Kim Hemsley

List of Publications by Citations

71 papers	1,473 citations	23 h-index	35 g-index
73 ext. papers	1,630 ext. citations	4.2 avg, IF	4.39 L-index

#	Paper	IF	Citations
71	Functional correction of CNS lesions in an MPS-IIIA mouse model by intracerebral AAV-mediated delivery of sulfamidase and SUMF1 genes. *Human Molecular Genetics*, **2007**, 16, 2693-702	5.6	90
70	Endo-lysosomal and autophagic dysfunction: a driving factor in Alzheimer's disease?. *Journal of Neurochemistry*, **2017**, 140, 703-717	6	79
69	Characterization of a C57BL/6 congenic mouse strain of mucopolysaccharidosis type IIIA. *Brain Research*, **2006**, 1104, 1-17	3.7	79
68	Injection of recombinant human sulfamidase into the CSF via the cerebellomedullary cistern in MPS IIIA mice. *Molecular Genetics and Metabolism*, **2007**, 90, 313-28	3.7	75
67	Development of motor deficits in a murine model of mucopolysaccharidosis type IIIA (MPS-IIIA). *Behavioural Brain Research*, **2005**, 158, 191-9	3.4	64
66	Effect of high dose, repeated intra-cerebrospinal fluid injection of sulphamidase on neuropathology in mucopolysaccharidosis type IIIA mice. *Genes, Brain and Behavior*, **2008**, 7, 740-53	3.6	61
65	Examination of intravenous and intra-CSF protein delivery for treatment of neurological disease. *European Journal of Neuroscience*, **2009**, 29, 1197-214	3.5	58
64	Open field locomotor activity and anxiety-related behaviors in mucopolysaccharidosis type IIIA mice. *Behavioural Brain Research*, **2008**, 191, 130-6	3.4	58
63	Intracerebral injection of sulfamidase delays neuropathology in murine MPS-IIIA. *Molecular Genetics and Metabolism*, **2004**, 82, 273-85	3.7	57
62	Effect of cisternal sulfamidase delivery in MPS IIIA Huntaway dogs--a proof of principle study. *Molecular Genetics and Metabolism*, **2009**, 98, 383-92	3.7	50
61	Enzyme replacement reduces neuropathology in MPS IIIA dogs. *Neurobiology of Disease*, **2011**, 43, 422-34	4.5	43
60	In vitro characterization of genetically modified embryonic stem cells as a therapy for murine mucopolysaccharidosis type IIIA. *Molecular Genetics and Metabolism*, **2004**, 81, 86-95	3.7	40
59	Reduction in open field activity in the absence of memory deficits in the App knock-in mouse model of Alzheimer's disease. *Behavioural Brain Research*, **2018**, 336, 177-181	3.4	35
58	An animal model of extrapyramidal side effects induced by antipsychotic drugs: relationship with D2 dopamine receptor occupancy. *Progress in Neuro-Psychopharmacology and Biological Psychiatry*, **2001**, 25, 573-90	5.5	35
57	Impact of high-dose, chemically modified sulfamidase on pathology in a murine model of MPS IIIA. *Experimental Neurology*, **2011**, 230, 123-30	5.7	34
56	A Preclinical Study Evaluating AAVrh10-Based Gene Therapy for Sanfilippo Syndrome. *Human Gene Therapy*, **2016**, 27, 363-75	4.8	28
55	Allogeneic stem cell transplantation does not improve neurological deficits in mucopolysaccharidosis type IIIA mice. *Experimental Neurology*, **2010**, 225, 445-54	5.7	28

54	Validation of a heparan sulfate-derived disaccharide as a marker of accumulation in murine mucopolysaccharidosis type IIIA. *Molecular Genetics and Metabolism*, **2006**, 87, 107-12	3.7	27
53	Delivery of therapeutic protein for prevention of neurodegenerative changes: comparison of different CSF-delivery methods. *Experimental Neurology*, **2015**, 263, 79-90	5.7	24
52	Changes in muscle tone are regulated by D1 and D2 dopamine receptors in the ventral striatum and D1 receptors in the substantia nigra. *Neuropsychopharmacology*, **2001**, 25, 514-26	8.7	24
51	Axonal dystrophy in the brain of mice with Sanfilippo syndrome. *Experimental Neurology*, **2017**, 295, 243-255	5.7	23
50	Treatment of canine fucosidosis by intracisternal enzyme infusion. *Experimental Neurology*, **2011**, 230, 218-26	5.7	23
49	SGSH gene transfer in mucopolysaccharidosis type IIIA mice using canine adenovirus vectors. *Molecular Genetics and Metabolism*, **2010**, 100, 168-75	3.7	23
48	Lessons learnt from animal models: pathophysiology of neuropathic lysosomal storage disorders. *Journal of Inherited Metabolic Disease*, **2010**, 33, 363-71	5.4	23
47	Determination of the role of injection site on the efficacy of intra-CSF enzyme replacement therapy in MPS IIIA mice. *Molecular Genetics and Metabolism*, **2015**, 115, 33-40	3.7	20
46	Helper-dependent canine adenovirus vector-mediated transgene expression in a neurodegenerative lysosomal storage disorder. *Gene*, **2012**, 491, 53-7	3.8	20
45	Intracisternal enzyme replacement therapy in lysosomal storage diseases: routes of absorption into brain. *Neuropathology and Applied Neurobiology*, **2011**, 37, 414-22	5.2	19
44	Neonatal Bone Marrow Transplantation in MPS IIIA Mice. *JIMD Reports*, **2013**, 8, 121-32	1.9	18
43	Evaluation of enzyme dose and dose-frequency in ameliorating substrate accumulation in MPS IIIA Huntaway dog brain. *Journal of Inherited Metabolic Disease*, **2015**, 38, 341-50	5.4	17
42	Delivery of recombinant proteins via the cerebrospinal fluid as a therapy option for neurodegenerative lysosomal storage diseases. *International Journal of Clinical Pharmacology and Therapeutics*, **2009**, 47 Suppl 1, S118-23	2	17
41	The effects of an irreversible dopamine receptor antagonist, N-ethoxycarbonyl-2-ethoxy-1,2-dihydroquinoline (EEDQ), on the regulation of muscle tone in the rat: the role of the substantia nigra. *Neuroscience Letters*, **1998**, 251, 77-80	3.3	16
40	Survival and engraftment of mouse embryonic stem cell-derived implants in the guinea pig brain. *Neuroscience Research*, **2005**, 53, 161-8	2.9	16
39	Raclopride and chlorpromazine, but not clozapine, increase muscle rigidity in the rat: relationship with D2 dopamine receptor occupancy. *Neuropsychopharmacology*, **1999**, 21, 101-9	8.7	15
38	Exocytosis is impaired in mucopolysaccharidosis IIIA mouse chromaffin cells. *Neuroscience*, **2012**, 227, 110-8	3.9	14
37	Emerging therapies for neurodegenerative lysosomal storage disorders - from concept to reality. *Journal of Inherited Metabolic Disease*, **2011**, 34, 1003-12	5.4	13

36	Low-dose, continuous enzyme replacement therapy ameliorates brain pathology in the neurodegenerative lysosomal disorder mucopolysaccharidosis type IIIA. *Journal of Neurochemistry*, **2016**, 137, 409-22	6	13
35	Glycosphingolipid analysis in a naturally occurring ovine model of acute neuronopathic Gaucher disease. *Neurobiology of Disease*, **2016**, 91, 143-54	7.5	12
34	Disease stage determines the efficacy of treatment of a paediatric neurodegenerative disease. *European Journal of Neuroscience*, **2014**, 39, 2139-50	3.5	12
33	Slow, continuous enzyme replacement via spinal CSF in dogs with the paediatric-onset neurodegenerative disease, MPS IIIA. *Journal of Inherited Metabolic Disease*, **2017**, 40, 443-453	5.4	11
32	Neuronal-specific impairment of heparan sulfate degradation in Drosophila reveals pathogenic mechanisms for Mucopolysaccharidosis type IIIA. *Experimental Neurology*, **2018**, 303, 38-47	5.7	11
31	Low-dose, continual enzyme delivery ameliorates some aspects of established brain disease in a mouse model of a childhood-onset neurodegenerative disorder. *Experimental Neurology*, **2016**, 278, 11-21	5.7	11
30	Intracisternal enzyme replacement therapy in lysosomal storage diseases: dispersal pathways, regional enzyme concentrations and the effect of posttreatment posture. *Neuropathology and Applied Neurobiology*, **2013**, 39, 681-92	5.2	11
29	AAVrh10 Vector Corrects Disease Pathology in MPS IIIA Mice and Achieves Widespread Distribution of SGSH in Large Animal Brains. *Molecular Therapy - Methods and Clinical Development*, **2020**, 17, 174-187	6.4	10
28	Embryonic stem cell-derived glial precursors as a vehicle for sulfamidase production in the MPS-IIIA mouse brain. *Cell Transplantation*, **2010**, 19, 985-98	4	9
27	A novel conditional Sgsh knockout mouse model recapitulates phenotypic and neuropathic deficits of Sanfilippo syndrome. *Journal of Inherited Metabolic Disease*, **2017**, 40, 715-724	5.4	8
26	Development of cerebellar pathology in the canine model of mucopolysaccharidosis type IIIA (MPS IIIA). *Molecular Genetics and Metabolism*, **2014**, 113, 283-93	3.7	8
25	Behavioural characterisation of the alpha-mannosidosis guinea pig. *Behavioural Brain Research*, **2008**, 186, 176-84	3.4	8
24	MPS-IIIA mice acquire autistic behaviours with age. *Journal of Inherited Metabolic Disease*, **2018**, 41, 669-677	5.4	7
23	Adeno-associated viral gene therapy for mucopolysaccharidoses exhibiting neurodegeneration. *Journal of Molecular Medicine*, **2017**, 95, 1043-1052	5.5	7
22	Primary culture of neural cells isolated from the cerebellum of newborn and adult mucopolysaccharidosis type IIIA mice. *Cellular and Molecular Neurobiology*, **2008**, 28, 949-59	4.6	7
21	Atropine acts in the ventral striatum to reduce raclopride-induced catalepsy. *European Journal of Pharmacology*, **2001**, 424, 179-87	5.3	7
20	Lysosomal Dysregulation in the Murine App Model of Alzheimer's Disease. *Neuroscience*, **2020**, 429, 143-155	3.9	7
19	Evaluation of Disease Lesions in the Developing Canine MPS IIIA Brain. *JIMD Reports*, **2019**, 43, 91-101	1.9	6

18	Dopamine receptors in the subthalamic nucleus are involved in the regulation of muscle tone in the rat. *Neuroscience Letters*, **2002**, 317, 123-6	3.3	6
17	Synthetic Disaccharide Standards Enable Quantitative Analysis of Stored Heparan Sulfate in MPS IIIA Murine Brain Regions. *ACS Chemical Neuroscience*, **2019**, 10, 3847-3858	5.7	5
16	Continual Low-Dose Infusion of Sulfamidase Is Superior to Intermittent High-Dose Delivery in Ameliorating Neuropathology in the MPS IIIA Mouse Brain. *JIMD Reports*, **2016**, 29, 59-68	1.9	5
15	Early disease course is unaltered in mucopolysaccharidosis type IIIA (MPS IIIA) mice lacking -synuclein. *Neuropathology and Applied Neurobiology*, **2019**, 45, 715-731	5.2	4
14	Variables influencing fluorimetric -sulfoglucosamine sulfohydrolase (SGSH) activity measurement in brain homogenates. *Molecular Genetics and Metabolism Reports*, **2015**, 5, 60-62	1.8	4
13	Atropine reduces raclopride-induced muscle rigidity by acting in the ventral region of the striatum. *European Journal of Pharmacology*, **2002**, 434, 117-23	5.3	4
12	-Mannosidosis in German Shepherd Dogs. *Veterinary Pathology*, **2019**, 56, 743-748	2.8	3
11	Neurological Examination of Sheep (Ovis aries) with Unilateral and Bilateral Quinolinic Acid Lesions of the Striatum Assessed by Magnetic Resonance Imaging **2019**, 05,		2
10	Canine adenoviral vector-mediated gene transfer to the guinea pig brain. *Gene Reports*, **2019**, 16, 100432	1.4	1
9	Genetic manipulation of murine embryonic stem cells with enhanced green fluorescence protein and sulfatase-modifying factor I genes. *Cytotherapy*, **2010**, 12, 400-7	4.8	1
8	Directed differentiation and characterization of genetically modified embryonic stem cells for therapy. *Methods in Molecular Biology*, **2006**, 329, 471-84	1.4	1
7	MUCOPOLYSACCHARIDOSIS II (MPS II) IN A FREE-LIVING KAKA (NESTOR MERIDIONALIS) IN NEW ZEALAND. *Journal of Wildlife Diseases*, **2021**, 57, 884-890	1.3	1
6	Is the eye a window to the brain in Sanfilippo syndrome?. *Acta Neuropathologica Communications*, **2020**, 8, 194	7.3	1
5	Longitudinal Magnetic Resonance Spectroscopy and Diffusion Tensor Imaging in Sheep (Ovis aries) With Quinolinic Acid Lesions of the Striatum: Time-Dependent Recovery of N-Acetylaspartate and Fractional Anisotropy. *Journal of Neuropathology and Experimental Neurology*, **2020**, 79, 1084-1092	3.1	1
4	Is SGSH heterozygosity a risk factor for early-onset neurodegenerative disease?. *Journal of Inherited Metabolic Disease*, **2021**, 44, 763-776	5.4	1
3	Lysosomal gene displays haploinsufficiency in a knock-in mouse model of Alzheimer's disease.. *IBRO Neuroscience Reports*, **2022**, 12, 131-141		0
2	Intracerebral gene therapy for mucopolysaccharidosis type IIIB syndrome. *Lancet Neurology, The*, **2017**, 16, 681-682	24.1	
1	Animal medical genetics: a historical perspective on more than 50 years of research into genetic disorders of animals at Massey University. *New Zealand Veterinary Journal*, **2021**, 69, 255-266	1.7	