15370-5	11.5	101
184	Cystic fibrosis transmembrane conductance regulator splice variants are not conserved and fail to produce chloride channels. *Nature Genetics*, **1993**, 4, 426-31	36.3	101
183	A microRNA network regulates expression and biosynthesis of wild-type and DeltaF508 mutant cystic fibrosis transmembrane conductance regulator. *Proceedings of the National Academy of Sciences of the United States of America*, **2012**, 109, 13362-7	11.5	100
182	Chloride secretion by canine tracheal epithelium: III. Membrane resistances and electromotive forces. *Journal of Membrane Biology*, **1983**, 71, 209-18	2.3	99
181	Gel-forming mucins form distinct morphologic structures in airways. *Proceedings of the National Academy of Sciences of the United States of America*, **2017**, 114, 6842-6847	11.5	98
180	ppk23-Dependent chemosensory functions contribute to courtship behavior in Drosophila melanogaster. *PLoS Genetics*, **2012**, 8, e1002587	6	98
179	Biochemical basis of touch perception: mechanosensory function of degenerin/epithelial Na+ channels. *Journal of Biological Chemistry*, **2002**, 277, 2369-72	5.4	98
178	Gene transfer of CFTR to airway epithelia: low levels of expression are sufficient to correct Cl- transport and overexpression can generate basolateral CFTR. *American Journal of Physiology - Lung Cellular and Molecular Physiology*, **2005**, 289, L1123-30	5.8	97
177	Number of subunits comprising the epithelial sodium channel. *Journal of Biological Chemistry*, **1999**, 274, 27281-6	5.4	97

176	Intestinal CFTR expression alleviates meconium ileus in cystic fibrosis pigs. *Journal of Clinical Investigation*, **2013**, 123, 2685-93	15.9	96
175	Human cystic fibrosis airway epithelia have reduced Cl- conductance but not increased Na+ conductance. *Proceedings of the National Academy of Sciences of the United States of America*, **2011**, 108, 10260-5	11.5	95
174	Origins of cystic fibrosis lung disease. *New England Journal of Medicine*, **2015**, 372, 1574-5	59.2	94
173	Cystic fibrosis-associated mutations at arginine 347 alter the pore architecture of CFTR. Evidence for disruption of a salt bridge. *Journal of Biological Chemistry*, **1999**, 274, 5429-35	5.4	94
172	The osmolyte xylitol reduces the salt concentration of airway surface liquid and may enhance bacterial killing. *Proceedings of the National Academy of Sciences of the United States of America*, **2000**, 97, 11614-9	11.5	93
171	Regulation of the cystic fibrosis transmembrane conductance regulator Cl- channel by its R domain. *Journal of Biological Chemistry*, **2001**, 276, 7689-92	5.4	90
170	Chemosensory functions for pulmonary neuroendocrine cells. *American Journal of Respiratory Cell and Molecular Biology*, **2014**, 50, 637-46	5.7	89
169	Interaction of the synaptic protein PICK1 (protein interacting with C kinase 1) with the non-voltage gated sodium channels BNC1 (brain Na+ channel 1) and ASIC (acid-sensing ion channel). *Biochemical Journal*, **2002**, 361, 443-450	3.8	89
168	Correction of cAMP-stimulated fluid secretion in cystic fibrosis airway epithelia: efficiency of adenovirus-mediated gene transfer in vitro. *Human Gene Therapy*, **1994**, 5, 585-93	4.8	89
167	Loss of Bardet-Biedl syndrome proteins alters the morphology and function of motile cilia in airway epithelia. *Proceedings of the National Academy of Sciences of the United States of America*, **2008**, 105, 3380-5	11.5	88
166	Normal sweat chloride values do not exclude the diagnosis of cystic fibrosis. *American Journal of Respiratory and Critical Care Medicine*, **1995**, 151, 899-903	10.2	86
165	Pigs and humans with cystic fibrosis have reduced insulin-like growth factor 1 (IGF1) levels at birth. *Proceedings of the National Academy of Sciences of the United States of America*, **2010**, 107, 20571-5	11.5	85
164	Adenovirus-mediated gene transfer in vivo to cerebral blood vessels and perivascular tissue. *Circulation Research*, **1995**, 77, 7-13	15.7	84
163	Effect of cystic fibrosis-associated mutations in the fourth intracellular loop of cystic fibrosis transmembrane conductance regulator. *Journal of Biological Chemistry*, **1996**, 271, 21279-84	5.4	81
162	Drosophila DEG/ENaC pickpocket genes are expressed in the tracheal system, where they may be involved in liquid clearance. *Proceedings of the National Academy of Sciences of the United States of America*, **2003**, 100, 2128-33	11.5	80
161	Normal gating of CFTR requires ATP binding to both nucleotide-binding domains and hydrolysis at the second nucleotide-binding domain. *Proceedings of the National Academy of Sciences of the United States of America*, **2005**, 102, 455-60	11.5	80
160	Changes in either cytosolic or nucleoplasmic inositol 1,4,5-trisphosphate levels can control nuclear Ca2+ concentration. *Journal of Biological Chemistry*, **1995**, 270, 4959-62	5.4	80
159	ASIC2 subunits target acid-sensing ion channels to the synapse via an association with PSD-95. *Journal of Neuroscience*, **2009**, 29, 8438-46	6.6	79

158	DRASIC contributes to pH-gated currents in large dorsal root ganglion sensory neurons by forming heteromultimeric channels. *Journal of Neurophysiology*, **2002**, 87, 2835-43	3.2	79
157	Adenoviral-mediated gene transfer to fetal pulmonary epithelia in vitro and in vivo. *Journal of Clinical Investigation*, **1995**, 95, 2620-32	15.9	79
156	Post-transcriptional regulation of cystic fibrosis transmembrane conductance regulator expression and function by microRNAs. *American Journal of Respiratory Cell and Molecular Biology*, **2013**, 49, 544-51	5.7	78
155	Vaccinia Virus Entry, Exit, and Interaction with Differentiated Human Airway Epithelia. *Journal of Virology*, **2007**, 81, 13278-13278	6.6	78
154	ENaC subunits are molecular components of the arterial baroreceptor complex. *Annals of the New York Academy of Sciences*, **2001**, 940, 42-7	6.5	78
153	Lysozyme secretion by submucosal glands protects the airway from bacterial infection. *American Journal of Respiratory Cell and Molecular Biology*, **2005**, 32, 548-52	5.7	78
152	Depression of ventilation by dopamine in man. Evidence for an effect on the chemoreceptor reflex. *Journal of Clinical Investigation*, **1978**, 61, 708-13	15.9	78
151	Interaction of the synaptic protein PICK1 (protein interacting with C kinase 1) with the non-voltage gated sodium channels BNC1 (brain Na+ channel 1) and ASIC (acid-sensing ion channel). *Biochemical Journal*, **2002**, 361, 443-50	3.8	77
150	cAMP-dependent protein kinase phosphorylation of the acid-sensing ion channel-1 regulates its binding to the protein interacting with C-kinase-1. *Proceedings of the National Academy of Sciences of the United States of America*, **2003**, 100, 2029-34	11.5	75
149	Curcumin stimulates cystic fibrosis transmembrane conductance regulator Cl- channel activity. *Journal of Biological Chemistry*, **2005**, 280, 5221-6	5.4	74
148	ASIC3 and ASIC1 mediate FMRFamide-related peptide enhancement of H+-gated currents in cultured dorsal root ganglion neurons. *Journal of Neurophysiology*, **2003**, 89, 2459-65	3.2	74
147	A shortened adeno-associated virus expression cassette for CFTR gene transfer to cystic fibrosis airway epithelia. *Proceedings of the National Academy of Sciences of the United States of America*, **2005**, 102, 2952-7	11.5	72
146	Engineering novel cell surface receptors for virus-mediated gene transfer. *Journal of Biological Chemistry*, **1999**, 274, 21878-84	5.4	72
145	The calcium signal and neutrophil activation. *Clinical Biochemistry*, **1990**, 23, 159-66	3.5	65
144	Protons activate brain Na+ channel 1 by inducing a conformational change that exposes a residue associated with neurodegeneration. *Journal of Biological Chemistry*, **1998**, 273, 30204-7	5.4	64
143	Group D adenoviruses infect primary central nervous system cells more efficiently than those from group C. *Journal of Virology*, **1999**, 73, 2537-40	6.6	63
142	Enhancing glycolysis attenuates Parkinson's disease progression in models and clinical databases. *Journal of Clinical Investigation*, **2019**, 129, 4539-4549	15.9	63
141	Oxidant regulated inter-subunit disulfide bond formation between ASIC1a subunits. *Proceedings of the National Academy of Sciences of the United States of America*, **2009**, 106, 3573-8	11.5	61

140	Association of domains within the cystic fibrosis transmembrane conductance regulator. *Biochemistry*, **1997**, 36, 1287-94	3.2	61
139	Antibacterial properties of the CFTR potentiator ivacaftor. *Journal of Cystic Fibrosis*, **2014**, 13, 515-9	4.1	60
138	Requirements for ion and solute transport, and pH regulation during enamel maturation. *Journal of Cellular Physiology*, **2012**, 227, 1776-85	7	60
137	Block by MOPS reveals a conformation change in the CFTR pore produced by ATP hydrolysis. *American Journal of Physiology - Cell Physiology*, **1997**, 273, C1278-89	5.4	60
136	Single apical membrane anion channels in primary cultures of canine tracheal epithelium. *Pflugers Archiv European Journal of Physiology*, **1986**, 407 Suppl 2, S116-22	4.6	60
135	Lentiviral-mediated phenotypic correction of cystic fibrosis pigs. *JCI Insight*, **2016**, 1,	9.9	56
134	Neonates with cystic fibrosis have a reduced nasal liquid pH; a small pilot study. *Journal of Cystic Fibrosis*, **2014**, 13, 373-7	4.1	55
133	Effects of C-terminal deletions on cystic fibrosis transmembrane conductance regulator function in cystic fibrosis airway epithelia. *Proceedings of the National Academy of Sciences of the United States of America*, **2003**, 100, 1937-42	11.5	55
132	Development of a porcine model of cystic fibrosis. *Transactions of the American Clinical and Climatological Association*, **2009**, 120, 149-62	0.9	55
131	Concentration of the antibacterial precursor thiocyanate in cystic fibrosis airway secretions. *Free Radical Biology and Medicine*, **2011**, 50, 1144-50	7.8	54
130	Ion transport by primary cultures of canine tracheal epithelium: methodology, morphology, and electrophysiology. *Journal of Membrane Biology*, **1985**, 88, 149-63	2.3	54
129	Sinus hypoplasia precedes sinus infection in a porcine model of cystic fibrosis. *Laryngoscope*, **2012**, 122, 1898-905	3.6	53
128	gene transfer with AAV improves early cystic fibrosis pig phenotypes. *JCI Insight*, **2016**, 1, e88728	9.9	53
127	Simultaneous disruption of mouse ASIC1a, ASIC2 and ASIC3 genes enhances cutaneous mechanosensitivity. *PLoS ONE*, **2012**, 7, e35225	3.7	52
126	Pyrophosphate stimulates wild-type and mutant cystic fibrosis transmembrane conductance regulator Cl- channels. *Journal of Biological Chemistry*, **1995**, 270, 20466-72	5.4	52
125	Acute regulation of tight junction ion selectivity in human airway epithelia. *Proceedings of the National Academy of Sciences of the United States of America*, **2009**, 106, 3591-6	11.5	51
124	Cystic fibrosis gene therapy using an adenovirus vector: in vivo safety and efficacy in nasal epithelium. *Human Gene Therapy*, **1994**, 5, 209-19	4.8	51
123	Anthracene-9-carboxylic acid inhibits an apical membrane chloride conductance in canine tracheal epithelium. *Journal of Membrane Biology*, **1984**, 78, 61-71	2.3	51

122	Targeting the urokinase plasminogen activator receptor enhances gene transfer to human airway epithelia. *Journal of Clinical Investigation*, **2000**, 105, 589-96	15.9	51
121	Glycaemic regulation and insulin secretion are abnormal in cystic fibrosis pigs despite sparing of islet cell mass. *Clinical Science*, **2015**, 128, 131-42	6.5	50
120	Assessing mucociliary transport of single particles in vivo shows variable speed and preference for the ventral trachea in newborn pigs. *Proceedings of the National Academy of Sciences of the United States of America*, **2014**, 111, 2355-60	11.5	50
119	Covalent modification of the regulatory domain irreversibly stimulates cystic fibrosis transmembrane conductance regulator. *Journal of Biological Chemistry*, **1997**, 272, 25617-22	5.4	49
118	PSD-95 and Lin-7b interact with acid-sensing ion channel-3 and have opposite effects on H+- gated current. *Journal of Biological Chemistry*, **2004**, 279, 46962-8	5.4	49
117	Cell surface expression and biosynthesis of epithelial Na+ channels. *Biochemical Journal*, **1998**, 336 (Pt 3), 705-10	3.8	49
116	Contribution of proline residues in the membrane-spanning domains of cystic fibrosis transmembrane conductance regulator to chloride channel function. *Journal of Biological Chemistry* , **1996**, 271, 14995-5001	5.4	48
115	Propagation of infectious human papillomavirus type 16 by using an adenovirus and Cre/LoxP mechanism. *Proceedings of the National Academy of Sciences of the United States of America*, **2004**, 101, 2094-9	11.5	47
114	Cystic fibrosis carriers are at increased risk for a wide range of cystic fibrosis-related conditions. *Proceedings of the National Academy of Sciences of the United States of America*, **2020**, 117, 1621-1627	11.5	47
113	Relationships among CFTR expression, HCO3- secretion, and host defense may inform gene- and cell-based cystic fibrosis therapies. *Proceedings of the National Academy of Sciences of the United States of America*, **2016**, 113, 5382-7	11.5	47
112	Effect of subunit composition and Liddle's syndrome mutations on biosynthesis of ENaC. *American Journal of Physiology - Cell Physiology*, **1999**, 276, C1346-51	5.4	46
111	Small-molecule ion channels increase host defences in cystic fibrosis airway epithelia. *Nature*, **2019**, 567, 405-408	50.4	45
110	Apical localization of the coxsackie-adenovirus receptor by glycosyl-phosphatidylinositol modification is sufficient for adenovirus-mediated gene transfer through the apical surface of human airway epithelia. *Journal of Virology*, **2001**, 75, 7703-11	6.6	45
109	Paradoxical stimulation of a DEG/ENaC channel by amiloride. *Journal of Biological Chemistry*, **1999**, 274, 15500-4	5.4	45
108	An intrinsic adenylate kinase activity regulates gating of the ABC transporter CFTR. *Cell*, **2003**, 115, 837-50	50.2	44
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