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	Innate Immunity in Mucopolysaccharide Diseases. International Journal of Molecular Sciences, 2022, 23, 1999.	4.1	13
2	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
3	The Inflammation in the Cytopathology of Patients With Mucopolysaccharidoses- Immunomodulatory Drugs as an Approach to Therapy. Frontiers in Pharmacology, 2022, 13, .	3.5	10
4	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
5	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
6	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
7	Early defects in mucopolysaccharidosis type IIIC disrupt excitatory synaptic transmission. JCI Insight, 2021, 6, .	5.0	8
8	High dose genistein in Sanfilippo syndrome: A randomised controlled trial. Journal of Inherited Metabolic Disease, 2021, 44, 1248-1262.	3.6	24
9	Targeting the MAPK7/MMP9 axis for metastasis in primary bone cancer. Oncogene, 2020, 39, 5553-5569.	5.9	20
10	Actinomycin D downregulates Sox2 and improves survival in preclinical models of recurrent glioblastoma. Neuro-Oncology, 2020, 22, 1289-1301.	1.2	27
11	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
12	The role of innate immunity in mucopolysaccharide diseases. Journal of Neurochemistry, 2019, 148, 639-651.	3.9	53
13	Delivering Hematopoietic Stem Cell Gene Therapy Treatments for Neurological Lysosomal Diseases. ACS Chemical Neuroscience, 2019, 10, 18-20.	3.5	10
14	Post-transplant laronidase augmentation for children with Hurler syndrome: biochemical outcomes. Scientific Reports, 2019, 9, 14105.	3.3	7
15	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
16	Metabolism of Non-Enzymatically Derived Oxysterols: Clues from sterol metabolic disorders. Free Radical Biology and Medicine, 2019, 144, 124-133.	2.9	39
17	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
18	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

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19	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
20	Hampering brain tumor proliferation and migration using peptide nanofiber:siPLK1/MMP2 complexes. Nanomedicine, 2019, 14, 3127-3142.	3.3	7
21	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
22	A nonmyeloablative chimeric mouse model accurately defines microglia and macrophage contribution in glioma. Neuropathology and Applied Neurobiology, 2019, 45, 119-140.	3.2	18
23	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
24	Macrophage enzyme and reduced inflammation drive brain correction of mucopolysaccharidosis IIIB by stem cell gene therapy. Brain, 2018, 141, 99-116.	7.6	64
25	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
26	A novel adeno-associated virus capsid with enhanced neurotropism corrects a lysosomal transmembrane enzyme deficiency. Brain, 2018, 141, 2014-2031.	7.6	80
27	Anatomical changes and pathophysiology of the brain in mucopolysaccharidosis disorders. Molecular Genetics and Metabolism, 2018, 125, 322-331.	1.1	71
28	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
29	Brainâ€targeted stem cell gene therapy corrects mucopolysaccharidosis type II via multiple mechanisms. EMBO Molecular Medicine, 2018, 10, .	6.9	66
30	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
31	Recommendations on clinical trial design for treatment of Mucopolysaccharidosis Type III. Orphanet Journal of Rare Diseases, 2017, 12, 117.	2.7	27
32	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
33	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
34	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
35	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
36	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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37	Obstructive Sleep Apnea in MPS. FIRE Forum for International Research in Education, 2015, 3, 232640981561639.	0.7	7
38	Neuroinflammation, mitochondrial defects and neurodegeneration in mucopolysaccharidosis III type C mouse model. Brain, 2015, 138, 336-355.	7.6	113
39	Quantitative Charge-Tags for Sterol and Oxysterol Analysis. Clinical Chemistry, 2015, 61, 400-411.	3.2	89
40	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
41	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
42	Is it congenital or acquired von Willebrands disease?. Haemophilia, 2015, 21, e113-5.	2.1	1
43	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
44	Assessment of Sleep in Children with Mucopolysaccharidosis Type III. PLoS ONE, 2014, 9, e84128.	2.5	33
45	An investigation of the middle and late behavioural phenotypes of Mucopolysaccharidosis Type-III. Journal of Neurodevelopmental Disorders, 2014, 6, 46.	3.1	19
46	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
47	Mucopolysaccharide diseases: A complex interplay between neuroinflammation, microglial activation and adaptive immunity. Journal of Inherited Metabolic Disease, 2014, 37, 1-12.	3.6	77
48	Novel approaches and mechanisms in hematopoietic stem cell gene therapy. Discovery Medicine, 2014, 17, 207-15.	0.5	10
49	Myeloid/Microglial Driven Autologous Hematopoietic Stem Cell Gene Therapy Corrects a Neuronopathic Lysosomal Disease. Molecular Therapy, 2013, 21, 1938-1949.	8.2	96
50	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
51	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
52	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
53	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
54	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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55	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
56	Busulfan Conditioning Enhances Engraftment of Hematopoietic Donor-derived Cells in the Brain Compared With Irradiation. Molecular Therapy, 2013, 21, 868-876.	8.2	95
57	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
58	Hematopoietic Stem Cell and Gene Therapy Corrects Primary Neuropathology and Behavior in Mucopolysaccharidosis IIIA Mice. Molecular Therapy, 2012, 20, 1610-1621.	8.2	94
59	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
60	Neuropathology in Mouse Models of Mucopolysaccharidosis Type I, IIIA and IIIB. PLoS ONE, 2012, 7, e35787.	2.5	148
61	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
62	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
63	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
64	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
65	Trial and Error: How the Unclonable Human Mitochondrial Genome was Cloned in Yeast. Pharmaceutical Research, 2011, 28, 2863-2870.	3.5	12
66	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
67	Genistein Improves Neuropathology and Corrects Behaviour in a Mouse Model of Neurodegenerative Metabolic Disease. PLoS ONE, 2010, 5, e14192.	2.5	121
68	Circadian rhythm and suprachiasmatic nucleus alterations in the mouse model of mucopolysaccharidosis IIIB. Behavioural Brain Research, 2010, 209, 212-220.	2.2	27
69	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
70	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
71	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
72	Genistein reduces lysosomal storage in peripheral tissues of mucopolysaccharide IIIB mice. Molecular Genetics and Metabolism, 2009, 98, 235-242.	1.1	90

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73	Cellular Therapy of Lysosomal Storage Disorders: Current Status and Future Prospects. Current Pediatric Reviews, 2009, 5, 147-159.	0.8	2
74	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
75	Murine leukemia following irradiation conditioning for transplantation of lentivirally-modified hematopoietic stem cells. European Journal of Haematology, 2007, 78, 303-313.	2.2	10
76	The Bone Marrow Functionally Contributes to Liver Fibrosis. Gastroenterology, 2006, 130, 1807-1821.	1.3	467
77	Minimized, CpG-Depleted, and Methylated DNA Vectors: Towards Perfection in Nonviral Gene Therapy. , 2006, , 43-54.		2
78	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
79	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
80	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
81	Oncogenesis Following Delivery of a Nonprimate Lentiviral Gene Therapy Vector to Fetal and Neonatal Mice. Molecular Therapy, 2005, 12, 763-771.	8.2	224
82	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
83	Hepatic stem cells: from inside and outside the liver?. Cell Proliferation, 2004, 37, 1-21.	5.3	145
84	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
85	Permanent phenotypic correction of hemophilia B in immunocompetent mice by prenatal gene therapy. Blood, 2004, 104, 2714-2721.	1.4	132
86	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
87	Perspectives on Gene Therapy for Cystic Fibrosis Airway Disease. BioDrugs, 2001, 15, 615-634.	4.6	12
88	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
89	Tipping the scales in favour of mitochondrial gene therapy. Gene Therapy, 1999, 6, 1909-1910.	4.5	3