

# Seng H Cheng

## List of Publications by Year in descending order

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81  
papers

12,065  
citations

46918

47  
h-index

66788

78  
g-index

81  
all docs

81  
docs citations

81  
times ranked

10542  
citing authors

#	ARTICLE	IF	CITATIONS
1	Defective intracellular transport and processing of CFTR is the molecular basis of most cystic fibrosis. <i>Cell</i> , 1990, 63, 827-834.	13.5	1,697
2	Phosphorylation of the R domain by cAMP-dependent protein kinase regulates the CFTR chloride channel. <i>Cell</i> , 1991, 66, 1027-1036.	13.5	651
3	Expression of cystic fibrosis transmembrane conductance regulator corrects defective chloride channel regulation in cystic fibrosis airway epithelial cells. <i>Nature</i> , 1990, 347, 358-363.	13.7	649
4	Sustained Therapeutic Reversal of Huntington's Disease by Transient Repression of Huntingtin Synthesis. <i>Neuron</i> , 2012, 74, 1031-1044.	3.8	635
5	Detailed Analysis of Structures and Formulations of Cationic Lipids for Efficient Gene Transfer to the Lung. <i>Human Gene Therapy</i> , 1996, 7, 1701-1717.	1.4	453
6	Antisense Oligonucleotides Delivered to the Mouse CNS Ameliorate Symptoms of Severe Spinal Muscular Atrophy. <i>Science Translational Medicine</i> , 2011, 3, 72ra18.	5.8	437
7	Targeting nuclear RNA for in vivo correction of myotonic dystrophy. <i>Nature</i> , 2012, 488, 111-115.	13.7	435
8	Repeated nebulisation of non-viral CFTR gene therapy in patients with cystic fibrosis: a randomised, double-blind, placebo-controlled, phase 2b trial. <i>Lancet Respiratory Medicine</i> , 2015, 3, 684-691.	5.2	344
9	Expression and characterization of the cystic fibrosis transmembrane conductance regulator. <i>Nature</i> , 1990, 347, 382-386.	13.7	337
10	Hypoxia-Inducible Factor-1 Mediates Activation of Cultured Vascular Endothelial Cells by Inducing Multiple Angiogenic Factors. <i>Circulation Research</i> , 2003, 93, 664-673.	2.0	292
11	CNS expression of glucocerebrosidase corrects $\alpha$ -synuclein pathology and memory in a mouse model of Gaucher-related synucleinopathy. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2011, 108, 12101-12106.	3.3	282
12	Acid $\alpha$ -glucosidase mutants linked to gaucher disease, parkinson disease, and lewy body dementia alter $\alpha$ -synuclein processing. <i>Annals of Neurology</i> , 2011, 69, 940-953.	2.8	276
13	CpG-free plasmids confer reduced inflammation and sustained pulmonary gene expression. <i>Nature Biotechnology</i> , 2008, 26, 549-551.	9.4	269
14	CNS-targeted gene therapy improves survival and motor function in a mouse model of spinal muscular atrophy. <i>Journal of Clinical Investigation</i> , 2010, 120, 1253-1264.	3.9	252
15	Improved Cationic Lipid Formulations for In Vivo Gene Therapy. <i>Annals of the New York Academy of Sciences</i> , 1995, 772, 126-139.	1.8	249
16	Basis of Pulmonary Toxicity Associated with Cationic Lipid-Mediated Gene Transfer to the Mammalian Lung. <i>Human Gene Therapy</i> , 1997, 8, 689-707.	1.4	239
17	Cystic fibrosis transmembrane conductance regulator: A chloride channel with novel regulation. <i>Neuron</i> , 1992, 8, 821-829.	3.8	226
18	Comprehensive Analysis of the Acute Toxicities Induced by Systemic Administration of Cationic Lipid:Plasmid DNA Complexes in Mice. <i>Human Gene Therapy</i> , 2000, 11, 2493-2513.	1.4	218

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19	Generation and characterization of a $\Delta$ F508 cystic fibrosis mouse model. <i>Nature Genetics</i> , 1995, 10, 445-452.	9.4	215
20	Reduced Inflammatory Response to Plasmid DNA Vectors by Elimination and Inhibition of Immunostimulatory CpG Motifs. <i>Molecular Therapy</i> , 2000, 1, 255-262.	3.7	211
21	CpG-Depleted Plasmid DNA Vectors with Enhanced Safety and Long-Term Gene Expression in Vivo. <i>Molecular Therapy</i> , 2002, 5, 731-738.	3.7	203
22	Augmenting CNS glucocerebrosidase activity as a therapeutic strategy for parkinsonism and other Gaucher-related synucleinopathies. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2013, 110, 3537-3542.	3.3	203
23	Contribution of Plasmid DNA to Inflammation in the Lung after Administration of Cationic Lipid:pDNA Complexes. <i>Human Gene Therapy</i> , 1999, 10, 223-234.	1.4	199
24	Dimorphic Effects of Transforming Growth Factor- $\beta$ Signaling During Aortic Aneurysm Progression in Mice Suggest a Combinatorial Therapy for Marfan Syndrome. <i>Arteriosclerosis, Thrombosis, and Vascular Biology</i> , 2015, 35, 911-917.	1.1	189
25	Glucosylceramide synthase inhibition alleviates aberrations in synucleinopathy models. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2017, 114, 2699-2704.	3.3	165
26	Silencing Mutant Huntingtin by Adeno-Associated Virus-Mediated RNA Interference Ameliorates Disease Manifestations in the YAC128 Mouse Model of Huntington's Disease. <i>Human Gene Therapy</i> , 2014, 25, 461-474.	1.4	135
27	High and Sustained Transgene Expression in Vivo from Plasmid Vectors Containing a Hybrid Ubiquitin Promoter. <i>Molecular Therapy</i> , 2001, 4, 75-82.	3.7	129
28	Hypoxia Up-regulates Expression of Peroxisome Proliferator-activated Receptor $\beta$ 3 Angiopoietin-related Gene (PGAR) in Cardiomyocytes: Role of Hypoxia Inducible Factor $1\alpha$ . <i>Journal of Molecular and Cellular Cardiology</i> , 2002, 34, 765-774.	0.9	128
29	Stabilization of Vascular Endothelial Growth Factor mRNA by Hypoxia-Inducible Factor 1. <i>Biochemical and Biophysical Research Communications</i> , 2002, 291, 908-914.	1.0	125
30	Fetal gene therapy for neurodegenerative disease of infants. <i>Nature Medicine</i> , 2018, 24, 1317-1323.	15.2	117
31	Optimization of Plasmid Vectors for High-Level Expression in Lung Epithelial Cells. <i>Human Gene Therapy</i> , 1997, 8, 575-584.	1.4	116
32	Intracellular protein trafficking defects in human disease. <i>Trends in Cell Biology</i> , 1992, 2, 145-149.	3.6	99
33	A Concentrated and Stable Aerosol Formulation of Cationic Lipid:DNA Complexes Giving High-Level Gene Expression in Mouse Lung. <i>Human Gene Therapy</i> , 1997, 8, 765-773.	1.4	93
34	Expression of constitutively stable hybrid hypoxia-inducible factor- $1\alpha$ protects cultured rat cardiomyocytes against simulated ischemia-reperfusion injury. <i>American Journal of Physiology - Cell Physiology</i> , 2005, 288, C314-C320.	2.1	89
35	Optimization of Formulations and Conditions for the Aerosol Delivery of Functional Cationic Lipid:DNA Complexes. <i>Human Gene Therapy</i> , 1997, 8, 313-322.	1.4	85
36	Partial restoration of cAMP-stimulated CFTR chloride channel activity in $\Delta$ F508 cells by deoxyspergualin. <i>American Journal of Physiology - Cell Physiology</i> , 1998, 275, C171-C178.	2.1	82

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37	Efficiency of Cationic Lipid-Mediated Transfection of Polarized and Differentiated Airway Epithelial Cells <i>In Vitro</i> and <i>In Vivo</i> . <i>Human Gene Therapy</i> , 1998, 9, 1531-1542.	1.4	81
38	Contribution of Toll-like Receptor 9 Signaling to the Acute Inflammatory Response to Nonviral Vectors. <i>Molecular Therapy</i> , 2004, 9, 241-248.	3.7	81
39	Translational Fidelity of Intrathecal Delivery of Self-Complementary AAV9 "Survival Motor Neuron 1 for Spinal Muscular Atrophy. <i>Human Gene Therapy</i> , 2014, 25, 619-630.	1.4	79
40	Hypoxia-inducible factor 1 mediates hypoxia-induced cardiomyocyte lipid accumulation by reducing the DNA binding activity of peroxisome proliferator-activated receptor $\beta$ /retinoid X receptor. <i>Biochemical and Biophysical Research Communications</i> , 2007, 364, 567-572.	1.0	67
41	A randomised, double-blind, placebo-controlled phase IIB clinical trial of repeated application of gene therapy in patients with cystic fibrosis: Table A1. <i>Thorax</i> , 2013, 68, 1075-1077.	2.7	66
42	Gaucher-related synucleinopathies: The examination of sporadic neurodegeneration from a rare (disease) angle. <i>Progress in Neurobiology</i> , 2015, 125, 47-62.	2.8	63
43	Inefficient cationic lipid-mediated siRNA and antisense oligonucleotide transfer to airway epithelial cells <i>in vivo</i> . <i>Respiratory Research</i> , 2006, 7, 26.	1.4	59
44	Antisense Oligonucleotide-Mediated Correction of Transcriptional Dysregulation is Correlated with Behavioral Benefits in the YAC128 Mouse Model of Huntington's Disease. <i>Journal of Huntington's Disease</i> , 2013, 2, 217-228.	0.9	58
45	Expression of angiopoietins in renal epithelial and clear cell carcinoma cells: regulation by hypoxia and participation in angiogenesis. <i>American Journal of Physiology - Renal Physiology</i> , 2004, 287, F649-F657.	1.3	55
46	Gene expression profiles in human cardiac cells subjected to hypoxia or expressing a hybrid form of HIF-1 $\beta$ . <i>Physiological Genomics</i> , 2002, 8, 23-32.	1.0	54
47	Systemic Delivery of a Peptide-Linked Morpholino Oligonucleotide Neutralizes Mutant RNA Toxicity in a Mouse Model of Myotonic Dystrophy. <i>Nucleic Acid Therapeutics</i> , 2013, 23, 109-117.	2.0	52
48	Glucocerebrosidase modulates cognitive and motor activities in murine models of Parkinson's disease. <i>Human Molecular Genetics</i> , 2016, 25, ddu124.	1.4	44
49	Correction of the cystic fibrosis defect by gene complementation in human intrahepatic biliary epithelial cell lines. <i>Gastroenterology</i> , 1995, 108, 584-592.	0.6	42
50	Electroporation enhances reporter gene expression following delivery of naked plasmid DNA to the lung. <i>Journal of Gene Medicine</i> , 2007, 9, 369-380.	1.4	42
51	IGF-1 delivery to CNS attenuates motor neuron cell death but does not improve motor function in type III SMA mice. <i>Neurobiology of Disease</i> , 2012, 45, 272-279.	2.1	41
52	Production of Cystic Fibrosis Transmembrane Conductance Regulator in the Milk of Transgenic Mice. <i>Nature Biotechnology</i> , 1992, 10, 74-77.	9.4	39
53	Adenovirus-Transduced Lung as a Portal for Delivering $\beta$ -Galactosidase A into Systemic Circulation for Fabry Disease. <i>Molecular Therapy</i> , 2002, 5, 745-754.	3.7	37
54	DNA Sequences in Cationic Lipid:pDNA-Mediated Systemic Toxicities. <i>Human Gene Therapy</i> , 2003, 14, 203-214.	1.4	36

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55	Cationic Lipid Structure and Formulation Considerations for Optimal Gene Transfection of the Lung. <i>Journal of Drug Targeting</i> , 2000, 7, 453-469.	2.1	35
56	Inhibition of osteoclastogenesis by prolyl hydroxylase inhibitor dimethyloxallyl glycine. <i>Journal of Bone and Mineral Metabolism</i> , 2010, 28, 510-519.	1.3	33
57	Purification and Characterization of Recombinant Cystic Fibrosis Transmembrane Conductance Regulator from Chinese Hamster Ovary and Insect Cells. <i>Journal of Biological Chemistry</i> , 1995, 270, 17033-17043.	1.6	32
58	Aerosolization of Cationic Lipid:pDNA Complexes—In Vitro Optimization of Nebulizer Parameters for Human Clinical Studies. <i>Human Gene Therapy</i> , 1998, 9, 43-52.	1.4	31
59	Characterization of the oligosaccharide structures associated with the cystic fibrosis transmembrane conductance regulator. <i>Glycobiology</i> , 2000, 10, 1225-1233.	1.3	31
60	The safety profile of a cationic lipid-mediated cystic fibrosis gene transfer agent following repeated monthly aerosol administration to sheep. <i>Biomaterials</i> , 2013, 34, 10267-10277.	5.7	30
61	Prospects for the gene therapy of spinal muscular atrophy. <i>Trends in Molecular Medicine</i> , 2011, 17, 259-265.	3.5	29
62	Secreted Gaussia luciferase as a sensitive reporter gene for in vivo and ex vivo studies of airway gene transfer. <i>Biomaterials</i> , 2011, 32, 2614-2624.	5.7	29
63	The use of carboxymethylcellulose gel to increase non-viral gene transfer in mouse airways. <i>Biomaterials</i> , 2010, 31, 2665-2672.	5.7	27
64	A hypoxic inducible factor-1 hybrid enhances collateral development and reduces vascular leakage in diabetic rats. <i>Journal of Gene Medicine</i> , 2009, 11, 390-400.	1.4	26
65	Mutant $\beta$ -GAL1; Expression and Synucleinopathy Risk: First Insights from Cellular and Mouse Models. <i>Neurodegenerative Diseases</i> , 2012, 10, 195-202.	0.8	26
66	Partial correction of the $\beta$ -galactosidase A deficiency and reduction of glycolipid storage in Fabry mice using synthetic vectors. <i>Journal of Gene Medicine</i> , 2004, 6, 85-92.	1.4	22
67	Gene Transfer to the CNS Is Efficacious in Immune-primed Mice Harboring Physiologically Relevant Titers of Anti-AAV Antibodies. <i>Molecular Therapy</i> , 2012, 20, 1713-1723.	3.7	22
68	A randomised, double-blind, placebo-controlled trial of repeated nebulisation of non-viral cystic fibrosis transmembrane conductance regulator (CFTR) gene therapy in patients with cystic fibrosis. <i>Efficacy and Mechanism Evaluation</i> , 2016, 3, 1-210.	0.9	22
69	Nitric oxide inhibits heterologous CFTR expression in polarized epithelial cells. <i>American Journal of Physiology - Lung Cellular and Molecular Physiology</i> , 1999, 277, L89-L96.	1.3	20
70	Increased Duration of Transgene Expression in the Lung with Plasmid DNA Vectors Harboring Adenovirus E4 Open Reading Frame 3. <i>Human Gene Therapy</i> , 1999, 10, 1833-1843.	1.4	20
71	Identification of transfected cell types following non-viral gene transfer to the murine lung. <i>Journal of Gene Medicine</i> , 2007, 9, 184-196.	1.4	18
72	Limitations of the Murine Nose in the Development of Nonviral Airway Gene Transfer. <i>American Journal of Respiratory Cell and Molecular Biology</i> , 2010, 43, 46-54.	1.4	18

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73	Cationic Lipid-Mediated Gene Delivery to the Airways. , 1999, , 39-68.		16
74	In vivo imaging of gene transfer to the respiratory tract. Biomaterials, 2008, 29, 1533-1540.	5.7	13
75	The role of doxorubicin in non-viral gene transfer in the lung. Biomaterials, 2009, 30, 1971-1977.	5.7	12
76	Low-frequency ultrasound increases non-viral gene transfer to the mouse lung. Acta Biochimica Et Biophysica Sinica, 2010, 42, 45-51.	0.9	11
77	Rapid identification of novel functional promoters for gene therapy. Journal of Molecular Medicine, 2012, 90, 1487-1496.	1.7	9
78	Restoration of Cyclic Adenosine Monophosphateâ€“Stimulated Chloride Channel Activity in Human Cystic Fibrosis Tracheobronchial Submucosal Gland Cells by Adenovirus-Mediated and Cationic Lipidâ€“Mediated Gene Transfer. American Journal of Respiratory Cell and Molecular Biology, 1999, 20, 1107-1115.	1.4	7
79	Assessment of the nuclear pore dilating agent transâ€“cyclohexaneâ€“1,2â€“diol in differentiated airway epithelium. Journal of Gene Medicine, 2012, 14, 491-500.	1.4	6
80	Formulation of Synthetic Vectors for Cystic Fibrosis Gene Therapy. , 2002, 70, 585-598.		0
81	Formulation of Synthetic Gene Delivery Vectors for Transduction of the Airway Epithelium. , 2004, 245, 95-114.		0