Brian Bigger

List of Publications by Year
in descending order

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89 papers	4,134 citations	126907 33 h-index	123424 61 g-index
93 all docs	93 docs citations	93 times ranked	4272 citing authors

#	Article	IF	Citations
1	The Bone Marrow Functionally Contributes to Liver Fibrosis. Gastroenterology, 2006, 130, 1807-1821.	1.3	467
2	Oncogenesis Following Delivery of a Nonprimate Lentiviral Gene Therapy Vector to Fetal and Neonatal Mice. Molecular Therapy, 2005, 12, 763-771.	8.2	224
3	Neuropathology in Mouse Models of Mucopolysaccharidosis Type I, IIIA and IIIB. PLoS ONE, 2012, 7, e35787.	2.5	148
4	Hepatic stem cells: from inside and outside the liver?. Cell Proliferation, 2004, 37, 1-21.	5.3	145
5	An araC-controlled Bacterialcre Expression System to Produce DNA Minicircle Vectors for Nuclear and Mitochondrial Gene Therapy. Journal of Biological Chemistry, 2001, 276, 23018-23027.	3.4	142
6	Permanent phenotypic correction of hemophilia B in immunocompetent mice by prenatal gene therapy. Blood, 2004, 104, 2714-2721.	1.4	132
7	Improved Metabolic Correction in Patients with Lysosomal Storage Disease Treated with Hematopoietic Stem Cell Transplant Compared with Enzyme Replacement Therapy. Journal of Pediatrics, 2009, 154, 609-611.	1.8	125
8	Genistein Improves Neuropathology and Corrects Behaviour in a Mouse Model of Neurodegenerative Metabolic Disease. PLoS ONE, 2010, 5, e14192.	2.5	121
9	Neuroinflammation, mitochondrial defects and neurodegeneration in mucopolysaccharidosis III type C mouse model. Brain, 2015, 138, 336-355.	7.6	113
10	Myeloid/Microglial Driven Autologous Hematopoietic Stem Cell Gene Therapy Corrects a Neuronopathic Lysosomal Disease. Molecular Therapy, 2013, 21, 1938-1949.	8.2	96
11	Busulfan Conditioning Enhances Engraftment of Hematopoietic Donor-derived Cells in the Brain Compared With Irradiation. Molecular Therapy, 2013, 21, 868-876.	8.2	95
12	Hematopoietic Stem Cell and Gene Therapy Corrects Primary Neuropathology and Behavior in Mucopolysaccharidosis IIIA Mice. Molecular Therapy, 2012, 20, 1610-1621.	8.2	94
13	Genistein reduces lysosomal storage in peripheral tissues of mucopolysaccharide IIIB mice. Molecular Genetics and Metabolism, 2009, 98, 235-242.	1.1	90
14	Xenogenic Transfer of Isolated Murine Mitochondria into Human <i>Ï‹/i>⁰ Cells Can Improve Respiratory Function. Rejuvenation Research, 2007, 10, 561-570.	1.8	89
15	Quantitative Charge-Tags for Sterol and Oxysterol Analysis. Clinical Chemistry, 2015, 61, 400-411.	3.2	89
16	A novel adeno-associated virus capsid with enhanced neurotropism corrects a lysosomal transmembrane enzyme deficiency. Brain, 2018, 141, 2014-2031.	7.6	80
17	Mucopolysaccharide diseases: A complex interplay between neuroinflammation, microglial activation and adaptive immunity. Journal of Inherited Metabolic Disease, 2014, 37, 1-12.	3.6	77
18	Anatomical changes and pathophysiology of the brain in mucopolysaccharidosis disorders. Molecular Genetics and Metabolism, 2018, 125, 322-331.	1.1	71

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19	Brainâ€targeted stem cell gene therapy corrects mucopolysaccharidosis type II via multiple mechanisms. EMBO Molecular Medicine, 2018, 10, .	6.9	66
20	Macrophage enzyme and reduced inflammation drive brain correction of mucopolysaccharidosis IIIB by stem cell gene therapy. Brain, 2018, 141, 99-116.	7.6	64
21	Mucopolysaccharidosis Type I, Unique Structure of Accumulated Heparan Sulfate and Increased N-Sulfotransferase Activity in Mice Lacking Î±-l-iduronidase. Journal of Biological Chemistry, 2011, 286, 37515-37524.	3.4	58
22	Parental social support, coping strategies, resilience factors, stress, anxiety and depression levels in parents of children with MPS III (Sanfilippo syndrome) or children with intellectual disabilities (ID). Journal of Inherited Metabolic Disease, 2013, 36, 281-291.	3.6	58
23	Analytical strategies for characterization of oxysterol lipidomes: Liver X receptor ligands in plasma. Free Radical Biology and Medicine, 2013, 59, 69-84.	2.9	56
24	Permanent partial phenotypic correction and tolerance in a mouse model of hemophilia B by stem cell gene delivery of human factor IX. Gene Therapy, 2006, 13, 117-126.	4.5	54
25	The role of innate immunity in mucopolysaccharide diseases. Journal of Neurochemistry, 2019, 148, 639-651.	3.9	53
26	Enzyme replacement therapy prior to haematopoietic stem cell transplantation in Mucopolysaccharidosis Type I: 10year combined experience of 2 centres. Molecular Genetics and Metabolism, 2016, 117, 373-377.	1.1	51
27	Biomarker responses correlate with antibody status in mucopolysaccharidosis type I patients on long-term enzyme replacement therapy. Molecular Genetics and Metabolism, 2015, 114, 129-137.	1.1	49
28	Hematopoietic stem cell transplantation improves the high incidence of neutralizing allo-antibodies observed in Hurler's syndrome after pharmacological enzyme replacement therapy. Haematologica, 2012, 97, 1320-1328.	3.5	48
29	Highly efficient EIAV-mediated in utero gene transfer and expression in the major muscle groups affected by Duchenne muscular dystrophy. Gene Therapy, 2004, 11, 1117-1125.	4.5	46
30	Heparin cofactor IIâ€thrombin complex and dermatan sulphate:chondroitin sulphate ratio are biomarkers of shortâ€and longâ€term treatment effects in mucopolysaccharide diseases. Journal of Inherited Metabolic Disease, 2011, 34, 499-508.	3.6	44
31	Plasma and urinary levels of dermatan sulfate and heparan sulfate derived disaccharides after longâ€term enzyme replacement therapy (ERT) in MPS I: correlation with the timing of ERT and with total urinary excretion of glycosaminoglycans. Journal of Inherited Metabolic Disease, 2013, 36, 247-255.	3.6	44
32	Sleep disordered breathing in mucopolysaccharidosis I: a multivariate analysis of patient, therapeutic and metabolic correlators modifying long term clinical outcome. Orphanet Journal of Rare Diseases, 2015, 10, 42.	2.7	43
33	Metabolism of Non-Enzymatically Derived Oxysterols: Clues from sterol metabolic disorders. Free Radical Biology and Medicine, 2019, 144, 124-133.	2.9	39
34	Pre-clinical Safety and Efficacy of Lentiviral Vector-Mediated ExÂVivo Stem Cell Gene Therapy for the Treatment of Mucopolysaccharidosis IIIA. Molecular Therapy - Methods and Clinical Development, 2019, 13, 399-413.	4.1	37
35	Development of a Self-assembling Nuclear Targeting Vector System Based on the Tetracycline Repressor Protein. Journal of Biological Chemistry, 2004, 279, 5555-5564.	3.4	35
36	Central and haematopoietic interleukin-1 both contribute to ischaemic brain injury in mice. DMM Disease Models and Mechanisms, 2013, 6, 1043-8.	2.4	35

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37	Heparan Sulfate Inhibits Hematopoietic Stem and Progenitor Cell Migration and Engraftment in Mucopolysaccharidosis I. Journal of Biological Chemistry, 2014, 289, 36194-36203.	3.4	34
38	Sterols and oxysterols in plasma from Smith-Lemli-Opitz syndrome patients. Journal of Steroid Biochemistry and Molecular Biology, 2017, 169, 77-87.	2.5	34
39	Evaluation of heparin cofactor IIâ€“thrombin complex as a biomarker on blood spots from mucopolysaccharidosis I, IIIA and IIIB mice. Molecular Genetics and Metabolism, 2010, 99, 269-274.	1.1	33
40	Assessment of Sleep in Children with Mucopolysaccharidosis Type III. PLoS ONE, 2014, 9, e84128.	2.5	33
41	Female Mucopolysaccharidosis IIIA Mice Exhibit Hyperactivity and a Reduced Sense of Danger in the Open Field Test. PLoS ONE, 2011, 6, e25717.	2.5	31
42	Haematopoietic stem cell gene therapy with <scp>IL</scp> â€¹Ra rescues cognitive loss in mucopolysaccharidosis <scp>IIIA</scp>. EMBO Molecular Medicine, 2020, 12, e11185.	6.9	31
43	The role of antibodies in enzyme treatments and therapeutic strategies. Best Practice and Research in Clinical Endocrinology and Metabolism, 2015, 29, 183-194.	4.7	30
44	Comparison of HIV- and EIAV-Based Vectors on Their Efficiency in Transducing Murine and Human Hematopoietic Repopulating Cells. Molecular Therapy, 2005, 12, 537-546.	8.2	28
45	Successful allogeneic bone marrow transplant for Niemannâ€“Pick disease type C2 is likely to be associated with a severe â€˜graft versus substrateâ€™ effect. Journal of Inherited Metabolic Disease, 2010, 33, 171-173.	3.6	28
46	Circadian rhythm and suprachiasmatic nucleus alterations in the mouse model of mucopolysaccharidosis IIIB. Behavioural Brain Research, 2010, 209, 212-220.	2.2	27
47	Recommendations on clinical trial design for treatment of Mucopolysaccharidosis Type III. Orphanet Journal of Rare Diseases, 2017, 12, 117.	2.7	27
48	Actinomycin D downregulates Sox2 and improves survival in preclinical models of recurrent glioblastoma. Neuro-Oncology, 2020, 22, 1289-1301.	1.2	27
49	Hyperactive behaviour in the mouse model of mucopolysaccharidosis IIIB in the open field and home cage environments. Genes, Brain and Behavior, 2011, 10, 673-682.	2.2	25
50	High dose genistein in Sanfilippo syndrome: A randomised controlled trial. Journal of Inherited Metabolic Disease, 2021, 44, 1248-1262.	3.6	24
51	The Fetal Mouse Is a Sensitive Genotoxicity Model That Exposes Lentiviral-associated Mutagenesis Resulting in Liver Oncogenesis. Molecular Therapy, 2013, 21, 324-337.	8.2	21
52	Identification of unusual oxysterols and bile acids with 7-oxo or 3Î²,5Î±,6Î²-trihydroxy functions in human plasma by charge-tagging mass spectrometry with multistage fragmentation. Journal of Lipid Research, 2018, 59, 1058-1070.	4.2	21
53	Introduction of Chloramphenicol Resistance into the Modified Mouse Mitochondrial Genome: Cloning of Unstable Sequences by Passage through Yeast. Analytical Biochemistry, 2000, 277, 236-242.	2.4	20
54	Actigraphic investigation of circadian rhythm functioning and activity levels in children with mucopolysaccharidosis type III (Sanfilippo syndrome). Journal of Neurodevelopmental Disorders, 2015, 7, 31.	3.1	20

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55	The impact of the immune system on the safety and efficiency of enzyme replacement therapy in lysosomal storage disorders. Journal of Inherited Metabolic Disease, 2016, 39, 499-512.	3.6	20
56	Targeting the MAPK7/MMP9 axis for metastasis in primary bone cancer. Oncogene, 2020, 39, 5553-5569.	5.9	20
57	Identification of age-dependent motor and neuropsychological behavioural abnormalities in a mouse model of Mucopolysaccharidosis Type II. PLoS ONE, 2017, 12, e0172435.	2.5	20
58	An investigation of the middle and late behavioural phenotypes of Mucopolysaccharidosis Type-III. Journal of Neurodevelopmental Disorders, 2014, 6, 46.	3.1	19
59	A nonmyeloablative chimeric mouse model accurately defines microglia and macrophage contribution in glioma. Neuropathology and Applied Neurobiology, 2019, 45, 119-140.	3.2	18
60	Enzyme replacement therapy and hematopoietic stem cell transplant: a new paradigm of treatment in Wolman disease. Orphanet Journal of Rare Diseases, 2021, 16, 235.	2.7	18
61	Modifying inter-cistronic sequence significantly enhances IRES dependent second gene expression in bicistronic vector: Construction of optimised cassette for gene therapy of familial hypercholesterolemia. Non-coding RNA Research, 2019, 4, 1-14.	4.6	16
62	High content screening of patient-derived cell lines highlights the potential of non-standard chemotherapeutic agents for the treatment of glioblastoma. PLoS ONE, 2018, 13, e0193694.	2.5	13
63	An Improved Adeno-Associated Virus Vector for Neurological Correction of the Mouse Model of Mucopolysaccharidosis IIIA. Human Gene Therapy, 2019, 30, 1052-1066.	2.7	13
64	Non-myeloablative busulfan chimeric mouse models are less pro-inflammatory than head-shielded irradiation for studying immune cell interactions in brain tumours. Journal of Neuroinflammation, 2019, 16, 25.	7.2	13
65	Innate Immunity in Mucopolysaccharide Diseases. International Journal of Molecular Sciences, 2022, 23, 1999.	4.1	13
66	Perspectives on Gene Therapy for Cystic Fibrosis Airway Disease. BioDrugs, 2001, 15, 615-634.	4.6	12
67	Trial and Error: How the Unclonable Human Mitochondrial Genome was Cloned in Yeast. Pharmaceutical Research, 2011, 28, 2863-2870.	3.5	12
68	Bile acid biosynthesis in Smith-Lemli-Opitz syndrome bypassing cholesterol: Potential importance of pathway intermediates. Journal of Steroid Biochemistry and Molecular Biology, 2021, 206, 105794.	2.5	12
69	RecET driven chromosomal gene targeting to generate a RecA deficient Escherichia coli strain for Cre mediated production of minicircle DNA. BMC Biotechnology, 2006, 6, 17.	3.3	11
70	Substrate accumulation and extracellular matrix remodelling promote persistent upper airway disease in mucopolysaccharidosis patients on enzyme replacement therapy. PLoS ONE, 2018, 13, e0203216.	2.5	11
71	Murine leukemia following irradiation conditioning for transplantation of lentivirally-modified hematopoietic stem cells. European Journal of Haematology, 2007, 78, 303-313.	2.2	10
72	In vivo Tâ€cell depletion using alemtuzumab in family and unrelated donor transplantation for pediatric nonâ€malignant disease achieves engraftment with low incidence of graft vs. host disease. Pediatric Transplantation, 2015, 19, 211-218.	1.0	10

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73	Delivering Hematopoietic Stem Cell Gene Therapy Treatments for Neurological Lysosomal Diseases. ACS Chemical Neuroscience, 2019, 10, 18-20.	3.5	10
74	Novel approaches and mechanisms in hematopoietic stem cell gene therapy. Discovery Medicine, 2014, 17, 207-15.	0.5	10
75	The Inflammation in the Cytopathology of Patients With Mucopolysaccharidoses- Immunomodulatory Drugs as an Approach to Therapy. Frontiers in Pharmacology, 2022, 13, .	3.5	10
76	Strategies for the Induction of Immune Tolerance to Enzyme Replacement Therapy in Mucopolysaccharidosis Type I. Molecular Therapy - Methods and Clinical Development, 2019, 13, 321-333.	4.1	9
77	Early defects in mucopolysaccharidosis type IIIC disrupt excitatory synaptic transmission. JCI Insight, 2021, 6, .	5.0	8
78	Obstructive Sleep Apnea in MPS. FIRE Forum for International Research in Education, 2015, 3, 232640981561639.	0.7	7
79	Post-transplant laronidase augmentation for children with Hurler syndrome: biochemical outcomes. Scientific Reports, 2019, 9, 14105.	3.3	7
80	Hampering brain tumor proliferation and migration using peptide nanofiber:siPLK1/MMP2 complexes. Nanomedicine, 2019, 14, 3127-3142.	3.3	7
81	Characterisation of the T cell and dendritic cell repertoire in a murine model of mucopolysaccharidosis I (MPS I). Journal of Inherited Metabolic Disease, 2013, 36, 257-262.	3.6	6
82	Signal One and Two Blockade Are Both Critical for Non-Myeloablative Murine HSCT across a Major Histocompatibility Complex Barrier. PLoS ONE, 2013, 8, e77632.	2.5	5
83	Current and Future Treatment of Mucopolysaccharidosis (MPS) Type II: Is Brain-Targeted Stem Cell Gene Therapy the Solution for This Devastating Disorder?. International Journal of Molecular Sciences, 2022, 23, 4854.	4.1	5
84	Tipping the scales in favour of mitochondrial gene therapy. Gene Therapy, 1999, 6, 1909-1910.	4.5	3
85	Minimized, CpG-Depleted, and Methylated DNA Vectors: Towards Perfection in Nonviral Gene Therapy. , 2006, , 43-54.		2
86	Cellular Therapy of Lysosomal Storage Disorders: Current Status and Future Prospects. Current Pediatric Reviews, 2009, 5, 147-159.	0.8	2
87	Immune activation or immunomodulation in the brains of MPS IIIB mice? Commentary on â€œinnate and adaptive immune activation in the brain of MPS IIIB mouse modelâ€. Journal of Neuroscience Research, 2010, 88, 233-233.	2.9	2
88	Ex-vivo autologous stem cell gene therapy clinical trial for mucopolysaccharidosis type IIIA: Update on phase I/II clinical trial. Molecular Genetics and Metabolism, 2021, 132, S56-S57.	1.1	2
89	Is it congenital or acquired von Willebrands disease?. Haemophilia, 2015, 21, e113-5.	2.1	1